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August 29 – September 2, 2010
New York Hilton • New York, NY

# CONGRESS ABSTRACTS

*Sponsored by Albert Einstein College of Medicine and the Children's Hospital at Montefiore in joint sponsorship with the International Pediatric Nephrology Association.*

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# The 15th Congress of the International Pediatric Nephrology Association

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# Schedule of Activities
# August 29 – September 2, 2010
# Hilton New York
# New York, NY, USA

## On-Site Registration Hours

| | |
|---|---|
| Friday, August 27 | 4:00 PM – 6:00 PM |
| Saturday, August 28 | 6:30 AM – 6:00 PM |
| Sunday, August 29 | 6:30 AM – 7:00 PM |
| Monday, August 30 | 6:30 AM – 7:00 PM |
| Tuesday, August 31 | 6:30 AM – 7:00 PM |
| Wednesday, September 1 | 6:30 AM – 7:00 PM |
| Thursday, September 2 | 6:30 AM – 12:00 PM |

## Speaker Ready Room Hours

| | |
|---|---|
| Friday, August 27 | 4:00 PM – 6:00 PM |
| Saturday, August 28 | 6:30 AM – 6:00 PM |
| Sunday, August 29 | 6:30 AM – 7:00 PM |
| Monday, August 30 | 6:30 AM – 7:00 PM |
| Tuesday, August 31 | 6:30 AM – 7:00 PM |
| Wednesday, September 1 | 6:30 AM – 7:00 PM |
| Thursday, September 2 | 6:30 AM – 12:00 PM |

## Exhibit Hours

| | |
|---|---|
| Sunday, August 29 | 6:30 PM – 7:30 PM |
| *Opening of Exhibits, Opening Reception* | |
| Monday, August 30 | 9:00 AM – 3:45 PM |
| Tuesday, August 31 | 9:00 AM – 3:45 PM |
| Wednesday, September 1 | 9:00 AM – 3:45 PM |

## Poster Viewing Hours

| | |
|---|---|
| Sunday, August 29 | 6:30 PM – 7:30 PM |
| Monday, August 30 | 9:00 AM – 5:30 PM |
| Tuesday, August 31 | 9:00 AM – 5:30 PM |
| Wednesday, September 1 | 9:00 AM – 5:30 PM |

## Poster Session (Presenters in Attendance)

| | |
|---|---|
| *Poster Session I* | |
| Sunday, August 29 | 6:30 PM – 7:30 PM |
| Monday, August 30 | 12:15 PM – 1:00 PM |
| *Poster Session II* | |
| Tuesday, August 31 | 12:15 PM – 1:00 PM |
| *Poster Session III* | |
| Wednesday, September 1 | 12:15 PM – 1:00 PM |

## Congress Opening Ceremony

| | |
|---|---|
| Sunday, August 29 | 5:00 PM – 6:30 PM |

## Congress Reception

| | |
|---|---|
| Wednesday, September 1 | 7:00 PM – 10:00 PM |

# Awards

## Trainee Travel Scholarship Awards

Yonca Acikgoz
*Abstract #321*

Banu Aykanat
*Abstract #589*

Nora Banki
*Abstract #193*

Surabhi Choudhary
*Abstract #563*

Elif Comak
*Abstract #105*

Orsolya Cseprekal
*Abstract #798*

Melina Chã d´Oliveira
*Abstract #29*

Martin Cuk
*Abstract #353*

Cagla Dogan
*Abstract #354*

Mohammad Hossein Fallahzadeh
*Abstract #360*

Andrea Fekete
*Abstract #111*

Quancheng Feng
*Abstract #648*

Yolanda Fuentes
*Abstract #544*

Paul Joseph Galutira
*Abstract #401*

Na Guan
*Abstract #366*

Ashima Gulati
*Abstract #396*

Kaan Gulleroglu
*Abstract #121*

Anna Kaczmarek
*Abstract #373*

Ina Kazyra
*Abstract #61*

Agata Korzeniecka-Kozerska
*Abstract #381*

Dusan Kostic
*Abstract #734*

Flavia Leao
*Abstract #407*

Aleksandra Mazo
*Abstract #389*

Md. Ariful Haque Mollik
*Abstract #298*

A Ohri
*Abstract #746*

Augustina Okpere
*Abstract #643*

Iva Palčić
*Abstract #617*

Cintia Rogow
*Abstract #751*

Krisztina Rusai
*Abstract #181*

Joana Santos
*Abstract #439*

Qian Shen
*Abstract #880*

Diamant Shtiza
*Abstract #156*

Magdalena Silska
*Abstract #443*

Aditi Sinha
*Abstract #678*

Vesna Stojanovic
*Abstract #903*

Qiang Sun
*Abstract #327*

Katarzyna Taranta-Janusz
*Abstract #683*

Prayong Vachvanichsanong
*Abstract #457*

Lucy Yaguo Ide
*Abstract #255*

Onder Yavascan
*Abstract #769*

Ilona Zagozdzon
*Abstract #771*

Mona Zahrane
*Abstract #257*

Yihui Zhai
*Abstract #717*

Xin Zhang
*Abstract #265*

# Non-Trainee Travel Scholarship Awards

Asiri Abeyagunawardena
*Abstract #640*

Adebowale Ademola
*Abstract #330*

Indira Agarwal
*Abstract #188*

Aamir Al Mosawi
*Abstract #335*

Laura Alconcher
*Abstract #843*

Sampson Antwi
*Abstract #849*

Csaba Bereczki
*Abstract #794*

Wattana Chartapisak
*Abstract #24*

A Delucchi
*Abstract #106*

Osman Donmez
*Abstract #206*

Ana Rose Dy
*Abstract #357*

Ayah Elmaghrabi
*Abstract #805*

Ahmed El-Refaey
*Abstract #730*

Fatemeh Ghane Sharbaf
*Abstract #117*

Wei Guo
*Abstract #934*

Wen-Yan Huang
*Abstract #935*

Augustina Jankauskiene
*Abstract #813*

Dafina Kuzmanovska
*Abstract #405*

Maria del Carmen Laso
*Abstract #641*

Ming-Lee Lee
*Abstract #737*

Ali Reza Merrikhi
*Abstract #297*

Israel Odetunde
*Abstract #869*

Saroj Patnaik
*Abstract #234*

Maria-Goretti Penido
*Abstract #570*

Zvonimir Puretic
*Abstract #507*

Jose Reyes
*Abstract #899*

Emilija Sahpazova
*Abstract #752*

Paulina Salas
*Abstract #515*

Ashot Sarkissian
*Abstract #441*

Ana Cristina Simoes eSilva
*Abstract #875*

Ertug Toroslu
*Abstract #760*

Vu Tru
*Abstract #687*

Alexey Tsygin
*Abstract #453*

Sandor Turi
*Abstract #250*

Anna Wasilewska
*Abstract #691*

Alev Yilmaz
*Abstract #262*

Igor Zorin
*Abstract #391*

# Trainee Research Awards

Best Clinical
Thurid Ahlenstiel
*Abstract #555*

Best Translational
J Jackson
*Abstract #467*

Best Basic Science
Eriko Tanaka
*Abstract #9*

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*All names listed are as of the date of printing.

# PRE CONGRESS PROGRAMMING

| Time | Tuesday, August 24 - Friday, August 27, 2010 |
|---|---|
| All Day Session | 11th INTERNATIONAL DEVELOPMENTAL NEPHROLOGY WORKSHOP<br>Mohonk Mountain House<br>New Palz, New York, USA |
| **Time** | **Saturday, August 28, 2010** |
| 6:30 AM - 6:00 PM | *Registration Open* |
| 8:00 AM - 5:00 PM | ALPORT SYNDROME FOUNDATION SYMPOSIUM<br>For Physicians, Researchers and Families |
| 8:00 AM - 5:00 PM | PRIMARY HYPEROXALURIA WORKSHOP |
| 8:00 AM - 6:30 PM | RENAL PATHOLOGY COURSE FOR PEDIATRIC NEPHROLOGISTS: FROM BASIC CONCEPTS TO RECENT ADVANCES<br>New York University Langone Medical Center<br>560 First Avenue, New York, USA<br>Smilow Seminar Room, First Floor |
| **Time** | **Sunday, August 29, 2010** |
| 6:30 AM - 6:00 PM | *Registration Open* |
| 8:00 AM - 5:00 PM | PRIMARY HYPEROXALURIA WORKSHOP |
| 8:00 AM - 4:30 PM | RENAL PATHOLOGY COURSE FOR PEDIATRIC NEPHROLOGISTS: FROM BASIC CONCEPTS TO RECENT ADVANCES<br>New York University Langone Medical Center<br>560 First Avenue, New York, USA<br>Smilow Seminar Room, First Floor |
| 11:00 AM - 3:00 PM | PEDIATRIC DIALYSIS: BASIC CONCEPTS FOR OPTIMAL CARE |
| 1:00 - 4:45 PM | INTERNATIONAL WORKSHOP ON HYPERTENSION IN CHILDREN AND ADOLESCENTS |

All sessions will take place at the Hilton New York Hotel unless otherwise stated.

# CONGRESS PROGRAMMING

| Time | Sunday, August 29, 2010 |
|---|---|
| 6:30 AM - 6:00 PM | *Registration Open* |
| 5:00 - 6:30 PM | OPENING CEREMONIES<br>Welcome<br>John E. Lewy Lecture: Caring for Children With Kidney Disease: Perspective of International Pediatrics |
| 6:30 - 7:30 PM | OPENING RECEPTION, OPENING OF EXHIBITS |

| Time | Monday, August 30, 2010 | | | | |
|---|---|---|---|---|---|
| 6:30 AM - 7:00 PM | *Registration Open* | | | | |
| 7:30 - 8:20 AM | **EARLY CLINICAL SESSION**<br>Clinical Approach to Kidney Stones | **EARLY CLINICAL SESSION**<br>Critical Care Nephrology | **EARLY CLINICAL SESSION**<br>Newer Technologies for Pathologic Diagnosis: Viral Infections | | |
| 8:30 - 9:15 AM | **STATE-OF-THE-ART-LECTURE**<br>Cystic Kidney Disease is Caused by Defects of the Cilia-Centrosome Complex | | | | |
| 9:15 - 9:30 AM | *Break, Exhibits Open* | | | | |
| 9:30 - 11:15 AM | **SYMPOSIUM**<br>Chronic Dialysis | **SYMPOSIUM**<br>Glomerular Disease | **SYMPOSIUM**<br>Metabolic Disorders | **SYMPOSIUM**<br>Value and Implications of Screening for Kidney Disease | |
| 11:15 - 11:30 AM | *Break, Exhibits Open* | | | | |
| 11:30 AM - 12:20 PM | **ORAL POSTER SESSION**<br>General Nephrology I<br>*Abstracts O-1 -- O-8* | **ORAL POSTER SESSION**<br>Glomerular Disease I<br>*Abstracts O-9 -- O-16* | **ORAL POSTER SESSION**<br>Transplantation I<br>*Abstracts O-17 -- O-24* | **ORAL POSTER SESSION**<br>Acute & Chronic Renal Failure<br>*Abstracts O-25 -- O-32* | **ORAL POSTER SESSION**<br>Urinary Tract Infection<br>*Abstract O-33 -- O-40* |
| 12:20 - 1:00 PM | **POSTER SESSION I**<br>*Abstracts 1 - 319* | | | | |
| 1:00 - 2:30 PM | **CLINICOPATHOLOGICAL CONFERENCE** | **INTERNATIONAL REGISTRIES MEETING** | **JOINT PEDIATRIC NEPHROLOGY/ UROLOGY SYMPOSIUM** | | |
| 2:30 - 3:15 PM | **STATE-OF-THE-ART-LECTURE**<br>Transplant Tourism: Falling Through the Cracks: Who is Responsible for Continuing Care? | | | | |
| 3:15 - 3:45 PM | *Break, Exhibits Open* | | | | |
| 3:45 - 5:30 PM | **SYMPOSIUM**<br>Progress in AKI | **SYMPOSIUM**<br>Tubular Transport | **SYMPOSIUM**<br>Hypertension and Obesity | **SYMPOSIUM**<br>Growth | **JOINT PEDIATRIC NEPHROLOGY/ UROLOGY SYMPOSIUM** |
| 5:30 - 6:00 PM | *Break* | | | | |
| 6:00 - 10:00 PM | **PHARMACEUTICAL SPONSORED SATELLITE SYMPOSIUM** | | | | |

| Time | Tuesday, August 31, 2010 | | | | |
|---|---|---|---|---|---|
| 6:30 AM - 7:00 PM | *Registration Open* | | | | |
| 7:30 - 8:20 AM | **EARLY CLINICAL SESSION**<br>Anemia/Erythropoiesis-Stimulating Agents | **EARLY CLINICAL SESSION**<br>New Immunosuppressant's and Their Application | **EARLY CLINICAL SESSION**<br>Role of Genetic Testing in Diagnosing and Treating Kidney Disease | | |
| 8:30 - 9:15 AM | **STATE-OF-THE-ART-LECTURE**<br>Development of the Renal Glomerulus-Good Neighbors and Good Fences | | | | |
| 9:15 - 9:30 AM | *Break, Exhibits Open* | | | | |
| 9:30 - 11:15 AM | **SYMPOSIUM**<br>Podocyte Biology | **SYMPOSIUM**<br>CKD | **SYMPOSIUM**<br>Inductive Interactions: The Role of Genes, Transcription Factors, and Stem Cells in Urinary Tract Development | **SYMPOSIUM**<br>Renal Disease Around the World | **SYMPOSIUM**<br>Oxalosis |
| 11:15 - 11:30 AM | *Break, Exhibits Open* | | | | |
| 11:30 AM - 12:20 PM | **ORAL POSTER SESSION**<br>General Nephrology II<br>*Abstracts O-41 -- O-48* | **ORAL POSTER SESSION**<br>Glomerular Disease II<br>*Abstracts O-49 -- O-56* | **ORAL POSTER SESSION**<br>Transplantation II<br>*Abstracts O-57 -- O-64* | **ORAL POSTER SESSION**<br>Nutrition, Growth and Bone & Mineral Metabolism<br>*Abstracts O-65 -- O-72* | **ORAL POSTER SESSION**<br>Genetics<br>*Abstracts O-73 -- O-80* |
| 12:20 - 1:00 PM | **POSTER SESSION II**<br>*Abstracts 320 - 639* | | | | |
| 1:00 - 6:00 PM | **The NephCure Foundation Symposium**<br>Focus on Focal | | **The Cystinosis Research Network Symposium** | | |

| Time | Wednesday, September 1, 2010 | | | | |
|---|---|---|---|---|---|
| 6:30 AM - 7:00 PM | *Registration Open* | | | | |
| 7:30 - 8:20 AM | **EARLY CLINICAL SESSION**<br>Fundamental Acid-Base | **EARLY CLINICAL SESSION**<br>Estimating GFR: Clinical Utility | **EARLY CLINICAL SESSION**<br>Hypertension and Ambulatory Blood Pressure Monitoring | | |
| 8:30 - 9:15 AM | **STATE-OF-THE-ART-LECTURE**<br>Novel Approaches to Erythropoiesis in CKD | | | | |
| 9:15 - 9:30 AM | *Break, Exhibits Open* | | | | |
| 9:30 - 11:15 AM | **SYMPOSIUM**<br>Hot New Issues in Genetics | **SYMPOSIUM**<br>Advanced Imaging for Research and Practice | **SYMPOSIUM**<br>Clinical Transplantation | **SYMPOSIUM**<br>Bone and Mineral Metabolism | **SYMPOSIUM**<br>Translational Medicine |
| 11:15 - 11:30 AM | *Break, Exhibits Open* | | | | |
| 11:30 AM - 12:20 PM | **ORAL POSTER SESSION**<br>Renal Replacement Therapy<br>Abstracts O-81 -- O-88 | **ORAL POSTER SESSION**<br>Hypertension & Cardiovascular Disease<br>*Abstracts O-89 -- O96* | **ORAL POSTER SESSION**<br>Tubulointerstitial Disorders & Immunology of Renal Disease<br>*Abstracts O-97 -- O-104* | **ORAL POSTER SESSION**<br>Congenital Abnormalities & Perinatal Nephrology<br>*Abstracts O-105 -- O-112* | **ORAL POSTER SESSION**<br>Developmental Nephrology, Renal Physiology, and Perinatal Nephrology<br>*Abstract O-112 -- O-120* |
| 12:20 - 1:00 PM | **POSTER SESSION I**<br>*Abstracts 640 - 959* | | | | |
| 1:00 - 2:30 PM | *Break / IPNA Business Meeting* | | | | |
| 2:30 - 3:15 PM | **STATE-OF-THE-ART-LECTURE**<br>Uric Acid and Hypertension | | | | |
| 3:15 - 3:45 PM | *Break, Exhibits Open* | | | | |
| 3:45 - 5:30 PM | **SYMPOSIUM**<br>Critical Care Nephrology | **SYMPOSIUM**<br>HUS and MPGN Overlap Syndromes | **SYMPOSIUM**<br>Renal Cystic Disease | **SYMPOSIUM**<br>Transplantation Biology | **SYMPOSIUM**<br>Clinical Bone Disease in Children with CKD |
| 7:00 - 10:00 PM | *Congress Reception* | | | | |

| Time | Thursday, September 2, 2010 | | | |
|---|---|---|---|---|
| 6:30 AM - 12:00 NN | *Registration Open* | | | |
| 7:30 - 8:20 AM | **EARLY CLINICAL SESSION**<br>Transitioning to Adult Care | **EARLY CLINICAL SESSION**<br>Challenges of Chronic Dialysis | **EARLY CLINICAL SESSION**<br>Therapeutic Approaches to Metabolic Bone Disease | |
| 8:30 - 9:15 AM | **STATE-OF-THE-ART-LECTURE**<br>Progress & Problems: Review of IPNA Fellowship Training Program | | | |
| 9:15 - 10:00 AM | **AWARDS CEREMONY** | | | |
| 10:00 - 10:15 AM | *Break* | | | |
| 10:15 - 12:00 PM | **SYMPOSIUM**<br>Cardiovascular Disease in CKD | **SYMPOSIUM**<br>Ethics | **SYMPOSIUM**<br>Interventional Genetics | **SYMPOSIUM**<br>Fluid-Electrolyte Pathophysiology and Management (Physiology) |

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# The 15th Congress of the International Pediatric Nephrology Association

# ABSTRACTS

**All presenters are required to disclose relevant conflicts of interest.**
**All such disclosures are published within the Abstract Book following each abstract.**
**Any presenters who have nothing to disclose have been omitted from the disclosure listing.**

## Glomerular Disease

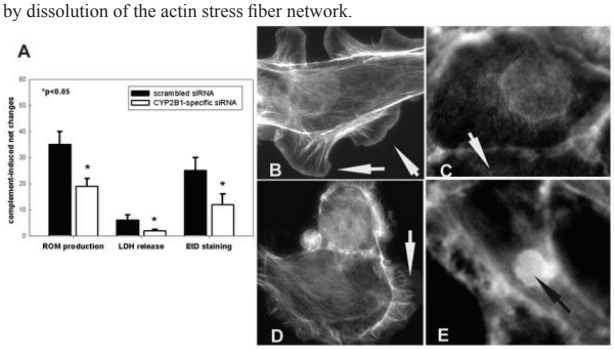
### Abstract# 1
**(O-1)**

**Cytochrome P450 2B1 (CYP2B1) Mediates Complement-Induced Sublytic Injury to Glomerular Epithelial Cells (GEC)** R. Baliga, N. Tian, I. Arany. *Pediatrics, University of Mississippi Medical Center, Jackson, MS, United States.*

**Objectives:** CYP2B1 is an important source for reactive oxygen metabolites (ROM) mediated injury in Passive Heymann Nephritis (PHN) (JASN, 18:295A, 2007). Effacement of the foot processes and disorganization of the cytoskeleton are characteristic features leading to proteinuria.The current study was to determine whether CYP2B1 mediates anti-Fx1A-induced ROM production and collapse of the actin cytoskeleton in GEC.

**Methods:** GEC that overexpress the CYP2B1 gene were transfected with scrambled or CYP2B1-specific siRNA prior to treatment with anti-Fx1A followed by incubation with heat-inactivated (HIS) or normal human serum (NS). ROM production, LDH release, Ethidium homodimer (EtD) staining (to quantitate membrane permeabilization) and disorganization of the actin filament was determined.

**Results:** NS(complement) significantly increased ROM production, LDH release, EtD staining(A)and collapse of the actin cytoskeleton (B-C) with loss of lamellopodia ( arrows in B-C). Knockdown of the CYP2B1 gene attenuated ROM production, LDH release and EtD staining (A) as well as preserved the actin filament and lamellopodia (D). EtD staining (arrow in E) was accompanied by dissolution of the actin stress fiber network.




**Conclusions:** CYP2B1 plays a pivotal role in oxidative stress leading to sublethal injury and collapse of the cytoskeleton in antiFx1A-induced PHN.

### Abstract# 2
**(O-2)**

**Efficacy of Rituximab in Childhood Nephrotic Syndrome** S. Adalat, J. Taylor, C. Booth, M. McCullough, S. Waller, S. Rigden, M. Sinha, A. Koziell. *Paediatric Nephrology, Evelina Children's Hospital, London, United Kingdom.*

**Objectives:** To explore the efficacy of rituximab, a CD20 specific B-lymphocyte depleting antibody in childhood nephrotic syndrome.
We studied clinical response in 25 cases of idiopathic NS in children pre and post rituximab for 36 months. Immunological investigations were performed to acquire clues as to mechanism of action.

**Methods:** 25 cases (8F:17M) were treated with rituximab (1-4 courses). 10 were steroid resistant and 15 steroid dependent. Histological diagnoses were minimal change disease (MCND) 15; focal segmental glomerulosclerosis (FSGS) 9, C1q nephropathy 1. Median duration of therapy prior to rituximab was 24 months (range 1-156mo). Cases were examined for clinical and immunological response.

**Results:** 20/25 cases responded to the 1st course of rituximab. Non-responders were either steroid resistant and/or in a protacted nephrotic state: 3 had longstanding FSGS and advanced disease.1 did not deplete B cells. 1 patient had a partial response. Only one experienced significant adverse side effects. Nephrotic remission was directly correlated with CD20 depletion and relapse with CD20 recovery. 90% of cases required re-treatment. Re-emergence of sensitivity to steroids +/- calcineurin inhibitors was observed. No reduction in rituximab efficacy occurred on repeat dosing. No consistent changes in immunological parameters were detected.

**Conclusions:** Rituximab is safe and efficacious in childhood NS, and appears to be more effective in steroid sensitive than steroid resistant disease. This data indicates an integral role for B lymphocytes in the pathogenesis of NS. The mechanism is likely to be more complex than simply depletion of peripheral B lymphocyte subsets.

### Abstract# 3
**(O-3)**

**Oxidative Stress and Aberrantly Glycosylated IgA1 as Risk Factors for IgA Nephropathy** R. Camilla,[1] H. Suzuki,[2] V. Daprà,[1] E. Loiacono,[1] L Peruzzi,[1] A. Amore,[1] G.M. Ghiggeri,[3] G. Mazzucco,[4] F. Scolari,[5] A.G.G. Gharavi,[6] G. Appel,[6] J. Novak,[2] B.A. Julian,[2] S. Troyanov,[7] R. Coppo.[1] *[1]Nephrology Dial Transplant, R Margh Hosp, Turin, Italy; [2]Microbiology Medicine, Un of Alabama at Birmingham, Birmingham, AL, United States; [3]Nephrology Lab, Gaslini Inst, Genoa, Italy; [4]Biom Science Oncology, Un, Turin, Italy; [5]Nephrology, Montichiari Hosp, Brescia, Italy; [6]Medicine, Columbia Un, New York, NY, United States; [7]Nephrology, Hop Sacré Coeur, Montreal, QC, Canada.*

**Objectives:** Aberrantly glycosylated IgA1 is thought to be involved in pathogenesis of IgA nephropathy (IgAN) and a role for oxidative stress has been suggested. Aim of the study was to evaluate these markers with respect to clinical activity and progression of IgAN.

**Methods:** We measured levels of galactose-deficient IgA1 (total levels,Gd-IgA1 and % of IgA, %HAA), advanced oxidation protein products (AOPPs) and albumin free SH groups (SH-Alb) in 292 IgAN patients (67 children) and 69 controls (HC) and correlated with clinical data.

**Results:**

| | IgAN | HC |
|---|---|---|
| Gd-IgA1 (U/ml) | 166±139* | 56±34 |
| %HAA | 46±16* | 33±12 |
| AOPPs (µmol/L) | 99±49* | 48±37 |
| SH-Alb (AU) | 3.6±2.4* | 7.7±3.5 |

*p<0.0001 vs HC

AOPPs correlated with Gd-IgA1 and %HAA (p<0.01). Patients with positive AOPPs, SH-Alb, Gd-IgA1 or %HAA had greater proteinuria (UP) at sampling and time average (TA) UP at follow-up (p<0.01). By multivariate analysis %HAA and AOPPs were independent predictors of TA-UP. AOPPs correlated with GFR decline (p<0.01). The association of high levels of AOPP and SH-Alb or AOPPs and %HAA were prognostic factors for GFR decline (p<0.01).

**Conclusions:** Increased levels of aberrantly glycosylated IgA1 and elevated markers of oxidative stress in IgAN are candidate prognostic factors for GFR decline.

### Abstract# 4
**(O-5)**

**The Administration of the Agonist of Angiotensin-(1-7), AVE0991, Improved Inflammation and Proteinuria in Experimental Nephrotic Syndrome** K.D. Silveira,[1] R.A.S. Santos,[1] L.C. Barroso,[2] C.X. Lima,[2] M.M. Teixeira,[2] A.C. Simoes e Silva.[3] *[1]Physiology, Federal University of Minas Gerais, Belo Horizonte, Minas Gerais, Brazil; [2]Biochemistry and Immunology, Federal University of Minas Gerais, Belo Horizonte, Minas Gerais, Brazil; [3]Pediatrics, Federal University of Minas Gerais, Belo Horizonte, Minas Gerais, Brazil.*

**Objectives:** The aims were to investigate the role of the oral agonist of Angiotensin-(1-7) receptor Mas, AVE0991, in experimental nephrotic syndrome and to compare it with losartan.

**Methods:** Nephrotic syndrome was induced by a single injection of adriamicin(10mg/kg) in male BALB/c mice. After 7 days, animals were allocated into 3 groups: oral administration of AVE0991 (3mg/kg), or losartan (10mg/kg), or vehicle. All treatments were daily given for 7 days per gavage and after that 24-hour urine and blood samples were collected. Then, animals were sacrificed to remove the kidneys. Renal function parameters, urinary cytokines, renal histology and the relative number of neutrophils were evaluated.

**Results:** AVE0991 and losartan partially improved renal function parameters. The albumin:creatinin ratio was significantly and similarly reduced by both treatments if compared to vehicle administration. Free water and osmolal clearance were normalized by both treatments, whereas creatinine clearance remained unchanged. Both treatments promoted a similar reduction (~25%) in the number of renal tissue neutrophils, but AVE0991 produced a more pronounced reduction in urinary concentrations of IL-6 (49%) and of TGFβ (57%) than losartan. Renal histology was also significantly improved by AVE0991.

**Conclusions:** AVE0991 emerges as a potential therapeutic agent for nephrotic syndrome.

## Abstract# 5
**(O-6)**

**PodoNet: International Registry for Steroid-Resistant Nephrotic Syndrome (SRNS)** A. Trautmann,[1] F. Ozaltin,[2] A. Noyan,[3] M. Azocar,[5] F. Emma,[4] F. Schaefer,[1] The PodoNet Consortium. *[1]Univ.of Heidelberg, Heidelberg, Germany; [2]Hacettepe Univ., Ankara, Turkey; [3]Curukova Univ., Adana, Turkey; [4]Ospedale Pediatrica Bambino Gesu, Rome, Italy; [5]Hosp.L.C.Mackenna, Santiago de Chile, Chile.*

**Objectives:** Classification of SRNS is challenging due to variable responsiveness to intensified immunosuppression (IS) and incomplete knowledge of genetic causes. Also, existing empiric therapies largely lack a trial evidence base.

**Methods:** The PodoNet consortium has recently established an Internet-based SRNS registry collecting clinical, pharmacological, histopathological and genetic information (www.podonet.org).

**Results:** Among 480 children reported to date by 30 centers in 12 countries, SRNS first manifested at age <1y in 6%, 1-5 y in 49%, 6-12 y in 29% and >12y in 17% of pts. Histopathology was FSGS in 65%, MCGN in 17%, MesPGN in 13%, global GS in 4%, and DMS in 1 %. At latest update, 45% of children had normal GFR and 17% had reached ESRD. 35% of conservatively treated children were in remission. 43% received RAS antagonists, 35% steroids, 28% CNI inhibitors, 6% MMF.

24% of patients originate from consanguineous families. Among the 276 pts with family history information, 56(20%) were suggestive of recessive and 13 (4.7%) of dominant inheritance. Disease-causing mutations were found in 28 of 206 subjects tested in *NPHS2*, 5/15 in *NPHS1*, and 6/169 in *WT1*.

**Conclusions:** The information collected so far allows preliminary characterization of IS-responsive and multidrug-resistant (including genetic) forms of SRNS. The PodoNet registry will help defining the indications, efficacy and safety of pharmacological therapies and hopefully lead to the discovery of new genetic causes of SRNS.

## Abstract# 6
**(O-7)**

**ADAMTS13 Expression in Glomerular Endothelial Cells** R. Tati, A. Kristoffersson, A.-l. Ståhl, D. Motto, S. Satchell, P. Mathieson, M. Manea, D. Karpman. *Department of Pediatrics, Clinical Sciences Lund, Lund University, Lund, Sweden; Lund University, Lund, Sweden; Lund University, Lund, Sweden; University of Iowa College of Medicine, Iowa City, IA, United States; University of Bristol, Bristol, United Kingdom; University of Bristol, Bristol, United Kingdom; Lund University, Lund, Sweden; Lund University, Lund, Sweden.*

**Objectives:** The aim of this study was to investigate expression and function of the von Willebrand factor (VWF) cleaving protease ADAMTS13 in glomerular endothelial cells.

**Methods:** Immunohistochemistry was performed on renal tissue from human controls and from ADAMTS13 wild-type (*Adamts13$^{B/129+/+}$*) and knock-out (*Adamts13$^{B/129-/-}$*) mice. ADAMTS13 mRNA and protein expression in human glomerular endothelial cells and human dermal microvascular endothelial cells were investigated by real-time PCR, flow cytometry and immunofluorescence. Bioactivity was demonstrated by cleavage of VWF assayed by multimer structure analysis and immunoblotting.

**Results:** Results showed ADAMTS13 in the glomerular endothelial cells from normal human tissue and wild-type mice but not in tissue from knock-out mice. ADAMTS13 mRNA and protein were detected in both glomerular and dermal microvascular endothelial cells. ADAMTS13 bioactivity was demonstrated in lysates of both endothelial cells. The protease was secreted to the glomerular endothelial cell membrane.

**Conclusions:** The results indicate that biologically active ADAMTS13 is expressed in glomerular endothelial cells. The proteolytic activity may have a local protective effect under the conditions of high shear stress present in glomerular capillaries.

## Abstract# 7
**(O-8)**

**Natural Remission Rate and Prognostic Factors in Childhood IgA Nephropathy with Minimal Change or Focal Mesangial Proliferation** Y. Shima,[1] K. Nakanishi,[1] H. Togawa,[1] K. Nozu,[2] R. Tanaka,[3] K. Iijima,[2] N. Yoshikawa.[1] *[1]Pediatrics, Wakayama Medical University, Wakayama, Wakayama, Japan; [2]Pediatrics, Kobe University Graduate School of Medicine, Kobe, Hyogo, Japan; [3]Pediatric Nephrology, Hyogo Prefectual Kobe Children's Hospital, Kobe, Hyogo, Japan.*

**Objectives:** It is generally considered that the prognosis of childhood IgA nephropathy (IgAN) is unpredictable even in cases showing minimal change or focal mesangial proliferation (MC/FMP). Some of them show natural remission without medication. However, details of the incidence and prognostic factors remain unclear. The purpose of this study is to clarify them in childhood IgAN with MC/FMP.

**Methods:** We analyzed retrospectively consecutive 98 children with newly diagnosed childhood IgAN with MC/FMP from January 1972 to December 2000 and observed without medication.

**Results:** Mean observation period was 7.5±4.6 years. Among 98 children, 44 children (44.9%) showed natural remission. Median remission time was 5.5 years. There was no significant difference in clinical findings depending on remission. On the other hand, there were significant differences in pathological findings (scleroses, crescents, mesangial proliferation (MP), and extracapillary lesions (EL)). The prognostic factor related to natural remission was the total ratio of glomeruli showing MP and EL in both univariate and multivariate analyses. The cut off point of the ratio in ROC curve was 25%.

**Conclusions:** The natural remission rate in childhood IgAN with MC/FMP was higher than expected. If the total ratio of glomeruli presenting MP and EL is under 25%, we should refrain from aggressive treatment, because such patients are likely to show natural remission.

## Abstract# 8
**(O-23)**

**Insights into the Uptake and Release of Complement Factor H by Platelets** V. van Eimeren,[1] F. Pluthero,[1] W. Kahr,[1] C. Licht.[2] *[1]Haematology/Oncology, The Hospital for Sick Children, Toronto, ON, Canada; [2]Nephrology, The Hospital for Sick Children, Toronto, ON, Canada.*

**Objectives:** Factor H (CFH), key inhibitor of the alternative complement pathway, plays a role in platelet stimulation as shown by platelet aggregation via plasma of an atypical HUS patient with CFH antibodies (*Licht et al, Blood 2009*). We have also shown *in vivo* / *in vitro* platelet uptake and release of exogenous CFH. Glycoprotein IIb/IIIa (GPIIb/IIIa) is involved in platelet uptake of exogenous proteins (e.g. fibrinogen) via endocytosis. - We investigated the mechanisms of platelet uptake and release of CFH.

**Methods:** To determine whether GPIIb/IIIa is involved in CFH uptake we incubated platelets of Glanzmann thrombasthenia patients lacking functional GPIIb/IIIa with labeled fibrinogen or CFH and visualized uptake via confocal laser microscopy. To investigate CFH release from normal platelets we stimulated platelets with various agonists and detected the release of specific proteins from various platelet compartments via immunoblotting.

**Results:** Fibrinogen uptake was absent in platelets lacking functional GPIIb/IIIa, however uptake and distribution of labeled CFH was not affected, suggesting that CFH uptake in platelets does not involve GPIIb/IIIa. Platelet stimulation studies demonstrated differential CFH release in the absence of α-granule secretion and release of other cytoplasmic proteins.

**Conclusions:** We conclude that CFH uptake in platelets is not GPIIb/IIIa dependent but mediated by a distinct mechanism. Furthermore, CFH release from platelets is independent from α-granule secretion and overt cytoplasmic release mechanisms. Further studies will elucidate the precise mechanism of uptake and release of CFH from platelets.

**DISCLOSURE:** *Licht, C.*: Grant/Research Support, Optherion; Consultant, Optherion; Alexion.

## Abstract# 9
**(O-24)**

**The Inhibition of Notch Pathway Reactivation in Podocytes** E. Tanaka,[1,2] K. Asanuma,[1] M. Takagi,[1] F. Kodama,[1] A. Kawasaki,[3] H. Yagita,[3] S. Mizutani,[2] Y. Tomino.[1] *[1]Division of Nephrology, Department of Internal Medicine, Juntendo University School of Medicine, Tokyo, Japan; [2]Department of Pediatrics and Developmental Biology, Tokyo Medical and Dental University, Tokyo, Japan; [3]Department of Immunology, Juntendo University School of Medicine, Tokyo, Japan.*

**Objectives:** Notch signaling pathway regulates cell differentiation in multiple developing organ systems, including kidney. It was recently reported that reactivation of Notch1 in podocytes caused proteinuria, apoptosis and podocyte loss, leading to glomerulosclerosis. Here we report that inhibition of Notch pathway by antibodies(Abs) against Notch1 ligand Jagged1 prevents Notch1 reactivation in cultured podocytes.

**Methods:** We examined Notch1 reactivation in adriamycin(ADR) induced nephropathy, a mouse model of nephrosis and FSGS, and expressions of Notch receptors and ligands on cultured podocytes. Moreover, we investigated the expressions of Jagged1 and Notch1 reactivation in vitro under ADR induced podocyte damage with anti-Jagged1 Abs.

**Results:** Notch1 reactivation in podocytes and massive proteinuria were induced by ADR injection in mice. Cultured podocytes under physiological condition had only Notch2 receptor on the surface. ADR induced podocyte injury increased Jagged1 expression, and Notch1 reactivation was caused within 2 hrs. Although injured podocytes by ADR with anti-Jagged1 Abs showed up-regulated Jagged1, Notch1 reactivation was not detected.

**Conclusions:** Our results revealed the relation of Notch pathway to progression of nephrosis and FSGS. Successful inhibition of Notch1 reactivation by anti-Jagged1 Abs in podocytes may shed light on the novel therapy for nephrosis and FSGS.

## Abstract# 10

**Occurence of Rheumatic Heart Disease and Acute Glomerulonephritis in an African Child** O.T. Adedoyin, O.P. Fatoye, S. Anoba, A. Saka, A. Adeniyi. *Paediatrics and Child Health, University of Ilorin Teaching Hospital, Ilorin, Kwara, Nigeria; Paediatrics and Child Health, University of Ilorin Teaching Hospital, Ilorin, Kwara, Nigeria; Paediatrics and Child Health, University of Ilorin Teaching Hospital, Ilorin, Kwara, Nigeria; Paediatrics and Child Health, University of Ilorin Teaching Hospital, Ilorin, Kwara, Nigeria; Paediatrics and Child Health, University of Ilorin Teaching Hospital, Ilorin, Kwara, Nigeria.*

**Objectives:** To report a 13 year old African girl with features suggestive of rheumatic heart disease and acute glomerulonephritis.

**Methods:** The case file of the patient was reviewed for relevant clinical and laboratory findings.

**Results:** We report a 13 year old girl from South West Nigeria who presented in our hospital with 3 month history of cough, orthopnea and breathlessness and three weeks history of bilateral pitting oedema. There was previous history of leg swelling. There was associated raised jugular venous pressure and pan systolic murmur radiating to the axilla. Blood pressure was 170/120mmHg at admission. Proteinuria was 3+ at admission, however serum protein, albumin and triglyceride were within normal range. Serum creatinine was on the upward trend from 315µmol/l at admission. Erythrocyte sedimentation rate was 45mm/hr. Urine output was between 0.3-1.1mls/kg/hr and the oedema never subsided. Echocardiography suggested rheumatic heart disease and mitral incompetence.

**Conclusions:** In view of the presence of cardiac symptom and increased erythrocyte sedimentation rate with oedema, hypertension, azotaemia and previous history of body swelling, a suspicion of rheumatic heart disease and acute glomerulonephritis is suspected.

## Abstract# 11

**Progression to Chronic Kidney Disease in Children with FSGS: A Single Center Study** F. Amanullah,[1] F. Akhtar,[2] A.H. Rizvi,[2] M. Mubarak,[2] A. Sohaila.[3] *[1]Pediatrics, The Indus Hospital, Karachi, Sindh, Pakistan; [2]Nephrology, Sindh Institute of Urology and Transplantation, Karachi, Sindh, Pakistan; [3]Pediatrics, Aga Khan University Hospital, Karachi, Sindh, Pakistan.*

**Objectives:** To study the clinical course of childhood FSGS and determine the possible predictive factors of chronic kidney disease (CKD) in Pakistani children presenting to the Sindh Institute of Urology and Transplanatation.

**Methods:** We retrospectively reviewed the records of 102 children and adolescents with biopsy proven idiopathic FSGS, who were treated at the Sindh Institute of Urology and Transplantation between 8/1995- 8/2007. Clinical and laboratory parameters at baseline, response to treatment, development of CKD (chronic kidney disease) and histopathological details were analyzed.

**Results:** A cohort of 102 children with a mean age of 5.9 +/- 4.1 years and a male: female ratio of 1.37:1 was identified. After a mean follow-up time of 5 years, 36/102 (35%) developed CKD. On univariate analysis, age greater than 6 years at onset (50% vs. 27% p-0.02), microhematuria at presentation (64% vs. 21% p-0.001), steroid resistant course (92% vs. 41 % p-0001) were significantly associated with risk of developing CKD. Patients who progressed to CKD were more likely to be partially responsive to cyclosporine (80% vs. 22% p-0.0001).

**Conclusions:** In terms of histopathological features, the patients with CKD were more likely to have moderate tubular atrophy and a higher percentage of segmentally and globally sclerosed glomeruli.

Renal survival rates for childhood FSGS were somewhat worse in our study, perhaps indicating a more aggressive disease.

## Abstract# 12

**A Randomized Controlled Trial on the Effect of Oral vs. Intravenous Furosemide on the Resolution of Volume Overload in Patients with Acute Post-Streptococcal Glomerulonephritis** C.L. Cifra, F.E. Anacleto, J. Mantaring III. *Pediatrics, College of Medicine-Philippine General Hospital University of the Philippines Manila, Manila, Philippines.*

**Objectives:** To determine if the administration of oral versus intravenous furosemide will lead to any significant difference in resolution of volume overload in children with post-streptococcal glomerulonephritis.

**Methods:** Children 4-12 years old clinically diagnosed with acute post-streptococcal glomerulonephritis and admitted at the Philippine General Hospital were considered for inclusion. Twenty-four patients (12 in the intravenous group, 12 in the oral group) were recruited and completed the study. Participants were randomized into treatment (oral) and control (intravenous) groups, after which they were given 2 mg/kg/dose of either the oral or intravenous preparation every 6 hours until the 24th hour of hospital stay. The differences in mean urine output, weight loss from admission, and blood pressure reduction were determined using the student's t-test. Outcomes were analyzed by intention-to-treat. A p value of < 0.05 was considered statistically significant.

**Results:** Results show no significant difference in all outcomes measured: urine output (2.31 ± 0.97 vs. 2.28 ± 0.82, p = 0.943), weight loss (1.73 ± 0.60 vs. 1.71 ± 1.23, p = 0.959), systolic BP reduction (29.82 ± 9.99 vs. 28.00 ± 9.64, p = 0.653) and diastolic BP reduction (17.28 ± 10.47 vs. 22.76 ± 9.66, p = 0.197).

**Conclusions:** The study shows no significant difference in the reduction of volume overload, mean urine output, weight loss, and blood pressure given oral vs. intravenous furosemide after 24 hours of treatment.

## Abstract# 13

**Increased Prevalence of Focal Segmental Glomerulosclerosis in Childhood Nephrotic Synrome in Ibadan Nigeria** A.O. Asinobi,[1] A.D. Ademola,[2] B.N. Yusuf,[3] C.A. Okolo.[4] *[1]Department of Paeditrics, University College Hospital, Ibadan, Nigeria; [2]Department of Paeditrics, University College Hospital, Ibadan, Nigeria; [3]Department of Paediatrics, University College Hospital, Ibadan, Nigeria; [4]Department of Pathology, University College Hospital, Ibadan, Nigeria.*

**Objectives:** To identify any changes in the histologic pattern of childhood nephrotic syndrome (NS) in Ibadan.

**Methods:** Between 1997 and 2000, consecutive presenting paediatric NS patients with no contraindications were biopsied. Between 2006 and 2009, those who failed to go into remission following the administration of oral prednisolone at 60mg/m$^2$/day for at least 4weeks and those with suspected secondary NS were biopsied. Only light microscopy was feasible.

**Results:** Of the 20 biopsied between 1997 and 2000, FSGS predominated (35%), followed by Membranoproliferative glomerulonephritis (MPGN) in 25%. Focal glomerulonephritis, Diffuse glomerulonephritis, Chronic glomerulonephritis(GN) were seen in 10% each while MCNS and Chronic glomerulosclerosis were found in 5% each.

Of the 20 biopsied between 2006 and 2009, the indication was SRNS in 9; others were secondary NS.

Overall, FSGS was seen in 40%, MCNS 25%, MPGN 20%, and 5% each for Diffuse Proliferative GN, Membranous nephropathy and Chronic GN. Among the SRNS patients, 66.7% were due to FSGS, 22.2% MPGN and 11.1% MCNS.

**Conclusions:** FSGS predominated in the total population studied and was clearly the predominant histology in the SRNS patients. This shows a change in pattern when compared with a previous report from the same unit in which MPGN was predominant.The lack of facilities for IF studies and EM is a big limitation in our setting and help is needed.

## Abstract# 14

**Efficacy of Rituximab (RTX) in Difficult Steroid Resistant (SRNS) & Steroid Dependent Nephrotic Syndrome (SDNS): Multicentric Experience** A. Bagga,[1] A. Gulati,[1] P. Hari,[1] A. Moudgil,[2] S. Jordan.[3] *[1]All India Institute of Medical Sciences, New Delhi, India; [2]Childrens National Medical Center, Washington, DC, United States; [3]Cedars Sinai Medical Center, Los Angeles, CA, United States.*

**Objectives:** To examine the efficacy & safety of RTX in inducing & maintaining remission in difficult SRNS & SDNS.

**Methods:** Patients with SRNS or SDNS, refractory to standard therapy or having calcineurin inhibitor (CNI) nephrotoxicity were treated with RTX & followed ≥12-mo. RTX (375 mg/m$^2$ IV q wk) was given in 2 & 4 doses for SDNS & SRNS respectively. Therapy with prednisone & CNI was tapered; all received ACE inhibitors.

**Results:** Baseline features of 57 patients (36 boys) with SRNS/SDNS are shown

| | SRNS (n=33) | SDNS (n=24) |
|---|---|---|
| Age, yr | 13±9 | 12±3 |
| Duration of illness, yr | 9±3 | 8±2 |
| PREVIOUS THERAPY | | |
| Long-term/IV steroids | 33/9 | 24/2 |
| Levamisole | 2 | 16 |
| Mycophenolate mofetil | 3 | 15 |
| Cyclophosphamide | 20 | 22 |
| CNI | 24 | 15 |
| Duration of CNI use, mo | 23±17 | 21±10 |
| CNI toxicity | 11 | 5 |
| Cushingoid/cataract | 2/0 | 24/8 |

***SRNS.*** 9 patients had complete & 7 had partial remission. Response was similar in initial *vs.* late resistance; those with minimal change (MCD) did better than FSGS.

| % | Initial resistance(n=24) | Late resistance(n=9) | MCD*(n=17) | FSGS*(n=16) |
|---|---|---|---|---|
| Complete remission | 21 | 44 | 41 | 12 |
| Partial remission | 25 | 11 | 23 | 19 |
| Non response % | 54 | 44 | 35 | 68 |

*P 0.08

***SDNS.*** Remission was sustained in 20 (84%) at 12-mo & 17 (71%) at 17±6 mo. Relapses before & 12-mo after RTX were 4±0.4 & 0.2±0.3 episode/pt/yr [mean difference 3.8; P 0.00]. There were no adverse events.

**Conclusions:** Therapy with rituximab was promising, since it induced a sustained partial/complete remission in 48% patients with refractory SRNS & highly significant reduction in relapse frequency in difficult SDNS.

## Abstract# 15

**Cyclosporine A Plus Low-Dose Steroid Treatment in Children with Idiopathic Nephrotic Syndrome** A. Bal,[1] M. Anil,[1] O.D. Kara,[2] O. Yavascan,[1] S. Sen,[1] N. Aksu.[1] *[1]Pediatric Nephrology, Izmir Tepecik Teaching and Research Hospital, Izmir, Turkey; [2]Pediatric Nephrology, Ege University Faculty of Medicine, Izmir, Turkey.*

**Objectives:** Cyclosporine A (CsA) is widely considered as the treatment of choice for steroid-resistant (SRNS) or dependent (SDNS) nephrotic children. However, long-term clinical outcome of patients treated with CsA is unclear. The aim of this study was to evaluate the efficacy of CsA plus low-dose steroid treatment in children with nephrotic syndrome (NS).

**Methods:** We reviewed the clinical outcome in children with idiopathic NS under CsA (5 mg/kg/day) plus low-dose steroid (30mg/m$^2$/day) treatment. Previously, all children had received corticosteroids. Indications for treatment included SDNS in 34 patients and SRNS in 16 patients. The median age at onset of NS was 25 months. Renal histology showed focal segmental glomerulosclerosis in 21(42%) patients, diffuse mesangial glomerulonephritis in 18(36%), minimal change disease in 8(16%), immunoglobulin M nephropathy in 2(4%) and membranous glomerulonephritis in 1(2%).

**Results:** The observation periods were 38.2±31.1 (1910 pt-mos) and 50.4±42.9 (2524 pt-mos) months before and after CsA treatment, respectively. The relapse rate was significantly higher before CsA (one episode (ep)/9.84 pt-mos, 3.8±3 relapse per year) than after CsA (one ep/33.65 pt-mos, 1.5± 1.8 relapse per year) ($p<0.05$). Under CsA therapy, the relapse rates showed no differences between SDNS and SRNS patients ($p>0.05$).

**Conclusions:** We conclude that CsA plus low-dose steroid treatment in inducing remission in both SDNS and SRNS patients seems to be a good choice. Therefore, CsA therapy should be considered in these patient groups to achieve remission.

## Abstract# 16

**The Study of PBMCs T-Helper Cell Subpopulation Th1/Th2 for Children with Primary Nephrotic Syndrome** Y. Bao, N. Zhou, H.K. Huang, W. Ren. *Dept. of Nephrology, Xi'an Children's Hospital, Xi'an, Shaanxi Province, China.*

**Objectives:** Aims of the study are to detect the expression of T-helper cell subpopulation $CD^+_4CD_{45}RA^+$ and $CD^+_4CD_{45}RO^+$ for children with primary nephrotic syndrome(PNS),to know the change and the relationship of those cells, discuss the immune mechanism of PNS.

**Methods:** All subjects are children with PNS hospitalized in Xi'an Children's hospital. Subject group has 37 cases, male 29 cases, famale 8 cases, the youngest is 1 year and 3 months old, the oldest is 13 years old, mean age 5 year and 6 month. Control group has 18 cases; male 11 cases, famale 7 cases, mean age 5 year and 3 month. Comparing with subject group, age and sex have no difference in statistics. Study procedure: The $CD^+_4CD_{45}RA^+$, $CD^+_4CD_{45}RO^+$ cell is detected in fresh blood sample by FCM. using PE/FITC double marked McAb.

**Results:** $CD^+_4$ cell of PNS increase slightly comparing with control group, but has no difference in statistics, $P>0.05$, 2.$CD_{45}RA^+$ expression increase obviously, $P<0.01$. $CD_{45}RO^+$ expression decrease obviously, $P<0.01$.3.$CD^+_4CD_{45}RA^+$ cell increase greatly, $P<0.01$. $CD^+_4CD_{45}RO^+$ cell decrease obviously than control, $P<0.01$. The ratio of $CD^+_4CD_{45}RA^+/CD^+_4CD_{45}RO^+$ increase obviously than control, $P<0.01$.

**Conclusions:** The studies show that children with PNS exist many kinds of immunal disord than control, 1.$CD_{45}RA^+$ expression increase, $CD_{45}RO^+$ expression decrease, it shows that there is lower immunologic function in children with PNS. 2.In children with PNS T-helper cell subpopulation $Th_1$ cell increase obviously, $Th_2$ cell decrease obviously. The ratio of $Th_1/Th_2$ is increase greatly comparing with control group, it shows a $Th_1$ dominance response.

## Abstract# 17

**Polyarteritis Nodosa Glomerulopathy in the Childhood – Case Report** M.J. Barcia, F. Hamamoto, J. Rosa, F. Leão, M.A. Cançado, J.T. Carvalhães. *Nefrologia Pediátrica, Universidade Federal de São Paulo, São Paulo, São Paulo, Brazil.*

**Objectives:** Polyarteritis nodosa (PAN) is a rare systemic vasculitis defined as the presence of necrotizing vasculitis affecting the wall of vessels, with little or no immune process, evolving with aneurysms and their complications.The prognosis of PAN depends on the severity and reversibility of renal involvement, which has a variety of presentations. The objective is to describe a case of 2 years old boy, at the time, with gross hematuria and hypertensive crises.

**Methods:** Diagnostic exams in sequence, based on cost-effectiveness and invasiveness to define etiologic case - Urianalysis, blood exams, US, citoscopy, renal biopsy, Angioresonance and arteriography.

**Results:** The patient presented in urianalysis with countless erythrocyte, leucocytes and proteinuria (26mg/kg/d) and negative urine culture. He had no hypercalciuria or hyperuricosuria and normal albumin and cholesterol. Renal biopsy showed segmental sclerosis and discrete area of interstitial fibrosis, absence of fluorescent deposits in glomeruli and basal membrane intact. There was no abnormal findings in US and the cistoscopy evidenced hematuria. Angioresonance showed an image suggestive of an aneurysm in medium lobe of right kidney. Then, renal and mesenteric arteriography was performed and we found microaneurysms diffusely distributed throughout the renal parenchyma and microaneurysms in first jejunal branches of superior mesenteric artery.

**Conclusions:** Based on the findings of arteriography the diagnosis was made and treatment has began. PAN is a rare vasculitis that can affect children and cause serious complications, including the possibility of bleeding.

## Abstract# 18

**Henoch-Schonlein Purpura Nephritis in Children** F. Bastug,[1] S. Tülpar,[1] R. Düsünsel,[1] Z. Gündüz,[1] H.M. Poyrazoglu,[1] H. Akgün,[2] T.E. Patiroglu,[2] K. Yilmaz,[1] S. Köse.[1] *[1]Pediatric Nephrology, Erciyes University Medical Faculty, Kayseri, Turkey; [2]Pathology, Erciyes University Medical Faculty, Kayseri, Turkey.*

**Objectives:** Henoch-Schonlein purpura (HSP) is the most common vasculitis in childhood. The long-term prognosis is variable and depends on renal involvement. The aim of this study was to evaluate the presentation, clinical and pathological manifestations and outcome of the HSP nephritis (HSPN) in children.

**Methods:** Clinical and laboratory data of 74 children with HSP nephritis treated from 1994 until 2008 at the Department of Pediatric Nephrology, Erciyes University Medical Faculty, Kayseri, Turkey, were retrospectively analyzed. The biopsy findings were graded according to the classification developed by the ISKDC.

**Results:** The median occuring time of renal findings was 20 days (between one day and 7.6 years) after HSP onset. Mean follow-up duration was 3.2 ±2.8 years. Renal biopsy was performed in 50 of the 74 patients with HSPN. The most common presenting clinical finding in patients was microscopic hematuria with non-nephrotic proteinuria (38%) which was followed by microscopic hematuria (18.3%). The biopsy findings according to the ISKDC were as follows: class I: 6 %; II: 44 %; III: 38 %; IV: 4%; V: 2 %; VI: 6%. Of 74 patients, 74.8% improved and 25.2% had microscopic hematuria and/or abnormal proteinuria. Chronic renal failure did not develop in any patient during study. There was no correlation between severity of clinical findings and histopathological features.

**Conclusions:** HSPN has favorable outcome when diagnosed early and treated properly. The severity of clinical findings at onset may not predict the severity of renal involvement.

## Abstract# 19

**Kaposi Sarcoma (KS) in a Boy with Steroid Dependant Nephrotic Syndrome (NS) Treated with Ciclosporine (CsA)** V. Baudouin,[1] T. Kwon,[1] S. Azib,[1] A.L. Sellier-Leclerc,[1] G. Deschênes,[1] C. Lebbe.[2] *[1]Nephrology, Robert Debré Hospital, Paris, France; [2]Dermatology, Saint Louis Hospital, Paris, France.*

**Objectives:** KS has been reported in organ transplantation. The cornerstone of treatment is to reduce immunosuppressive regimen and add mTOR inhibitors. Additional chemotherapy can be required, while rituximab (RTX) had been reported to worsen KS.

**Methods:** We report the case of a 9 year-old boy, with NS who developed KS while undergoing CsA and prednisone (P) treatment.

**Results:** NS began when the boy was 2 and because of numerous relapses CsA was associated to P when he was 3. At 9, cutaneous KS lesions occurred. HHV8 serology was positive while blood PCR was negative. CT scan revealed pulmonary nodules and abdominal adenopathies. CsA was switched to sirolimus and P was maintained at 0.5mg/kg eod. A relapse of NS occurred 1 month later and P was increased to 2mg/kg/d. KS lesions spread rapidly within few weeks and all treatment was withdrawn. During the following 6 months, KS lesions regressed but he experienced life threatening complications of NS. KS being in remission, 2 mg/kg/d of P allowed a NS remission. A RTX injection was then done and P stopped in 3 months. Remission of NS went on for 5 months, until blood $CD19^+$ $CD20^+$ B cells reappeared. The flare of NS was treated with P for 1 month and a second injection of RTX. Remission of both NS and KS maintained the following year without any treatment and despite reappearance of $CD19^+CD20^+$ B cells.

**Conclusions:** This is the first report of KS in a patient with NS. Its management is challenging because of both risks of NS and KS relapses. In our case, RTX infusion while KS in remission allowed a sustained remission of NS.

## Abstract# 20

**Membranous Nephropathy (MN) and Focal Segmental Glomerulosclerosis (FSGS) in Nephrotic Child after 2 Years of Bone Marrow Transplantation (BMT)** P. Pasztor,[1] C. Bereczki,[2] E. Kemény,[1] G. Kriván,[3] S. Turi.[1] *[1]Pediatrics, University of Szeged, Szeged, Hungary; [2]Pathology, University of Szeged, Szeged, Hungary; [3]BMT Unit, St.Laszlo Hospital, Budapest, Hungary.*

**Objectives:** Haematopoietic cell transplantation is a common treatment option for haematopoietic malignancies, but the late heavy proteinuria and nephrotic syndrome are rare complication. Most cases are attributed to graft-versus-host disease (GVHD) associated with membranous nephropathy. We report a case of membranous nephropathy (MN) and focal segmental glomerulosclerosis in nephrotic child without GVHD.

**Methods:** 4.5-year-old girl presented with nephrotic syndrome. Her high risk acute lymphoblastic leukemia was diagnosed at age 14 month and because of disease relapse 1 year later matched unrelated donor allogenic BMT was performed.

**Results:** At presentation 2 years after BMT she was therapy free and had no clinical manifestation of GVHD. She had oedema, heavy proteinuria ( 5.1 g/day), microscopic hematuria, hypoalbuninemia (26 g/L), normal renal function and normal complements levels. Renal biopsy showed MGN and FSGS. Cyclosporine and prednisolone were started and she went to complete remission after 3 weeks. Prednisolone was tailed after 4 weeks to 0,5mg/bwkg / days and stopped after 6 month with cyclosporine. In January 2010 she was in complete remission with microscopic hematuria ( 10-12 rbc/hpf).

**Conclusions:** We present a morphologically unique combination of membranous glomerulopathy and focal segmental glomerulosclerosis after BMT. Successful treatment options include glucocorticosteroids and CsA. Several studies suggested that the presence of FSGS in the renal biopsy predicted a worse outcome in patients with idiopathic MN.

## Abstract# 21

**The Concurrent Presentation of Idiopathic Thrombocytopenia and Focal Segmental Glomerulosclerosis** M. Buyukcelik,[1] B. Demircioglu Kilic,[1] A. Bay,[2] F. Ozaltin,[3] I. Sari,[4] A. Balat.[1] *[1]Pediatric Nephrology, Gaziantep University, Gaziantep, Turkey; [2]Pediatric Hemotology, Gaziantep University, Gaziantep, Turkey; [3]Pediatric Nephrology, Hacettepe University, Ankara, Turkey; [4]Pathology, Gaziantep University, Gaziantep, Turkey.*

**Objectives:** Focal segmental glomerulosclerosis (FSGS) is one of the most heterogeneous glomerular diseases in children and has been associated with many autoimmune diseases.

**Methods:** Herein, we report a patient with FSGS presenting concurrently with thrombocytopenia, subsequently diagnosed as idiopathic thrombocytopenic purpura (ITP). A 3 year-old boy was admitted to our hospital with thrombocytopenia (15,000/$mm^3$).

**Results:** Bone marrow aspiration showed ITP findings, and his platelet count came back to normal limits (288.000/$mm^3$) after intravenous immunoglobulin therapy. His urine analysis revealed proteinuria (++) and hematuria (+++). There were findings of focal segmental glomerulosclerosis in renal biopsy specimen. Conventional steroid treatment did not reduce the proteinuria, and the patient accepted as steroid-resistant nephrotic syndrome. DNA analysis showed homozygous c353C>T mutation in 2th egzone of NPHS2.

**Conclusions:** To our knowledge, this is the first case of familial FSGS associated with ITP.

## Abstract# 22

**Atypical Hemolytic Uremic Syndrome and Membranoproliferative Glomerulonephritis: A Case Report** M. Buyukcelik,[1] B. Demircioglu Kilic,[1] I. Sari,[2] A. Balat.[1] *[1]Pediatric Nephrology, Gaziantep University, Gaziantep, Turkey; [2]Pathology, Gaziantep University, Gaziantep, Turkey.*

**Objectives:** It has been suggested that atypical hemolytic uremic syndrome (aHUS) and membranoproliferative glomerulonephritis (MPGN), especially subtype II, represent a spectrum of related disorders. Thrombi are common in both, and develop in different microvascular beds including kidney and appear relevant for organ dysfunction.

**Methods:** Herein, we report a 10 year-old patient with aHUS and clinically rapidly progressive glomerulonephritis diagnosed as MPGN based on the pathological findings.

**Results:** In initial laboratory analysis; blood urea nitrogen was 120 mg/dL, while creatinine was 3,6 mg/dL, total protein 4,2 g/dL, albumin 2,2 g/dL, hemoglobin 5 g/dL, platelet 75.000/$mm^3$, and $C_3$ 0.53 mg/dL (N:90-120). Proteinuria (++++) and hematuria (+++) were detected in urine. In immunological tests, antinuclear antibody (ANA) was positive, anti-dsDNA, P-ANCA and C-ANCA were negative. Renal biopsy specimen revealed glomerular hypertrophy, mesangial proliferation, narrowing of capillary lumen, glomerular lobulation, and rare small necrosis in the glomerular tuft area. There were depositions of IgA, IgG and C3c in basement membrane, IgA and IgG in mesangium in immunofluorescence microscopy. These results were consistent with MPGN.

**Conclusions:** Based on the existence of acquired immune inhibitors and the shared thrombus formation in aHUS and MPGN as seen in our patient, we emphasize that aHUS and MPGN may share the same pathophysiological mechanisms responsible for the occurrence of disease.

## Abstract# 23

**Predictors of Outcome in Steroid Resistant Nephrotic Syndrome (SRNS)** R. Chanchlani, P. Hari, A. Dinda, R.N. Srivastava, A. Bagga. *Pediatric Nephrology, All India Institute of Medical Sciences, Delhi, India.*

**Objectives:** To analyze the clinical & pathological predictors of outcome in patients with SRNS.

**Methods:** Records of patients diagnosed with idiopathic SRNS secondary to minimal change disease (MCD) & focal segmental glomerulosclerosis (FSGS) between 1995 and 2008 were reviewed. Complete remission, steroid sensitive relapses & non nephrotic proteinuria were considered as favourable outcomes; chronic kidney disease and nephrotic range proteinuria were considered unfavourable.

**Results:** Of 467 patients (74% boys), 64% had initial steroid resistance; biopsy showed FSGS in 36%. The histopathology was similar in patients with initial and late resistance. The mean age at onset of nephrotic syndrome was 55±41 months and mean duration of follow up was 31±35 months. Therapy was with high dose IV corticosteroids & PO cyclophosphamide (37%); IV cyclophosphamide (25%) and calcineurin inhibitors (37%). 57% patients had a favourable outcome which was significantly higher for patients with MCD (65%) *vs.* FSGS (52.5%) (P 0.02). On univariate analysis, early age of onset (P 0.03), biopsy showing MCD (P 0.02), and therapy with cyclosporine (P 0.006) or tacrolimus (P 0.0001) were associated with a favourable outcome. On multiple logistic regression, biopsy showing MCD (OR 1.8, P 0.03; 95% CI 1.1, 3.0), and therapy with cyclosporine (OR 3.9, P=0.009; 1.4, 10.7) or tacrolimus (OR 3.4, P=0.001; 1.7, 6.8) were independent predictors of outcome.

**Conclusions:** Therapy was associated with favourable outcome in more than half the patients with SRNS. The presence of MCD and therapy with cyclosporine or tacrolimus were independent predictors of a favourable outcome.

## Abstract# 24

**A Retrospective Study of Outcome in Pediatric Lupus Nephritis after the Maintenance Therapy by Azathioprine or ECMPS** W. Chartapisak, S. Opastirakul. *Department of Pediatrics, Faculty of Medicine, Chiang Mai University, Chiang Mai, Thailand.*

**Objectives:** To report the outcome of lupus nephritis patients after the maintenance phase of oral immunosuppressive.

**Methods:** As this is a retrospective study, we recruited the children who had lupus nephritis and had been treated by IVCY followed by Azathioprine or ECMPS between 2006 to 2010. The data includes age, sex, class of lupus nephritis, time to relapse and the complications. For the children who had relapsed, the time to relapse after the maintenance therapy was analyzed by the log rank test.

**Results:** 35 children ( F:M = 29 : 6 ) were recruited. The histopathology included 2 children in class II (with crescent), 5 in class III, 22 in class IV and 4 in class V. The everage age is 10.8 yrs ( 5-14 yrs). 29 children received Azathioprine and 6 received ECMPS. There is none in ECMPS group who has relapsed before 2 years whereas 24 % ( 7 cases) in Azathioprine group has relapsed while undertaking the Azathioprine. The over all relapse rate in Azathioprine group is 34.5 % and 16 % in ECMPS which is not statistically significant (P= 0.08). The most frequent complications in Azathioprine group were neutropenia and hepatitis. There were fewer complications in the ECMPS group.

**Conclusions:** Both Azathioprine and ECMPS were the maintenance therapy for Lupus nephritis. The patients in Azathioprine group tends to relapse more frequently while the patients not having ceased the treatment whereas ECMPS group has less relapse and still in the remission while the patient was continued the treatment.The RCT needs to prove the efficacy of ECMPS for maintenance therapy in pediatric lupus nephritis.

## Abstract# 25

**Genetic Polymorphisms of CYP3A5 and ABCB1 Genes in Steroid Treatment of Children with Idiopathic Nephrotic Syndrome** Y.-H. Chiou,[1] L.-Y. Wang,[2] T.-H. Wang,[1,2] S.-P. Huang.[2] *[1]Division of Pediatric Nephrology, Departments of Pediatrics, Kaohsiung Veterans General Hospital, Kaohsiung, Taiwan; [2]Department of Medical Technology, Fooyin University, Kaohsiung, Taiwan.*

**Objectives:** This study is to analyze the correlation between outcome of steroid treatment and the genetic polymorphisms of steroid-metabolism related genes

including CYP3A5 (G6986A) and ABCB1 (C1236T, C3435T, and G2677T/A) in children with idiopathic nephrotic syndrome (INS).
**Methods: Methods:** Seventy four patients with INS were enrolled contain 58 steroid-sensitive (SS) and 16 steroid-resistant (SR) patients. The genetic polymorphisms were analyzed with SNP genotyping assay. The genotype and allele frequencies were calculated.
**Results: Results:** The genotype frequencies of the C1236T in ABCB1 gene showed significant difference between SS and SR group (p = 0.042). The allele frequency of C in C1236T is significantly higher in SS group (0.38) than that in SR group (0.19). The relative risk analysis showed positive correlation between SR and C1236T in ABCB1 (odds ratio: 2.65) and negative correlation between SR and G6986A in CYP3A5 (odds ratio: 0.38). There was no significant association between the steroid treatment and the other polymorphisms including C3435T and G2677T/A in ABCB1 gene.
**Conclusions: Conclusion:** The C1236T in ABCB1 and G6986A in CYP3A5 appear to be associated with the steroid treated children with INS. It will be provided to predict the clinical course and to improve the prognosis in children with INS.

## Abstract# 26

**The Impact of Mass School Screening: IgA Nephropathy in Korean Children** H.J. Choi,[1] Y.H. Jung,[1] H.K. Lee,[1] I.S. Ha,[1] H.I. Cheong,[1] Y. Choi,[2] H.G. Kang.[1] *[1]Pediatrics, Seoul National University Children's Hospital, Seoul, Republic of Korea; [2]Pediatrics, Inje University Haeundae Paik Hospital, Busan, Republic of Korea.*
**Objectives:** It is still debated whether a mass screening program can alter the prognosis of children with renal disease. We evaluated the prognosis of IgA nephropathy (IGAN) according to the clinical presentation to assess the effect of early detection of IGAN.
**Methods:** This retrospective study included children diagnosed as IGAN at our center from January 1981 to December 2009. The patients were categorized as group presented with asymptomatic urine abnormalities (AUA group) and symptomatic group according to the clinical sign and symptom on diagnosis. Correlations between clinical presentation, renal histology and stages of chronic kidney disease based on estimated glomerular filtration rate were examined.
**Results:** A total of 270 children with IGAN were reviewed (M:F=1.7:1). The age at the time of the biopsy was 10.7 ± 3.5 years and follow up duration was 5.5 ± 5.0 years. The proportion of AUA group increased after 1998 (32%) when the nationwide school screening of urinalysis started (before 1998, 9%) and the overall proportion was 25%. In the histologic grading, patients with grade 4 or 5 were 1.5% of AUA group and 6.9% of symptomatic group. On the last examination, 1.5% of AUA group and 4.9% of symptomatic group belonged to CKD stage 3 or higher.
**Conclusions:** The proportion of AUA group increased after the nationwide screening program. The clinical status on final examination was not significantly different between the groups. To define the effect of urinalysis screening more long-term follow up is needed.

## Abstract# 27

**Fish Oil in IgA Nephropathy: A Systematic Review and Meta-Analysis** H.-H. Chou, Y.-Y. Chiou. *Pediatrics, Chiayi Christian Hospital, Chiayi, Taiwan; Pediatrics, National Cheng Kong University Hospital, Tainan, Taiwan.*
**Objectives:** Fish oil rich in omega-3 polyunsaturated fatty acids has been tested in patients with IgAN with conflicting results. We therefore conduct a review summarizing currently available evidence pertaining to the effect of fish oil for the treatment of IgAN.
**Methods:** We searched Pubmed, Medline, CINAHL, and the Cochrane Central Register of Controlled Trials from 1966 to 2010, for randomized controlled trials on the efficacy of fish oils for patients with IgAN. The quality of studies to be included was assessed independently by two reviewers. The reviewers extracted data pertaining to deterioration in renal function (defined by a 50% increase in creatinine clearance level from baseline value). Risk ratios with 95% confidence intervals were pooled and random effect model was used for heterogeneity.
**Results:** We identified 6 clinical trials providing data for total 304 patients with mean follow up period about 2.1 years. No data has been published on the use of fish oil for the treatment of pediatric patients with IgAN. Overall, fish oil is not beneficial because the cumulative RR and its 95% CI cross the null hypothesis (4 trials, 234 patients; RR, 0.57; 95% CI, 0.17 to 1.97; heterogeneity chi-square = 11.56; P = 0.009). A sensitivity analysis was carried out and no statistically significant all-studies effect was obtained by excluding any single study from the analysis.
**Conclusions:** The benefit of fish oil can not be established based on currently available evidence either in adults or pediatric population. More properly designed and reported trials are necessary to reach a definitive assessment of this matter.

## Abstract# 28

**Clinicopathologic Characteristics and Prognosis of Primary IgM Nephropathy in Children** M. Chu, L. Cao. *Nephrology, Capital Institute of Pediatrics, Beijing, China; Nephrology, Capital Institute of Pediatrics, Beijing, China.*
**Objectives:** Immunoglobulin M nephropathy (IgMN) is a special glomerular disease and it is thought to be a variant of other renal disease's pathology. IgM nephropathy is an idiopathic glomerulonephritis characterized by mesangial deposits of IgM. IgM nephropathy presenting with proteinuria, especially nephrotic syndrome, frequently is steroid dependent or steroid resistant. Because no clinical investigation was done for pediatric patients with IgM nephropathy,and we also need a treatment proposal. Our study explore the relations between clinical manifestations,pathological features,and prognosis of IgM nephropathy in children.
**Methods:** The clinical manifestations, pathological features, therapeutic methods and the long term followup information of 14 children with IgM nephropathy from October,2004 to March,2009 in Capital Institute of Pediatrics were retrospective analyzed.
**Results:** 50% of our patients's were nephrotic syndrome. The main features of renal pathologies were mesangial proliferative. Immunofluorescence indicated IgM deposits in 8 cases, IgM+IgG deposits in 5 cases. 81.8% patients were effective to our therapy. No one reach end-stage renal disease in 5 years followup.
**Conclusions:** The clinical manifestations of IgM nephropathy in children are correlated with pathological changes.Early and regular therapy was well effective in our study. Massive proteinuria indicates poor prognosis.

## Abstract# 29

**Response Therapy in Children with IgM Nephropaty** M.C. d´Oliveira, T.F. Santos, J.R.P. Santos, F.V. Leão, M.A.P. Cançado, J.T.A. Carvalhaes. *Division of Pediatric Nephrology, Federal University of São Paulo, São Paulo, Sao Paulo, Brazil.*
**Objectives:** Evaluate therapeutic response and clinical outcome of children with IgM nephropathy. IgM nephropathy is a primary diffuse mesangioproliferative glomerulonephritis, with predominant IgM deposition in the mesangium, characterized by massive proteinuria with normal creatinine and unsatisfactory response to steroids and immunosuppressants. Some authors consider it a distinct entity with tendency to corticoresistency and evolution to progressive renal failure.
**Methods:** Fourteen (14) patients treated and followed with IgM nephropathy, were evaluated according to age of onset, initial presentation, treatment, renal biopsy, response to therapy and progression to renal failure.
**Results:** Edema and proteinuria were found as the initial presentation of 11 patients (78.57%). Of these, 2 evolved with hypertension. Three (3) had only hematuria. Renal biopsies showed: minimal lesions in 8 (57.14%), mesangial proliferation in 4 (28.57%), focal segmental glomerulosclerosis in 2 (14.28%). None of the patients progressed to loss of renal function. Treatment: 3 (21.42%) used corticosteroid (CS), 5 (35.71%) received CS and cyclophosphamide, 3 (21.42%) used CS, cyclophosphamide and cyclosporine (CSA), 1 (7, 14%) had CS, cyclophosphamide, CSA and mycophenolate mofetil; 1 (7.14%) remained untreated for display only microscopic hematuria, and 1 (7.14%) received CS and azathioprine for the treatment of Henoch-Schoenlein.
**Conclusions:** Of 14 patients, 9 received CS associated with immunosuppressive drugs with unsatisfatory response. There was no progression to chronic renal failure in an average of 9 years of follow-up.

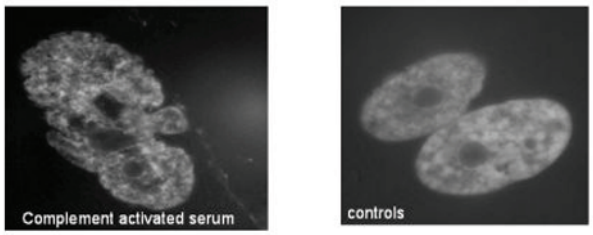
## Abstract# 30

**Hypothyroidism in Children with Steroid Resistant Nephrotic Syndrome** A. Dagan, R. Kleper, I. Krause, D. Blomental, M. Davidovits. *Institute of Pediatric Nephrology, Schneider Children's Medical Center, Petach Tiqva, Israel.*
**Objectives:** Patient with nephrotic syndrome, are usually considered to be euthyroid. However there is scanty information on the thyroid function in children with steroid resistant nephrotic syndrome especially on patients who suffer from long standing proteinuria.
**Methods:** In this case series we present five children with steroid resistant nephrotic syndrome duo to either focal segmental glomerulosclerosis ( FSGS) or diffuse mesangial proliferation (DMP).
**Results:** All the patients were resistant to therapy, developed chronic renal failure and end stage renal disease (ESRD). The patients were found to have depressed free $T_4$ (FT4) and elevated thyrotropin (TSH) during the course of their disease without relation to the duration of nephrotic syndrome or level of renal function. At diagnosis of hypothyroidism must of the patients have non-specific complaints that could be attributed either to their neprotic syndrome or hypothyroidism. There was no evidence for autoimmune thyroiditis in any of the patients. All patients were treated with thyroxine, but their thyroid hormone profile usually normalized only when they reached ESRD and started hemodialysis. The patients could be weaned from thyroxine therapy while on dialysis or after

kidney transplantation. Three patients were already underwent kidney transplant. Despite recurrence of FSGS in the transplanted kidney none developed recurrent hypothyroidism.
**Conclusions:** In conclusion non autoimmune hypothyroidism should be actively sought for and treated in patient with steroid resistant nephrotic syndrome. This state is usually transient and resolves when the patients reach ESRD and require dialysis.

## Abstract# 31

**Shigatoxin and Complement Activation Are Toxic to Human Podocytes in Different Ways** E. Binder,[1] A. Dettmar,[1] E. Feifel,[2] G. Gstraunthaler,[2] H. Karch,[3] L. Zimmerhackl.[1] *[1]Department für Kinder- und Jugendheilkunde, Medizinische Universität, Innsbruck, Austria; [2]Department für Physiologie, Medizinische Universität, Innsbruck, Austria; [3]Institut für Hygiene, Westfälische Wilhelms- Universität, Münster, Germany.*
**Objectives:** Resulting in similar clinical presentation different pathomechanisms are responsible for typical and atypical hemolytic uremic syndrome (tHUS, aHUS). Shigatoxin (Stx), produced by EHEC 0157:H7, is responsible for renal damage in tHUS and activation of complement for thrombotic microangiopathy and renal failure in aHUS.
**Methods:** Cultured human podocytes were incubated with Stx or complement activated human serum. Cells were stained with Hoechst and lysates were tested using assays for caspase 1,4 and 3 activity, and LDH- release after incubation.
**Results:** Podocytes changed their shape after both treatments. Hoechst staining showed blurring of borders of the nuclei compared to controls (figure), but no change in caspase activity after incubation with complement activated serum.




Stx application results in an increase of caspase 3 activity.
**Conclusions:** Podocytes, as an important part of the glomerular filter, are propably damaged in HUS. In vitro they reacted to both, Stx and complement activation, but in different ways. Further studies will be done to determine this difference and combine the influence of complement and shigatoxin.

## Abstract# 32

**Nephrotic Syndrome in 10 to 15 Year-Old Children at the Children's Hospital No 2, HCMC, VN** T.H. Huynh,[1] D.M. Duong,[2] T.H. Vu.[3] *[1]Nephrology, Children's Hospital No2, HCM, Viet Nam; [2]Pediatrics, Franco-Vietnamese, HCM, Viet Nam; [3]Pediatrics, University of Medicine-Pharmacy, HCM, Viet Nam.*
**Objectives:** Nephrotic Syndrome (NS) is a common renal disease reported at the Children's Hospital No2, HCMC, VN. The aim is to describe manifestations of NS children at the age of 10-15.
**Methods:** The study population consisted of 32 NS children (age of 10-15). Clinical and laboratory findings, complications were discribed. The study was one-year prospective cohort. Epi.Info 2002 was used for statistics.
**Results:** From 2007 to 2008, there were 32 NS children (10-15 years old) recruited to the study. The male to female ratio was 1.6:1. The family history with NS was 6.3%. The percentages of edema, oliguria, gross hematuria, and hypertension were 50; 62.5; 6.3 and 21.9, respectively. Laboratory abnormalities included severe hypoalbuminemia (12.5%), decreased GFR (21.9%), hypocalcemia (68.8%), high HDL (46.9%), microscopic hematuria (50%). The number of children positive for HBsAg was 15.6%, compared with 12.5% of CMV, 9.3% of EBV. Low serum C3 and/C4 level amounted to 15.6%. The number of positive Ds DNA in the study was 6.3%. ANA and LE cell were positive for 6.3% and 9.4%, respectively. Renal biopsy done in 19 out of 32 patients presented with 6.3% of SLE, 3.1% of minimal change, 42.9% of mesangial proliferation, 3.2% of FSGS. Complications recorded in the study comprised cellulitis (3.1%), peritonitis (3.1%), acute kidney injury (6.3%), hypovolemic shock (3.1%).
**Conclusions:** The children with NS in our study presented with outstanding features of gross and microscopic hematuria, hypertension, decreased GFR, low C3/C4 levels. The percentages of positive HBV, CMV, EBV were high. Minimal change was not predominant.

## Abstract# 33

**Predictors of Outcome in IgA Nephropathy** S.F. Edstrom Halling, U.B. Berg. *Dep of Pediatrics, Karolinska University Hospital, Stockholm, Sweden.*
**Objectives:** Clinical predictors of a poor outcome in pediatric IgA nephropathy.
**Methods:** 99 children with a mean age of 9.7 ± 3.9 years at onset followed for > 5 - 36 years, mean follow-up (FU) time 13.4 ± 8.2 years, in those not having developed terminal renal failure (TRF). Patients were investigated with clearances of inulin or iohexol, mean arterial blood pressure (MAP) and Ualbumin/Ucreatinine (Ualb/c) and UIgG/Ucreatinine (UIgG/c) ratios at time of biopsy and during FU. All patients had a renal biopsy performed at median 1.6 years from onset.
**Results:** GFR at time of biopsy (GFR bio) was 100 ± 31 ml/min per 1.73 $m^2$, significantly ($p<0.0001$) lower than that of 59 healthy controls. 17 patients had CKD stage 4-5 at the last visit.15 patients developed TRF after median 6.1 (0.08-14.8) years FU. Patients with CKD 4-5 at the last visit had significantly lower GFR, higher MAP, higher Ualb/c and UIgG/c at biopsy, compared to those with CKD stage 1-3. GFR at the last visit (GFR last) correlated directly to GFR at biopsy, and at 1, 3 and 5 years FU and also correlated inversely to MAP and to Log Ualb/c at biopsy.
There was a significant difference in outcome (CKD 4-5 vs 1-3) between patients who were hypertensive vs normotensive at biopsy ($p= 0.005$) and between those with nephrotic proteinuria vs non- nephrotic or no proteinuria at biopsy ( $p=0.00002$). Logistic regression (univariate analysis) showed that GFR bio ($p=0.0002$) and Log Ualb/c ($p=0.0004$) at biopsy discriminated between CDK 4-5 and CDK 1-3 at the last visit. The two variables included in the multivariate model showed a high agreement (91 %) between observed and expected values.
**Conclusions:** GFR and Ualb/c ratio at time of biopsy were predictors of poor outcome.

## Abstract# 34

**Clinical Features and Outcome of Biopsied Henoch-Schönlein Purpura Nephritis (2000-2009)** T.M. Eison,[1] M.C. Hastings,[1] K.K. Lau,[2] N.M. Delos Santos,[1] B.H. Ault,[1] D.P. Jones,[1] R.J. Wyatt.[1] *[1]University of Tennessee Health Sciences Center, Memphis, TN, United States; [2]McMaster University, Hamilton, ON, Canada.*
**Objectives:** To describe the clinical and pathologic features and outcome for patients (pts) < 18 years old who were biopsied for Henoch-Schoenlein purpura nephritis at the Le Bonheur Children's Medical in Memphis from 1/1/2000 to 12/31/2009.
**Methods:** Descriptive, retrospective review of medical records.
**Results:** We identified 20 pts (11 male; 9 female) (14 causasian; 5 African-American; 1 caucasian/asian mixed) with mean age at onset of 7.79 years. Indication for biopsy was usually moderate to severe proteinuria with or without nephrotic syndrome (serum albumin <2.5). Eleven had nephrotic syndrome, while 8 of the 9 remaining had urine protein:creatinine ratio (Ur pr:cr) >2.0 at initial presentation. Renal biopsy showed >25% glomeruli with crescents in one pt and 1-25% crescents in 6. Treatment was with IV methylprednisolone (MP) followed by oral prednisone in 4 and oral prednisone in 13. Two pts also received mycophenolate mofetil. Eighteen pts were treated with lisinopril which was the only therapy for 2. Length of treatment varied widely among pts. Mean time from onset to last followup was 2.26 years. All patients had normal renal function at last follow-up (estimated GFR by Schwartz formula >90). Of 9 pts followed >2 years, 2 had Ur pr:cr >1; 1 had >0.5<1; 1 had >0.2<0.5, and 5 had ratios <0.2.
**Conclusions:** The short to medium term outcome of recent patients from our cohort appears better than that for past studies from major tertiary centers.

## Abstract# 35

**Nephrotic Syndrome in Haitian Children: About the Prevalence and the Evolution** J. Exantus, R. Dall'Amico. *Hopital St Damien (Nos Petits Freres et Soeurs), Port-au-Prince, Haiti; Pediatria, Azienda Ospedaliera S Maria degli Angeli, Pordenone, Italy.*
**Objectives:** Haiti is a Caribbean country with a 95% black population and strong genetic similarities with the African. Nephrotic Syndrome in children is one of the most common kidney disorders in Africa with different prevalence and response to therapy according to geographical area. There is no data about nephrotic syndrome in Haitian children. We retrospectively study his profile in a small cohort of children.
**Methods:** We reviewed the records of patients hospitalized with a diagnosis of nephrotic syndrome at the Hospital St. Damien in the period 1990-2008.
**Results:** We identified 83 children with a mean age of 6.4 years (3 months-14 years), 37 females and 46 males.
Four had been treated previously for pulmonary tuberculosis. Two/38 were HIV positive, 1/17 HBsAg positive. Hemoglobin electrophoresis made it possible to identify: 5 Hb AS, Hb SC 1, 1 Hb AC, Hb 1 CC.
Nephrotic syndrome was steroid responsive in 67/83 cases and steroid resistant in 16/83 (19%). Five patients died: 3 with a nephrotic syndrome occurred before 3 months of age, one with HIV, one for unspecified reasons. Thirteen patients were lost in follow-up.

None of the children underwent biopsy in the country. Three patients received a biopsy abroad.
All patients have been treated with cortisone orally. Two patients with frequent recurrences were treated with levamisole.
**Conclusions:** The data shows that the prevalence of steroid resistant nephrotic syndrome is lower if compared to other series in black population.
We recommend another study for a larger population of nephrotic patients, to appreciate their evolution and the influence of nephrotic syndrome in chronic renal failure.

## Abstract# 36

**A Massive Pulmonary Embolism as a First Symptom of Nephrotic Syndrome in 18-Years Old Female Patient** V. Stará,[1] F. Fencl,[1] K. Rücklová,[1] K. Bláhová,[1] J. Zieg,[1] J. Charvát.[2] *[1]Dpt. of Paediatrics, 2nd Faculty of Medicine Charles University nad Motol Univ. Hospital, Prague, Czech Republic; [2]Dpt. of Internal Medicine, 2nd Faculty of Medicine Charles University nad Motol Univ. Hospital, Prague, Czech Republic.*
**Objectives:** The incidence of thromboembolic complications (TEC) in patients with nephrotic syndrome (NS) is about 1.8-5%, but may be underestimated. Most patients with TEC are asymptomatic.
**Methods:** We report a case of an 18-years old female patient on hormonal contraception with irrelevant family and personal history presenting a clinical picture of dyspnea, chest pain, collapse and discrete oedemas of eyelids, arms and legs. On echocardiography were indirect signs of pulmonary hypertension, dilation and dysfunction of right ventricle, on ECG was right bundle branch block. At this point, CT-angiography was performed and confirmed the bilateral massive pulmonary embolism (PE). Anticoagulation therapy with heparin and subsequently low-molecular heparin was started. Ultrasound examination focused on detection of the source of embolism was negative, as well the clinical symptomps of previous deep vein thrombosis. Further examinations proved nephrotic proteinuria (6 g/m2/day), biochemical parameters corresponded to NS.
**Results:** Renal biopsy showed a retraction of the epithelial foot processes characteristic for minimal change disease and confirmed the diagnose of NS. Therapy with prednisone was started according to standard protocol with good effect.
**Conclusions:** The NS should be considered in differential diagnoses in patients with PE, all these patients should undergo urine testing for proteinuria. As we demonstrate, massive PE can be rarely the first symptom of NS.

## Abstract# 37

**Clinicopathological Characteristics and Outcome in Children with Nephrotic Syndrome** d.P. Fonseca Leal,[1,2] A. Rangel Castilla.[1,2] *[1]Hospital Central Ignacio Morones Prieto, San Luis Potosi, Mexico; [2]San Luis Potosi University, San Luis Potosi, Mexico.*
**Objectives:** To identify the clinical differences and outcome between patients on whom kidney biopsy was or wasn't performed.
**Methods:** Retrospective study involving 70 consecutive children with nephrotic syndrome, they were classified into two groups: kidney biopsy performed (G1) and without kidney biopsy performed (G2). Statistical analysis was done with the software R version 2.10.1.
**Results:** Kidney biopsy was performed in 47%. Significant differences between groups were: mean age at onset, G1 6.04±4.09 and G2 4.85±3.42 years, OR 1.29 (CI 95% 1.01 – 1.62); hematuria at presentation 63.6% in G1 and 43.2% in G2, OR 5.29 (CI 95% 1.09 – 34.7); mean follow-up 35.8±32.64 in G1 and 13.9±23.51 in G2, OR 1.05 (CI 95% 1.01 – 1.62); end stage renal disease 9% in G1 and 2.7% in G2, OR 24.3 (CI 95% 106 – 1238.5).
Second line drugs prescribed were: Cyclosporine in 54.5% G1 and 21.6% in G2; Azathioprine in 24.2% in G1 and 13.5% in G2; Cyclophosphamide in 12.1% in G1 and 19.8% in G2.
Histopathological profile: focal segmental glomerulosclerosis in 39.4%, minimal change disease in 30.3%, IgM nephropathy in 12.1%, There was an increased probability of FSGS at older ages, MCD had a 0.31 probability for remission and FSGS 0.32 probability for no remission.
**Conclusions:** It would be useful to have at presentation, clinical predictors that could guide us to decide in which patients it would be necessary to perform a kidney biopsy, but we found only few significant differences in clinical presentation between groups. The outcome: remission and renal failure were statistically different.

## Abstract# 38

**Chronic Lesion Index – Prognostic Marker in Pediatric Nephrotic Syndrome** P.M. Fortes,[1] M.A.R. Moreira,[2] A.V. Naghettini,[3] C.M. Salgado.[4] *[1]Pediatria e Puericultura, Faculdade de Medicina - Universidade Federal de Goias, Goiânia, Goias, Brazil; [2]Patologia e Imaginologia, Faculdade de Medicina - Universidade Federal de Goias, Goiânia, Goias, Brazil; [3]Pediatria e Puericultura, Faculdade de Medicina - Universidade Federal de Goias, Goiânia, Goias, Brazil; [4]Pediatria e Puericultura, Faculdade de Medicina - Universidade Federal, Goiânia, Goias, Brazil.*
**Objectives:** The use of the chronic lesion index (CLI) assessed as a marker of worst prognosis, relating this to steroid therapy response, evolution to end stage renal disease (ESRD) and arterial hypertension (AH).
**Methods:** Responses to steroid therapy were steroid-resistant, steroid-dependent or frequent relapser. ESRD was defined as estimated creatinine clearance < 15 ml/minute and AH based on the 4th TASK FORCE, 2004. The CLI (Hill et al., 2000) was constituted by: tubular atrophy, interstitial fibrosis, glomerulosclerosis and glomerular scars (quantitatively graded 0 – 3, with maximum sum of 12). Slides were reviewed by light microscopy and immunofluorescence for CLI application.
**Results:** Glomeruli number per slides totaled 20,34 ± 16.31. Mminimal change disease (MCD) frequency was 9/46 (19.5%) and focal segmental glomerulosclerosis (FSGS) was 37/46 (80.5%). CLI presented a higher sum for FSGS (0 to 7), in patients unresponsive to steroid therapy (p=0.001), where ESRD developed regardless of glomerular lesion (p=0.004), in FSGS bearers that developed ESRD (p=0.005) presenting correlation with higher systolic AP percentile levels at biopsy time (r=0.335 p=0.024).
**Conclusions:** CLI application presented relation with worst prognosis indices such as: non response to steroid therapy, higher AP levels and development of ESRD.

## Abstract# 39

**Comparison of Azathioprine vs. Mofetil Mycofenolate for Henoch-Schönlein Nephritis Treatment** Y. Fuentes, S. Valverde, L. Velasquez-Jones, B. Romero, A.M. Hernández, I.E. Del Moral, R. Maldonado, E. Faugier, G. Ramón, M. Medeiros. *Hospital Infantil de México Federico Gómez, Mexico, DF, Mexico.*
**Objectives:** The aim of the study was to compare if Mofetil mycophenolate (MMF) is better than Azathioprine (AZA) for the treatment of Henoch Schonlein Purpura nephritis.
**Methods:** Prospective randomized trial in children with biopsy proven Henoch-Schönlein Purpura nephritis, two treatment groups: Group I: prednisone + AZA, Group II: Prednisone + MMF. Monthly visits for serum creatinine, proteinuria, liver function test, urine for MCP1by ELISA. Second renal biopsy after 12 months of treatment.
**Results:** 17 children have completed one year of follow up. Two patients (25%) persisted with proteinuria in the AZA group; all patients presented remission of proteinuria in the MMF group. Demographic and outcome data is depicted in table 1.

Demographic data and outcome

| | AZA (n=8) | MMF (n=9) | Total (n=17) |
|---|---|---|---|
| Age | 6.4 ± 1.5 | 7.4 ± 2 | 7.4 ± 2 |
| ISKDC Histologic lesion | | | |
| Class I | | 1 (11%) | 1(6%) |
| Class II | 4(50%) | 2 (22%) | 6(35%) |
| Class III | 4(50%) | 6 (67%) | 10(59%) |
| Baseline Creatinine clearance (ml/min/1.73 m2BSA | 154 ± 34 | 148 ± 23 | 151 ± 28 |
| Basal proteinuria (median, range) | 98 (10, 274) | 28 (14, 333) | 50 (10, 333) |
| Regression of histological lesion at 1 year (regression/no change) | 5/3 | 7/2 | 12/5 |
| Basal urine MCP1 pg/mL | 316 (26, 2728) | 65 (14, 665) | 188 (14, 2728) |
| Urine MCP1 pg/mL at 12 months | 128 (0,2357) | 17 (0, 143) | 23.5 (160, 2357) |

**Conclusions:** There are more remissions of proteinuria in MMF group, the difference is not statistically significant. More patients and prolonged follow up is needed.
**DISCLOSURE:** *Medeiros, M.*: Consultant, Novartis, Mexico.

## Abstract# 40

**Incidence of FSGS in Children over the Last 15 Years in the Czech Republic** A. Kolsky, E. Jancova, J. Dusek, M. Hladik, S. Skalova, K. Vondrak, P. Geier, J. Starha, I. Rychlik, V. Tesar. *Department of Pediatrics, 3rd Medical Faculty, Charles University, Prague, Czech Republic; Czech Registry of Renal Biopsies, Prague, Czech Republic; Division of Nephrology, Children's Hospital of Eastern Ontario, Ottawa, ON, Canada.*
**Objectives:** The aim of our study was to analyze the incidence of FSGS based on data collected over the last 15 years from a nationwide (Czech Republic) prospective registry of renal biopsies.
**Methods:** Data were collected from 1994 to 2008 from all 10 pediatric nephrology centres in the country reporting kidney biopsy results of children 0-18

years of age. Annual demographic population data were obtained from the Czech Statistical Office. The 15-year study period was retrospectively divided into an early period 1994-2000 (A) and a more recent period 2001-2008 (B).
**Results:** Out of a total of 1903 biopsies performed in the study period 1994-2008, 106 biopsies showed FSGS (5.6%). Fifty five children were diagnosed in period A and 51 in period B. Over the study period, the child population has decreased from 2.7 millions in 1994 to 2.0 millions in 2008. The mean±SD annual incidence (per 100 000 child population) of FSGS was 0.32±0.07 in period A and 0.31±0.19 in period B (p=0.82) and did not change over time. The cumulative 7-year period incidence of FSGS was also not different between period A and B (p=0.67; relative risk = 0.95, 95% CI = 0.79 to 1.15).
**Conclusions:** The annual incidence of 0.3/100000 is similar to other reports for Caucasian population in North America. However, the incidence of FSGS has not significantly changed over the last 15 years.

## Abstract# 41

**Tubular Dysfunction in Idiopathic Nephrotic Syndrome** E. Girardin,[1] J. Ngoué Epée,[1] H. Chehade,[2] P. Parvex.[1] *[1]Ped. Nephr., HUG, Geneva, Switzerland; [2]Ped. Nephr., CHUV, Lausanne, Switzerland.*
**Objectives:** To evaluate the degree of tubular involvement in INS at various stage of the disease.
**Methods:** 19 patients with INS were studied. 13 were steroid responders (group 1). 5 of them had biopsy which showed MCD. 6 patients were non responder to steroid or were steroid dependant with frequent relapses (group 2). Biopsies showed 3 FSGS and 3 MCD. They were treated with prednisone, ciclosporin and/or mycofenolate mofetil. Protein, microalbumin (ALB), alpha-microglobulin (AMG), N-acetyl-beta-D-glucosaminidase (NAG) and creatinine (cr) were measured in each urine sample. Patients were considered in remission if prot/cr ratio (g/mol) was < 20 (group 1a and 2a), and in relapse if the ratio was > 200 (group 1c and 2c). Some patients in group 1 had non nephrotic proteinuria (group 1b). Tubular dysfunction was defined by NAG/cr ratio (mg/mmol) > 0.86 or by AMG/cr ratio (mg/mmol) > 1.58.
**Results:**

| | Prot/cr | ALB/cr | NAG/cr | AMG/cr |
|---|---|---|---|---|
| Group 1a | 10.3 ± 4.1 | 1.1 ± 1.0 | 0.19 ± 0.12 | 1.40 ± 0.97 |
| Group 1b | 60.4 ± 63.4 | 42.8 ± 66.7 | 0.39 ± 0.21 | 1.20 ± 0.56 |
| Group 1c | 713.3 ± 276.8 | 799.8 ± 534.9 | 2.25 ± 1.86* | 4.25 ± 2.09* |
| Group 2a | 11.3 ± 6.1 | 4.7 ± 5.7 | 0.26 ± 0.19 | 1.18 ± 0.60 |
| Group 2c | 914.9 ± 718.6 | 682.9 ± 589.3 | 3.00 ± 2.72* | 5.47 ± 4.30* |

Results are mean ± SD, p < 0.001 compared to group 1a and 2a
No difference was observed between group 1 and group 2 neither in remission nor in relapse.
**Conclusions:** These data indicate that tubular dysfunction occurs in INS but only in patients in relapse. In this population, tubular dysfunction was independent of the severity of the nephrotic syndrome, the treatment protocol and the histopathology.

## Abstract# 42

**Pharmacokinetic Tools for, and Clinical Findings in Favour of, the Therapeutic Drug Monitoring of Mycophenolate Mofetil in Children with Idiopathic Nephrotic Syndrome** V. Guigonis,[1] F. Saint-Marcoux,[2] B. Ranchin,[3] J.B. Palcoux,[4] F. Bouissou,[5] J. Harambat,[6] F. Broux,[7] E. Berard,[8] P. Marquet.[2] *[1]Pediatrics Nephrology, CHU, Limoges, France; [2]INSERM U850, CHU, Limoges, France; [3]CHU, Lyon, France; [4]CHU, Clermont-Ferrand, France; [5]CHU, Toulouse, France; [6]CHU, Bordeaux, France; [7]CHU, Rouen, France; [8]CHU, Nice, France.*
**Objectives:** The present study aimed at: (i) looking for potential relationships between mycophenolic acid (MPA) and clinical status; in children with INS; (ii) modelling MPA PK profiles and developing a Bayesian estimator (BE) for individual MPA AUC prediction.
**Methods:** Informations and full PK profiles were collected in paediatric inpatients already treated with MMF (with no CNI). A one-compartment open model where the absorption is described by a double gamma law was used to model the data. The performance of the BE was evaluated by the bias and precision of predicted AUCs and by the proportion of predicted AUCs with absolute error > 20%.
**Results:** Sixty PK profiles of MPA were studied. The median AUC was significantly higher in remission than in relapse. (62 mg.h/L [32 to 139]) versus 24 mg.h/L [7 to 41]; p< 0.001). Forty-five of these PK profiles were used to develop a PK model and a BE, and 15 for their validation. The PK model fitted accurately the PK profiles of MPA and a BE which allowed the estimation of AUC on the basis of a 20min-60min-180min LSS was developed. In the independent group of 15 patients, its MRE versus reference was -0,036 0,145 (-0.205 to 0.189).
**Conclusions:** A BE for the estimation of MPA AUC based on a limited sampling protocol has been developed. Upcoming prospective trials in INS should test the potential impact of the therapeutic drug monitoring of MMF.

## Abstract# 43

**Randomized, Multicenter Study on Tacrolimus & Alternate Day (AD) Prednisolone *vs.* Intravenous (IV) Cyclophosphamide & ad Prednisolone for Steroid Resistant Nephrotic Syndrome (SRNS)** A. Gulati,[1] P. Hari,[1] M. Kanitkar,[2] J. Sharma,[3] M. Mantan,[4] A. Bagga.[1] *[1]All India Institute of Medical Sciences, Delhi, India; [2]Armed Forces Medical College, Pune, India; [3]Bharti Vidyapeeth, Pune, India; [4]MAMC, Delhi, India.*
**Objectives:** To compare the efficacy & safety of treatment with tacrolimus & ad prednisolone *vs.* IV cyclophosphamide & ad prednisolone for inducing partial or complete remission in patients with SRNS.
**Methods:** Following consent, 120 newly diagnosed patients with SRNS (lack of remission to prednisolone @2 mg/kg/d for 4 wk), 1-16 yr old, are randomized to therapy with IV cyclophosphamide (500 mg/m$^2$/dose q mo x 6) or tacrolimus (trough levels 5-8 ng/ml) & prednisolone (1.5 mg/kg/ad, tapered 0.5 mg/kg/ad for 1-yr). Therapy is stratified for initial or late resistance; former are sequenced for mutations in *NPHS1, NPHS2* & *WT1* genes. Patients with eGFR <60 ml/min/1.73m$^2$ or non-steroid immunosuppression excluded.Primary outcome is to compare the rate of complete/partial remission at 6-mo.The time to first relapse, frequency of relapses, side effects and GFR shall be compared at 1-yr.
**Results:** Baseline data on 105 patients (67 boys) with SRNS.
Table 1

| | IV cyclophosphamide (n=54) | Tacrolimus (n=51) |
|---|---|---|
| Age at onset, yr | 5±3 | 4±2 |
| Initial /Late resistance | 34/20 | 33/18 |
| MCD | 31 | 32 |
| Mesangial cellularity | 3 | 2 |
| FSGS | 20 | 17 |
| Se creatinine, mg/dl | 0.5±0.1 | 0.5±0.1 |
| eGFR ,ml/min/1.73m2 | 86±14 | 89±10 |
| Se albumin, g/dl | 2.4±0.6 | 2.14±0.6 |
| Se cholesterol, mg/dl | 348±137 | 354±145 |
| Urine protein/creat,mg/mg | 4.2±1 | 3.7±1 |

**Conclusions:** Results of this study shall have implications for guiding initial treatment of children with SRNS, especially where costs are a major factor determining therapy.

## Abstract# 44

**Puromycin Aminonucleoside Modulates p130Cas Via Oxidative Stress** T.-S. Ha, E.-M. Ahn, W.-G. Kim, J.-S. Lee. *Chungbuk National University, Cheongju, Korea.*
**Objectives:** Puromycin aminonucleoside (PAN) specifically injured podocytes, leading to foot process effacement, actin cytoskeleton disorganization, and abnormal distribution of slit diaphragm proteins, resulting in a well-described model of podocyte injury. p130Cas is a docking protein interacting with FAK and c-Crk as well as its localization to focal adhesions indicate a role for p130Cas in signaling pathways from cell adhesion sites to the cytoskeleton. p130Cas localized subcellulary in podocytes.
**Methods:** To investigate the mechanism of proteinuria, we observed the changes of p130Cas in cultured podocytes treated with PAN and analyzed by confocal imaging, Western blotting, and PCR.
**Results:** In immunofluorescence study p130Cas showed a diffuse cytoplasmic distribution with accumulation at distinct sites visible as short stripes and colocalized with P-cadherin. The fluorescences of p130Cas protein were internalized and became granularly by PAN in a dose-dependent manner which had been augmented by EGCG, an anti-oxidant. PAN also increased the amount of p130Cas protein in a dose-dependent manner which had been also reversed by EGCG.
**Conclusions:** We suggest that PAN induced the changes of podocytes p130Cas partly through oxidative stress resulting in podocyte dysfunction.

## Abstract# 45

**Lipocalin-2 as a Marker of Severe Lupus Nephritis in Children** A. Hammad,[1] Y. Mosaad,[2] S. El-Hanbly,[3] H. Youssef,[4] A. El-Refaey,[1] F. El-Husseini,[5] A. Bakr.[1] *[1]Nephrology Unit, Mansoura University Children's Hospital, Mansoura, Egypt; [2]Clinical Pathology Department, Faculty of Medicine, Mansoura, Egypt; [3]Dermatology Department, Faculty of Medicine, Mansoura, Egypt; [4]Rheumatology Department, Faculty of Medicine, Mansoura, Egypt; [5]Pathology Department, Faculty of Medicine, Mansoura, Egypt.*
**Objectives:** Lupus nephritis (LN) occurs in about 65% of children with Systemic Lupus Erythematosus (SLE). Lipocalin-2 is a protein secreted by leukocytes during inflammation. It is over expressed in the kidneys following ischemic and nephrotoxic damage.This work was done to study the level of urinary and serum Lipocalin-2 in children with SLE and to investigate its possible role as a marker of renal involvement.

**Methods:** Urinary and serum Lipocalin-2 levels were assessed in 33 children with active SLE (22 with LN and 11 with no renal involvement) and compared to 15 age and sex matched controls.
**Results:** Children with SLE had elevated urinary Lipocalin-2 levels when compared to controls ($P < 0.001$). Urinary lipocalin-2 levels were higher in LN patients in comparison to those without LN ($p < 0.001$). In LN children; serum Lipocalin-2 levels were not significantly different from controls ($P = 0.4$) and urinary lipocalin-2 correlated with the renal score of SLE Disease Activity Index ($r = 0.5$, $P = 0.02$), but not with serum Lipocalin-2 levels ($P = 0.5$). Urinary Lipocalin-2 level was found to be statistically significant to detect class III & IV LN ($P = 0.005$). A cutoff point of 10.07 ng/mg creatinine with sensitivity of 91% and specificity of 70% was noted in our cohort.
**Conclusions:** Urinary lipocalin-2 seems to be a sensitive marker for severe nephritis in children with SLE.

## Abstract# 46

**Effect of Calcineurin Inhibitors in Nephrin and Podocin Gene Expression in Experimental Nephrotic Syndrome** A.M. Hernandez, M. Medeiros, R. Hernandez-Mote, Y. Fuentes, L. Ortiz, F. Velásquez-Forero, A. Mendoza, G. Ramon, B. Rodriguez, R. Muñoz. *Hospital Infantil de México Federico Gómez, Mexico, DF, Mexico.*
**Objectives:** To determine if the administration of calcineurin inhinitors (CI) ) modifies the gene expression of nephrin and podocin in puromycine aminonucleoside (PAN) induced nephrotic syndrome (NS).
**Methods:** There were six groups of male Wistar rats (n=9 each group). Three groups were nonnephrotic controls, they received saline solution 1.5ml SC, Group I: corn oil, Group II: CsA 20m/Kg/day, Group III Tac 0.3mg/Kg/day. Three groups received PAN 150mg/kg SC: Group IV: PAN+corn oil, Group V: PAN+ CsA, Group VI: PAN+ Tac.
Drugs and the vehicle were adiminestered from day 6 to day 14 after PAN injection. Proteinuria was quantified daily. The animals were sacrificed on day 14 and blood saved for cholesterol, tryglicerides, serum albumin. Kidney gene expression of nephrin and podocin was obtained by real time PCR.
**Results:** See Table.

Gene expression

| | I | II | III | IV | V | VI |
|---|---|---|---|---|---|---|
| Nephrin copy/µg RNA | 10.9 ± 0.5 | 9.9 ± 0.6* | 10.7 ± 0.4 | 10.3 ± 0.5 | 10.3 ± 0.5 | 10.2 ± 0.95 |
| Podocin copy/µg RNA | 10.8 ± 0.7 | 10.8 ± 0.8 | 10.7 ± 0.4 | 10.4 ± 0.2 | 10.6 ± 0.4 | 10.9 ± 0.8 |

ANOVA $p = 0.03$
**Conclusions:** CsA and Tac treatment did not improve proteinuria in PAN induced NS animals.
The gene expression of nephrin was reduced in PAN induced NS. The treatment with calcineurin inhibtors in PAN induced NS increases the expression of nephrin. Podocin gene expression was not modified by the PAN induced model neither by the CI treatment.
**DISCLOSURE:** *Medeiros, M.*: Consultant, Novartis, Mexico.

## Abstract# 47

**Efficacy of Prednisone –Tacrolimus vs. Prednisone – Cyclosporine in Steroid-Resistant Nephrotic Syndrome** S. Valverde, A.M. Hernandez, L. Velasquez, B. Romero, A. Mendoza, G. Ramon, M. Medeiros. *Hospital Infantil de México Federico Gómez, Mexico, Mexico.*
**Objectives:** To determine if the treatment with Prednisone (PDN) and Tacrolimus (FK) in pediatric patients with steroid resistant nephrotic syndrome (SRNS) for 12 months is superior to PDN and Cyclosporine(CyA) to induce and maintain remission.
**Methods:** A comparative, randomized clinical trial was conducted in children with SRNS. Group I receive PDN+CsA, Group II PDN+FK for 12 months. Blood and urine samples were drawn monthly for serum creatinine, total protein albumin, lipids, calcineurin inhibitor levels and proteinuria.
**Results:** 17 patients were included. Response rate is depicted in Table 1.

Response to treatment

| | Group I CyA+PDN (n=10 ) | Group II Tac+PDN (n= 7 ) |
|---|---|---|
| Remission (n,%) | 10 (100%) | 7 (100%) |
| Partial | 6 (60%) | 6 (85.7%) |
| Complete | 4 (40%) | 1 (14.3%) |
| Hypertension | 8 (80%) | 1 (14.3%)* |
| Time to achieve remission (weeks mean ± SD) | 24 ± 18 | 16 ± 7 NS |

*Chi square $p < 0.05$. NS: No statistically significant.
One patient in Group I had a relapse 20 weeks after initial remission,associated to infection and remitted after PDN reintroduction.
**Conclusions:** Both calcineurin inhibitors are useful in the treatment of SRNS. PDN+FK have a greater proportion of complete remissions and lower incidence of hypertension.
**DISCLOSURE:** *Medeiros, M.*: Consultant, Novartis, Mexico.

## Abstract# 48

**Cyclosporin A Treatment for Dense Deposit Disease with Steroid Resistant Nephrotic Syndrome** M. Hiramatsu. *Pediatric Nephrology, National Nishibeppu Hospital, Beppu Souenn 6-5, Oita, Japan; Pediatric Nephrology, National Nishibeppu Hospital, Beppu Souenn 6-5, Oita, Japan; Nephrology, Kumamoto Welfare University, Kumamoto, Kumamoto, Japan.*
**Objectives:** Dense Deposit Disease(DDD) is rare and only a relatively smallnumber of cases and studies in Japan.
A steroid resistant nephritic syndrome with densedeposit deposit disease was treated withcyclosporin A(CSA) and low dose predonisolone.
**Methods:** An18-year old Italian boy had initially demonstrated microhematuria and proteinuria and hypocomplementemia at theage of 14years.
The histological findings on renal biopsy revealed mesangial interposition and doublecontours and ultrastructurally intensive electron dense substance in the lamina dense of the GBM.
He was diagnosed with DDD and treated with pulse therapy followed by an alternate-day prednisolone regimen(1mg/kg).
**Results:** At the age of 17years,with discontinue predonisolone, he developed the severe nephrotic syndrome, hypertension, renal dysfunction(creat.0.98mg/dl).
He was not successfully treated with steroid therapy. So,he was additionally treated with CSA combined with predonisolone (1mg/kg). The trough level was 100-120 ng/ml.
**Conclusions:** Following recovery from the nephrotic syndrome, CSAand predonisolone was reduced to alternate day low dose.
However second renal biopsy revealed increased mesangial interposition and double contouts and fibro cellular crescent, anddense deposit by EM.

## Abstract# 49

**Retrospective Investigation of Incentive in Children with Recurrent Henoch-Schonlein Purpura** Q.-M. Huang, H.H. Chen, D. LI. *Pediatric Department, Second Affiliated Hospital of Guanzhou University of Traditional Chinese Medicine, Guangdong, Guangzhou, China.*
**Objectives:** Objective To investigate the incentive features of recurrent Henoch-Schonlein purpura (AP) in Children.
**Methods:** 76 cases of AP in children were studied, including Survey indicators of incentives including:the infection rate, ASO-positive rate, Mycoplasma pneumoniae(MP) positive rate, the Food and drug allergy history-positive rate, positive rate of serum total IgE, serum allergen-specific IgE-positive rate.
**Results:** 1, The male to female ratio of AP in early-onset group (1.25:1 ) was lower than that in the recurrent group (1.82:1), $P < 0.05$; 2, Infection rate of AP in early-onset group( 73.33%) was higher than that in the recurrence group (51.61%), $P < 0.05$; MP positive rate in the early onset group and ASO-positive rate were 22.22%, 13.33%,respectively, for the recurrent group, they were 29.03%, 6.5%, respectively, there were no difference in the corresponding rate, $P > 0.05$; 3,Drug and food allergy history-positive rates in early onset and recurrent group were 17.78%, 19.36%,There was no difference between the two groups, $P > 0.05$; serum total IgE Higher than normal levels in early-onset and recurrent group were 46.67%, 42.86%, There was no difference between the two indicators, $P > 0.05$; The positive rates of Food and inhaled allergens in two groups were 40%, 33.33%, no difference between the two indicators, $P > 0.05$.
**Conclusions:** 1,It seems that boy has more chance to relapse than girl in AP ; 2, The proportion of non-infectious incentive was risen in recurrent AP; 3,there were no diffrences in MP infection, streptococcal infection, allergic factors in AP at stage of early-onset and recurrent.

## Abstract# 50

**Intravenous Cyclophosphamide for Lupus Nephritis in Children** L. Huynh Thoai, D. Nguyen thi Ngoc. *Nephrology, Children's Hospital 1, Ho Chi Minh, Viet Nam.*
**Objectives:** This study evaluates the clinical efficacy of CYC IV to achieve remission in children with severe lupus nephritis
**Methods:** prospective descriptive study. Patients SLE who had 4 /11 ARA, biopsy proven lupus nephritis with either diffuse proliferative glomerulonephritis (WHO class IV) or focal proliferative glomerulonephritis (WHO class III) and who had received no immunosuppressive agent other than oral steroids were enrolled in the study.
**Results:** 47 pateints were diagnosed lupus nephritis WHO III, IV (8 WHO III, 38 WHO IV). 1 did not attend follow-up, non died. 46 patients completed 6 months therapy with CYC. 41 (89%) girls, 4 (11%) boys. The mean age was 12 ± 2.5 years (range 8–16 years). At the time of diagnosed: 16 (34.7%) of patients were hypertention (blood pressure ≥ 140/90 mmHg), 2 (4%) macro hematuria, 6 (13%) renal failure (serum creatinine ≥ 1.2mg/dl). Clinical significance of lupus nephritis: 17 (37%) nephrotic syndrome (proteinuria > 50mg/kg/day or proteinuria/creatininuria > 200 mg/mmol), 1 (2%) nephritis (hematuria or cellular

casts, hypertention, ± renal failure), 11 (24%) nephotic – nephritis syndrome, 17 (37%) abnormal urinalysis. After 6 times CYC IV: 3(6.5%) non response, 43 (93.5%) response: 25 (54.3%) partial remission, 18 (39.1%) complete remission.
**Conclusions:** CYC IV seems to be effective in the treatment of severe lupus nephritis with minor side-effects. However, the long-term outcomes and side-effects of CYC need to be followed up.

## Abstract# 51

**Prevalence of Hivan in an African Paediatric HIV Population** E.E. Ikpeme, M.U. Akpan, U. Ekrikpo, O. Etukudo. *Department of Paediatrics, University of Uyo Teaching Hospital, Uyo, Akwa Ibom, Nigeria.*
**Objectives:** HIVAN is the most common form of chronic kidney disease resulting directly from HIV infection. Its true prevalence in Africa is unknown. The study set out to determine the prevalence of HIVAN in a tertiary institution in Nigeria using persistent proteinuria with renal ultrasound changes. Its correlation, with CD4 count, duration of treatment with highly active antiretroviral therapy (HAART) and association with clinical staging of the disease was also examined.
**Methods:** This was a prospective study over a six month period conducted in the Infectious Diseases Unit of the Department of Paediatrics, University of Uyo Teaching Hospital, Uyo, Nigeria involving all HIV positive children. Urine microalbumin-creatinine ratio > 30µg albumin/mg creatinine was considered positive for micro albuminuria. These were measured at baseline and at 4 weeks. Renal ultrasound, CD4, clinical staging of HIV and treatment with HAART were done in all the patients and assessed.
**Results:** Prevalence rate of HIVAN was 31.6%, out of which 3.1% had abnormal ultrasound findings. There was a significant correlation ($r = -0.22$, $p = 0.03$ ) between CD4 count and HIVAN. There was no correlation between HIVAN and duration on HAART ( $r = -0.10$, $p = 0.31$) and no association between HIVAN and clinical staging of HIV ( $p=0.14$).
**Conclusions:** Prevalence of childhood HIVAN in Nigeria is high. Screening for urine microalbuminuria is essential for the early diagnosis of HIVAN and prevention of progression to CKD. Identified risk factors were low CD4 count and late clinical stages of HIV infection.

## Abstract# 52

**Efficacy and Safety of Tacrolimus in Children with Refractory Nephrotic Syndrome** X.Y. Jiang, R.H. Lin, L.Z. Chen, Y.H. Ling, Y. Mo. *Pediatrics, Sun Yat-sen University, Guangzhou, China.*
**Objectives:** To observe the efficacy and safety of a 12-24 months course of tacrolimus(FK506) therapy in children with refractory NS.
**Methods:** Of the 16 patients enrolled in our study, 2 were SDNS, 10 were SRNS and 4 were FRNS. All patients initially received prednisone 2 mg/(kg·d). Fifteen of 16 patients were nonrespnsive to other therapies included CsA, CTX and mycophenolate mofetil(n=4). All these patients received a combined FK506 [(0.10~0.15) mg/(kg·d)] / prednisone treatment. The dose of FK506 was adjusted to maintain a blood level of (5.0~10.0) µg/L during the initial 10 months of treatment, then tapered off gradually until the total course of the treatment was 12-24 months. 24 hours urine protein and the blood level of Scr, BUN, Alb, ALT, Ccr, Chol, Glu, PLT were measured before and after FK506 treatment. Side effects of FK506 were observed at the same time.
**Results:** Remission was achieved in 15(complete remission 12, partial remission 3) of 16 patients (93.8%). The remission rate was 100% in simple type NS, while 87.5% in nephritis type NS. And the remission rates in FRNS, SDNS and SRNS groups were 100%, 100% and 90.0%, respectively. In MCD group and MsPGN group, the remission rates were both 100%, while 75.0% in FSGS group. Two cases(12.5%) relapsed when FK506 was tapering. One patient had gastrointestinal tract reaction and anorexia,while another had insomnia during the combined FK506/prednisone treatment.
**Conclusions:** FK506 was effective and well-tolerated in children with refractory NS, especially to NS children who were FRNS or had MCD. Besides, Our study also suggested that FK506 were effective for NS in children who were nonrespnsive to CsA and CTX.

## Abstract# 53

**Effects of Prednisone and Benazepril on Proteinuria and Podocyte-Associated Molecules in Rats with IgA Nephropathy** H.-y. Lu, X.-Y. Jiang, L.-z. Chen, Y. Mo, S.-m. Chen. *Pedaitrics, The First Affiliated Hospital of Sun Yat-sen University, Guangzhou, Guangdong, China.*
**Objectives:** To observe the effects of prednisone and benazepril on proteinuria in rats with IgAN and changes of expression and distribution of nephrin, podocin, CD2AP and desmin, and to explore the mechanism of proteinuria of IgAN and try to find the approaches to the therapy for IgAN.
**Methods:** SD rats were divided into control group, model group, prednisone group and benazepril group. The IgA intensity was examined by direct immunofluorescence. Expression and distribution of nephrin, podocin and desmin were examined by indirect immunofluorescence. The mRNA expression of nephrin, podocin, CD2AP and desmin were examined by RT-PCR.
**Results:** (1) Hematuria and proteinuria in prednisone and benazepril group were less than model group. (2)Nephrin, podocin and desmin in model, prednisone and benazepril group were higer than control group. Nephrin in prednisone and benazepril group were higher than model group, but podocin and desmin were lower than model group. Abnormal discontinuous distribution of nephrin and podocin in prednisone and benazepril group were slighter than model group. (3) There was no difference of mRNA expressions of nephrin, podocin and CD2AP between control group and model group, however, desmin mRNA in model group was higher than control group. In prednisone group and benazepril group, mRNA of nephrin, podocin and CD2AP expressed significantly higher than model group, but desmin mRNA was lower than that in model group.
**Conclusions:** Both prednisone and benazepril are good for decreasing urinary protein by the way of regulating the expression and distribution of nephrin, podocin, CD2AP and desmin.

## Abstract# 54

**Effect of Huaiqihuangkeli on Proteinuria and Podocyte-Associated Molecules in Rats with IgA Nephropathy** H.-y. Lu, X. Jiang, L.-z. Chen, Y. Mo, S.-M. Chen. *Pedaitrics, The First Affiliated Hospital of Sun Yat-sen University, Guangzhou, Guangdong, China.*
**Objectives:** To observe the effect of huaiqihuangkeli on proteinuria in rats with IgAN and changes of expression and distribution of nephrin, podocin, CD2AP and desmin, and to explore the mechanism of proteinuria of IgAN and try to find the approache to the therapy for IgAN.
**Methods:** SD rats were divided into control group, model group, and huaiqihuang group. IgAN model was induced by the method of BSA+$CCl_4$+LPS. The IgA intensity was examined by direct immunofluorescence. The protein expression and distribution of nephrin, podocin and desmin were examined by indirect immunofluorescence. The mRNA expression of nephrin, podocin, CD2AP and desmin were examined by RT-PCR.
**Results:** (1) Hematuria and proteinuria in huaiqihuang group was less than model group. (2) Expression of nephrin, podocin and desmin in model were higer than control group. In huaiqihuang group, nephrin was higher than model group. Podocin had no difference from that in model group, but was still higher than control group. Desmin was lower than model group, but higher than that group. Abnormal discontinuous distribution of nephrin and podocin in huaiqihuang group was slighter than model group. (4) There was no difference of mRNA of nephrin, podocin and CD2AP between control and model group, however, desmin mRNA in model group was higher than control group. In huaiqihuang group, mRNA of nephrin, podocin and CD2AP expressed higher than that in model group, but desmin mRNA has no difference from that in model group.
**Conclusions:** Huaiqihuangkeli is good for decreasing urinary protein by the way of regulating the expression and distribution of nephrin, podocin, CD2AP and desmin.

## Abstract# 55

**Effect of Huangqi on Proteinuria and Podocyte-Associated Molecules in Rats with IgA Nephropathy** H.-y. Lu, X.-Y. Jiang, L.-z. Chen, Y. Mo, S.-m. Chen. *Pedaitrics, The First Affiliated Hospital of Sun Yat-sen University, Guangzhou, Guangdong, China.*
**Objectives:** To observe the effect of huangqi on proteinuria in rats with IgAN and changes of expression and distribution of nephrin, podocin, CD2AP and desmin, and to explore the mechanism of proteinuria of IgAN and try to find the approache to the therapy for IgAN.
**Methods:** SD rats were divided into control group, model group, and huangqi group. IgAN model was induced by the method of BSA+$CCl_4$+LPS. The IgA intensity was examined by direct immunofluorescence. The protein expression and distribution of nephrin, podocin and desmin were examined by indirect immunofluorescence. The mRNA expression of nephrin, podocin, CD2AP and desmin were examined by RT-PCR.
**Results:** (1) Hematuria and proteinuria in huangqi group was less than in model group. (2) Nephrin, podocin and desmin in model were higer than control group. In huangqi group, nephrin had no difference from that in model group, but was higher than control group. Podocin was lower than model group, but had no difference from that in control group. Desmin was higher than model group and control group. Abnormal discontinuous distribution of nephrin and podocin in huangqi group was slighter than model group. (3) There was no difference of mRNA expression of nephrin, podocin and CD2AP between control group and model group, however, desmin mRNA in model group was higher than control group. In huangqi group, mRNA of nephrin, podocin and CD2AP expressed higher than model group, but desmin mRNA has no significant difference from that in model group.
**Conclusions:** Huangqi is good for decreasing urinary protein by the way of regulating the expression and distribution of nephrin, podocin, CD2AP and desmin.

## Abstract# 56

**Improved IgA Nephropathy Model** H.-y. Lu, X.-Y. Jiang, L.-z. Chen, Y. Mo, S.-m. Chen. *Depatment of Pedaitrics, The First Affiliated Hospital of Sun Yat-sen University, Guangzhou, Guangdong, China.*

**Objectives:** To improve the method of constructing IgA nephropathy (IgAN) model and seek a more reliable and stable method.

**Methods:** SD rats (n=96) were divided into control group (n=48) and model group (n=48). Reforming method: the dose of oral immunogen bovine serum albumin (BSA) increased one time (400mg/kg every other day), lasting 8 weeks; the method of injecting carbon tetrachloride ($CCl_4$) was subcutaneous injection instead of intraperitoneal injection, dose of which was one third of content to induce hepatic fibrosis (0.1mL), weekly, continuing 9 weeks; combined with lipopolysaccharide (LPS) 0.05mg through tail vein once in the 6th week. Urine, serum and renal tissue samples were collected at the end of the 2nd, 4th, 6th, 8th, 10th and 12th week.

**Results:** Hematuria and proteinuria appeared at the end of the 6th week in model group. Count of urinary red cells and urinary protein continued increasing till the end of the 12th week. There were no significant differences of ALT, AST, ALB, TP, CHOL, TG, TBIL, Cr, BUN and CK-MB between model group and control group(all P>0.05). Under LM, renal histopathologic changes became obvious after the end of the 8th week in model group, that was moderate to severe mesangial proliferation. Under EM, foot processes fusion appeared at the end of 4th week in model group, that was more and more severe till the end of the 12th week. Immunofluorescence showed intensity of IgA deposition in glomeruli in model group was ++~+++.

**Conclusions:** Rat IgAN model is successfully established by the improved method of BSA+$CCl_4$+LPS, whose clinical index and pathology are similar to the human IgAN.

## Abstract# 57

**Changes and Significance of Podocyte-Associated Molecules in Rats with IgA Nephropathy** H.-y. Lu, X.-Y. Jiang, L.-z. Chen, Y. Mo, S.-m. Chen. *Depatment of Pedaitrics, The First Affiliated Hospital of Sun Yat-sen University, Guangzhou, Guangdong, China.*

**Objectives:** To observe the expression and distribution of nephrin, podocin, CD2AP and desmin in rats with IgAN, and to explore the mechanism of proteinuria of IgAN.

**Methods:** SD rats were divided into control group and model group. IgAN model was induced by the method of BSA+$CCl_4$+LPS. Expression and distribution of nephrin, podocin and desmin were examined by indirect immunofluorescence. The mRNA expression of nephrin, podocin, CD2AP and desmin were examined by RT-PCR.

**Results:** (1) In model group, proteinuria increased at the 6th week and persisted up to the 12th week. (2) Podocin and nephrin increased from the 2nd and 4th week individually to the 8th week, then decreased, but higher than control group at the 12th week. Podocin and nephrin staining gradually shifted from a linear-like pattern along the capillary loop of glomeruli to a discontinuous patch or gobbet pattern. Expression of desmin increased at the 2nd week and elevated up to the 12th week. (3) Nephrin, podocin and CD2AP mRNA were up-regulated at the 2nd, 2nd and 4th week individually, which persisted increasing to the 8th week, and thereafter returned to control level at the 12th week. Desmin mRNA increased at the 8th week, then recovered again, but it was still higher than control group at the 12th week. (4) Nephrin, podocin and mRNA, CD2AP mRNA and desmin mRNA were all positively correlated with urinary protein.

**Conclusions:** Changes of expression and abnormality of distribution of podocyte-associated molecules may be the cause of proteinuria of IgAN. Change of podocin may be the trigger of proteinuria, and change of CD2AP may be caused by the change of nephrin and podocin.

## Abstract# 58

**The Protean Nature of Childhood SLE May Cause a Delay in the Diagnosis of Lupus Nephritis** R. Jodorkovsky, K. Bromberg. *Pediatrics, The Brooklyn Hospital Center, Brooklyn, NY, United States.*

**Objectives:** Lupus nephritis occurs in 80% of childhood-onset SLE. Systemic symptoms of lupus are non-specific. Given non-specific symptoms clinicans may not look for renal dysfunction and possibly delay the treatment of lupus nephritis.

**Methods:** Demographics, pre-renal biopsy systemic and renal presenting symptoms, their duration, and pathology diagnosis were reviewed in all patients diagnosed with lupus nephritis from 2005-2009.

**Results:** There were 11 new cases. The diagnosis was confirmed between 5 months to 6 years after the onset of systemic symptoms in the majority of cases. During this time, several children did not appear to have had renal testing. Renal pathology was severe in most cases.

| Age | Sex/Eth | Pre-Bx syst.symp | Time | Pre-Bx renal symp | Time | C3-4/ Antibod | Class |
|---|---|---|---|---|---|---|---|
| 16 | M/AA | Abd pain | 6 mo | NephrSynd, ↑BP,↓GFR | 2 wks | ↓/⊕ | 5 |
| 7 | M/AA | Fever,↑lymph | 10 mo | NephrSynd,↓GFR | 1 wk | ↓⊕ | 4 |
| 14 | F/AA | Arthr,↓WBC | 4 yr | Uprot | 4 wks | N/⊕ | 2 |
| 9 | F/Chin | ↓Plat,rash | 6 yr | NephrSynd | 1 wk | ↓/⊕ | 5 |
| 16 | F/AA | Myalg,fever | 10 mo | Uprot,↓GFR | 1 wk | N/- | 4 |
| 17 | F/His | Arthr,RVT,lung thrombi | 6 mo | NephrSynd | 5 wks | ↓/⊕ | 5 |
| 19 | M/His | Arthr,rash,fever | 6 mo | NephrSynd,↓GFR | 2 wks | ↓/⊕ | 4 |
| 15 | F/AA | Edema | 6 wks | NephrSynd | 2 wks | ↓/⊕ | 5 |
| 17 | F/AA | Malaise | 2 wks | NephrSynd,↑BP | 2 wks | ↓/⊕ | 4 |
| 13 | F/AA | Abd pain,vomit | 6 mo | NephrSynd,↓GFR,↑BP | 1 wk | ↓/⊕ | 4 |
| 18 | M/AA | Diarrh,fever | 8 days | NephrSynd,↓GFR,↑BP | 3 days | ↓/⊕ | 4 |

**Conclusions:** Lupus nephritis can be preceded by a protean assortment of systemic symptoms lasting from several months to years. During this time, renal dysfunction may already be present and its detection could be renal preserving. A high index of suspicion for nephritis should be considered when children present with prolonged imprecise symptoms that could be lupus.

## Abstract# 59

**Descriptive Study of Biopsy Proven IgA and Henoch Schonlein Purpura (HSP) Nephropathy in Two Government Hospitals in Johannesburg, South Africa** J.G. Mitchell,[1] U.K. Kala,[1] D. Hahn.[2] *[1]Paediatrics, University of the Witwatersrand and Chris Hani Baragwanath Hospital, Johannesburg, Gauteng, South Africa; [2]Paediatrics, University of the Witwatersrand and Charlotte Maxeke Johannesburg Academic Hospital, Johannesburg, Gauteng, South Africa.*

**Objectives:** Determine if IgA and Henoch Schonlein Purpura(HSP) nephropathies occur less frequently in South African Black Children and the disease progression is worse.

**Methods:** A retrospective review of biopsy proven IgA or HSP nephropathy presenting at two paediatric nephrological services in two academic hospitals namely Chris Hani Baragwanath and Charlotte Maxeke Johannesburg Academic Hospitals in Johannesburg from January 1985 to December 2008 in children ≤ 16 years.

**Results:** Total of 1835 renal biopsies of which 51 (3%) confirmed HSP or IgA nephropathy. 1 excluded from analysis due to inadequate records. Average age of presentation was 9.5 years. All children well nourished. M:F ratio 2.2:1. HSP group n=17(34%) and IgA group n=33 (66%).Racial breakdown, Black n=28 (56%), Caucasian n=15(30%), Asian n=4(8%) and Mixed races n=3(9%). Average calculated GFR (Swartz formula) was 100ml/min/1.73m$^2$.Commonest presenting symptom was haematuria macroscopic >macroscopic.50% had nephrotic range proteinuria. 22% (11) presented in acute renal failure 10 (91%) of which were IgA. Histological grading, 26% grade 1, 28% grade 2, 28% grade 3, 12% grade 4 and 6% grade5. Average follow up of 3 years.5 children dialysed, 2 in ESRF received transplants, 3 demised. Various treatment regimes used.

**Conclusions:** Neither race nor sex indicated poorer prognosis. No difference in outcome of IgA and HSP nephritis. No correlation between presenting symptoms and outcome based on disease activity.

## Abstract# 60

**5% vs 20% Albumin in Nephrotic Syndrome** M. Kanitkar, A. Garg, V. Venkateshwar. *Department of Pediatrics, Armed Forces Medical College, Pune, India.*

**Objectives:** Compare efficacy of 5% Vs 20% albumin for edema in nephrotic syndrome.

**Methods:** 24 nephrotics 15 to144 months with anasarca randomised to two groups; Group A(n=14) received 20% albumin followed by 5% and group B(n=10) vice versa with a wash out period of 48 hours.Baseline and post therapy vitals, fluid intake, urine output and biochemistry noted. Frusemide given midway to all. Data tabulated for 20% Group 1(n=24) and 5% Group2(n=24). Paired 't' test used for efficacy of either 5% or 20% and unpaired 't' test to compare difference between the two. The effect of sequential use of 5% and 20 % was analysed for Groups A and B.

**Results:** Baseline and post transfusion FeNa was 0.7±0.72%, 0.73±0.76% and 1.46±1.1%,1.73±2.3% in groups 1 & 2.Urine output and change in weight in the two groups depicted and the effect of sequential use of albumin.

Table 1 Effect of Albumin Infusion (n=24)

| Variables | Group1 (5%Albumin) | Group2 (20%Albumin) | 't' | p value |
|---|---|---|---|---|
| Δ%Wt loss | 2.25±2.12 | 3.68±3.84 | 1.6 | .12 |
| ΔUrine output(ml/kg/hr) | 1.52±1.11 | 1.66±0.95 | .47 | .64 |

Table 2 Effect of sequential use of albumin Gp A Vs Gp B*

| Variable | t value | p value |
|---|---|---|
| ΔUrine output treatment effect | -1.768 | 0.0909 |
| ΔUrine output periodic effect | 2.876 | .0088 |
| ΔWt loss treatment effect | -2.5318 | 0.01 |
| ΔWt loss periodic effect | 1.8895 | .07 |

*Gp A(14)- 5% followed by 20% albumin Gp B(10)- 20% followed by 5% albumin

**Conclusions:** Both 20% and 5% albumin equally safe and effective in anasarca due to nephrotic syndrome.There is an additive effect of sequential use of both strengths seen after the second infusion which is more if 5% albumin is given first followed by 20%.

## Abstract# 61

**Mycophenolate Mofetil (MMF) Treatment in Paediatric Onset Systemic Lupus Erythematosus (SLE)** I. Kazyra, C. Pilkington, S.D. Marks, K. Tullus. *Paediatrics, 2nd Children's Hospital, Belarus State Medical University, Minsk, Belarus; Paediatric Nephrology, Great Ormond Street Hospital, London, United Kingdom.*

**Objectives:** We present our safety and efficacy data on the use of MMF treatment in children with SLE and lupus nephritis (LN).

**Methods:** 31 children and adolescents, aged 5-22 (median 16) years with SLE were treated with MMF at GOSH, London, UK. 22/31 patients had biopsy-proven LN. Treatment outcome was monitored according the British Isles Lupus Assessment Group (BILAG) index, blood and urine parameters. Group 1 were commenced on MMF induction and/or maintenance therapy (n=15) and Group 2 were converted from azathioprine (AZA) due to inadequate disease control (n=16).

**Results:** 77% of all (67% group 1 and 88% group 2) patients experienced an improvement in BILAG at 12 months after initiation of MMF treatment ($p < 0.05$). Group 1 with low C3, C4 increased their C3 (from 0.53 to 1.15) and C4 (0.08 to 0.17g/l) levels significantly ($p<0.05$), whereas group 2 patients' C3, but not C4 showed increased levels (0.56 to 0.12, $p<0.05$). Renal function and albuminuria improved in children with active LN ($p\leq0.01$). Significant improvements were seen in both groups in Hb, ESR and lymphocyte counts. Prednisolone dose was weaned in Group 1 from 25 (range 5-60) mg, $p<0.05$ to 7 (0-10) and in Group 2 from 13 (0-60) to 5 (0-20), $p<0.05$. Side-effects of nausea, diarrhoea, leucopenia and viral infection were seen in 7(22.5%) but none were judged to be severe enough to discontinue treatment.

**Conclusions:** MMF treatment in our cohort of children with SLE and LN seemed to be safe, well-tolerated and effective. Randomized controlled trials are urgently needed to further define the role of MMF in childhood SLE.

## Abstract# 62

**Efficacy of Pulse Methylprednisolone & Cyclophosphamide for Steroid Resistant Nephrotic Syndrome** J. Smuk, S.E. Kennedy, F.E. Mackie, A.R. Rosenberg, G. Kainer. *Nephrology, Sydney Children's Hospital, Randwick, NSW, Australia.*

**Objectives:** To examine the effectiveness of pulse methylprednisolone (MP) and cyclophosphamide (CPA) therapy in steroid resistant nephrotic syndrome (SRNS).

**Methods:** Retrospective review of patients with SRNS treated with pulse MP in our unit. SRNS defined as failure to enter remission after 4 weeks of oral prednisone. MP treatment was 30mg/kg second daily for 6 doses then weekly for 8 doses and then fortnightly for 8 doses. Those with FSGS received prolonged treatment over 18 months. CPA was commenced after 6 MP doses. Full remission was defined as ≤ trace protein on UA and normal serum albumin.

**Results:** 26 patients were included. Median age at onset of NS 3.3 years (range 1.2 to 14.4 years). 12 (46%) had FSGS; all others had minimal change disease (MCD).

Full remission occurred during MP therapy in 20 patients (77%). Median number of MP doses before remission was 3.5 (range 1 to 12). 14 patients subsequently relapsed. Median duration of remission was 10 months (range 1 to 65).

All but 2 patients with MCD responded (86%) vs 67% with FSGS (NS). All responders with MCD responded within 1 month of MP therapy; those with FSGS tended to take longer (median 5.5 doses, range 3 to 12).

23 (88%) patients received CPA. 74% (17/23) of patients who received both MP and CPA achieved full remission. Nine (53%) of these patients achieved remission prior to receiving CPA.

There was no significant change in height or BMI z scores after MP treatment.

**Conclusions:** MP induced remission in the majority of patients with SRNS regardless of pathology. Pulse MP and oral CPA should be considered as an initial treatment option for SRNS. Exploration of long-term outcomes is warranted.

## Abstract# 63

**Clinical Features of Five Children with Mental Retardation and Massive Proteinuria** E. Kikuchi,[1] K. Kamei,[1] H. Kaito,[1] M. Ogura,[1] K. Matsuoka,[2] S. Ito.[1] *[1]Nephrology, National Center for Child Health and Development, Tokyo, Japan; [2]Pathology, National Center for Child Health and Development, Tokyo, Japan.*

**Objectives:** The combination of mental retardation and nephrotic syndrome is rare. Galloway-Mowat syndrome (GMS) is an autosomal recessive disorder characterized by early onset nephrotic syndrome and microcephary. Clinical manifestations and histological findings of GMS show a wide variation.

**Methods:** We reviewed the medical records of five children with unexplained mental retardation and nephrotic syndrome.

**Results:** All patients showed microcephary with diffuse cortical atrophy. Four of the five had infantile spasms. Three developed nephrotic syndrome before two years of age and died from renal failure at early ages(cases 1, 2 and 3). The other two children developed nephrotic syndrome after four years of age and remain alive without renal dysfunction. Nephrotic syndrome was steroid-resistant in all cases. Renal pathology on light microscopy showed diffuse mesangeal hypercellularity (cases 1 and 2), FSGS (case 3), MPGN like with mesangiolysis (case 4) and extracapillary glomerulonephritis (case 5). All patients had effacement of foot processes and two had irregular thickness of the glomerular basement membranes (cases 1 and 4).

**Conclusions:** The three patients with early onset nephrotic syndrome were clinically diagnosed with typical GMS while the other two patients may have had a milder form. Some podocyte proteins involved in the formation of cellular processes and signal transduction are known to be expressed in the kidneys and brain. It is possible that some unknown mutations in podocyte proteins are responsible for the proteinuria and brain anomalies seen in GMS.

## Abstract# 64

**Early Intensive Plasma Exchange (PEX) for Atypical Haemolytic Uraemic Syndrome (aHUS) May Slow Progression to End-Stage Renal Failure (ESRF)** J. Kim,[1] T. Goodship,[2] J. Tizard,[1] C. Inward.[1] *[1]Bristol Children's Hospital, Bristol, United Kingdom; [2]Institute of Human Genetics, Newcastle University, Newcastle, United Kingdom.*

**Objectives:** We report a single centre analysis of renal response to PEX in aHUS.

**Methods:** Retrospective case series.

**Results:** 3 patients with diarrhoea negative aHUS presented with acute renal failure and severe hypertension aged 4 months, 22 months and 6 months (Patients 1-3 respectively). Daily PEX was commenced early following presentation in addition to dialysis. This resulted in HUS remission and cessation of dialysis after 2 weeks, 9 days and 2 weeks. Relapses were common and associated with increasing time interval between PEX. All relapses responded to intensification of PEX therapy. Patient 1 only recovered 50% of renal function after first presentation. She had 4 relapses and 32 months after presentation she remained dialysis free but was approaching ESRF. Mutation screening of *CFH* showed a missense mutation (c.3546G>T, p.Arg1182Ser) in exon 23. Patient 2 had slow tapering of PEX over 4 months to fortnightly sessions and relapsed when PEX was extended to four weekly. Renal function remained normal 12 months post-presentation. Mutation screening of *CFH* showed a mutation in exon 23 (c.3590T>C, p.Val1197Ala) and two sequence variants in exon 3 and 4. Patient 3 had 2 relapses associated with intercurrent illnesses concurrently with reducing to weekly PEX. Renal function was normal 5 months post-presentation. Genetic results pending.

**Conclusions:** Our series of aHUS infants had good renal response to PEX with improvement after both initial and relapse management. Further research is necessary to determine best maintenance strategy to delay progression to ESRF.

**DISCLOSURE:** *Goodship, T.*: Other, Member of Scientific Advisory Boards of Taligen Therapeutics and LFB.

## Abstract# 65

**Sustained Remission of Refractory Nephrotic Syndrome with Combined Rituximab and Mycophenolate Mofetil Therapy** W. Seeherunvong, C. Abitbol, J. Chandar, M. Freundlich, G. Zilleruelo. *Division of Pediatric Nephrology, University of Miami/Holtz Children's Hospital, Miami, FL, United States.*

**Objectives:** To evaluate the efficacy of Rituximab (RTX) and Mycophenolate Mofetil (MMF) in achieving remission in refractory idiopathic nephrotic syndrome (INS).

**Methods:** Retrospective analysis of patients with refractory INS who received RTX as a rescue therapy. All had frequent relapses or persistent nephrotic syndrome despite multiple immunosuppressants including prednisone, alkylating agents, calcineurin inhibitors, pulse solumedrol, and MMF. All received angiotensin receptor blockers and/or angiotensin converting enzyme inhibitors. All had experienced severe complications including episodes of acute renal failure, thrombosis and morbid obesity. Rituximab was given as 4 weekly infusions of 375 mg/m$^2$ (first as a half dose). All patients and/or parents provided consent for therapy.

**Results:** Nine patients (6 males) aged 12±6 years were studied. Age at diagnosis was 4±3 years. Four were steroid dependent and 5 resistant. Five had FSGS, 3 focal mesangial proliferation, and 1 minimal change. Eight (88%) achieved sustained remission after RTX and were maintained with MMF and ACEI/ARB for a follow up of 10± 3 months. Only one relapsed 3 months after the first cycle of RTX, associated with non-adherence to any medications. After the 2nd course of RTX, he underwent remission. No serious adverse effects were observed during treatment.

**Conclusions:** Combined RTX and MMF is effective in sustained remission in children with refractory idiopathic nephrotic syndrome. A large prospective controlled study is warranted to clarify the role of B cell depletion in the management of these patients.

## Transplantation

### Abstract# 66
**(O-9)**

**Cardiorespiratory Fitness Is a Marker of Cardiovascular Health in Renal Transplanted Children** T. Tangeraas,[1] K. Midtvedt,[2] P.M. Fredriksen,[3] M. Cvancarova,[4] A. Bjerre.[1] *[1]Department of Pediatrics, Oslo University Hospital (OUH), Oslo, Norway; [2]Department of Medicine, OUH, Oslo, Norway; [3]Clinical Trials Office, OUH, Oslo, Norway; [4]Department of Oncology, National Resource Center for Late Effects OUH, Oslo, Norway.*

**Objectives:** Traditional cardiovascular (CV) risk factors are known to increase due to immunosuppressive medication in renal transplanted (TX) children. Purpose: Assess level of cardiorespiratory fitness (CRF) and daily physical activity (PA) in stable TX children and adolescents in relation to traditional CV risk factors.

**Methods:** Laboratory testing including assessment of : CRF by treadmill exercise testing ($V0_{2peak}$), 24-hour ambulatory blood-pressure, oral glucose tolerance test, anthropometrics, lipids and measured glomerular filtration rate (mGFR). PA was self-reported by questionnaire.

**Results:** Twenty-two TX patients were tested: Median (range) age 14.5 (9-18) years, time since TX 5.2 ( 2-16) years, mGFR 49.5 (22-77) ml/ min/1.73m$^2$, Hb 12 (10-16) g/dL.$V0_{2peak}$ was 66% of expected compared to healthy controls. Nineteen (86%) children reported < 60 minutes of daily moderate to vigorous activity. Fourteen (64%) were hypertensive (SBP> 95p) and 8 (34%) were overweight/obese.

Four children fulfilled criterias for the metabolic syndrome.Children with ≥ 2 of 3 CV risk factors (hypertension, overweight and impaired glucose tolerance, n=7) obtained $V0_{2peak}$ 45% of expected compared to $V0_{2peak}$ 73% of expected in children with 1 or 0 risk factors (n=15, p=0.003).

**Conclusions:** Renal TX children and adolescents have severely impaired CRF and PA. Reduced CRF is associated with clustering of CV risk factors. Routine counseling for increased PA is strongly recommended.

### Abstract# 67
**(O-10)**

**Aldosterone Blockade in Children with Chronic Allograft Nephropathy** M. Medeiros,[1] S. Ramirez,[1] L. Velasquez,[1] A.M. Hernandez,[1] S. Valverde,[1] A. Vargas,[1] K. Sanchez,[2] J. Sosa,[1] N. Bobadilla.[2] *[1]Hospital Infantil de México Federico Gómez, Mexico, Mexico; [2]Instituto Nacional de Ciencias Médicas y la Nutrición, Mexico, Mexico.*

**Objectives:** To investigate the effect of aldosterone blockade in renal allograft function in children with biopsy proven chronic allograft nephropathy CAN.

**Methods:** A prospective, randomized and blind to the patient study is performed. Patients aged 6 to 17 years, with CAN and glomerular filtration rate >40 ml/min/1.73m²SC were randomized to receive either placebo (Group I) or Eplerenone, initial dose 12.5mg/day increased to 25mg/day in two weeks (Group II). Eight visits were scheduled in the first 24 weeks. A clinical examination was performed and blood sample was drawn for complete blood cell count, serum creatinine (SCr), electrolytes, transaminases. A 24h urine collection for proteinuria and urine nitrates is also obtained.

**Results:** Thirteen patientes have completed 24 weeks of follow up, six in Group I and seven in Group II. There was no changes in serum electrolytes, hepatic enzymes and blood cell count in both groups. Patients in group I had no changes in SCr, from 1.2 ± 0.5 at baseline to 1.3 ±0.6 mg/dL at 24 weeks (paired t test 0.46), whereas patients in Group II had a baseline SCr of 0.97 ± 0.4 and diminished to 0.72 ± 0.3 at 24 weeks of eplerenone treatment (paired t test p= 0.01). There was a non significant reduction in proteinuria in the eplerenone treated group. There was no changes in urine nitrates in the first four weeks of treatment in both groups.

**Conclusions:** Eplerenone is safe and well tolerated in children with CAN, and induces a statistically significant reduction of SCr at 24 weeks of treatment.

**DISCLOSURE:** *Medeiros, M.*: Consultant, Novartis, Mexico.

### Abstract# 68
**(O-11)**

**Influence of CYP3A5 Polymorphism on Tacrolimus Doses and Serum Level after Renal Transplantation: Pharmacological Interaction with Steroids** J.R. Ferraris, J.M. Larriba, J. Torres Fuenzalida, G. Gimenez, P.A. Coccia, L.F.R. Ghezzi, V. Ferraris, M.A. Redal. *Hospital Italiano de Buenos Aires, Buenos Aires, Argentina.*

**Objectives:** Tacrolimus (Tac), mycophenolate mofetil (MMF) and methylprednisone (MP) are used in pediatric kidney transplantation (tx). The cytochrome P450 (CYP) 3A5 enzyme appear to play a role in the Tac metabolism. The aims of this study were to investigate CYP3A5 polymorphism's effect on Tac dose by testing blood concentrations and the interaction between steroids and Tac during the first year after tx.

**Methods:** Genomic DNA was extracted amplified with specific primers. CYP3A5 alleles was confirmed by direct sequencing of PCR products on an automated ABI3100 capillary sequencer.

**Results:** We studied 40 renal transplant patients (age at tx 12 +/- 0.5 years, 22 male) receiving Tac, MMF, MP. Of these, 82.5 % were CYP3A5*3/*3 (nonexpressors homozygotes) and 17.5% were CYP3A5*1/*3 (expressors). The Tac trough levels were 7+/-0.1 in CYP3A5*3/*3 and 6.6+/-0.2 in CYP3A5*1/*3 ($p<0.05$). CYP3A5*1/*3 patients presented lower levels of dose-adjusted Tac (ng/ml / mg/kg/day, 40+/-2) to achieve target blood concentration of Tac and required higher daily dose per weight (0.2+/-0.04) than CYP3A5*3/*3 patients (82+/-18 and 0.1+/-0.02, $p<0.01$and $<0.01$ respectively). Tac dose and dose-adjusted correlated with daily MPdose CYP3A5*1/*3 (r 0.9, $p<0.05$and r-0.9, $p<0.05$) and in CYP3A5*3/*3 (r 0.9, $p<0.05$and r -0.8, $p<0.05$) patients.

**Conclusions:** CYP3A5 polymorphisms performed before tx could contribute to a better individualization of Tac therapy. Pharmacological interactions between MP and TAC have different mechanism in CYP3A5*3/*3 and CYP3A5*1/*3 patients.

### Abstract# 69
**(O-12)**

**Improved Intra-Individual Variability of the New GFR Equations (CKiD) in Pediatric Renal Transplant Recipients** A. Tsampalieros,[1] P. Geier,[1] N. Lepage,[2] J. Feber.[1] *[1]Pediatrics, Children's Hospital of Eastern Ontario, Ottawa, ON, Canada; [2]Laboratory Medicine, Children's Hospital of Eastern Ontario, Ottawa, ON, Canada.*

**Objectives:** New GFR formulas have recently been developed from the CKiD cohort* using a) height and SCr (mSGFR); b) height, SCr, CysC and serum urea (CKiD GFR). The objective of this study was to compare intra-patient variability of mSGFR and CKiD GFR with the currently used cystatin C GFR (Cys GFR)** among pediatric renal transplant (Tx) patients. We also analyzed the agreement between the Cys GFR and the new CKiD GFR.

**Methods:** A retrospective chart review of 25 Tx children (12 males, median age = 8.9 years) was performed. All available serum urea, SCr and CysC values were collected from the last 3 years of follow-up. Mean and SD GFR (ml/min/1.73 m2) were calculated in each patient individually; CKiD GFR and mSGFR were then compared with Cys GFR using coefficient of variation (CV) and Bland-Altman analysis.

**Results:** A total of 811 samples were collected (32 ± 13 samples per patient). Median Cys GFR (76.4) was significantly higher compared to CKiD GFR (61.4) and mSGFR (59.6), p=0.007. The Cys GFR overestimated the CKiD GFR by a mean of 25% (95% limits of agreement = 0.9-1.6). Median CV of both CKiD GFR (7.6%) and mSGFR (8.9%) were significantly lower than the CysC GFR (10.7%); p=0.02.

**Conclusions:** The new formulas for GFR yield lower GFR than Cys GFR but show a significantly lower intra-individual variation; therefore they may be better suited for longitudinal follow-up of patients post Tx.

*Schwartz GJ et al, J Am Soc Nephrol 2009;20:629

**Filler et al. Pediatr Nephrol 2003; 18:981

### Abstract# 70
**(O-13)**

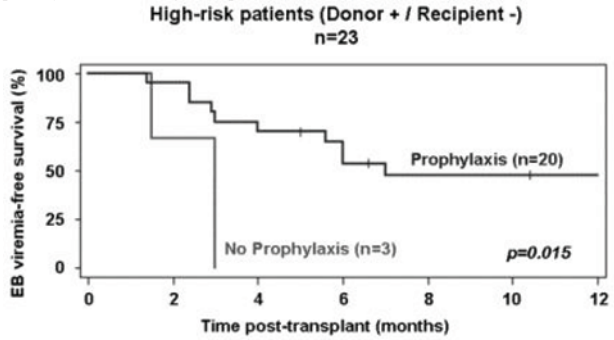
**Effect of CMV Hyperimmunoglobulin and/or (Val-)Ganciclovir on EBV in Pediatric Renal Transplantation: Subgroup Analysis of a Prospective Multicenter Trial** B. Höcker,[1] U. Küsters,[1] S. Böhm,[1] H. Fickenscher,[1] M. Pohl, M. Holder, U. John, M. Kemper, H. Fehrenbach, M. Wigger, M. Schröder, E. Wühl,[1] B. Tönshoff.[1] *[1]Univ., Heidelberg, Germany; [2]German Society of, Pediatric Nephrology, Germany.*

**Objectives:** Data on potential effects of antiviral prophylaxis on the development of EB viremia in pediatric renal transplant patients are lacking.

**Methods:** In a prospective trial (n=114 patients), we performed a subgroup analysis of the influence of chemoprophylaxis with CMV hyperimmunoglobulin and/or (val-)ganciclovir on the occurrence of EB viremia.

**Results:** Of 73 patients with known donor (D)/recipient (R) EBV serostatus, 23 were at high risk (D+/R-) of EB viremia, and 50 at moderate (D+/R+, n=39) or

low risk (D-/R+, n=8; D-/R-, n=3). Prophylaxis with CMV hyperimmunoglobulin and/or (val-)ganciclovir led to a significantly (p=0.015) lower incidence of EBV primary infection in high-risk patients.




Patients on (val-)ganciclovir only, experienced EBV primary infection numerically less often than patients without prophylaxis (5/13 vs. 3/3, p=0.051). A potential additive effect of CMV hyperimmunoglobulin on the development of EB viremia could not be shown so far (3/7 *vs*. 5/13, p=0.651).

**Conclusions:** Antiviral prophylaxis with (val-)ganciclovir significantly reduces the emergence of EBV primary infection in high-risk patients, thereby potentially lowering the risk of EBV-associated PTLD.

## Abstract# 71
**(O-14)**

**Biomarkers of Endothelial Injury and Repair in the Development of Allograft Vasculopathy Following Pediatric Cardiac Transplantation**

M. Stack, K. Daly, M. Seifert, B. Blume, D. Briscoe. *Nephrology and Cardiology, Children's Hospital Boston, Boston, MA, United States.*

**Objectives:** Chronic rejection remains a major challenge in the management of allograft recipients. Cardiac allograft vasculopathy (CAV) is a relatively silent disease with major morbidity. Circulating angiogenic factors may reflect endothelial injury in rejection. Erythropoietin has tissue-protective properties and promotes vascular repair following injury. Epo and angiogenic factors stimulate the mobilization of endothelial progenitor cells (EPCs).

**Methods:** We collected plasma and mononuclear cells from pediatric cardiac transplant recipients (> 2 years of age, >2 years post transplant) at the time of their annual angiogram. We performed analysis on 36 patients, 7 with moderate to severe CAV and 29 controls without disease.

**Results:** By ELISA, we found that mean plasma levels of Epo were slightly lower in patients with moderate to severe CAV (p=0.10). We found higher plasma VEGF levels in patients with CAV vs. without CAV in luminex ($p<0.05$) and elisa ($p<0.05$). We also see trends in changes in endostatin, VEGF-D and thrombospondin-2 in plasma. We examined mRNA profiles of molecules associated with vascular repair in PBMCs in a pilot group (n=13). 44 genes were evaluated by qPCR. Of all genes examined to date, only the expression of PDGF-B, TIE-1 and RANTES demonstrated trends for higher levels in patients with CAV. The mRNA expression of VEGF was not different between patients with/without CAV.

**Conclusions:** These observations demonstrate that plasma levels of angiogenic molecules reflecting endothelial injury and repair may serve as biomarkers of allograft vasculopathy in pediatric recipients of transplants.

**DISCLOSURE:** *Briscoe, D.*: Grant/Research Support, Grant funding for non-related projects in the lab.

## Abstract# 72
**(O-15)**

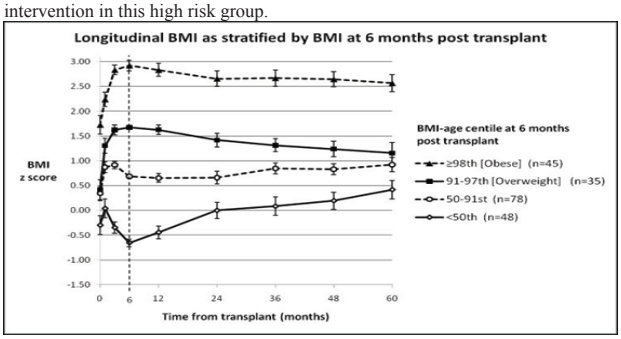
**Longitudinal Change in Body Mass Index (BMI) Following Renal Transplantation in UK Children** Y. Tse, L. Kerecuk, C. Inward, R. Sinha, J. Shield, M. Sinha. *for the, British Association of Paediatric Nephrology, United Kingdom.*

**Objectives:** Children with chronic kidney disease are often malnourished. Post transplant obesity adds to their cardiovascular morbidity. We describe BMI over the first 5 years post transplant in UK children.

**Methods:** Retrospective longitudinal study that included <18 year olds who received transplants 1990-2003. 206 patients with minimum 4-years BMI and eGFR data were included. 153 had data to 5 years. UK clinical definitions of overweight (91-97th) and obesity (≥98th) BMI age percentile were used.

**Results:** Subjects were 71% male, transplanted median age 6.9 years (range 0.9-14.3) and 33% from living donors. Mean BMI z score increased from 0.48 pre transplant to 1.03 at 6 months ($p<0.001$) and remained similar (1.19) at 5 years (p=0.26). When subjects were stratified by their BMI at 6 months (figure), those with BMI <50th centile increased z score +1.07 ($p<0.001$), those overweight and obese decreased z score -0.52 (p=0.012) and -0.35 (0.10). At 5 years, 43% were overweight or obese.

**Conclusions:** Significant numbers of UK children became obese rapidly after transplant and remained so at 5-years. There is a need for early identification and intervention in this high risk group.




| BMI centile | Pre-transplant | 6 months | 5 years |
|---|---|---|---|
| Underweight <3rd | 4.1% | 0.5% | 0% |
| Overweight 91-97th | 15.1% | 17.0% | 15.7% |
| Obese ≥98th | 11.6% | 21.8% | 26.8% |

## Abstract# 73
**(O-16)**

**FGF-23 Predicts Progression of Renal Dysfunction in Pediatric Transplant Recipients** K. Wesseling-Perry,[1] E. Tsai,[1] R. Ettenger,[1] H. Juppner,[2] I.B. Salusky.[1] *[1]Pediatrics, UCLA, Los Angeles, CA, United States; [2]Endocrine Division, Mass General Hospital, Boston, MA, United States.*

**Objectives:** In adult CKD, elevated FGF-23 is linked to progressive renal failure. This study evaluated the relationship between FGF-23 levels, rjxn, and progression of renal disease in pediatric renal transplant pts.

**Methods:** FGF-23, Ca, P, creatinine, Alk P'tase, PTH, $1,25(OH)_2D$, 25(OH)D, and urine Ca, P, and creatinine, were measured in 68 pediatric pts aged 15.3 ± 0.5 yrs with stable renal allograft fxn. Pts were followed for 2 years for >50% deterioration in graft fxn and for bx-proven rjxn. Immunosuppression included CNI and antimetabolites w/ or w/o steroids.

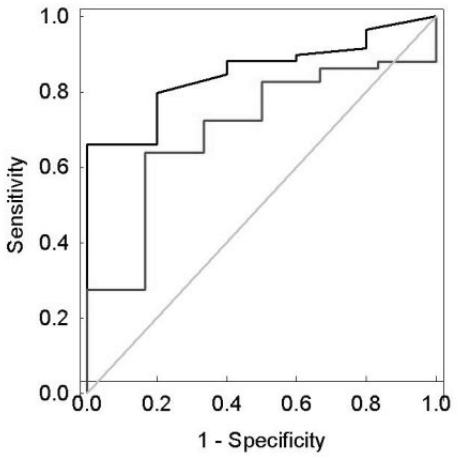
**Results:**

| Biochemical parameter | CKD 1 (n=6) | CKD 2 (n=27) | CKD 3 (n=32) | CKD 4 (n=3) |
|---|---|---|---|---|
| Calcium (mg/dL) | 9.6 ± 0.1 | 9.6 ± 0.1 | 9.6 ± 0.1 | 9.3 ± 0.2 |
| Albumin (g/dL) | 4.2 ± 0.1 | 4.1 ± 0.1 | 4.1 ± 0.1 | 4.5 ± 0.3 |
| Phosphorus (mg/dL) | 4.4 ± 0.4 | 4.4 ± 0.2 | 4.1 ± 0.1 | 4.6 ± 0.2 |
| Alk P'tase (IU/L) (median, IQ range) | 240 (224-290) | 156 (82-223) | 112 (77-190) | 125 (94-136) |
| 1st PTH-IMA (pg/ml) (median, IQ range) | 84 (70-170) | 77 (48-137) | 67 (54-116) | 104 (46-319) |
| 25(OH)D (ng/ml) | 24 ± 4 | 27 ± 2 | 36 ± 2 | 24 ± 6 |
| $1,25(OH)_2D$ (pg/ml) | 57 ± 8 | 54 ± 5 | 41 ± 3 | 24 ± 5 |
| FGF-23 (RU/ml) (median, IQ range) | 40 (8-56) | 96 (43-115)* | 151 (78-214)* | 99 (68-504)* |
| Urine Ca/Cr (mg/mg) | 0.10 ± 0.02 | 0.07 ± 0.02 | 0.05 ± 0.01 | 0.05 ± 0.03 |
| TRP (%) | 91 ± 2 | 88 ± 1 | 82 ± 2‡ | 82 ± 3‡ |

*p<0.05 from nl range, ‡p<0.05 from CKD stages 1 and 2

GFR declined >50% in 6 pts and 12 rjxn episodes occurred.

The AUC for FGF-23 > 110 RU/ml and for baseline GFR as predictors of graft loss were 0.7 and 0.85, respectively (NS).




Red: FGF-23, Black: GFR

More acute rjxn episodes occurred in pts w/ FGF-23 values >110 RU/ml (33% v. 9%, respectively, $p<0.05$).
**Conclusions:** FGF-23 may predict progression and rjxn in renal Tx recipients.
**DISCLOSURE:** *Wesseling-Perry, K.*: Other, Honorarium - Genzyme. *Tsai, E.*: Grant/Research Support, Novartis. *Ettenger, R.*: Grant/Research Support, Novartis. *Salusky, I.B.*: Other, Honoraria - Genzyme and Johnson & Johnson.

## Abstract# 74

**The Integral Role of Renal Allograft Biopsies** S. Adalat, N.J. Sebire, S.D. Marks. *Paediatric Nephrology, Great Ormond Street Hospital for Children, London, England, United Kingdom.*
**Objectives:** To emphasise the importance of performing renal allograft biopsies.
**Methods:** A 12 year old female with ESRF secondary to nephronophthisis had a 3-Ag matched deceased donor renal transplant with good primary function using triple immunosuppression with corticosteroids, azathioprine and tacrolimus. However on the 4th post-operative day, she suffered a gastro-intestinal haemorrhage requiring blood transfusion for hypovolaemia. She underwent rapid weaning of her steroids but continued to have intermittent GI bleeds.10 days later, she developed fever, graft tenderness and renal allograft dysfunction. Renal ultrasound, blood and urine cultures were unremarkable. Transplant biopsy was performed as acute rejection was suspected.
**Results:** Histopathology revealed marked intimal large vessel thickening suggesting chronic vasculopathy with ATN and ischaemic tubular changes indicative of recent hypovolaemia. Neutrophilic infiltrate within many tubules and patchy CD3 infiltration suggested possible infection. Large nuclei and vacuolation suggested possible viral infection without viral inclusions.There were no features of acute rejection with negative C4d staining. She was treated with IV steroids and antibiotics. Her tacrolimus dose was increased due to low trough levels but she developed CMV and EBV viraemia requiring treatment with oral valganciclovir. Her symptoms and renal function improved with chronic vascular changes only on repeat biopsy at 6 weeks.
**Conclusions:** We highlight the importance of performing transplant renal biopsies and careful co-ordinated clinicopathological correlation. Multiple histological abnormalities may be found to provide clues to potential aetiologies of allograft dysfunction.

## Abstract# 75

**Cytomegalovirus (CMV)-Specific T Cells as a Parameter for Individual CMV-Specific Immune Response after Pediatric Kidney Transplantation (Tx)** T. Ahlenstiel,[1] U. Sester,[2] M. Sester,[2] L. Pape.[1] *[1]Pediatric Nephrology, Medical School of Hannover, Hannover, Germany; [2]Nephrology, University of the Saarland, Homburg (Saar), Germany.*
**Objectives:** After Tx immunosuppression leads to impaired cellular immune defense resulting in increased risk of CMV-disease. Post-Tx follow-up of CMV-specific T cells (CMV-s T cells) may serve as a predictive marker for the necessity of antiviral therapy.
**Methods:** Within a prospective study we monitored CMV-s T cells in 36 children during first year after kidney Tx. Antiviral therapy was administered in case of significant CMV-DNA-detection. CMV-s CD4-/CD8-T cells were determined by flow cytometry.
**Results:** Pre-Tx prevalence of CMV-s CD4-T cells (25%) correlated with CMV-seropositivity. After Tx 3 CMV-seronegative children with seropositive donors showed a primary CMV-infection. Two of them were characterized by initial DNA-boost: CMV-s CD4-T cells secondarily increased simultaneously with decrease of CMV-DNA after start of antiviral therapy. In case of asymptomatic primary infection with initial boost of CMV-s CD4-T cells (n=1), we found CMV-seroconversion without detection of DNA. Two children with symptomatic CMV-reactivation showed transient disappearance of CMV-s CD4-T cells (<3/µl) combined with increase of CMV-DNA, whereas in case of asymptomatic reactivations (n=2), low CMV-DNA-level vanished in presence of sufficient level of CMV-s CD4-T cells (>3/µl). There was no significant detection of CMV-s CD8-T cells.
**Conclusions:** Sufficient levels of CMV-s CD4-T cells (>3/µl) protect against CMV-disease. Serving as prognostic marker for individual susceptibility to CMV-disease, CMV-s CD4-T cells may improve post-Tx management and optimize antiviral therapy.

## Abstract# 76

**Polyoma BK-Virus (BKV)-Specific T Cells as a Prognostic Marker for Polyomavirus-Associated Nephropathy (PVAN) after Pediatric Kidney Transplantation** T. Ahlenstiel,[1] U. Sester,[2] J.H.H. Ehrich,[1] L. Pape.[1] *[1]Pediatric Nephrology, Medical School of Hannover, Hannover, Germany; [2]Neprhology, University of the Saarland, Homburg (Saar), Germany.*
**Objectives:** After transplantation (Tx) immunosuppression causes impaired cellular immune defense resulting in increased risk of PVAN. Prognostic markers for the outcome of PVAN are missing. Follow-up of BKV-specific T cells (BKV-s T cells) may serve as a predictive marker.
**Methods:** After kidney Tx BKV-s T cells were examined in 10 children with detection of BKV-DNA in blood at different points in time. Leucocytes were stimulated with BKV-antigen (VP1 and large T); BKV-s CD4-/CD8-T cells were determined by flow cytometry.
**Results:** The majority of our study group (8/10) showed -at least temporarily- BKV-s CD4 T cells, whereas only 4 patients had BKV-s CD8 T cells. In case of persistent BKV-DNA (>2000 cop/ml) only a small number of BKV-s CD4 T cells (≤600/ml) were detectable. In contrast patients with high levels of BKV-s CD4 T cells (>1500/ml) were characterized by asymptomatic BKV-infections with rapid disappearance of BKV-DNA. If BKV-DNA was not longer detected for several months, BKV-s CD4 T cells were only found in small quantities (≤1000/ml) or completely vanished. A temporary detection of BKV-s CD8 T cells was only found in patients without persistent BKV-DNA.
**Conclusions:** In case of low levels of BKV-s CD4 T cells patients are not able to overcome BKV-infection, whereas a sufficient number of BKV-s CD4 T cells was associated with successful defense. Serving as prognostic marker for individual BKV-specific defense, levels of BKV-s T cells may represent the risk of BKV-associated complications and optimize individual timing of therapeutic interventions.

## Abstract# 77

**Posterior Reversible Encephalopathy Related to Tacrolimus in a Kidney Transplanted Adolescent** E. Comak,[1] M. Koyun,[1] A.U. Gökceoglu,[1] C.S. Dogan,[1] ö. Duman,[2] K. Karaali,[3] S. Akman.[1] *[1]Pediatric Nephrology, Akdeniz University, Antalya, Turkey; [2]Pediatric Neurology, Akdeniz University, Antalya, Turkey; [3]Radiology, Akdeniz University, Antalya, Turkey.*
**Objectives:** Posterior reversible encephalopathy syndrome (PRES) may develop in patients with hypertension, renal insufficiency, hypomagnesemia, hypocholesterolemia or due to immunosuppressive drugs such as tacrolimus.
**Methods:** We report a renal transplant recipient who developed PRES.
**Results:** A 15 year old boy, who had underwent renal transplantation three weeks previously because of Prune Belly syndrome and was tacrolimus, mycophenolat mofetil and prednisolone therapy, presented with headache and mental status changes. He was afebrile, blood pressure was 123/70 mmHg; systemic examination findings were normal. He suffered a generalized tonic-clonic seizure at the emergency room and complained of significant visual disturbance on recovery. Serum creatinine was 1.2 mg/dl and tacrolimus trough level was 15 ng/ml. Serum electrolytes, glucose, liver enzymes, calcium, phosphate, albumin, magnesium, and acid-base status were all normal. Lumbar puncture findings were normal. Cranial magnetic resonance imaging was compatible with PRES (bilateral low-density areas on cortical and subcortical regions of parieto-occipital lobes). It was thought to be due to tacrolimus therapy; so it was switched to cyclosporine, which improved the clinical situation. The majority of lesions on magnetic resonance imaging disappeared within 5 weeks.
**Conclusions:** Transplantation teams should be aware of this uncommon complication of immunosuppression, which may be associated with significant morbidity and mortality if it is not expeditiously recognized.

## Abstract# 78

**BK Virus Nephropathy in Pediatric Renal Transplantation: Tacrolimus Versus Cyclosporine** M. Koyun,[1] A.U. Gökceoglu,[1] C.S. Dogan,[1] E. Comak,[1] D. Mutlu,[2] A. Gürkan,[3] S. Akman.[1] *[1]Pediatric Nephrology, Akdeniz University, Antalya, Turkey; [2]Microbiology, Akdeniz University, Antalya, Turkey; [3]Akdeniz University, Antalya, Turkey.*

**Objectives:** The aim of this study was to analyze the prevalance and risk factors for BK viruria, viremia and nephropathy in children who underwent renal transplantation.

**Methods:** Urine and blood were collected to study BKV by polymerase chain reaction method biweekly for the first three months, monthly between 3-6 months and trimonthly thereafter.

**Results:** 115 children were enrolled in the study. Mean age at the time of transplantation was 11.7±3.9 years; median follow-up period was 18 months. 76 children were receiving tacrolimus, 24 cyclosporine and two sirolimus; 13 patients received both tacrolimus and cyclosporine at different periods because of adverse effects of either drug. Also, all children used mycophenolate mofetil and glucocorticoids. BK viremia was observed in 42 children (36.5%); it was transient in half. Viremia of patients receiving tacrolimus and cyclosporine were similar. BK viruria occurred in 63 of all patients (54.8%). Patients using cyclosporine had more viruria than that using tacrolimus (79.2% vs 43.4%, p=0.002); however, sustained viruria was similar between the two groups (29.2% vs 24.3%, p>0.05). Definite nephropathy (biopsy-proven) was observed in 4 (3.5%). Although not statistically significant, nephropathy was more common in patients receiving tacrolimus than cyclosporine (3.9% vs 0%).

**Conclusions:** We conclude that, though transient viruria is more common in children receiving cyclosporine, BKV nephropathy seems to be more frequent in patients receiving tacrolimus.

## Abstract# 79

**Latin American Registry of Pediatric Renal Transplantation 2004-2009** Latin American Pediatric Nephrology Association ALANEPE. *Brazil, Argentina, Chile, Mexico, Venezuela, Paraguay, Guatemala, Ecuador; Honduras, Cuba, Costa Rica, Colombia, Peru.*

**Objectives:** Latin American Pediatric Nephrology Association reports the first kidney transplant registry 2004-2009.

**Methods:** Between 30-75% of the LA recipients under 21 y-o were included. Demographic characteristics,immunosuppression, patient and graft survival were analyzed. 20 countries were invited to participate,14 performed renal transplant,11 had universal financial support from their governments.

**Results:** Centers:Brazil(9),Argentina(4),Chile(4),Venezuela (4),Mexico(2)1 in:Cuba,Colombia,Costa Rica,Nicaragua,Guatemala,Ecuador,Honduras, Paraguay, Peru.The registry included 2,600 patients, 301 per year, 55% male. Mean follow-up was 23.4±17 months.Mean age 11.7±4.3 ys(1-21),11% under 5 ys.Etiology: uropathy/reflux nephropathy 27%,glomerulopathies 24% (12% FSGS),hypo/dysplasia(11%), vascular (6%), congenital/hereditary (5%), unknown (19%). DD 54.3%, in Venezuela and Chile(70%), Mexico 30%, Brazil and Argentina 50%. Anti-IL2RAb/71%), ATG/TIMO(13%),noninduction (14%).Maintenance therapy was TAC 64%,CsA 32%,MMF 54%,MPS 20%,noTORi 96%,steroids 90%, withdrawal/steroid avoidance 10%.
Loss of graft 155/1,458(11%), death with functioning graft(3.4%),vascular thrombosis(2.8%),AR (2.8%), recurrence (1%).Forty-eight patients died (3.3%); infection 23 (2.1%).Global patient survival rate at 1,3,5 years was 97%,96%,96%. Graft survival rate at 1,3,5 years for living related donors (LRD) 96%,93%,89%; DD 92%,86,76% respectively. Patient and graft survival rates were higher in LRD (p<0.008;p<0.001).

**Conclusions:** A collaborative study in pediatric renal transplant has started; on-line registry is in progress to make information available shared between the centers and the rest of the world.

## Abstract# 80

**Growth Hormone (GH) Administration Improves Longitudinal Growth in Rats with Growth Retardation Induced by Rapamycin** O. Alvarez-Garcia, E. García-Lopez, V. Loredo, H. Gil-Peña, J. Rodríguez, F.A. Ordoñez, F. Santos. *Hospital Universitario Central de Asturias & University of Oviedo, Oviedo, Spain.*

**Objectives:** To find out the effect of GH therapy on longitudinal growth of rapamycin treated rats.

**Methods:** Four groups (N=10) of young rats were studied: treated with rapamycin (R), treated with rapamycin and GH (RGH), control pair fed with R (C), and control pair fed with R and treated with GH (CGH). Animals received 1 mg/kg of rapamycin or vehicle during 14 days. Last week of the protocol, animals received 3.3 mg/kg of GH or vehicle and longitudinal growth was measured. Tibial growth plates were embedded in methil-methacrilate for morphological study, measurement of osseous front advance as an index of longitudinal growth rate, and detection of BrdU incorporation to assess chondrocyte proliferation.

**Results:** GH significantly increased (X±SD) longitudinal growth rate (C:164±7; CGH:182±1;R:82±6;RGH:102±5 µm/day) and length gain (C:2.4±0.2;CGH:2.8 ±0.2;R:2.0±0.2;RGH:2.5±0.1 cm) in CGH and RGH animals. Growth cartilage ( C:396±27;CGH:392±19;R:471±10;RGH:579±40 µm) and its hypertrophic zone ( C:232±21;CGH:199±20;R:271±11;RGH:345±28 µm) were greater in R animals and this expansion was more marked in RGH group. An increase of the height of the terminal chondrocyte was found in both groups treated with GH (C:25±0;CG H:26±0;R:25±0;RGH:27±1µm) whereas chondrocyte proliferation was increased only in CGH animals (C:32.3±3.6;CGH:44.1±2.6;R:28.5±2.5; RGH:30.7±1.9 BrdU labeled cells/100 cells).

**Conclusions:** GH administration improves longitudinal growth in young rats with rapamycin-induced growth retardation. This effect is likely related to an increase in chondrocyte hypertrophy within the epiphyseal growth plate.

## Abstract# 81

**Growth Following Renal Transplantation in Children and Adolescents with End-Stage Renal Disease** M.C. Andrade, C. Mangia, J.T. Abreu Carvalhaes. *Pediatrics, UNIFESP-EPM, São Paulo, São Paulo, Brazil.*

**Objectives: Objective:** To evaluate growth in children and adolescents after kidney transplantation in a high middle income country.

**Methods:** We evaluated children and adolescents consecutively during a period of 5 years after kidney transplantation. Demographic data, age of transplantation, gender, donor source, allograft function, kind of treatment before the kidney transplantation, etiology of chronic kidney disease (CKD) and anthropometric data.

**Results: Results:** The analysis of 48 recipients was stratified by age group (< 6 years(ys), 6-12 ys and 12-18 ys). The overall demographic analysis were: age of transplantation: 10.6±3.4, weight: 29±12 kg, height: 130.5± 21.4, height Z score (HAZ): after transplant – 1.85±1.34; HAZ 6 months: -1.8±1.2; HAZ 1year: -1.7±1.2; HAZ 2 years: -1.7±1.1; HAZ 2 years: -1.7±1.2; HAZ 3 years: - 1.7±1.1; HAZ: -1.7±1.1; HAZ: -1.7±1.2.There were differences between standardized height Z score and groups (p<0.001), but not in within groups. There were differences between gender (p<0.00) without catch-up, final HAZ and type of donor (life vs death; p<0.05) and HAZ 5y vs length of dialysis ( LOD >13months; p=0.04). There were not differences between allograft function, etiology of CKD, kind of treatment before kidney transplantation. The factors that influenced in the growth at 5 years after transplantation were: HAZ 4 years, height at 4 and 5 years, length of dialysis (< 13 months), weight at 2 years and gender (multivariate linear regression).

**Conclusions: Conclusion:** There weren't improvements in height Z score during the period of study. The kidney transplantation didn't have positive impact on final adolescent height.

## Abstract# 82

**Postural Alterations in Children and Adolescents after Kidney Transplantation** M.C. Andrade,[1] A.C. Gama,[2] J.T. Carvalhaes,[1] J.O. Medina,[2] D. Diniz.[2] *[1]Pediatrics, UNIFESP-EPM, Sao Paulo, Sao Paulo, Brazil; [2]Neprhology, UNIFESP-EPM, Sao Paulo, Sao Paulo, Brazil.*

**Objectives:** To evaluate presence and kind of muscle-skeletal alterations in children and adolescents after kidney transplantation.

**Methods:** Evaluation of 41 out patients in our post-renal transplantaion ambulatory from April to November, 2007, determining muscular strength, orthopedic deviations and weight and height measurements.

**Data** : Age, creatinin clearance, chronical kidney disease (CKD) ethiology, time and kind of previous treatment and time of transplantation were taken from charts.

**Results:** Patients' age varied from 6 to 21 years, mean 13 years (IC95%=11.9-14.39 years), without any significant difference in genders. Mean z-score values for W/A, H/A and BMI indicators were -1.22, -2.29 and 2.23, respectively.
All patients presented alterations in the head and scapula alignment, and mean in number of orthopedic/patient alterations was 9.68. Muscular strength was considered good only for 5 (12.20%) patients.
Relating to ethiology, most frequent diagnostic was uropathy (41.5%). Before kidney transplantation 56.1% of patients were under hemodialysis, 36.6% on peritoneal dialysis and 7.3% on conservative treatment. About. 24% of patients who were submitted to kidney transplantation were less than one year old, 29% between 1 and 2 years and 47% between 3 and 7 years.

**Conclusions:** Muscle-skeletal alterations were observed in all patients, reinforcing the need of intervention and early treatment.

## Abstract# 83

**Clinical Presentation of Septic Shock Induced by Influenza A (H1N1) in Children Immunocompromised Transplant Patients** M.C. Andrade,[1] C. Mangia, J.T. Carvalhaes, J.O. Medina. *[1]Pediatrics, UNIFESP-EPM, Sao Paulo, Sao Paulo, Brazil; [2]Nephrology, UNIFESP-EPM, Sao Paulo, Sao Paulo, Brazil.*

**Objectives:** Case of immunocompromised patient due to renal transplant who developed septic shock due to infection by H1N1 reporting.

**Methods:** patient data were taken from charts.

**Results: Case Report**: Child with cystinosis, underwent a kidney transplant in March 2009. On July he was seen at hospital with cough and fever. Initial diagnosis was bacterial pneumonia, and it was introduced treatment with amoxycillin. Eight days after he returned with fever, cough, dyspnea and significant impairment of general condition. Chest X-rays showed interstitial and broncho-alveolar bilateral diffuse infiltrates. Large spectrum antibiotis therapy was introduced. After 3 hours there was worsening of respiratory failure and he was transferred to the ICU with respiratory isolation and ventilatory support.He progressed to septic shock and multiple organ systems failure at subsequent 24 hours. Sample of nasopharyngeal secretion collected (RT-PCR) swwed infection with influenza H1N1, and oseltamivir therapy was introduced. At 4th day of admission he showed improvement in hemodynamic and ventilatory parameters.

**Conclusions: Comments:** H1N1 infection in an immunocompromised patient was presented quickly and with rapid evolution to septic shock and multiple organ systems dysfunction. Wide information to parents and caregivers about disease characteristics, early recognition of septic viral shock, adequate transport to appropriate centers of reference, and hospital rapid and aggressive goal-directed proceedings by a specialized team are recommended for the care of these patients.

## Abstract# 84

**BK Viremia and Nephropathy: Significance of Prospective Viral Screening in Pediatric (Ped) Renal Transplant (RTx) Recipients (Rec)** E.I. Anyaegbu, S.I. Al-Akash. *Kidney Center, Driscoll Children's Hospital, Corpus Christi, TX, United States.*

**Objectives:** BK nephropathy (BKN) is an important cause of allograft dysfunction and accounts for up to 50% of graft loss. The objective was to determine the significance of prospective BK viral screening in Ped RTx Rec.

**Methods:** A retrospective study using multivariate analysis to assess risk factors associated with BK viremia, and BKN. 27 Ped RTx Rec were prospectively screened for BK virus in urine and plasma or by quantitative PCR at regular intervals.

**Results:** Mean follow up period was 15.1 (2-25) months. Mean Rec age was 12.3 (1.9-9.8) years, 58% were male, and 85% received deceased-donor Tx. 96% were primary Tx. Induction therapy was a 4-day Methylprednisone taper and either Daclizumab (56%) or r-Thymoglobulin (Thymo) (44%). Maintenance immunosuppression (IS) consisted of Tacrolimus (TAC)/ Mycophenolic acid (MPA) in 88%, TAC/ Prednisone (P) in 8% and TAC/ MPA/ P in 4%. Screening began 2 weeks post Tx and monthly afterwards. Frequency of testing was increased once viremia developed or after treatment of acute rejection (AR). 16 (59%) pts developed BK viruria, 7 (26%) viremia and 1 (4%) was found to have BKN on surveillance biopsy. Reduction in IS was the first line therapy in pts who developed viremia.

**Conclusions:** Thymo use and higher TAC trough level were significantly associated with the development of viremia (p= 0.0158 and 0.0269 respectively). None of the pts developed AR following IS reduction. No graft loss due to BKN occurred in spite of 26% having viremia. Early screening and reduction in IS appears to be associated with a reduced incidence of BKN. The optimal timing, frequency of testing, and the most appropriate therapy need further study.

## Abstract# 85

**BK Viremia and Nephropathy: Response to Treatment with Intravenous Immunoglobulin (IVIg) in Pediatric (Ped) Renal Transplant Recipients (RTx)** E.I. Anyaegbu,[1] S.I. Al-Akash.[2] *[1]Driscoll Children's Hospital, Corpus Christi, TX, United States; [2]Kidney Center, Driscoll Children'S Hospital, Corpus Christi, TX, United States.*

**Objectives:** BK virus nephropathy (BKVN) is a significant cause of allograft dysfunction and loss in Ped RTx Recs. Therapeutic options are limited. Pre-emptive reduction in immunosuppression (IS) has been found to reduce viral load and improve graft function. This report demonstrates viral clearance and histological resolution of BKVN with IVIg therapy.

**Methods:** Our BK surveillance protocol involves screening Ped RTx Recs for BK by whole blood quantitative PCR at regular interval. The frequency of testing is increased once BK viremia (BKV) is detected or following treatment for acute rejection.

Gradual reduction of IS was instituted after detection of BKV until clearance was achieved. The indication for IVIg therapy was persistent viremia or development of BKVN.

**Results:** 27 Ped RTx Recs were screened for BKV. Mean follow up was 15.11 ± 8.17 months. 22% developed BKV 33- 631 (149.17 ± 236.62) days post transplant. PCR ranged from 400-5200 (2049 ± 1747) copies/ml.

IVIg (2 gm/kg) therapy was given in 3 pts (11%) due to persistent BKV. Another pt (4%), diagnosed with BKVN on protocol biopsy, received IVIg with resolution confirmed by biopsy 27 days later, with excellent graft function at last follow up Graft function remained stable in all our pts. Nephrotoxic anti-viral therapy was not required for our pts.

**Conclusions:** Our surveillance protocol with IS reduction might account for our low incidence of BKVN. IVIg therapy seems to be effective for treatment of persistent BKV and BKVN.

IVIg therapy should be considered in pts with persistent BKV after IS reduction and in pts with BKVN.

## Abstract# 86

**Successful Kidney Transplantation in 4 Patients with Factor H Deficiency-HUS** G. Ardissino, S. Testa, N. Borsa-Ghiringhelli, M. Belingheri, F. Paglialonga, P. Castorina, A. Edefonti. *Center for HUS Control, Fondazione IRCCS Ca' Granda - Osp Maggiore Policlinico, Milan, Italy.*

**Objectives:** Factor H deficiency hemolitic uremic syndrome (FHD-HUS) has a very high risk of recurrence after kidney transplantation (KTx). Refreining from KTx, combined liver-KTx or KTx associated to lifelong plasmaexchange (PLE) have been proposed for patients in ESRD due to FHD-HUS with contrasting results. Herein we describe our protocol for KTx in FHD-HUS which proved to be efficacious in all the patients we have treated so far.

**Methods:** Four patients (age range 5-36 yrs) with CKD-V due to documented FHD-HUS underwent KTx following one PLE before KTx and several maintenance PLEs and fresh frozen plasma infusions according to the protocol shown in the table:

| Time | PLE | Plasma Infus. | Frequency |
|---|---|---|---|
| Pre-Tx | 75ml/kg | 1000 ml | Once |
| POD 1-5 | 75ml/kg | no | Daily |
| POD 6-7 | 50ml/kg | no | Daily |
| POD 8-17 | 50ml/kg | 25ml/kg | Alternate day |
| POD 18 to 26 | 50ml/kg | no | Every other day |
| POD 27-41 | 50ml/kg | no | Every 5 days |
| POD 42 to 180 | STOP | 20ml/kg | Weekly |

Immunosuppressive protocol included basiliximab, prograf or cyclosporine and prednisone. We emphatize that all patients were addressed to KTx with a significant fluid overload (as much as 3% above optimal body weight) obtained with plasma infusion.

**Results:** Over a cumulative observation period of 61 mos., we only observed 2 recurrences (in 2 different patients) which were managed with PLE and Eculizumab with immediate recovery of the recurrence.

**Conclusions:** Our therapeutic approach to TKx in FHD-HUS represents a less aggressive solution in the meantime that Factor H becomes available for maintenance treatment.

## Abstract# 87

**The Meaning of Anemia in Children during the First Year of Kidney Transplantation (KTx)** G. Ariceta,[1,2] R.A. Cohn,[1] M. Cuevas,[3] C.B. Langman.[1] *[1]Kidney Diseases, Children's Memorial Hospital, Northwestern University, Chicago, IL, United States; [2]Nefrologia Pediatrica, Hospital Cruces.Basque Country University, Baracaldo-Bilbao, Spain; [3]Nefrologia Pediatrica, Pontificia Universidad Catolica Chile, Santiago, Chile.*

**Objectives:** To assess the prevalence of post transplant anemia in pediatric KTx recipients, and the relationship between anemia, graft function & hospitalizations.

**Methods:** Retrospective cohort study. 85 consecutive KTx in 2004-2008. Data analyzed at 0,1,3,6,&12 months (m) after KTx. Anemia definition: Hb <5$^{th}$ percentile or erythrocyte-stimulating-agents (ESA) treatment. Data reported are mean±SD.

**Results:** 85 patients (55 boys), 13 ± 5.4y with KTx (80, 1$^{st}$ graft) studied. Preemptive KTx 32 (38%). Living donor 37(43%). Hb at 0-1-3-6-12 m: 12.8±1.8; 11.2±1.3; 11.9±1.3; 12.7±1.5; 12.7±1.7 g/dL. Anemia observed in 47%-68%-67%-52%-51%; & ESA prescribed in 42%-54%-52%-55% of them at each study-point. Likelihood of anemia/ESA-use: no differences found based on pre-emptive vs prior-dialysis KTx, or living vs deceased donor. More boys were anemic at baseline ($p<0.005$) & 1m after KTx ($p<0.008$). Graft function, judged by eGFR did not differ between patients with/without anemia at 1 & 3m, but eGFR was reduced in anemic patients at 6 ($p<0.002$) & 12m ($p<0.012$) post KTx. Anemic patients were admitted more often between 3 to 6 ($p<0.001$) & 6 to 12m ($p<0.0001$). From 6m post-KTx, anemic patients had a 6.7 fold [CI: 2.01-22.03] greater risk of hospitalization compared to non-anemic patients.

**Conclusions:** Post KTx anemia was frequent in a pediatric cohort within the 1$^{st}$ year. Anemia beyond the 3$^{rd}$ month after KTx was associated with longer-term impaired graft function & increased risk of hospitalization.

## Abstract# 88

**Rhodococcus equi: An Underestimated Cause of Necrotizing Pneumonia after Renal Transplant** S. Azib,[1] V. Baudouin,[1] P. Mariani,[2] A. Maisin,[1] V. Houdouin.[3] *[1]Pediatric Nephrology, Hopital Robert Debré, Paris, France; [2]Microbiology, Hopital Robert Debré, Paris, France; [3]Pneumolgy, Hopital Robert Debré, Paris, France.*

**Objectives:** Rhodococcus Equi is a mycobacterium-like organism witch is normally pathogen in foals and horses. Human infection is rare but increasing number of cases are reported in immunocompromised patients such as transplant recipients with fatal outcome up to 30%.

**Methods:** We report the case of a bacteriemic lung abcess caused by Rhodococcus Equi in a 20-years-old renal transplant patient occurring 9 years after his graft.

**Results:** Patient was treated 3 months before for a graft rejection leading to an increase in the immunosuppressive regimen. The patient presented with a 4 week history of weight loss and chest pain. Chest x-ray showed a rounded consolidation in the left lower lumb. Computerized tomography (CT) of the chest demonstrated necrotizing pneumoniae in the left lower lumb. Quantitative culture of bronchoalveolar lavage showed $10^6$/ml of Rhodococcus Equi with bacterial blood culture positive. No animal contact was found. Patient received a prolonged antibiotic therapy with rifampicine and minocycline.

**Conclusions:** Diagnosis of Rhodococcus Equi infection is unusual. The antibiotic treatment is difficult as no standard treatment has been established. Therapy should be prolonged, at least during 4 months. Relapses are common and could occur at the initial site or at distant locations. In case of incomplete resolution, surgery intervention could be required.

## Abstract# 89

**Conversion to Sirolimus in a Pediatric Renal Transplant Population** M. Azocar, A. Delucchi, A.M. Lillo, J.L. Guerrero, F. Cano. *Pediatrics, Hospital Luis Calvo Mackenna/Universidad de Chile, Santiago, Chile.*

**Objectives:** To present our experience with sirolimus in a pediatric population after conversion from a calcineurin-based inmmunosuppression.

**Methods:** Retrospective study in transplant recipients at the Luis Calvo Mackenna Children's Hospital. Fourteen patients were included, 7 males. Age at transplant 6±3.73 years. Immnunosupresion at the time of intervention: Ciclosporin-Azathioprin-steroids: 5/14, tacrolimus (FK)-mycophenolate (MMF)-steroids:4/14, FK-MMF: 5/14. Patients were converted to sirolimus because biopsy findings of drug toxicity or chronic allograft nephropathy.

**Results:** Mean glomerular filtration rate (GFR, Schwartz formula) increased from 47,7 ml/min to 54,3 ml/min at month 6, a non-significant change ($p > 0.05$). In those converted with a GFR>40 ml/min, clearence was found to be better than the group converted < 40 ml/min: 59,3 at month 1, 58.6 at month 3 and 60 at month 6, vs 39.8 at month 1, 33 at month 3 and 30 at month 6 ($p<0.05$ for each month). Total cholesterol (TC) was 189 mg/dl prior starting sirolimus and increased to 196 at month 6 ($p>0.05$,n.s). Mean serum triglyceride (TG) prior to the change in medication was 124 mg/dl, with a non-significant change at month 6. Mean proteinuria/creatininuria ratio (P/C) prior to sirolimus conversion was 0.83, increasing to 1.03 at month 6 ($p>0.05$, n.s). No rejection was observed. Other adverse effects were not observed.

**Conclusions:** Our experience with sirolimus shows that most of the patients tolerate the drug well and potentially will benefit from long-term avoidance of calcineurin inhibitors. GFR level at the time of change should be carefully considered.

## Abstract# 90

**Using mTOR Inhibitor in Paediatric Renal Transplantation** F.D. Bandin,[1] A. Garnier,[1] V. Baudouin,[2] A. Dallocchio,[1] K. Brochard,[1] M. Cailliez,[3] M. Dehennault,[4] A. Masin,[2] B. Ranchin,[5] G. Roussey,[6] S. Decramer.[1] *[1]Pædiatric Nephrology, Children's Hospital, Centre de Référence des Maladies Rénales Rares, Toulouse, France, Metropolitan; [2]Pædiatric Nephrology, Robert Debré Hospital, Paris, France, Metropolitan; [3]Pædiatric Nephrology, Children's Hospital, Marseille, France, Metropolitan; [4]Pædiatric Nephrology, Jeanne de Flandre Hospital, Lille, France, Metropolitan; [5]Pædiatric Nephrology, Children's Hospital, Centre de Référence des Maladies Rénales Rares, Lyon, France, Metropolitan; [6]Pædiatric Nephrology, Children's Hospital, Nantes, France, Metropolitan.*

**Objectives:** In paediatric renal transplantation, the mTOR inhibitors are seldom used. We evaluated the possible benefits of mTOR inhibitors switched to prevent or treat calcineurin inhibitors nephrotoxicity.

**Methods:** We retrospective studied 20 renal transplant patients switched to mTOR inhibitors. We collected urine protein excretion rate and estimated glomerular filtration rate (EGFR) before and after swtich.

**Results:** Immunosuppressive protocol most frequently used: cyclosporine, mofetil mycophenolate and steroids.

The main reason for switching was chronic allograft nephropathy.

For nine patients we analyzed EGFR and Proteinuria before and after switch with a follow-up more than 12 months: 7 showed stabilization or improvement of EGFR.

Treatment was stopped in two patients because proteinuria closes to nephrotic rate.

For the other 11 patients : seven needed to stop treatment due to secondary effects.

**Conclusions:** mTor inhibitor treatment seems to be associated with a GFR stabilization. Nevertheless, these results must be confirmed and validated by prospective studies in bigger cohorts.

## Abstract# 91

**Association of Hearing Loss and Calcineurin Inhibitors in Pediatric Renal Transplant Recipients** K. Gulleroglu,[1] E. Baskin,[1] E. Aydin,[2] U. Bayrakci,[1] S. Sevmis,[3] E. Melek,[1] L. Ozluoglu,[2] H. Karakayali,[3] M. Haberal.[3] *[1]Pediatric Nephrology, Baskent University, Ankara, Turkey; [2]Otorhinolaryngology, Baskent University, Ankara, Turkey; [3]General Surgery, Baskent University, Ankara, Turkey.*

**Objectives:** The knowledge about hearing impairment after renal transplantation is limited. Hearing loss in pediatric renal transplant group has not been evaluated yet. Calcineurin inhibitors may have potential ototoxic side effects.

**Methods:** We prospectively investigated the relationship between hearing impairment and calcineurin inhibitors in 27 pediatric renal transplant recipients. All patients underwent audiological assessment by means of pure-tone, impedance audiometry and otoacoustic emission tests.

**Results:** Mean age of recipients was 14,05±4,11 years old at the time of transplantation. Mean posttransplant follow-up was 2,48±1,87 years. 22 patients received cyclosporine A (CycA), 17 patients received tacrolimus and 12 patients was switched from CycA to tacrolimus. Sensorineural hearing loss was found in 17 patients (62,9%) and speech discrimination was decreased in 8 (29,6%). There was a marked hearing impairment for the higher frequencies between 4 000 and 8 000 Hz. CycA levels were significantly higher in patients with hearing impairment (mean: 1016,2±558,9 ng/ml) than the group without hearing impairment (mean:807,7±46,8 ng/ml) ($p=0,005$). CycA levels were found to be higher in patients with low speech discrimination ($p=0,00$). This kind of relationship was not valid for tacrolimus levels.

**Conclusions:** We conclude that dose-dependent CycA toxicity might be one of the responsible agents which cause hearing impairment after renal transplantation.

## Abstract# 92

**Tuberculosis in a Renal Transplant Recipient after Rituximab Treatment** E. Baskin,[1] K.S. Gulleroglu,[1] U.S. Bayrakci,[1] F. Eyuboglu,[2] H. Arslan,[3] S. Sevmis,[4] H. Karakayali,[4] M. Haberal.[4] *[1]Pediatric Nephrology, Baskent University, Ankara, Turkey; [2]Pulmonary Diseases, Baskent University, Ankara, Turkey; [3]Infectious Diseases, Baskent University, Ankara, Turkey; [4]General Surgery, Baskent University, Ankara, Turkey.*

**Objectives:** Tuberculosis is still one of the most serious infections among renal transplant recipients. It has been reported that rituximab treatment not increase the risk of tuberculosis, but this situation is not exactly clear. We reported a renal transplant recipient that developed tuberculosis after using rituximab for the treatment of disease recurrence.

**Results:** A 19-year-old male patient diagnosed for FSGS at the age of 12. At the age of 14 he had his 1st kidney transplant but recurrence of FSGS occurred and few months later he had to re-start dialysis. Two years later he got his 2nd kidney transplant. Early onset FSGS recurrence was controlled by rituximab treatment and pre-and posttransplant plasmapheresis after the 2nd transplatation. After seven months of taking the last dose of rituximab, on the 19th months of transplantation, he was diagnosed for pneumonia. In the beginnig it was thought that the rituximab treatment may have been one of the factors that may have faciliated the development of infection and treatment's failure. In spite of antibiotherapy bronchoscopy was applied at the end of constancy of his fever and coughing. *Mycobacterium tuberculosis* produced in the culture of bronchoalveolar lavage sample. Clinical signs were resolved after the antituberculoid treatment.

**Conclusions:** In spite of as rituximab accepted safe for tuberculosis infection, it must be allerted for synchronous infections especially in renal transplant recipients.

## Abstract# 93

**Renal Transplantation in Children with Abnormal Lower Urinary Tract** E. Baskin,[1] E. Melek,[1] U. Bayrakçi,[1] K. Gülleroglu,[1] S. Sevmis,[2] H. Karakayali,[2] M. Haberal.[2] *[1]Pediatric Nephrology, Baskent University, Ankara, Turkey; [2]General Surgery, Baskent University, Ankara, Turkey.*

**Objectives:** Renal transplantation in children with chronic renal failure (CRF) due to abnormal lower urinary tract (LUT) has been associated with a poor outcome compared to transplantation in those with a normal LUT. The optimal urological

treatment for such children remains unclear and reports had conflicting results. We retrospectively evaluated graft survival, graft functions and post transplant urological complications in our renal transplant patients with abnormal LUT.
**Methods:** 71 renal transplant recipients were included to study (mean age: 13.3±4.25 years). The etiology of CRF was the abnormality of LUT in 24 (32.4%) recipients.
**Results:** The mean follow-up was 34.72±28.54 months in patients with LUT abnormalities. There was no statistically significant difference between patients with normal and abnormal LUT when regarding graft survival, immunosuppressive treatment, type of donor, sex, mean ages and mean serum creatinine levels. Though it was not statistically significant the incidence of urinary tract infection was found to be slightly higher in patients with lower urinary tract problems (50.0% and 32% respectively, p>0.05). There was no statstically significant diference between the two groups regarding allograft lost (p>0.05).
**Conclusions:** Although these children display a high incidence of urologic and infectious complications, renal transplantation in patients with abnormal lower urinary tracts is safe and effective. Pre- and posttransplant urologic management is very important for good results.

## Abstract# 94

**Cardiac Dysrhythmias Secondary To Usage of Tacrolimus in 2 Renal Transplant Patients** U. Bayrakçi,[1] E. Melek,[1] E. Baskin,[1] K. Gülleroglu,[1] N. Cindik,[2] B. Varan,[2] S. Sevmis,[3] M. Haberal.[3] *[1]Pediatric Nephrology, Baskent University, Ankara, Turkey; [2]Peditric Cardiology, Baskent University, Ankara, Turkey; [3]General Surgery, Baskent University, Ankara, Turkey.*
**Objectives:** Tacrolimus is a potent macrolide widely used in transplanted patients. One of the side effects of Tacrolimus is cardiovasculer complications including dysrhytmias. Here we report two renal transplant patients experienced tacrolimus releted dysrythmia.
**Methods:** 16 years old female with the diagnosis of systemic lupus erythematosus and 15.5 years old boy with the diagnosis of posterior uretral valve and reflux nephropathy undervent renal tarnsplantation (living donor and cadaveric donor respectively). 1st patient at 20th day and 2nd patient at 50th day of post-transplantation period admitted to our unit with the complaint of palpitation.
**Results:** On their electrocardiograms, sinus taccycardia was detected. Holter monitorization of both patients revealed sinus tachycardia and ventricular extrasystoles (VES). Both patients had elevated tacrolimus level and electrolite levels were normal. After regulation of tacrolimus dosage and starting of metoprolol, palpitation disappear and patients were to get better.
**Conclusions:** Although dysritmias are very rare after oral administration and reversible in most cases, sometimes fatal arrytmias can be triggered by Tacrolimus. After regulation of tacrolimus dosage, improvement of patients complaint show that this effect is dose related. We report these cases to point that patients using tacrolimus should be followed for dysrhytmias and tacrolimus level should be checked If there is suspicion of dysrhytmias.

## Abstract# 95

**Sirtuin 1 Targeting Increases T Regulatory (Treg) Cell Suppression and Prolongs Allograft Survival** U.H. Beier,[1,2] L. Wang,[2] T. Bhatti,[2] Y. Liu,[2] R. Han,[2] G. Ge,[2] K.E. Meyers,[1] W.W. Hancock.[2] *[1]Department of Pediatrics, Division of Nephrology, The Children's Hospital of Philadelphia, Philadelphia, PA, United States; [2]Department of Pathology and Laboratory Medicine, The Children's Hospital of Philadelphia, Philadelphia, PA, United States.*
**Objectives:** Sirtuin 1 (Sirt1), a class III histone deacetylase, is an important regulator of the caloric restriction response, life span and cancer formation. We investigated its role in T-cell functions.
**Methods:** Mismatched BALB/c hearts were transplanted to wild-type (C57BL/6) mice or mice with targeted deletion of Sirt1 in CD4+ T-cells (SC) or Foxp3+ Tregs (SF). Treg function was analyzed by in vitro suppression assays and inhibition of homeostatic proliferation in RAG1-/- mice. Effector T cell function was analyzed by injection of CFSE-labeled SC or wild-type lymphocytes injected into B6/DBA2 mice (parent->F1).
**Results:** Allograft survival was improved 2-8 fold in SF and SC mice (p=0.014 and p=0.0011 on Mantel-Cox test, respectively), and data were reproduced in wild-type mice treated with Sirt1 inhibitors. Graft histology indicated a similar degree of lymphocyte infiltration, albeit significantly improved preservation of cardiac myocytes, in SC recipients. Consistent with this, SC and SF Treg cells showed enhanced function in Treg suppression assays, and 4-6 fold upregulation of CTLA4 and Foxp3, respectively (p<0.001), whereas SC effector T cells showed normal function in vitro and in vivo.
**Conclusions:** Sirt1 targeting enhances Treg suppression in vitro and in vivo, leading to prolongation of allograft survival.

## Abstract# 96

**Pharmacodynamic Monitoring of Cyclosporine A by NFAT-Regulated Gene Expression and the Relationship with Infectious Complications in Pediatric Renal Transplant Recipients** H. Billing,[1] T. Giese,[2] C. Sommerer,[3] M. Zeier,[3] R. Feneberg,[1] S. Meuer,[2] B. Toenshoff.[1] *[1]1University Children's Hospital Heidelberg, Heidelberg, Germany; [2]Institute of Immunology, University of Heidelberg, Heidelberg, Germany; [3]Department of Nephrology, University of Heidelberg, Heidelberg, Germany.*
**Objectives:** PK of cyclosporin A (CsA) is unsatisfactory, because the frequency and severity of adverse effects vary considerably among patients (p). We have therefore developed a whole blood assay that measures the suppression of CsA-target genes in T lymphocytes (NFAT). Different characteristics of CsA PK in children and the higher propensity for infectious complications, this assay requires validation in the pediatric patient population.
**Methods:** We therefore quantified in a prospective study of 45 pediatric renal transplant recipients NFAT in lymphocytes by RT-PCR and correlated these findings with the frequency of recurrent infections in the maintenance period post-transplant.
**Results:** P with infections showed a stronger inhibition of NFAT (18%) than patients without recurrent infections (31%; P=0.012). This difference was specific, because various PK parameters of CsA and the concomitant immunosuppressive therapy were comparable. Multivariate regression analysis showed that age and NFAT were the only independent determinants of recurrent infections. ROC curve analysis, a cut-off value of 23% NFAT had the highest sensitivity (71%) and specificity (65%).
**Conclusions:** PD monitoring of CsA by measurement of NFAT has the potential to identify over-immunosuppressed pediatric renal transplant recipients and is therefore a useful tool for the optimization of CsA therapy.

## Abstract# 97

**Extended Experience with IVIG and Rituximab for Treatment of Chronic Antibody-Mediated Rejection in Pediatric Renal Transplant Recipients** H. Billing,[1] C. Suesal,[2] J. Ovens,[2] R. Waldherr,[3] G. Opelz,[2] B. Toenshoff.[1] *[1]Children's Hospital, University Heidelberg, Heidelberg, Germany; [2]Department of Transplantation Immunology, University Heidelberg, Heidelberg, Germany; [3]Institute for Clinical Pathology, Heidelberg, Germany.*
**Objectives:** Chronic antibody-mediated rejection (CAMR) of renal allografts is assotiated with poor outcome and there is no established treatment protocol. We have published the results of a pilot study in 6 patients on treatment of CAMR with IVIG and rituximab (Billing et al, Transplantation 2008).
**Methods:** 11 patients (p) with CAMR, follow-up in median 18 months (mo); range, 6 to 36 mo. Antihumural therapy consisted of 4 weekly doses of IVIG (1 g/kg body weight per dose), and rituximab (375 mg/m2). We used the Banff '05 classification. HLA-antibodies were detected by solid phase ELISA assays and Luminex assay.
**Results:** In p without transplant glomerulopathy TG (n=6), median eGFR 6 months prior to intervention dropped by 8.4 (range, 5.9 16.0) ml/min/1.73 m2 (P<0.05) and increased by 8.1 (0.6 to 14.5) ml/min/1.73 m 12 months (P=0.029) after treatment, while p with TG remained stable. After 36 mo, transplant survival in p with CAMR and TG was 40% compared to 100% in p with CAMR without TG (P<0.02). Prior to therapy, 7 of 11 p had donor-specific class I (n=2) and/or class II (n=8) HLA antibodies by ELISA/Luminex. Six mo after therapy, HLA ab were no longer detectable in 3 of 9 p and persisted in 6 of 9.
**Conclusions:** This pilot study demonstrates that CAMR without TG in pediatric renal transplant recipients can be treated successfully and safely with a combination of IVIG and rituximab. P with CAMR complicated by TG did not show a persistent response.

## Abstract# 98

**Evaluation of Leflunomide (LEF) Therapy for Polyomavirus (PV) Nephropathy (PVN) in Pediatric Kidney Transplant (KT) Recipients** M. Bitzan,[1] F. Ghane Sharbaf,[1,2] L. Bell,[1] T. Dumonceaux,[3] A. Severini,[4] C. Bernard.[1] *[1]Montreal Children's Hospital, Montreal, QC, Canada; [2]Dr. Sheikh Pediatric Hospital, Mashhad, Islamic Republic of Iran; [3]Agriculture/Agri-Food Canada, Saskatoon, SK, Canada; [4]Public Health Agency of Canada, Winnipeg, MB, Canada.*
**Objectives:** PVN is an important risk for graft failure. We evaluated efficacy & adverse effects (AEs) of antiviral LEF therapy in children with PVN.
**Methods:** Retrospective cohort study of 8 consecutive KT recipients with biopsy proven (5) or presumed (3) PVN. Data analysis/reporting: median (range); Spearman's rank correlation.
**Results:** Diagnosis was 11 (1-28) mo post KT at age 10 (3-17) yrs. Peak BK viral load was 7.8 x $10^4$/ml plasma (3.3 x $10^3$ - 4.8 x $10^6$). 2 pts with histological PVN demonstrated JCV replication alone (1.7 x $10^6$/ml plasma) or combined with

BKV. S-creatinine (Cr) was 25 (12-72) % above baseline. MMF was stopped in 6/7 & reduced in 1, tacrolimus reduced in 6/7, & sirolimus in 1/1 pts. Median steady state LEF plasma levels per pt ranged from 29 to 166 mg/l with dose requirements inversely related to age (r = - 0.35). 6/8 pts cleared PV from blood after 17 (1-30) mo of treatment; 5/8 ceased shedding virus in urine. Cr returned to baseline in 4, remained stable in 1 & increased gradually in 3 pt. Rising transaminases led to LEF discontinuation in 1 with preexisting mild hepatopathy. 3 pts had LEF-associated (moderate) platelet and 2 neutrophil decline.
**Conclusions:** LEF was generally well tolerated. 75% cleared BKV from blood. LEF dose requirements correlated inversely with age, a new finding. There was no graft loss due to PVN nor rejection with immunosupression reduction and LEF, and most maintained good graft function.

## Abstract# 99

**Generic Tacrolimus (T- Inmun) in Pediatric Renal Transplant Recipients in Chile. Preliminary Experience** L. Bolte, M. Aglony, A. Vogel, V. Perez. *Pontificia Universidad Católica de Chile, Santiago, Chile.*
**Objectives:** Tacrolimus has long been used for prevention and treatment of acute rejection in solid organ transplantation. Its efficacy and safety in children has been proved. We report our experience in pediatric renal transplantation using a generic formulation(T-Inmun).
**Methods:** Descriptive study, 3 patients(14-19 years) receiving T-Inmun as part of their immunosuppressive therapy for renal transplantation.
**Results:** All had induction therapy with basiliximab(in days 0 and 4 post transplantation), methylprednisolone and T-Inmun immediately after surgery.

Maintenance immunosuppressive therapy

| Patient ID | 1 | 2 | 3 |
|---|---|---|---|
| Primary renal disease | FSGS | Alport | Unknown |
| Donor source | LD | LD | CD |
| Time since transplant (mo) | 16 | 25 | 29 |
| Immunosuppressive therapy | Ti-S-P | Ti-MMF-P | Ti-MMF-P |
| Estimated clearence (mL/min/1.73m²) | 114 | 104 | 81 |
| Ti Plasma levels T0 (ng/mL) | 1,8 | 7,1 | 9,4 |
| Ti Dosis (mg/kg) | 0,06 | 0,16 | 0,12 |

LD:living donor; CD:cadaveric donor; Ti: T-Inmun; S: sirolimus; MMF: mofetil mycophenolate; P: prednisone

One patient (ID 1) was switched one year post transplantation to sirolimus, prednisone and low dose of Tinmun because of secondary effects of tacrolimus (severe tremor). Initial doses of T-Inmun were 0.15 mg/kg/d, and were adjusted to maintain trough plasma levels between 10-15 ng/mL in the first year after transplantation and 5-10 ng/mL thereafter. To date, no rejection episodes have been observed and all patients have a stable renal function. Two have controlled arterial hypertension. None has developed proteinuria, hyperglicemia or hyperkalemia.
**Conclusions:** T-Inmun has been a safe and efficient immunosuppressive drug in this little group of children with renal transplant.

## Abstract# 100

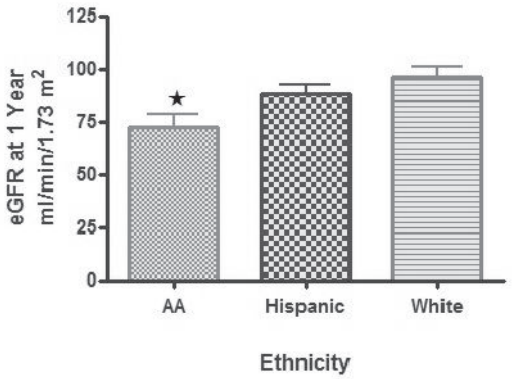
**Methylmalonic Acidemia: A New Indication for Preemptive Renal Transplantation?** O. Boyer, P. Krug, G. Guest, V. Valayannopoulos, P. Niaudet. *Hôpital Necker Enfants Malades, Paris, France.*
**Objectives:** Methylmalonic acidemia (MMA), an inborn error of organic acid metabolism often leads to severe central nervous system damage and end-stage renal disease (ESRD). When medical treatment is ineffective, liver or kidney transplantation are discussed.
**Methods:** We report a series of 4 children who received a cadaveric renal transplant in order to partially correct the enzymatic activity (methylmalonyl CoA mutase).
**Results:** The four boys were diagnosed with a severe form of vitamin B12-unresponsive MMA before the age of 2 months, responsible for frequent metabolic decompensations and neurologic complications despite low-protein diet by exclusive enteral feeding and carnitine. Patient (Pt) 1 reached ESRD at 8 years and was transplanted at 9. Pt2 was started on peritoneal dialysis at age 6 years and transplanted 4 months later. Pt3 and 4 with multiple metabolic decompensations and neurological deterioration received a preemptive renal transplant at 10 and 5 years despite an eGFR of 37-105 ml/min/1.73m$^2$ respectively. In all patients, creatinine and MMA levels quickly decreased allowing a safe increase in protein intake. All had a functioning graft with a follow-up of 0.5-3 years. The number of metabolic decompensations per year dramatically decreased, occurring mostly in the setting of an acute rejection episode. All of them started oral alimentation and attended school. Neurological complications stabilized, and no further deterioration was noted. Patient 1 died of a hepatocarcinoma 2 years after transplantation.
**Conclusions:** Renal transplantation partially corrects the enzymatic activity, stabilizes neurological status and improves the quality of life in children with severe MMA.

## Abstract# 101

**Live Renal Transplantation Versus Cadaveric Renal Transplantation** E. Laura,[1] N. Mercedes,[1] G. Carmen,[1] A. Angel,[1] M. Marta,[1] F. Carlota,[1] M.U. Mª José,[2] J. Enrique.[2] *[1]Nefrología Infantil, Hospital La Paz, Madrid, Spain; [2]Urología Infantil, Hospital La Paz, Madrid, Spain.*
**Objectives:** We report our experience with live renal transplantation of related donors (LKT) in paediatric patients versus cadaveric kidney transplantation (CKT).
**Methods:** From 1994 to 2009 a kidney transplantation was performed in 231 pediatric patients, 72 (31%) LKT from 21 fathers and 51 mothers. The standard immunosuppressive protocol is: induction (baxilisimab/Thymoglobuline) and triple therapy with steroids, mycophenolate mofetil/azathioprine and cyclosporine/tacrolimus.
**Results:** Ethiology of ESRD in LKT and CKT was:renal dysplasia 23% vs 21%, obstructive uropathy 23% vs 37%,FSGS 1% vs 7% and others 46% vs 50%.The transplantation´s age was non significant (NS) 10,9 ± 5,5 y (LKT) vs 10,6±5,3 (CKT); donor´s age for LKT was 39,7±7 y vs 9,4±9,3 (p<0,0001). Preemptive transplantation: 50% of LKT vs 13,2% for CKT (p<0,0001).The time in dialysis for the first graft in LKT has been 5,75±10,7 months vs 10,6±11 m in CKT (p=0,005). Cold ischemia time was lesser in LKT 1,8±0,5 h vs 17,6±5h in CKT (p<0,0001). There have been lost 8/72 grafts (11%) in LKT vs 42/117 (26%) in CKT (p=0,009). The failure´s age was 15,8±5,6 y (LKT) vs 14,3±4,5 y (CKT) (NS).The cumulative acute cellular rejection was 25% (LKT) vs 33,8% (CKT) (NS).Patient survival was 98,3% vs 98,1 for LKT vs CKT. Graft survival was 97,5%, and 81,3% to 1 and 7 years follow up (LKT) vs .90,4 and 72,8 in CKT (p=0,039).
**Conclusions:** Live kidney transplantation in children is the best treatment for ESRD: 50% preemptive transplantation, significant decrese of time on dialysis and better long time survival (p=0,039).

## Abstract# 102

**Ethnic Disparities in Pediatric Kidney Transplantation: A Single Center Experience** J. Chandar, S. Muneeruddin, C.L. Abitbol, W. Seeherunvong, M. Freundlich, G. Ciancio, G.W. Burke III, G. Zilleruelo. *Pediatric Nephrology, University of Miami/Holtz Children's Hospital, Miami, FL, United States.*
**Objectives:** To examine a 20 year experience in kidney transplantation in children from a predominantly Hispanic community.
**Methods:** A retrospective analysis was performed in 124 pediatric kidney transplants with 81 (65%) from living donors (LD) during the period from 1985 to 2005.
**Results:** Ethnic distribution was 48% Hispanic, 24% African American (AA), and 26% Caucasian. First year allograft survival was similar in LD and deceased donors (DD). eGFR <60 ml/min/1.73 m$^2$ at 1 year post transplant was associated with a median allograft survival of 3.3 years, compared to 16 years in those with eGFR ≥ 60 ml/min/1.73m$^2$ (p<0.0001). Graft loss in the first 5 years was from non-adherence, recurrent disease and infections. Those of AA race were more likely to receive a DD and have the poorest median allograft survival compared to Hispanics and Caucasians (6 versus ≥15 years; p<0.001). Hispanics showed similar allograft survival regardless of donor source with a median survival of 13 years for LD and 15.5 years for DD.




**Conclusions:** This predominantly Hispanic cohort emphasizes the disadvantaged profile of AA's compared to other ethnic groups. Strategies to improve supportive services and living donations in minority populations need to be developed.

## Abstract# 103

**Posttransplantation Lymphoproliferative Disorder (PTLD) in Pediatric Kidney Transplant (KT) Recipients – A National Study**

R. Cleper,[1] E. Ben Shalom,[3] D. Landau,[5] I. Weissman,[6] I. Krause,[1] R. Rahamimov,[2] E. Mor,[2] N. Bar Nathan,[2] Y. Frishberg,[3] M. Davidovits.[1]
*[1]Pediatric Nephrology and Dialysis Institute, Schneider Children's Medical Center of Israel, Petah Tiqva, Israel; [2]Department of Transplantation, Rabin Medical Center, Beilinson Hospital, Petah Tiqva, Israel; [3]Division of Pediatric Nephrology, Shaare Zedek Medical Center, Jerusalem, Israel; [4]Department of Pediatric Radiology, Schneider Children's Medical Center of Israel, Petah Tiqva, Israel; [5]Department of Pediatrics A and Pediatric Nephrology Clinic, Soroka Medical Center, Beer Sheva, Israel; [6]Department of Pediatric Nephrology and Dialysis, Nahariya, Western Galilee Hospital, Nahariya, Israel.*

**Objectives:** Examine prevalence, profile, risk factors and outcome of PTLD in Israeli pediatric KT recipients.

**Methods:** File search of all pediatric(<19yrs)KT recipients in the National Israeli Kidney Transplant Registry during1991-2008.

**Results:** 12 pts(5F,7M)1st time KT recipients were diagnosed with PTLD from 300(112F,188M) KT patients. PTLD pts were younger at KT: **4.2vs13yrs** (p=0.002), had **AB** blood group: 30% vs 6.2%(p=0.017) and had received **OKT3** 25% vs 3.8% (p=0.015). **50%** PTLD patients were **EBV+** at KT. PTLD occurred at **˜3.2yrs** post-KT- 3/12: 1st yr. Presenting features: 8/12 **fever**, 5/12 **abdominal pain**,6/12 **GFR loss**(transient).PTLD site:10/12 abdomen: 6/10- nodal, 1/12-kidney graft. PTLD type: 8/12 **Bc**, 3/12 **Tc**(at 5-15.5 yrs postKT) and 1 **Kaposi type**. Mortality was **25%,** for **EBV+: 60%**. 25% grafts were lost in survivors. Mortality risk factors: EBV+ status,older age at KT,cadaveric donor, late-onset(˜6.3yrs),Tc PTLD.

**Conclusions: EBV+** KT recipients are at risk for late-onset, highly malignant,↑mortality PTLD.

## Abstract# 104

**Metabolic Syndrome in Pediatric Renal Transplant Patients Steroid vs Non-Steroid Group – Analysis at 2 Years Post Transplant**

R.M. Coelho, G. Hidalgo, L. Fornell, L. Briars, R. Bottke, J. Siciliano, E. John. *Pediatrics, University of Illinois at Chicago, Chicago, IL, United States.*

**Objectives:** Cardiovascular (CV) disease is the main cause of morbidity and mortality in pediatric renal transplant (tx) patients(pts.). The objective of the study was to investigate the presence of metabolic syndrome (MS) in pediatric pts in the steroid (SG) and steroid withdrawal group (SWG) at 2 years post tx and to analyze the glomerular filtration rate (GFR) in SG, SWG and in MS and nonmetabolic syndrome group (NMS).

**Methods:** We retrospectively reviewed metabolic parameters, GFR and demographics in pediatric tx pts.between 1997-2008. The immunosuppressive protocol utilized was similar in SG and SWG except SWG received only 5 days of steroid post tx.

MS was defined as having three or more of the following criteria: z-BMI ≥ 2, fasting blood glucose ≥100 mg/dl, triglycerides ≥ 110 mg/dl, HDL cholesterol ≤ 35 mg/dl and systolic/diastolic blood pressure ≥ 90th % for height or taking antihypertensive medications.

Statistical analysis was done by student t test. * p <0.05 was considered significant.

**Results:** Twelve pts (35%) were in SG and 22 (64%) in SWG; 16 (47%) were female and 18 (52%) were male. Mean age ranged from 6-17 years.

The incidence of MS for the whole group was 35%; 58% in SG and 27% in SWG. The mean serum cholesterol*, triglycerides* and BMI* was high in SG with MS. At 2 years mean GFR was lower in MS vs NMS pts in both SG and SWG but not significant.

**Conclusions:** CV risk factors were high in SG and SWG with MS which can lead to morbidity and mortality. Vigorous treatment of MS in pediatric tx pts is warranted to avoid CV morbidity and mortality.

## Abstract# 105

**EBV Infection in Children with Renal Transplantation: 16 Years' Experience in a Single Center**

E. Comak,[1] A.U. Gökceoglu,[1] M. Koyun,[1] C.S. Dogan,[1] D. Colak,[2] A. Dinckan,[3] S. Akman.[1] *[1]Pediatric Nephrology, Akdeniz University, Antalya, Turkey; [2]Microbiology, Akdeniz University, Antalya, Turkey; [3]Akdeniz University, Antalya, Turkey.*

**Objectives:** The aim of this study was to detect the frequency and risk factors of Epstein-Barr virus (EBV) infection in pediatric renal transplant recipients.

**Methods:** The children who underwent renal transplantation at our center since 1994 were evaluated retrospectively. EBV was detected by serologic methods before November 2007, after this period by EBV DNA levels measured by polymerase chain reaction, which was studied biweekly in the first 2 months post-transplant, monthly between 2-6 months and trimonthly thereafter. All patients received anti-viral prophylaxis by acyclovir or valacyclovir for 6 months.

**Results:** A total of 117 children, 65 boys, mean age of 11.2±3.6 years, were included. Prior to transplantation, 100 patients (85.5%) were EBV seropositive and 17 were seronegative. Within a median follow-up period of 18 months, 3 of 17 (17.6%) seronegative children developed primary EBV infection and 22 of 100 children (22%) developed EBV reactivation. No patient had serum creatinine elevation accompanying EBV infection. EBV infection rates were similar between patients receiving tacrolimus and cyclosporin (18.9% vs 28%, p>0.05). Two of three patients with primary infection developed post-transplant lymphoproliferative disorder (PTLD), one of whom died.

**Conclusions:** The most important risk factor for EBV infection and PTLD after renal transplantation is EBV seronegativity prior to transplantation; so, these patients should be screened closely at the post-transplantation period.

## Abstract# 106

**Early Steroid Withdrawal in Pediatric Kidney Transplantation. Five Years of Follow-Up**

A. Delucchi,[1] M. Valenzuela,[2] M. Ferrario,[1] J. Godoy,[1] F. Cano,[1] A.M. Lillo,[1] J.L. Guerrero,[1] M. Azocar,[1] P. Zambrano,[3] P. Salas,[3] V. Pinto,[3] J. Rodriguez,[1] G. Gonzalez,[1] G. Cavada,[4] Fondecyt 1080166.
*[1]Nephrology, CMachenna, Santiago, Chile; [2]Nephrology, GBenavente, Concepción, Chile; [3]Nephrology, EGCortes, Santiago, Chile; [4]Public Health, UChile, Santiago, Chile.*

**Objectives:** A prospective controlled multicenter study 60months follow-up investigated early steroid withdrawal on growth (SDS) safety and efficacy.

**Methods:** 100 recipients (PRA<30%) were assigned to two groups: SW (steroid withdrawal n =60) SC (steroid control n=40). Initial dose TAC was 0.15 mg/kg twice daily,basal levels 10-15 ng/ml until the Day30 and then 5-7 ng/ml. MMF 800 mg/m2/d until the Day30 then 600 mg/m2/d and after third month 400 mg/m2/d. Basiliximab 20 mg/m2 IV on Day0 and 4. In SW, steroid doses decreased progressively to suspension at Day6. In SC, steroid were decreased to 10 mg/m2 within 2 months post-TX.

**Results:** 53/60 (88.3%) SW and 34/40 (85 %) SC completed 12months,20/60 (33.3%) SW and 11/40 (27.5 %) SC-60months of follow-up. Doses, TAC levels were comparable in both groups.Z height/age was significantly higher in SW (0.75) vs SC (0.12), (p=0.000) and delta Z height/age SW (0.88) vs SC (0.19) (p=0.000)at Month12 and remained significant until Month60.The biopsy-proved AR rate was (5/40) 8.3% SW vs. (7/16) 17.5% SC (p:ns).Patient and graft survival 1, 3 and 5 years was 99%,95%,95% vs. 94.6%/89.5%/86.1%. respectively. The average GFR at Month12 for SW and SC was 82.8 mL/min/1.73m2 and 82.1 mL/min/1.73m2 (p: ns) and 75.8 mL/min/1.73m2 and 71.7 mL/min/1.73m2 (p:ns), at Month60. PTLD incidence 2/53(3.3 %) SW and 1/34 (2.5 %) in SC (p:ns).

**Conclusions:** Early SW showed positive impact on growth until Month60 without AR risk, GFR change, diabetes or PTLD.

## Abstract# 107

**Biomarkers in Early Acute Rejection in Pediatric Renal Transplantation**

A. Delucchi,[4] M.I. Bunster,[1,2,3] M. Gonzalez,[1,2,3] F. Cano,[4] V. Pinto,[5] P. Salas,[5] L Michea.[1,2,3] *[1]Biomedical Science Institute, University Chile, Santiago, Chile; [2]Molecular Cell Studies, UCh, Santiago, Chile; [3]Millenium Centre Nucleus on Immunology and Immunotherapy, UCh, Santiago, Chile; [4]Luis Calvo Mackenna, FM,UCh, Santiago, Chile; [5]Exequiel G Cortes, FM,UCh, Santiago, Chile.*

**Objectives:** In children protocol biopsy, the best predictor of AR is difficult to perform. Non-invasive analyzing markers in plasma and urine have been explored. The objective was to assess quantitatively mRNA expression of IL-17 and FoxP3 in MNSP, urinary cells and graft biopsies in pediatric Tx.

**Methods:** Fifteen recipients were randomized and completed one year of follow-up. Blood and urine samples were obtained monthly until month 12 post-TX, RT-PCR (qPCR) was used for extraction of total mRNA and measure mRNA IL-17 and Foxp3. Biopsies were performed when AR was suspected and a protocol biopsy at month 12 post-TX.

**Results:** The FoxP3/IL-17 in MNSP (0.009±0.01) and (0.002±0.003) in urine, no association between specific donor anti HLA and AR in biopsies. One patient increased 3.200 times mRNA FoxP3/IL-17 in urinary cells and 165 times in mRNA FoxP3/IL-17 MNSP at month4 post-TX,with clinical AR, ab anti HLA and a decreased in renal function. The biopsy confirmed *Bordeline* AR, received steroids in high doses and renal function became normal, and FoxP3/IL-17 decreased. The biopsy at month12 post-TX detected a new episode of *Bordeline* AR, increase in plasma creatinine, no variation in the FoxP3/IL-17 reason in this opportunity.

**Conclusions:** Our results showed an increase in mRNA IL-17/FoxP3 in MNSP, urine and graft biopsy, suggesting as a promising bookmark in AR. Financed by FONDECYT 1080166, FONDAP 15010006, Nucleus Millennium P04/030F.

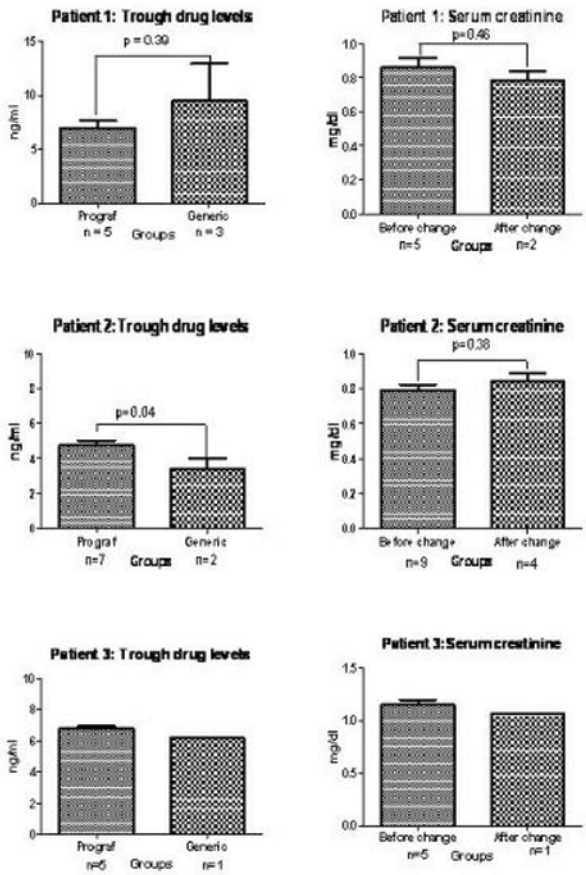
## Abstract# 108

**Comparison of Generic Tacrolimus and Prograf Drug Levels in Children with Kidney Transplants** V. Dharnidharka, H. Abdulnour, C. Araya. *Division of Pediatric Nephrology, Shands Children's Hospital and University of Florida, Gainesville, FL, United States.*

**Objectives:** A generic version of tacrolimus was approved for use in the USA in 2009. Since these generics are tested for bioequivalence only in adults, pediatricians are hesitant to use narrow therapeutic index generics in children right after approval. No data are yet available on trough generic tacrolimus levels in children with organ transplants in the USA.

**Methods:** Three patients in our pediatric kidney transplant program, all with stable allograft function, were inadvertently switched to generic tacrolimus. We retrospectively analyzed pre-and post-switch 12-hour trough tacrolimus levels and serum creatinine values.

**Results:** Tacrolimus levels (mean + S.E.) before and after switch were a) Patient 1 (12 year old female): 7.0 ± 0.69 and 9.7 ± 3.5 (p = NS); b) Patient 2 (8 year old male): 4.7 ± 0.68 and 3.4 ± 0.84 (p =0.04); c) Patient 3 (22 year old female): 6.8 ± 0.7 and 6.1 (no SE). Serum creatinine levels were virtually identical pre- and post-switch in all three patients, eGFR above 75 ml/min/1.73 m2 in all cases. No subject required a biopsy.




**Conclusions:** Though serum creatinine levels were unchanged, the mean trough tacrolimus level was statistically significantly lower in patient 2 and clinically different in patient 1. We suggest careful monitoring of pediatric patients who get switched to generic tacrolimus.

**DISCLOSURE:** *Dharnidharka, V.*: Consultant, BristolMyersSquibb; Other, Genzyme.

## Abstract# 109

**BK Viraemia and Nephropathy in Paediatric Renal Transplant Recipients** N.M. Dolan, D. Cubitt, N.J. Sebire, S.D. Marks. *Great Ormond Street Hospital for Children NHS Trust, London, United Kingdom.*

**Objectives:** Evaluate incidence of BK viraemia and nephropathy in a single centre paediatric renal transplant population.

**Methods:** Children who received renal transplants between 1993-2008 were monitored during the period May 2007-June 2008. Patient population was divided into 2 groups:Group 1 were transplanted during study period and prospectively monitored with regular plasma BK PCR DNA.Group 2 included all patients transplanted from 1993.Patients were screened at routine clinic appointments or if any evidence of renal allograft dysfunction.Renal biopsy was performed if any graft dysfunction.

**Results:** 729 samples were analysed from 130 patient with 18 patients in Group 1 and 112 in Group 2.Median ages at transplantation for Group1 and 2 were 10.6(1.6-16.6) years and 8.1(1.25-17.3) years respectively.BK viraemia was evident in four(22%) from group1 and seven(6%) from group 2.Median time to detection of BK viraemia post transplant in Group 1 was 52(30-100)days and 470(41-1450)days in Group 2.Six patients in BKV group received induction therapy with monoclonal antibodies; three patients from group 1 and three from group 2.Three patients(two in group 1,one in group 2)had biopsy proven BKVAN.Time to detection post transplant in these patients was 2.7-14.5 months. All three were receiving triple immunosuppression with corticosteroids, MMF and tacrolimus at time of BKVAN detection.There was a 100% patient and graft survival at 2.4-169.3(median 26.6)months post-transplantation.

**Conclusions:** Viral complications are commoner after transplantation due to increasing immunosuppression.Paediatric renal transplant recipients should be screened for BK viraemia in order to reduce immunosuppression and avoid BKVAN.

## Abstract# 110

**Recurrence of MPGN Type II in Pediatric Renal Transplant: Early Diagnosis & Successful Therapy** E. Elenberg. *Pediatrics, Baylor College of Medicine, Houston, TX, United States.*

**Objectives:** Risk of recurrence of membranoproliferative glomerulonephritis type II (MPGN II) with graft loss after renal transplant (RTX) is high (>50%). Early detection allowing early treatment has not been reported previously. In this case report, MPGN II recurrence was diagnosed after onset of new proteinuria prior to rise in serum Cr (Scr) & successfully treated.

**Methods:** Retrospective case review of a 10 yo girl with recurrent MPGN II in RTX was done. Pt presented at age 6 yo with Scr 1.5 mg/dl, low C3 (22mg/dl), nl C4, positive C3 nephritic factor (C3NeF+), nl factor H, & renal biopsy (Bx) with crescentic MPGN II. Pt was resistant to all Rx & developed ESRD in 3 wk. She had deceased donor RTX at age 10 yo, treated with basilizimab induction + prednisone (pred), tacrolimus (FK) & mycophenolate.

**Results:** Within 2 wk of RTX, pt had new proteinuria (3+ protein, 0.9 g protein/24h), Scr 0.6mg/dl, albumin 3.2mg/dl, C3 17mg/dl & C3NeF+, with MPGN II recurrence by RTX Bx. Pt received methylprednisolone 6mg/kg IV x3d, followed by slow pred taper from 3mg/kg/d. After 2 m, proteinuria resolved, but pt developed IDDM, which resolved with changing FK to cyclosporine, & tapering pred to 17.5mg/d at 6m post RTX. 12m after RTX, pt again had proteinuria 334mg/24h, which resolved with pred 20mg/d. Pred was slowly tapered to 12.5mg by 18 m. Currently, 3 y post RTX, Scr is 0.6mg/dl & no proteinuria, despite persistent low C3 (11mg/dl) & C3NeF+.

**Conclusions:** This case highlights early detection of MPGN II recurrence in RTX by early Bx at first appearance of new onset proteinuria. Early detection & early pulse steroid Rx led to resolution of proteinuria & maintenance of good allograft function despite active serologic indices of MPGN II.

## Abstract# 111

**Heat Shock Protein (HSP) Polymorphism Predisposes to Urinary Tract Malformations and Renal Transplantation in Children?** A. Fekete,[1] K. Rusai,[1] N.F. Banki,[1] E. Karoly,[2] G.S. Reusz,[1] T. Tulassay,[1] A.J. Szabo.[1] *[1]1st Department of Pediatrics, Semmelweis University, Budapest, Hungary; [2]Department of Pediatrics, Hetenyi Geza Hospital, Szolnok, Hungary.*

**Objectives:** Anatomical malformations of the kidney and urinary tract (UT) account for 17% of pediatric renal transplantations (Tx). HSPs activates innate immunity through toll-like receptor (TLR)4. HSPA1B A(1267)G polymorphism leads to lower HSP72 mRNA and is associated with kidney diseases in adults and children. TLR4 A(896)G polymorphism is associated with a higher risk of infections.

**Methods:** HSPA1A G(190)C, HSPA1B A(1267)G and TLR4 A(896)G polymorphisms were analyzed in 41 pediatric Tx recipients, 103 children with recurrent UT infections and 235 healthy controls (CON). Clinical data were also evaluated.

**Results:** HSPA1B (1267)GG genotype and (1267)G allele were more frequent in Tx (p=0.014; p=0.011) and in UTI (p=0.0001) vs. CON. HSPA1B (1267)GG and (1267)G allele were associated with higher risk of renal scarring after UTI (p=0.012 and 0.049). VUR was associated with a higher incidence of HSPA1B (1267)GG genotype in Tx (p=0.007).

TLR4 (896)AG genotype and (896)G allele were more frequent in UTI vs. CON (p=0.031; 0,041), while there was no difference in Tx. TLR4 (896)G was more often in UTI without VUR vs. UTI with VUR (p=0.03).

**Conclusions:** HSPA1B (1267)G allele might be a risk for UTI, VUR and influences the development of renal scarring and ESRD leading to Tx.TLR4 (896)G allele might be associated with UTI independently of other renal abnormalities. These findings raise further questions about the clinical relevance of these polymorphisms in pediatric nephrology.

Supported by OTKA, ETT,TAMOP.

## Abstract# 112

**A One Year Prospective Comparison of Kidney Growth and Function in Children Recipients of Grafts from Children and Adults**

L.S. Feltran,[1] P.C. Koch Nogueira,[2] A. Pacheco-Silva.[2] *[1]Nephrology, UNIFESP, Sao Paulo, Brazil; [2]Pediatric Nephrology, UNIFESP, Sao Paulo, Brazil.*

**Objectives:** We conducted a one year prospective study comparing the growth of renal grafts from children and adults transplanted in children, and correlated this growth with graft function and recipient's growth.

**Methods:** Two groups were studied: a) Group 1 – comprising 32 children transplanted with pediatric deceased kidneys from donors < 16 years old and b) Group 2 comprising 31 children transplanted with organs from adult living donors. Anthropometric measurements, sonographic study of the graft and serum creatinine at 1 week, 1, 6 and 12 months post-transplantation were performed.

**Results:** Children from Group 1 presented an 18% increase in graft volume after the sixth month of transplant, whereas in Group 2 grafts presented a 14% reduction in volume, mainly during the first month. The variation in renal diameters was not uniform. GFR increase was detected during the follow up of children from Group 1 (46 to 102 ml/min/1,73m$^2$) and GFR decrease was seen in Group 2 (104 to 99 ml/min/1,73m$^2$). Growth of individuals from both groups was comparable.

**Conclusions:** During the first year of transplant, pediatric kidneys displayed a slight increase in volume after six months, which was in parallel with a GFR increase in children recipients. Adult kidneys evidenced a slight reduction in both volume and GFR. Variation in graft sizes was independent of children's growth. Taking our and other studies results into consideration, we can hypothesize that besides compensatory hypertrophy, pediatric grafts are likely capable of continued somatic growth.

## Abstract# 113

**Comparison between Valganciclovir and Aciclovir/Valaciclovir for CMV Prophylaxis in Pediatric Renal Tranplantation**

M. Fila, A. Deschartres, A. Maisin, S. Azib, G. Deschenes, V. Baudouin. *Nephrologie Pediatrique, CHU Robert Debré, Paris, France.*

**Objectives:** Systematic prophylaxis has dramatically decreased CMV infection in transplanted patients but treatment options are multiple and few data are available for pediatric population. The aim of this work was to evaluate efficiency of valganciclovir *vs* aciclovir/valaciclovir in a pediatric renal transplant population.

**Methods:** Data from 88 renal transplantations done between 12/1999 and 07/2008 were retrospectively analyzed. All patients except those R-/D- (n=17) received prophylaxis for 90 days either with aciclovir/valaciclovir (G1, n=40) or valganciclovir (G2, n=31). Incidence of positive CMV antigenemia (Ag+) with CMV disease (CMVd) or not as well as the delay in relation to prophylaxis were collected during at least 6 months after end of treatment.

**Results:** Thirty patients (34%) experienced Ag+: 18 were from G1 (45%), and 11 (35.4%) from G2. CMVd occurred in 11 patients (12.5%) (4 in G1 and 7 in G2) (ns). Ag+ occurred during the 90 days of prophylaxis in 12 patients in G1 (30%) *vs* only 1 in G2 (3.2%) (p=0.01). The others experienced Ag+ after cessation of prophylaxis (6/40 in G1, 10/31 in G2, (ns), without any difference in delay between G1 and G2 (ns).

**Conclusions:** In this study, incidence of Ag+ or CMVd is similar with aciclovir or valganciclovir prophylaxis. However valganciclovir is more efficient to prevent early infection as incidence of infection is much less high than with aciclovir during time of prophylaxis. Increase incidence of Ag+ after the end of treatment point out the issue of modality and duration of prophylaxis with valganciclovir.

## Abstract# 114

**Typhilitis: A Rare Cause of Fever of Undetermined Origin Post Kidney Transplant**

J.W. Foreman, D.R. Wigfall, S. Nagaraj, L. Patterson, C. Hollingsworth, R. Vinson, K. McGann, R. Gbadegesin. *Pediatrics, Duke University, Durham, NC, United States.*

**Objectives:** The discovery of potent immunosuppressive agents has dramatically improved the outcome of kidney and other solid organ transplantation by decreasing the rate of acute rejection and chronic allograft dysfunction. However, a trade off for this is the emergence of infectious complications as a leading cause of morbidity and mortality in solid organ recipients. Careful documentation of these unusual infections will increase the index of suspicion leading to early diagnosis, treatment, reduced morbidity, and mortality. This is a report of an unusual cause of fever in a renal transplant patient.

**Methods:** The clinical course of a kidney transplant recipient with fever of unknown origin was extracted from his medical records.

**Results:** He is a 15 year old boy with CKD due to renal dysplasia. He developed allograft dysfunction 8 months post transplant due to primary CMV, 2 months later, he developed fever, abdominal pain and leucopenia. All his bacterial, viral and fungal cultures were negative. CT scan with contrast revealed bowel wall thickening which is confined to the region of the cecum along with minimal surrounding fat stranding and a small pericecal fluid collection consistent with typhilitis. He was managed with antibiotics and nil per oris. He defervesced within 48 hours of starting antibiotics and his abdominal pain got better.

**Conclusions:** The favorable outcome in this patient is probably because of early initiation of treatment. This report suggests that typhilitis should be considered as a differential diagnosis of unexplained fever, GI symptoms and leucopenia in a post transplant patient.

## Abstract# 115

**Anti-HLA Antibodies and Renal Acute Rejection in Children**

R. Galeas, S. Valverde, B. Romero, R. Gomezchico, L. Velasquez, C. De Leo, M. Medeiros. *Nefrologia, Hospital Infantil de Mexico Federico Gomez, Mexico, DF, Mexico; Laboratorio de Trasplantes, Instituto Nacional de Ciencias Medicas y la Nutricion Salvador Zubiran, Mexico, DF, Mexico.*

**Objectives:** The aim of the study was to determine the presence of HLA antibodies in children with acute renal graft rejection.

**Methods:** 21 children with biopsy proven acute rejection were included, renal tissue was evaluated according to Banff Classification; C4d immunostaining was performed in all cases. A serum sample was drawn at the time of biopsy, HLA antibodies Class I and Class II were determined by Luminex single antigen (One Lambda).

**Results:** Seventeen patients had cellular rejection (CR) and four patients antibody mediated rejection (AMR). 11/21 patients had donor specific antibodies (DSA), 4/21 were DSA vs. Class I, 5/21 DSA vs. Class II, and 2/21 against both Class II and Class II. All the patients with AMR had DSA vs. Class I. (Table 1).
20/21 patients had non-specific antibodies (95.2%). One patient with cellular rejection Class IIA had no HLA antibodies.
After a mean follow up of 46 months, both groups had a reduction in glomerular filtration rate (GFR) vs. baseline. One patient with cellular rejection lost the graft due to non-adherence. 3/4 patients with AMR were successfully treated with plasmapheresis, IVIG and rituximab. CR were treated with methylprednisolone.

Donor-specific antibodies and type of rejection

| Type of antibodies | Cellular rejection | Antibody mediated rejection |
|---|---|---|
| Class I donor- specific antibodies | 2 | 4 |
| Class I non donor-specific antibodies | 15 | 0 |

Chi Square p <0.05

**Conclusions:** HLA antibodies are frequent in acute rejection episodes (95.2%). DSA Class I was present in all the patients with AMR.

**DISCLOSURE:** *Medeiros, M.*: Consultant, Novartis, Mexico.

## Abstract# 116

**Brazilian Pediatric Renal Transplant Registry: 5 Year Report**

J. Medina-Pestana, P. Koch, M.C. Andrade, C. Garcia, V. Bittencourt, R. Meneses, J. Fontes, V. Belangero, L. Prates, D. Carvalho, T. Matuck, V. Benini, S. Laranjo-Martins, E. Lima, J.M. Penido, M. Abbud Filho, H. Ramalho, I. Fernandez, L. Feltran, M.F. Camargo, M.d.F. Gesteira, N. Leao. *Registro Brasileiro de Transplante, ABTO & ALANEPE, Sao Paulo, Brazil.*

**Objectives:** In 2003, the pediatric renal transplant registry started and a growing number of centers are participating each year.

**Methods:** Data from RTx performed in pts < 21 years-old, from January 1st 2004 to December, 31st 2009 were analyzed. Demographics and patient and graft survival are reported.

**Results:** 897 pediatric RTx were performed in 11 centers. Mean age at RTx was 11.9+/-1.5 y (53.2% male). Etiology of ESRD was uropathy (30.1%), glomerulopathy (26.3%) - 40.7% FSGS, and other causes (43.6%). Deceased donor 54.4%. Initial immunosupression consisted mainly of tacrolimus (79.6%), mycophenolate sodium (49.7%) and steroids (92.2%). Induction: IL2RAb (81.8%) and ATG (4.7%). One year graft survival was 89.0% in DD and 92.5% in LD (p=0.01). There were 115 graft losses: acute rejection (n=15), recurrence of kidney disease (n=15), arterial or venous thrombosis (n=14), chronic allograft nephropathy (n=13), death with functioning kidney (n=13), and other causes or unknown (n=45). One year patient survival was 97.2%.From the 30 patient deaths, 16 were due to infection.

**Conclusions:** In Brazil, efforts are being made to provide an on-line entry of data in order to facilitate the report and provide an exchange of information, either among the participating centers as with other international registries.

## Abstract# 117

**Native Nephrectomy (Nx) in Children Prior to Kidney Transplant (KT)** F. Ghane Sharbaf,[1,2] L. Bell,[1] I. Gupta,[1] G.-P. Capolicchio,[3] M. Bitzan.[1] *[1]Pediatrics, McGill Univ, Montreal, QC, Canada; [2]Mashhad Univ of Med Sciences, Mashhad, Islamic Republic of Iran; [3]Urology, McGill Univ, Montreal, QC, Canada.*

**Objectives:** To review indications, complications, urine output (UO) & laboratory changes pre/post-Nx in a pediatric KT recipient cohort.

**Methods:** From our database (1992-2009), we identified all patients (pts) with uni- (U) or bilateral (B) native pre-KT Nx and extracted pertinent data (mean, SD, paired t-test).

**Results:** 82 kidneys were removed from 52 pts, age 8.5 yrs, 77% male (22 U, 30 B Nx; 75% retroperitoneoscopically). Main indications: polyuria (PU;13), proteinuria (PrU;13), PU&PrU (7), frequent UTI (9), hypertension (HTN;2), Denys Drash syndrome (2). 8/22 UNx were performed at KT, one 4 mo post KT. 12/30 with BNx had a 2-stage procedure. Complications were rare: peritoneal laceration requiring temporary PD to HD switch (1), subcutaneous emphysema (1), fluid overload/HTN (5), hyperkalemia (2). UTI decreased in 7/9. Table shows available UO & biochemical changes.

| Indication | Nx | N | | Pre | Post | P |
|---|---|---|---|---|---|---|
| Polyuria | U | 12 | ml/kg/h | 4.4 ± 2.6 | 2.9 ± 1.7 | 0.108 |
| | B | 5 | | 6.2 ± 3.9 | 0 | 0.007 |
| Proteinuria | U | 10 | Upr (mg/h/m²) | 188 ± 108 | 125 ± 90 | 0.172 |
| | B | 7 | | 356 ± 181 | 0 | 0.000 |
| | U | 10 | S alb (g/l) | 30 ± 10 | 34 ± 9 | 0.341 |
| | B | 7 | | 22 ± 5 | 36 ± 4 | 0.002 |

**Conclusions:** In this large cohort study, predominant Nx indications were PU and heavy PrU (62%). Complication rate was low. UNx alone was insufficient to decrease UO or PrU. Nx can be safely performed pre KT, even for PD pts, or in 2 stages before & at KT. Prospective studies are to determine risk/benefits for long-term graft function, particularly in young pts with high UO.

## Abstract# 118

**Is the Treatment of Chronic Antibody-Mediated Rejection with IVIg and Rituximab Successful?** L. Ghio,[1] M. Belingheri,[1] A. Edefonti,[1] M. Giani,[1] A. Innocente,[1] D. Cresseri,[1] A. Sementa,[2] G. Barbano,[2] A. Parodi,[2] A. Nocera,[2] F. Ginevri.[2] *[1]Pediatric Nephrology Unit – Organ and Tissue Transplantation Immunology - Nephrology Unit, Found. IRCCS Ca'Granda, Milano, MI, Italy; [2]Clinical and Experimental Transplantation Unit, G. Gaslini Institute, Genova, GE, Italy.*

**Objectives:** Chronic antibody-mediated rejection (CAMR) of renal transplant is now a defined nosologic entity. Some success has been described with the use of IVIg and Rituximab, but there is no established treatment protocol for this condition.

**Methods:** Herein we describe our experience with 12 young patients (18+5.6yrs) with CAMR treated with IVIg and Rituximab, associated to Plasmapheresis in two cases. The median follow up post-transplantation at diagnosis of CAMR was 14 months (10-27). All patients but one presented a GFR reduction. Transplant glomerulopathy and C4d positivity were present in all biopsies. Donor specific antibodies were positive in 6/12 patients.

**Results:** After the treatment only one patient improved GFR; in this patient the diagnosis of CAMR was performed after a protocol biopsy and without laboratory tests suggestive for GFR impairment. One patient had a progressive graft dysfunction, 5 needed dialysis treatment after 18 months and the last 5 maintained a stable renal function. At the end of follow up a mean GFR reduction of 8.4+9.2 ml/min was observed.

**Conclusions:** CAMR is confirmed to be a clinical condition with a severe outcome. The currently available treatments are not always able to reverse the CAMR. A better outcome is obtained when the treatment is well-timed and in patients with mild tubular and interstitial damages.

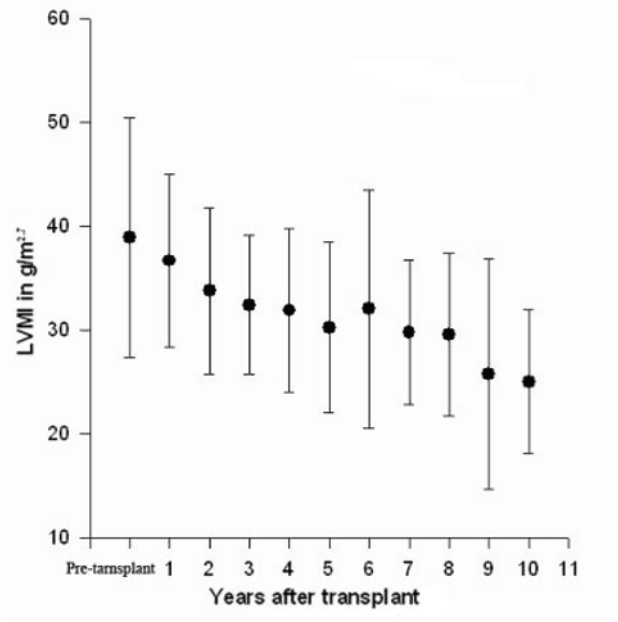
## Abstract# 119

**Effect of Renal Transplantation on Left Ventricular Mass in Children with End Stage Renal Disease** R. McLaughlin, L. Hamiwka, S. Samuel, D. Fruitman, S. Grisaru. *Pediatrics, University of Calgary, Calgary, AB, Canada.*

**Objectives:** Childhood end-stage renal disease (ESRD) is associated with an increased risk for early adulthood cardiovascular (CV) morbidity and mortality. Increased left ventricular mass (LVM) is an early indicator of this high risk. Previous studies suggested that in children LVM decreases after renal transplantation however, trends have been inconsistent. Our objective was to evaluate the longitudinal effect of transplantation on LVM, in children with ESRD.

**Methods:** In our centre, children routinely undergo echocardiograms prior to transplant and yearly thereafter. LVMI values from these measurements, starting pre-transplant and annually post-transplant, were averaged among included patients.

**Results:** Twenty seven children were followed for a median period of 5 years. Mean LVMI values graphically represented in the attached figure, decreased after renal transplant for up to a maximum of 10 years.




**Conclusions:** A sustained decline in LVMI following renal transplantation was observed in a cohort of children post-renal transplantation. We plan to investigate the correlation between blood pressure (BP) control and the observed decline in LVMI, however since BP targets in this cohort, before and after transplant, remained similar; we speculate that other factors associated with renal transplantation may have been responsible for this observation.

## Abstract# 120

**Steroid Free Immunosuppression in Asian Children – A Pilot Study** S. Gulati, P.N. Gupta, V. Kher, R. Ahlawat. *Neprhology, Fortis Institute of Renal Sciences adn Transplantation, New Delhi, Delhi, India.*

**Objectives:** The aim of this study was to assess the safety of steroid free protocol in Asian pediatric renal transplant recipients receiving Tacrolimus based immunosuppression.

**Methods:** We evaluated three children who received live related kidney transplant and were on steroid free (SF) protocol. All of them received 2 doses of IL2RA as induction. They were given IV Methylprednisolone in an initial dose 500 mg followed by oral prednisolone which was stopped on day 5. All of them received Tacrolimus (0.1 mg/kg) and MMF (1100 mg/m2) as maintenance immunosuppression. We compared these 3 children with three other children who received steroid based protocol during the same time period. The outcome measures that were evaluated were number of acute rejections, graft loss, PTDM, antihypertensive medications, S. Cholestrol, eGFR, height velocity and height SDS.

**Results:** The study group comprised of two boys and one girl. The mean age was 11.7 years and mean follow up was 14 months (5-24 months). We compared these 3 children (Group 1) with 3 age matched controls who were on steroid protocol (Group 2). None of these 6 children has any episode of acute rejection. We observed that the growth velocity in group 1 (6.8 cm/yr) was greater than group 2 (4.7 cm/yr). Similarly the improvement in Z-score was greater in group 1 as compared to group 2 (1.48 vs 1.07). The mean cholesterol level in group 1 was lower as compared to group 2 (149 mg/dl vs 248 mg/dl). None of the children in Gp 1 had PTDM while transient PTDM was seen in 1 child in Gp 2.

**Conclusions:** Complete steroid free immunosuppression is efficacious and safe in this selected group of children with no early clinical acute rejection episodes.

## Abstract# 121

**Early Proteinuria after Renal Transplantation and Allograft Outcomes** K. Gulleroglu,[1] E. Baskin,[1] U. Bayrakci,[1] E. Melek,[1] N. Cengiz,[1] S. Sevmis,[2] H. Karakayali,[2] M. Haberal.[2] *[1]Pediatric Nephrology, Baskent University, Ankara, Turkey; [2]General Surgery, Baskent University, Ankara, Turkey.*

**Objectives:** Posttransplant clinical management may be changed with early detection of graft damage. Late-onset proteinuria after renal transplantation has been associated with poor allograft outcomes. Relation between early proteinuria and posttransplant prognosis is not well understood.

**Methods:** Sixty-seven pediatric renal transplant recipients divided into 2 groups based on the 24-hour urine protein excretion during the 3rd posttransplant month. [Proteinuria≤ 4mg/m²/hour (group1, n=28), proteinuria >4mg/m²/hour (group2, n=39)]. The impact of early proteinuria on the various outcomes was performed.

**Results:** Mean age of recipients was 13,7±4,2 years old. Mean follow-up time after transplantation was 3,2±2,7 years. There was no significant difference for age, donor type, follow-up time between 2 groups. Early proteinuria was significantly higher for the cadaver donor group (mean 14,25mg/m$^2$/hour versus 7,08mg/m$^2$/hour, p=0,002). Median proteinuria of the rejection group (11,25 mg/m$^2$/hour, min-max:2,3-101,8) was significantly higher than these of non-rejection group (4,13mg/m$^2$/hour, min-max:0,13-39,42) (p<0,05). Significant positive correlation between early proteinuria and acute rejection was shown (r=0,34, p=0,005). Another positive correlation between early proteinuria and increased serum creatinine on 3rd year, as a marker of graft injury was shown (r=0,36, p=0,045). This kind of relationship was not valid for graft loss, GFR and immunosuppressive regiment.
**Conclusions:** Early proteinuria can be used for prediction of the posttransplant acute rejection and graft injury.

## Abstract# 122

**The First Case of MMF Administration to a Pediatric Renal Transplant Recipient with Partial HPRT Deficiency** R. Hamada,[1] H. Hataya,[1] S. Okamoto,[1] T. Sakai,[1] M. Muramatsu,[2] H. Satou,[2] Y. Hamasaki,[1] K. Ishikura,[1] M. Honda.[1] *[1]Nephrology, Tokyo Metropolitan Children's Medical Center, Tokyo, Japan; [2]Urology, Tokyo Metropolitan Children's Medical Center, Tokyo, Japan.*
**Objectives:** Background
Hypoxanthine-guanine phosphoribosyltransferase (HPRT) is an enzyme in the purine *salvage* pathway. Severe HPRT deficiency causes Lesch-Nyhan syndrome, while partial deficiency, also known as Kelley-Seegmiller syndrome, is associated with gouty arthritis, renal problems from nephrolithiasis, and rarely neurological dysfunction. Here, we report the first case of mycophenolate mofetil (MMF) administration to a pediatric renal transplant recipient with partial HPRT deficiency.
**Results:** Case
A 12-month-old boy presented with severe renal failure and hyperuricemia. Peritoneal dialysis was initiated. There were no signs of neurological or behavioral abnormalities. Enzymatic analysis was performed and partial HPRT deficiency (HPRT activity, 30% of normal) was diagnosed.
At the age of 4 years and 11 months, he underwent renal transplantation with immunoprophylactic basiliximab, MMF, cyclosporin A, and methylprednisolone. MMF was initiated 2 weeks prior to transplantation without producing adverse drug reactions possibly associated with reduced HPRT activity. Although MMF inhibits the *de novo* pathway of purine synthesis, no dose-limiting adverse drug reactions of MMF were detected and he was discharged on day 74.
**Conclusions:** To our knowledge, this is the first report of MMF treatment in a child with partial HPRT deficiency. Careful assessment of residual HPRT activity is recommended to ensure safe administration of MMF with low toxicity to patients with HPRT deficiency.

## Abstract# 123

**Renal Transplantation in Children Less Than 12 kg. A Single Center Experience** M. Herthelius, J. Sandberg, G. Tydén, U. Berg. *Department of Clinical Science, Intervention and Technology, Karolinska Institutet, Stockholm, Sweden.*
**Objectives:** To study underlying diseases, graft function and survival and patient survival and growth.
**Methods:** A cross-sectional analysis of 44 kidney transplants (24 boys and 20 girls) performed at the Karolinska University Hospital Huddinge 1980-2006 in children less than 12 kg. Mean age was 1.4 (0.4-3.7) years and mean weight was 9.4 (3.3-11.5) kg at transplantation. 35 (80%) patients received kidneys from a living donor and 9 (20%) from a deceased donor. All grafts were placed intra-abdominally. The immunosuppression was cyclosporine or tacrolimus, azathioprine or MMF, and prednisolone.
**Results:** The underlying diseases were obstructive uropathy (12 patients), dysplasia (5), congenital nephrotic syndrome (12), Drash syndrome (6), atypical HUS (1), ischemic events (5), others (3). 39 patients (89%) suffered from congenital disorders and 5 (11%) from acquired disorders. Graft function deteriorated from a mean GFR of 84 ml/min/1.73 m$^2$ three months after transplantation to 50 ml/min/1.73 m$^2$ within ten years. Ten year patient survival was 71% before 1992 and 97% after 1992. Ten year graft survival was 71% and 89% respectively before and after 1992. There was a rapid growth catch-up within the first years post transplant with a mean height of -2,89 SD at transplantation, -1,45 SD at 1 year and -0,83 SD at 3 years post-transplant respectively.
**Conclusions:** In small children, patient and graft survival are at least as good as in older ones. Renal function deteriorate during the first years post transplant but stabilize within a few years. In most children there is a substantial improvement of growth within the first years after transplantation.

## Abstract# 124

**Living Related Renal Transplantation in a 14 Year Old Boy with Factor H Antibody Associated aHUS** J. Hofer,[1] M. Riedl,[1] T. Jungraithmayr,[1] A. Rosales,[1] R. Würzner,[3] R. Mitkiene,[4] B. Pundziene,[4] M. Walter,[2] L.B Zimmerhackl.[1] *[1]Pediatrics, Medical Ubiversity, Innsbruck, Austria; [2]Operative Medizin, Medizinische Universität, Innsbruck, Austria; [3]Hygiene, Mikrobiologie und Sozialmedizin, Medizinische Universität, Innsbruck, Austria; [4]Pediatrics, Medical University, Kaunas, Lithuania.*
**Objectives:** A 12 year old boy was admitted to hospital on April 2007. Laboratory examination showed hemolytic anemia, thrombocytopenia and acute renal failure. The patient was diagnosed with Factor H antibody (FH-Ab) associated aHUS. The patient did not regain renal function. The initial FH-Ab level was high (1600 AU/ml) and even increased during the following months (1800 AU/ml). Under HD the titer dropped to 800 AU/ml. However, renal function did not recover and the patient required chronic dialysis, thus renal transplantation from a living donor (father) was planned. To reduce FH-Ab titers prior to kidney transplantation plasma exchange and a single infusion of i.v. IgG on the day before transplantation were planned. In 11/2009 the living related renal transplant was done. Induction therapy with ATG and continues immunosuppression with ATG, Tacrolimus, MMF and Steroids was done. 5 months after transplantation complement levels (C3, terminal complement complex) are normal, the FH-Ab titers are in the low range (<200).
**Conclusions:** At present, evidence based therapy recommendations for FH-Ab aHUS are missing. In this case report antibody titers dropped during frequent HD, but stayed clearly positive over a 2 year period. In our successfully transplanted patient FH-Ab titers dropped below 100 AU/ml and complement system remained normal.

## Abstract# 125

**How To Enhance Deceased Donor Kidney Transplantation (KT) in Children? – Importance of Education for Future Health Professionals** S. Hoshii, Y. Araki. *Pediatrics, Hokkaido National Medical Center, Sapporo, Hokkaido, Japan.*
**Objectives:** Because of severe organ shortage in Japan, 91% of KT in children is from living donors. To enhance organ donation, appropriate education is required for health professionals and the general public, especially the young. To learn how to educate, we surveyed young future health professionals' views on KT, compared with pediatric nephrologists' (PN).
**Methods:** Local medical students (MS, n=301), and nursing students (NS, n=305), and PN nationwide (n=239) answered questionnaires. PN were categorized; those caring for children with CKD (n=143) and those not (n=96). For comparison, $\chi^2$ test was used.
**Results:** More than 80% agreed on transplantation. Only 24%, 16% and 37% of MS, NS, and PN expressed their wishes in a donor card, higher than 8.8% in the general public. When asked if they accept their family member's wish to donate organs, less MS and NS accept, compared with PN (66% and 50 % vs. 80%). Only 27% and 32% of MS and NS, and 64% of PN had enough information about KT, and 85%, 93%, and 66% needed more. As for legal aspects of deceased donor, less MS and NS knew correctly compared with PN (43% and 15% vs. 73%). Only 34% and 10% of MS and NS thought KT could be successful, and 25% and 15% would chose deceased donors KT for their own renal replacement therapy if needed. More PN caring children with CKD knew legal aspects, and positive to preemptive KT, compared with those not (80% vs. 68% and 89% vs. 66%).
**Conclusions:** Because of limited information, future health professionals and even PN not treating CKD didn't have positive views on KT. These results indicate that more priority to education of future health professionals facilitate deceased donor KT.

## Abstract# 126

**Linear Growth in Renal Transplant Recipients Is Similar with Sirolimus vs. Calcineurin Inhibitors** L. Hymes, B. Warshaw. *Children's Healthcare of Atlanta and Emory University School of Medicine, Atlanta, GA, United States.*
**Objectives:** Sirolimus (SRL), potent macrolide immunosuppressant, can replace calcineurin inhibitors(CNI) for prevention of rejection and avoidance of CNI nephrotoxicity. However, animals studies indicate the SRL may inhibit linear growth. We compared height-z scores in a group of children maintained on SRL to patients receiving CNI to determine if SRL is detrimental to growth.
**Methods:** Records of patients who received renal transplants from 1999-2007 were retrospectively evaluated. The SRL and CNI groups were matched for age and gender among children with stable graft function, and compared for height-z scores, total mg of prednisone and eGFR (Schwartz equation) at 24 months post-transplantation.
**Results:** (Table) 20 children, age 1-15 years, were compared in each group. Initial height-z scores and eGFR were similar. At 24 mo, 9 children in each group displayed improved growth velocity. Height-z scores and cumulative doses of prednisone were similar. eGFR was higher in the SRL patients at 24 mo.

**Conclusions:** Linear growth in children on SRL was similar to those maintained on CNI. The potential inhibitory growth effects of SRL may be offset by avoidance of CNI nephrotoxicity and better graft function.

PATIENT DATA and RESULTS

| | CNI n=17 | SRL n=17 | p value |
|---|---|---|---|
| AGE years | 7 ± 5 | 7 ± 5 | ns |
| MALE (n) | 14 | 14 | ns |
| Initial HEIGHT- z scores | -2.2 ± 1.6 | -2.2 ± 1.3 | ns |
| 24 months HEIGHT- z scores | -1.7 ± 1.3 | -1.6 ± 1.2 | ns |
| IMPROVED z-scores (n) | 9 | 9 | ns |
| PREDNISONE total mg | 3045 ± 1000 | 3150 ± 1426 | ns |
| eGFR ml/min initial | 89 ± 19 | 88 ± 12 | ns |
| eGFR ml/min 24 mo | 85 ± 21 | 103 ± 23 | < .05 |

## Abstract# 127

**p16$^{INK4a}$ Expression in Zero Hour Biopsies Correlates with Renal Function 12 Months after Transplantation** C. Jacobi,[1] J. Schröder,[4] A. Bohlmann,[2] K. Theophile,[3] V. Bröcker,[3] O. Bock,[3] M. Wittau,[4] C. Brockschmidt,[4] A. Melk.[1] *[1]Pediatric Nephrology, Medical School, Hannover, Germany; [2]Nephrology, Medical School, Hannover, Germany; [3]Pathology, Medical School, Hannover, Germany; [4]Surgery, University Clinic, Ulm, Germany.*

**Objectives:** Somatic cellular senescence (SCS) results in reduced regenerative capacity, but does vary in kidneys with age. Since organ shortage forces us to accept older donor kidneys even for pediatric recipients, it would be helpful to develop tools to better assess the biological age of an organ.

**Methods:** 42 zero hour biopsies from adult renal transplantations were evaluated. Expression of SCS markers (p16 isoform1=p16i1, p16 isoform4=p16i4 and p21 measured by RT-PCR from formalin fixed tissue) were correlated with clinical data.

**Results:** Zero hour biopsies were derived from 6 living related donors and 36 deceased donors. Median follow up time was 24 months. Mean donor and recipient age was 54±15 yrs and 53±13 yrs. The expression of p16i1 was significantly higher in kidneys from donors that had smoked (p=.002) or had had cardio-/cerebrovascular diseases (p<.05). p16i1 expression significantly correlated with renal function calculated by MDRD at time of discharge from the hospital (r=.43, p=.007), at 3 (r=.46, p=.006), 6 (r=.39, p=.03) and 12 months (r=.44, p=.01) after renal transplantation. Neither p16i4 nor p21 correlated with any of the former clinical values.

**Conclusions:** Assessment of SCS markers allows further evaluation of donor kidneys and provides insight into renal function after transplantation. The use of such markers could facilitate chosing organs even in pediatric transplantation.

## Abstract# 128

**Mycophenolate Mofetil in Paediatric Renal Transplantation: Challenging Chests** H.E. Jones, M.I. McCulloch, J.O. Taylor. *Paediatric Nephrology, Evelina Children's Hospital, London, United Kingdom.*

**Objectives:** Mycophenolate mofetil (MMF) has been used in 50% (n=43) of our paediatric renal transplant patients. 12% of these patients have developed chronic respiratory symptoms requiring cessation of MMF.

**Methods:** Retrospective casenote review of patients who developed respiratory complications during treatment with MMF which required its withdrawal.

**Results:**

Chronic respiratory symptoms on MMF

| Case | 1 | 2 | 3 | 4 | 5 |
|---|---|---|---|---|---|
| Age, Sex | 12y9m, F | 12y4m, M | 11y7m, M | 11y4m, M | 8y, M |
| Diagnosis | Dysplasia | Congenital nephrotic | Congenital nephrotic | Dysplasia | FSGS, asthma |
| Age transplanted | 2y6m | 3y11m | 2 9m | 2y4m | 4y4m |
| Age MMF started | 9y2m | 4y5m | 6y6m | 7y8m | 4y4m |
| Reason for MMF | Sirolimus intolerance | Rejection | CNI toxicity | CNI toxicity | Rejection |
| Clinical Picture | Chronic sinusitis, persistent cough | Recurrent pneumonia | Recurrent pneumonia | Recurrent pneumonia | Recurrent pneumonia |
| Radiological fndings | Maxillary sinusitis | Bronchiectasis | Pulmonary PTLD | Bronchiectasis | Bronchiectasis |
| Bronchoscopy | N/A | No | Yes | Yes | Yes |
| Antibiotics* | 1, 2 | 1,2 | 1,3,4,5,6,7 | 1,2 | 1,2,3,4,7 |
| Time from starting MMF to symptoms developing | 2m | 4m | 1m | 2y1m | 9m |
| Duration of MMF | 1y9m | 5y5m | 10m | 2y3m | 2y11m |
| Current status | Clinically well | Ongoing infections | Lost graft | Chest improved | Recurrent infections |

*1=co-amoxiclav, 2=azithromycin, 3=cephalosporin, 4=co-trimoxazole, 5=ciprofloxacin, 6=gentamicin, 7=amphotericin

**Conclusions:** More potent immunosuppressive agents have improved graft outcomes but led to an increase in infections in the paediatric renal transplant population. Our 5 patients failed to respond to conventional antibiotic therapy for recurrent or persistent respiratory infections that required extensive investigation, prolonged antimicrobial treatment, and MMF withdrawal.

Tubulointerstitial Disorders

## Abstract# 129

**(O-17)**

**The Protective Effects of Mizoribine on Cyclosporine A Nephropathy in Rats** S. Hara,[1] D. Umino,[1] T. Someya,[1] S. Fujinaga,[1] H. Murakami,[2] Y. Ohtomo.[1] *[1]Pediatrics, Juntendo University Faculty of Medicine, Tokyo, Japan; [2]Pathology, Saitama Children's Medical Center, Saitama, Japan.*

**Objectives:** The therapeutic benefits of Cyclosporine A (CsA) are often limited by the chronic nephrotoxicity of its long-term use.Chronic nephrotoxicity is manifested by renal function impairment and progressive histopathological kidney lesions characterized by tubular vacuolization, tubular necrosis, interstitial fibrosis, and afferent arteriolopathy. This study tested the hypothesis that the concurrent administration of Mizoribine (MZR) may improve chronic CsA nephrotoxicity.

**Methods:** Sprague-Dawley male rats were divided into the following four groups: group 1, control (n = 6); group 2, treated with CsA alone (n = 5); group 3, treated with CsA and MZR (n = 4); and group 4, treated with MZR alone (n = 6). The anti-inflammatory and antifibrotic effects of MZR were studied by evaluating the concentrations of the inflammatory mediator, osteopontin, renal function, and histopathology. The interstitial fibrosis was stained blue with Elastica-Massontrichrome and the sections were quantified.

**Results:** The CsA-treated rats showed decreased renal function and increased histologic parameters in comparison with the control rats and also showed significantly increased interstitial fibrosis area and macrophage in comparison with the control rats. The CsA MZR treatment significantly improved the interstitial fibrosis area and macrophage in comparison with the CsA-treated rats.

**Conclusions:** On the basis of these findings, we suggest MZR effectively attenuates renal macrophage accumulation and the progression of interstitial fibrosis.

## Abstract# 130

**(O-18)**

**Nephrocystin-4 Modulates the Phosphorylation State and Regulates the Subcellular Localisation of Nephrocystin-1** M.C. Liebau,[1] K. Hoepker,[1] F. Fabretti,[1] S. Zank,[1] B. Schairer,[1] I. Schmedding,[1] H. Zentgraf,[2] G. Walz,[3] T. Benzing,[1] B. Schermer.[1] *[1]Department of Nephrology, University Hospital, Cologne, Germany; [2]German Cancer Research Center, Heidelberg, Germany; [3]Department of Nephrology, University Hospital, Freiburg, Germany.*

**Objectives:** Nephronophthisis (NPHP) is the most common genetic cause of ESRD in children and adolescents. The most frequently affected gene is NPHP1 encoding for the ciliary protein nephrocystin-1. Mutations of the NPHP4 gene encoding for nephrocystin-4 account for a smaller proportion of NPHP cases, yet the clinical presentation of patients with NPHP1 or NPHP4 mutations is quite similar. The cellular function and regulation of the proteins nephrocystin-1 and nephrocystin-4 remain poorly understood.

We have recently shown that nephrocystin-1 interacts with the focal adhesion kinase Pyk2. Here we show that Pyk2 induces phosphorylation of nephrocystin-1 at three defined tyrosine residues and that this phosphorylation regulates binding of nephrocystin-1 to the trans-golgi sorting protein PACS-1. Interestingly, fullength nephrocystin-4, but not different patient mutations, abolishes the Pyk2-induced tyrosine phosphorylation of nephrocystin-1. Knockdown of nephrocystin-4 leads to changes of the subcellular localisation of nephrocystin-1 in ciliated epithelial cells.

Our data suggest a model in which nephrocystin-4 acts upstream of nephrocystin-1 and modulates its tyrosine phosphorylation state. Thus nephrocystin-4 dynamically regulates the composition of the nephrocystin protein complex and defines its subcellular localisation.

## Abstract# 131

**(O-19)**

**Clinical Situation of Patients wth Cystinosis in Germany** K. Latta, A. Latta, C. Selbsthilfe e.V. *Clementine Kinderhospital, Frankfurt, Germany.*

**Objectives:** It was the aim to assess the current situation of cystinosis patients in Germany.

**Methods:** A questionnaire was sent to all patients of the German cystinosis group (n=89). 59 responded.

**Results:** There were 23 adults, and the children had a mean age of 10.9 years. In 39, diagnosis was made before age of two, while 7 were diagnosed beyond 3 years of age. Cysteamine treatment was started at over 2 years of age in 23 patients, and in 15 even after the age of 6 years. Glycosuria and Fanconi´s syndrome are almost universally present. Only 10 patients have a normal GFR with their native kidneys. 3 patients are on dialysis, 20 are successfully

transplanted. While 15/30 beyond age 15 have a reduced height, only 6/25 children (4 girls, 2 boys) have a height <-2SD for age.
Visual acuity is reduced in 10/58, 37 are photophobic. 16 suffer from hypothyroidism, 4 have pulmonary dysfunction, 2 diabetes, and 2 pancreatic insufficiency respectively. 33 patients developed orthopedic problems, 8 difficulties in swallowing, and 18 muscle weakness. 17 participate regularly in sports activity.
46 have a legally recognized degree of disability. Most of the patients attend(ed) school at normal age. While 17/19 pursue professional education or are professionally active, in 2 such activity is no longer possible.
**Conclusions:** This is the first attempt to describe the clinical situation of cystinosis patients in Germany. While none of the children less than 10 years is on dialysis, all adults have reached terminal kidney failure. Disability is much more pronounced in adults, most likely reflecting the lack or late start of cystine depleting treatment in infancy. However, even with early treatment marked signs of the disease are present in everyday life.

## Abstract# 132
**(O-20)**

**Cysteamine Toxicity in Cystinosis Patients** M.T. Besouw,[1] L.P. van den Heuvel,[1] J.P. Dutertre,[2] A. Awan,[3] W.G. van 't Hoff,[4] M.A. Cornelissen,[5] F. Emma,[6] E.N. Levtchenko.[1] *[1]Pediatrics, University Hospitals, Leuven, Belgium; [2]Orphan Europe, Paris, France; [3]Nephrology, Children's University Hospital, Dublin, Ireland; [4]Nephro-Urology, GOS Hospital, London, United Kingdom; [5]Pediatrics, RUMC, Nijmegen, Netherlands; [6]Nephrology & Urology, Bambino Gesù Children's Hospital, Rome, Italy.*

**Objectives:** Cystinosis is an autosomal recessive disease, caused by intralysosomal cystine accumulation. It initially affects kidneys followed by eyes, endocrine organs and neuro-muscular system. Cysteamine preserves renal function and postpones extra-renal complications. Recently 8 European patients were reported with muscular-skeletal weakness, skin striae and bruising-like lesion on elbows. One patient deceased.
**Methods:** Detailed information was obtained from physicians and/or Orphan Europe.
**Results:** WBC cystine levels were below 1 nmol ½cystine/mg protein in all patients, indicating compliance with cysteamine therapy. Daily cysteamine dose was within the previously recommended range (60-90 mg/kg) in 7 patients, but above the currently recommended max dose (1.9g/m$^2$) in 5 patients. Skin biopsies in 6 patients showed angioendotheliomatosis with randomly arrangement of the collagen fibres. One patient died from cerebral ischemia, brain imaging showed acute ischemia and extensive vascular abnormalities, no autopsy was performed.
**Conclusions:** We present 8 cystinosis patients who developed adverse reactions of cysteamine that were not reported thus far. Most patients initially improved after dose reduction, indicating a causal relation between cysteamine administration and these lesions. The mechanisms of these adverse events are unknown and require further investigation.
**DISCLOSURE:** *Dutertre, J.P.*: Other, Personnel of Orphan Europe.

## Abstract# 133

**Renal Tubular Dysfunction in Pediatric Patients with beta-Thalassemia Major** A. Ahmadzadeh, S. Assar. *Pediatrics, Ahvaz Jundishapour University of Medical Sciences, Ahvaz, Khuzestan, Islamic Republic of Iran.*

**Objectives:** The aim was to evaluate the prevalence of renal tubular dysfunction in children with β-T major, referring to Shafa hospital.
**Methods:** From May 2007 to April 2008), glomerular and tubular function were assessed in 140 normal and 140 children(7-16 years) with of β-T major. The morning samples were collected from each patient and analyzed for electrolytes, protein, uric acid (UA), creatinine and urinary N-acetyl-β- D- glucosaminidase activity (UNAG). Blood samples were also collected for serum creatinine (SCr), electrolytes and ferritin before transfusion. Data were evaluated by SPSS software. A $P$ value <0.05 was considered to be significant.
**Results:** Among 140 patients 72 were males. The mean age was 11.5 years. All of them had a normal SCr level.The mean UNAG in the patients was 17.8 IU/L (normal 0.15-11.5 IU/L) whereas in control group was 3.2 IU/L ($P$ < 0.001). No body in control group had an abnormal UNAG whereas 82 (52.6%) of the patients had an elevated UNAG. Of the 82 patients, 58 (62.4%) had also a high blood level of ferritin ($P$ <0.001),13 (15.9%) had hypercalciuria (UCa/UCr> 0.21) ($P$ : 0.006)and 9 (6.4%) had proteinuria. Sixty-nine (49.3%) of the 140 patients and 45 (65.2%) of patients having UNAG had also uricosuria. Ten (7%) patients had microscopic hematuria.
**Conclusions:** Although glomerular involvement is not a frequent complication in patients with β-T major, tubular dysfunction is relative a common complication of the disease. Nevertheless, this abnormality is not detected by routine biochemical tests. Although, UNAG is the best to detect renal tubular dysfunction in these patients, currently at least periodic measurement of UCa/Ucr and U UA/Ucr ratios are recommended.

## Abstract# 134

**A Case with Bartter Syndrome Accompanying Severe Growth Hormone Deficiency and Focal Segmental Glomerulosclerosis** I. Akil, S. Ozen, A.R. Kandiloglu, B. Ersoy. *Pediatric Nephrology, Celal Bayar University, Manisa, Turkey; Pediatrics, Celal Bayar University, Manisa, Turkey; Pathology, Celal Bayar University, Manisa, Turkey; Pediatric Endocrinology, Celal Bayar University, Manisa, Turkey.*

**Objectives:** Ten year old male child has severe growth retardation (height standard deviation score: -8.15).
**Methods:** He has thin triangular face, prominent ears and forehead, big eyes. Megacystis, bilateral hydroureteronephrosis and residual urine were detected in ultrasonography, but there was no vesicoureteral reflux. Lumbo-sacral MR showed posterior disc bulging at L4-5 level. Serum sodium and chloride levels were normal, but mild hypokalemia was overlooked at the beginning.
**Results:** In follow-up, hypokalemic hypochloremic metabolic alkalosis developed; there were high urinary chloride and potassium excretion (52 mEq/L and 43 mEq/L respectively). The patient with renal salt losing was thought that he might have classic Bartter syndrome due to absence of nephrocalcinosis, presence of persistent hypercalciuria and sensorineural deafness and presence of relatively mild clinical and laboratory findings except polyuria at the beginning. The child was treated with indomethacine, sprinolactone and oral potassium besides growth hormone. By the use of this treatment he had considerable increase in his weight and height compared to growth hormone therapy period.
**Conclusions:** We presented this case because, although growth retardation is a major feature of Bartter syndrome, associated GH deficiency is reported rarely in the literature; proteinuria associated with focal segmental glomerolosclerosis responded to the treatment of Bartter syndrome.

## Abstract# 135

**Omeprazole Induced Acute Interstitial Nephritis in Paediatrics – A Serious Side Effect** D. Noone,[1] O. Murwan,[1] T. Dorman,[2] M. Riordan,[1] A. Awan.[1] *[1]Nephrology, Children's University Hospital, Temple Street, Dublin, Ireland; [2]Renal Pathology, Beaumont Hospital, Dublin, Ireland.*

**Objectives:** Drug induced Acute Interstitial Nephritis (AIN) is common cause of acute renal failure and is reversible in half of cases. Proton Pump Inhibitors (PPIs) are aetiological agent in most cases of AIN in adults. We report two cases of adolescents with biopsy proven AIN due to Omeprazole. To our knowledge there is no published cases of PPI related AIN in paediatrics.
**Methods:** Case1 A15 year old adolescent girl presented with 3-week history of abdominal pain, anorexia and vomiting. She had acute renal failure, serum creatinine 974µmol/L (range 27-60) and urinary protein of 1046mg/l. She had been taking Omeprazole 20mg daily for three weeks. Biopsy confirmed AIN. Omeprazole was discontinued and treated with steroids for six weeks. Six months later she has mild renal impairment Cr 94µmol/L and minimal proteinuria.
Case 2 A15 year old male transplant recipient was given Omeprazole at transplantation. An allograft biopsy because of delayed graft function on day 8 showed mild acute tubular necrosis but also, severe tubulointerstitial inflammation. He was not taking Trimethoprim-sulfamethoxazole,other agent associated with AIN especially in renal allograft.No features of antibody-mediated rejection or Tacrolimus toxicity were present AIN resolved with pulse steroid therapy and cessation of Omeprazole.
**Conclusions:** Use of PPIs for Gastro Oesophageal Reflux Disease in paediatrics is widespread. We present two cases of AIN in adolescents, in a native kidney and a renal allograft. This rare but severe adverse event needs to be considered early and offending agent withdrawn to prevent permanent renal damage.

## Abstract# 136

**Isolated Proteinuria and Bartter Syndrome as Initial Presentation of Nephropathic Cystinosis** V.M.S. Belangero, T.V.R. Valim, L.C. Prates, A.C.G. Britto. *Pediatrics (Pediatric Nephrology Unit), State University of Campinas, Campinas, Sao Paulo, Brazil.*

**Objectives:** To present rare initial manifestations of Nephropathic Cystinosis
**Methods:** Description of two children (brothers) that had as initial presentation of Cystinosis only proteinuria. The former had diagnosis of Nephrotic Syndrome. His brother has non-nephrotic proteinuria as the only sign of Cystinosis.
Both presented cystine crystals on the slit-lamp. The third case had his first presentation as a Bartter Syndrome.

**Results:**

| Cases | Patient 1 | Patient 2 | Patient 3 |
|---|---|---|---|
| History | No symptons untill 5 years old | No symptons untill now | Failure of growth |
| Age on first problem | 5 yo | 5 yo | one yo |
| First evaluation here | 8 yo | 5 yo | 2,5 yo |
| Urine density | 1024 | 1023 | 1007 |
| Proteinuria | ++++ | ++ | ++ |
| Glucose urine | + | negative | negative |
| Metabolic acidosis | No | No | No |
| Metabolic alkalosis | No | No | Yes |
| Phosphatemia | 2,5 mg% | 3,9 mg% | 4,0 mg% |
| Natremia | 139 mEq/L | 133 | 135 |
| Kalemia | 3,4 mEq/L | 4,3 | 2,8 |
| Renal function | 35 m//min/1,73m2 | 93 | 25 |
| Slit-Lamp | Cystine crystals | Cystine crystals | Cystine crystals |

**Conclusions:** The etiologic investigation of isolated proteinuria is difficult. The slit-lamp evaluation must be included since can early give a diagnostic of Nephropathic Cystinosis.

## Abstract# 137

**Growth Hormone Deficiency in Bartter's Syndrome; Three Cases**

B. Demircioglu Kilic,[1] Y. Kor,[2] M. Buyukcelik,[1] M. Keskin,[2] A. Balat.[1] *[1]Pediatric Nephrology, Gaziantep University, Gaziantep, Turkey; [2]Pediatric Endocrinology, Gaziantep University, Gaziantep, Turkey.*

**Objectives:** Bartter's syndrome is a rare autosomal recessive, salt-losing disorder characterized by hypokalemia, hyponatremia, metabolic alkalosis, hyperaldosteronizm, hyperreninemia, and normal blood pressure. Although growth retardation is one of the major features of Bartter syndrome, associated growth hormone (GH) deficiency is rarely reported in the literature.

**Methods:** Herein, we report three patients with Bartter's syndrome having severe growth retardation.

**Results:** Their heights were under the 3th percentile compared with same aged children. Growth hormone levels were 0.068 ng/mL, 0,139 ng/mL, and 0,112 ng/mL respectively, and all under normal range (>0.3 ng/mL). Growth hormone stimulation test revealed growth hormone deficiency. The growth hormone levels in 60th and 90th minutes following L-dopa stimulation test were 4, 7 ng/mL (N: >10 ng/mL) and 1, 2 ng/mL (N: >10 ng/mL) in the first patient, 0.5 ng/mL and 0, 5 ng/mL in the second, and 0,7 ng/mL and 0.2 ng/mL in the third patients. Clonidine stimulation test showed similar results as in L-dopa test; 1, 39 ng/mL and 1.79 ng/mL in the first, 4, 49 ng/mL and 3, 66 ng/mL in the second, and 1, 64 ng/mL and 1,96 ng/mL in the third patients.

**Conclusions:** We suggest that children with Bartter syndrome may have growth retardation because of GH deficiency. Therefore, GH deficiency should be noted especially in the presence of severe growth retardation during childhood despite of effective treatment of this syndrome.

## Abstract# 138

**Bi-Nephrectomy as a Treatment for Inherited Tubulopathy**

A.A.D. Chen, A.A.G. Garnier, K.K.B. Brochard, F.F.B. Bandin, C.C.P. Pajot, P.P.G. Galinier, L.L.P. Percheron, S.S.D. Decramer. *Pediatric Nephrology,Internal Medecine and Hypertension, Centre de Référence des Maladies Renales Rares, Children Hospital, Toulouse, France; Children Hospital, Toulouse, France; Pediatric Nephrology, Internal Medecine and Hypertension, Children Hospital, Toulouse, France; Children Hospital, Toulouse, France; Children Hospital, Toulouse, France; Children Hospital, Toulouse, France; Children Hospital, Toulouse, France; Children Hospital, Toulouse, France.*

**Objectives:** First described case of renal tubulopathy requiring bi-nephrectomy.

**Methods:** We report the case of a three years old girl that had been treated since the age of two for a proximal tubulopathy in relation to a Pearson-like syndrome. Severe electrolytical abnormalities were noted (hypokaliema 2mmol/l, metabolic acidosis with bicarbonatemia 6 mmol/l, hypophosphoremia 0.3 mmol/l). Enteral nutrition was needed due to recurrent vomiting. Digestive's involvement secondary occurred (recurrent nauseas and chronic pancreatitis) requiring exclusive parenteral nutrition. No improvement was seen despite daily administration of 12 mEq/kg potassium, 22 mEq/kg of bicarbonates and supportive treatment. Adjunction of treatment by IEC failed to reduce polyuria. Ponderal stagnation, and developpement of rachitism led to unilateral nephrectomy which didn't change the course of the disease.

**Results:** Normalisation of biological disturbances occured following bi-nephrectomy and hemodialysis

**Conclusions:** Proximal tubulopathy might sometimes be refractory and radical surgery can be mandatory.

## Abstract# 139

**Fanconi Syndrome Due to Deferasirox in a Child with Aplastic Anemia**

A. Endo, T. Someya, S. Fujinaga, Y. Ohtomo, T. Shimizu. *Department of Pediatrics, JUNTENDO University Faculty of Medicine, Bunkyo-ku, Tokyo, Japan.*

**Objectives:** [Background] Deferasirox (DFX) is a novel oral iron chelator developed for the treatment of chronic iron overload due to blood transfusions. DFX can be administered once daily and orally, resulting in better acceptance in patients. However, preliminary data have suggested that renal toxicity may be a major issue in this drug. Although fatal, acute, irreversible renal failure in adult patients has been described in post marketing reports, there is no such reports in children treated with DFX.

**Results:** [Case]We report here on a case of Fanconi syndrome due to DFX in a 9 year-old boy with aplastic anemia. Aplastic anemia was diagnosed at the age of 3. Since 6 years old, he has been on monthly transfusion programs. After a year later, his serum ferritin level has begun to increase, suggesting chronic iron overload due to frequent blood transfusions. DFX was started at the age of 8. He responded well to the therapy. However, one year after the start of DFX, clinical symptoms such as malaise and appetite loss developed. Laboratory evaluations demonstrated the presence of pan-aminoaciduria and hypo-kalemic/phosphatemic hyper-potassiuria/phosphaturia. Based on these findings, he was diagnosed as Fanconi syndrome. Therefore, DFX was immediately discontinued. Several months later, his clinical symptoms and abnormal laboratory data were improved.

**Conclusions:** [Discussion]To our current knowledge, this is the first pediatric case of the Fanconi syndrome due to DFX. Since the use of DFX in children has been popular, careful monitoring of kidney function should be necessary in those patients.

## Abstract# 140

**Epidemiology of Hantavirus Infections in Pediatric Patients Compared to Adults in Germany in the Last 10 Years**

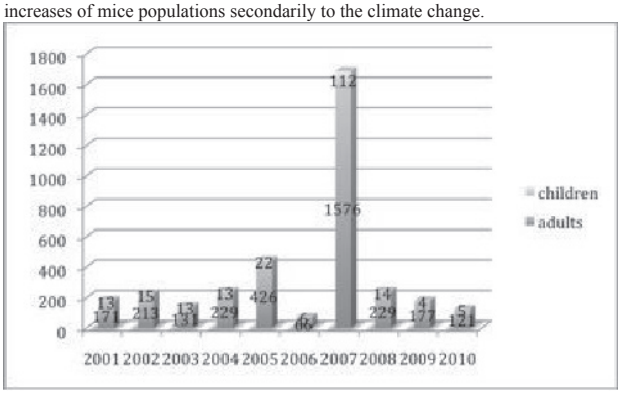
R. Goldwasser,[1] M. Zeier,[1] K. Stark,[2] B. Tönshoff.[1] *[1]University Children's Hospital, Heidelberg, Germany; [2]Infections and Pathogens, Robert Koch Institute, Berlin, Germany.*

**Objectives:** Infection with hantaviruses is associated with significant morbidity and mortality worldwide, causing beside others the hemorrhagic fever with renal failure. Hantavirus infection is acquired by inhalation of aerosolized virus containing particles from infected mice. Data on the epidemiology in pediatric patients are scarce.

**Methods:** We therefore analyzed data on Hantavirus infections in pediatric and adult patients in Germany reported to the German Institute for Infectious Disease Epidemiology (Robert Koch-Institute in Berlin) from 2001 to mid February 2010.

**Results:** The reported overall incidence of Hantavirus infections varied significantly between 72 cases in 2006 and 1688 cases in 2007 (Figure). The proportion of pediatric patients ranged from 2.2% in 2009 to 9% in 2003, being significantly lower than expected (20% of the entire population in Germany belongs to the pediatric age group). 76% of pediatric cases occurred in the age group 15-19 yrs, although individual cases in toddlers have been reported. In 2007 there was an Outbreak of Hantavirus infection.

**Conclusions:** Hantavirus infections in Germany affect mainly adult patients and adolescents. Outbreaks as observed in 2007 are most likely due to regional increases of mice populations secondarily to the climate change.



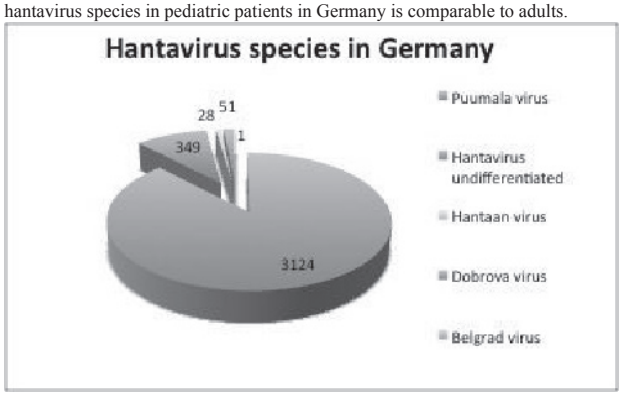

## Abstract# 141

**Prevalence of Different Hantavirus Species in Children and Adults in Germany in the Last 10 Years**

R. Goldwasser,[1] M. Zeier,[1] K. Stark,[2] B. Tönshoff.[1] *[1]University Children's Hospital, Heidelberg, Germany; [2]Infections and Pathogens, Robert Koch Institute, Berlin, Germany.*

**Objectives:** Hantavirus infection may lead to life threatening diseases such as the hemorrhagic fever with renal syndrome (HFRS) or the hantavirus cardiopulmonary

syndrome (HCPS) with high morbidity rate and renal failure. Little is known on the epidemiology of different hantavirus species in the pediatric population in Germany.
**Methods:** We analyzed data on hantavirus infections in general and the various species in adult and pediatric patients in Germany reported to the German Institute for Infectious Disease (Robert Koch-Institute in Berlin) from 2001 to February 2010.
**Results:** Infections with the Puumala virus accounted for the majority of hantavirus infections both in adult (88%, Figure) and pediatric patients (91%). Other detected virus species comprised Dobrova virus (adult 1.4%, pediatric 1.8%), Hantaan virus (adult 0.8%, pediatric 0.9%) and Belgrad virus (adult 0.03%, none pediatric). All these virus species are associated with HFRS. Andes virus infections, the only hantavirus transmitted by person-to-person contact, as well as species causing the HCPS did not occur.
**Conclusions:** The hantavirus species most frequently observed in Germany is the Puumala virus causing HFRS via contact to mice. The distribution of various hantavirus species in pediatric patients in Germany is comparable to adults.




## Abstract# 142

**Familial Hypomagnesaemia with Hypercalciuria and Nephrocalcinosis (FHHNC) Due to Claudin 19 (CLDN19) Gene Deletion in an Asian Child** S. Hegde, C.R. Iyer, R. Krishnan. *Department of Paediatric Nephrology, University Hospital of Wales, Cardiff, South Wales, United Kingdom.*
**Objectives:** Familial hypomagnesaemia with hypercalciuria and nephrocalcinosis due to mutations in CLDN19 has been described in Swiss and Spanish/Hispanic families. We describe this condition in an 11 years old Asian boy born to consanguineous parents.
**Methods:** He was well until 3.5 months of age when nephrocalcinosis and hypercalciuria was noted following work up for an episode of UTI. He was not acidotic, hypocalcaemic or hypokalaemic and his renal function was normal. Hypomagnesaemia, noted during second year and high myopia was detected at three years of age.
**Results:** Claudin 16 mutation analysis at 5 years of age was negative. Further screening showed homozygous deletion of exons 1-4 in CLDN19, which lead to complete loss of function. Hypercalciuria, nephrocalcinosis and hypomagnesemia persist despite appropriate therapy but renal function has remained stable.
**Conclusions:** FHHNC due to CLDN19 mutation has not been described in Asian children. Claudins are the major components of renal tight junctions and contribute to epithelial barrier function by restricting free diffusion of solutes through the paracellular pathway. Claudin 19 is expressed in retina in addition to renal tubules and hence it is associated with various eye changes like macular colobomata, significant myopia, and horizontal nystagmus. Mutations in the Claudin genes result in a progressive renal calcium and magnesium wasting leading to chronic kidney disease at variable rate. Renal impairment is thought to be due to nephrocalcinosis and some other unidentified factors.

## Abstract# 143

**Clinical and Genetic Analysis of Dent's Disease in 6 Chinese Children with Low Molecular Weight Proteinuria** B. Zhu, P. Li, J. Huang. *Peking University first Hospital, Beijing, China.*
**Objectives:** Dent's disease is an X-linked proximal tubulopathy characterized by low molecular weight proteinuria, hypercalciuria, nephrocalcinosis, nephrolithiasis, and renal failure. To date, two responsible genes for the development of Dent disease have been identified: CLCN5 and OCRL1. The purpose of this study was to investigate the phenotypes and genotypes of six Chinese boys with Dent's disease.
**Methods:** six patients from six unrelated families were studied. Genomic DNA was extracted from peripheral white blood cells using a simple salting out procedure after informed consent. Thirteen pairs of primers were used to amplify all coding exons and exon–intron boundaries of the *CLCN5* gene by polymerase chain reaction (PCR). All PCR products were sequenced directly on an autosequencer.
**Results:** Low molecular weight proteinuria and hypercalciuria were found in all patients, nephrocalcinosis in four patients, hematuria in three patients, hypophosphataemia in three patients and rickets in one patient. This study also describes a case of Dent's disease due to a missense mutation of CLCN5 gene S244L, associated with a Bartter-like syndrome that is characterized by hypokalaemic metabolic alkalosis and hyper-reninaemic hyperaldosteronism. six mutations of the CLCN5 gene were revealed, including L594fsX595,R637X, R467X,IVS4-2A>G,S244L and V505G. The mutation IVS4-2A>G and V505G was never reported before.
**Conclusions:** Low molecular weight proteinuria and hypercalciuria were the main clinical features of the six Chinese boys with Dent's disease. Dent's disease could be associated with a Bartter-like syndrome, which make the gene diagnosis more important.

## Abstract# 144

**Clinical Characteristics of Genetically-Proven Gitelman's Syndrome** H. Kaito,[1] K. Nozu,[1] Y. Hashimura,[1] M. Oka,[1] T. Ninchoji,[1] K. Nakanishi,[2] N. Yoshikawa,[2] K. Iijima,[1] M. Matsuo.[1] *[1]Pediatrics, Kobe University Graduate School of Medicine, Kobe, Hyogo, Japan; [2]Pediatrics, Wakayama Medical University, Wakayama, Wakayama, Japan.*
**Objectives:** Gitelman's syndrome (GS), recognized as a variant of Bartter's syndrome (BS), is a salt-losing tubulopathy which is characterized by hypokalemic metabolic alkalosis with hypomagnesemia and hypocalciuria. Hypomagnesemia and hypocalciuria with no growth impairment has been believed to allow differentiation between GS and BS. GS is associated with inactivating mutations in *SLC12A3* gene and it is becoming clearer that diagnostic confirmations by only clinical manifestations often lead to misdiagnosis. We herein investigated the clinical characteristics of genetically-proven GS patients.
**Methods:** We retrospectively analyzed clinical data from GS patients with homozygous or compound heterozygous mutations in *SLC12A3* gene. Serum potassium and magnesium levels were investigated. Urinary excretion of calcium was determined by morning urinary calcium-to-creatinine ratio.
**Results:** Thiry-one patients were investigated. At their first visit, 71% patients were below average in body height, and 41% of them fell below -2SD. All the patients had below normal for serum potassium levels, while 26% patients were thoroughly within normal limits for serum magnesium levels. Morning urinary calcium-to-creatinine ratio was as low as $0.03 \pm 0.04$ g/gCre.
**Conclusions:** Our study revealed that it was not unusual for GS patients to show severe growth impairment, and what was more noteworthy was that many patients had no overt hypomagnesemia. These findings suggest that it is essential for definite diagnosis of GS to perform genetic analysis.

## Abstract# 145

**Albumin Induced Production of Proinflammatory Substances in Cystinotic Proximal Tubular Cells** M.J. Wilmer,[1,2] T. Velden van der,[1] L.A. Monnens,[3] L.P. Heuvel, van den,[1,2] E.N. Levtchenko.[2] *[1]Department of Pediatrics, Radboud University Nijmegen Medical Centre, Nijmegen, Netherlands; [2]Department of Pediatric Nephrology, University Hospital Leuven, Leuven, Belgium; [3]Department of Physiology, Radboud University Nijmegen Medical Centre, Nijmegen, Netherlands.*
**Objectives:** Lysosomal cystine accumulation is the hallmark of the autosomal recessive disorder cystinosis. Patients usually develop Fanconi syndrome in the first year of life leading to progressive renal damage. Whehter tubular chemokine and cytokine production is involved in the pathogenesis of cystinosis is investigated.
**Methods:** Conditionally immortalized proximal tubular cell lines (ciPTEC) of controls (n=4) and cystinotic patients (n=8) were developed by transfection with SV40T and hTERT. Production of IL-8, MCP-1 and TGF-β1 were measured after incubation with bovine serum albumin (BSA; range 20-500μg/ml) using ELISA and corrected for intracellular protein. Urinary levels of IL-8, MCP-1 and RANTES were measured and corrected for creatinine in cystinotic patients (n=11) and controls (n=6). Data are presented as pg/μmol creatinine.
**Results:** In control and cystinotic ciPTEC, BSA dependent IL-8, MCP-1 and TGF-β1 production was demonstrated. No alterations between the two groups were observed at basal or stimulated conditions. IL-8 (control 1.5 versus cystinosis 7.5), MCP-1 (10.2 versus 107.9) and RANTES (0.7 versus 4.0) were increased in cystinotic urine ($p<0.05$).
**Conclusions:** Cystinotic ciPTEC are susceptible to albumin induced production of proinflammatory substances comparable to control cells. Because our previous studies showed glomerular proteinuria in cystinosis, treatment targeting RAAS inhibition is indicated.

## Abstract# 146

**Medullary Nephrocalcinosis in an Infant with Incomplete Distal Renal Tubular Acidosis and Idiopathic Hypercalciuria: A Case Report** S. Lazova, M.I. Lilova. *Department of Pediatrics, Tokuda Hopsital, Sofia, Bulgaria.*

**Objectives:** Nephrocalcinosis (NK) is common complication in incomplete distal renal tubular acidosis (idRTA), which is characterized with an absence of metabolic acidosis, hypocitraturia and alkaline urine pH. Idiopathic hypercalciuria(IH) might be complicated with NK and secondary idRTA.

**Methods:** We observed 6-mo-old girl with bilateral medullary NK, detected by abdominal ultrasound examined because of febrile UTI.

**Results:** The results from lab tests are as follows: Blood- Na, K, Cl, Ca, P, Mg- normal, creatinine 24 µmol/l, urea 2.1 mmol/l uric acid 149 µmol/l, alk. phosph.235 U/l, PTH 31.17 pg/ml (N15 – 65), TSH 1.187 mU/l (N 0.35 – 4.5), 25(OH), vitD 76 mg/l (N20 – 70), 1,25 (OH) vitD 104 mg/l (N45 – 270), PRA 9.21 ng/ml/h (N1.9 – 3.7). Urine **:** spec.grav. 1.005-1.010, pH 7 ;6.5;7, glucose, proteins- negative, citrate 30 mg/24h (N≤ 600), oxalate, glycolate, glycerate-normal, Ca/Cr index -2.76 - 0.93 mg/mg ( N<0.8 -0.6) Persistent urine pH >6.5, hypocitraturia, abnormal response of frusemide test in absence of metabolic acidosis were strongly consistent with idRTA. IH with NK and secondary idRTA cannot be excluded. CT showed right double drainage renal system. Thiazide diuretic 1mg/kg/day and potassium citrate 1mmol/kg/day were started. After 6 mo normal for age urinary Ca/Cr index (0.29-0.34 mg/mg) was acheived. Increasing serum creatinine up to 59 µmol/l and PTH were measured.

**Conclusions:** In some cases more than one coexisting factors might be responsible for development of NK and exact cause is difficult to distinguish.

## Abstract# 147

**Clinical Usefulness of Urinary beta2-Microglobulin in Children with Glomerulonephritis** I.S. Lim. *Department of Pediatrics, College of Medicine, Chung-Ang University, Seoul, Korea.*

**Objectives:** Beta2-microglobulin (b2-MG) is a marker of tubulointerstitial injury and considered as predictor of the prognosis in idiopathic membranous nephropathy. There have been numerous researches on urinary b2-MG in nephrotic syndrome and their predictive value of responsiveness to steroid therapy, but not much on those in glomerulonephritis, especially of children. This study was performed to investigate the clinical usefulness of urinary b2-MG in children with one of three common childhood glomerulonephritis; APSGN, IgA nephropathy, and HSP nephritis.

**Methods:** Fifty-one patients with APSGN (n=12), IgA nephropathy (n=14) and HSP nephritis (n=25) were enrolled in this study. And healthy children without urologic disorder (n=44) and patients with idiopathic MCNS (n=45) were included as control group. In their 24-hr urine samples, b2-MG levels were checked and then compared to clinical features.

**Results:** In glomerulonephritis patients, the mean b2-MG level was significantly higher than those in normal control and MCNS group (168.9±101.3 vs. 61±28 and 78±55 µg/g-Cr, respectively, $P<0.05$). When each glomerulonephritis groups were compared, the mean urinary b2-MG level in IgA nephritis group was significantly higher than others ($P<0.05$).

**Conclusions:** These results suggest urinary b2-MG excretion might be a reliable marker in diagnosing childhood glomerulonephritis, especially IgA nephropathy. Although b2-MG value is not appropriate as an indicator of general renal function and pathology, it seems to be useful in the differential diagnosis of the pediatric nephropathy and in the prediction of their treatment period. However further larger study will be needed.

## Abstract# 148

**Spectrum of Tubulopathies in Indian Children** M.G. Matnani. *Pediatirc Nephrology, KEM Hospital, Pune, Maharashtra, India.*

**Objectives:** To assess the nature of renal tubular disorders in Indian children in a tertiary care centre.

**Methods:** Clinical records of patients attending the Pediatric Nephrology Clinic (from January 2007 to December 2009) were retrospectively reviewed to get a descriptive data about the nature of renal tubular disorders in children.

**Results:** Total of 170 children with tubulopathy were assessed.
100 children had distal Renal Tubular Acidosis (dRTA)- 38- primary, 62- secondary. Of the Primary - 17(44.7%) had associated hypercalcuria. Secondary causes include- posterior urethral valves in 30, Vesico-ureteric reflux in 24, metabolic liver disease in 8. 10 children had proximal RTA-4- primary, 6- secondary to metabolic liver disease (2-glycogen storage disease,1-Wilson's disease, 2-chronic liver disease, 1- congenital glucose and galactose intolerance) 10 children had Fanconi's syndrome-6- primary, 4- secondary (2-Wilson's disease, 1- tyrosinemia, 1-cystinosis). 8 children had Bartter's syndrome while 1 had Pseudo Bartter's due to furosemide toxicity. 38 children had idiopathic hypercalciuria, all of which presented with one of the following 4 clinical features- recurrent episodes of gross hematuria, recurrent abdominal pain, recurrent UTI's and secondary nocturnal enuresis. 2 children had nephrogenic DI (Diabetes Insipidus), 1 had chronic SIADH and 1 had ?Dent's with hypercalcuria and salt wasting.

**Conclusions:** The study shows that the commonest tubulopathy (58.8%) in children is dRTA. Obstructive uropathy (PU valves and VUR) account for majority (87%) of the secondary causes of dRTA. Metabolic liver disease, though uncommon also forms a sizable portion of secondary causes for both proximal and distal RTA.

## Abstract# 149

**Assessment of Urinary N-Acetyl-beta-D-Glucoseaminidase (NAG) in Epileptic Children before & after Treatment with Antiepileptic Drugs** M. Mazaheri, A. Samaee. *Pediatric Nephrology -Pediatric Neurology, Semnan University of Medical Science, Semnan, Semnan, Islamic Republic of Iran.*

**Objectives:** Antiepileptic drugs have various side effects on renal function. This study is planned to evaluate reliability of NAG index (urinary NAG/cr)for screening of renal injury in patients who recieved carbamazepin or valproat sodium.

**Methods:** In this study 105 epileptic patients whose age were 3-16 yrs old were selected & divided in three sex &age matched group: **A ;**patients who did not take any teatment.(tx) **B**:patients who took min.6 month tx. with carbamazepin (monotherapy). **C**:patients who recieved min. 6 month tx. with valproat sodium (monotherapy).4th is selected as healthy control group( **D**).patients number was the same in all groups.Blood sample was drawn for biochemical tests,urine sample for all patients was sent for assessing NAG & creatinine & urinalysis.

**Results:** Biochemical tests like BUN.Cr.Eectrolytes,in all children was normal. Sonographic finding in all was normal too.Mean urinary NAG index was 0.68 & 0.74 in control & group A respectively.There was not significant difference between them.Mean NAG index in groupB was 1.71& it was 2.3 times of group A.(P=0.000).Also in group C it was 2.45 & 3.3 times of group A. (P=0.000). Comparing group B & C showed that NAG index in group C was1.5 times of group B.(P=0.000).

**Conclusions:** Treatment with carbamazepin & valproat sodium can increase urinary NAG significantly in comparison with control group &group who did not take any drugs.NAG index in patients can be raised by taking valproat sodium more than carbamazepin. It means both of drugs can cause renal tubular function deterioration but valproat sodium can cause more sever damage in kidney.

## Abstract# 150

**Nephrocalcinosis (NC) in Lowe's Syndrome – A Possible Role for Hypervitaminosis D?** L.S. Milner, L. Mazzola. *Pediatric Nephrology, Floating Hospital for Children, Tufts University School of Medicine, Boston, MA, United States.*

**Objectives:** The objective was to see if Hypervitaminosis D is associated with NC in children with Lowe's Syndrome (LS),an X-linked recessive disorder characterized by renal tubular dysfunction.
Hypercalciuria in LS is thought to be due to a genetically determined chloride channel defect, thought to be associated with Dent's disease also characterized by NC.Since elevated Vitamin D (VD) levels causes hypercalciuria, predisposing to NC,we evaluated serum VD status in children with LS who had NC,since moderation of VD intake could theoretically ameliorate NC.

**Methods:** Four children with LS were studied.Serum creatinine (CR),calcium(CA),25 Hydroxy (Vit D) and 1,25 Dihydroxy Vitamin D (DHVD) levels,the active form of VD as well as spot urine protein (PR), and CA to CR ratios were measured. Each child received a renal utrasound. All the children were on an unrestricted CA diet. One received low dose VD supplements.

**Results:** The mean age of the patients was16.3 years. Reduced renal function occurred in 2 of the children. All had evidence of renal tubular dysfunction and proteinuria with a mean urine PR to CR ratio of 4.3 (normal <0.2).
Gross hematuria occurred in 2. Hypercalciuria with elevated DHVD levels and NC were noted in all of the children.

Maximum serum VitD,DHVD levels and urine CA/Cr ratio

| LS Patients (4) | VitD (ng/ml) | DHVD( pg/ml) | UCA/CR ratio |
|---|---|---|---|
| Mean ± SD | 34.5 ± 22 | 102.2 ± 14 | 0.56 ± 0.2 |
| Upper limit of normal | 100 | 62 | 0.2 |

SD= standard deviation

**Conclusions:** Elevated DHVD levels appears to be common in LS, possibly causing hypercalciuria, promoting NC.Abnormal metabolism of VD in LS could possibly account for this finding.

## Abstract# 151

**Urinary Calcium to Creatinine Ratio in Children in Limestone Area, Korea** E.G. Soon, H.J. Cho, M.K. Namgoong. *Pediatrics, Yonsei University, Wonju College of Medicine, Wonju, Korea; Pediatrics, Yonsei University, Wonju College of Medicine, Wonju, Korea; Pediatrics, Yonsei University, Wonju College of Medicine, Wonju, Korea.*

**Objectives:** The purpose of this study aims to compare the Urine calcium to creatinine ratio (U Ca/Cr) of school children between nonlimestone and limestone area.

**Methods:** The study was performed on 398 healthy school children of age 7-12. Urine samples were collected just before lunch hour. In a questionnaire, on the

same day, the parents and caregivers filled out diet habit, disease history, family income, morning diet, and the type of drinking water of the children. U Ca/Cr ratio of 350 healthy children in a non-limestone area was detected.
**Results:** The mean value for U Ca/Cr ratio of the limestone group and the control group were 0.09±0.076 and 0.11±0.133, respectively (P<0.001). The 95th percentile for U Ca/Cr ratio of the limestone group and the control group were 0.25±0.076 and 0.30±0.134, respectively. The U Ca/Cr ratio >0.20 was 9% in the limestone group and 18.9% in the control group(P<0.05). A negative correlation R=-0.15, P=0.002 was observed between age and U Ca/Cr ratio. There was no significant difference in sex. The U Ca/Cr ratio of the limestone group was irrelevant to diet habit, disease history, family income, morning diet, and BMI (B.wt/m²). No significant variations in U Ca/Cr ratio according to the type of drinking water such as filtered water, ground water and tap water were observed in the samples of the limestone group.
**Conclusions:** The expected value of U Ca/Cr ratio was not significantly high enough to compare with that in the non-limestone area. There were not different U Ca/Cr ratio according to the drinking water types.

## Abstract# 152

**Gitelman Syndrome Accompanied by Nephrotic Syndrome** E.G. Soon,[1] H.J. Cho,[1] M.K. Namgoong.[1] *[1]Pediatric Department, Yonsei University, Wonju College of Medicine, Wonju, Korea; [2]Pediatric Department, Yonsei University, Wonju College of Medicine, Wonju, Korea; [3]Pediatric Department, Yonsei University, Wonju College of Medicine, Wonju, Korea.*
**Objectives:** The typical symptoms of Gitelman syndrome are hypokalemia and metabolic alkalosis associated with hypereninemia via NaCl cotransporter dysfunction in renal distal tubule. This syndrome has a tendency to show only tubulopathy but only several cases have been reported with the concomitant glomerulopathy. We present the Gitelman syndrome with the concomitant minimal change nephrotic syndrome, confirmed with gene study.
**Results:** A 2 year old male child was admitted for a 10 days history of generalized edema. Urinalysis showed severe proteinuria. Blood chemistry test showed hypoalbuminemia (1.1g/dL), high cholesterol (490mg/dL) and high triglyceride (641mg/dL). Complement test was normal C3 and C4. Since the 1st admission, he has shown one or two times relapse every year. He was diagnosed as a typical steroid dependent nephrotic syndrome. His electrolytes were hypokalemia and metabolic alkalosis. Hypokalemia was not correlated with prednisolone administration. Blood renin and aldosterone level were high renin (42.9ng/dL) and normal aldosterone (2.6ng/dL). There were normal serum magnesium (2.2mg/dL) and hypocalciuria (UCa/Cr 0.08%). T3, FT4 and TSH value were within normal limits. Gitelman syndrome was confirmed with SLC12A3 gene DNA analysis. He had a compound heterozygous mutation on SLC12A3 gene.
**Conclusions:** Although the function of SCL12A3 gene mutation in proteinruia has not found out, we would like to suggest that proteinuria could be one of the symptoms of Gitelman syndrome.

## Abstract# 153

**Renal Manifestations in Children Co-Infected with HIV and Disseminated Tuberculosis** P. Nourse,[1] M.F. Cotton,[2] W.D. Bates.[3] *[1]Pediatrics, School of Child and Adolescent Health, University of Cape Town, Cape Town, South Africa; [2]Pediatrics, Department of Pediatrics and Child Health, Tygerberg Children's Hospital and Stellenbosch University, Cape Town, South Africa; [3]Histopathology, Tygerberg Hospital, Stellenbosch University, National Health Laboratory Service, Cape Town, South Africa.*
**Objectives:** In Cape Town many children are co-infected with human immunodeficiency virus (HIV) and tuberculosis. Our objective was to establish if tuberculosis played a role in renal disease in HIV-infected children.
**Methods:** We identified children co-infected with Tuberculosis and HIV from our database and reviewed their biopsies and clinical notes.
**Results:** From 2002 to 2008 12 renal biopsies or postmortem examinations were done in HIV infected children at our institution. In four cases, median age 73(24 to 108) months, the clinical scenario and renal biopsies were consistent with tuberculous involvement. Mean CD4 count and percentage: 508 cells/µl (23%). All four cases presented with culture proven disseminated tuberculosis (not yet on treatment). All four had nephrotic range proteinuria and hypoalbuminemia. Three patients had renal impairment. Renal biopsies: Prominent features were a severe interstitial inflammatory infiltrate and mild to moderate mesangial proliferation. An interstitial granuloma was seen in one patient. In all four patients proteinuria resolved and renal function improved after tuberculosis treatment.
**Conclusions:** Tuberculosis contributes to proteinuric renal disease in HIV-infected children and improves on tuberculosis treatment.

## Abstract# 154

**A Rare Cause of Proteinuria: Lysinuric Proteine Intolerance** C. Pietrement,[1] A. Babik,[1] A. Gobe,[2] M. Abely,[1] G. Roselyne.[3] *[1]Paediatrics, American Memorial Hospital, Reims, France; [2]Paediatrics, Hopital de Charleville-Mézières, Charleville Mézières, France; [3]Laboratory of Paediatric Biochemistry, CHU de Reims, Reims, France.*
**Objectives:** Lysinuric proteine intolerance (LPI) is an autosomal recessive metabolic disorder characterized by defective transport of dibasic amino acids at the basolateral membrane of epithelial cells in intestine and kidney tubule. The dysfunction leads to low plasma and high urine concentration of dibasic amino acids. Hyperammonaemia is caused by functional impairment of the urea cycle. Clinical symptoms are variable and non specific leading often to misdiagnosis.
**Methods:** We report the case of a 13 year old boy, seen in paediatric nephrology consultation after one episode of macroscopic haematuria with mild proteinuria.
**Results:** He had no positive family history and was born after an uneventful pregnancy. Because of cataract, hepatosplenomegaly and hyperferritinemia, he had had lots of explorations for suspicion of galactosemia, Gaucher disease, Niemann-Pick disease, spherocytosis and haemochromatosis. Urinary amino acid chromatography revealed increased excretion of lysine, arginine, ornithine and citrulline, subsequently plasma analysis has shown low concentration of them and urinary organic acid chromatography elevated acid orotic. Elevated levels of ferritin, liver transminases and mild leucopenia with anemia were also present. Detailed dietary history revealed aversion to protein diet.
**Conclusions:** The treatment has been started consisting in strict avoidance of protein rich food and supplementation of citrulline to replenish the urea cycle. The renal function will be carefully monitored since chronic renal failure has been reported in LPI.

## Abstract# 155

**Growth Hormone (GH) Increases Body Weight of Potassium Depleted Young Rats but Does Not Modify Renal Enlargement** H. Gil-Peña, E. Garcia-López, O. Alvarez-García, V. Loredo, E. Carbajo-Perez, F. Santos. *Hospital Universitario Central de Asturias & University of Oviedo, Oviedo, Spain.*
**Objectives:** To investigate GH effect on renal enlargement induced by chronic potassium (K) deficiency.
**Methods:** Young rats were grouped (n=10) in Normal diet (C), Low K diet (KD), KD+3.3 mg/kg/d of GH for 7 days (KDGH), C paired with KD (CPF). At sacrifice, the following variables were analyzed.
**Results:**

| | C | KD | KDGH | CPF |
|---|---|---|---|---|
| Serum K(mEq/L) | 3.81±0.41 | 1.78±0.18a | 1.60±0.23a | 3.73±0.33b,c |
| Serum IGF-I(ng/ml) | 196.0±81.2 | 119.5±37.7a | 112.3±34.8a | 85.2±20.4a |
| Weight gain(g) | 13.95±4.47 | 5.78±2.02a | 10.56±1.78b | 6.23±3.12a,c |
| Kidney weight(g) | 0.50±0.03 | 1.03±0.12a | 0.97±0.09a | 0.52±0.04b,c |
| BrdU labelling | + | ++ | +++ | + |
| GHR fold change* | 1±0.05 | 0.52±0.02a | 0.8±0.02b | 1.62±0.12a,b,c |
| IGF-I fold change* | 1±0.05 | 0.32±0.21a | 0.38±0.02a | 0.51±0.05a |
| IGF-IR fold change* | 1±0.1 | 1.67±0.10a | 2.78±0.12a,b | 1.33±0.10c |

Numerical values are X ± SD. aStatistically different from C. bStatistically different from KD. cStatistically different from KDGH. P≤0.05. *Assessed by qPCR
**Conclusions:** Compared with C, K deficiency reduces body weight gain and increases kidney size associated with low expression of GHR and IGF-I and upregulation of IGF-IR. Treatment normalized weight gain of DK animals but did not modify the exaggerated renal growth in spite of further increasing cell proliferation. GH also normalized GHR expression, intensified IGF-IR upregulation and did not modify low IGF-I mRNA levels caused by K depletion.

## Abstract# 156

**Nephrocalcinosis in Children: Preliminary Data of Prospective Study in Albania** D. Shtiza,[1] R. Xhepa,[1] O. Xhango,[1] A. Bulo,[2] N. Marku.[2] *[1]Department of Pediatrics, Service of Nephrology, University Hospital Centre of Tirana; [2]Department of Laboratories, Biochemical and Clinical Service, University Hospital Centre of Tirana, Tirana, Albania.*
**Objectives:** To study the data of children with nephrocalcinosis (NC) and to analyze etiology, presenting complaints, clinical findings, growth, renal function at presentation, treatment, and to relate growth and renal function to changes in NC in patients with a follow-up for a period from 3-5 years.
**Methods:** Eighteenn children, followed with the diagnosis of NC in our hospital were included in the study. Prospective evaluation. A complete blood count, biochemical evaluation, urinalysis, blood pH and HCO3 levels, hand-wrist x-ray, abdominal ultrasonographie were performed in all pts. The patients were evaluated every six months. Audiometric test were performed in all patients.
**Results:** The median age at the time of diagnosis was 11 months. Presenting symptoms: failure to thrive in the first year of life (62%), UTI, vomiting, bladder voiding dysfunction or psychomotor delay (15%). In 28% of cases NC was detected incidentally. In 36 % of children NC was associated with idiopathic hypercalciuria, 38% with various hereditary tubular disorders and 13% to prophylactic bolus administration of vitamin D. Renal function at diagnosis was normal in 79% of children. Growth standard score improved from a median of

-2,8 to -1,4 and renal function remained stable in 86% of patients. GFR changed only slightly during this period of observation.
**Conclusions:** The most frequent causes of NC were hereditary tubulopathies and vitamin D intoxication in childhood in Albania. Our results show that the treatment of the underlying conditions is associated with catch-up growth and stabilization of renal function in many children, but not with reduction in the degree of NC in the majority of cases.

## Abstract# 157

**Persistent Hypouricemia in a Child with Distal Renal Tubular Acidosis and URAT1 Mutation** V. Tasic,[1] Z.S. Gucev,[1] N. Ristoska-Bojkovska,[1] V.J. Lozanovski,[1] H.I. Cheong,[2] N. Anzai,[3] K. Kitamura.[4] *[1]University Children's Hospital, Skopje, Macedonia, The Former Yugoslav Republic of; [2]University Children's Hospital, Seoul, Korea; [3]Kyorin University School of Medicine, Tokyo, Japan; [4]Kumamoto University Graduate School of Life Sciences, Kumamoto, Japan.*
**Objectives:** Distal renal tubular acidosis (dRTA) may be associated with transitory proximal tubular abnormalities. In this work we describe a female child with dRTA and persistent hypouricemia.
**Methods:** The index patient underwent assessment of the proximal and distal renal tubular function, uric acid handling and mutational analysis of URAT1 gene.
**Results:** A 5 year old female presented with hyperchloremic metabolic acidosis (pH 7.23, HCO3 13.6 mmol/l, Cl 122 mmol/l, urine pH 6.77), severe staturoponderal deficit and proximal tubular abnormalities (low molecular proteinuria, hyperaminoaciduria, hypophosphatemia, hypouricemia). After correction of acidosis all abnormalities normalized except hypouricemia (highest value 78 micromol/l, FEUric acid 30%). Analysis of the URAT1 revealed heterozygous mutation R434C.
**Conclusions:** Persistent hypouricemia in our patient with dRTA was due to URAT1 mutation. We believe that acidification defect in dRTA (persistently alkaline urine) protects her from urate lithiasis.

## Abstract# 158

**The Role of Y-Box Binding Protein-1 (YB-1) in Collecting Duct (CD) Epithelial-Mesenchymal Transition (EMT)** P. Trnka, L. Ivanova, S.E. Dunn, D.G. Matsell. *Pediatrics, Child and Family Research Institute, Vancouver, BC, Canada.*
**Objectives:** The transcription factor YB-1 has been implicated in carcinogenesis and in epithelial phenotypic transition. The aim of this project was to study the role of YB-1 in insulin-like growth factor-1 (IGF-1)-induced CD EMT.
**Methods:** Wild-type and YB-1 siRNA-transfected mouse kidney mIMCD3 cells were stimulated with IGF-1 in a dose- (0-100 ng/ml) and time- (0-72 hours) dependent fashion to induce EMT. Intracellular signaling and efficiency of YB-1 knockdown were determined by Western blot (WB) analysis, while cell phenotype was determined by phase contrast microscopy, immunohistochemistry, and WB analysis.
**Results:** Stimulation of mIMCD3 cells by IGF-1 resulted in a dose- and time-dependent change in cell phenotype, from a cobblestone to a spindle-shape morphology, dislocation of E-cadherin and β-catenin from the cell membrane to the cytoplasm and nucleus, a significant decrease in E-cadherin and β-catenin expression (0.52 and 0.32-fold respectively), and a significant increase in vimentin and α-smooth muscle actin expression (2.23 and 4.1-fold respectively) compared to unstimulated control cells. Early molecular events included the activation of PI3K/Akt, MAPK/ERK, and RSK pathways, with predominantly nuclear localization of phosphorylated YB-1. Knockdown of YB-1 to 18% of control abrogated the effects of IGF-1 stimulation on CD EMT, with preservation of epithelial morphology, and normalization of the expression of epithelial and mesenchymal proteins back to 109-150% of control values.
**Conclusions:** YB-1 modulates IGF-1-induced CD EMT, and may be an important player in congenital obstructive CD injury.

Immunology of Renal Disease

## Abstract# 159

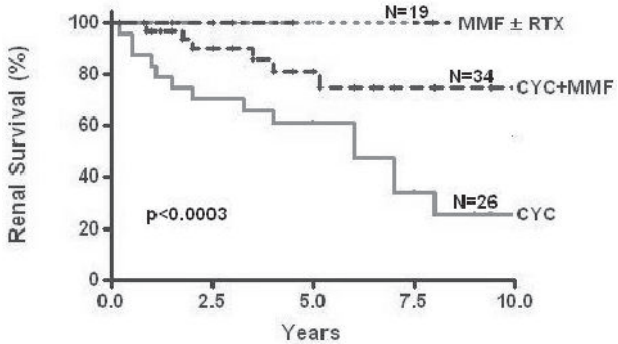
**(O-4)**

**Plasmatherapy in Atypical Hemolytic Uremic Syndrome, French Cohort** T. Kwon,[1] N. Biebuyck,[2] M. Cailliez,[3] F. Broux,[4] M.A. Macher,[1] P. Niaudet,[2] M. Tsimaratos,[3] G. Deschenes,[1] V. Fremeaux Bacchi,[5] C. Loirat.[1] *[1]P.Nephrology, Hop. Debré, Paris, France; [2]P.Nephrology, Hop. Necker, Paris, France; [3]P.Nephrology, Hop. Timone, Marseille, France; [4]Pediatrics, Hop. Nicolle, Rouen, France; [5]Immunobiology, Hop. Pompidou, Paris, France.*
**Objectives:** Plasmatherapy (PT) is the first line treatment in atypical hemolytic uremic syndrome (aHUS), but little is known on its efficiency to prevent end stage renal disease (ESRD). We report the follow up of 10 children treated by long term PT.
**Results:** Median age at diagnosis was 6 months (0.5 to 51 months). 8 patients have factor H (*CFH*) mutations: homozygous in 3 (quantitative deficiency), heterozygous in 4 (type 1 -quantitative- in 1; type 2 -functional- in 3), compound (type 1 and 2) in 1. One patient has a heterozygous factor B mutation and 1 patient a heterozygous factor I (*CFI*) mutation (type 2), combined to an unknown risk factor. All patients received PT (fresh frozen plasma infusions in 6, plasma exchanges in 4) within the first month after onset and were maintained on long term prophylactic PT. Median follow up is 31 months (5 to 67 months). All patients except 2 have had relapses during infections or PT tapering, rescued by treatment intensification. All patients have a normal glomerular filtration rate at last follow up, except 1 with hybrid *CFH* (type 2) mutation. Another patient (*CFI* mutation) had persistent anemia after 1 year on PT, and has been switched to eculizumab therapy.
**Conclusions:** Prophylactic PT appeared to be efficient to prevent ESRD in 8/10 patients. Therefore it should be recommended in aHUS although the complement blocker eculizumab may be another option in near future.

## Abstract# 160

**(O-21)**

**Three Decades of Progress in Treating Childhood Onset Lupus Nephritis** T. Pereira, C.L. Abitbol, W. Seeherunvong, J. Chandar, M. Freundlich, G. Zilleruelo. *Pediatric Nephrology, University of Miami/ Holtz Children's Hospital, Miami, FL, United States.*
**Objectives:** To compare renal survival in a cohort of pediatric patients with childhood onset systemic lupus erythematosus (cSLE) and severe lupus nephritis (LN) with progressive treatment regimens that included pulse solumedrol, cyclophosphmide (CYC), mycophenolate mofetil (MMF) and rituximab (RTX).
**Methods:** A retrospective analysis was performed on 117 patients with cSLE. Of the initial cohort, 79 (63 female; 80%) had ≥ WHO class 3 and were designated as having severe LN. The end point for renal survival was progression to end stage kidney disease (ESKD).
**Results:** Average age of onset was 12.5 ± 3.0 years with a mean follow-up of 5.6 ± 3.5 years. There was a nonwhite racial predominance of 92%. Twenty-six (33%) progressed to ESKD. During the first 2 decades, all patients received corticosteroids and CYC. When MMF was added, renal survival improved from a median of 6 to 12 years (p<0.003). Patients treated with MMF alone or with RTX have experienced no patient or renal demise.




**Conclusions:** Renal survival in childhood onset LN has improved during the past 3 decades with the addition of MMF to the treatment regimen. Continued experience with this as an induction agent with or without the use of RTX requires further investigation. Collaborative trials in pediatric patients are very much warranted.

## Abstract# 161
**(O-22)**

**Functional Toll-Like Receptor (TLR) System in Podocytes** T. Srivastava,[1] K. Yew,[1] P.A. Cudmore,[1] R. Sharma,[2] E.T. McCarthy,[3] M. Sharma,[2] C.J. Harrison.[1] *[1]Children's Mercy Hospital, Kansas City, MO, United States; [2]Kansas City VA Medical Center, Kansas City, MO, United States; [3]University of Kansas Medical Center, Kansas City, KS, United States.*

**Objectives:** TLR activation has been shown to be important in models of renal injury. TLR signal via (1) MyD88 pathway which activates NF-κB and (2) TRIF pathway which activates IRF genes. LPS is a potent ligand for TLR4 in cells with functional TLR. We investigated if podocytes have functional TLR.

**Methods:** We determined baseline TLR expression in podocytes by RT-PCR. We then studied the effect of LPS (0-50 μg/ml) and puromycin (PAN, 0-90 μg/ml) on podocyte cell morphology by crystal violet (0.1%) and F-actin cytoskeleton by phalloidin stain. LPS (25 μg/ml) or PAN (60 μg/ml) were used to study (a) TLR mRNA expression by qRT-PCR at 1hr and 6hr, and (b) concentrations of IL-6, TNFα, and IFNβ in cell supernatants at 6hr and 24hr using Luminex assay.

**Results:** Podocytes constitutively express TLR1-6, TLR10 and MD2, but not TLR7-9 or MD1. LPS and PAN caused similar blunting or loss of cytoplasmic processes and disorganization of F-actin fibers. LPS but not PAN upregulated the expression of TLR1,2,4,6 plus MyD88 and NF-κB (MyD88 pathway). Neither agent induced TLR 7-9, or upregulated TLR3, TLR5, TRIF or IRF3. Supernatant IL-6 (pg/ml) increased above controls at 6hr (361±82, p=NS) and 24hr (747±316, p=0.002) post LPS exposure, but not after PAN exposure. There was no detectable TNFα or IFNβ in supernatants.

**Conclusions:** We demonstrate that podocytes express non-endosomal TLRs and have a functional TLR4 signaling pathway. We believe that alterations in TLR signaling (or innate immune response) may play a crucial role in podocyte's response to injury in glomerular diseases.

**DISCLOSURE:** *Harrison, C.J.*: Grant/Research Support, Cubist, Astellas, J&J, Biosynexus, GSK.

## Abstract# 162

**Uncontollable IgA Glomerulonephritis and Homozygous Disruption of the CD81 Gene** B. Adams,[1] M.C. Van zelm,[2] J. Smet,[3] F. Mascart,[3] F. Janssen,[1] A. Ferster,[1] M. Van den Burg,[2] J.J. van Dongen.[2] *[1]Pediatric Nephrology, HUDERF, Brussels, Belgium; [2]Immunology, Erasmus MC, Rotterdam, Belgium; [3]Clinical Immunobiology, Hopital Erasme, Brussels, Belgium.*

**Objectives:** Henoch shonlein purpura (HSP) is a systemic vasculitis characterized by tissue deposition of IgA containing immune complexes. Although associated renal involvement is common it is usually mild and of favorable outcome. The tetraspan molecule CD81 is widely expressed on immune cells, but also on most stromal and epithelial cells. In B-cells it is required for signaling upon antigen recognition. Its functions on other cells are still unclear.

**Methods:** We herein report the case of a 7-year-old girl who presents an association of severe and uncontrollable IGA glomerulonephritis, profound hypogammaglobulinemia, immunopathology and a CD81 mutation.

**Results:** The patient is the first child of consanguineous parents. At the age of 3.5 years she developed HSP. Renal biopsy revealed diffuse mesangial proliferation, crescent formation and strong deposition of IgA and C3.She also presented recurrent episodes of thrombocytopenia associated with anti-platelet antibodies and seric levels of IgG were persistently low.She was treated with repeated full course immunosuppressive therapy but progressed to end stage renal failure. Flow cytometric immunophenotyping of blood showed no expression of CD19 on the patient's B-cells. Additional studies showed that all cells lacked CD81 expression. Sequencing of the CD81 gene showed a homozygous G>A substitution.

**Conclusions:** We present the first case of immunodeficiency syndrome, due to a CD81 mutation. The nephropathy is likely an indirect consequence of the CD81 defect.

## Abstract# 163

**Varicella Zoster Virus (VZV) Infection in Immunocompromised Patients: Treatment with Specific Human VZV Polyclonal Immunoglobulins** A.-L. Sellier-Leclerc,[1] B. Aoun,[2] V. Baudouin,[1] G. Deschênes,[1] T. Ulinski.[2] *[1]Pediatric Nephrology, Robert Debré Hspital, Paris, France; [2]Pediatric Nephrology, Armand Trousseau Hospital, Paris, France.*

**Objectives:** Infection with the varicella-zoster virus (VZV) is more dangerous in immunocompromised patients than it is in the general population. High doses of acyclovir and immediate reduction of immunosuppression may improve the prognosis of severe forms, but multiorgan failure and death may occur despite early antiviral treatment.

Here, we report four cases of VZV infection in children receiving steroid therapy and immunosuppressive drugs for renal allograft, idiopathic nephrotic syndrome, and systemic lupus.

**Methods:** The only clinical manifestation in three patients was general malaise, fever, and disseminated vesicular rash, whereas one patient also showed severe and diffuse visceral involvement with multiorgan failure.

**Results:** Adjuvant treatment with specific human varicella zoster polyclonal immunoglobulins led to a dramatic improvement of VZV infection in all four patients within two days after administration. Simultaneous acyclovir therapy may jeopardize interpretation, but there was a rapid clinical improvement 24 to 48 hours after administration of specific human varicella zoster polyclonal immunoglobulin, suggesting its favourable role for disease outcome in these four patients.

**Conclusions:** Specific human varicella zoster polyclonal immunoglobulin is probably a useful adjuvant therapy to acyclovir in declared and severe varicella in immunocompromised children.

## Abstract# 164

**Regulatory T Cells (Treg) and Interleukin 17 Producing T Cells (TH17) in Iga Nephropathy (IgAN)** R. Camilla,[1] V. Daprà,[1] E. Loiacono,[1] L Peruzzi,[1] C. Rollino,[2] G. Beltrame,[2] M. Ferro,[2] R. Gallo,[1] L Garassino,[1] A. Amore,[1] R. Coppo.[1] *[1]Nephrol Dial Transplant, R Margherita Hosp, Turin, Italy; [2]Nephrol, Dial, G Bosco Hosp, Turin, Italy.*

**Objectives:** We aimed at investigating Treg/TH-17 producing cells balance in IgAN and the correlation with TLRs expression, as marker of innate immunity.

**Methods:** Circulating mononuclear cells were isolated from 28 IgAN patients, 82% males, e-GFR 92.6±46.5 ml/min, median protein/creatinine 0.2, and10 healthy controls (HC). mRNA expression of Foxp3, TH17-related factors (IL-17 and retinoid orphan nuclear receptor (RORc), TFG-beta1 and TLR 2, 4 and 7 were assessed by Taqman and normalized to Abelson gene (Abl).

**Results:** Transcriptional levels of Foxp3 were significantly lower in IgAN vs HC (0.82±0.30 vs 1.05±0.37, p= 0.041), while those of IL-17 and of its regulatory factor RORc were slightly, but not significantly increased (IL-17 1.17±1.07 vs 1.05±0.41 in HC, RORc 1.25±0.85 vs 1.14±0.71 in HC). A significant correlation was found between IL-17 and RORc mRNAs (p<0.0001). Transcriptional levels of TGFβ1 were similar to HC in IgAN patients and directly correlated with RORc (p=0.0015) and IL-17 (p=0.031) mRNAs. Patients with urinary protein/creatinine >1 (median 1.62) had Foxp3 levels lower than those with protein/creatinine <1, (median 0.18), p=0.06. ROR-c levels had a trend inverse correlation with e-GFR (p=0.06).

Of interest, a significant inverse correlation was found between Foxp3 and TLR4 expression (p= 0.022).

**Conclusions:** These results indicate in IgAN a functional defect in Tregs, which correlated with signs of hyperactive innate immunity. TGFβ1 seems to favour the expansion of TH17 cells. A trend correlation with proteinuria was observed.

## Abstract# 165

**Slit2 Impairs Neutrophil Adhesion in Ischemia Reperfusion Injury** S. Chaturvedi, I. Mukovozov, S. Patel, Y. Huang, G.Y. Liu, L. Robinson. *Nephrology, The Hospital for Sick Children, Toronto, ON, Canada.*

**Objectives:** Acute Kidney Injury (AKI) occurs in approximately 5% of all hospitalized patients and leads to significant morbidity, mortality and financial costs. Inflammation marked by recruitment of circulating leukocytes particularly neutrophils into the injured kidney is a key component of AKI caused by ischemia-reperfusion injury (IRI). Recruited neutrophils exacerbate injury by releasing inflammatory mediators. The neuronal guidance cue, Slit2 and its transmembrane receptor roundabout (Robo) prevent axonal migration during development of central nervous system. Recently, we showed that Slit2 also inhibits neutrophil chemotaxis towards diverse chemoattractants. The objectives of this study were to determine whether Slit2 affects neutrophil adhesion in an *in vitro* model of IRI.

**Methods:** Human umbilical endothelial cells (HUVECs) grown to confluence were exposed to 1% oxygen (hypoxia), followed by variable periods of re-perfusion. HUVEC were then incubated with fluorescent labelled human neutrophil in presence or absence of Slit2. Nonadherent neutrophils were removed by washing and adherent neutrophils were counted using a fluorescent plate reader.

**Results:** Slit2 reduced neutrophil adhesion in a cell culture model of IRI ranging from 30 minutes to 3 hrs of re-perfusion (p<0.05).

**Conclusions:** Our findings suggest that in IRI, Slit2 inhibits neutrophil adhesion. Thus Slit2 may have a potential role in prevention and treatment of AKI.

## Abstract# 166

**The Role of Th17/IL-17 in the Pathogenesis of Primary Nephrotic Syndrome in Children** Q. Li, L. Wang. *Children's Hospital of Chongqing Medical Universtiy, Chongqing, China; 136 Second Zhongshan Road, Yuzhong District, Chongqing 400014, People's Republic of China, Chongqing, China.*

**Objectives:** To investigate the role of Th17 cell and IL-17 in pathogenesis of primary nephrotic syndrome(PNS) of children.

**Methods:** 35 Children with PNS and 20 healthy subjects were selected. The frequency of Th17 cells, mRNA expressions of RORc,IL-23p19 in peripheral blood mononuclear cells, plasma concentration of IL-1β,IL-6 and expression of IL-17 in renal biopsy tissue were examined. Also the release of inflammatory factors by human mesengial cells in response to recombinant human(rh) IL-17 and expression of NF-κB were detected. In addition, the effect of apopotosis on mouse podocyte co-cultured with recombinant murine(rm) IL-17 was measured. The espressions of casepase-3 and Nephrin,WT1, Synaptopodin mRNA in mouse podocyte co-cultured with 100ng/ml rmIL-17after 72h were detected.

**Results:** The frequency of Th17 cells, mRNA levels of RORc,IL-23p19 and plasma concentration of IL-1β,IL-6 were higher in PNS group than control group,and expression of IL-17 in renal biopsy tissue were higher gradually in MCNS,MsPGN and FSGS than contol group(all $P<0.05$).RmIL-17 had no effect on the expression of Nephrin,WT1 and Synaptopodin mRNA of mouse podocyte, but promoted apoptosis in a dose- and time-dependent fasion by activating casepase-3. Also rhIL-17 increased the productions of IL-1β and TNF-a in human mesengial cells in a dose- and time-dependent style by activating NF-κB, but the role can be blocked by NF-κB inhibitor.

**Conclusions:** These data show that Th17/IL-17 might contribute to the pathogenesis of PNS in children.

## Abstract# 167

**The Relation between NF-κB on Regulation of Dendritic Cell and Imbalance of T Helper Cell Differentiation in Henoch-Schonlein Purpura** Q. Li, H. Tao. *Children's Hospital of Chongqing Medical Universtiy, Chongqing, China.*

**Objectives:** To investigate The relation between nuclear transcription factor-κB on regulation of dendritic cell(DC) and imbalance of T helper cell differentiation in the pathogenesis of Henoch-Schonlein purpura (HSP).

**Methods:** Inflammatory cytokines IL-4, INF-γ in blood plasma and IL-12,IL-10 in the supernatants of cultured DC were detected by ELISA. The cell count of $CD_{86}$,$CD_{80}$,$CD_{83}$,HLA-DR,CD-209 expression on DC were determined by cell flow cytometry. RT-PCR detection of NF-κB isoforms p50,p65mRNA expression. cells immunohistochemical detection the abnormal NF-κB expression in DC.

**Results:** Compared with the control group, DC cultured in HSP and HSPN group showed: IL-12 levels and INF-γ levels were lower than the control group,IL-10 levels and IL-4 levels were significantly increased than the control group;$CD_{86}$,$CD_{83}$, HLA-DR were significantly higher but $CD_{80}$ was significantly lower than the control group, ($P<0.05$).NF-κB p50,p65 mRNA expression intracellular also increased ($P<0.05$),and had a large number of NF-κB activation. HSP and HSPN group detected had no statistical relationship between the above ( $P>0.05$), When NF-κB inhibitors to join most of the above indicators are back to normal.

**Conclusions:** The function of $T_H1$ was decreased while the function of $T_H2$ was increased in HSP,which were related to DC abnormal discussion.NF-κB is a key regulatory genes of DC immune function, inhibit activation of NF-κB is a valid target to blocking DC function and depends on the T-cell immune response, It will be an effective way to treatment HSP earlier.

## Abstract# 168

**Altered Regulation of Gatekeeper Molecules (*OSM* and *ABCG5*) Affect Cholesterol Metabolism in an *IL-13* Overexpression Rat Model of Minimal Change Nephrotic Syndrome (MCNS)** I.D. Liu, L.D. Low, C.Y. Chan, T.K. Maheshwari, H.H. Yang, H.K. Yap. *Pediatrics, National University Health System, Singapore, Singapore.*

**Objectives:** We have previously shown that rats overexpressing *IL-13* developed MCNS, accompanied by proteinuria, hypoalbuminemia and hypercholesterolemia. This study aimed to examine the key molecules affecting cholesterol metabolism in this disease.

**Methods:** RNA isolated from liver tissue of *IL-13*-transfected rats with MCNS whose serum cholesterol levels were >3.10 mmol/L was reverse-transcribed and hybridized into Illumina Rat Ref12 microarray chips. Gene ontology and pathway analysis were carried out using MetaCoreTM, and validated using real-time PCR (RT-PCR).

**Results:** Out of 13,517 genes that were differentially expressed, 778 genes showed >2-fold change in expression. MetaCoreTM pathway analysis showed downregulation of Oncostatin M (OSM) and ABCG5 molecules that regulate cholesterol uptake. Analysis of (OSM) signaling pathway showed upregulation of IL-13 receptor molecules, IL-13Rα1 and IL-13Rα2, downregulation of downstream molecules Jak2 and STAT5, LIFR, SHP-2, MAP kinases and LDLR. Key molecules in cholesterol metabolism such as SREBP, ACLY, HMG-CoAR and CYP7A1 were upregulated in *IL-13* transfected rats compared to controls. RT-PCR showed down-regulation of ABCG5, as well as 5-fold downregulation of LIFR, the receptor for OSM.

**Conclusions:** This study suggests that hypercholesterolemia in *IL-13* overexpression rats with MCNS, could result from diminished circulatory clearance of cholesterol secondary to diminished hepatic clearance of cholesterol arising from inhibition of ABCG5 transcription and/or down-regulation of OSM inhibiting LDLR transcription.

## Abstract# 169

**Activation of Complement Classical Pathway in PODOCYTE Involved in the Pathogenesis of Puromycin Aminonucleoside (PAN)-Induced Proteinuria** J. Miao,[1] Q. Fan,[1] Q. Cui,[2] Y. Tan,[1] M.-h. Zhao,[1] J. Ding.[1] *[1]Peking University First Hospital, Beijing, China; [2]Peking University Health Science Center, Beijing, China.*

**Objectives:** Mounting data demonstrated that some podocyte molecules play critical role in the pathogenesis of proteinuria, while the underlying mechanism is still elusive.

**Methods:** Glomeruli microarray analysis was performed in PAN rat at different days. Microarray results were further analyzed by real-time PCR and immunohistochemistry.

**Results:** Functional enrichment analysis showed that the complement classical pathway was activated obviously in PAN rats. Real-time PCR confirmed that *C1qb*, *C4b*, *C2, C3*, *C5* and *Serping1* increased significantly at day 10 in PAN rats, while *C3* and *C5* recovered to normal level at day 15, and that *Mbl1* and *Cfb* displayed no difference between control and PAN rats. By immunohistochemistry, C3d signal was clearly observed along GBM in a linear-granular pattern at day 10, suggestive of podocyte localization. In addition, *C1qb*, *C4b*, *C2, C3* and *Cfb* mRNA were detected in normal cultured podocytes. More importantly, *C1qb*, *C4b, C2* and *C3* showed a PAN dosage-dependent increment. Nevertheless, *C1s*, *Serping1*, *Mbl1*, *C5* and *C9* was not detected both in normal and PAN-treated podocytes. *Cfb* showed no differences between normal and PAN-treated podocytes.

**Conclusions:** Our results firstly demonstrated that some complement molecules expressed by podocytes contribute to the activation of the complement classical pathway in nephrotic rats, which might be associated with the development of proteinuria. These findings prompt us to know podocyte as an immune cell, and open a new field in exploring the function of podocyte in the pathogenesis of proteinuria.

## Abstract# 170

**Successful Treatment of Recurrent Focal Segmental Glomerulosclerosis after Renal Transplantation by Lymphocytapheresis and Rituximab** S. Nishio, M. Shimizu, T. Yokoyama, K. Ohta, A. Yachie. *Department of Pediatrics, School of Medicine, Institute of Medical, Pharmaceutical, and Health Sciences, Kanazawa University, Kanazawa, Japan.*

**Objectives:** Treatment of patients with recurrent FSGS after renal transplantation remains a significant clinical challenge. We describe a case of sustained remission of recurrent NS after living-related donor renal transplantation using lymphcytapheresis (LCAP) and rituximab.

**Methods:** Subsets of circulating lymphocytes were analyzed by flowcytometry using monoclonal antibodies before and after LCAP and rituximab in each procedure.

**Results:** The patient is a 20-year-old woman who developed nephritic syndrome (NS) due to FSGS at the age of 6 year. She progressed to end-stage renal failure and received renal transplantation from his father at the age of 15 year. One year after renal transplantation, she had recurrence of FSGS. She was treated with 4 times of LCAP with steroid pulse therapy (methylprednisolone, 500mg/day, 3days, 3weeks). After these treatments, proteinuria had decreased to the range between 1-2g/day. However, she had second recurrence of FSGS two years later. To achieve further immunomodulation, she was treated with 4 times of LCAP and two doses of rituximab(375mg/m2). After these treatments, complete remission was achieved. She had no infectious episodes during this period. After lymphocytapheresis and rituximab, the total number of B cells and HLA-DR positive activated T cells decreased.

**Conclusions:** LCAP combined with rituximab may be an effective treatment of recurrent FSGS in the graft. Our results suggest that it might be important to regulate the interaction between T cells and B cells by B cell depletion.

## Abstract# 171

**Interleukin 17 (IL17) and B Cell Activating Factor (BAFF) Serum Levels in Children with Systemic Lupus Erythematosus (SLE)** B. Ranchin,[1] A. Doreau-Bastid,[2] B. Riche,[3] C. Ludwig,[4] A.L. Adra,[4] P. Cochat,[1] N. Bonnefoy-Berard.[2] *[1]Pediatric Nephrology Unit, Hospices Civils de Lyon, Bron, France; [2]Inserm U851, Lyon, France; [3]Biostatistic Unit, Hospices Civils de, Lyon, France; [4]Pediatric Department, Arnaud de Villeneuve Hospital, Montpellier, France.*

**Objectives:** IL7 and BAFF serum levels are elevated in adults with SLE. These cytokines act synergistically to control survival, proliferation and differentiation of B cells. Aims of this study were to assess IL7 and BAFF serum levels in children with SLE and to compare with adults.

**Methods:** IL-17 and BAFF were measured by ELISA in a prospective cross-sectional cohort of 20 children (3 boys, mean±SD for age 14.7±1.9 years) and 52 adults with SLE, 12 children (3 boys, 14.2±3.3 years) and 38 adult controls. 13/20 children presented lupus nephritis (active in 6). Statistic comparisons used Wilcoxon test.

**Results:**

Serum IL17 and BAFF levels (pg/mL, median [Q25-Q75])

| | Children | | | Adults | | |
|---|---|---|---|---|---|---|
| | SLE | Controls | p | SLE | Controls | p* |
| N | 20 | 12 | | 52 | 38 | |
| IL17 | 243 [72-441] | 11 [6-17] | <0,001 | 149 [41-328] | 14 [6-25] | 0.064 |
| BAFF | 368 [247-511] | 90 [34-123] | <0,001 | 325 [173-470] | 99 [21-121] | 0.277 |

*: children vs adults patients Mean±SD for SLEDAI score, serum C3, C4, anti-dsDNA Ab and urinary protein/creatinine ratio were respectively: 8.3±8.0, 0.83±0.37 g/L, 0.11±0.07 g/L, 156±239 IU/L and 196±146 mg/mmol in children.

**Conclusions:** IL-17 and BAFF serum levels are both increased in children with SLE, such as in adults. We plan to study their potential correlation with SLE activity markers and to follow longitudinally their levels in our patients since this may lead to further new therapeutic approaches.

## Abstract# 172

**Antibody Response to IgA-Binding Streptococcal M Proteins in Children with IgA Nephropathy** R. Schmitt, G. Lindahl, D. Karpman. *Department of Pediatrics, Clinical Sciences Lund, Lund University, Lund, Sweden; Division of Medical Microbiology, Department of Laboratory Medicine, Lund University, Lund, Sweden; Department of Pediatrics, Clinical Sciences Lund, Lund University, Lund, Sweden.*

**Objectives:** IgA nephropathy (IgAN), the most common glomerulonephritis worldwide, is characterized by mesangial deposits containing predominantly IgA. IgAN occurs or exacerbates after upper respiratory tract infections such as streptococcal pharyngitis. Certain group A streptococci have IgA-binding regions (IgA-BR) in their M protein. We have previously shown that these streptococcal IgA-BRs co-localize with mesangial IgA in IgAN.

**Methods:** In this study we used blood samples from patients with IgAN (n=20) and age-matched controls (n=83) to study the antibody response to the IgA-BRs of M-protein serotypes M4, M22 and M60, and to M4-N (the non-IgA binding N-terminal region of M4).

**Results:** Levels of IgG against the IgA-BR of M4 protein were significantly higher in IgAN patients than controls (p=0.023) and correlated significantly with antibodies to M4-N (r=0.639, p=0.002) indicating an immunological reaction to the entire M4 protein. Antibodies to the IgA-BR of streptococcal serotypes M22 and M60 were not significantly elevated in patients, but when antibody levels to all three IgA-BRs were combined, the levels in patients were increased to an extent that approached significance (p=0.058).

**Conclusions:** The results suggest that many children with IgAN had a previous infection with a streptococcal strain expressing an IgA-binding M protein. The low antibody responses may be explained by a long time-span between the infection and the debut of IgAN, and/or by low immunogenicity of the IgA-BRs.

## Abstract# 173

**Increased Aberrant IgA Production and Development of Immune-Complex Mediated Glomerulonephritis in Wiskott-Aldrich Syndrome Protein Deficiency** M. Shimizu. *Department of Pediatrics, School of Medicine, Institute of Medical, Pharmaceutical, and Health Sciences, Kanazawa University, Kanazawa, Japan.*

**Objectives:** Wiskott-Aldrich syndrome (WAS) is a rare X-linked primary immunodeficiency syndrome caused by mutations in the WAS protein (WASP) gene. IgA nephropathy is a kidney complication in WAS, however, kidney histopathology information is scarce. We set out to investigate kidney histopathology and immunopathological characteristics in WASp deficient mice.

**Methods:** Kidney samples were used for histological study and immunofluorescence (IF) study. Serum IgA, C3, urea, urinary albumin and creatinine, IgA-containing immune complexes were measured using ELISA. Aberrant glycosylation of IgA molecule were examined using lectin-binding assay. IgA production was assessed using LPS stimulasting splenic B cells.

**Results:** Histological analysis showed mesangial proliferative changes in mice older than six months and the severity of these findings increased with age. IF study showed significantly more intense glomerular IgG, IgA and C3 deposition in the kidneys of Wasp-KO mice compared to wild type mice (WT). Electron microsciopic analysis showed mesangial and paramesangial deposits were present. IgA and circulating IgA-containing immune complexes in the sera showed significantly higher titers in Wasp-KO mice compared to WT. A lectin-binding study revealed a reduced ratio of sialylated and galactosylated IgA in the sera of from old Wasp-KO mice. IgA production by LPS-stimulating B cells was increased in Wasp-KO mice compared to WT.

**Conclusions:** Increased IgA production induced by the intrinsic defect of B cells and aberrant glycosylation of IgA may be critically involved in the pathogenesis of glomerunephritis in WAS.

## Abstract# 174

**Sclerosing Peritonitis as a Complication of Lupus Peritonitis in Childhood Systemic Lupus Erythematosus** J. Dehoorne,[1] R. Joos,[1] A. Raes,[2] A. De Guchtenaere,[2] J. Vande Walle.[2] *[1]Pediatric Nephrology/ Rheumatology, UZ Gent, Gent, Belgium; [2]Pediatric Nephrology/ Rheumatology, UGent, Gent, Belgium.*

**Objectives:** Sclerosing peritonitis (SP) is a rare chronic inflammatory disease characterised by fibrosis and adhesion of the peritoneum. SP is an unusual complication observed in continuous ambulatory peritoneal dialysis, or secondary to the use of beta-blockers and peritoneo-venous shunts.

**Methods:** We describe an 11 year old Black-African female with systemic lupus erythematosus (SLE) who developed SP with chylous ascites secondary to chronic lupus peritonitis.

**Results:** At the age of 9, she was diagnosed with SLE when she presented with severe skin lesions pancytopenia, hypocomplementemia and positive serology. One year after diagnosis, lupus peritonitis appeared with massive ascites and pleural effusion, responding to treatment with IV pulse methylprednisolone. 22 months later she redeveloped asymptomatic progressive abdominal distension. Abdominal CT demonstrated multiple adhesions of the small bowel, with thickening of the bowel wall and massive ascites. Abdominal tap revealed a milky fluid with triglycerides 1180 mg/dL, 2470 leukocytes/µL albumin 2.29 g/dL, and glucose 1.10 g/L; without malignant cells or cultured bacteria. Laparoscopy showed a sclerosing peritonitis with multiple adhesions of the intestines. Histology indicated diffuse fibrosis and sclerosis with a chronic inflammatory infiltrate and small vessel vasculitis. Lymphoscintigraphy showed partial obstruction of the abdominal ductus thoracicus and lympaticus dexter.

**Conclusions:** A diagnosis of SP was established based on these features. To the authors' knowledge this is the first case report of SP in childhood SLE.

## Abstract# 175

**No Genetic Abnormalities in Complement System Regulators in Children with C3NF + Acquired Partial Lipodystrophy** J. vd Deure,[1] D. Westra,[2] N. vd Kar.[2] *[1]Pediatrics, Deventer Ziekenhuis, Deventer, Netherlands; [2]Ped. Nephrol., Radboud University MC, Nijmegen, Netherlands.*

**Objectives:** Acquired partial lipodystrophy (S. Barraquer-Simons; APL) is characterized by gradual loss of adipose tissue of the upper extremities and trunk. Many patients have low C3 plasma concentrations and pos. C3 nephr factor (C3NF+), an autoantibody stabilizing C3 convertase C3bBb. Lipodystrophy is explained as the effect of the C5-9 membrane attack complex, due to stabilized C3bBb, on the adipocyte, which in turn produces C3, factor B and factor D, enhancing complement activation. 20% of the patients develop MPGN a decade after onset of APL. It is not understood why some patients develop MPGN. Serological levels of complement factors and proteins other than C3 have been reported to be normal, but a dysregulation of the alternative complement pathway might be involved. We have looked in three unrelated patients for mutations in genes encoding regulatory proteins of the alternative pathway: complement factors H (CFH), I (IF), D (FD), and membrane cofactor protein (MCP).

**Methods:** Mutational screening on genomic DNA, by means of PCR and DNA sequencing, was performed on genes encoding CFH, IF, FD, and MCP in three patients and controls.

**Results:** Patient characteristics: three female patients age 9-14 yrs, (age at onset 7-9 yrs), C3NF+ with low C3 values, showing no signs of MPGN.
No mutations in CFH, IF, FD, or MCP could be identified.

**Conclusions:** To our knowledge, this is the first study in which genomic DNA for complement regulatory genes in APL patients are examined. In our three APL patients no mutations in CFH, IF, FD, or MCP were identified. No genetic explanation for complement activation of the alternative pathway was found.

## Abstract# 176

**Clinicopathological Study of Childhood Lupus Nephritis in Malaysia**

Y.-C. Yap, Y.N. Lim. *Department of Paediatrics, Hospital Kuala Lumpur, Kuala Lumpur, Malaysia; Hospital Kuala Lumpur, Kuala Lumpur, Malaysia.*

**Objectives:** To assess clinical characteristics, pathological findings and identify risk factors predicting chronic kidney disease (CKD) among children with lupus nephritis.

**Methods:** Data for children below 15 years old who have lupus nephritis and who underwent renal biopsies from 1991 to 2008 were abstracted from the Malaysia Renal Biopsy Registry.

**Results:** There was a total of 189 children (161 girls and 28 boys) with lupus nephritis, mean age 11.5±2.8 years. The racial distribution was Malay (61.4%), Chinese (25.4%), Indian (4.7%) and others (8.5%). 83% fulfilled 4 or more ARA criteria. The presentation were nephrotic syndrome (32.3%), nephritic syndrome (17.5%), urinary abnormalities (19.6%), nephritic and nephrotic syndrome (15.9%). At the time of biopsy, 5.8% of children were on dialysis. Renal biopsy revealed Class II, III, IV, V and VI nephritis in 8.5%, 17.5%, 66.5%, 6.9% and 0.5% patients, respectively. The prevalence of renal impairment (GFR <60ml/min/1.73m2, by Schwartz Formula) correlated with histology findings. The median duration of follow up was 12 months. Of the 189 patients, 13 died, 27 had CKD (stage 3-5) and 10 had end stage renal failure. Renal survival rate was 97.1% at 3 years and 5 years. Risk factors for progressing to CKD were dialysis at time of biopsy (p=0.04) and moderate tubulointerstitial inflammation in histology (p=0.04). The complication rate of biopsy procedure was 4.9%, higher risk among those on dialysis therapy and haemoglobin level ⟨10g/dL.

**Conclusions:** The characteristic of childhood lupus nephritis in Malaysia are comparable with other series. Dialysis at the time of biopsy and moderate tubulointersitial inflammation were the risk factors for CKD.

## Abstract# 177

**Urinary Heme Oxygenase-1 Is a Novel Biomarker for an Inflammatory Tubulointerstitial Damage in Acute Kidney Injury**

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**Objectives:** In this study, we examined intracellular HO-1 expression in urine sediments (uHO-1) of patients with various renal diseases, to assess whether uHO-1 can be an alternative biomarker to detect tubulointerstitial inflammatory damages in acute kidney injury (AKI).

**Methods:** A total of 61 urine samples from patients with various renal diseases and healthy children were examined. Lysates of the urinary sediments were prepared and the levels of uHO-1 were measured by enzyme-linked immunosorbent assay.

**Results:** The levels of uHO-1 in patients with hemolytic uremic syndrome (HUS), acute tubulointerstitial nephritis (TIN), acute glomerulonephritis and IgA nephritis (IgAN) were high. In contrast, the majority of patients with minimal change nephrotic syndrome, prerenal failure, non-glomerular bleeding and Kawasaki diseases showed normal uHO-1 levels. IgAN patients with interstitial cellular infiltrations showed higher levels of uHO-1 than the patients without these findings. In AKI patients, the levels of uHO-1 were increased only in patients with inflammatory tubulointerstitial changes such as HUS and TIN. The patterns of kinetics of uHO-1 in patients with AKI were different among each background diseases.

**Conclusions:** These results indicate that the potential usefulness of uHO-1 as a novel, noninvasive biomarker to evaluate the degree of tubulointerstitial inflammatory damages in AKI.

## Abstract# 178

**Soluble Interleukine 2 Receptor and MDR1 Gene Expression Levels as Inflammatory Biomarkers for Prediction of Steroid Response in Children with Nephrotic Syndrome**

D.M. Youssef,[1] R.M. Elbehidy,[1] H.S. Abdelhalim,[2] G.E. Amr.[2] *[1]Pediatrics, Zagazig University, Zagazig, Zagazig, Egypt; [2]Clinical Biochemistry, Zagazig University, Zagazig, Zagazig, Egypt.*

**Objectives:** The present study was designed to estimate serum (*sIL2R*) levels and *MDR*-1 gene expression on lymphocytes in NS to elucidate relationship between there level and clinical relevance to corticosteroids therapy.

**Methods:** We examined 40 patients with NS group A; they were 15 cases with recent onset NS and 25 known cases of NS and 20 healthy children as a control group B. We examined every patient twice 1st with activity and 2nd within one week of remission.

**Results:** we found a significant increase in *sIL-2R* level and *MDR1* gene expression in patients group in comparison to control group whether in *activity* or *remission* and also they were significantly higher in *activity* than in *remission* as *sIL-2R* was (1197.2183.3) Iu\L in group A in *activity*, (791192.6) Iu\L in *remission* and (44927.3) Iu\L in control group B and *MDR1* gene was (8.70.86) % in group A in *activity*, (6.31.4) % in *remission* and (2.80.87) % in control group and p value was <0.05 in comparing groups. Levels of *sIL-2R* and *MDR1* gene expression in different subgroups are higher in old cases than new ones both in *activity* and *remission* and relatively higher levels in steroid resistant NS than steroid sensitive ones.

**Conclusions:** We propose using *sIL2R* and /or *MDR1*gene expression levels as early predictors of steroid resistance in NS to promote early control of disease, and as this is the first report providing new insight into use of *sIL2R* as a predictor of steroid resistance a wide scale study is needed to determine a cut off level of *sIL2R* above it cytotoxic drugs are introduced.

## Acute and Chronic Renal Failure

## Abstract# 179

**(O-25)**

**Vitamin D Receptor Activation (VDRA) Ameliorates Cachexia and Inflammation in Chronic Kidney Disease (CKD)**

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**Objectives:** We showed that cachexia in CKD is associated with inflammation and maladaptive energy homeostasis(Cheung et al JCI 2005). Vitamin D(VD) deficiency is prevalent in CKD. We studied the effect of VDRA in a mouse model of CKD.

**Methods:** 8-wk old c57BL/6J mice underwent 5/6 nephrectomy(N) or sham operation(S). N mice received either vehicle or paracalcitol(PC)(N-PC) (0.15 mg/kg ip 3X per week). S mice received vehicle. N mice were fed ad lib, the other 2 groups of mice were pair-fed for 2 weeks.

**Results:** Serum creatinine was higher in N and N-PC than in S mice($p<0.01$). Serum 25-VD3 and 1,25-VD3 were lower in N mice than in S mice($p<0.01$). S & N-PC mice gained more weight than N mice ($p<0.01$) with equal food intake. Basal metabolic rate was higher in N than S and N-PC mice($p<0.01$). Efficiency of food consumption was lower in N mice than S and N-PC mice. N mice lost both lean body mass and fat mass whereas S and N-PC mice gained lean body mass and fat mass. mRNAs of uncoupling proteins(UCP)-1 and 2, which control energy expenditure, were upregulated in both skeletal muscle and adipose tissue in N compared with S and normalized in N-PC mice. mRNAs of myogenic pathway genes, IGF-I, MyoD and PAX3, were all downregulated in the skeletal muscles in N compared with S and normalized in N-PC mice. IL-6 mRNAs in skeletal muscle and adipose tissue was upregulated in N compared with S and normalized in N-PC mice.

**Conclusions:** VDRA ameliorated cachexia and reversed cytokine over-expression in a mouse model of CKD. VD deficiency may be an important factor in the pathogenesis of inflammation and cachexia in CKD.

## Abstract# 180

**(O-26)**

**Puromycin-Induced Mitochondrial Alterations in Podocyte Apoptosis**

S. Jeruschke,[1] A. Peters,[1] S. Kummer,[1] V. Wegerich,[1] A. Seibt,[1] J. Weiss,[2] K. Jeruschke,[2] E. Mayatepek,[1] J. Oh.[1] *[1]Department of General Pediatrics, University Children's Hospital, Duesseldorf, Germany; [2]Institut for Clinical Biochemistry, German Diabetes Center, Duesseldorf, Germany.*

**Objectives:** Cellular stress of podocytes results in proteinuria, apoptosis and glomerular sclerosis. During apoptosis podocyte mitochondria appear to have a central role in the development of chronic kidney disease. Dexamethasone (DEX) is known to protect podocytes from apoptosis induced by puromycin (PAN). The underlying mechanism is still unknown. We studied the effects of PAN and DEX on mitochondrial function and morphology in murine and human podocytes.

**Methods:** Podocytes were treated with PAN, DEX or both in combination. Apoptosis was confirmed by FACS-analysis and Hoechst staining. To visualize mitochondrial morphology and membrane potential, cells were stained with the fluorescent cation TMRM and anti-Cox-4 antibody.To study morphological details, transmission electron microscopy (TEM) was performed.

**Results:** After treatment with PAN, TMRM fluorescence intensity decreased in a time-dependent manner, indicating mitochondrial depolarisation ($p\leq0.02$; compared to untreated podocytes). Combination of DEX and PAN fully prevented this effect. The mitochondrial morphology showed an increase of shorter mitochondria under PAN. In addition, destruction of mitochondrial cristae and modification of intracellular distribution were seen (TEM and anti-Cox-4 staining). The morphological alterations could not be fully prevented by DEX.

**Conclusions:** PAN induced time-dependent alterations in membrane potential and morphology of mitochondria, which were followed by apoptosis in podocytes. These effects could be partially prevented by DEX treatment.

## Abstract# 181
**(O-27)**

**The Serum and Glucocorticoid-Regulated Kinase-1 (SGK1) in Hypoxic Renal Injury** K. Rusai,[1] C. Schmaderer,[2] M. Strobl,[2] K.M. Boini,[3] A. Grenz,[3] D. Kuhl,[4] U. Heemann,[2] A.J. Szabo,[1] F. Lang,[3] J. Lutz.[2] *[1]Department of Pediatrics, Semmelweis University, Budapest, Hungary; [2]Department of Nephrology, Technical University, Munich, Germany; [3]Department of Physiology and Pharmacology, University of Tübingen, Tübingen, Germany; [4]Department of Biology, Chemistry, and Pharmacy, Free University, Berlin, Germany.*

**Objectives:** SGK1 is a novel serine-threonine kinase up-regulated by different cell stress stimuli such as hyperosmosis, heat shock, oxidative stress, and counteracts apoptosis. Although effects of SGK1 have been intensively studied, to date, nothing is known about its role in renal hypoxic injury.

**Methods:** *In vitro*, HEK293 cells were subjected to hypoxia/reoxygenation (H/R), where expression and effects of SGK1 using plasmid-mediated overexpression were studied. *In vivo*, SGK1 up-regulation and activation were explored in rat renal ischemia/reperfusion (I/R), and its effect on tissue injury was investigated using *sgk1-/-* knock-out mice.

**Results:** *In vitro*, H/R increased SGK1 transcript and protein levels and phosphorylation. H/R further enhanced apoptosis of cells, an effect inhibited by prior SGK1 overexpression. *In vivo* I/R injury increased SGK1 transcript and protein levels, and activation. I/R furthermore enhanced apoptosis of tubular cells, which was more pronounced in gene targeted mice lacking *sgk1*.

**Conclusions:** In conclusion, SGK1 is up-regulated by and counteracts apoptosis following H/R *in vitro* and ischemia *in vivo*. Our results strongly indicate that the kinase participates in the machinery fostering cell survival under stress conditions representing thereby an attractive target of intervention under ischemia in the kidney.

## Abstract# 182
**(O-28)**

**Prevalence of Dyslipidemia, Carotid Intima Media Thickness and Endothelial Dysfunction in Children with Chronic Kidney Disease (CKD)** V. Murugan,[1] S. Hari,[2] P. Hari,[1] A. Bagga.[1] *[1]Pediatrics, All India Institute of Medical Sciences, Delhi, India; [2]Radiodiagnosis, AIIMS, Delhi, India.*

**Objectives:** To determine lipid profile, carotid intima media thickness (CIMT) and brachial artery flow mediated dilatation (FMD) in children with CKD and compare them with controls.

**Methods:** Cases included 80 children with CKD III-IV of >2 yr duration while 42 age and sex matched healthy children served as controls. All children underwent CIMT and brachial artery FMD determination using high resolution ultrasonography. Total cholesterol (Tc), low density lipoprotein (LDL), very low density lipoprotein (VLDL), high density lipoprotein (HDL) and triglycerides (Tg) were estimated.

**Results:** The mean age of cases was 9.5±3.4 yr. 45% were malnourished and 87% had hypertension. 35% cases had at least one abnormal lipid parameter. The mean maximum CIMT was significantly higher and brachial artery FMD was significantly lower in cases than in controls. Tc, Tg, LDL, VLDL were significantly higher in cases than controls.

Comparison of CIMT, FMD and lipid profile [mean (SD)]

| | Cases | Controls | P |
|---|---|---|---|
| Maximum CIMT (mm) | 0.433 (0.05) | 0.397 (0.03) | 0.0000 |
| FMD (% increase) | 12.0 (8.8) | 18.6 (14.3) | 0.002 |
| Tc (mg/dl) | 181.5 (45.4) | 138.9 (20.3) | 0.0001 |
| Triglyceride (mg/dl) | 138.3 (41.5) | 86.5 (21.8) | 0.0000 |
| LDL (mg/dl) | 111.0 (41.5) | 77.5 (21.1) | 0.0007 |

On regression analysis, Tc, BMI, systolic blood pressure and LDL were independent determinants of CIMT.

**Conclusions:** Dyslipidemia is seen in 35% children with CKD. These children have significantly increased CIMT and decreased brachial artery FMD.

## Abstract# 183
**(O-29)**

**Acute Kidney Injury Related to Toxic Exposures** A.M. Vilay, C.S. Wong, R.M. Schrader, R.C. Mercier, S.A. Seifert. *University of New Mexico, Albuquerque, NM, United States.*

**Objectives:** To characterize acute kidney injury (AKI) associated with pediatric exposures documented in the American Association of Poison Control Centers (AAPCC) database.

**Methods:** This was a retrospective, case-controlled study of single-substance exposures reported to the AAPCC database between 2001-2007 in individuals <20 years of age and coded with an exposure-related renal effect. AKI was defined as elevated serum creatinine, oliguria/anuria, and/or renal failure. Substances identified as renal toxins a priori and those associated with ≥50% incidence of AKI were compared to other substances in the database. A chi-squared analysis identified agents associated with AKI.

**Results:** 5332 pediatric exposures were coded with a related renal effect. The case fatality rate was >500 times greater in pediatric exposures with related renal effects than those without. There were 2711 single-substance exposures, with AKI occurring in 26% (males 56%; females 43%). The highest incidence of AKI was in 13-19 year olds (42%). The vast majority (94%) of exposures in ≤12 year olds were unintentional while for 13-19 year olds the most common reason was intentional/suspected suicide (41%). Of single-substance medication exposures with related renal effects, the highest rates of AKI occurred with acetaminophen (84%), lithium (52%), non-steroidal anti-inflammatories (74%), and salicylates (81%).

**Conclusions:** Substances in the database associated with high rates of AKI among children are known nephrotoxins. This is the first large database study associating AKI with acetaminophen. This data will direct future areas of research aimed at prevention and early intervention of kidney injury resulting from pediatric toxic exposures.

## Abstract# 184
**(O-30)**

**Hemolytic Uremic Syndrome Associated to Sorbitol Fermenting O157: H$^-$ Shiga Toxin Producing Escherichia coli** A. Rosales,[1] J. Hofer,[1] M. Riedl,[1] T. Jungraithmayr,[1] R. Würzner,[2] H. Karch,[3] L.-B. Zimmerhackl.[1] *[1]Department of Pediatrics 1, Medical University of Innsbruck, Innsbruck, Austria; [2]Division of Hygiene and Medical Microbiology, Medical University of Innsbruck, Innsbruck, Austria; [3]Institut für Hygiene, Universität Münster, Münster, Germany.*

**Objectives:** Infections with Shiga toxin producing *Escherichia coli* (STEC) are the main cause of hemolytic uremic syndrome. Recently, a new STEC strain, which unlike O157:H7 ferments sorbitol and has been found only in humans, was associated to HUS cases in Europe. The clinical presentation of HUS due to sorbitol fermenting (SF) O157:H$^-$ is not well described.

**Methods:** From January 1st 1997 to December 31st 2002, 628 patients <21 years of age with the clinical diagnosis of HUS were registered in a prospective multicenter study in Austria and Germany.

**Results:** SF O157:H$^-$ STEC were found in 44 out of 298 cases in which STEC serotype could be determined. SF O157:H$^-$ did not show a seasonal distribution. These patients had a severe acute illness presenting with hypertension (22.6%) and neurological symptoms (41.5%) more frequently than O157:H7 cases. 38 of 44 cases presented with diarrhea, which was bloody in 22 of them. Among the patients with SF O157:H$^-$ STEC, 62.2% needed hemodialysis and 27.3% plasmapheresis, both significantly more frequently than in O157:H7 patients ($p<0.05$, $x^2$). Regarding the long term outcome, a significant association between SF O157:H$^-$ STEC and the presence of symptoms after one year was found.

**Conclusions:** The detection of STEC should not be limited to the O157:H7 serotypes, since SF O157:H$^-$ and other non-O157:H7 serotypes are highly virulent and are associated to severe course of disease.

## Abstract# 185
**(O-31)**

**Urinary Nitrate Detects Acute Kidney Injury in Children Presenting to the Emergency Department** A.I. Mian,[1] Y. Du,[2] H. Garg,[3] A.C. Caviness,[1] S. Goldstein,[2] N. Bryan.[3] *[1]Pediatrics-Emergency Medcine, Texas Children's Hospital /Baylor College of Medicine, Houston, TX, United States; [2]Pediatrics-Nephrology, Texas Children's Hospital /Baylor College of Medicine, Houston, TX, United States; [3]Brown Foundation Institute of Molecular Medicine, University of Texas-Houston Health Science Center, Houston, TX, United States.*

**Objectives:** Nitric oxide (NO) is perturbed during kidney injury. Our objective was to assess the ability of the urinary NO metabolite nitrate to detect early acute kidney injury(AKI) in the emergency department (ED).

**Methods:** Patients (pts) were recruited if they had a urinalysis and serum creatinine (SCr) obtained. Using SCr in the ED and after hospitalization, AKI was defined by validated pediatric pRIFLE (Risk[R], Injury[I], Failure[F]) criteria; pts without AKI were controls. Urinary nitrate was determined by HPLC. Statistical comparison between groups was done using non-parametric methods and t-test. Diagnostic accuracy was assessed using ROC curves.

**Results:** Urine nitrate was assayed from 252 pts. Mean (SD) age was 11.4(4.8) yrs, (50% male), 138 (60%) admitted. AKI results: 233 controls, 12 pRIFLE-R, 6 pRIFLE-I. Absolute median urinary nitrate and urinary creatinine normalized mean log nitrate were significantly lower for the injury group (pRIFLE-I) compared with the combined risk (pRIFLE-R) and no AKI groups ($p=0.012$ and $p=0.026$, respectively). Urinary nitrate demonstrated good accuracy (AUC 0.726; 95%CI 0.609-0.844) to predict pts with pRIFLE-I versus pts with pRIFLE-R or without AKI.

**Conclusions:** Low urinary nitrate detects AKI early in the pediatric ED. Future studies will determine the biomarker's ability to predict AKI progression in hospitalized patients.

## Abstract# 186

**(O-32)**

**The Effect of Losartan on NA/K-Atpase (NKA) and Heat Shock Protein (HSP) 72 in Rats with Diabetic Nephropathy** L. Wagner,[1] N.F. Banki,[2] A. Prokai,[2] A.J. Szabo,[2] A. Ver,[3] A. Fekete.[2] *[1]Dept. of Transplantation and Surgery, Semmelweis University, Budapest, Hungary; [2]1st Dept of Pediatrics, Semmelweis University, Budapest, Hungary; [3]3Dept. of Med. Chem., Mol. Biol. and Pathobiochem, Semmelweis University, Budapest, Hungary.*

**Objectives:** Angiotensin II (AII) is an important mediator of renal disease in diabetic nephropathy. The angiotensin receptor blocker losartan delays complications. In physiologic conditions the effective NKA is bound to the tubular basal membrane. This connection is protected by HSP72. AII and diabetes increase the activity of NKA, however the effect of losartan is unkonown in this situation.

**Methods:** Control (C), streptozotocin(STZ)-diabetic (D), AII-treated C and D (CA and DA), and losartan treated D (DL) Wistar rats were studied after 7 weeks of treatment (STZ: iv 65 mg/bwkg, AII: 24h sc 33 µg/bwkg/h, losartan: po 5 mg/bwkg/day, 2 weeks). We determined the expression and intracellular localization of renal NKA and HSP72.

**Results:** The expression and localisation of NKA in the cytosolic fraction increased in D and CA animals vs. C, and we saw a further increase in DA rats. Losartan prevented this effect of diabetes. The expression of HSP72 increased in D, more in DA rats vs. C, which effect was reduced by losartan (D vs. DL).

**Conclusions:** Diabetes and AII lead to NKA increase, however the internalisation of the enzyme increases as well, thus the proportion of effective NKA decreases. Though these changes were reversible by losartan, we assume that this effect is mediated by the Ang 1 receptor. The parallel changes of HSP72 may reflect compensatoric-protective processes.

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## Abstract# 187

**Cystometry Findings in Children with Chronic Renal Failure and Suspected Bladder Dysfunction** H. Öborn, M. Herthelius. *Department of Clinical Science, Intervention and Technology, Karolinska Institutet, Stockholm, Sweden.*

**Objectives:** To examine children with chronic renal failure (CRF) and suspected bladder dysfunction with cystometry in order to explore the need for invasive urodynamics in pre-transplant evaluation.

**Methods:** During 2006-2008 all children at our center with CRF of grade 3 or more were evaluated for bladder dysfunction by means of a comprehensive history, a bladder diary, uroflowmetry and bladder ultrasound. Bladder dysfunction was suspected in 29 of 40 patients. These patients were recommended further investigation with cystometry. Four patients received a kidney transplant before being approached, 6 refused investigation and 4 were lost to follow up, which left 15 for investigation.

**Results:** The most common finding was a combination of large cystometric bladder capacity (CBC) and reduced bladder sensation (8 patients, 53%). These results were similar to the results obtained by non-invasive urodynamics (9 patients, 60%, with a large bladder capacity). A large CBC and normal sensation was found in 2 patients and a large CBC and detrusor overactivity in one. Severe abnormalities, i.e. reduced compliance +/- detrusor activity, were found in 2 patients only. Cystometry was normal in 2 patients. Both of these patients had a slightly elevated physiological bladder capacity of 160-165% of expected for age as the only sign of bladder dysfunction.

**Conclusions:** Cystometry confirmed that, in this group of children, the most common finding was a large bladder capacity and reduced bladder sensation. Cystometry did not add much information to the one obtained by non-invasive urodynamics in those cases.

## Abstract# 188

**The Profile of Acute Kidney Injury in Children with Shock and the Utility of Cystatin C as an Early Marker of Acute Kidney Injury** I. Agarwal,[1] J. Mohan,[1] K. Ranjini,[2] E. Jacob,[2] P. Prashanth.[1] *[1]Child Health and Pediatric Nephrology, Christian Medical College, Vellore, Tamil Nadu, India; [2]Pediatric Intensive Care Unit, Christian Medical College, Vellore, Tamil Nadu, India.*

**Objectives:** To study the profile of Acute Kidney Injury (AKI) in shock and the utility of Cystatin-C as an early marker of AKI.

**Methods:** Children admitted in PICU with fluid refractory shock were evaluated. Cystatin C levels were checked at admission and 24 hours later and compared to Serum Creatinine and Creatinine clearance. Data was entered on Epidata and statistical analysis was done using SPSS-12.

**Results:** Of the 547 admissions to PICU, 93 children with refractory shock were studied further. 73% had septic shock, 18% hypovolemic and 9% had cardiogenic shock. The incidence of Acute Kidney Injury (by pRIFLE) was 43 % (40/93). 31% had risk, 6% renal injury and 5% had renal failure. Mortality was 53.7% (50/93). Mortality with AKI (65%) was higher than the non AKI group (45%).Of the total mortality, 48% (24/50) had normal renal function, 38% had Risk, 10% had Injury and 4% had Failure. The significant factors associated with mortality in AKI were prolonged illness prior to presentation, coagulopathy, elevated liver enzymes, need for > 2inotropes, prolonged usage, need for ventilation and prolonged ventilation(p=<0.05) Creatinine was more sensitive (64.7%) vs Cystatin C (59% ) but less specific( 63%) than Cystatin C (79%).Cr Cl was highly sensitive (100%) but poorly specific (18%). Correlation of Cystatin C with creatinine was 44%.

**Conclusions:** Mortality in shock with AKI is higher. Recognition and appropriate management of early stages may help to decrease mortality. Cystatin C was a good marker for Acute Kidney Injury.

## Abstract# 189

**The Effect of a Multidisciplinary Clinic on the Outcomes in Pediatric Chronic Kidney Disease** S. Ajarmeh, G. Brin, L. Er, O. Djurdjev, J. Dionne. *Pediatric Nephrology, BC Children's Hospital, Vancouver, BC, Canada.*

**Objectives:** To describe the effect of multidisciplinary care (MDC) on the clinical outcomes of children with chronic kidney disease (CKD) in British Columbia.

**Methods:** The MDC clinic started at BC Children's Hospital in 2006. In this cross-sectional retrospective study, we analyzed the data of all patients seen in 2003 (n=73) and 2009 (n=125). Patient demographics and laboratory results were analyzed.

**Results:** Patient demographics were similar though CKD stage is significantly lower in 2009 (P<0.05). Hemoglobin level was significantly better in 2009 (130 g/L vs. 122 g/L, P<0.05) and varied by cohort and CKD stage. In 2009,78% of patients reached KDOQI calcium target compared to 55% in 2003 (P<0.05). Calcium level varied by cohort and CKD stage (P<0.05). Phosphate and PTH varied only by CKD stage. Albumin level was significantly better in 2009 (44 g/L vs. 40 g/L, p<0.05) and varied by CKD stage and cohort. Blood pressure control was better in 2009 with 17% hypertensive on evaluation compared to 30% in 2003 (P<0.05). There was no significant difference in growth or renal disease progression. Patients were significantly more likely to see social worker or pharmacist in 2009 and a dietician if they had CKD stages 3-5 (P<0.05). Allied health support was essential as the median number of medications per patient in 2009 was 4.0 (max 10) and 30% were receiving caloric supplements. Hospitalization events were comparable in the two groups but the total length of stay in 2009 was shorter (median days 0.2 vs. 3.0, P<0.05).

**Conclusions:** The multidisciplinary care clinic improved the outcome of children with CKD especially in anemia, bone metabolism and blood pressure control.

## Abstract# 190

**Retrospective Evaluation of Fluid Status and Clinical Outcomes in Critically Ill Children with Sepsis** J.R. Angelo,[1] J.M. Graf,[2] E.A. Williams,[2] C.E. Kennedy,[2] S.L. Goldstein.[1] *[1]Renal Section, Department of Pediatrics, Baylor College of Medicine, Houston, TX, United States; [2]Critical Care Medicine, Department of Pediatrics, Baylor College of Medicine, Houston, TX, United States.*

**Objectives:** To describe the epidemiology of acute resuscitative and late fluid administration, and assess potential associations between degree of daily and cumulative fluid accumulation and outcomes in septic PICU pts.

**Methods:** Retrospective chart review of all patients with sepsis admitted to the Texas Children's Hospital (TCH) PICU from 2007 to 2009 (n=300). Demographics, anthropometric measures, vital signs, lab data and fluid balance at specified time points were extracted by integration of the TCH VPS and PICU databases with medical records. Logistic regression analyses will evaluate predictors of clinical outcomes, controlling for disease severity by PRISM-III and shock index scores. Predictor variables included in the analyses are acute resuscitative (defined as 60cc/kg over the first 1 hr of admission) and late (defined by daily percentage of fluid overload (FO) as validated from the Prospective Pediatric CRRT registry) fluid administration. Selected pulmonary outcomes are FiO2, mean airway pressure, and PEEP. Cardiac outcome variables are SBP, heart rate, and CVP. Serum creatinine changes define renal function by pRIFLE and AKIN criteria.

**Results:** Multiple databases were successfully integrated for a pilot of three patients.

**Conclusions:** This is the largest pediatric population studied to date with respect to fluid management in septic ICU patients. The results of this study could inform treatment guidelines for optimal fluid administration over the course of pediatric critical illness.

## Abstract# 191

**Rasburicase in Acute Renal Failure-Related Hyperuricemia of the Newborns** A. Mastrangelo,[1] S. Ghirardello,[2] G. Ardissino,[1] A.C. Lonati,[2] A. Edefonti,[1] F. Mosca.[2] *[1]Pediatric Nephrology and Dialysis Unit, Clinica Pediatrica G. e D. De Marchi, Fondazione IRCCS Ca' Granda Ospedale Maggiore Policlinico, Milan, Italy; [2]Institute of Pediatrics and Neonatology, Fondazione IRCCS Ca' Granda Ospedale Maggiore Policlinico, Milan, Italy.*

**Objectives:** Serum uric acid is a well-known endogenous nephrotoxic agent that accumulates in course of acute renal failure (ARF).Rasburicase (RU) converts urate into allantoin,that is rapidly excreted in the urine.

**Methods:** We retrospectively reviewed 10 newborns treated with RU for hyperuricemia secondary to ARF; 6 of them had a gestational age 29,4 wks±30 day, birth weight 1227± 644 gr.RU was administered i.v., at a single dose of 0.2 mg/kg in 30 minutes;2 pts received a second dose.Seric parameters,diuresis and side effects were monitored before and 24 hrs after RU.

**Results:** Student's T-test analysis for paired data was performed and a probability <5% was considered significant.Following RU dose, a statistically significant decrease in serum uric acid concentration from (mean±sd) 14.5±3.6 mg/dL to 6.6 ± 7.48 mg/dl (p<0.002) was observed.

Table 1. Biochemistry details

| Parameter | Baseline | After rasburicase | p value |
|---|---|---|---|
| Uric acid (mg/dL) | 14,50 (± 3,56) | 6,57 (± 7,48) | p < 0.002 |
| Creatinine (mg/dL) | 4,35 (± 2,09) | 3,92 (± 1,83) | p = 0.522 |
| BUN (mg/dL) | 82.16 (± 35.5) | 88.35 (± 39.2) | p = 0.324 |
| Phosphorus (mg/dL) | 4,60 (± 2,01) | 4,06 (± 1,98) | p = 0.376 |
| Urine output (ml/kg/h) | 1,90 (± 2,28) | 2,58 (± 2,51) | p = 0.331 |

We did not find any improvement in renal function nor side effects.

**Conclusions:** RU appears to be safe and efficacious in the treatment of hyperuricemia in low birth weight neonates.

## Abstract# 192

**Low Rate of Peritonitis in Children on Peritoneal Dialysis, 5 Year Review from a Single-Centre** D. Noone, L. Edwards, S. Boyle, M. Kinlough, M. Riordan, A. Awan. *Nephrology, Children's University Hospital, Dublin, Ireland.*

**Objectives:** Report low rates of peritonitis in our center over past 5 years and discuss characteristics and outcomes.

**Methods:** Retrospective chart review of children on PD between 2004 – 2008. Episodes of peritonitis, laboratory data reviewed. Number of Peritonitis Episodes per Patient Months of PD recorded for each year. Peritonitis was diagnosed >100 WBC x 10^6/litre, >50% polymorphs ± organisms on Gram stain, supported by history and examination.

**Results:** 23 patients included in study. All on cycler dialysis. Age 8.8 ± 5.7 years. Mean Time on PD 18 ± 13 months, range 1 – 54 months. All received intravenous antibiotics at time of Double cuff Tenckhoff catheter insertion. Over 5 year period mean number of Peritonitis Episodes was 1 per 73 Patient Months of PD, range 1 per 14.3 to 1 per 95.5 months.10 episodes of peritonitis over 5 years. Mean number of peritonitis episodes per patient was 1.3 (range 1-3). Median time to onset of peritonitis post insertion of catheter was 1 month (range 2 days to 29 months).Median number of WCC was 1,050 (range 160-100,000). In 5 no organism was cultured. *Staphylococcus aureus* accounted for 4 ( 1 was relapse) and *Staphylococcus epidermidis in* 2 episode, 1 by *Pantoea spp*.2 Tenckhoff catheters removed because of *Staphylococcus aureus* peritonitis. Mean duration of treatment with intraperitoneal antibiotics 12 days (range 3-21). Vancomycin and ciprofloxacin are antibiotics used.

**Conclusions:** Low rate of peritonitis due to intense training of parents by dedicated trained nursing staff. No case of gram negative peritonitis and only 2 catheters lost over 5 years.

## Abstract# 193

**The Difference between Renin-Angiotensin-Aldosterone-System (RAAS) Inhibitors in Diabetic Nephropathy** Nó.F. Bánki,[1] Lá. Wagner,[2] Á. Prókai,[1] Á. Vér,[3] Á. Vannay,[1] A.J. Szabó,[1] A. Fekete.[1] *[1]1st. Dep. of Pediatrics, Semmelweis University, Budapest, Hungary; [2]Transplant Surgery, Semmelweis University, Budapest, Hungary; [3]Med.Chem. Mol. Biol. and Pathobiochem., Semmelweis University, Budapest, Hungary.*

**Objectives:** Diabetic (DM) nephropathy leads to end-stage renal failure. Its pathophisiology is unclear, but the role of the RAAS is presumptive. Previously we found elevated expression and translocation of the renal Na/K ATPase (NKA) in a DM rat model, which worsens renal function. Heat-shock protein (HSP) 72 provides a stable, membrane associated NKA.

**Methods:** We induced DM in male Wistar rats with iv. streptozotocin (60 mg/kg). After 5 weeks of DM we daily treated them with ACE inhibitor enalapril (40 mg/kg/day), renin inhibitor aliskiren (30 mg/kg/day) and aldosteron antagonist spironolactone (50 mg/kg/day) for 2 weeks p.o. Untreated DM, and treated non-DM animals served as controls (n=6/group). To evaluate protein levels and intrarenal localization of NKA and HSP72 we used Western blot, and immunofluorescent staining.

**Results:** DM rats had higher NKA and HSP72 protein expressions than the controls. Aliskiren, and spironolactone reduced the elevated NKA and HSP72 protein levels in DM rats (p<0.05 vs. DM). NKA moved from the basolateral membrane to the cytoplasm in tubular cells in DM group vs controls, which was prevented only by aliskiren treatment.

**Conclusions:** Higher glucose levels might be the reason for altered protein expression and distribution in DM nephropathy. Various RAAS inhibitors influence the expression and distribution of NKA, which could explain their various clinical efficacies in moderating DM nephropathy.

## Abstract# 194

**Intestinal Damage in Enterohemorrhagic *Escherichia coli* (EHEC) Infection** Z.D. Békássy,[1] C. Calderon Toledo,[1] G. Leoj,[1] A. Kristoffersson,[1] S.R. Leopold,[2] M.-T. Perez,[3] D. Karpman.[1] *[1]Pediatrics, Clinical Sciences Lund,Lund University, Lund, Sweden; [2]Pediatrics, Washington University School of Medicine, St Louis, MO, United States; [3]Ophthalmology, Clinical Sciences Lund,Lund University, Lund, Sweden.*

**Objectives:** EHEC infection leads to marked intestinal injury. The aim of the study was to investigate intestinal damage during EHEC infection in patients with hemolytic uremic syndrome and to correlate EHEC virulence factors Shiga toxin (Stx), intimin and the type III secretion system to symptoms and intestinal damage in a mouse model.

**Methods:** Sigmoideum was available from two children who underwent sigmoidectomy due to colonic perforation. C3H/HeN mice were infected with Stx-producing (86-24) and non-producing (87-23) *E. coli* O157:H7 strains as well as mutants lacking *eae,* encoding intimin (UMD619) or *escN,* regulating the expression of type III secretion effectors (CVD451). Tissues were investigated by light microscopy and TUNEL to identify dying cells.

**Results:** Human sigmoid exhibited abundant TUNEL-positive cells. Severe symptoms developed in mice inoculated with strains 86-24 and 87-23. Few mice inoculated with the mutant strains developed severe symptoms. Strain 86-24 colonized well, followed by 87-23, whereas strains UMD619 and CVD451 colonized minimally. Significantly more TUNEL-positive cells were found in proximal and distal colons of mice inoculated with strain 86-24 compared to strains 87-23, CVD451 and UMD619.

**Conclusions:** Strains 86-24 and 87-23 exhibited better colonization and more symptoms, presumably due to the presence of intimin and type III secretion effectors. Extensive intestinal mucosal cell death was related to the presence of Stx.

## Abstract# 195

**Pharmacokinetic Modeling of Renal Function Markers in Acute Renal Failure** P. Slort,[1] A. Wilhelm,[2] A. Bökenkamp.[1] *[1]Pediatric Nephrology, VUmc, Amsterdam, Netherlands; [2]Clinical Pharmacology&Pharmacy, VUmc, Amsterdam, Netherlands.*

**Objectives:** The RIFLE classification provides standardized definitions of acute kidney injury (AKI). The pediatric adaptation (pRIFLE) defines risk (R) as a decrease in estimated Schwartz-GFR by 25%, incipient (I) by 50% and failure (F) by 75%. This corresponds to a rise in creatinine (Crea) by 1.33, 2 and 4 x baseline concentration. Aim: To develop a corresponding definition for cystatin C (Cys), an alternative marker of GFR, using a pharmacokinetic model.

**Methods:** Simulation of Cys and Crea concentrations during AKI using a one-compartment model with zero-order input. Baseline GFR entered as 100 ml/min/1.73m$^2$, volume of distribution as total body water (Crea) or extracellular fluid (Cys), production rate as 20 mg/kg/day and 0.117 mg/min/1.73m$^2$, respectively. Clearance taken linear with GFR. In a second analysis, extra-renal elimination of Cys (i.e. 22.7 ml/min/1.73m$^2$) taken into account. The corresponding relative rise in Cys concentrations calculated using Cys-based GFR-estimation equations by Grubb and by Filler for comparison.

**Results:** Following an acute decrease in GFR, Cys concentrations reach a new steady state after 4 (R), 6 (I) and 13 (F) vs. 13, 19 and 39 hours (Crea). Taking only renal elimination into account, the relative rise (RR) of Cys is 1.33 (R), 2 (I) and 4 (F) x baseline concentration. If extra-renal elimination is included, RR is only 1.28, 1.79 and 2.97. The latter corresponds better with in vivo data (Grubb 1.19, 1.51, 2.28; Filler 1.29, 1.85, 3.44).

**Conclusions:** Owing to the lower volume of distribution, Cys reaches a new steady state earlier than Crea. The RR in Cys is attenuated in severe AKI as a result of constant extrarenal elimination.

**DISCLOSURE:** *Bökenkamp, A.*: Grant/Research Support, Siemens Healthcare Paid Lectures.

## Abstract# 196

**Chronic Kidney Disease in Egyptian Children: A Report from a Tertiary Centre** A.M. Bakr, A. Sarhan, A. Hammad, A. Al-Refaey, A. Al-Mougy. *Pediatric Nephrology, Mansoura University Children's Hospital, Mansoura Faculty of Medicine, Mansoura, Egypt.*

**Objectives:** Chronic kidney disease (CKD) in children is a major health problem. There are distinct geographic differences in the reported information on CKD due to environmental, racial, genetic, and cultural differences. The aim of this study was to analyze the characteristics of CKD children in our unit over the last 9 years.

**Methods:** Medical records of all CKD children referred to nephrology unit at Mansoura University Children's Hospital from 2001-2008 were revised. Patients with stages 2-5 were included.

**Results:** In the last 9 years, 353 children with CKD stages 2-5 were referred to our unit. At presentation the median age was 5 years and growth retardation, malnutrition and anemia were evident in 58.1%, 47.9% and 87.8% respectively. Patients presented in stage 5 (35.7%), 4 (25.8%), 3 (27.5%), and 2 (11%). Urologic problems were the leading causes (42.8%) with obstructive uropathy, mainly due to posterior urethral valve, was the most frequent subclass (29%). Etiology was unknown in 16.1%. After a median follow up period of 50 months, 27.5% of patients were lost to follow up, 22.7% died and transplantation was undertaken in only three patients.

**Conclusions:** Most CKD patients referred late to our center with evident morbidities. The leading cause of CKD was obstructive uropathy which is preventable. The majority of ESRD patients were treated by hemodialysis and almost one fifth of them died. This profile of CKD indicates the need for great efforts to increase the publics' and health care personnel's awareness of CKD in children and to improve living related transplant program for these children.

## Abstract# 197

**Association of CKD Stage and Glomerular Disease with Medication Use in Children with CKD** T. Blydt-Hansen,[1] C. Pierce,[2] R. Zack,[2] C. White,[2] M. Moxey-Mims,[2] V. Dharnidarka,[2] B. Warady,[2] S. Furth.[2] *[1]U of Manitoba, Winnipeg, MB, Canada; [2]CKiD Investigators, United States.*

**Objectives:** To describe factors associated with the frequency and type of medications used by children with CKD.

**Methods:** Cross-sectional analysis of baseline data from the CKiD cohort, aged 1-16 yrs. We assessed the association between medication usage (classified by problem) and clinical characteristics, including glomerular (GD)/non-glomerular (NGD) diagnosis and GFR (CKD stage): ≥60 (II), 45-60 (IIIa), 30-45 (IIIb), ≤30 (IV).

**Results:** 552 subjects (GD=119) self-reported medication use. Patients with NGD were younger, male, Caucasian and had longer duration of CKD. Median (1st-3rd quartile) number of unique medications was 3 (2-5) and 4 (2-7) for NGD and GD respectively (p=0.02). The most prevalent medications were ACEi/ARB (52%), active vitamin D (37%), iron (30%), alkali (29%) and anti-bacterials (28%). Medication treating CKD-specific complications (active vitamin D, PO4 binders, alkali, growth hormone, ESA, iron, lipid control, diuretics, non-ACEi/ARB BP meds) increased progressively as GFR declined. Use of growth hormone, PO4 binders and ESA increased predominantly in stages IIIb & IV of CKD. For GD vs NGD, adjusted prevalence ratios (95% CI) were significant for lipid medication 6.3, 2.1-19.5, ACEi/ARB 1.6, 1.4-2.0, diuretics 2.9, 1.3-6.5, corticosteroids 7.2, 3.2-16.0, other immunosuppressants 25.0, 5.3-117.8, antacids 2.0, 1.2-3.4, alkali 0.6, 0.4-1.0, bladder medications 0.1, 0.0-0.5, stool softeners 0.2, 0.0-1.0 and anti-bacterials 0.5, 0.3-0.8.

**Conclusions:** The prevalence of medication use increased in a problem-restricted fashion with advancing CKD stage and with secondary to GD.

**DISCLOSURE:** *Dharnidarka, V.*: Consultant, BristolMyersSquibb; Other, Genzyme.

## Abstract# 198

**Anticipating Treatment Needs and Complexity in Children with CKD** T. Blydt-Hansen,[1] C. Pierce,[2] R. Zack,[2] G. Schwartz,[2] N. Benador,[2] P. Hmiel,[2] R. Mak,[2] B. Warady,[2] S. Furth.[2] *[1]U of Manitoba, MB, Canada; [2]CKiD Investigators, United States.*

**Objectives:** To describe factors associated with treatment intensity and inter-relatedness in children with CKD.

**Methods:** Cross-sectional analysis of baseline data from the CKiD cohort, aged 1-16 yrs to assess medication use by CKD stage (GFR): II (≥60), IIIa (45-60), IIIb (30-45), IV (≤30). Adjusted *relative* changes in number of medication subclasses associated with CKD stage and glomerular disease (GD) using multivariate generalized linear models. Pairwise comparisons (2x2 contingency tables) of medication groups. Significant associations defined as having $p<0.05$, >3:1 prevalence ratio and conditional prevalence of medication usage >25%.

**Results:** N=552 had available self-reported medication data classified into 19 CKD-related problem groups. The mean number (±SD) of treated problems increased by CKD stage: 1.9±1.6 (II), 2.3±1.8 (IIIa), 3.3±2.2 (IIIb), 4.5±2.3 (IV). Adjusted relative counts increased ($p<0.05$) from CKD stage II to IIIa (1.2), IIIb (1.8) and IV (2.5), and with GD (1.5). 22/171 medication group pairings showed significant conditional associations. Bi-directional conditionality was noted for three groupings: corticosteroid/immunosuppressant/antacid, ESA/iron supplement & ESA/growth hormone (GH). Unidirectional conditional prevalence for med A (on med B) included ESA (alkali, active vitamin D), GH (alkali), non-ACEi/ARB BP med (K binder, diuretic) and PO4 binder (K-binder, iron supplement, active vitamin D).

**Conclusions:** Treatment intensity increases with CKD stage and GD. Conditional associations of treatments suggest inter-relatedness and may anticipate impending treatment needs, regardless of CKD stage.

## Abstract# 199

**Evaluation of Acute Kidney Injury in Children in the Intensive Care Unit Using pRIFLE Criteria** N.L. Bresolin, A.P. Pereira, J.E. Goes, F.L. Carvalho. *Pediatric, Universidade Federal Santa Catarina, Florianopolis, Santa Catarina, Brazil; Pediatric, Universidade Federal Santa Catarina, Florianopolis, Santa Catarina, Brazil; Pediatric, Universidade Federal Santa Catarina, Florianopolis, Santa Catarina, Brazil.*

**Objectives:** The aim was to apply the pRIFLE criteria to a pediatric population at risk for AKI and to analyze the prevalence and association of AKI as defined by pRIFLE with mortality, and with length of stay in both the hospital and ICU. It also evaluates the applicability of pRIFLE as a prognostic tool in the ICU.

**Methods:** Prospective, descriptive observational study. Single-center, 8-bed PICU facility. Participants: One-hundred twenty-six consecutive patients admitted between September 2008 and December 2009.

**Results:** Data of 126 patients were studied. Median age was 2 years (range 1 month-15 years). Twenty-three (18.3%) patients died. Fifty-eight patients (46%) developed AKI as defined by pRIFLE. Hospital and ICU length of stay was significantly longer for patients with AKI (median 1 vs. median 5, $P<0.001$). Higher pRIFLEmax result was associated with greater ICU and hospital length of stay ($P<0.05$) and also was associated with higher PIM II score. Median PIM II scores for controls, R, I, and F were 1.5, 2.05, 9.05, and 15.06 respectively ($P<0.05$). Patients with AKI had an in-hospital mortality rate twelve times higher than patients without AKI (36 vs. 3% $P<0.001$).

**Conclusions:** The prevalence of AKI was significant, and was directly associated with in-hospital mortality, and length of stay in both the hospital and ICU. The pRIFLE classification was useful in defining AKI in the PICU, and, according to these criteria, AKI was a significant prognosis predictor.

## Abstract# 200

**The Haiti Earthquake Experience** M. Ferris,[1] F. Kaskel,[3] G. Hidalgo,[4] E. Mena, I. Salusky,[5] N. Orta,[9] G. Zilleruelo,[8] M. Bonilla-Felix,[6] L Satlin,[7] T. Bunchman.[2] *[1]UNC, Chapel Hill, NC, United States; [2]HDCH, Grand Rapids, MI, United States; [3]Montefiore, Bronx, NY, United States; [4]U of I, Chicago, IL, United States; [5]UCLA, LA, CA, United States; [6]U of PR, SJ, Puerto Rico; [7]Mt Sinai, NY, NY, United States; [8]U of Miami, Miami, FL, United States; [9]Nefrologia, Valencia, Venezuela; [10]UDIAT, Barcelona, Spain.*

**Objectives:** The response from international pediatric nephrologists to the Haiti earthquake and future activities are described.

**Methods: Communications:** Members of IPNA, ALANEPE and ASPN communicated daily with the ASN Disaster Relief Task Force & with Dr. Mena as on-site coordinator. A major barrier was the inability to have a local communication system.

**Manpower & Patients:** We registered 27 international pediatric nephrologists willing to be deployed or receive patients in their home institutions. Dr. Hidalgo provided medical services in Haiti. Less than 5 children were diagnosed with AKI

**Equipment:** Pediatric-specific dialysis equipment was supplied by the Fresenius Disaster Relief Global Program, arriving to the Dominican Republic 7 days post-earthquake. Other companies (Baxter, MedComp, Cook Critical Care & Nexus) also participated.

**Results: Future Activities**

1. Maintain a registry focusing on technical/language capacity, ability to rapidly deploy & apriori identification of institutions willing to receive patients
2. Coordinate with the dialysis industry and first responders, specifying the unique needs of children
3. Train first responders emphasizing pediatric AKI prevention
4. Create patient registries to identify needs and action plans.

**Conclusions:** Response to the 2010 Haiti earthquake was reactive and the delivery of dialysis supplies/manpower difficult. Proactive, coordinated efforts are underway.

## Abstract# 201

**Acute Renal Failure Due to Bilateral Hypoxantine Urolithiasis in a Boy with Lesch-Nyhan Syndrome** C. Candan,[1] P. Turhan,[1] N. Yildiz,[1] M. Mutus,[2] C. Ulukaya Durakbasa,[2] M. Erguven,[1] H. Okur.[2] *[1]Pediatric Nephrology, Ministry of Health Istanbul Goztepe Education and Investigation Hospital, Istanbul, Turkey; [2]Pediatric Surgery, Ministry of Health Istanbul Goztepe Education and Investigation Hospital, Istanbul, Turkey.*

**Objectives:** Lesch-Nyhan syndrome (LNS) is a very rare X-linked recessive disorder caused by a deficiency of the enzyme *hypoxanthine-guanine phosphoribosyltransferase* (HPRT). A complete deficiency of HPRT leads to severe purine overproduction and to uric acid stone formation. This may be effectively prevented by administration of allopurinol; however, its overdosage may result in xanthinuria, hypoxanthinuria and urolithiasis as a consequence.

**Methods:** We report on a 7-year-old boy with LNS who developed acute renal failure due to bilateral obstructive urolithiasis. The serum creatinine and urea levels were elevated with 2.7 mg/dl and 48 mg/dl, respectively, but the serum uric acid level was particularly low at 1.4 mg/dl. After stones were surgically removed, his renal function returned to normal.

**Results:** In contrast to hypoxanthine, xanthine is very poorly soluble in urine, and therefore practically only xanthine is responsible for stone formation. But, interestingly, infrared spectroscopy examination revealed pure hypoxantine stones in our patient.

**Conclusions:** In conclusion, allopurinol dosage should be carefully adjusted and closely monitorized in order to avoid iatrogenic xanthine or hypoxantine crystal formation.

## Abstract# 202

**Kinetics of Cysteamine Bitartrate in an Anephric Patient on Hemodialysis** J.Y. Chung,[1] T.E. Bunchman,[1] B.A. Barshop,[2] G.-M. Barletta,[1] J.M. Steinke.[1] *[1]Pediatric Nephrology, Dialysis, and Transplantation, Helen DeVos Children's Hospital/Michigan State University, Grand Rapids, MI, United States; [2]Department of Pediatrics, University of San Diego, San Diego, CA, United States.*

**Objectives:** Since the introduction of cysteamine bitartrate (Cystagon™), renal outcome and life expectancy of patients with nephrogenic cystinosis have improved significantly. Despite the common use of this medication for cystinosis treatment, pharmacological data in dialysis patients is limited. The aim of the study was to understand the kinetics of cysteamine bitartrate on an anephric patient on hemodialysis (HD).

**Methods:** Cysteamine bitartrate dose (50 mg/kg/day) was administered orally to a 13 year-old anephric patient with nephropathic cystinosis every 6 hours. Plasma concentrations of cysteamine were measured at pre-HD (3-hr after a dose administration), post-HD (6-hr after and before the next dose), 2-hr post-HD, 4-hr post-HD, and 6-hr post-HD, using a liquid chromatography/mass tandem spectrometer.

**Results:** The pre-HD plasma cysteamine level was 16.4 µM. The level at the end of HD was 5.99 µM (baseline level). The patient was given the next dose of cysteamine at the end of HD. The cysteamine levels at 2, 4, and 6-hr were 24.7, 8.26, and 5.98 µM respectively.

**Conclusions:** Our data demonstrated that cysteamine levels 6 hours post dialysis returned to the baseline level and that cysteamine bitartrate is metabolized and eliminated in an anephric hemodialysis patient similar to a non-dialysis patient. This suggests that dosing frequency should remain every 6 hours in patients without residual renal function. However, further pharmacokinetic studies in this population would be warranted.

## Abstract# 203

**Haemolytic Uraemic Syndrome in Scotland – 1987-2009** M.C. Convery,[1] B.D. Oates,[2] I.J. Ramage.[1] *[1]Renal Unit, RHSC, Yorkhill, Glasgow, United Kingdom; [2]Paediatrics, Crosshouse Hospital, Kilmarnock, United Kingdom.*

**Objectives:** To describe the clinical features, treatment and outcome of Haemolytic Uraemic Syndrome (HUS) in Scotland over the past 23 years.

**Methods:** A retrospective case note review of children presenting with a clinical diagnosis of HUS to a national paediatric nephrology centre between 1987 and 2009.

**Results:** 307 children were identified(145(47%) males) with a median age of 3.2yrs(range 0.2-14.9). A diarrhoeal prodrome was present in 298/307(97%) with E.coli 0157 identified in 180/250(72%). At presentation, associated clinical features were anuria 118/273(43%), hypertension 105/304(35%), hyperkalaemia 71/307(23%), hypovolaemia 40/258(16%)and clinical fluid overload 59/260(23%). Dialysis was required in 237/306(77%). Peritoneal dialysis was undertaken in 197/303(65%) and haemodialysis in 76/301(25%) with 38/301(13%) receiving both. 2 patients did not recover renal function; of those that did the average dialysis duration was 12 days for haemodialysis and 10 days for peritoneal dialysis.Median hospital stay was 14 days(2-137). A median of 2 transfusions(1-12) were undertaken in 295/303 patients. Seizures occurred in 9% of children, with a significant association with hyponatraemia p=0.007(2-sample t-test). 11/306(4%) required a laparotomy and 4/306(1.3%) developed diabetes. There were 5 deaths(1.6%). Follow up EDTA GFR data was available in 131/307, mean 111.2mls/min/1.73m$^2$ with 16/131(12%) having a GFR < 80. Blood pressure was documented as normal in 250/302(83%) and proteinuria resolved in 200/302(66%).

**Conclusions:** We report data from a large cohort of patients treated in a single centre and found death or endstage renal disease in 2.3% with 12% having evidence of an impaired GFR.

## Abstract# 204

**Childhood Diarrhea-Associated Hemolytic Uremic Syndrome in Georgia: Single Center Experience** T. Davitaia,[1,2] O. Rusadze,[1] G. Megrelishvili,[2] L. Aragveli,[1] M. Dzidziguri.[1] *[1]Division of Pediatric Nephrology, M.Iashvili Children Central Hospital, Tbilisi, Georgia; [2]Tbilisi State Medical University, Department of Pediatrics, Tbilisi, Georgia.*

**Objectives:** In Georgia, the majority of cases of childhood HUS are linked to diarrheal illness and are mainly of sporadic occurrence. We present single center experience on 14 cases of D(+) HUS, which were admitted to our hospital between June and September 2009.

**Methods:** Clinical and laboratory data of patients were collected and analyzed.

**Results:** A total of 14 children (<17 years) with D(+) HUS were diagnosed at our hospital. The mean age at presentation was 7.3± 5.1 years. 57% of children were male vs 43% female. Bloody diarrhea was reported in 85.7%. The mean interval between onset of diarrhea and the diagnosis of HUS was 4 ±0.99 days. Twelve children received antibiotics during their antecedent illness. A causative infectious agent STEC O157 serotype was identified in only 2 cases (14%). On admission, all children had evidence of hemolytic anemia (mean serum HB level of 96.1 g/l), various degrees of thrombocytopenia (178 X 109/l, range 87± 292 X 109/l) and renal failure (mean serum creatinine level 442 mol/l). Eleven (78.6%) patients required dialyses. Two patients (14%) died during the acute phase of HUS. 11 of surviving children fully recovered renal function. Cases were hospitalized for a median length of 20.8±12.1 days (range, 4± 46 days).

**Conclusions:** We present the sole known outbreak of D(+) HUS in Georgia. Considering significant morbidity and mortality of HUS, we encourage public awareness of this pathology as well as establishment of effective preventive measures and future close follow-up of these patients in Georgia.

## Abstract# 205

**Significant Acute Renal Failure Due to Non-Steroidal Anti-Inflammatory Drugs: Inpatient Setting** M.P. Dixit, T. Doan, R.C. Kirschner, N.M. Dixit. *Pediatric Nephrology, Florida Children's Hospital, Orlando, FL, United States.*

**Objectives:** In United States non-steroidal anti-inflammatory drugs (NSAID) are freely available over-the-counter. Because of the adverse effects on the kidneys and the popularity of these drugs, unregulated use of NSAIDs is an under recognized and potentially dangerous problem.

**Methods:** Fifteen inpatients, mean age of 15.1 ± 2.72 years (5 males, 10 females), were referred to nephrology for acute renal failure. All patients admitted to taking ibuprofen and six also consumed naproxen. None of the patients had underlying renal diseases at the time of admission. Nine patients had proteinuria and 12 had hematuria (including one with gross hematuria). One patient had nephrotic syndrome but resolved spontaneously without steroids and has remained in remission for 4 years. Two patients required dialysis. Only one of the dialyzed patient required steroid therapy for recovery of renal function.

**Results:** The mean duration of hospitalization was 7.4 ± 5.5 days. The serum creatinine peaked at 4.05 ± 4.53 mg/dL (range 1.3-16.6). All patients recovered renal function with normalization of serum creatinine to 0.71 ± 0.15 mg/dL. However, the duration from onset to normalization of serum creatinine was 37 ± 42 days. This indicates that many patients had abnormal renal function for a prolonged period.

**Conclusions:** In conclusion, NSAIDs pose significant risk of renal failure for significant duration and as an entity may be under recognized.

## Abstract# 206

**The Comparison of Serum Cystatin C and Creatinine Levels in Determination of Glomerular Filtration Rate in Children** O. Donmez,[1] N. Yildiz,[2] O. Durmaz.[3] *[1]Pediatric Nephrology, Uludag University, Bursa, Turkey; [2]Pediatric Nephrology, Uludag University, Bursa, Turkey; [3]Pediatric Nephrology, Uludag University, Bursa, Turkey.*

**Objectives:** The aim of this study was to establish serum cystatin C and creatinine levels in patients with chronic kidney disease. We also aimed to determine glomerular filtration rate (GFR) both associated with serum cystatin C and creatinine levels and compare them.

**Methods:** Data were collected from 166 patients with chronic kidney disease followed-up at the Uludag University Medical Faculty.
**Results:** The mean age was 10.3±0.4 years. The mean serum creatinine level, cystatin C level and Schwartz GFR were found 0.69±0.03 mg/dl, 0.76±0.03 mg/L and 121.5±2.6 ml/min/1.73m$^2$, respectively. The mean serum creatinine and cystatin C levels of the control group were 0.55±0.01 mg/dl and 0.60±0.02 mg/L. Receiver Operating Characteristic analysis was used to evaluate sensitivity of the serum creatinine and cystatin C for determining low GFR. For cystatin C, cut-off level was 0.62 mg/L, sensitivity was 70.8%, and area under curve was 0.63±0.05 (p<0.05). For creatinine cut-off level was 0.58 mg/dl, sensitivity was 66.7% and area under curve was 0.52±0.06 (p>0.05). Cystatin C was found to be more sensitive than creatinine for determining low GFR. The correlation between glomerular filtration rate result and creatinine clearance was evaluated. It was found that cystatin C associated GFR was in accordance with creatinine clearance.
**Conclusions:** It is concluded that serum cystatin C, which is not affected from age, height and body mass index, might be used as a more practically and appreciate marker in diagnosis of chronic kidney disease.

## Abstract# 207

**The Effects on the Quality of Life of the Sociodemographic Characteristics of Patients Undergoing Peritoneal Dialysis** O. Donmez,[1] A. Beyazit,[2] O. Durmaz.[3] *[1]Pediatric Nephrology, Uludag University, Bursa, Turkey; [2]Uludag University, Bursa, Turkey; [3]Uludag University, Bursa, Turkey.*
**Objectives:** The study aimed to define the sociodemographics of pediatric patients undergoing chronic peritoneal dialysis (PD) and to evaluate the effect of the sociodemographic characteristics on their quality of life (QOL).
**Methods:** The QOL in Children Measurement Form was requested from the patient and The QOL in Children Parent's Form from the parents.
**Results:** The study comprised 40 patients and their parents; 24 male and 16 female. The mean age was 12.6±3.6 years. The QOL forms completed by the patients themselves showed a mean score of 55.7±18.6 points whereas the forms completed by the parents showed a mean score of 51.2±18.7 points. A significant relationship was determined between the QOL total and lowest measurements and gender, health insurance, type of PD, parents' status, duration of chronic renal malfunction, age at starting PD, duration of dialysis, incidence of peritonitis attack, Kt/V, hemoglobin and serum albumin levels. A significant relationship was determined between the educational level of the mother and the total child measurement points and the child's physical health total points. A significant relationship was determined between the educational level of the father and the total child measurement points, the child's physical health total points, the child's psychosocial total points, parent measurement total points and parents' physical health total points.
**Conclusions:** Our perceptions of the low QOL of our patients undergoing PD and their parents were determined. An increase in the educational level of the parents was seen to have a parallel increase in the child's QOL.

## Abstract# 208

**Metabolic Syndrome (MS) in Children with Chronic Kidney Disease (CKD). Preliminary Results from 5 Polish Centers** D. Drozdz,[1] J.A. Pietrzyk,[1] M. Zajaczkowska,[2] B. Leszczynska,[3] M. Szczepanska,[4] A. Wasilewska.[5] *[1]JUMC, Krakow, Poland; [2]MUL, Lublin, Poland; [3]MUW, Warsaw, Poland; [4]MUS, Zabrze, Poland; [5]MUB, Bialystok, Poland.*
**Objectives:** MS remains recognizable risk factor of cardiovascular morbidity and progression of renal failure.
The assessment of the prevalence of MS in children with CKD was the aim of the study.
**Methods:** Study was conducted in a group of 120 CKD stage 1-5 children (75 M, 45 F) aged 11.6 years, with body weight – 36.6 kg and eGFR – 27.5 ml/min/1,73m$^2$ on the average, respectively. In each child the fasting serum levels of glucose, insulin, creatinine, HbA1c and lipid profile were evaluated and blood pressure (BP) and waist circumference were measured. MS was diagnosed according to A. Ferranti and B. IDF criteria.
**Results:** If A. criteria were applied, MS was diagnosed in 27% of studied pts and if B. - only in 9.8%. Accordind to A. MS was diagnosed in 28% of children with CKD stage 2, in 22% pts. stage 3, in 21% stage 4 and in 47% stage 5, respectively. Increased TGL levels were observed in 79 children, hypertension in 64 but according to IDF criteria (BP≥130/85) - only in 12%; decreased HDL levels in 55, whereas an increased glucose serum levels only in 5 pts. HbA1c levels exceeding 6 mmol/l were found in 10 out of 98 pts. The waist circumference >75 pc. was found in 20% of all studied children. An increased insulin levels were found in 14 out of 67 pts.
**Conclusions:** Metabolic signs typical for MS occur more frequently in children with CKD stage 2 and up and Ferranti criteria seems to be more appropriate for MS detection, however we should start to workup on therapeutical interventions in children in whom the above disturbances were detected.

## Abstract# 209

**A Case Report: Hyperkalemic Acute Renal Failure Due to Cystine Nephrolithiasis** S. Hosseinzadeh, D. Fahimi, N. Parvin. *Pediatric, Shahrekord University of Medical Sciences, Shahrekord, Chaharmahal Va Bakhtiary, Islamic Republic of Iran; Pediatric Nephrology, Tehran University of Medical Sciences, Tehran, Tehran, Islamic Republic of Iran; Nursing Department, Shahrekord University of Medical Sciences, Shahrekord, ChaharmAhal Va Bakhtiary, Islamic Republic of Iran.*
**Objectives:** Cystinuria is a rare autosomal recessive disease that reabsorption of four dibasic amino acids (cystine, ornithine, lysine and arginine) is impaired. Impairment of proximal tubular reabsorption of filtered cystine causes cystine nephrolithiasis. The median age of onset of stones is usually 12 years old. We presented a 5-month old infant with hyperkalemic acute renal failure due to bilateral ureteral obstruction by cystine stones.
**Results:** *Case presentation*: Five-month old male infant, single child of nonrelative parents, presented with acute renal failure and bilateral ureteral obstruction by cystine stones. The patient underwent peritoneal dialysis due to the worsening hyperkalemia and after performing bilateral nephrostomy and resolution of obstruction the patient discharged with good condition after 1 week.
**Conclusions:** Cystinuria may manifest in early life and bilateral ureteral obstruction due to cystine stones is a rare but notable cause of acute renal failure in infancy.

## Abstract# 210

**Progression of Chronic Renal Disease (CRD) during Puberty?** C. Fernandez, M. Navarro, A. Angel, E. Laura, P. Antonia. *Pediatric Nephrology, Hospital Infantil "La Paz", Madrid, Madrid, Spain.*
**Objectives:** Analyze progression of CRD (Stages 1-4) during puberty.
**Methods:** Retrospective study of 114 patients (87♂ 27♀) with a structural disease in 71% of the boys. Data of GFR, microalbuminuria and use of ACE inhibitors are recorded 2 years prior and after Tanner III stage onset.
**Results:** Mean age of onset of Tanner III stage was 13.6± 1.2 years for boys and 12.4± 1.4 years for girls. Mean GFR (Schwartz) at -2 years, Tanner III and + 2 years was: 75±24 ml/min /1,73 m$^2$;76±26 ml/min /1,73 m$^2$;69±24 ml/min /1,73 m$^2$ (♂) and 79±25 ml/min /1,73 m$^2$;76±26 ml/min /1,73 m$^2$;67±29 ml/min /1,73 m$^2$ (♀).Mean GFR (Cr-EDTA) at -2 years, Tanner III and + 2 years was: 48±23 ml/min /1,73 m$^2$;50±23 ml/min /1,73 m$^2$;39±20 ml/min /1,73 m$^2$ (♂) and 50±13 ml/min /1,73 m$^2$;53±10 ml/min /1,73 m$^2$;46±15 ml/min /1,73 m$^2$ (♀).Mean microalbuminuria at -2 years, Tanner III and + 2 years was: 43±69; 113±185; 205±368(♂) and 75±166;91±176; 94±99
The percentage of patients treated with ACE inhibitors at - 2 years, Tanner III and + 2 years was: 25%. 30%, 47%.
Significant worsening (p<0.05) of GFR over the 2 years after Tanner III onset. Significant worsening ((p<0,001) of microalbuminuria in the same period of puberty. The worst GFR evolution correlated with worse initial GFR(p< 0,001) and greater microalbuminuria(p= 0,004) and was independent of neither sex nor age of onset of puberty. 16% of the boys and 11% of the girls evolved to ESRD.
**Conclusions:** GFR and microalbuminuria were stable over the two years before Tanner stage III onset, worsening significantly over the next two years.The worsening in GFR correlates with microalbuminuria and initial GFR.

## Abstract# 211

**Idiopathic Capillary Leak Syndrome: And Under-Recognized Caused of Acute Kidney Injury and Edema** E.F. Fossali,[1] F.M. Patria,[1] G.P. Milani,[1] R.M. Dellepiane,[1] C. Persico,[1] M.G. Bianchetti.[2] *[1]Pediatric Emergency Department, Fondazione IRCCS Ca' Granda, Ospedale Maggiore Policlinico, Milan, Italy; [2]Division of Pediatrics, Ospedale San Giovanni, Bellinzona, Switzerland.*
**Objectives:** To recognize a rare cause of acute kidney injury in childhood, which can be fatal or cause cerebral edema or compartment syndrome.
**Methods:** We describe a case of a girl 4-year-old with acute kidney injury secondary to idiopathic capillary leak syndrome (ICLS). In few time (less than 5 hours) diffuse edemas and hemoconcetration appeared without proteinuria or signs of anaphylactic reaction.
**Results:** Five reports describe episodic idiopathic capillary leak syndrome in childhood. The present case meets the diagnostic criteria for ICLS: 1) diffuse swelling and rapid weight gain; 2) circulatory shock; 3) hemoconcentration and hypoalbuminemia. Diagnosis can be difficult, and ICLS can be fatal or causes severe irreversible damage. It is probably under-recognized or confused with other entities like anaphylactic shocks or protein-losing enteropathy. Elevated infusion with colloids is useless and often harmful (the girl we report, had a compartment syndrome of the left leg caused by excessive infusion with colloids). Corticosteroids and diuretics are necessary for acute phase, whereas verapamil and aminophylline can contribute to prevent relapses.
**Conclusions:** This case contributes to recognize and manage a rare, but important cause of acute kidney injury in childhood.

## Abstract# 212

**Evaluation of pRIFLE To Predict Need for Acute Renal Replacement Therapy (RRT) in Children** T. Almaslamani,[1] J. Sheth,[2] M. Frieling,[1] J. Hutchison,[2] D. Geary.[1] *[1]Department of Paediatrics, The Hospital for Sick Children, Toronto, ON, Canada; [2]Department of Critical Care, The Hospital for Sick Children, Toronto, ON, Canada.*

**Objectives:** Objective; Acute Kidney Injury (AKI) is classified by pRIFLE criteria as 'risk', 'injury' or 'failure' based on both urine output (UOP), and change in estimated creatinine clearance (eCCl),

However, the ability of pRIFLE to predict the need for continuous RRT (CRRT) is unproven. We reviewed our experience to determine if pRFILE scores reliably predict a need for RRT.

**Methods: Methods**: This 5-year retrospective study examined children admitted to PICU and treated ≥ 1 day with CRRT and/or intermittent hemodialysis (IHD). Pre-existing ESRD, in-born errors of metabolism, and treatment with ECMO were excluded.

**Results: Results**: 95 RRT courses (87 patients) were included; 75 CRRT, and 20 IHD. 43.7% of patients expired; 94.7% on CRRT, and 5.2% on IHD.

pRIFLE classification demonstrated no AKI in 19.4% and 1.3%, and 'failure' in only 37.3% and 51.3% of RRT courses according to UOP and eCCl, respectively. UOP and eCCl measures agreed only 21.1% of the time. Lack of height measure was frequent and absence of historical creatinine values necessitated assignment of a normal eCCl value of 100 ml/min/1.73m$^2$, 42.1% of the time. Only 70.5% and 80% of RRT courses had adequate data to fulfill UOP, and eCCl criterion, respectively.

**Conclusions: Conclusion:** pRIFLE classification is not a good predictor of need for RRT. Agreement between UOP and eCCl criteria is poor, and data for classification by pRIFLE is inaccurate or unavailable for many PICU patients.

## Abstract# 213

**Carnitine Palmitoyl Transferase II Deficiency with Renal Failure** F. Gok,[1] D.O. Hacihamdioglu,[1] S. Kalman,[1] F. Ezgu.[4] *[1]Pediatric Nephrology, GMMA Med School, Ankara, Turkey; [2]Pediatric Nephrology, GMMA Med School, Ankara, Turkey; [3]Pediatric Nephrology, GMMA Med School, Ankara, Turkey; [4]Pediatric Metabolism, Faculty of Medicine, Gazi University, Ankara, Turkey.*

**Objectives:** Herein we report a 12 years old boy who was born from unconsanguineous Turkish parents. He developed severe paleness and fatigue 4 months ago and presented with chronic renal disease.

**Results:** Non member of family was affected with renal disease in the medical history; there were no meaningful finding except paleness and fatigue on physical examination. His weight, length and blood pressure were normal. On the laboratory study; serum creatinine, BUN, total lipids, CPK and transaminase were increased, uric acid and lactic acid were normal. Proteinuria, ketonuria were not detected but hematuria on the dipstick test. Red blood cell was not seen on the microscopic examination of the urine. Renal US were normal. With these findings the patient considered that he has oxidation defect of long chain fatty acid. The responses of increased serum lactate and ammonia levels of front arm ischemic test were normal. The increased result of CPK levels was considered long-chain fatty acid oxidation defect.

**Conclusions:** CPT type 2 deficiency is a disorder of mitochondrial fatty acid oxidation. These patients are at risk of rhabdomyolysis which was triggered by exercises, infections, low nutritional intake, exposure of cold. It could be presented with renal failure cause of degree of enzyme deficiency and frequency of rhabdomyolysis attacks. We concluded that the diagnosis of CPT type 2 deficiency should be consider in ARF or CRF patient who presented with hyperlipidemia, myoglobinuria with no ketonuria.

## Abstract# 214

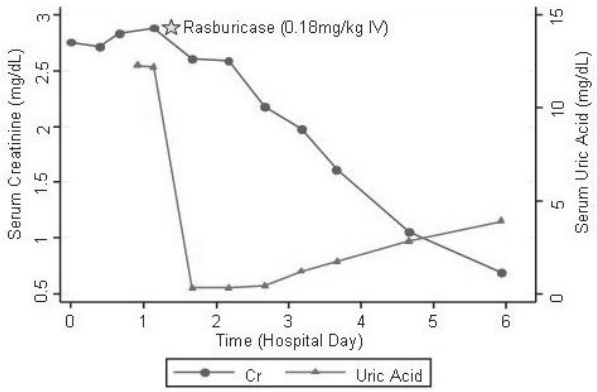
**Rasburicase for Hyperuricemia in Hemolytic Uremic Syndrome (HUS)** A. Acosta, R. Hogg. *Pediatrics, Children's Hospital at Scott & White, Temple, TX, United States.*

**Objectives:** Acute kidney injury (AKI) with elevated serum uric acid (UA) levels (10.2–29mg/dL) has been reported in patients with HUS. AKI is thought to occur from tubular obstruction with UA crystals. Inducing a diuresis may ameliorate the oligo-anuria in such patients.

**Methods:** We describe a child with HUS in whom reducing UA with rasburicase appeared to accelerate recovery of renal function.

**Results:** A 9 month old Caucasian male presented with 6 days of diarrhea, 3 days of vomiting, and 24 hours of oliguria. On admission, hemoglobin was 8.3g/dL, platelet count 36,000/L, blood urea nitrogen 73mg/dL, and serum creatinine (SCr) 2.7mg/dL. Diarrhea associated HUS was diagnosed. The day after admission, SCr was 2.9mg/dL and UA 12.3mg/dL. On hospital day 2, he received a dose of rasburicase 0.18mg/kg IV, and less than 12 hours later, UA was 0.3mg/dL. SCr started declining (Figure), and urine output progressively increased without the use of diuretics. Renal function continued to improve and UA remained normal despite ongoing hemolysis requiring a 2nd RBC transfusion on hospital day 5. The patient was discharged on hospital day 7 in good condition. Two months later, he was well with SCr 0.2mg/dL and UA 4.2mg/dL.

**Figure. Trend of serum creatinine and uric acid**




**Conclusions:** We postulate aggressive management of the high UA with rasburicase accelerated renal recovery. More studies are needed to determine the role of rasburicase for treatment of hyperuricemia in patients with HUS.

**DISCLOSURE:** *Acosta, A.*: Consultant, Pfizer Pharmaceuticals (One time consultant regarding high blood pressure; no involvment with any medications described in this abstract).

## Abstract# 215

**Single Dose of Rasburicase for Treatment of Hyperuricemia in Acute Kidney Injury – A Novel Option** N. Hooman, M. Javadi-Larijani, H. Otoukesh. *Pediatric Nephrology, Ali-Asghar Children Hospital,Iran University of Medical Sciences, Tehran, Islamic Republic of Iran.*

**Objectives:** Hyperuricemia is one of the indications of dialysis in acute renal failure. Rasburicase metabolize uric acid to more soluble form allantoin. We report three hyperuricemic cases with acute kidney injury who treated by single dosage of urate oxidaze effectively.

**Methods:** Between 2008 and 2009, three cases were referred for acute kidney injury and the need for dialysis. Acute Kidney Injury was defined as serum creatinine x2,or urine output <0.5 ml/kg/12 hour, uric acid level higher than 10 mg/dl was considered for dialysis. Rasburicase was administered 0.1mg/kg single dose in two cases and 0.05 mg/kg in one case. Urine output, BUN, Cr, Hco3,and uric acid level was measured at least 12 hour after injection. Consent was taken from parents.

**Results:** Three male children were included. The mean age was 5.3 year (range 23 month -9 year).The cause of renal injury was mutli-organ dysfunction, rapidly progressive glomerulonephritis due to systemic lupus erythematosis, and recurrent acute kidney injury that finally diagnosed as T-cell acute lymphoblastic leukemia. Mean urine output was 0.63 ml/kg/h, uric acid 28 mg/dl, creatinin 2.4 mg/d, and bicarbonate 21meq/L. None of them responded to hydration and bicarbonate therapy, After rasburicase injection the uric acid level declined to 2.7 mg/dl in average and remained stable afterward, urine output increased to 1.85ml/kg/h, and the other parameters has no significant changes.

**Conclusions:** Single dosage of rasburicase can be effective in reducing serum uric acid and increase urination.

## Abstract# 216

**Prognosis for Children with Acute Kidney Injury: A Series of 118 Cases** L.Z. Chen, L. Chen, X.Y. Jiang, Y. Mo, H.Y. Lu. *Pediatrics, Sun Yat-sen University, Guangzhou, China.*

**Objectives:** To explore the risk factors of prognosis for children with acute kidney injury(AKI).

**Methods:** To discuss the causes, clinical characteristics, laboratory features, renal pathological findings, treatment and outcomes of AKI pediatric patients retrospectively, and risk factors of prognosis were to be defined. AKI was defined by the new classification criteria of the Acute Kidney Injury Network. Prognostic factors were determined by univariate methods and stepwise multiple logistic regression analysis.

**Results:** One hundred and eighteen patients(83male;35female) were enrolled in our study, admitted between January 1, 2005 and May 31, 2008 to the First Affiliated Hospital of Sun Yat-sen University in China. Median age at the time of AKI was 7.5 years(range 1 day-14 years). Patients'AKI was classified according to the staging system as follows: 52.5% stage 1, 32.2% stage 2 and 15.3% stage 3. The most common causes of AKI in patients were infectious and autoimmune diseases(39.8%), kidney diseases(27.1%) and circulatory disturbance(11.9%). Hospital mortality rate was 21.2%. Multivariate analysis showed that dependent risk factors for death were need for mechanical ventilation(*OR*=51.75, *P*<0.01),

sepsis/septic shock(*OR*=14.76, *P*<0.01), severe acidosis(*OR*=11.38, *P*<0.01), and white blood cells(WBC) count more than $20\times10^9$/L(*OR*=8.51, *P*<0.01).
**Conclusions:** Infectious and autoimmune diseases, kidney diseases and circulatory disturbance were the most common causes of AKI in children. The important risk factors for death in children with AKI were need for mechanical ventilation, sepsis/septic shock, severe acidosis, and WBC count more than $20\times10^9$/L.

## Abstract# 217

**Cause of Chronic Renal Failure and Clinical Status of Children during Initiation of Hemodialysis in China (Single Center Experience)** L. Jiao, Y. Shen, L. Yuan. *BeiJing Children's Hospital, BeiJing, China.*
**Objectives:** The aim of this study was to evaluate the cause of chronic renal failure (CRF) and clinical status of children during the onset of hemodialysis (HD) in one center of Beijing in China for nine years experience. To investigate the main cause of end stage renal disease (ESRD) and the ratio of kidney transplantation.
**Methods: Methods** Medical documentation of 77 children initiating HD therapy from January 2001 to February 2010 was analysed. The cause of ESRD, concomitance of hypertension, serum concentration of creatinine (Cr), serum level of BUN, potassium, calcium, phosphorus and hemoglobin were assessed. Fifty-two patients were followed up on March 2010.
**Results: Results** Seventy-seven children (41 males, 36 females) aged from 30 months to 15.0 years were analysed. The causes of ESRD were: chronic glomerulonephritis (22.08%), reflux nephropathy (16.88%), congenital renal disorder(16.88%), nephrotic syndrome (12.99%) and unknown origin (19.48%). 44.16% represented hypertension. Hyperphosphatemia, hyperpotassium, hypocalcemia, and anemia were found in 77.27%, 16.88%, 58.44% and 96.10% respectively. The mean level of BUN and Cr was 40.18+20.57mmol/L and 987.67+381.46μmol/L. Within patients who followed up, 50% died. The survival cases who accepted kidney transplantation was 9 in 12, and the longest living time was 4.5 years post transplantation.
**Conclusions: Conclusions** Most of patients with CRF were referred to the nephrologists at the advanced stage. The high mortality and low kidney transplant rate was mostly due to the costly expense of treatment and renal source. Lack of systematic monitoring might be a major reason of missed diagnosis or diagnostic errors.

## Abstract# 218

**Tumor Lysis Syndrome in Korean Children: Change in Last Decade** H.G. Kang, H.K. Lee, I.S. Ha, H.I. Cheong, Y. Choi. *Pediatrics, Seoul National University College of Medicine, Seoul, Korea.*
**Objectives:** As a cause of acute renal failure, tumor lysis syndrome (TLS), a serious complication of highly proliferating malignancy, is one of the important underlying conditions. The strategy against TLS had been hyperhydration, urine alkalinization, and allopurinol to protect the kidney. Recently, rasburicase was added to the armament against this life-threatening condition. In Korea, rasburicase is used as a rescue therapy for cases with allopurinol-resistant hyperuricemia due to the restriction by the National Health Insurance. We reviewed our experiences throughout the last decades to re-assess the risk factors of childhood TLS and the efficacy of rasburicase in our country.
**Methods:** Medical records were retrospectively reviewed for 396 children who were diagnosed as acute leukemia and non-Hodgkin lymphoma (NHL) at our center between 2000 and 2009. The risk factors for TLS were analyzed statistically, and those before and after the availability of rasburicase were compared.
**Results:** Sixty eight patients (17.2%) had TLS with no mortality. Multivariate analysis showed that pre-chemotherapy hypophosphatemia was a significant independent risk factor for TLS, in addition to the known risk factors of hyperuricemia and high LD concentration. The availability of rasburicase as a rescue therapy did not negate the importance of uric acid as a risk factor of TLS.
**Conclusions:** Rasburicase as a second line treatment for intractable hyperuricemia was not effective in reducing the incidence of TLS. Since pre-chemotherapy hypophosphatemia was a significant independent risk factor for TLS in addition to the traditional risk factors, hypophosphatemia may be a warning sign for emergency dialysis.

## Abstract# 219

**Complement Activation on Platelet-Leukocyte Complexes and Microparticles in Enterohemorrhagic *Escherichia coli*-Induced Hemolytic Uremic Syndrome** D. Karpman, L. Sartz, A.-l. Ståhl. *Pediatrics, Clinical Sciences Lund, Lund University, Lund, Sweden.*
**Objectives:** Hemolytic uremic syndrome (HUS) is commonly associated with Shiga toxin (Stx)-producing *Escherichia coli* O157:H7. This study examined patient samples for complement activation on leukocyte-platelet complexes and microparticles as well as donor samples for Stx and lipopolysaccharide (O157LPS)-induced complement activation on platelet-leukocyte complexes and microparticles.
**Methods:** Whole blood and plasma analyzed by flow cytometry.
**Results:** Whole blood from a child with EHEC-associated HUS showed surface-bound C3 on 27% of platelet-monocyte complexes compared to 3% of platelet-monocyte complexes from a healthy volunteer. Plasma samples from 10 patients with EHEC-induced HUS were analyzed for the presence of microparticles derived from platelets, monocytes and neutrophils. Acute phase samples exhibited high levels of platelet microparticles and, to a lesser extent, monocyte microparticles, both bearing C3 and C9. Levels decreased significantly at recovery. Stx or O157LPS incubated with whole blood from healthy donors increased the population of platelet-monocyte and platelet-neutrophil complexes with surface-bound C3 and C9. Simultaneous incubation with Stx and O157LPS enhanced C3 and C9 deposition on complexes. Both Stx and O157LPS induced the release of microparticles, mostly from platelets and monocytes, which had C3 and C9 deposited on their surfaces.
**Conclusions:** Complement deposition on leukocytes and platelets would promote their activation, and the presence of complement on platelet-leukocyte complexes, and microparticles-derived from these cells, suggests a role in the inflammatory and thrombogenic events occurring during HUS.

## Abstract# 220

**Pediatric Chronic Kidney Disease (p-CKD) Prevalence Is Higher Than Reported** D. Landau, R. Schreiber, H. Shalev. *Pediatric Nephrology, Soroka University Medical Center, Beer Sheva, Israel.*
**Objectives:** CKD is defined as evidence for bilateral kidney damage for more than 3 months, which can be structural or functional, with or without a decrease in GFR. The presence of CKD at any stage is a strong risk factor for renal function deterioration during lifetime, many times beyond the pediatric age. The prevalence of p-CKD (stage $\geq$2) has been estimated to be as high as 75 cases/$10^6$ population at risk (Pediatrics 2003;111:e382), but this number is much lower than reports from American young adults (up to 40,000/$10^6$ by NHANES). Thus, the extent of p-CKD may be underestimated. The purpose of this study was to assess the prevalence and etiology of p-CKD in Southern Israel.
**Methods:** Being the only Pediatric Nephrology center in Southern Israel (with an ESRD incidence of 19 cases/$10^6$/yr) and serving a pediatric population of 250,000, we reviewed all cases of p-CKD among inhabitants of this area during the 1994-2008 periods.
**Results:** We identified 258 children (47% Jews, 58% males) with CKD during the study period, 188 of them were living CKD patients aged < 19 yrs at Dec 31 2008, providing a point prevalence of 752 cases/ $10^6$. CKD main etiologies were: hypodysplasia: 36%; obstructive uropathy: 12%; genetic renal diseases: 30%, glomerulonephritis: 14%; other: 9%. CKD stage (defined according to the worst eGFR by the end of 2008) was: 1: 52%, 2: 16%, 3: 8%, 4: 7%, 5: 18%. CKD etiologies were equally represented along its stages.
**Conclusions:** This higher reported p-CKD prevalence: a) may still be an underestimation due to the retrospective nature of the study; b) may contribute significantly to the higher CKD prevalence in the young adult population.

## Abstract# 221

**Neonates with Severe Asphyxia Exist Acute Kidney Injury, Not Acute Renal Failure** X. Liu, Y. Wang, X. Zhang, J. Ding. *Department of Pediatrics, Peking University First Hospital, Beijing, China.*
**Objectives:** Acute Kidney Injury was proposed in the last few years. The purpose of this study was to analyze the features and short-term outcome in neonates of severe asphyxia with or without AKI in order to validate the applicability of the criteria and classification of AKI.
**Methods:** Thirty neonates were enrolled and retrospectively analyzed. The criteria proposed by Acute Kidney Injury Network were applied. All neonates enrolled with severe asphyxia were divided into two groups, AKI group and non-AKI group. Clinical features and short-term outcome were analyzed and compared between the two groups.
**Results:** Among neonates with severe asphyxia, 17/30 (56.7 %) were diagnosed with AKI and divided into AKI group, 13/30 did not reach the criteria of AKI who were divided into non-AKI group. There were 6/30 (20%) neonates whose serum creatinine transitionally increased to more than 1.5mg/dl, fulfilled the diagnostic criteria of acute renal failure. For 13/17 patients in AKI group, the grade of the classification according to urine output was higher than that according to creatinine criteria. However, the number of death (P=0.565) and the average hospital length of stay (14.65±13.92 days vs 13.69±9.57 days, P=0.83) were not statistical different between the AKI group and non-AKI group in this study.
**Conclusions:** More than a half of severe asphyxia neonates reached the criteria of AKI, most of them did not exist acute renal failure. The criteria of diagnosing AKI by measuring urine output seemed more sensitive than that by detecting the increase of serum creatinine. Studies with large sample size and long term follow-up are needed for further investigation on AKI in neonates.

## Abstract# 222

**Factor H Antibody of Japanese Children with Atypical Hemolytic Uremic Syndrome** K. Maekawa, T. Shibano, N. Takagi, J. Sawaki, H. Mae, M. Hattori, T. Tanizawa, F. Kawashima, M. Nishimura. *Department of Pediatrics, Hyogo College of Medicine, Nishinomiya, Japan.*

**Objectives:** Typical hemolytic uremic syndrome (HUS) is a self-limited disorder, but atypical disease, not associated with diarrhea, has significant acute morbidity and high rates of recurrence and progression to end-stage renal disease. Multiple complement pathway mutations are thought to predispose to atypical HUS. The purpose of this study was to determine the incidence and the efficacy of anti complement factor H (CFH) antibody related atypical HUS.

**Methods:** We retrospectively analyzed CFH and anti CFH antibody in 13 children with atypical HUS from 2005 to 2008. Age ranged from 2-11(mean age of 5.3), 5 boys and 8 girls. They were divided two groups; anti CFH antibody positive group and anti CFH antibody negative group. We compared two groups. These character were positive group (2 boys and 1 girl) and negative group (3 boys and 7girls).

**Results:** Three children have high titer of anti CFH antibody (positive group), case 1 has 2094.6ug/ml, case 2 has 1403.4ug/ml, case 3 has 427.6ug/ml. Negative group(other ten children) have low titer of anti CFH antibody mean 96.4ug/ml. Positive group was performed plasma pheresis or hemodialysis or peritoneal dialysis. Now they were administered immunosuprresive therapy and low anti CFH antibody and normal complement, renal function was almost normal. Familial hypocomplementaemia was not found in two groups.

**Conclusions:** We consider that analysis of anti CFH antibody is effective and necessary for atypical HUS. Atypical HUS patients who found to have autoimmune antibody may receive benefits from treatments with corticosteroids with or without immunosuprresive therapy, and plasma exchange.

## Abstract# 223

**A Database for Epidemiology of CRF and Development of Pediatric Nephrology in Nicaragua** G. Marra,[1] M. Sandoval,[2] Y. Silva,[2] G. Selvaggio,[3] F. Sereni,[4] A. Edefonti.[1] *[1]Pediatric Nephrology Unit, Fondazione IRCCS Ca'Granda, Milano, Italy; [2]Pediatric Nephrology Unit, Hospital Infantil Manuel de Jesus Rivera, Managua, Nicaragua; [3]Pediatric Surgery Unit, Hospital Buzzi, Milano, Italy; [4]University of Milan, Milano, Italy.*

**Objectives:** In the context of a project aimed at developing pediatric nephrology in Nicaragua, data on children with CRF were collected by 7 Departmental Hospitals, covering 61% of the pediatric population, with the aim of asessing the epidemiology of CRF and using such data to improve the allocation of resources.

**Methods:** Between 2002-2008, 131 patients (< 19 yrs old) with CRF (gfr <75 ml/min/1,73 m²) were entered in the database.

**Results:** Comparison of Nicaraguan vs Italian (ItalKid) data shows a lower incidence and prevalence of CRF in Nicaragua: 9.3 *vs* 12.1 new cases/yr/million age-related population (marp) and 42.9 *vs* 74.7 marp. These data are repectively due to the under-diagnosis of kidney diseases in particular of VUR (0,8 vs 25,8 % of causes of CRF) and to the higher mortality rate (39,1 vs 1,4%). The high grade of CRF at first diagnosis (IV-V NKF-CKD in 56% of pts) and the high rate of unknown causes of CRF (30.8 vs 3,3%) indicate late referral. A higher percentage of neurogenic bladder (15 vs 3.7%), lupus nephritis (4,2 vs 1,1%) and glomerulopathies (19,2vs 5.8%) suggests different etiology of CRF due to geographic/genetic reasons and the need of improving primary and secondary care over the territory.

**Conclusions:** The database played an important role to gather epidemiological information and allocate project resources over the territory for a bettere diagnosis and prevention of CRF.

## Abstract# 224

**Incidence of Acute Kidney Injury (AKI) in Sick, Hospitalized Children: A Prospective Observational Study** P. Mehta, A. Sinha, R. Lodha, P. Hari, A. Bagga. *Division of Pediatric Nephrology, All India Institute of Medical Sciences, New Delhi, India.*

**Objectives:** To determine, at a tertiary care center, the incidence of AKI [defined by Acute Kidney Injury Network (AKIN) staging] in critically ill & non-critically ill patients, 1 month to 18-yr of age.

**Methods:** Patients, 1 month-18 yr-old, admitted to the PICU or wards from Feb 2008 to Aug 2009 were screened for AKI by serial serum creatinine levels. Cases with stage 5 chronic kidney disease, serum bilirubin >5mg/dl, AKI at admission, or those with <24 hr hospital stay were excluded. AKI was diagnosed and staged using AKIN staging. Patients with AKI were evaluated for its etiology, need for dialysis, duration of hospitalization & outcome.

**Results:** Fifty five (25.9%) of 212 critically ill patients and 20 of 468 (4.3%) who were not critically ill showed AKI (P<0.001). The incidence density of AKI in the former was 22.5 cases/1000 patient days and in the latter 5.4 cases/1000 patient days. AKI stages 1, 2 & 3 were seen in 76%, 14.6% & 9.3% patients respectively. ATN was most common etiology (96%) for AKI. Most patients showed AKI within the 1st wk of hospital stay; dialysis was required in 9 AKI patients (12%). Patients with AKI had a significantly longer duration of hospital stay (P<0.001). On multivariate analysis, risk factors for AKI & mortality were need for mechanical ventilation (respective OR 8.7 & 21.6) & shock (OR 174.3 & 18). The mortality in patients with AKI & non AKI was 45.3% & 11% respectively, P< 0.001.

**Conclusions:** AKI is common in critically ill patients, results in increased hospital stay & is associated with higher mortality.

## Abstract# 225

**Modified RIFLE Criteria in Critically Ill Children with Acute Kidney Injury** A. Mehta, Y.S. Sudan, U. Acharya. *Pediatrics, SMS Medical College, Jaipur, India.*

**Objectives:** Modified RIFLE criteria(pRIFLE)to describe the epidemiology and clinical course of Acute Kidney Injury (AKI) in critically ill children.

**Methods:** In this prospective study 100 critically ill children admitted in Pediatric Intensive Care Unit (PICU) either on mechanical ventilation and/or on vasopressor medications were included.CKD or pre-existing renal disorders were excluded.Prism III score was applied for severity of illness.Serum creatinine and urine output were measured for initial 3 days.Estimated creatinine clearence( eCCL)was calculated using Schwartz formula.

**Results:** 80 patients developed AKI by pRIFLE criteria,28(35%)patients reached pRIFLEmax for risk,29(36.2%) for injury and 23(28.7%) for failure. AKI occurred early in the course of PICU stay (First day 62 cases, second day 12 and third day 6 cases).Common admitting diagnoses were encephalitis(23%) septicemia(20%)and congestive heart failure(19%).Mean PRISM III score was 11.01±8.30with a maximum of 18.35±6.93 in failure group.61.2% patients with AKI required mechanical ventilation.Mean duration of ventilation was 4.02±1.88 days.91.2%AKI patients were on vasopressors.Mean number of medications was 2.55±77. Serum creatinine showed stronger association than urine output alone.5(6.2%) AKI patients required renal replacement therapy and all belonged to failure group.Mean PICU length of stay and hospital stay in AKI group were5.92±2.64 days and 7.72±4.66 days respectively.A significant mortality of 61.25% was observed in AKI patients.AKI was independent risk factor irrespective of admitting diagnosis.

**Conclusions:** pRIFLE criteria is an important method to detect AKI for early intervention.The score is comparable to other ICU scores in predicting the outcome.

## Abstract# 226

**Views of Adults Who Presented in Infancy with CKD 4/5** D. Mekahli, A. Gulett, S.E. Ledermann, L Rees. *Renal Unit, Gt Ormond St Hospital, London, United Kingdom.*

**Objectives:** Renal replacement therapy for infants began in 1980s.Before this, nephrologists believed treatment was unjustified because of ethical dilemmas and uncertain outcome.Data on long-term outcome of these young children is emerging, but there little is known about psychosocial aspects.We have previously reported the outcome of 101 infants who presented with CKD stage 4/5 after1986 (Mekahli et al. CJASN2010;5:10-7).We now describe the psychosocial outcome of those who are now young adults.

**Methods:** Forty patients now aged >16 years were sent a 71-item questionnaire:36 from the Rand-36 Health survey and 35 about environment and social life.Patient scores were compared according to gender,duration of dialysis and/or transplant, number and treatment modality changes and compared with general UK population. Results are given as median(range).

**Results:** All 40 patients(12 females) completed the questionnaire at age 19.2(16.3-23.4) years.Five were on dialysis and 35 were transplanted.Ages at diagnosis of CKD and start of RRT were 0.1(0.0-1.4) and 4.5(0.0-15.6) years and cumulative times on dialysis or transplant were 1.4(0.0-7.7) and 12.8(1.0-21.3) years respectively.Numbers of changes of modality were 2(1-7).Compared with the general UK population our cohort fared worse in physical and social functioning and general health.Patients who had more than one transplantation and/or period of dialysis had a worse health score(p<0.05). 28(70%) were still living at home and 16(40%) of the patients' parents were separated or divorced.

**Conclusions:** Long term outcome of infant with CKD is continuously improving, however,we need to pay more attention to their social and emotional well-being and encourage parents to stimulate social contacts and autonomy.

## Abstract# 227

**Do Urine and Serum NGAL and Selected Interleukins' Concentrations Help in Acute Kidney Injury (AKI) Prediction in ICU Children?** M. Miklaszewska,[1] K. Zachwieja,[1] K. Przemyslaw,[2] J.A. Pietrzyk,[1] D. Drozdz,[1] A. Moczulska.[1] *[1]Paediatric Nephrology, JUMC, Kraków, Poland; [2]AGH, Kraków, Poland.*

**Objectives:** The aim of the study was an assessment of the concentrations of the early markers of AKI in intensive care unit children.

**Methods:** In 47 children during 6 consecutive days the following urine (u) and serum (s) concentrations were determined: uNGAL and sNGAL, uIL-6 and sIL-6, uIL-8 and uIL-18.
**Results:** The study group was divided into two AKI(+) (N=26) and AKI (-) (N=21) subgroups according to RIFLE criteria. In AKI(+) subgroup only the average results of sNGAL and uNGAL were statistically significantly higher that in AKI(-) group: (sNGAL: 3,84 ± 5,5 vs 1,66 ± 2,14 ng/ml; p<0,00001 and uNGAL:13,43 ± 29,6 vs 5 ± 18,52 ng/ml; p<0,00001). This difference [between AKI(+) and AKI(-) subgroup] was also noted for the average sNGAL values in the 1-st (4,31 vs 1,6 ng/ml, p=0,038), 2-nd (4,99 vs 1,72 nl/ml, p = 0,009) and 3-rd (4,29 vs 1,49 ng/ml, p = 0,011) day of screening, respectively. The mean concentrations of other cytokines did not vary, however the higher average values of sIL-6 and uIL-6 were noted in AKI (+) patients. In group of children with fatal outcome (N=11) mean level of sNGAL concentration was statistically higher than in survivors' group (N=36). sNGAL levels survivors vs non-survivors was: 2,52 ± 4,12 vs 4,20 ± 5,5 ng/ml; p<0,00001.
**Conclusions:** In AKI (+) pediatric ICU patients the uNGAL and sNGAL levels were statistically higher than in AKI (-) group, unlike the studied interleukins. The uNGAL and sNGAL may be useful markers of AKI in ICU pediatric patients.

## Abstract# 228

**Non-Invasive Urinary Markers of Renal Dysfunction in Sickle Cell Disease** D. Mohtat,[1] R. Thomas,[1] T. Moulton,[2] C. Driscoll,[3] R. Woroniecki.[1] *[1]Ped Nephrology, Children's Hospital at Montefiore, Bronx, NY, United States; [2]Ped Hematology/Oncology, Bronx Lebanon Hospital, Bronx, NY, United States; [3]Ped Hematology/Oncology, Children's Hospital at Montefiore, Bronx, NY, United States.*
**Objectives:** Renal dysfunction is a major complication of sickle cell disease (SCD). We hypothesized that subjects with SCD will have increased urinary excretion of Transforming Growth Factor β-1 (TGFβ1) and Neutrophil Gelatinase Associated Lipocalin (NGAL) as compared to healthy controls (CTR).
**Methods:** 51 Patients (42 with hemoglobin SS, 8 with hemoglobin SC/SD) and 21 controls (CTR) had spot urine samples collected. ELISA was used to detect urinary TGFβ1/NGAL. Exclusion criteria: subjects who received blood transfusion in the last 90 days, estimated glomerular filtration rate (eGFR) of <60ml/min/1.73m², or presence of any other glomerular disease.
**Results:** Mean age, gender, and race were not statistically different between SCD and CTR. Urinary excretion of TGFβ1was 26.4 pg/mg Cr +/-1.5 in SCD vs 14.9 pg/mg Cr +/-2.4 in CTR, (p<0.00001). TGFβ1was higher in both SS and SC subjects (SS vs CTR, p=0.001, and SC vs CTR, p=0.01), but there was no statistical difference between SS and SC TGFβ1 levels. SCD patients with hemoglobin less than 9gm/dl had higher urinary TGFβ1 than SCD patients with milder anemia, (p=0.002). There was no difference in urinary NGAL between the SCD subjects vs CTR. There were no correlation between urinary TGFβ1 and levels of urinary microalbumin (p=0.15) or estimated glomerular function.
**Conclusions:** Urinary TGFβ1, a biomarker of renal injury, may serve as a better guide than microalbuminuria to monitor progression of renal disease. This proposition needs to be tested prospectively.

## Abstract# 229

**Thrombotic Microangiopathy in Allogenic Bone Marrow Transplantation in Acute Leukemia** Y. Motoyoshi,[1] A. Endo,[1] M. Takagi,[1] M. Nagasawa,[1] T. Morio,[1] S. Mizutani,[1] M. Nagata.[2] *[1]Pediatrics, Tokyo Medical and Dental University, Tokyo, Japan; [2]Pathology, University of Tsukuba, Ibaragi, Japan.*
**Objectives:** A ten year old boy with high risk AML received cord-blood transplantation using CY 120mg/kg + kidney uncovered TBI 12Gy as conditioning regimen on July 2008. Leukemic relapse was developed on March 2009 and he received second transplantation with allogenic unrelated HLA DRB1 mismatched bone marrow. After 2nd BMT, serum creatinine and cystatin C had elevated to 0.40 mg/dl and 2.11 mg/l, respectively, and although there was no hematuria observed, his urinary protein increased to 1g/day.
**Results:** By the symptoms of GVHD appeared in September 2009, the dose of FK506 was increased although his serum Cre level was 2.97 mg/dl. FK506 improved GVHD paralleled with serum Cre level to 2.0 mg/dl. Interestingly, tapering of FK506 was accompanied with relapse of renal dysfunction. Kidney biopsy on November 2009 showed diffuse interstitial fibrosis and edema, tubular atrophy, remarkable glomerular mesangiolysis and features of TMA those of which seemed responsible for renal dysfunction. Immunofluorescence study was negative for IgA, IgG, IgM, C3, C4 and C1q. Presence of TMA was confirmed by electron microscopy.
**Conclusions:** Clinical profiles and kidney pathology suggested that both interstitial fibrosis and TMA by radiation nephropathy as well as GVHD might be the base of kidney dysfunction of the patient. Re-elevation of serum Cre after reduction of FK506 administration is likely explained by remedy of renal circulation including attenuation of interstitial edema and improvement of TMA changes via modulation of systemic GVHD.

## Abstract# 230

**Chronic Kidney Disease in Children: The National Pediatric Hospital Experience in Hanoi, Vietnam** Q.H.T. Nguyen,[1] L.D. Tran,[1] L.T. Nguyen,[2] F. Bouissou,[3] J. Bascands.[4] *[1]Nephrology Department, Hanoi Medical University, National Hospital of Pediatrics, Hanoi, Viet Nam; [2]Surgical Department, National Hospital of Pediatrics, Hanoi, Viet Nam; [3]Nephrology Department, Hopital Des Enfants, Toulouse, France; [4]Inserm U 858, Toulouse III Paul Sabatier University, Toulouse, France.*
**Objectives:** The goal of this descriptive study was to evaluate the aetiology and the socioeconomic status in hospitalized children in Hanoi and propose solutions to improve prevention and basic health care of patients with chronic kidney disease in Hanoi City.
**Methods:** We analyzed the records of all the 152 hospitalized children with chronic kidney disease in the Hanoi hospital from January 2001 to December 2005.
**Results:** The incidence of paediatric chronic kidney disease native to Hanoi City was estimated to be 5.1 per million-child population (pmcp). Median age was 11.29 years; 60.5% were boys and 39.5% were girls; 65% of patients were in end-stage renal disease. Cause of chronic kidney disease included glomerulonephritis (66.4%) and congenital/hereditary anomalies (13%). In 19.8% of children, the aetiology was unavailable. During hospitalization, 5 patients died and 76 patients (50%) refused the treatment although beneficiary of health insurance. Thirty patients (19.74%) received peritoneal dialysis and hemodialysis, 7 patients received renal transplantation with a familial living donor.
**Conclusions:** Late referral and limited facilities for renal replacement therapy explain the poor outcome in this study. We need a program to delineate the burden of chronic kidney disease and improve primary health care for health promotion and prevention of paediatric chronic kidney disease.

## Abstract# 231

**Clinical Profile and Long Term Outcome of HUS** A. Ohri, S. Desai, U. Ali. *Nephrology Division, B J Wadia Hospital for Children, Mumbai, India.*
**Objectives:** To evaluate the clinical and laboratory profile, mortality and long term outcome of HUS.
**Methods:** Retrospective study of 42 patients of HUS over 13 years. Data on demography, CNS involvement, proteinuria, severe hypertension (HT), duration of oliguria, dialysis, recorded. On followup CNS impairment, proteinuria, BP, GFR and deaths noted.
**Results:** 42 patients( 37 boys) had mean age of onset of 5.4 yrs, 6.4 yrs for D- & 3.5 yrs for D+,(p.005). 27 (64%) were D- & 15 (36%) D+. 23(55%) had U Alb/Cr > 3 ; 74% in D- vs 20% in D+ (p 0.001). 25(59%) had CNS involvement, 74% in D- vs 26% in D+ (p 0.003). 21 (50%) had severe HT, 70.4% in D- and 13.3% in D+ (p .0009). D- HUS had longer oliguria, 24.6 vs 7.33 days(p .001), dialysis 20.1 vs 6.57 days (p.001) and hospital stay 36.7 vs 14 days(p.0002) than D+. 6 cases didn't followup.7 died., 6/24 in. D- (25%) & 1/12 in D+.(8.3%). Severe HT was seen in all deaths 7/7 vs 14/29 in suvivors(p=.027). Fatal cases had longer oliguria 32.4 than survivors 17 days(p=0.04).
23/29 cases(79%) had sequelae.16(55%) had HT, 83.3% in D- and 9% in D+ (p = 0.0002). Proteinuria on follow up was present in 17 (59%), 77.8% of D- vs 27.3% of D+.(p=0.017). HT was more in D- 83% vs 9% in D+(p.0002). CNS impairment was seen in 4(14%). 19(64.5%) had GFR<90ml/min, 8(28%) had GFR <60ml/min. No correlation found between oliguria>14 days, dialysis>14 days, severe HT, severe proteinuria, as risk factors for CKD.
**Conclusions:** D- HUS was seen in 65% of cases and in older age. Severe HT, proteinuria & CNS involvement was commoner. They also had longer oliguria & more days on dialysis. Mortality was 19%. 79% of survivors had sequelae, proteinuria in 59%,HT in 55%,GFR<60ml/min in 28% & CNS impairment in 14%.More than 75% of D- HUS had renal sequelae.

## Abstract# 232

**First 28 Days of Acute Kidney Injury Is Critical to Survival** W.A. Olowu,[1] O. Adefehinti,[1] A.L. Bisiriyu.[2] *[1]Pediatric Nephrology & Hypertension, Obafemi Awolowo University (OAU) Teaching Hospitals Complex, Ile-Ife, Nigeria; [2]OAU, Ile-Ife, Nigeria.*
**Objectives:** Clinical characteristics of hospital-acquired acute kidney injury (hAKI) including 60-day outcome were determined.
**Methods:** Clinicolaboratory data of hAKI patients were retrospectively analysed. The AKI Network Workgroup's serum creatinine (Scr) crteria were used. hAKI was staged both at onset and peak (highest recorded Scr) of AKI.
**Results:** Annual hAKI incidence and prevalence rates were 0.17% (or 3.7 per million children population [pmcp]/year) and 0.84% (or 18.3 pmcp), respectively. Male (20): female (8) ratio was 2.5: 1. Median age was 5 (0.063-15.0) years. At hAKI onset there were 14 (50.0%), 5 (18.0%) and 9 (32.0%) patients in AKI stages 1, 2, and 3, respectively. At peak of injury there were 4 (14.3%), 7 (25.0%), and 17 (60.7%), respectively in AKI-1, -2 and -3. None progressed to permanent kidney function loss. AKI-3 was most anuric with high dialysis

requirement (p=0.0329). Nephrotoxics (42.87%): frusemide, cytotoxics, captopril and lisinopril caused hAKI most. All deaths were in AKI-2 and -3. 75% of deaths were in the first 28 hAKI days. Median survival time was 23.5 admission (11-52) days. Maximum Scr means for survivors (486.0 ± 382.0 µmol/L or 5.5 ± 4.3 mg/dL) and non-survivors (353.0 ± 160.0 µmol/L or 4.0 ± 1.8 mg/dL) were similar (p>0.20). The 60-day cumulative mortality was 36.7%.
**Conclusions:** Progression from one AKI stage to the next severe stage occurred commonly. Scr level was not a reliable mortality determinant. Maximal mortality in the first 28 hAKI days and high mortality rate called for high level of clinical vigilance and informed therapeutic interventions to limit AKI progression during this period.

## Abstract# 233

**Acute Kidney Injury in a Pediatric Intensive Care Unit** Z.B. Ozcakar,[1] A. Kavaz,[1] T. Kendirli,[2] S. Altugan,[1] M. Ekim,[1] F. Yalcinkaya.[1] *[1]Pediatric Nephrology, Ankara University Medical School, Ankara, Turkey; [2]Pediatric Intensive Care Unit, Ankara University Medical School, Ankara, Turkey.*
**Objectives:** Acute kidney injury (AKI) is a common complication in the intensive care unit (ICU)s. However, clinical information about the characteristics of AKI in the pediatric population is limited. The aim of this sudy was to determine the incidence and outcome of AKI in a tertiary pediatric ICU in Turkey.
**Methods:** An analysis of all patients admitted to our ICU (with 7 beds) between the dates of July 2009-January 2010 was performed. Patients were classified according to proposed staging system for AKI (modified from RIFLE).
**Results:** Ninety two patients (46 M; 46 F- mean age 56.25 ± 63.33 months) were enrolled. Acute kidney injury developed in 17 (18.5%) patients. Eight of them had stage I and 9 had stage III AKI. Twelve patients with AKI had underlying chronic diseases and 2 had renal diseases. Acute kidney injury developed at the first week of ICU admission in all patients. Major cause of AKI was circulatory failure ± nephrotoxic medications. Peritoneal dialysis was performed in 5 patients. Overall mortality rate in the ICU was 18% during this period. Mortality rate was 41% in patients with AKI and 13% in patients without AKI. Three patients with stage I AKI and 4 patients with stage III AKI were died. 76% of the patients with AKI were mechanically ventilated whereas, 28% of the patients without AKI were mechanically ventilated. Intensive care unit length of stay did not differ between patients with and without AKI.
**Conclusions:** Acute kidney injury developed in a significant number of ICU patients. There was a close association between AKI and ICU outcome in pediatric patients.

## Abstract# 234

**Study of AKIN Criteria in Nonventilated Neonates in NICU** S.K. Patnaik, A.K. Patra, N.K. Biswas. *Pediatrics, Command Hospital (Eastern Command), Kolkata, West Bengal, India.*
**Objectives:** To assess the applicability of AKIN criteria for Acute Kidney Injury (AKI) in neonates admitted to Neonatal Intensive Care Unit (NICU) not requiring mechanical ventilation.
**Methods:** Prospective followup of all consecutive admissions to a level II NICU with serial serum creatinine and urine output monitoring till discharge and at 3 months of age. Neonates were classified for AKI as per AKIN criteria as well as using a serum Creatinine cutoff of >1.5 mg/dl as per previously published studies.
**Results:** 125 neonates were recruited (28-42 wks;median 36 wks) over 18 months. Comorbidities: MAS (9.6%), sepsis (35.2%), asphyxia (23.2%), RDS (8.8%), shock (2.4%), neonatal jaundice (40%) and antibiotics in 79 (63.2%) babies - aminoglycosides 34.4%; vancomycin 12%. 67(53.6%) neonates showed a difference of >=0.3 mg/dl between peak and baseline creatinine values with a median time gap of 120 hours. Peak creatinine values ranged from 0.3to 4.2 mg/dl; baseline values ranged from 0.2 to 2.3 mg/dl. Oliguria of >= 6 hours was observed in 16 (12.8%) cases; 11 in first 24 hours of life; 10 had serum creatinine rise of at least 0.3 mg/dl (range 0.3-1.4 mg/dl). Serum creatinine cutoff of > 1.5 mg/dl to define AKI led to a prevalence of 7/125 (5.6%). Using AKIN criteria, 10/125 (8%) cases met the serum creatinine criteria and 05 (4%) cases met urinary output criteria leading to AKI prevalence of 12% (stage I -10; II- 3; III-2). Mortality was 2.4%; progression to CKD was observed in 04 cases (3.2%) ; all had oliguria and serum creatinine > 0.9 mg/dl at end of 02 weeks.
**Conclusions:** AKIN criteria detected twice the number of cases than a cutoff of >1.5 mg/dl serum creatinine. Majority of neonatal renal failure remains nonoliguric.

## Abstract# 235

**Causes and Outcomes of Acute Renal Failure in 1998-2008. Data of Kaunas Medical University Hospital Clinic of Children Diseases** B. Pundziene, D. Dobiliene, S. Rudaitis. *Pediatric Nephrology Department, Kaunas Medical University Hospital, Kaunas, Lithuania.*
**Objectives:** To determine causes of acute renal failure in children and to compare outcomes between periods: 1998-2003 and 2004-2008.
**Methods:** Retrospective analysis of medical records data of all children treated for acute renal failure (ARF) in Kaunas Medical University Hospital between 1998-2008 years was made. Causes of ARF, sex and age at the onset of the disease, the need for renal replacement therapy (RRT), duration and methods of RRT were evaluated. Comparison between periods (1998-2003 and 2004-2008) was made.
**Results:** 179 children were included in the study. The age range of patients was 1 month to 18 years. 75 were treated in year 1998-2003 (I group) and 104 in 2004-2008 (II group). 124 (69.3%) children were treated in PICU. Basic characteristics (according to child sex and age) between the groups didn't differ. Causes of ARF between the groups did't differ (p>0.05): primary kidney disease 22 (29.3%) in the I group vs 27 (26.0%) in the II group, sepsis 11 (14.7%) vs 28 (26.9%), HUS 7 (9.3%) vs 8 (7.7%) and other causes. Multi organ failure (MOF) was diagnosed in 33 (44.0%) vs 40 (38.5%) patients (p<0.05). 55 (30.7%) childred needed RRT: 29.3% in the I group and 30.8% in the II group. RRT procedures performed: HD 37 (67.3%), PD 13 (23.6%) and CRRT 5 (9.1%). 28 (37.3%) children with ARF died in the period of 1998-2003 and 15 (14.4%) – in 2004-2008 (p<0.05). The most common cause of the death was sepsis with MOF: 78.8% in the I group and 37.5% in the II group (p<0.001).
**Conclusions:** 1/3 of children with ARF needed RRT. Mortality decreased 2.5 times and mortality with MOF decreased twice compared periods 1998-2003 and 2004-2008.

## Abstract# 236

**Mean Serum Uric Acid Levels Predict Left Ventricular Hypertrophy in Children with Chronic Kidney Disease** L.P.R. Resontoc, I.D. Liu, M. Than, K.-H. Ng, Y.-H. Chan, H.K. Yap. *University Children's Medical Institute, National University Health System, Singapore, Singapore.*
**Objectives:** Cardiovascular events are important causes of morbidity in pediatric patients with chronic kidney disease (CKD). This study aimed at determining factors present during CKD stages 2-5 that may affect cardiovascular status at the time of end-stage renal disease (ESRD).
**Methods:** 46 pediatric patients, mean age 9.70±6.48 years at diagnosis of CKD and mean CKD duration of 3.65±3.74 years were retrospectively reviewed. The following parameters were studied: age, sex, time-averaged systolic and diastolic blood pressure index (SBPi and DBPi), hemoglobin (Hb), serum calcium, phosphate, uric acid and intact parathyroid hormone levels. 2-D echocardiographic parameters obtained at time of ESRD included: left ventricular mass index (LVMI), ejection fraction (EF) and fractional shortening (FS). Pearson's correlation and multivariate linear regression analysis were performed.
**Results:** 54.3% of patients had at least one echocardiographic abnormality at onset of ESRD. Severe LVMI (>51g/m$^2$) was present in 21.7%. There was significant correlation between SBPi and LVMI (r=0.33, p=0.027), DBPi and both FS (r=-0.40, p=0.006) and EF (r=-0.42, p=0.004), and uric acid and LVMI (r=0.46, p=0.001). Only time-averaged uric acid (p=0.002) was a significant predictor of LVMI on multivariate linear regression.
**Conclusions:** Time-averaged serum uric acid independent of elevated blood pressures, appeared to be an important predictor of LVMI at onset of ESRD. This may be related to long-term proinflammatory effects on vascular cells. Control of uric acid in CKD patients may have a role in improving long-term cardiovascular outcomes.

## Abstract# 237

**Application of the Criteria of Acute Kidney Injury Networt and the Criteria pRIFLE in Critically Ill Children** M.C. Riyuzo, C.S. Macedo, H.D. Bastos, J.R. Fioretto. *Pediatrics, Faculdade de Medicina de Botucatu, UNESP-Univ Estadual Paulista, Botucatu, Sao Paulo, Brazil.*
**Objectives:** The objective was to apply the classifications criteria proposed by the Acute Kidney Injury Network (AKIN) and the criteria pRIFLE in ctitically ill children and to determine the clinical characteristics, laboratory features and outcomes of AKI.
**Methods:** We retrospectively studied children with sepsis and AKI admitted to Pediatric Care Intensive Unit (PICU) (Faculdade de Medicina de Botucatu,UNESP-Univ Estadual Paulista) in a tertiary hospital.
**Results:** Seventy seven patients (47 male; 30 female) were enrolled. Median age at the time of AKI was 4 months (range 1 to 132 months). Twenty six patients (33,8%) received invasive mechanical ventilation and 76 received vasoactive medications. Mean PICU lenght of stay was 7,33±0,16days and 66 patients (85,7%) had the diagnosis od AKI at the first day of hospitalization in the PICU.Twenty patients (25,9%) presented oligo-anuria and peritoneal dialysis was perfomed in 33 (42,8%). Patients ' AKI classification according to the AKIN criteria was: 11(14,3%) stage 1; 23(29,9%) stage 2 and 43 (55,8%) stage3. The patients' pRIFLE was 4(5,2%) had I (injury) and 73(94,8%) had F (failure). Mortality rate was 33,7%. Hypoalbuminemia, metabolic acidosis, thrombocitopenia, PICU length of stay, invasive mechanical ventilation, dialysis need and stage 2 and stage 3 patients' AKI were independent risk factors of mortality.

**Conclusions:** The children presented at hospitalization with severe AKI. Mortality rate was higher in stage 2 and stage 3 patients of the criteria of the AKIN. The underlying cause of AKI seemed to be an important risk factor for death.

## Abstract# 238

**The Role Serum and Glucocorticoid-Regulated Kinase-1 (SGK1) in Renal Ischemia/Reperfusion (I/R) Injury after Erythropoietin (EPO) and Dexamethasone (Dexa) Treatment** K. Rusai,[1] A. Prokai,[1] B. Szebeni,[1] A. Fekete,[1] A. Vannay,[1] V. Mueller,[1] G. Reusz,[1] U. Heemann,[2] J. Lutz,[2] A.J. Szabo.[1] *[1]Department of Pediatrics, Semmelweis University, Budapest, Hungary; [2]Department of Nephrology, Technical University, Munich, Germany.*

**Objectives:** EPO and Dexa are widely used drugs in different clinical settings. Recently, as novel actions, both have been suggested to protect from I/R injury; however, the exact signalling and effects are not fully described. SGK1 is a novel anti-apoptotic kinase up-regulated by cell stress stimuli, and by hormones and growth factors. In our study, the association between EPO / Dexa treatment and SGK1 was tested in I/R kidney injury.

**Methods:** *In vitro*, HEK293 cells were exposed to hypoxia, and effect of EPO and Dexa on cell death and on SGK1 expression / activation was tested. Moreover, we explored the impact of SGK1 downregulation on EPO effects. In an *in vivo* model of renal I/R, rats were treated with EPO or Dexa prior to ischemia, and tissue injury, SGK1 up-regulation and localization were demonstrated.

**Results:** EPO and Dexa protected cells from apoptosis paralleled by up-regulation and phosphorylation of SGK1. Downregulation of SGK1 by small interfering RNA ameliorated the anti-apoptotic effects of EPO. In rats, EPO and Dexa resulted in less severe tissue injury with up-regulation and activation of SGK1. Moreover, Dexa changed the distribution of SGK1 in tubular cells with a shift from apical to basal membrane localization.

**Conclusions:** In summary, our results provide novel data on the signalling mechanism of EPO and Dexa demonstrating that SGK1 contributes to their renoprotective effects under hypoxia / ischemia.

## Abstract# 239

**The Hemolytic Uremic Syndrome: A Challenging Disorder** H. Safouh, A. Elsisi. *Pediatric Nephrology Unit, Cairo University, Cairo, Egypt.*

**Objectives:** Data about the presentations and outcome of HUS from developing countries is lacking. The aim of this study was to assess the clinical presentations and outcomes of HUS in a single center in Egypt.

**Methods:** Between 4/2005 and 12/2009, 23 patients (14 females) with HUS were admitted to the Pediatric Nephrology Unit, Cairo University, Egypt, constituting 28 % of all cases with ARF. Thirteen were from urban and 10 from rural areas. Age ranged from 2 months to 14 years (mean 46 +/- 48.8 months).

**Results:** There was a history of bloody diarrhea in 11 (48%), watery diarrhea in 6 (26 %) and no diarrhea in 6 (26 %).

Table 1: Symptoms and Signs

| | |
|---|---|
| Abdominal pain | 19 (83 %) |
| Edema | 17 (74%) |
| Vomiting | 17 (74%) |
| Oliguria | 15 (65 %) |
| Fever | 13 (57 %) |
| Anuria | 8 (35 %) |
| Lethargy | 7 (30 %) |
| Petechiae | 6 (26%) |
| Bleeding | 6 (26%) |
| Jaundice | 4 (17%) |
| Convulsions | 4 (17%) |

Admission creatinine and urea were 7.1 +/- 3.41 and 121.8 +/- 47 mg/dl, respectively. Proteinuria and hematuria were universally found. Urine cultures were positive in 6, negative in 10 and not done in 7. Admission Hb was 5.8 +/- 1.6 gm/dl; platelets 188000 +/- 126 /mm3. Fragmented RBCS were seen in all but 2 cases. Treatment included antihypertensives in 14, steroids in 3, blood transfusion in 18, plasma transfusion in 15, plasmapheresis in 3, acute PD in 15 and HD in 8. Four recovered, 5 died and 14 ended up with CKD.

**Conclusions:** HUS is still a major cause of morbidity and mortality in developing countries.

## Abstract# 240

**High-Dose Candesartan and Erythropoietin Combination Therapy in Diabetic Nephropathy with Nephrotic Syndrome** H. Saito, A. Yoshida, J. Suzuki, M. Ishige, T. Urakami, S. Takahashi. *Department of Pediatrics, Nihon University School of Medicine, Tokyo, Japan.*

**Objectives:** Background

Three patients with diabetic nephropathy with nephrotic syndrome due to juvenile-onset type 1 diabetes mellitus were treated with a high-dose angiotensin II receptor blocker (ARB) and erythropoietin (EPO) combination therapy, which resulted in the remission of nephrotic syndrome and long-term suppression of renal impairment.

**Methods:** Patients

Patients are 37~40-year-old female who developed type 1 diabetes mellitus at the age of 12~17. They developed nephrotic syndrome at the age of 29~33 and received candesartan at a dose gradually increased up to 11~16 mg/day with concomitant EPO.

All 3 patients achieved remission of nephrotic syndrome after combination therapy, and the progression of renal impairment was retarded in two patients. Since subsequent reduction of candesartan dose in one patient resulted in a relapse of nephrotic syndrome, she is currently undergoing an escalated dose of candesartan again.

**Results:** Discussion

All 3 patients were at risk for early transition to end-stage renal disease. Dose escalation of ARB up to double the maximum dose for treatment of renal impairment with careful monitoring of changes in serum electrolyte and renal clearance function aimed at elimination of proteinuria resulted in remission of nephrotic syndrome and suppressed progression of renal impairment. It should be noted that this high-dose ARB therapy has the potential to suppress progression of Stage IV diabetic nephropathy.

**Conclusions:** The high-dose ARB/EPO combination therapy has the potential to serve as an effective treatment strategy for chronic kidney disease (CKD).

## Abstract# 241

**Catch-Up Growth of Uremic Rats Is Not Associated with Changes in the Growth Plate Expression of Factors Involved in the Regulation of Endochondral Growth** H. Gil, V. Loredo, N. Mejía, O. Álvarez, F. Santos. *Hospital Universitario Central de Asturias-University of Oviedo, Oviedo, Spain.*

**Objectives:** To gain understanding on the underlying mechanism of both the abnormal catch-up growth described in young uremic rats (Kidney Int 2006;70,1955–61) and the normalizing effects of GH treatment on this process, the growth plate expression of factors potentially involved in the endochondral growth regulation was analyzed during a catch-up growth period.

**Methods:** IGF-1, IGFBP5, GH receptor, BMP2, TGFβ, PTHrP, caspase 3, VEGF, chondromodulin 1 and collagen X expressions were explored by immunohistochemistry, in situ hybridization and western blot in proximal tibiae of uremic rats either untreated or GH treated and control animals, also treated or untreated, fed ad libitum or pair-fed with the former ones. Animals were sacrificed when the uremic rats exhibited catch-up growth after food restriction (Kidney Int 2006;70,1955–61).

**Results:** Consistent quantitative differences were not found in any of the analyzed peptides, although protein expressions of BMP2 and VEGF showed a tendency towards repression by GH treatment. The immunohistochemical distribution pattern of IGF-I, IGFBP5, TGFβ, PTHrP and caspase 3 was patched in the widened hypertrophic zones of uremic rats with GH instead of being uniform as in the other groups.

**Conclusions:** The catch-up phenomenon in uremia is not accompanied by relevant changes in the growth plate expression of the analyzed factors. Normalization of catch-up growth induced by GH treatment is not associated with consistent quantitative differences on the analyzed peptides. The pathophysiological meaning of these findings deserves further investigation.

## Abstract# 242

**Risk Factors for Progression to End Stage Renal Disease in Children with Renal Hypo/Dysplasia without Posterior Urethral Valves** K. Satomura, Y. Santo. *Pediatric Nephrology and Metabolism, Osaka Medical Center for Maternal and Child Health, Izumi, Osaka, Japan.*

**Objectives:** Renal hypo/dysplasia (RHD) is the most common cause of end stage renal disease in children. We evaluated risk factors for progression to end stage renal disease (ESRD: requiring renal replacement therapy or having an estimated glomerular filtration rate (eGFR) less than 15 ml/min/1.73m2) in pediatric patients with RHD.

**Methods:** This study retrospectively analyzed children presenting to our department at less than three years of age with a diagnosis of bilateral RHD without posterior urethral valve. Incidence of ESRD was compared by potential risk factors such as nadir serum creatinine (nCr) greater than 1.0 mg/dl at three years of age, low birth weight less than 2500 g (LBW), severe bladder dysfunction and severe vesicoureteral reflux (VUR), using Kaplan-Meier analysis and logrank tests. Patients who were followed less than two years and those who progressed to ESRD before three years of age were excluded.

**Results:** Twenty-six patients (male/female=17/9) were included in the analysis. The mean age at last follow-up was 11.6 years of age. A total of seven patients progressed to ESRD during childhood at a mean age of 8.9 years. Among the risk factors, only nCr greater than 1.0 mg/dl at three years of age was a risk factor for progression to ESRD (logrank $p < 0.001$).

**Conclusions:** Nadir serum creatinine is the main risk factor affecting renal outcome in patients with RHD. A value of nCr > 1.0 mg/dl at three years of age predicts a high probability of progression to ESRD before the age of 16 years in patients with RHD.

## Abstract# 243

**Acute Kidney Injury Following Cardiopulmonary Bypass for Congenital Heart Disease** S.K. Sethi,[1] D. Yadav,[1] D. Goyal,[1] U. Shukla,[1] R. Khurana,[1] V.K. Gupta.[2] *[1]Department of Pediatrics, PGIMER and associated RML Hospital, New Delhi, India; [2]Department of Cardiothoracic Vascular Surgery, PGIMER and associated RML Hospital, New Delhi, India.*

**Objectives:** There is paucity of data on incidence, epidemiology and risk factors for the development of acute kidney injury (AKI) in children post cardiopulmonary bypass for cardiac surgery. The objective was to investigate the incidence, implicating factors and outcome of AKI (AKIN criteria) after cardiopulmonary bypass.

**Methods:** Design: Retrospective review study. Patients: One hundred and twenty five children (less than 18 years of age) admitted to the cardiothoracic intensive care unit following cardiopulmonary bypass between January 2007 and December 2009. Methods: Age, sex, diagnosis, Jenkins score, duration of cardiopulmonary bypass, ischemia time, Baseline and post surgery: biochemical investigations, urea (mg/dl), creatinine (mg/dl), urine output (ml/kg/hr), frusemide dose, inotrope requirement, and systemic dysfunctions (cardiac, renal, hepatic and neurological) were recorded.

**Results:** Results: Seven (5%) children developed AKI I, 5 (4%) developed AKI II and 2 developed AKI stage III (2%). The patients with AKI stage II and III had a longer ICU stay and increased mortality. Two children required dialysis and none developed chronic renal impairment. All patients with AKI III died during the ICU stay. Using stepwise regression, younger age (< 1 year), low cardiac output, sepsis and duration of cardiopulmonary bypass were the significant risk factors identified for developing AKI.

**Conclusions:** Conclusions: AKI is common and occurred in 11% of children following cardiac surgery, but AKI requiring renal replacement therapy is uncommon.

## Abstract# 244

**Chronic Renal Sequelae after Sporadic Diarrhea Positive Hemolytic Uremic Syndrome: Results of a Case-Control Study** A.P. Sharma,[1] G. Filler,[1] P. Dwight,[1] W.F. Clark.[2] *[1]Dept. of Pediatrics, University of Western Ontario (UWO), London, ON, Canada; [2]Dept. of Medicine, UWO, London, ON, Canada.*

**Objectives:** Many uncontrolled studies and a meta-analysis based on these studies led to the opinion that diarrhea positive (D+) hemolytic uremic syndrome (HUS) is associated with a significant risk for chronic renal sequelae. Two recent controlled studies that followed children with D+ HUS after an *E coli* O157:H7 outbreak, with controls selected from the population exposed in the outbreak, have conflicting results. To clarify this apparent difference in renal outcome, we evaluated renal sequelae after sporadic D+HUS with unexposed case matched controls.

**Methods:** Thirty children after sporadic D+ HUS were compared in long-term follow-up with 30 children unexposed to *E coli* O157:H7 that were age and gender matched. Both groups had measurements of albuminuria, blood pressure (BP) and estimated GFR (Cystatin C). For children, BP measurements were converted to *z* scores and percentiles.

**Results:** In the follow-up (median 6.2 years, range 3.3- 20.1), 40% of the HUS patients showed albuminuria, and one-third of albuminuric subjects had macroalbuminuria. In contrast, microalbuminuria was present in only 3% of the matched controls, and none had macroalbuminuria. The proportion of children with hypertension and prehypertension was three times higher and eGFR was 30 ml/min/1.73 $m^2$ lower in the HUS group versus the controls.

**Conclusions:** Unlike the interpretation in two controlled studies on D+HUS with controls exposed in the outbreaks, children with sporadic D+ HUS compared to unexposed controls had a higher prevalence of chronic renal sequelae.Future studies with unexposed controls can expand on this issue.

## Abstract# 245

**Optic Neuritis after H1N1 Flu Vaccination in a Young Child with Peritoneal Dialysis: A Case Report** J. Shimizu,[1] S. Mori,[1] Y. Higuchi,[1] M. Wakatsuki,[1] K.-i. Ohshima,[2] T. Kubo.[1] *[1]Pediatrics, NHO Okayama Medical Center, Okayama, Okayama, Japan; [2]Ophthalmology, NHO Okayama Medical Center, Okayama, Okayama, Japan.*

**Objectives:** We report a case of 3-year-old patient with optic neuritis after H1N1 Influenza vaccination.

**Methods:** A 3-year-old boy with chronic renal failure secondary to bilateral hypoplastic kidneys became ill tempered. He had been on peritoneal dialysis (PD) since 10-month-old of age. Two days later, his parents noticed his visual impairment and he was hospitalized the next day. It was found he had received a vaccination of H1N1 flu ten days before the admission.

**Results:** On examination, his general condition was good; however, his pupil was dilated and didn't respond to light stimulation in both eye. On fundus examination bilateral papilledema was observed. He had no other abnormal neurological findings including ocular movement. A brain magnetic resonance imaging and an electroencephalogram were normal.

We made a diagnosis of optic neuritis and employed methylprednisolone pulse (MP) therapy three times and supplementation of oral vitamins with continuing PD. However, he was left with severe visual impairment one month later. He had transient hyponatremia during the MP therapy.

**Conclusions:** Reports of optic neuritis after vaccination are infrequently found; however, the relation between optic neuritis and chronic renal failure has not been pointed out specifically. We will report because of an unusual and serious presentation.

## Abstract# 246

**Characterization of an Experimental Model of Progressive Renal Disease in Rats** N.V. Baracho,[1] R.M. Pereira,[2] E.A. Vargas,[1] L.P. Ferreira,[1] R.A.S. Santos,[3] A.C. Simões e Silva.[4] *[1]Faculty of Medicine of Itajubá, Itajubá, Minas Gerais, Brazil; [2]Hospitalar Foundation of the State of Minas Gerais- FHEMIG, Belo Horizonte, Minas Gerais, Brazil; [3]Physiology, Federal University of Minas Gerais, Belo Horizonte, Minas Gerais, Brazil; [4]Pediatrics, Federal University of Minas Gerais, Belo Horizonte, Minas Gerais, Brazil.*

**Objectives:** This study aimed to characterize functional changes produced by progressive nephrectomy in rats.

**Methods:** Male Wistar rats were anesthetized and submitted to progressive degrees of nephrectomy. According to surgical procedure, animals were allocated into experimental groups and placed in individual metabolic cages for two weeks: control(sham operated animals, n=10), unilateral nephrectomy(n=10), 3/4 nephrectomy(n=10) and 5/6 nephrectomy(n=10). Daily measurements of urinary output, water and food intake were performed. At the end of experiment, blood and 24-hour urine samples were collected to determine serum and urinary concentrations of creatinine, urea, Na, K and microalbuminuria.

**Results:** Unilateral nephrectomy did not change water intake and urinary output, but increased serum creatinine and urea and reduced creatinine clearance($p<0.05$). 3/4 nephrectomy also did not change water and food intake. However, this procedure increased urinary output, microalbuminuria and serum levels of urea, creatinine, Na and K($p< 0.05$). 5/6 nephrectomy did not change food intake, but increased water intake and urinary output and produced the highest serum levels of urea, creatinine, Na, K and microalbuminuria and the lowest creatinine clearance($p< 0.05$).

**Conclusions:** Progressive degrees of nephrectomy seem to be a helpful model for evaluating chronic renal disease.

## Abstract# 247

**The Prevalence of Mental Disorders and the Quality of Life in a Pediatric Population with Chronic Kidney Disease** R.C. Marciano,[1] A.C. Simoes e Silva,[2] C.D. Melo,[1] E.M. Lima,[2] J.S.S. Diniz,[2] M.R. Canhestro,[2] C.M.B.M. Soares,[2] H. Correa,[1] E.A. Oliveira.[2] *[1]Mental Health, Federal University of Minas Gerais, Belo Horizonte, Minas Gerais, Brazil; [2]Pediatrics, Federal University of Minas Gerais, Belo Horizonte, Minas Gerais, Brazil.*

**Objectives:** This study aimed to estimate the prevalence of mental disturbances and to analyze the quality of life (QL) in a pediatric population with chronic kidney disease (CKD).

**Methods:** This is a transversal study with 136 pediatric CKD patients regularly assisted in our Pediatric Nephrology Center under various types of treatment: conservative (n=75), dialysis (n=38) and transplant (n=23). The Strengths and Difficulties Questionnaire (SDQ) and PedsQL were applied to both patients and caregivers, in their specific versions. An instrument was also standardized to analyze sociodemographic and economic characteristics of the studied population. Clinical control data were collected from medical charts.

**Results:** There was a high prevalence of mental disturbances (60.2%) and impairment of quality of life in all aspects, with lower scores in the physical and educational aspects. There were no statistically significant alterations between therapies. Better QL was associated with: Catholic religion, non-Caucasian race and age under 10 years. Emotional disturbances predominate among Caucasians over the age of 10, and among those in the initial stages of CKD.

**Conclusions:** The present study stresses the various emotional and social repercussions of CKD and its treatment.

## Abstract# 248

**Chronic Renal Disease in Children (CREDIC): A Population-Based Field Study** O. Soylemezoglu,[1] F. Yalcinkaya,[2] A. Duzova,[3] A. Gur-Guven.[4] *[1]Pediatric Nephrology, Gazi University Faculty of Medicine, Ankara, Turkey; [2]Pediatric Nephrology, Ankara University School of Medicine, Ankara, Turkey; [3]Pediatric Nephrology, Hacettepe University Faculty of Medicine, Ankara, Turkey; [4]Pediatric Nephrology, Antalya University Faculty of Medicine, Antalya, Turkey.*

**Objectives:** To assess the prevalence of chronic kidney disease (CKD) and associated disorders in children in Turkey.

**Methods:** A population-based field study in which individuals are accessed by house visits throughout Turkey has been conducted. As the prevalence of CKD is expected to vary according to geographical region, gender and age, the study sample is selected to represent Turkish population regarding these characteristics. A total of 3622 children (50.4% M, 62.4% living in urban areas) aged 5 to 18 years were enrolled.

**Results:** GFR was <75ml/min/1.73m$^2$ in 10.6%; 1.5% of the study group had Stage III-V CKD; 64% of the patients who had Stage III-V CKD were in 5 to 12 years of age. The prevalence of obesity, hypertension, macroalbuminuria, hyperuricemia and metabolic syndrome were 4.5%, 5.6%, 1.7%, 2.5% and 0.6%, respectively. The prevalence of obesity was higher in males and in young ages. There were statistically significant correlations between body mass index, uric acid level, systolic and diastolic blood pressures. The prevalence of CKD increased with obesity: the prevalence of hypertension was 16% in obese children.

**Conclusions:** This first population-based field study in Turkey showed that CKD is an important burden for childhood health; hypertension and obesity should be focused on in order to decrease CKD prevalence in children and in early adulthood.

The Study is supported by The Scientific and Technological Research Council of Turkey.

## Abstract# 249

**Urinary and Serum NGAL Correlations with Cystatin C, and eGFR in Critially Ill Pediatric Patients** O. Soylemezoglu, M. Polat, K. Fidan. *Pediatric Nephrology, Gazi University Hospital, Ankara, Turkey; Immunology and Allergy dept, Ankara University Children's Hospital, Ankara, Turkey; Pediatric Nephrology, Gazi University Hospital, Ankara, Turkey.*

**Objectives:** However, serum creatinin- the main ARF biomarker used in the clinical setting-demonstrates major limitations. The aim to evaluate the utility of a new kidney damage marker -NGAL- and its correlations with cystatin C, serum creatinine, and eGFR at the time and follow up of ARF in critially ill pediatric patients.

**Methods:** We prospectively eveluated 28 pediatric patients with ARF. When ARF was detected, we took spot urine and blood samples at 24-48 h intervals until the recovery of ARF or anuria occured. Urine and blood samples from 20 healthy children analysed for a control group.NGAL was measured using an ELISA method.

**Results:** At the first day of ARF, uNGAL significantly correlated with sNGAL ( P<0.001), serum cystatin C ( P<0.001) and eGFR ( P=0.012), but did not significantly correlate with serum creatinine ( P=0.114).

The first, second and third urine and serum NGAL, cystatin C,serum creatinin and eGFR values

| Markers | I | II | III |
|---|---|---|---|
| Serum creatinine(mg/dl) | 1.5 | 1.3 | 0.8 |
| sNGAL(ng/ml) | 795 | 684 | 324 |
| Cystatin C(mg/l | 1.2 | 1 | 0.9 |
| eGFR ml/min | 43 | 59 | 93 |

In the follow up, urinary and serum NGAL did not significantly correlate with other markers.

**Conclusions:** We conclude that urinary and serum NGAL are highly spesific and sensitive predictors of ARF in critially ill children. They are found to be useful markers for detecting ARF in the early period, however, they are not in the follow up.

## Abstract# 250

**Effects of Darbepoetin alfa in Different Frequencies on Erythropoiesis and Oxidative Stress in a 5/6 Nephrectomized Rat Model** S. Túri,[1] P. Monostori,[1] G.F. Kocsis,[2,3] Z. Ökrös,[1] P. Bencsik,[2,3] I. Ocsovszki,[2] J. Pálóczi,[2] S. Török,[2] I.S. Varga,[4] I. Kiss,[5] E. Fodor,[3] T. Csont,[2,3] P. Ferdinandy.[2,3] *[1]Department of Pediatrics, University of Szeged, Szeged, Hungary; [2]Department of Biochemistry, University of Szeged, Szeged, Hungary; [3]PharmaHungary Group, Szeged, Hungary; [4]Department of Biochemistry and Molecular Biology, University of Szeged, Szeged, Hungary; [5]Department of Nephrology-Hypertension, St Imre Teaching Hospital, Budapest, Hungary.*

**Objectives:** To study possible effects of different administration frequencies and withdrawal of darbepoetin alfa (DA) on red blood cell (RBC) production and oxidative stress in 5/6 nephrectomized (NX) rats.

**Methods:** Wistar rats were randomly assigned to: sham-operated; NX control; NX receiving 0,4 µg/kg DA weekly (DA1); or NX receiving 0,8 µg/kg DA every two weeks (DA2) groups (n=9-9). DA was withdrawn at Week 10.

**Results:** In NX controls, hemoglobin (Hb) decreased from Week 8 on. In DA1 and DA2, hematocrit (Htc) decreased at Week 4 and increased to the baseline at Week 8. In DA1, Htc increased at Week 10. At Week 12 (without DA), Htc decreased in all study groups. Ratio of oxidized/reduced glutathione (GSSG/GSH) decreased in all groups up to Week 10 and increased in the three NX groups at Week 12. Percentages of reticulocytes and CD59-positive RBCs decreased in all groups up to Week 12.

**Conclusions:** DA in different administration frequencies had similar effects on RBC production and GSSG/GSH in 5/6 NX rats. In addition to distinct trends in Hb and Htc, GSSG/GSH decreased to a slightly greater extent in the DA1 and DA2 groups than in NX controls. Further examinations are needed to explain this finding.

## Abstract# 251

**Irreversible Acute Renal Failure in MELAS with Renal Mitochondrial Abnormalities** H. Tamura, S. Tsuchida, T. Takahashi. *Pediatric Department, Akita University, Akita City, Akita Prefecture, Japan.*

**Objectives:** MELAS is one of a group of clinically syndromes defined mitochondrial dysfunction. Common kidney involvements in MELAS are chronic clinical patterns including Fanconi syndrome, renal tubular acidosis and FSGS, but a few reports with acute renal failure including ATN. Here we report a case who had diagnosed with MELAS developed irreversible acute renal failure. Rapidly evolving from normal renal function to ESRF meant relationship with mitochondrial dysfunction, histo- and ultrastructural analysis was performed.

**Methods:** We performed renal biopsy and analyzed histo- and ultrastructural pathology of the renal tissue, especially mitochondria in glomeruli and renal tubules.

**Results:** Histological findings showed features of ATN, extensive tubular dilation, degeneration of tubular structure, necrosis. Severe interstitial fibrosis and inflammatory cell infiltration were observed. In some glomeruli sclerotic changes were observed. Ultrastructural changes in proximal renal tubules included proliferation of enlarged mitochondria with irregular abnormal cristae were observed. Most of mitochondria in glomeruli and distal renal tubules are intact.

**Conclusions:** Histological analysis suggested that the acute renal failure may have been secondary to acute renal ischemic event, but features of chronic renal injury, FSGS and interstitial fibrosis may have been caused by continuous mitochondrial dysfunction. Most of ATN cases with normal renal function prior to ischemic event recover sufficient renal function, but our case rapidly developed to ESRF. This irreversible acute renal failure is thought to be related to chronic renal injury and regional mitochondrial abnormalities in proximal tubules.

## Abstract# 252

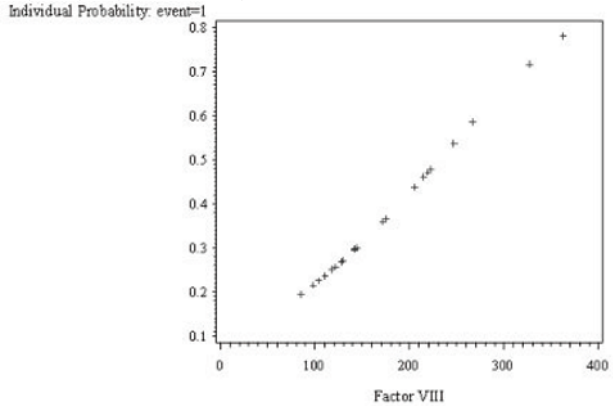
**Elevated Plasma Level of Factor VIII Predicts Venous Thrombosis in Children with End Stage Kidney Disease (ESRD)** H.K. Tawadrous, S. Mongia, R. Gorgy, M. Schoeneman, A. Mongia. *Pediatric Nephrology, SUNY Downstate Medical Center, Brookly, NY, United States.*

**Objectives:** Clotting factor abnormalities are common in children with ESRD. We studied the prevalence of factor VIII and clotting factor abnormalities and thrombotic events.

**Methods:** From Jan07 to Oct09, 29 pts with ESRD on HD or PD were screened for inherited and acquired thrombophilic disorder: protein C,S, factorVLeiden, thrombin III deficiency, prothrombin mutation, homocysteine, factorVIII, IX, XI, DRVVT, PTT-LA, and antiphospholipid antibodies.

**Results:** 22/29 pts on dialysis, 7/29 had stg 4 CKD. Age from3-21yrs. 55.1% male, 62% AA. Renal diseases, FSGS in 48.2%, dysplasia in 6, obstructive uropathy in 2 and others 7. 19 on HD, 3 on PD. 15 pts had CVC, and 4 had AV graft.51% had more than one thrombotic risk factor.Hyperhomocystinemia 48%and elevated factor VIII 44% were the commonest abnormalities. Thromboembolism occurred in 31.8% dialysis pts. We analyzed data with separate logistic regressions of thrombotic event on homocysteine, factor VIII.

There was a trend toward higher levels of homocysteine with less risk,95%CI= 0.7 to 1.02,OR= 0.8, P=0.08). Higher factor VIII associated with greater risk,OR=1,95%,CI= 0.99 to1.02,P = 0.15




**Conclusions:** There is a linear relation between factor VIII level and risk of thrombosis. Prophylactic anticoagulants should be considered in pts with high factor VIII.

## Abstract# 253

**Study of Acute Kidney Injury in Intensive Care Unit** A.S. Vasudev, A.S. Bakshi, S. Jain, R.N. Srivastava. *Dept. of Pediatrics, Apollo Indraprastha Hospital, New Delhi, India.*

**Objectives:** To study the etiology and outcome of acute kidney injury (AKI ) in pediatric intensive care unit.

**Methods:** 36 children with AKI were treated at our tertiary care center over last 5 years.

**Results:** The mean age of patients was 4.6 years (range 2 months to 15 years). There were 21 boys and 15 girls. The causes of AKI were septicemia (36%), hemolytic uremic syndrome (HUS) (14%), cardiac surgery (14%), primary renal disease (14% ): ( glomerulonephritis -2, polyangiitis -1, nephrotic syndrome-2 ), multiorgan failure (MOF) (11%), and tumor lysis syndrome and rhabdomyolysis (1 case each). 31 patients had AKI stage 3 while 5 had stage 1 and 2 ( Acute Kidney Injury Network criteria ). 19 patients (58%) had AKI stage 3 at presentation while others had mild renal dysfunction and developed renal failure subsequently. Dialysis was performed in 31(87% ) patients, peritoneal (PD) in 64% and hemodialysis (HD) in 23%. 4 patients initially started on PD were shifted to HD. The average duration of PD was 6.3 days (range 1 day to 28 days). Peritonitis developed in 2 cases. The overall mortality rate was 30%, ascribed to MOF in 3 patients (following pyopneumothorax, ARDS, pyogenic meningitis in 1 each) and underlying primary condition in rest (cardiomyopathy-2, HUS-1, head injury-1, sepsis-3, tumor lysis syndrome -1). 3 patients eventually developed ESRD and were put on maintainance HD (HUS -2, polyangiitis-1).

**Conclusions:** Children with severe AKI needing dialysis have diverse etiologies. Those with acute tubular necrosis due to a reversible cause fully recover. AKI following sepsis and multiorgan failure is associated with poor outcome.

## Abstract# 254

**Successful Treatment of a DEAP-HUS with Mycophenolate Mofetil: A Two Year Follow-Up** M. Wigger,[1] E. Drückler,[1] C. Skerka,[2] P.F. Zipfel,[2] D. Haffner.[1] *[1]Paediatric Nephrology, Children's Hospital, Rostock, Germany; [2]Hans-Knöll-Institute, Jena, Germany.*

**Objectives:** DEAP-HUS (Deficient for CFHR1 and CFHR3 proteins and Autoantibody Positive form of Hemolytic Uremic Syndrome) represents a novel type of haemolytic uremic syndrome. DEAP - HUS is characterized by aquired Factor H (FH) deficiency due to FH - autoantibodies (FH - ab) and deficiency of CFHR proteins 1 and 3 (CFHR1 and CFHR3) resulting in sustained activation of the alternative complement pathway.

**Methods:** We report on a 7 year old boy showing symptoms of atypical HUS with acute renal failure. The patient revealed low C3 levels (0.54 mg/L), FH - ab (704 U) and CFHR1/CFHR3 deficiency, whereas FH concentration and mobility (western blot analysis) were normal. The initial treatment with hemodialysis, plasmapheresis was followed by recurrent fresh frozen plasma applications during 24 months. Additionally the boy received a CD 20 antibody (Rituximab: 375 mg/m²) at month 4, 9 and 23, respectively.

**Results:** This treatment resulted in normalization of serum C3 levels, reduction in FH - ab levels (440 U) and clinical remission. At month 28 the therapy was switched to mycophenolate mofitil resulting in a constant FH - ab level (between 370 U and 400 U) during the next 24 months. C3 levels persisted in the normal range (0.84 – 1.19 mg/L) and estimated GFR ranged between 101 and 120 ml/min/1.73 m².

**Conclusions:** Patients suffering from aHUS due to FH - ab and deficiency of CFHR1/CFHR3 (DEAP-HUS) appear to benefit from immunosuppressive treatment with mycophenolate mofetil. Whether the previous treatment with Rituximab has contributed to this effect remains to be elucidated.

## Abstract# 255

**Prevalence of Microalbuminuria in Paediatric Patients with Sickle Cell Anaemia at the University of Port Harcourt Teaching Hospital** L.E. Yaguo Ide,[1] N.A. Akani,[2] C.A. Nwauche.[3] *[1]Paediatrics, University of Port Harcourt Teaching Hospital, Port Harcourt, Rivers State, Nigeria; [2]Paediatrics, University of Port Harcourt Teaching Hospital, Port Harcourt, Rivers State, Nigeria; [3]Haematology and Blood Transfusion, University of Port Harcourt Teaching Hospital, Port Harcourt, Rivers State, Nigeria.*

**Objectives:** : The objective of this study was to determine the prevalence of microalbumin in the urine of children with sickle cell anaemia and to identify early those that might be at risk of renal failure.

**Methods:** : This was a prospective study of children with sickle cell anaemia attending the anaemia clinic of the department of Paediatrics for a period of 8 months: June 2006 to February 2007, and was both cross sectional and descriptive.

Method: One hundred and ten children with sickle cell anaemia (in steady state) aged 0.5-16years were consecutively enrolled and evaluated. Employing a semi-quantitative method, Microalbumin in early morning urine was determined in each subject.

**Results:** : Of the one hundred and ten children (110) studied 58( 52.7%) were males while 52(47.3%) were females ( M:F ratio1.1: 1) Prevalence of Microalbuminuria in this study was 42.7%. Though not significant P=0.28, prevalence of Microalbuminuria was more in females (48.1%) than in males (37.9%) and tended to increase with age.

**Conclusions:** : The Prevalence of Microalbuminuria in children with sickle cell anaemia in this environment is high ( 42.7%). Routine screening for Microalbuminuria in patients with Sickle cell anaemia will identify patients with early involvement that will necessitate the institution of intervention measures.

## Abstract# 256

**Outcome of Children Treated with Hemodialysis for End Stage Renal Disease in 9 Year Single Center Experience in Egypt** D.M. Youssef, D.M. Tawfeek. *Pediatrics, Zagazig University, Zagazig, Zagazig, Egypt.*

**Objectives:** Is to evaluate cases of ESRD under regular hemodialysis in our unit,over the past 9 years (from 2001 to 2010) as regard, their age of onset, sex, main etiology, major problems, main causes of death and their final outcome.; where in developed countries transplantation is the logic solution other developing countries are lacking this choice, makes every single center experience is unique.

**Methods:** 75 children with ESRD studied by revising their files over the past 9 years, complete history taking, thorough clinical examination, routine investigations plus specific investigations in some selected patients.

**Results:** We found the mean age of our patients presentation was 7±1.4 yrs, there were (females/males) 1.5/1, the main etiologies of ESRD were showen in table

etiology of ESRD in our cases

| Etiology | total number of 75 | percent |
|---|---|---|
| Glomerular | 30 | 40% |
| unknown | 27 | 36% |
| urological | 12 | 16% |
| others | 6 | 8% |

Main complications in those patients are hypertensive attacks, cardiac problems, AVF complication and infections. Fate of our patients was transplantation in only two cases, death in 20 cases, and the 53 cases are under regular hemodialysis; the main cause of death was cardiovascular diseases (26.7%), followed by infection and sepsis (15%).

**Conclusions:** We conclude that maintaining an appropriate care for children with ESRD in developing countries is quit difficult due to many factors late referral of children, poor acceptance and adherence of parents to ask medical service, limitation of financial resources to supply a high cost medication and limited cases of transplantation. So our children supposed to be on hemodialysis for long durations which lead to appearance of more complications.

## Abstract# 257

**Impact of Recombinant Human Erythropoietin Treatment on Left Ventricular Mass and Cardiac Function in Patients with End Stage Renal Disease on Haemodialysis** M.A.B. Zahrane, S. Emam, N.A. Rahman. *Pediatric, Aboul Rich Hospital -Cairo University, Cairo, Egypt.*

**Objectives:** of this work was to demonstrate the effect of rHu EPO therapy on LVH and LV systolic function in patients with end stage kidney disease.

**Methods:** Thirty two patients were enrolled in this study, 14 females and 18 males. Their age ranged from 5 to 17 years along with 15 age and sex matched healthy subjects as controls. The inclusion criteria were; the presence of renal anemia, adequate serum iron status with serum ferritin level of 100 ng/ml or more and a transferrin saturation of >20%, normotension or controlled hypertension and no history of valid heart disease or other systemic illness. We analyzed the laboratory and echocardiographic data before starting EPO treatment and after treatment in period of follow up ranged between 4 and 9 months with a mean of 5.8+/-1.5 months.
**Results:** Hb level increased from 8.5+/-1.87 to 9.3+/-1.7 gm/dl, Hct level increased from 25.78+/-6.59% to 28.88+/-5.5%, LVMI showd reduction from 108.8+/-41.97 to 97.13+/-43.9 g/m$^2$ , SV decreased from 59.58+/-21.17 to 53.9+/-18.49 ml and finally CO decreased from 5.74+/-2.2 to 5+/-1.5 L/minute. No significant change was detected regarding the HR, EDV, &ESV. LV systolic function was normal at the start of the work and remained so in the follow up examination.
**Conclusions:** We concluded that in patients with ESRD on chronic hemodialysis, LVH regression can be obtained after partial correction of anemia with rHu EPO which can be also associated with reduction of the high CO encountered in these cases. Weather this regression would improve outcome in haemodialysis patients remain to be established.

## Abstract# 258

**Acute Renal Failure (ARF) in the Course of Nephrotic Syndrome (NS) in Children** M. Zaniew, T. Jarmolinski, D. Runowski. *Department of Pediatrics, Nephrology and Toxicology, District Children's Hospital, Szczecin, Poland.*
**Objectives:** The aim of the study was to investigate causes and clinical course of ARF in children with NS.
**Methods:** Nine patients (mean age: 8.3±5.2 yrs) with steroid-dependent (5 pts) and steroid-resistant (2 pts) NS, hospitalized between 2000-2009, experienced 12 episodes of ARF (17% of all ARF cases). Four of patients had biopsy-proven MCD, three had FSGS, one DMP and MPGN.
**Results:** Generalized oedema and preceding infection were common presenting features. In 7 cases antihypertensive agents were given on admission. Dehydration in a patient treated with cyclosporine A (CyA), nephrotoxicity of CyA (1 case) and hypotension (1 case) were found as the possible causes of ARF. In 4 cases ARF was present on admission, whereas in eight cases ARF was developed during hospitalization. The analysis of laboratory studies showed hyponatremia (< 135mmol/l) in 8 cases. The highest concentrations of serum creatinine ranged from 0.54 to 4.90 mg/dl (mean: 2.53±1.68 mg/dl). During 7 episodes of ARF, patients were treated with renal replacement therapy (RRT) and during five conservative management was introduced. The indications for RRT were: clinical signs of overhydration (5 cases), hyperkaliemia and high concentration of urea in 2 cases. The recovery from ARF was achieved in most patients. However, two youngest children progressed to end-stage renal disease (ESRD). In 4 children, RRT-related complications were: infections (in 2), arterial hypertension (in 1) and convulsions (in 1).
**Conclusions:** ARF in the course of NS is independent of underlying glomerulopathy. The prognosis for nephrotic children, who experienced ARF, seems to be good. In some patients, ARF episode may precede ESRD.
**DISCLOSURE:** *Zaniew, M.*: Other, Subinvestigator in Clinical Trial - Salary. *Runowski, D.*: Other, Subinvestigator in Clinical Trial - Salary.

## Abstract# 259

**Matrix Metalloproteinases (MMP-2, 9) and Their Tissue Inhibitors (TIMP-1,2) as Novel Markers of Atherosclerosis in Children with Chronic Kidney Disease (CKD) on Conservative Treatment** K. Musial, D. Zwolinska. *Department of Pediatric Nephrology, Wroclaw Medical University, Wroclaw, Poland.*
**Objectives:** CKD triggers acceleration of atherosclerosis, pictured by cardiovascular complications seen already in childhood. MMP/TIMP system, by the impact on matrix accumulation and endothelial destruction, may play a key role in atherogenesis. However, the data on its role in children with CKD is lacking. The aim of our study was to evaluate serum concentrations of MMP-9, MMP-2, TIMP-1 and TIMP-2, as well as their correlations with known markers of endothelial dysfunction (sE-selectin) and inflammation (IL-4, hsCRP), in CKD children treated conservatively.
**Methods:** 37 CKD children were divided into two groups (gr.I – 20 patients with CKD stage II-III, gr.II – 18 children with CKD stage IV-V). 24 healthy subjects served as controls. Serum concentrations of MMP-9, MMP-2, TIMP-1, TIMP-2, sE-selectin and IL-4 were assessed by ELISA. Kidney function and serum hsCRP were also evaluated.
**Results:** Median values of MMP-9, MMP-2, TIMP-1 and TIMP-2 were significantly higher in CKD children than in controls. Moreover, the concentrations of all parameters were increased in patients with CKD stage IV-V when compared to the group with CKD stage II-III. All parameters correlated with: GFR, sE-selectin and IL-4 in serum.
**Conclusions:** Chronic kidney disease in children is characterized by MMP/TIMP system dysfunction, aggravated by the disease progression. The presence of correlations between examined parameters and markers of endothelial damage and inflammation suggests the possibility of their application as novel markers of vascular changes characteristic for atherogenesis in children with CKD on conservative treatment.

# Urinary Tract Infection

## Abstract# 260

**(O-33)**
**Clinical Practice: A Prospective Study about the Application of NICE Guidelines for Children with Urinary Tract Infection** I. L'Erario, M. Pennesi, A. Ventura. *Pediatric Department, IRCCS Burlo Garofolo, Trieste, Italy.*
**Objectives:** NICE guidelines on urinary tract infections (UTI) in childhood were published in 2007.The children at the first episode of UTI with negative ultrasound (US) do not underwent a voiding cystourethrogram (VCUG), which is recommended only in case of relapse. Since 1997 our group has used an identical diagnostic protocol. Here we refer the results of 11 years of follow-up.
**Methods:** 406 children aged between 1 and 36 months at the first episode of UTI were enrolled and followed up prospectively.All were treated with antibiotic therapy and subjected to renal US.Children with negative renal US and no risk factors did not undergo further investigations nor prophylactic antibiotic therapy, starting a follow-up. Those who showed pathologic renal US and/or risk factors underwent to VCUG; if this was positive, a DMSA scintigraphy was performed.
**Results:** The average age at frist UTI was 9.5 months (range 1-36 months), the follow-up period was 47.3 months (range 12-133 months). 376 children (92.6%) showed normal US, 18 had a second episode of UTI, 48 VCUG were performed. A VUR was found in 14 children (29%), 12 with a pathological US and 2 with recurrent UTI. The DMSA scintigraphy showed kidney damage in 6 children (12.5%, all with VUR grade IV), no damage in 34 despite recurrent UTI.
**Conclusions:** In conclusion, our data confirm the effectiveness of new the NICE guidelines demonstrating a substantial saving of invasive investigations (88% of VUCG in our series), without any risk for renal damage and/or UTI relapses.

## Abstract# 261

**(O-34)**
**Establishment of a Non-Invasive Urinary Proteome Test To Identify Children with High Grade Vesicoureteric Reflux** J. Drube,[1] E. Schiffer,[2] E. Lau,[3] C. Petersen,[3] M. Kirschstein,[4] M.J. Kemper,[5] L. Pape,[1] H. Mischak,[2] J.H.H. Ehrich.[1] *[1]Pediatric Nephrology, Hannover Medical School, Hannover, Germany; [2]Mosaiques Diagnostics GmbH, Hannover, Germany; [3]Center of Pediatric Surgery Hannover, Hannover Medical School, Hannover, Germany; [4]Pediatric Department, Celle General Hospital, Celle, Germany; [5]Department of Pediatric Gastroenterology and Hepatology, University Medical Centre Hamburg-Eppendorf, Hamburg, Germany.*
**Objectives:** Vesicoureteric reflux (VUR) is a risk factor for renal scars, impairment of renal function and arterial hypertension. Voiding cystouretherography (VCU) is considered goldstandard to detect VUR. However, VCU is an invasive procedure exposing children to radiation. We conducted a urinary proteome analysis to establish a non-invasive test to identify children without high grade VUR in a population suspected for VUR.
**Methods:** Using capillary electrophoresis coupled to mass spectrometry we established a urinary proteome pattern on 18 patients with high grade primary VUR and 19 patients with no VUR. This pattern of nine polypeptides was then evaluated in a blinded study on 17 patients with high grade primary VUR and 19 patients with no VUR.
**Results:** Sensitivity to detect high grade VUR in the blinded study was 88% (15/17, 95%CI 64-98%), specificity was 79% (15/19, 95%CI 54-94%). The AUC of ROC analysis was 80% (95%CI 64-97%).
**Conclusions:** High grade VUR is found in 12% of children screened for VUR resulting in a calculated negative predictive value of 98% and a positive predictive value of 36.3% for this proteome pattern. Therefore, this non-invasive test can be used to avoid unnecessary invasive diagnostics in children with negative test results.
**DISCLOSURE:** *Schiffer, E.*: Other, E Schiffer is employee of Mosaiques Diagnostics which developed the CE-MS. *Mischak, H.*: Stockholder, H Mischak is Founder and co-owner of Mosaiques Diagnostics, which developed the CE-MS technology and the Mosaiquesvisu Software.

## Abstract# 262
**(O-35)**

**Can We Predict Renal Scar Using Urine Endotheline-1 Level?** A. Yilmaz,[2] A. Gedikbasi,[1] E. Sevketoglu,[1] S. Karyagar,[3] S. Hatipoglu,[1] A. Kiyak,[2] M. Mulazimoglu,[3] G. Aydogan,[2] T. Ozpacaci.[3] *[1]Bakirkoy Dr. Sadi Konuk Training and Research Hospital, Istanbul, Turkey; [2]Bakirkoy Maternity and Childrens Hospital, Istanbul, Turkey; [3]Okmeydani Training and Research Hospital, Istanbul, Turkey.*

**Objectives:** The aim of our study was to investigate whether urine Endothelin-1 (uET-1) could represent a novel biomarker of renal scarring and to determine the optimal cut-off level for uET-1 for prediction of renal scar.

**Methods:** Fourty four patients with renal scar and 44 patients without renal scar on dimercaptosuccinic acid (DMSA) scan were enrolled in the study. Serum urea and creatinine levels were normal in all children in the study. Urine ET-1 was measured by ELISA.

**Results:** Mean uET-1 level was significantly higher in children with scar than in without scar (2.65 fmol/ml vs 0.50 fmol/ml, p=0.001). According to receiver operating curve (ROC) analysis, the optimal cut-off level was 1.1 fmol/ml for uET1 to predict renal scar. Using a cut-off 1.1 fmol/ml for uET-1, sensitivity and specificity were 93.18% and 93.18% respectively. Area under curve (AUC) was found 0.975 (%95 CI 0.945-1.005) for uET-1.

Mean uET-1/creatinine (uET-1/cr) level was also significantly higher in children with scar than in without scar (3.36 fmol/mg vs 1.03 fmol/mg, p=0.001). According to ROC analysis, the optimal cut-off level was 2 fmol/mg for uET1/cr to predict renal scar. Using a cut-off 2 fmol/mg for uET1/cr, sensitivity and specificity were 86.36% and 90.91% respectively. Area under curve was found 0.946 (%95 CI 0.901-0.990) for uET-1/cr.

**Conclusions:** Urine ET-1 and urine ET-1/creatinine can be used as a sensitive marker for prediction of renal scar in patients with normal renal function.

## Abstract# 263
**(O-36)**

**Do Cranberry Capsules Prevent Recurrent Urinary Tract Infection in Children with Neurogenic Bladder?** Z. Bircan,[1] H. Mutlu,[1] S. Tanriverdi,[1] C. Ozkurkcugil.[2] *[1]Pediatric Nephrology, Kocaeli University School of Medicine, Kocaeli, Turkey; [2]Urology, Kocaeli University School of Medicine, Kocaeli, Turkey.*

**Objectives:** Cranberry capsules(CC) have been used as a prophylactic agent for urinary tract infection (UTI) but the efficacy in neurogenic bladder(NB) is questionable. The aim of this retrospective study is to evaluate the efficacy of CC for prevention of recurrent UTI in children with NB.

**Methods:** To be eligible for this study, patients had to have been followed up for a minimum of 6 months and urodynamic evaluation have been done. Patients with overactive detrusor (OAD) and hypocompliant bladder (HB) were elected for the evaluation of risk of infection rate according to bladder pathology.

CC were prescribed as 1 capsule/day for the prevention of UTI. Any other prophylactic agents have not been used during follow up. Infection rate for each patient was calculated before and after CC usage.

**Results:** The study population included 50 (F/M: 34/16) patients with NB (OAD/HB: 32/18) with the mean age of 7.08 ± 4.29 (1-18) years. The average rate of UTI were 4.36 / year in overall group with a mean follow up time of 25.94 ± 24.12 (3, 81) months. The infection rates between patients with OAD and patients with HB did not show any significant difference before and after CC usage ($p>0.05$) proving that infection rates are similar in both groups, independent from bladder pathology. Decrease in infection rate is significant during CC usage in overall group (p=0.000), OAD group (p=0.000), HB group (p=0.025) and in patients with/ or without reflux (p=0.008; 0.004) without any side effects and drop outs.

**Conclusions:** We concluded that CC is a good option for prevention of recurrent UTI in children with NB.

## Abstract# 264
**(O-37)**

**Should Reflux Nephropathy Be Re-Classified as Reflux Associated Damage (RAD)?** A.J.F. Lunn, A.R. Watson. *Children's Renal and Urology Unit, Nottingham University Hospitals NHS Trust, Nottingham, Nottinghamshire, United Kingdom.*

**Objectives:** "Reflux nephropathy" describes renal disease associated with vesico-ureteric reflux(VUR) but does not inform our understanding of the pathogenesis or prognosis. Our objective was to determine if a new classification of Reflux Associated Damage(RAD) would identify groups with different outcomes.

**Methods:** A database of patients with a diagnosis of VUR was prospectively maintained since 1985. A retrospective review recorded the baseline status and 4 outcomes measures; dialysis/transplant, hypertension, proteinuria. and radiographic progression(scars, lack of hypertrophy). The patients were classified into 4 groups.

CLASSIFICATION

| Classification of RAD | Definition |
|---|---|
| Type 1 | Congenital with no obstruction |
| Type 2 | Congenital with obstructive uropathy |
| Type 3 | Acquired with documented UTI |
| Type 4 | Acquired with voiding dysfunction |

**Results:** Notes were reviewed of 748 of 1080 patients on the database. Overall 3.5% were on dialysis or transplanted, 5.9% hypertensive, 6.2% proteinuric and 10% progressed. There was sufficient information to classify 725 patients according to our definitions; Type 1 - 209(28%) patients, Type 2 - 37(5%) patients, Type 3 - 464(62%) patients and Type 4 - 15(2%) patients. The different outcomes are shown in table 2.

OUTCOME MEASURES (%)

| RAD Classification | Dialysis or Transplant | Hypertension | Proteinuria | Progression |
|---|---|---|---|---|
| Type 1 | 3.3 | 9.3 | 8.5 | 11 |
| Type 2 | 22 | 11 | 25 | 10 |
| Type 3 | 0.9 | 5.4 | 6.7 | 12 |
| Type 4 | 5.6 | 6.7 | 38 | 31 |

**Conclusions:** Our classification identifies groups with different aetiology and outcomes of Reflux Associated Damage. This better informs our understanding of "Reflux Nephropathy" and the likelihood of progression following a diagnosis of VUR.

## Abstract# 265
**(O-38)**

**The Efficacy of Acute DMSA Scan in Predicting VUR in Chinese Young Children with Febrile UTI** X. Zhang, H. Xu. *Department of Nephrology and Rheumatology, Children's Hospital of Fudan University, Shanghai, China.*

**Objectives:** The aim of this study was to evaluate the efficacy of DMSA in predicting vesicoureteral reflux (VUR) in young children with febrile urinary tract infection (UTI) and to investigate an imaging procedure suitable to China's domestic condition.

**Methods:** The medical and radiologic records of children (age ≤ 2 years), presenting with febrile UTI between January 2000 and December 2009, were retrospectively reviewed. In all cases, acute DMSA renal scan and micturating cystourethrography (MCU) had been performed.

**Results:** A total of 370 children (233 boys and 137 girls) were included, of which 263 (71.1%) had abnormal DMSA result and 126 (34.1%) were identified as VUR on MCU. Among children with VUR, the number of high-grade (III–V grade) was 103 (81.7%). The sensitivity of DMSA for detecting high grade VUR was 99.0%. The negative predictive value was 99.1% and negative likelihood ratio was 0.03, respectively. In high-grade VUR group, the rate of abnormal DMSA result was significantly higher than those in non-VUR group and low-grade VUR group ($P<0.001$).

**Conclusions:** High-grade VUR remains an important risk factor for renal damage for young children with febrile UTI in China. The possibility to detect high-grade VUR on MCU is rather low when the result of DSMA is negative. DMSA should be the first strategy to investigate the first febrile UTI children at acute phase, which would predict the majority of children with high-grade VUR while presenting renal lesions.

## Abstract# 266
**(O-39)**

**Predictive Ability of Procalcitonin (PCT) as a Predictor for Acute Pyelonephrtitis (APN) and Late Renal Scars (RS) in Children with UTI: Meta-Analysis on Individual Patient Data** S. Leroy,[1] A. Fernandez,[2] R. Nifkar,[3] C. Romanello,[4] F. Bouissou,[5] A. Gervaix,[6] M.K. Gurgoze,[7] S. Bressan,[8] V. Smolkin,[9] D. Tuerlinkx,[10] C.J. Stefanidis,[11] S. Gardikis,[12] P. Leblond,[13] F. Gungor,[14] G. Breart,[15] D. Gendrel,[16] M. Chalumeau.[16] *[1]Univ. Oxford, Oxford, United Kingdom; [2]Joan de Deu Hosp., Barcelona, Spain; [3]Abuzar Hosp., Ahvaz, Islamic Republic of Iran; [4]Univ. Udine, Udine, Italy; [5]CHU Purpan, Toulouse, France; [6]Univ. Hosp. Geneva, Geneva, Switzerland; [7]Univ. Faculty Medicine, Elazig, Turkey; [8]Univ. Padova, Padova, Italy; [9]Med. Center, Afula, Israel; [10]Cliniques Univ. Mont-Godinne, Yvoir, Belgium; [11]Kyriakou Hosp., Athens, Greece; [12]Alexandroupolous Hosp., Thrace, Greece; [13]J. Flandre Hosp., Lille, France; [14]Akdeniz Univ., Antalya, Turkey; [15]INSERM U953, Paris, France; [16]St Vincent Hosp., Paris, France.*

**Objectives:** Prompt but high-quality diagnosis of UTI is important as well as identification of RS is also pertinent to keep children from future complications. We aimed to investigate the predictive ability of PCT for both APN and RS, in children with UTI.

**Methods:** A meta-analysis of individual patient data.

**Results:** 1011 patients (332 boys, VUR ≥3 in 11%, APN in 613 patients, LRS in 131/350 children) were included from 17 studies. PCT was associated to both APN and RS ($p<0.001$). A procalcitonin ≥0.5 ng/mL yielded to an aOR=6.4 [4.6-

8.8], p <0.001 with a sensitivity of 74% [71-78], and a specificity of 71% [66-75]. In the 350 patients for whom late DMSA was available, a procalcitonin ≥0.5 ng/mL was also associated to RS (aOR=2.1; [1.4-3.9], p=0.004) with a sensitivity of 79% [71-86], and a specificity of 41% [34-48].
**Conclusions:** PCT may predict reasonably well APN then RS.

## Abstract# 267
**(O-40)**
**The Antimicrobial Peptide Cathelicidin Interferes with Polymerization of Curli fimbrie and Thus Inhibits the Formation of Uropathogenic *E. coli* Biofilm** M. Chromek, Y. Kai-Larsen, P. Lüthje, X. Wang, Å.A. Holm, K.-O. Hedlund, J. Johansson, M.R. Chapman, S.H. Jacobson, U. Römling, B. Agerberth, A. Brauner. *Karolinska Institutet, Stockholm, Sweden; The Biomedical Center, Uppsala, Sweden; University of Michigan, Ann Arbor, MI, United States.*
**Objectives:** The antimicrobial peptide cathelicidin (LL-37) lyses bacteria and thereby protects the urinary tract against infection. We sought to investigate if low concentrations of LL-37 could affect formation of uropathogenic *E. coli* biofilm.
**Methods:** Urine from 118 patients with urinary tract infection was tested for biofilm. The impact of different concentrations of LL-37 on *E. coli* biofilm was investigated. Precipitation assay, surface plasmon resonance, Western blot analysis, CD spectroscopy, and Thioflavin Y assays were utilized to investigate the binding of LL-37 to curli, and the polymerization of curli fibers.
**Results:** LL-37 binds to curli fimbrie and to their subunit CsgA. The structure and levels of CsgA monomers remain stable in the presence of LL-37. Cathelicidin inhibits polymerization of curli fimbrie and formation of *E. coli* biofilm. This effect is specific for LL-37 and was not observed if a scrambled peptide or a control peptide (VIP) was tested instead of cathelicidin.
**Conclusions:** Here we describe a new mode of action of the antimicrobial peptide LL-37. Even at concentrations lower than bactericidal concentration, LL-37 inhibits the formation of *E. coli* biofilm. This might be an important mechanism in the protection against recurrent urinary tract infections as well as a target of new treatment strategies.

## Abstract# 268
**Renal Doppler Ultrasound in the Diagnosis of Acute Pyelonephritis** F. Cavagnaro,[1] L. Schonhaut,[1] B. Morales,[2] A. Espinoza.[2] *[1]Pediatrics, Clínica Alemana de Santiago-Universidad del Desarrollo, Santiago, Chile; [2]Radiology, Clínica Alemana de Santiago, Santiago, Chile.*
**Objectives:** The differentiation between upper and lower urinary tract infection (UTI) has important clinical implications in children. The aim of this study was to evaluate the ability of renal doppler ultrasound (RDU) in the diagnosis of acute pyelonephritis (APN) compared with Tc-99m dimercaptosuccinic acid (DMSA) renal scintigraphy, in children without renal or urinary malformations.
**Methods:** This propective study comprised 54 elegible children, aged 10 days to 12 years, admitted during 2009 to our pediatric unit with the first febrile UTI. In all of them a DMSA renal scintigraphy and a RDU were done during the first 72 hours into the hospital. Both studies were evaluated independently each other by pediatric radiologists, and the findings were compared. Also all children were evaluated with blood tests looking at elevations of acute-phase reactants and white blood cells. The renal scintigraphy was considered the gold standard for diagnosing APN.
**Results:** There were 42 children (77,8%) with abnormal renal scan, who also had significantly altered the blood tests when compared with those with normal scintigraphy. RDU showed matching findings with scintigraphy in 78%. The sensitivity and specificity of RDU for the detection of APN were 78,6% and 75%, and the positive and negative predictive value were 91,7% and 50%, respectively.
**Conclusions:** In clinically suspected APN, an abnormal RDU should avoid the use of DMSA renal scintigraphy. However, due to the low negative predictive value of RDU, we suggest to complete the study with a renal scintigraphy when the RDU is normal, mainly if they have altered blood tests.

## Abstract# 269
**Comparison of Extended Virulence Genotypes for Bacteria Isolated from Pediatric Patients with Urosepsis, Acute Pyelonephritis, and Acute Lobar Nephronia** C.-H. Cheng, Y.-K. Tsau, T.-Y. Lin. *Pediatrics, Chang Gung Memorial Hospital, Taoyuan, Taiwan; Pediatrics, National Taiwan University Hospital, Taipei, Taiwan; Pediatrics, Chang Gung Memorial Hospital, Taoyuan, Taiwan.*
**Objectives:** Despite recent advances in molecular epidemiology and pathogenecity analyses of extraintestinal *E. coli* infections, detailed analyses identifying virulence factors of *E. coli* isolates from pediatric urosepsis patients have not been reported. This study was conducted to explore and differentiate bacterial virulence factors associated with urosepsis and two other severe parenchymal infections, acute pyelonephritis (APN) and acute lobar nephronia (ALN).
**Methods:** Patients included were those fulfilled the diagnostic criteria of urosepsis, APN, and ALN, without underlying disease or structural anomalies (excluding VUR). Patients with cystitis were included as controls. *E. coli* isolates from urine (cystitis, APN and ALN) or blood (urosepsis) specimens were analyzed using polymerase chain reaction for 25 virulence genes.
**Results:** A total of 147 children (24 cystitis, 45 APN, 48 ALN, and 30 urosepsis) were enrolled. Distinct syndrome-specific differences in the distribution for certain virulence genes, but conservation across syndromes for others, were found. In addition, urosepsis isolates presented higher aggregate virulence factor scores ($p < 0.0001$) compared with cystitis, APN and ALN isolates. Cystitis isolates, rather, showed significantly lower aggregate virulence factor scores than all three invasive urinary bacterial infections.
**Conclusions:** Our findings suggested that urosepsis isolates carry more virulence factors and are therefore likely more urovirulent compared with cystitis, APN and ALN isolates.

## Abstract# 270
**Copenhagen Experience with the NICE-Guidelines for Children with Upper Urinary Tract Infection** D. Cortes,[1,3] R. Lytzen,[1] J. Thorup.[2,3] *[1]Pediatrics, Hvidovre Hospital, Copenhagen, Denmark; [2]Pediatric Surgery, Rigshospitalet, Copenhagen, Denmark; [3]Faculty of Health Science, University of Copenhagen, Copenhagen, Denmark.*
**Objectives:** The number of investigations at pyelonephritis in children has been questioned.
**Methods:** 98 consecutive patients median aged 2.1 years (0.05-14.9) treated for microbiologically verified first episode of pyelonephritis in accordance to AAP, and initially assessed with renal ultrasound examination (US) and MAG3 scinti-/renography ( R). Follow-up for median 4.8 years (3.1-8.2). No patient had prenatally diagnosed urological anomalies.
The results were analyzed in regard to the new British NICE-guidelines for pyelonephritis, which recommends investigations of patients at high risk of urological anomalies only.
**Results:** Initial US was abnormal in 32 patients and initial R was abnormal in 28 patients.
At follow-up prophylactic antibiotics (AB) was prescribed to 20 patients, 20 patients were diagnosed with urological anomalies, 10 of these with vesicoureteral reflux (VUR). Surgery was performed in 9 patients, 6 of these for VUR.
Based on NICE-guidelines, 5 patients operated on for VUR would not initially have been identified, 9 would not have received AB and 5 would still be ignorant to another urological diagnosis. This is unsatisfying considering the possibility of endoscopic therapy and the increasing problems with bacteria resistant to AB.

Negative consequences of NICE-guidelines

| Age group | 0-6 months | 0.5-3 years | 3-15 years |
|---|---|---|---|
| Number | 37 | 37 | 24 |
| No prophylactic AB | 1 | 6 | 2 |
| No VUR diagnosis | 0 | 4 | 1 |

**Conclusions:** We do not recommend the NICE-guidelines, especially not for children 0.6 to 3 years of age, but recommend initially US and R at first episode of pyelonephritis.

## Abstract# 271
**Urinary Tract Infection in Children with Chronic Liver Disease Waiting for Liver Transplantation** S.M. Dehghani, M. Basiratnia. *Shiraz University of Medical Sciences, Shiraz, Islamic Republic of Iran.*
**Objectives:** Urinary tract infection (UTI) is one of the most common infections in patients with chronic liver disease. Data on UTI frequency in children with chronic liver disease is scarce. Published studies only evaluated adult patients. This study is evaluated the UTI frequency in this group of children.
**Methods:** We prospectively evaluated the pediatric patients aged less than 18 year listed for liver transplantation (LT) between 2007 and 2009. Besides history taking and physical examination, blood chemistries, liver function tests, urinalysis, urine culture, and urinary tract ultrasonography was done for all patients.
**Results:** Totally 78 patients with mean age of 6.3±5.1 year (51.3% female) were studied. Urine cultures were positive in 8 (10.3%) cases (7 of them were female), E.coli in 3 (37.5%), Klebsiella in 3 (37.5%), Staphylococcus spp. in 2 (25%). All of them were asymptomatic. All organisms were sensitive to ciprofloxacin. The etiologies of liver disease in these patients were progressive familial intrahepatic cholestasis (n=2), Crigler-Najjar syndrome (n=2), neonatal hepatitis, biliary atresia, tyrosinemia, and autoimmune cirrhosis each in one.
The mean age of children with UTI was significantly lower than those children without UTI (4.6±3.6 Vs. 6.5±5.3).
UTI was significantly more common in patients with younger age, female gender, lower serum albumin (<2.5 g/dL), and higher platelet counts. (p<0.05)
**Conclusions:** Asymptomatic UTI is common in children with chronic liver disease waiting for liver transplantation and it is recommended to screen these children for UTI before liver transplant especially those with younger age, female sex, and low serum albumin.

## Abstract# 272

**Frequency of Urinary Tract Infection and Nocturnal Enuresis in Children with Chronic Functional Constipation** S.M. Dehghani,[1] M. Basiratnia.[2] *[1]Gastroenterohepatology Research Center, Shiraz University of Medical Sciences, Shiraz, Islamic Republic of Iran; [2]Pediatric Nephrology, Shiraz University of Medical Sciences, Shiraz, Islamic Republic of Iran.*

**Objectives:** There are conflicting results about the role of chronic constipation in the urinary tract disorders like urinary tract infection (UTI) and enuresis. Constipation may cause UTI and enuresis due to the uninhibited bladder contraction. The aim of this study was to investigate the frequency of UTI and nocturnal enuresis in children with chronic constipation.

**Methods:** 120 children (73 females) with chronic functional constipation according to the Rom III criteria were enrolled in the study. Detailed past and present history of UTI or symptoms pointing to this diagnosis and enuresis were obtained. Urinalysis, urine culture and abdominal ultrasonography were performed for all patients.

**Results:** The mean age of the patients was 7.4±3.2 years (range, 1-15 year). The most common urinary symptoms were dysuria (15%), urinary frequency (12.5%), and dribbling (4.1%). The frequency of nocturnal enuresis and daytime enuresis were 22.5% and 3.3%, respectively. Pyurias were seen in 11 (9.2%) patients and urine cultures were positive in 7 (5.8%) cases, that all of them were female. E.coli was the most common organism (71.4%) followed by S.aureus and Enterobacter each in 14.3%. All organisms were sensitive to ciprofloxacin.

**Conclusions:** Nocturnal enuresis was found in a significant number of children who had chronic constipation but UTI is not more prevalent than the general population. Therefore, we suggest that nocturnal enuresis should be questioned in children with chronic functional constipation, but screening for UTI is not mandatory in these patients.

## Abstract# 273

**The Prevelance and Clinical Associations of Idiopathic Hypercalciuria in Children with Urinary Tract Infections** H.T. Nacaroglu,[1] G. Demircin,[2] M. Bülbül,[3] Ö. Erdogan,[3] S.G. Akyüz,[3] A. Çaltik.[3] *[1]Pediatrics, Dr Sami Ulus Children's Hospital, Ankara, Turkey; [2]Pediatric Nephrology, Afyon Kocatepe University, Medical School, Afyon, Turkey; [3]Pediatric Nephrology, Dr Sami Ulus Children's Hospital, Ankara, Turkey.*

**Objectives:** To evaluate the association of idiopathic hypercalciuria (IH)with urinary tract infections (UTI) in children.

**Methods:** 224 children diagnosed to have UTI were investigated for the prevelance of IH. IH was defined as urinary calcium/creatinine ($U_{Ca}/U_{Cr}$) ratio over 0.6 for 0-1 year of age, 0.21 after 1 year and/or calcium excretion more than 4 mg/kg/day. The association of IH with demographic and clinical findings were investigated.

**Results:** Hypercalciuria was found in 36 patients (16%). The mean $U_{Ca}/U_{Cr}$ ratio was found to be 0,39±0,26 for hypercalciuric patients. There was no sex difference in the prevelance of IH. Frequency of IH was statistically higher in patients with a family history of urolithiasis and consanguinity and in patients having complaints of abdominal pain, loss of appetite and restlessness. A statistically significant but weak correlation was found between urinary calcium excretion and body weight, height and age. No association could be found between IH and the recurrence of UTI, presence of vesicoureteral reflux, scar formation and prognosis.

**Conclusions:** Since urinary tract infections and their complications are significant causes of morbidity and mortality, hypercalciuria should be kept in mind while investigating risk factors for UTI, especially in patients with hematuria, recurrent UTI and family history of IH though its effect on renal scarring and recurrence of UTI could not be shown in this study.

## Abstract# 274

**Analysis of Twenty Pediatric Cases of Recurrent UTIs** H. Eguchi, H. Fujimaki, S. Matsumoto, R. Hiramoto. *Department of Pediatrics, Matsudo City Hospital Children's Medical Centre, Matsudo, Chiba, Japan.*

**Objectives:** Recently, prophylaxis for low grade VUR has been controversial. We retrospectively evaluated the clinical features regarding pediatric recurrent UTI cases.

**Methods:** One hundred forty seven children were admitted to our hospital because of UTI from October 2005 to February 2009. All patients were followed for more than one year. Of 147, 20 cases (15 males, 5 females) presented UTIs at least twice and were analyzed in this study. The onset of the first UTI episode ranged from 20 days to 7 years (median 3.5 months). The data were collected from the medical records. The evaluated factors were as follows: grade of VUR, lower urinary tract abnormalities and usage of prophylaxis at the onset of another UTI.

**Results:** VCG revealed VUR in 12 out of 19 cases. Of 12, 7 showed high grade VUR(IV≤) and 8 lower urinary tract abnormalities (2 true phimosis, 1 ureterovesical junction stenosis, 1 utriculus prostaticus, 1 posterior urethral valve, 1 unstable bladder, 1 diverticulum of bladder, 1 urethral stenosis + phimosis). Of all 20, 12 caes were on prophylaxis at the onset of recurrent UTIs. Of all, 3 patients had neither VUR nor lower urinary tract abnormalities. Two of them were under one-year-old boys while the rest was seven-year-old girl whose toilet training was already completed. One male infant was on prophylaxis, on the other hand another infant and seven-year-old girl were not.

**Conclusions:** Recurrent UTIs can occur not only in children with some urinary tract abnormalities but also in those without any urological abnormalities. Therefore, once a child experiences a UTI, a close attention should be paid to them until a certain age not to overlook another UTI episode.

## Abstract# 275

**The Incidence of Renal Malformation and Scarring in Children with Urinary Tract Infection Referred to SQU Hospital, Muscat, Oman** I.B. Elnour,[1] D.A. Al-Nabhani,[2] M.A. Elnaggari,[3] A.E. Hassan,[4] M.A. Al Sadoon,[5] S.S. Hussein,[6] D.K. Sankhla.[7] *[1]Child Health, SQU Hospital, Sultan Qaboos University, Muscat, Muscat, Oman; [2]Medical Imaging, SQU Hospital, Sultan Qaboos University, Muscat, Muscat, Oman.*

**Objectives:** The aim of this study was to identify the incidence of congenital renal malformation and renal scarring in children referred to Sultan Qaboos University Hospital with UTI, and to plan the most appropriate protocol of investigations for our population.

**Methods:** Children referred to SQUH with UTI, 1991-2008 were investigated prospectively according to the practiced UK consensus-based guidelines, which were published by RCP 1991. All children less than 5 years with UTI had renal ultrasound, MCUG and DMSA. Older children would have only US and DMSA except if they had a positive history of recurrent UTI or abnormal finding on renal US and DMSA.

**Results:** 262 females and 52 males were enrolled in the study. A total of 93 patients had scarred kidneys (29.6%). 34 patients with renal scarring had no previous history of UTI (36.5%).The number of the left scarred renal units were significantly higher than the right ($P < 0.05$). Our data showed more renal scarring in the elder age group. The incidence of renal malformations was 41%.

**Conclusions:** high rate of renal scarring in the elder age group may be related to delay in recognition and referral of children with UTI. Renal scarring may occurs even after a single UTI. General practitioners in developing countries should be aware of the serious complications of UTI and the need for earlier recognition and management.

## Abstract# 276

**Protocol Study after Pyelonephritis: Changes in Ten Years** M. Espino,[1] T. Fernandez Soria,[1] M. Soria,[1] R. Mata,[1] R. Martin,[1] F. Echavarri,[1] M. Bueno,[1] M. Mijtavila.[2] *[1]Pediatrics and Neonatology, Hospital Universitario Fundacion Alcorcon, Madrid, Spain; [2]Nuclear Medicine, Hospital Universitario Fundacion Alcorcon, Madrid, Spain.*

**Objectives:** To determine the best protocol of study of the urinary tract (UT) after first episode of pyelonephritis (PN) to diagnose renal scar and uropathy, improve management and reduce radiation dosage without risk of renal damage.

**Methods:** Transversal analysis of 365 patients at follow-up after PN through 10 years. First protocol, patients under 5 years undergo sonography (US); 7 days intravenous treatment (IV), prophylactic treatment (PT), voiding cystourethrography (VCU) and renal scan (RS) after 9 months. 2º reduced IV to 3 days. Third protocol the age was reduced to 2 years old or bladder sphincter control. Fourth protocol, with the same age than previous, include only a RS and US, if US anormal they received PT and VCU was performed, if US normal no PT neither VCU.

**Results:** First protocol included 25; second 24, third 177 and fourth 139 patients. We didn't find difference in renal scars between 3 Vs 7 days IV; patients with VUR or not; recurrent pyelonephritis or not. We found VUR in 16%; 19%; 26% and 14%; renal scars in 25%; 18%; 18% and 28%. Only 6% of patients without VCU present new PN and when VCU was performed low degree vesicoureteral reflux (VUR) were diagnosed, so management would be the same.

**Conclusions:** There is not important to diagnose low degree VUR because it is not clear if it is useful PT or surgical treatment so VCU may be avoided. If the objective of the UT it is to prevent and diagnose renal scar, we think it is possible to reduce the number of VCU because it is not the main factor in renal scar genesis.

## Abstract# 277

**Idiopathic Hypercalciuria in Filipino Children with History of Urinary Tract Infection: A Preliminary Study** P.J. Galutira, B.B. Canonigo, R.D. Chan, R.F. Cabansag. *Pediatric Nephrology, University of Santo Tomas Hospital, Manila, Philippines.*

**Objectives:** The general objective is to screen for idiopathic hypercalciuria (IH) in Filipino children with history of UTI.The specific objectives are 1)To present the clinical profile of children with history of UTI; 2)To determine urine calcium

excretion in these children; 3)To establish the incidence of IH among these children; and 4)To identify the clinical manifestations of children with IH.
**Methods:** This prospective study included patients aged <19 y/o diagnosed with UTI. They were divided into 2 groups: single and recurrent UTI. Data were collected regarding gender, age, clinical manifestations and family history of urolithiasis. Serum calcium, urinalysis, urine calcium & creatinine (UCa/Cr) or 24-hr urine calcium excretion were determined once UTI has resolved. Data was expressed as mean ± standard deviation. Descriptive analyses was performed for the calculation of frequencies.
**Results:** There were 18 children diagnosed with UTI of whom 11 were females (61.1%) and 7 were males (38.9%). There was an equal distribution in the number of patients who had single vs. recurrent episodes of UTI. The mean age was 3.57 years (±2.70 years). Majority are in the 1-5 years old age group. Only 4 out of the 18 patients (22.2%) were symptomatic. Manifestations reported were nocturnal enuresis, terminal dysuria, abdominal pain, daytime incontinence, microscopic hematuria and urolithiasis. 6 out of the 18 patients (33.3%) had an elevated UCa/Cr and only 3 patients (16.7%) were confirmed to have an elevated 24-hr urine calcium excretion.
**Conclusions:** IH is not a rare finding among Filipino children with history of UTI.

## Abstract# 278

**Vesicoureteral Reflux (VUR): The Role of Cranberry Prophylaxis** V. Goj, L. Bernardo, G. Masnata. *Paediatrics, Fatebenefratelli and Oftalmico Hospital, Milan, Italy; Paediatrics, Brozu Hospital, Cagliari, Italy.*
**Objectives:** The prevention of urinary tract infection(UTI)in children with VUR is still matter of discussion.In vitro studies have confirmed the main mechanism of action by cranberry(inhibition of bacterial adhesion).The efficacy of this prophylaxis in pediatric patients with severe uropathy is still matter of study.Objective:1)to evaluate the protective effect of a commercially available standard concentrated cranberry juice(ivumir)on UTI in young children with VUR attended by two department of pediatric nephrology(Milano and Cagliari) 2)to verify possible side effects.
**Methods:** The group included sixty three children(24 boys and 39 girls)aged between 1 and 97 months years old(mean 20)with primary VUR receiving 0.5 ml/Kg ivumir per day.Medium follow-up:24 months.Fifty seven patients underwent voiding cystourethrogram after initial episode of UTI,six in consequence of fetal diagnosis of pyelectasia.In these children 90 refluxing ureteres were involved(grade I:3,II:31,III:39,IV:16,V:1).10 patients had other associated urogenital malformations.
**Results:** The incidence of UTI was 4.7% (3/63).Technetium-99m dimercaptosuccinic acid scan, performed in 36/63 children(58%) 6-12 months after urinary tract infection,found renal abnormality in 7 patients.No patient suffered by side effects.
**Conclusions:** We conclude that administration of standard concentrated cranberry juice is free from side effects and effective in preventing UTI in pediatric patients with VUR including high grade VUR and urinary tract malformations associated. Larger prospective randomized controlled studies are needed to confirm our preliminary results.

## Abstract# 279

**Urine Collection in Non Toilet-Trained Children: Is Catheter Always More Painful Than Bag?** J. Guinaud,[1] C. Lamy,[1] A. Tahir,[1] V. Gajdos,[2] V. Guigonis,[1] P. Blanc.[3] *[1]Pediatrics, Hopital de la Mère et de l'Enfant, Limoges, France; [2]Pediatrics, Hôpital Antoine Béclère, Clamart, France; [3]Pediatrics, Hôpital de Poissy, Poissy, France.*
**Objectives:** Despite its drawbacks, bag remains a widely used device to collect urine in non toilet trained children suspected of urinary tract infection (UTI), mostly because of practical matters and concern about the level of induced pain. We conducted a prospective study in order to compare, from this point of view, bag and catheter urine collection.
**Methods:** In our centers, every positive dipstick performed on bag obtained urine samples are confirmed with an urethral catheterization before the child is treated. Therefore we conducted a prospective study in two phases to compare these two methods in the same children. The first part aimed to compare the duration of the procedure between the application of the two devices and the collection of urines. The second part aimed to compare, with a FLACC scale, the pain level induced by each of the two devices.
**Results:** 56 children were involved in the first part of the study. Mean (± SD) duration of the procedure was 35.7 ± 28.5 min and 1.3 ± 2.2 min when bag and catheter were usedrespectively. 72 children were involved in the second part of the study. Mean FLACC result was 3.29 ± 2.97 and 5.24 ± 3.35 when, bag and catheter were used respectively. Unexpectedly bag related FLACC was higher than catheter related FLACC in 22.2% of cases, and catheter and bag related FLACC levels were equal in 16.7 % of cases.
**Conclusions:** We confirm that urethral catheterization is a feasible and tolerable procedure in an emergency setting in order to check for UTI in non toilet trained children.

## Abstract# 280

**Is It Necessary To Perform a Lumbar Puncture in Young Infant with Febrile Urinary Tract Infection?** V. Smolkin,[1,2] R. Halevy,[1,2] D. Karni,[2] W. Sakran,[2,3] A. Koren.[2,3] *[1]Pediatric Nephrology Unit, Ha'Emek Medical Center, Afula, Israel; [2]Pediatric Department "B", Ha'Emek Medical Center, Afula, Israel; [3]Baruch Rappaport School of Medicine, Technion, Israel Institute of Technology, Haifa, Israel.*
**Objectives:** We studied the frequency of bacterial meningitis in infants with primary diagnosis of febrile urinary tract infection.
**Methods:** The study comprised neonates and infants, age 0-60 days, admitted to the pediatric department with febrile UTI from 1999 to 2008. Inclusion in this study was confirmed by a diagnosis of UTI based on positive urine culture. All these patients also underwent a lumbar puncture for cerebrospinal fluid examination as a part of fever workout guidline. The study was based on the computerized medical record.
**Results:** During 1999-2008, 184 neonates and infants 0-60 days of age have been hospitalized in pediatric department with the diagnosis of febrile UTI. There was no case of bacterial meningitis in all patients included in our study.
**Conclusions:** The study has shown that the frequency of bacterial meningitis in young infants with febrile UTI is almost zero. This fact raises a question about expendiency of lumbar pubcture and possibility of revising a fever workup guidlines in this group of patients.

## Abstract# 281

**Toll-Like Receptor (TLR) 4 Polymorphism Is Associated with Increased Risk of Urinary Tract Infection (UTI) in Girls above Age 2** V. Jankó,[1] S. Pozsgayová,[1] L. Podracká,[2] Lá. Kovács.[1] *[1]Department of Pediatrics, Comenius University Medical School, Bratislava, Slovakia (Slovak Republic); [2]Department of Pediatrics, Safarik University Medical School, Košice, Slovakia (Slovak Republic).*
**Objectives:** Innate immunity plays a central role in the development of UTI, however the genes involved remain largely unknown. In this study we tested the hypothesis that TLR4, a key element of innate defense, is associated with susceptibility to UTI in children.
**Methods:** We determined TLR4 A(896)G polymorphism by means of PCR in 113 children with upper UTI. Of them, 63 aged <2 years (34 F, 29 M) and 50 were >2 years (43 F, 7 M). Genotypes were compared with that of 230 controls. Clinical data were statistically evaluated.
**Results:** Urinary microbiological analyses detected E. coli (95 cases), E. faecalis (12), P. mirabilis (5 ) and Citrobacter (1). CRP values (in mg/l) were 81,7 (median: 58) in patients <2 years and 93,4 (median: 56,7) in children >2 years (p=ns). In the whole UTI group, the prevalence of the TLR4 (896) G allele did not statistically differ from controls (98/15 and 218/17 respectively, p=0,104). However, it was significantly higher in patients >2 years than in those < 2 years (50/11 vs 63/4, p <0.03) or in the control group (50/11 vs 218/17, p<0.041). A more significant association found in older children (p< 0,001) was due to higher proportion of girls in this group.
**Conclusions:** Our findings show that carrier status of TLR4 (896)G is associated with altered ability of girls >2 years to respond to urinary pathogens. It is suggested, that this polymorphism acts in concert with other innate and acquired factors to influence the physiological defense against UTI.

## Abstract# 282

**Hypercalciuria Is an Important Cause of UTI in Children** M. Abdullah,[1] M.S. Hossain,[1] A.H. Khan,[2] M.M. Alam.[3] *[1]Department of Biochemistry, Dhaka Medical College, Dhaka, Bangladesh; [2]Department of Paediatric Nephrology, National Institute of Kidney Diseases and Urology, Dhaka, Bangladesh; [3]Department of Nephrology, Natioanl Institute of Kidney Diseases and Urology, Dhaka, Bangladesh.*
**Objectives:** To evaluate the association between hypercalciuria and urinary tract infection in children.
**Methods:** This case control study was done on 30 cases of diagnosed urinary tract infection (UTI) patients admitted in paediatric nephrology unit in National Institute of Kidney Diseases and Urology, Dhaka and 40 age-sex matched apparently healthy control. Blood samples of all study subjects were analyzed for serum total calcium concentration to exclude hyper or hypocalcaemia. Random urine samples were analyzed for urinary calcium creatinine ratio ( Uca/Ucr) to screen hypercalciuria.
**Results:** Mean total calcium concentration of cases and controls were 8.87±70 mg/dl and 9.27±39 mg/dl respectively. In this study, Uca/Ucr≥ 0.2 was considered as hypercalciuria. Hypercalciuria were reported in 46.7% cases whereas only in 20.0% controls which was significantly higher in cases to controls (p<0.05). Among the cases, 76.7% had recurrent UTI and 23.3% had first time UTI. Hypercalciuria were reported in 52.2% cases with recurrent UTI and only in 28.6% cases with first time UTI. Hypercalciuria in cases with recurrent UTI were also found significantly higher compared to controls (p<0.05).
**Conclusions:** Hypercalciuria is associated with UTI and is an important cause of UTI in children.

## Abstract# 283

**Acute Focal Bacterial Nephritis in Children** H. Hwang,[1] K.H. Kim.[2] *[1]Department of Pediatrics, Fatima Hospital, Changwon, Republic of Korea; [2]Department of Pediatrics, Ilsan Hospital, Koyang, Korea.*

**Objectives:** There is only limited knowledge of Acute Focal Bacterial Nephritis (AFBN) in children. To aware the clinical importance and the need of proper management of AFBN, we performed this study.

**Methods:** We analyzed 30 AFBN patients and 30 other upper urinary tract infection (UTI) patients by comparative studies.

**Results:** 1) The incidence of AFBN was 5.6% and more common in boys than girls. 2) Since both groups had similar symptoms, it was difficult to diagnose AFBN by clinical presentations alone. 3) ESR and CRP were significantly higher in AFBN patients. 4) The positive results of urine cultures were seen in 22 cases of AFBN patients, with no significant difference when compared to the control group. The most common causative organism was *E.Coli* in both groups. 5) On the sonographic findings, the most lesions were seen on the upper lobe of the kidney, more frequent on the left side. The lesions of 29 cases showed globular or wedge shaped increased echogenecity compared with the adjacent normal renal cortex. 6) 99mTc-DMSA scan could detect earlier lesion of AFBN which was not seen on ultrasonography at initial diagnosis. It showed the complete coincidence of the location, size and shape in all cases compared to the findings of renal sonography. 6) 14 of 18 cases who had radiologic follow up showed improvement by antibiotics therapy alone.

**Conclusions:** Ultrasonography is excellent as an initial detection tool in diagnosing AFBN. Since the degree of infection in AFBN is more severe than other UTIs and evolution into a renal abscess is possible, early diagnosis and appropriate antibiotics therapy is essential.

## Abstract# 284

**Features of Lysosome Enzymuria in Children with Pyelonephritis Depending on Age** I. Bagdasarova, L. Korol, L. Migal, O. Lavrenchuk. *SI "Institute of Nephrology AMS of Ukraine", Kyiv, Ukraine; SI "Institute of Nephrology AMS of Ukraine", Kyiv, Ukraine.*

**Objectives:** The purpose of workis to study the age features of changes in activity of enzymes lysosomal origins: N-acetyl - β-D-glukosaminidase (NAG), its isoenzyme is NAG B and galaktosidase (Gal) in children with PN.

**Methods:** 186 children at the age from 2 till 16 years (mainly girls) with an active stage of inflammatory process in kidneys and without infringement of a functional condition of kidneys which were derided in groups depending on the activity degree of PN (1-3), forms PN (obstructed -OPN and nonobstructed - NPN), character of a disease course (acute, chronic) and age features of patients (till 6 years-82, after 6 years-104 patients) are surveyed. Control group – was practically healthy 30 children.

**Results:** It was shown, that in all children with 3-rd degree of activity of inflammatory process in kidneys and practically in all children with the 2-nd degree, in patients with acute PN, and chronic OPN and NPN activity levels NAG, NAG B and Gal in children till 6 years statistically authentically exceeded similar indicators in children in above 6 years old. We noticed, that the maximum indicators of activity of lysosomal enzymes of urine among children are elderly till 6 years, and also among children above 6 years are registered in patients with chronic OPN with 2-nd and 3-rd degree of activity of inflammatory process in kidneys in comparison with patients with acute and chronic NPN.

**Conclusions:** So, we registered the dependence of changes in levels of activity of lysosomal enzymuria on the degree of activity of inflammatory process in kidneys and on the age of PNH children.

## Abstract# 285

**Probiotic Versus Antibiotic Prophylaxis in Infants with Primary Vesicoureteral Reflux** S.-J. Lee, M. Lee. *Pediatrics, Ewha Womans University School of Medicine, Seoul, Yangcheunku Mokdong, Korea; Pediatrics, Ewha Womans University School of Medicine, Seoul, Yangcheunku Mokdong, Korea.*

**Objectives:** Probiotics, beneficial living microorganisms, were deemed to be an alternative to antibiotics for prophylaxis but the preventive effect against UTI is inconclusive. To compare the effect of probiotic prophylaxis with antibiotic prophylaxis, a prospective randomized controlled study was done in infants with primary VUR.

**Methods: Methods**: 132 infants (age 5.2±4.5 months), who were diagnosed to have primary VUR after their first UTI, were randomly allocated into a probiotic (Antibio300$^{R}$*Lactobacillus acidophilus* $10^{8}$ CFU/g 1g twice daily, n=64) or an antibiotic (Septrin$^{R}$ trimethoprim/sulfimethoxazole (TMP-SMX) 2/10 mg/kg once at night, n=64) prophylaxis groups and were followed for one year. Four infants were dropped out and 128 infants were analyzed.

**Results:** The incidence of recurrent UTI in the probiotic group was 32.8% (21/64), which was not significantly different from 40.6% (26/64) in the antibiotic group ($P$=0.348). The causative organisms of recurrent UTI were not significantly different between the two groups ($P$=0.355). The resistant rates of causative uropathogens to TMP-SMX was 100% (26/26) in the antibiotic group, which was significantly higher than 14.3% (3/21) of the probiotic group ($P$<0.01). The resolution rate of primary VUR after one year was not significantly different between two groups [15.6% (10/64) in the antibiotic group vs. 18.8% (12/64)] in the probiotic group ($P$=0.676).

**Conclusions: Conclusion**: Probiotics prophylaxis have a similar effect as antibiotics prophylaxis in preventing recurrent UTI but was safer in infants with primary VUR.

## Abstract# 286

**Prediction of High-Grade Vesico-Ureteral Reflux after a First Urinary Tract Infection in Children: Construction and Internal Validation of a Clinical Decision Rule** S. Leroy,[1] C. Romanello,[2] V. Smolkin,[3] A. Galetto-Lacour,[4] B. Korczowski,[5] D. Tuerlinckx,[6] C. Rodrigo,[7] V. Gajdos,[8] F. Moulin,[9] P. Pecile,[2] R. Halevy,[3] A. Gervaix,[4] B. Duhl,[5] T. Vander Borght,[6] C. Prat,[7] F. Foix-l'Hélias,[8] D.G. Altman,[1] G. Bréart,[10] D. Gendrel,[9] M. Chalumeau.[9] *[1]CSM, Univ. Oxford, Oxford, United Kingdom; [2]Univ. Udine, Udine, Italy; [3]Ha'Emek Med Center, Afula, Israel; [4]Univ. Hosp. Geneva, Geneva, Switzerland; [5]Regional Hosp., Rzeszow, Poland; [6]Cliniques Univ. Mont-Godinne, Yvoir, Belgium; [7]Germans Trias i Pujol Hosp., Badalona, Spain; [8]Béclère Hosp., Clamart, France; [9]Saint-Vincent Hosp., Paris, France; [10]INSERM U953, Paris, France.*

**Objectives:** We aimed to derive a clinical decision rule to predict VUR≥3 in children with a first UTI to avoid unnecessary cystographies.

**Methods:** Secondary analysis of prospective series of children with a first UTI.

**Results:** 494 patients (8 centres, 197 boys, VUR ≥3 in 11%) were included. Procalcitonin (PCT) and ureteral dilation on US were significantly associated with VUR ≥3 and combined into a prediction model with a 0.75 [0.69-0.81] ROC area. Given the pre-specified constraint of achieving 85% sensitivity, our model led to the following clinical decision rule: cystography should be performed in cases with ureteral dilation and a serum PCT level ≥0.17 ng/mL, or without ureteral dilatation when the serum PCT level ≥0.63 ng/mL. The rule had a sensitivity of 86% [74-93] with a specificity of 47% [42-51]. Internal cross-validation produced 86% sensitivity [79 to 93] and 43% specificity [39 to 47].

**Conclusions:** A clinical decision rule was derived and predicted VUR ≥3 with ~85% sensitivity and avoids half of the cystographies that do not find high-grade VUR.

## Abstract# 287

**Prediction of High-Grade Vesico-Ureteral Reflux after a First Urinary Tract Infection in Children: External Validation of a Clinical Decision Rule** S. Leroy,[1] F. Bouissou,[2] A. Fernandez,[3] M. Gurgoze,[4] K. Karavanaki,[5] T. Ulinski,[6] S. Bressan,[7] S. Gardikis,[8] P. Leblond,[9] Y. Coulais,[2] C. Cubells,[3] A. Aygun,[4] C. Stefanidis,[5] A. Bensman,[6] L DaDalt,[7] G. Vaos,[8] S. Bigot,[9] D. Gendrel,[10] G. Bréart,[11] M. Chalumeau.[10] *[1]Univ. Oxford, Oxford, United Kingdom; [2]CHU Purpan, Toulouse, France; [3]Joan de Deu Hosp, Barcelona, Spain; [4]Univ. Medicine, Elazig, Turkey; [5]Kyriakou Hosp, Athens, Greece; [6]Trousseau Hosp, Paris, France; [7]Univ. Padova, Padova, Italy; [8]Alexandroupolous Hosp, Thrace, Greece; [9]Jeanne de Flandre Hosp, Lille, France; [10]Saint-Vincent Hosp, Paris, France; [11]INSERM U953, Paris, France.*

**Objectives:** We previously derived a clinical rule to predict VUR ≥3 at the time of the first UTI: cystography should be performed in cases with ureteral dilation and a serum procalcitonin (PCT) ≥0.17 ng/mL, or without ureteral dilatation when the PCT ≥0.63 ng/mL. We sought to validate this rule.

**Methods:** A secondary analysis of prospective series of children with a first UTI.

**Results:** 413 patients were included (8 centres, 157 boys, high-grade VUR in 11%). The rule had a sensitivity of 64% [50-76], with a specificity of 46% [41-51]. This led to a significant decrease of sensitivity between validation and derivation populations (20% [17-36]), but a non different specificity (0.4% [-7 to 7]. No major weakness in the validation study or difference in features of the populations could explain that resutl, neither the statistical rosbustness of the rule. However the lack of physiopathological link between VUR and PCT might lead to a refinement of the rule into a robuster one.

**Conclusions:** The rule was not enough reproducible, and some refinement might be warranted before clinical use.

## Abstract# 288

**Comparison of the Efficacy of Renal Ultrasonography, 99mtechnetium DMSA Renal Scan in Children with Urinary Tract Infection with Vesicourethral Reflux in Korea** I.S. Lim, S. Kim. *Department of Pediatrics, College of Medicine, Chung-Ang University, Seoul, Korea.*

**Objectives:** So far, renal ultrasonography, 99mTechnetium dimercaptosuccinic acid (99mTc-DMSA) renal scan, and voiding cystourethrography(VCUG) were recommended for detecting abnormalities. Recently, questions have been risen over the efficacy of renal ultrasonography in patients who have been previously diagnosed with UTI, and there has been controversy over the use of various imaging methods in the diagnosis of UTI. So therefore, we studied the efficacy of renal ultrasonography and 99mTc-DMSA renal scan in the diagnosis of UTI with VUR in Korea.

**Methods:** We retrospectively studied 169 children who were admitted to Chung-Ang university hospital for UTI from January 2005 to September 2009.

**Results:** 42 children(24.9%) had abnormal ultrasonographic findings; 36 with hydronephrosis, 2 with ureteropelvic junction obstruction, and 1 with ureterocele. 30 children(17.8%) had photon defect in 99mTc-DMSA renal scan. 17 children(30.9%) had VUR; 4 with Grade I, 2 children with Grade II, 3 children with Grade III, 5 children with Grade IV, 3 children with Grade V. Sensitivity of renal US for VUR was 0.55, and positive predictive value was 0.37. there was no relation between abnormal ultrasonographic finding and VUR(p=0.50). Sensitivity of 99mTc-DMSA renal scan for VUR was 0.55, and positive predictive value was 0.42. There was no relation between photo defect and VUR(p=0.50).

**Conclusions:** Our study showed that renal ultrasonography and 99mTc-DMSA renal scan may not replace VCUG in the diagnosis of VUR. Therefore, in UTI, routine renal ultrasonography, 99mTc-DMSA renal scan, and VCUG may not be always necessary.

## Abstract# 289

**Methods for Reliability of Urine Culture from Bag Specimens in Urinary Tract Infection in Infants** I.S. Lim. *Department of Pediatrics, College of Medicine, Chung-Ang University, Seoul, Korea.*

**Objectives:** Since infants with suspected urinary tract infections (UTIs) cannot control urination, urine cultures for diagnosis are usually performed via urine bags. This method is noninvasive but has a high contamination rate. We studied the contamination rate of bag urine culture in diagnosing UTIs in infants under one year and the factors responsible for contamination in Korea, in order to achieve additional reliability of this method.

**Methods:** We examined patients under one year in whom urine culture through the urine bag method yielded over $10^5$ colonies of a single pathogen. We defined UTIs by referring to the guidelines of 2009 report of the committee on infectious diseases. We examined the factors responsible for contamination according to sex, duration of urine collection, and whether diarrhea took place with contamination rate.

**Results:** We examined 537 patients (372 males and 162 females). The contamination rate of one bag urine culture was 31.3%. Gender was not related to the contamination rate, but duration of urine collection showed an association with it ($P$<0.05). When duration of urine collection was divided into three groups: first group, <2 hours; second group, 2-4 hours; and third group, ≥4 hours, the contamination rates was 27.5%, 41.2%, and 44.1%, respectively ($P$=0.001). And diarrhea at admission had no impact on the contamination rate.

**Conclusions:** In infants with a suspected UTIs, urine collection for culture by bag method should be finished within 2 hours. If it takes over 2 hours, the urine bag must be removed and a new bag should be reattached to the infant for reliable results.

## Abstract# 290

**Prevalence of Monosynptomatic and Nonmonosymptomatic Enuresis in 739 School Children** G.T.B. Vaz, M.M.A. Vasconcelos, T.M.L. Santos, P.K.S. Torres, E.M. Lima. *Pediatrics, Federal University of Minas Gerais, Belo Horizonte, Minas Gerais, Brazil.*

**Objectives:** To investigate the prevalence of nocturnal enuresis (NE) in school children.

**Methods:** This is a cross-sectional study, including 739 school children aged 6 to 12 years from 3 public schools, being 239 from school 1 (socioeconomic class C), located in Belo Horizonte, and two social class D schools; school 2 with 267 students and school 3 located in the metropolitan area of high social risk, with 233 children. We carried out an interview in the school environment, with questions about the voiding during the night. NE was considered when the child had at least 1 episode of bedwetting per month. Children with NE received an educational booklet on the function of the lower urinary tract and also a referral for medical evaluation.

**Results:** NE was detected in 110 (14.8%) children and classified as monosymptomatic enuresis (MNE) in 63 (57.2%), being more prevalent in boys; nonmonosymptomatic enuresis (NMEN) were detected in 47 (42.7%) children, with higher prevalence in girls (34). Both were more common in younger children and in schools with more socially disadvantaged.

**Conclusions:** Although NE is a condition associated with multiple causes, has several consequences for the psychosocial and behavioral development of children. The individual interviews at the school helped select the children with NE. Early detection can prevent complications related to lower urinary tract, and improve the emotional aspects of children.

## Abstract# 291

**Family History of Urolithisis in Children Diagnosed of Urinary Tract Infection** M.-I. Luis-Yanes,[1] M.-D. Rodrigo-Jiménez,[2] K. Dublán-García,[3] E. Ramos-Trujillo,[4] V.E. García-Rodríguez,[1] V.M. García-Nieto.[1] *[1]Pediatric Nephrology Unit, Hospital Nuestra Señora de Candelaria, Santa Cruz de Tenerife, Islas Canarias, Spain; [2]Pediatric Nephrology Unit, Hospital Son Dureta, Palma de Mallorca, Islas Baleares, Spain; [3]Nefrología Pediátrica, Centro Médico Nacional La Raza IMSS, Ciudad de México, Mexico; [4]Unidad de Investigación, Hospital Nuestra Señora de Candelaria, Santa Cruz de Tenerife, Islas Canarias, Spain.*

**Objectives:** Children with idiopathic hypercalciuria have a higher frequency of urinary tract infection (UTI) than the control population. Conversely, it has never studied family history of urolithiasis in children diagnosed of UTI.

**Methods:** In this study, we conducted a survey on the existence of a family history of stones in the relatives of 1st and 2nd grades of 116 children (45M, 71F) diagnosed one or more times of UTI or asymptomatic bacteriuria.

**Results:** In 81 children (69.8%) there were family history of urolithiasis (26 from Grade 1, 46 from Grade 2 and 9 from 1 and 2 degrees). In contrast, only in 36 families of 128 children in the control group, any of its members had urolithiasis (28.1%) (p <0.001). 47/97 of patients (48.5%) were carriers of hypercalciuria and/or hypocitraturia (prelithiasis). There was an association between this last condition and the formation of renal scars (p=0.03).

**Conclusions:** The family history of urolithiasis in children with UTI is very frequently. Since urolithiasis has a genetic basis, it is possible that genetically susceptible children that are more prone to form stones, also can suffer ITU from birth because they are inadequately protected from bacteria such as *E. coli.*

## Abstract# 292

**Study of Renal Function in Children under Two Years Old Diagnosed with Acute Pyelonephritis** V.M. García-Nieto, S. González-Cerrato, L. Pérez-Baena, J.R. Alberto-Alonso, M.I. Luis-Yanes. *Pediatric Nephrology Uniy, Hospital Nuestra Señora de Candelaria, Santa Cruz de Tenerife, Canary Islands, Spain.*

**Objectives:** The diagnosis of acute pyelonephritis is suspected by clinical criteria and acute phase reactants and confirmed by urine culture and diminished uptake of the trazer detected in the renal parenchymal scintigraphy performed with dimercaptosuccinic acid (DMSA). Rarely, renal function was assessed in these cases.

**Methods:** We studied 61 children (32M, 39F) with abnormal scintigraphy (diminished uptake of the trazer in acute phase). In acute phase, all of them had given the maximum urinary osmolality (Uosm) stimulation with desmopressin. Also sets the values of urinary elimination microalbumin (MAU) and N-acetylglucosaminidase (NAG).

**Results:** The most common bacteria was E. coli (n=53, 86.9%). 24.6% (n=15) of patients had vesicoureteral reflux (VUR) and subsequently developed a renal scar. 86.9% of children (n=53) showed a reduced Uosm. The ratio MAU/Creatinine was elevated in 34.1% of cases (15/44) and it was observed an inverse relation between Uosm y UNAG/Ucr (r: -0.35; p: 0.02). 100% of children with one month or less showed a defect in renal concentration capacity (≤ 453 mOsm/kg).

**Conclusions:** When using sensitive functional parameters, there is impaired renal function in an elevated percentage of cases of pyelonephritis. This feature can be useful for the topographic location of urinary tract infections in the absence of scintigraphic study, and to strengthen the diagnosis of pyelonephritis in the centers in which the urine cultures were collected by non-invasive methods.

## Abstract# 293

**Prevalence of Breakthrough Infections during Prophylactic Antibiotic Treatment in Children with Primary Vesico-Ureteral Reflux** I. Máttyus, É. Kis, I. Várkonyi, T. Verebély, A. Nyitrai, Z. Agócs. *1st Department of Pediatrics, Semmelweis University, Budapest, Hungary.*

**Objectives:** The conservative therapy of vesico-ureteral reflux (VUR) consists of antibiotic treatment and prophylaxis of urinary tract infections (UTI). On the other hand long term administration of antibiotics is disputed. The aim of the study was to determine the prevalence of pyelonephritis among the primary VUR patients cured in our clinic.

**Methods:** We checked the prevalence of febrile urinary tract infections among the patients treated by antibiotic prophylaxis (cefuroxime/cefaclor and amoxycillin alternated on a monthly basis) at our department between the years 1994 and 2007. The percentage of different pathogenic bacteria has been

determined. Patients: Group 1: 57 patients with primary VUR, age at diagnosis < 60 days. Group2: 83 children with primary VUR, age at diagnosis > 60 days.
**Results:** In group 1, pyelonephritis occurred once in 11 patients (19%), 4 patients (7%) had more than 1 time pyelonephritis. Altogether 15 children (26%) had pyelonephritis at least once. In group 2, pyelonephritis occurred once in 18 patients (21%), 6 patients (7%) had more than 1 time pyelonephritis. Altogether 24 children (28 %) had pyelonephritis at least once. All infections were treated by targeted antibiotic therapy based on bacterial culture.
**Conclusions:** The prevalence of breakthrough pyelonepritis during prophylactic antibiotic therapy of children with primary VUR was 26% in group 1 (age <61 days) and 28% in group 2 (age >61 days). These data on prevalence are lower than given in the literature when different antibacterial protocols were used. In our study the pathogenic bacteria were polyresistant.

## Abstract# 294

**Vesicoureteral Reflux in Children after the First Urinary Tract Infection** N. MarcunVarda, J. Zapusek, A. Gregoric. *Department of Pediatrics, University Medical Centre Maribor, Maribor, Slovenia.*
**Objectives:** Vesicoureteral reflux (VUR) is commonly diagnosed in children after the first urinary tract infection (UTI). We focused on what the management of children with the first UTI was like during six years period in our department. We also considered the quantity of diagnostic tests performed (urinary tract ultrasounds, voiding urosonographies – VUS), the amount of children with VUR diagnosis and the way of treatment.
**Methods:** Complete medical documentation of the children with the first UTI, hospitalized in our department from the beginning of year 2002 till the end of 2007, was reviewed. Under these criteria 524 children were included. Afterwards they have been followed till the end of 2009.
**Results:** Every child after the first UTI was sonographically inspected. We decided to take further diagnostic steps in 151 (28.8%) children. Throughout this period 210 VUS have been performed. 58 of them were positive (27.6%), 147 were negative (70.0%) and 5 were unsuccessfully carried out (2.4%). VUR has been diagnosed in 42 children (8.0% of children). All children have been followed-up clinically. In 23.3% of children UTI has been diagnosed once again. The results of VUR treatment turned out to be successful. In one third of affected children VUR maturated spontaneously, second third was successfully treated with Deflux application. Only 9 children required classic surgical approach. Reflux nephropathy was diagnosed only in 3 children.
**Conclusions:** Our results show that diagnostic procedures in children after the first UTI have been performed in 28.8% of these children and VUR diagnosed in 8.0% of them, mainly with VUS. They speak in favor of the new management algorithm of these children.

## Abstract# 295

**Cranberry in Children with Recurrent Urinary Tract Infections** S.S. Maringhini,[1] A.A. Costa,[2] V.V. Migliore,[2] G.G. Pavone,[1] F.F. Leone,[1] C.C. Corrado,[1] R.R. Cusumano.[1] *[1]Pediatric Neprology, ARNAS Civico, Palermo, Italy; [2]School of Pediatrics, Palermo University, Palermo, Italy.*
**Objectives:** Urinary tract infections (UTI) are frequent in children and often recur. Antibiotic treatment may produce side-effects and microbial resistance. Since Cranberry is effective in reducing the number of UTI in adults we performed a study in children with recurrent UTI.
**Methods:** Seventy-nine consecutive children (56 F, 23 M) mean age 5,2 years (6 months to 17 years) with recurrent UTI (more than two UTI in six months) received a cranberry product (IVUMIR ®) 0,5 ml/kg/die in two doses for a mean of 6.4 months (range 1 to 21). A dipstick urinalysis was performed every month (and in presence of symptoms evocative of UTI).
Vesico-ureteral reflux (VUR) was present in 17 out of 66 (26%) pts. An abnormal ultrasound (ABUS) was registered in 24 out of 79 pts (30%). Voiding disfunction (VD) was present in 30 out of 68 (44%) pts.
**Results:** UTI average rate during treatment was 0.03/month compared *with* 0.4/month in the preceding period. UTI recurred in 11 (14%) pts during IVUMIR® with a rate of 0.2/month; these patients were all females and, compared to the others, they were older, had a higher percentage of ABUS and a higher percentage of VUR which was more often bilateral and of higher degree. No side effects were registered.
**Conclusions:** Profilaxys with IVUMIR reduces the rate of UTI. It is difficult to predict which child will benefit of this treatment.

## Abstract# 296

**Assessment of Reliability of Serum Procalcitonin Level for Differentiating Upper & Lower UTI** M. Mazaheri. *Pediatric Nephrology, Semnan University of Medical Science, Semnan, Semnan, Islamic Republic of Iran.*
**Objectives:** Precise differentiation of upper UTI (PN) from the lower is very important because badtreated or not treated PN can cause severe renal injury.Markers like CRP,ESR,WBC count can't define the location of infection exactly.In this study we evaluate reliability of serum procalcitonin (PCT) level for this meaning.
**Methods:** The study was performed on 88 patients suspected to haveUTI.Blood sample was drawn forCBC,diff,CRP,ESR & serum PCT level ( in the first 6 hours).Urinalysis & urine culture was checked.DMSA scan was done for all. IsotopicVCUG was performed after obtaining negative urine culture.PCT levels were measured by immunoluminometric method.Levels above0.5 ng/ml were considered as elevated.
**Results:** 58 cases had known to have UTI.On the basis of parenchymal involvement on DMSA scan,patients were devided in group**A** with PN(30 Cases)&group**B** with lowerUTI(28 cases).MeanPCT level in group **A**&**B** was5. 537±1.566&. 238±0.780 respectively(P<0.002). Sonographic finding showed abnormality in 46.7%of group**A**&21.4% in group **B**(p<0.04). Patients with higher PCT level had more parenchymal injury on DMSA scan (P<0.001). There was significant relation between level of PCT & of vesicourethral reflux(VUR).There was no significant difference between two groups for CRP,WBCcounts but mean ESR in group**A** and **B**was 52.2 ±4.7 & 36.6±2.6 respectively(p<0.01).Diagnostic value of each marker was checked by ROC curve.Area under the curve for PCT test was more than others (0.980).
**Conclusions:** PCT test can help the clinicians todifferentiate upper UTI from the lower.It's sensitivity is more than other markers likeCRP,WBCcount& ESR. Patients with higher PCT level had more parenchymal injury & they had more chance for VUR.

## Abstract# 297

**Comparison between MRVCUG and Contrast VCUG in Diagnosis of Vesicoureteral Reflux in Children** A.R. Merrikhi. *Pediatric Nephrology, Esfahan University of Medical Science, Esfahan, Esfahan, Islamic Republic of Iran.*
**Objectives:** ***Background***:This study is evaluate the diagnostic value of MRVCUG as a non-invasive and non-radiating alternative method to standard VCUG for diagnosis, managing and follow up of patient with VUR.
**Methods:** A total of 60 kidneys of 30 patients (7 boys and 23 girls) and (the mean age 5.15 years) were performed VCUG and MRVCUG within period of less than one or two months. At the time of MRVCUG studies, radiologist wasnot aware from the result of the VCUG performed by pediatric nephrologist. If needed, mild sedation with Chloral hydrate was been prescribed under supervision of pediatrician. The children were asked to drink water before the examination until they had a full-bladder sensation. Subsequent to the scout image acquisition,1mg/kg Furosemide was been administered (IM).
**Results:** Reflux grading according to MRVCUG was (26.7% normal, 43.3% mild, 20% moderate, 10% severe). According to standard VCUG findings the sensitivity and specificity of MRVCUG by kidney-ureter units were 92.6% and 68.4%, respectively. Agreement regarding the presence or absence of VUR by kidney - ureter units between MRVCUG and VCUG was 93%.Regarding the assessment of severity of VUR, a high level of agreement was achieved between MRVCUG and VCUG.
**Conclusions:** We conclude that MRVCUG can show reflux especially high - grades VUR (grade III–V reflux) with proper sensivity without ionizing radiation or catheterization.

## Abstract# 298

**Utilization of Plants as a Source of Medicine for Urinary Tract Infections in Lakshmipur District of Bangladesh** A.H. Mollik. *Epidemiology, Biostatistics, Community Nutrition and Noncommunicable Diseases, Peoples Integrated Alliance, Dhaka, Bangladesh.*
**Objectives:** Plants are widely used world wide to address a variety of health problems. Because of living conditions in rural areas of Bangladesh, urinary tract infections (UTIs) are common. Traditional health practitioners (THPs) provide primary healthcare to most of the rural population of Bangladesh and they use plants for this purpose. The THPs rely on decoctions made from plants or plant parts to treat UTIs, which decoctions can vary widely between THPs in the various districts of the country. The present ethnopharmacological survey was carried out in Lakshmipur district of Bangladesh.
**Methods:** An open-ended semi-structured questionnaire was used in collecting field information. Descriptive statistics were used to analyze the ethnopharmacological data collected. Factor of informant consensus was used to analyze the ethnopharmacological importance of the plants. The THPs described the signs, symptoms, and causes of UTIs. Details of the preparation and use of plants for management of UTIs were recorded. All plant samples were later identified at the Bangladesh National Herbarium.
**Results:** A total of 53 plant species representing 48 genera and 38 families employed in the traditional indigenous therapeutic applications of the people in Lakshmipur district of Bangladesh; are recorded from 321 homes. It was noted in this ethnopharmacological survey that the patients were quite satisfied with treatment by the THPs.

**Conclusions:** The results suggest that modern scientific studies have the potential of discovering new antimicrobial compounds in the above-mentioned plants, which can be effective as remedy for microorganisms causing UTIs.

## Abstract# 299

**Outpatient Pelvic Floor Therapy (Biofeedback) in Girls with Repeated Urinary Tract Infection and Dysfunctional Voiding** C.C. Mourani, B. Gerbaka, S. Merhej, N. Nehme. *Pediatrics, Hotel Dieu de France, Beirut, Lebanon; Pediatrics, Hotel Dieu de France, Beirut, Lebanon; Urology, Hotel Dieu de France, Beirut, Lebanon.*

**Objectives:** We compare in a prospective study the treatment of girls with repeated UTI and dysfunctional voiding. First group (A) was treated by an outpatient pelvic floor therapy program. Second group (B) was treated by conventional treatment : associating anticholinergic drugs and preventive antibiotherapy.

**Methods:** We analyzed the files of 62 girls (5 to 12 years) with at least one episode of UTI and high suspicion of overactive bladder (OBA). OBA was suspected on clinical signs and radiological investigations including ultrasound and voiding cystogram.

Thirty one girls in the group A received conventional treatment and 31 girls were treated by biofeedback and toilet behavior. No significant difference was noted in both groups concerning percentage and degree of reflux neither in the number of previous episodes of UTI.

**Results:** After six months of treatment and follow-up in both group, Data were complete in only 21 patients from group A and 19 patients from the group B. Two UTI episodes were reported in Group A compared to 4 in group B. Bladder capacity and day time incontinence improved in 20 patients from group (A ) compared to 17 patients from group (B).

**Conclusions:** This study demonstrated that biofeedback therapy alone was as efficient as classical treatment on improvement of voiding dysfunction (VD ) and number of UTT. Pelvic-floor exercise is a physiotherapeutic, noninvasive treatment. It should be included in the treatment of children with VD and UTI and it allows avoiding antibioprophylaxis treatment which is source of serious resistant infections.

## Abstract# 300

**Extended-Spectrum Beta-Lactamase Producing Bacteria Associated Urinary Tract Infections: Microorganisms Change – Clinical Pattern Does Not** Z.B. Ozcakar,[1] F. Yalcinkaya,[1] A. Kavaz,[1] G. Kadioglu,[1] S. Altugan,[1] D. Aysev,[2] H. Güriz,[2] M. Ekim.[1] *[1]Pediatric Nephrology, Ankara University Medical School, Ankara, Turkey; [2]Microbiology Laboratory, Ankara University Medical School, Ankara, Turkey.*

**Objectives:** The aim of this study was to investigate the clinical and radiological findings in patients with community acquired urinary tract infection (UTI)s due to extended-spectrum beta lactamases (ESBL) producing bacteria.

**Methods:** Files of the patients that had UTI due to ESBL producing bacteria, between the dates of January 2008- December 2009 were retrospectively evaluated.

**Results:** 111 UTI episodes (acute pyelonephritis in 45%) in 94 patients (66 girls, 28 boys; mean age: 62±54 months), due to ESBL producing bacteria were included. 25% of the patients had only one UTI episode, the rest had recurrent UTIs. 101 episodes were due to E.coli and 10 due to Klebsiella. Patients were already on antibiotic prophylaxis before 46% of the UTI episodes. Ultrasound, DMSA scan and VSUG were found abnormal in 39%, 42% and 32% of the patients, respectively. Overall, 64% of the patients had an underlying predisposing factor for UTI (voiding dysfunction in 38%, anomaly in 32%). When we divided the patients according to the age of the first UTI episode due to ESBL producing bacteria; 34 (19 boys) were <12 (Group 1), 23 (7 boys) were in between 12-60 (Group 2) and 37 (2 boys) were >60 months of age (Group 3). Acute pyelonephritis occured in 68%, 52% and 27% of groups 1,2 and 3 respectively. 78% of the patients in group 3 had voiding dysfunction.

**Conclusions:** Clinical pattern of UTIs due to ESBL producing bacteria does not seem to be different from the UTIs due to non-ESBL producing bacteria.

## Abstract# 301

**Nonkeratinizing Squamous Metaplasia of the Bladder in a 13-Year Old Girl with Gross Haematuria and Ureplasma Urealyticum Infection** I. Palcic,[1] J. Delmiš,[1] Z. Bahtijarevic,[1] G. Petkovic,[1] B. Krušlin.[2] *[1]Pediatric, Children´s University Hospital Zagreb, Zagreb, Croatia; [2]Pathology, University of Zagreb, School of Medicine, Zagreb, Croatia.*

**Objectives:** We present a 13-year old girl with gross haematuria and nonkeratinizing squamous metaplasia of the bladder caused by Ureaplasma urealiticum urinary tract infection.

**Methods:** The girl was referred with history of gross haematuria and suprapubic pain. She had no dysuria, frequency, urgency, incontinence, history of trauma or bleeding disorder. Physical and genitourinary tract examinations were normal. Urinanalysis showed gross haematuria. Urine culture, renal and bladder ultrasound and urography were normal. Cystoscopy showed cystic changes and haemorrhagic erosions of the trigonal part of the bladder and urethra. Biopsy of lesions was performed and pathologic evaluation demonstrated nonkeratinizing squamous metaplastic cells. Urethral cultures revealed Ureaplasma urealitycum infection that was treated with appropriate antibiotics.

**Results:** After two cycles of antibiotics she had no haematuria and urethral smear culture was sterile. Control cystoscopy showed a mild regression of cystic changes and erosions of bladder mucosa.

**Conclusions:** Nonkeratinizing squamous metaplasia is a sign of chronic infection, in this case with Ureaplasma urealyticum. It can be present as gross haematuria. Ureaplasma urealyticum infection of genitourinary tract is considered a sexual transmitted disease affecting sexual active women or abuse victims. Gynecologic and psychological examination showed no signs of sexual activity or abuse. To the best of our knowledge, our patient is the youngest child reported with this lesion so far.

## Abstract# 302

**Low Predictive Value of Ultrasound for Detection of Vesicouretral Reflux in Infants after the First Acute Pyelonephritis** D. Paripovic,[1] B. Spasojevic,[1] D. Kruscic,[1] G. Lomic,[1] M. Cvetkovic,[1] D. Milovanovic,[2] M. Kostic,[1] A. Peco-Antic.[1] *[1]Nephrology Department, University Children's Hospital, Belgrade, Serbia; [2]DZ, Novi Beograd, Serbia.*

**Objectives:** To investigate predictive ability of ultrasonographic exam (US) in detecting vesicouretral reflux (VUR) in infants with the first detected acute pyelonephritis (APN).

**Methods:** All infants with the first detected APN hospitalized between 2005 and 2009 were included in the study. Kidney US was performed during APN and voiding cystourtehrography (VCUG) within next 2 months. The comparison between US and VCUG was performed using kappa coefficient and receiver operator curve (ROC).

**Results:** 292 infants (143 males) with the first APN were included in the study. Median age was 5 months (range 1-12 months). Majority of pts had normal kidney ultrasound (72%) and VCUG (74%). Pyelectasis (14%), hydronephrosis (6%) and megaureter (2%) were the most common structural abnormalities during APN. VCUG was performed in 192 of pts. Mild VUR (gradus I to II) was present in 22% and high-grade (III-V) only in 4% of pts. There was no difference in gender in prevalence of imaging abnormalities. Pyelectasis, increased parenchyma echogenicity, double contour of pyelons, renal swelling and retrovesically visible ureters were the most significant ultrasound findings that were typical for VUR. There was a poor agreement between US and VCUG for detection of VUR (kappa 0.09). Using the ROC, US was not significant method for prediction of VUR (AUC 0.55±0.05).

**Conclusions:** Ultrasound seems to have a low predictive value for detection of VUR in infants after the first APN. Ultrasound still can't replace VCUG in detection of VUR.

## Abstract# 303

**Urine Matrix Metalloproteinases-2 and -9 (MMP-2, -9) and Tissue Inhibitor-1 (TIMP-1) in Children with Pyelonephritis (PN)** S.S. Paunova,[1] Y.A. Savenkova,[1] A.G. Kucherenko,[2] L.A. Revenkova,[1] O.V. Anokhina,[1] N.L. Goltsova.[1] *[1]Chair of Pediatrics, Russian State Medical University, Moscow, Russian Federation; [2]Dept of Pathophisiology, Scientific Center of Healthy Children, Moscow, Russian Federation.*

**Objectives:** Acute inflammation of renal tissue is considered to be one of the most common diseases in childhood, that leads to fibrosis. Recent studies show the important role of extracellular Zn-containing proteases in renal damage.

In order to reveal the possible role of MMP-2,-9 and TIMP-1 in renal scarring formation 25 children (aged 1-12 years) with PN were examined. 8 of them with acute presentation of PN were examined twice (at onset and after 10-14 days of antibacterial treatment). Seven of 25 patients had a remission of PN.

**Methods:** ELISA-measured MMP-2, MMP-9, TIMP-1 in urine standardized to urinary creatinine (uCr) concentrations were evaluated.

**Results:** All children with acute PN demonstrated significant increase of MMP-2, -9 and TIMP-1 in the urine. After treatment MMP-2 level decreased in 2 times, MMP-9 level- in 10 times. Only TIMP-1 was still elevated ($p<0,05$). Out of 7 children with remission of PN, four had higher urinary level of MMP-2, -9 and TIMP-1 than others ($p<0,001$), close to patients with acute inflammation. Theremore TIMP-1 urinary level was higher than MMP-9. These children presented with severe onset and had VUR.

**Conclusions:** High level of TIMP-1 in children with acute PN after treatment may be a result of progressing macrophages infiltration of renal tissue, leading to fibrosis. Significant disturbances in extracellular proteases system in patients with abnormal urodynamics may be considered as a risk factor of developing reflux nephropathy.

## Abstract# 304

**Increasing Prevalence of Antibacterial Resistance among Enterobacteriaceae Uropathogens** A. Peco-Antic,[1] D. Paripovic,[1] B. Spasojevic,[1] G. Lomic,[1] M. Cvetkovic,[1] S. Laban,[2] G. Lazarevic,[2] D. Kruscic,[1] M. Kostic.[1] *[1]Nephrology Department, University Children's Hospital, Belgrade, Serbia; [2]Microbiology, University Children's Hospital, Belgrade, Serbia; [3]DZ Novi Beograd, Belgrade, Serbia.*

**Objectives:** To assess evolution of resistance to commonly used antibacterial drugs of Enterobacteriaceae strains isolated from urine of children with the first acute pyelonephritis (APN).

**Methods:** Study included all patients treated due to the first documented APN during 2 periods: A (2005–2007) and B (2008–2009). Enterobacteriaceae were Gram stained and then identified. Susceptibility of these strains to antibiotics was determined by diffusion method. Following antibiotics were tested: ampicillin (Amp), cephalosporine (CS) and trimethoprim/sulfamethoxazole (TS). Extended-spectrum beta-lactamase (ESBL) strains were searched for by a double-diffusion method with cefotaxime and amoxicillin/clavulonic acid.

**Results:** During the last 5 years 352 children (median age 6 months) were treated due to APN, 99 in period A and 253 in period B. There was no difference in age, while boys were more prevalent in group B. C reactive protein was higher in period B. Enterobacteriaceae strains were the most dominant cause of APN in both periods. The distribution of strains among species did not vary significantly during periods A and B (Escherichia coli 86% vs 90%). Resistance to Amp was very high (>86%) and stable over 5 years period, while resistance to CS (28% to 65%) and TS (38% to 55%) increased significantly.

**Conclusions:** Our findings confirmed increasing prevalence of resistance to antibiotics and emphasize the need to reconsider empiric antibacterial treatment of APN in children.

## Abstract# 305

**Differences between the First and the Second Renal Scintigraphy in Children with Recurrent UTI** M. Poropat,[1] D. Batinic,[2] M. Ciglar,[1] D. Dodig,[1] S. Tezak.[1] *[1]Department of Nuclear Medicine and Rdiation Protection, University Hospital Centre Zagreb, Zagreb, Croatia; [2]Department of Paediatrics, University Hospital Centre Zagreb, Zagreb, Croatia.*

**Objectives:** We analyzed DMSA scintigraphy in children with recurrent UTI to find parenchymal damage.

**Methods:** We analyzed 117 children (234 kidneys), 80 girls and 37 boys, aged from 3 months to 12 years at the time of the first DMSA. All children underwent US and voiding cystourethrography. VUR was detected in 140 kidneys. Second DMSA was performed one to five years after the first. All children were on the antibiotic uroprophylaxis, and some of them were treated surgically with antireflux procedures. At the time of second DMSA VUR were found in 95 kidneys. DMSA scans were graded as normal, pathologic with scars or equivocal, and compared with presence of VUR and US findings.

**Results:** On the first DMSA out of 234 kidneys 63 were normal, 116 equivocal, and 55 pathologic while on the second DMSA 87 kidneys were normal, 78 were equivocal and 69 pathologic ($p<0.01$). On second DMSA kidneys with VUR (140 vs 95) and without VUR (94 vs 139) were significantly different ($p<0.01$). Comparison of DMSA in kidneys without VUR showed significantly increased number of normal (31 vs 53), and pathologic (17 vs 46) findings. 25 kidneys had pathologic US, 19 with pelvic dilatation, and 16 with parenchymal damage.

**Conclusions:** On second DMSA higher pathologic kidneys without VUR could be results pontaneous or surgical disappearance of VUR in damaged kidneys and less equivocal and more normal findings might be the result of antibiotic prophylactic treatment which is obligatory and very important for the prevention of renal damage.

## Abstract# 306

**Value of Ultrasonography in Work-Up of Infants with First-Time Urinary Tract Infection** I. Preda,[1] U. Jodal,[1] R. Sixt,[2] E. Stokland,[3] S. Hansson.[1] *[1]Pediatrics, The Queen Silvia Children's Hospital, University of Gothenburg, Gothenburg, Sweden; [2]Pediatric Clinical Physiology, The Queen Silvia Children's Hospital, Gothenburg, Sweden; [3]Pediatric Radiology, The Queen Silvia Children's Hospital, Gothenburg, Sweden.*

**Objectives:** Evaluate the ultrasound in the management of infants with first time urinary tract infection with focus on important structural abnormality.

**Methods:** In a setting of limited antenatal ultrasound screening, this population-based prospective 3-year study included 290 infants, 161 boys and 129 girls. Ultrasound and DMSA scintigraphy were performed as acute investigations and cystourethrography within 2 months. Important structural abnormality of the urinary tract detected both in as outside of the study cohort were recorded.

**Results:** Ultrasound showed dilatation in 15% and increased kidney length in 28% of the patients. The sensitivity to detect scintigraphic abnormality was 48%. Renal length was significantly correlated to inflammatory parameters including scintigraphic abnormality. Important structural abnormality was detected in 40 infants of whom ultrasound identified 30, while 10 of 27 cases with dilating reflux were missed, mostly grade III. Outside of the study there were a further 28 infants with structural abnormality of whom 15 were antenatally detected.

**Conclusions:** Ultrasound detected most infants with structural abnormality with the exception of reflux grade III. Being noninvasive, ultrasound has a place in the work-up of infants with urinary tract infection, especially when antenatal ultrasound during late pregnancy is lacking. Kidney length in infants with acute infection correlated with inflammatory parameters and this finding needs to be studied further.

## Abstract# 307

**Prevalence of Vesico Ureteric Reflux (VUR) and Renal Scars in Children with Urinary Tract Infection** H. Rahman, M. Hossain, G. Muinuddin, A. Begum, A. Rahman. *Dept Pediatric Nephrology and Pulmonology, Bangabandhu Sheikh Mujib Medical University, Dhaka, Bangladesh.*

**Objectives:** To see the prevalence of VUR in children presented with UTI and also to correlate the grading of VUR with renal scar.

**Methods:** Sixteen children of both sexes from six months to twelve years were enrolled for the study. UTI associated with other systemic illness like nephrotic syndrome, acute glomerulonephritis and SLE were excluded from the study. Patients were diagnosed by relevant investigations like urine R/M/E and cultural sensitivity test, ultrasound of the kidney MCU, DTPA renorgram with total and split renal function and DMSA scan too see renal scar were done for each patient. Serum creatinine was evaluated for every patient to see renal function status.

**Results:** Findings were 4 (25%) patients with UTI had no VUR but 12(75%) had different grades of reflux. Out of 12 patients 7(58.33%) had grade iv reflux and only one patient (8.33%) had grade V reflux. When different grades of renal scar were evaluated by DMSA scan it was observed that out of 7 patients having grade IV VUR, 3(42.8%) and 4(51.15%) patients had grade III and IV renal scar respectively. One patient with grade V, VUR had grade IV renal scar. When renal scar was correlated with hypertension it was observed that patients with grade IV and grade V renal scar had hypertension in 5(71.42%) and 1(100%) cases respectively.

**Conclusions:** Though the sample size is too small to draw any definite conclusion but we may draw a conclusion from this study that higher grade of VUR is mostly related with UTI and in higher grade of VUR from grade III to V renal scar develop frequently.

## Abstract# 308

**Serum C Reactive Protein (s-CRP) and Serum Interleukin 6 (s-IL-6) and Childhood Renal Scarring Risk** L.M. Rodríguez,[1] J.M. Marugán,[2] B. Robles,[1] A. Suárez,[1] J.M. García,[3] M. Fernández.[1] *[1]Nefrología Pediátrica, CALE, León, Spain; [2]Pediatría, Hospital Clínico, Valladolid, Spain; [3]Inmunología, CALE, León, Spain.*

**Objectives:** Investigate whether s-CRP and s-IL6 predict the risk of renal scarring after urinary tract infection in children (UTI).

**Methods:** We present an study carried out with 32 children (9 male) (range: 0.6-153 months); diagnosed with UTI.

s-CRP and s-IL6 were measured at the time of UTI diagnosis and the patients forming renal scarring during follow up were identified by means of DMSA renal scan.

The effectiveness of the two parameters for diagnosis of renal scarring was evaluated using the indices of diagnostic quality: sensitivity (S) and specificity (Sp).

**Results:** Eight children (25%) (range: 2-144 months) showed renal scarring after the follow up and 24 children (range: 0.6-153 months;) did not present post UTI damage.

s-CRP was 110.23±59.69 and 52.46 ±63.13 mg/L respectively, in patients with, and without renal scarring. s-IL6 was 18.34±11.80 and 8.07 ± 9.51 pg/ml respectively, in patients with, and without renal scarring. The value of greatest S for CRP was ≥5mg/L (S:100%) and greatest Sp was ≥150mg/L (Sp: 95.83). The value of greatest S for s-IL6 was ≥4pg/ml (S: 100%) and maximum Sp was ≥ 40pg/ml (Sp: 100%). The cut off points for optimum diagnostic result in diagnosing renal scarring were 115mg/L for s-CRP and 20pg/ml for s-IL6.

**Conclusions:** The results confirm that children who will form renal scarring show higher levels of s-IL6 and s-CRP at the time of diagnosis of UTI and that very high values for these two parameters almost completely guarantee future formation of renal scarring. However, neither of the techniques provides sufficient information for predicting renal damage in all patients.

## Abstract# 309

**Recurrent Urinary Tract Infection in Girls: Do Urodynamic, Behavioral and Functional Abnormalities Play a Role?** S. Rudaitis, B. Pundziene. *Pediatric Nephrology Department, Kaunas Medical University Hospital, Kaunas, Lithuania.*

**Objectives:** To determine urodynamic, behavioral and functional abnormalities predisposing to recurrent urinary tract infection in 5- to 17-year-old girls.
**Methods:** Prospective case-control study was done. Evaluation included complete history, voiding-drinking diary, bowel questionnaire, physical investigation, sonography, voiding cystourethrogram, urodynamic investigation.
**Results:** Out of 148 girls, 104 had recurrent urinary tract infection (rUTI) (case group) and 44 nonrecurrent UTI (control group). Age ≤6.5 years (area under the ROC curve, 0.66; p<0.05) was a risk factor for rUTI. Association of rUTI with abnormal voiding frequency (case group - 47 (45.2%) vs control group 77 (61.4%)), postponing voiding-42 (40.4%) vs 6 (13.6%), functional stool retention-31 (29.8%) vs 2 (4.5%), poor fluid intake-35 (33.7%) vs 3 (6.8%), abnormal bladder capacity-32 (30.8%) vs 6 (13.6%), post-void residual urine volume >20 mL-3 (36.5%) vs 4 (9.1%) established (p<0.05). Of the 104 urodynamic investigations, none of the factors: detrusor overactivity, detrusor pressure (Pdet), maxPdet, EMG activity during voiding was associated with rUTI (p>0.05). In a multivariate model, independent risk factors for rUTI: age ≤6.5 years at the first UTI (OR=0.9; 95% CI, 0.85-0.98), abnormal voiding frequency (OR=5.3; 95% CI, 1.1-26.2), voiding postponement (OR=3.8; 95% CI, 1.4-10.1), poor fluid intake (OR=9.2; 95% CI, 2.5-33.6), residual urine >20 mL (OR=1,1; 95% CI, 1.0-1.1).
**Conclusions:** Independent risk factors for rUTI: age ≤6.5 years at first UTI, abnormal voiding frequency, voiding postponement, poor fluid intake, residual urine.
*The source. JN Vol.22(6):766-73.

## Abstract# 310

**Combined Use of Renal Scintigraphy and Ultrasound Scanning as First Line Screening after First Episode of Urinary Tract Infection in Children** I.G. Quirino, J.M. Penido Silva, E.M. Lima, J.S.S. Diniz, A.C. Simoes e Silva, E.A. Oliveira. *Pediatrics, Federal University of Minas Gerais, Belo Horizonte, Minas Gerais, Brazil.*

**Objectives:** The aim of this study was to evaluate the diagnostic accuracy of renal scintigraphy (CRE) and renal ultrasound scanning (US) in identifying high grade vesicoureteral reflux (VUR) in children after the first episode of urinary tract infection.
**Methods:** A total of 553 children after first urinary tract infection which were assessed by three diagnostic imaging studies (renal US, DMSA scanning and voiding cystourethrography) were included in the analysis. The main event of interest was the occurrence of high grade VUR (III-V). Diagnostic odds ratio (DOR), sensitivity, specificity, positive predictive value (PPV), negative predictive value (NPV), and likelihood ratio of renal scintigraphy and renal ultrasound scanning were determined for both methods separately and also in combination.
**Results:** From January 1985 until December 2007, 533 patients were included in the study. VUR was diagnosed in 246 (46.2%), 144 of them grade III –V. DOR values were lower for low grade VUR (I-II): 1.91 and 2.2, respectively by using the "OR rule" and "AND rule". On the other hand, higher DOR values were obtained in high grade VUR evaluation: 10.9 for DMSA (95% CI, 6.7 - 17.7) and 7.9 for US (95% CI, 4.9 – 12.78). The combination of US and DMSA features significantly increased DOR to 25.3 and 16.9, respectively by using the "OR rule" and "AND rule".
**Conclusions:** Our findings support that the combined use of US and DMSA scans are reliable predictors of clinical significant VUR.

## Abstract# 311

**The Incidence of Recurrent Urinary Tract Infection in Children with Vesicoureteral Reflux Hospitalized with Fever** R. Halevy,[1,2] V. Smolkin,[1,2] Y. Haim,[2] W. Sakran,[2,3] A. Koren.[2,3] *[1]Pediatric Nephrology Unit, Ha'Emek Medical Center, Afula, Israel; [2]Pediatric Department "B", Ha'Emek Medical Center, Afula, Israel; [3]3Baruch Rappaport School of Medicine, Technion, Israel Institute of Technology, Haifa, Israel.*

**Objectives:** UTI is one of the most frequent bacterial infections in children. Recently, a number of studies have contented that the presence of VUR does not increase the incidence of recurrent UTI. The aim of our study was to define the frequency of recurrent UTI in patients with VUR hospitalized with fever and also the influence of prophylactic treatment on the UTI's recurrency.
**Methods:** The study comprised 246 patients admitted to the pediatric department during 1999 to 2007. All the patients had UTI in the past and VUR was diagnozed in all cases. In the process of study we have analyzed both demographic, laboratory and rentgenologic data.
**Results:** 56.2% out of 246 patients hospitalized with febrile disease, had UTI. 80.4% of these patients got prophilactic treatment and 19.6% did not get it. As it turned out there was no difference in the frequency of repeated UTI between these 2 groups (57% vs 56.2% ). High percent of the *Pseudomonas* growth in urinary culture (18.6%) was found out in patients who had prophylactic treatment.
**Conclusions:** Patients with VUR have an increasing frequency of recurrent UTI. There is no difference in the incidence of recurrent UTI between children having prophylactic treatment and not having it.Prophylactic treatment increases the percent of *Pseudomonas* as a pathogen of UTI. All the above-mentioned findings lead us to the conclusion about the necessity of revising the treatment guidlines in patients who have suffered from UTI and diagnozed as having VUR.

## Abstract# 312

**Treatment Delay in Urinary Tract Infection and Permanent Renal Damage in Children with Urinary Tract Infection** S. Swerkersson, E. Stokland, Rune Sixt, U. Jodal, S. Hansson. *Department of Pediatrics, Sahlgrenska Academy,Gothenburg University, Gothenburg, Sweden.*

**Objectives:** Delayed treatment in children with UTI has been proposed as an important factor in development of renal damage. However few clinical studies support this statement. We studied the impact of treatment delay on renal damage.
**Methods:** Retrospective analysis of 402 boys and 394 girls < 2 years of age with a single episode of symptomatic UTI. All were examined with VCU and DMSA scan. Renal damage was classified as discrete, moderate or severe and as general or focal. Information of fever duration was available in 698 children.
**Results:** Median age in boys was 3.4 and in girls 8.5 months**.** The mean duration of fever at start of treatment was 2.4 days in boys and 3.1 days in girls (p<0.0001). Duration of fever was significantly associated with age and CRP level. Vesicoureteral reflux (VUR) was found in 148 (19%) children: grade I in 5%, II in 5%, III in 5%, grade IV in 4% and V in < 1%. At follow-up DMSA abnormalities were detected in 221 (28%) children, discrete in 16%, moderate in 8% and severe in 4%. Of those with scarring 162 (73%) were focal and 59 (27%) generalized. Grade of VUR and maximal CRP were significantly correlated to renal damage. In children with abnormal DMSA scan mean fever duration was 2.9 days and for those with normal DMSA scan 2.8 days (p=0.5).
**Conclusions:** Early treatment in children with UTI was more frequent in young age and in males. Treatment delay was not associated with increased risk of permanent renal damage. This is in contrast to the prevalent opinion that treatment delay is a major risk factor for renal scarring. However the results are in concordance with several recent clinical studies.

## Abstract# 313

**First Time Urinary Tract Infection in Children. Clinical, Laboratory Findings and Risk of Renal Scarring** D. Tramma,[1] G. Gerasimou,[2] A. Papadopoulou.[1] *[1]Pediatric, Aristotle University, Thessaloniki, Greece; [2]Nuclear Medicine, Aristotle University, Thessaloniki, Greece.*

**Objectives:** The aim of our prospective study was to evaluate the clinical and laboratory characteristics of children presented with first urinary tract infection (UTI) and developed renal scars.
**Methods:** Inclusion criteria : a) first incident of febrile UTI, b) two of the following findings : fever> 38.5 ° C, WBC > 10.000 cells/mm$^3$, ESR> 20mm/h, CRP> 20mg/dl, c) absence of congenital urinary abnormalities, except of VUR e) no UTI relapses between the two scintigraphies.
$^{99m}$Tc-Dimercaptosuccinic acid renal scintigraphy (DMSA) was performed within 3 days after admission and if abnormal, then follow-up DMSA was performed after 6-8 months. Cystourethrography was performed 1 month after UTI.
**Results:** A total of 70 children were enrolled in the study. The main pathogen was E.coli. VUR was found in 21.5% of the children. 75% of the children had findings of acute pyelonephritis (APN) in DMSA and there was a complete recovery in 68% of them. Scars were observed more frequently in older children, with VUR grade>III and not taking chemoprophylaxis.
**Conclusions:** 1) first episode of APN in children occurs in more than 50% during the first year of life. Boys are mostly affected, 2) the 'classic' clinical and laboratory findings of APN can serve as criteria for diagnosis of UTI level, 3) VUR does not appear to be associated with first episode of APN, 4) children>1 year of age had a higher risk of renal scarring, 5) chemoprophylaxis may serve as a protective agent to renal scars formation.

## Abstract# 314

**Nephromegaly Is a Highly Significant Risk Factor of Renal Scarring in Children with First Febrile Urinary Tract Infections** Y.-K. Tsau, C.-H. Cheng. *Pediatrics, National Taiwan University Hospital, Taipei, Taiwan; Pediatrics, Chang Gung Children's Hospital, Taoyuan, Taiwan.*

**Objectives:** The aim of this study was to examine whether ultrasonographic nephromegaly can be a way to pick out the patients with urinary tract infection (UTI) at increased risk of renal scarring and to reassess the value of renal ultrasonography.
**Methods:** Patients with a first febrile UTI were chosen for this study. All patients underwent renal ultrasonography. Follow-up urine-culture analyses

were performed to exclude recurrent infections before renal scintigraphy. All patients underwent DMSA scintigraphy at least 6 months later to assess any renal scarring.
**Results:** A total of 545 children (80 with nephromegaly and 465 without nephromegaly) were enrolled. UTI patients with nephromegaly have a more severe disease than patients without nephromegaly in terms of higher inflammatory indices and C-reactive protein and longer fever durations before treatment and after treatment. The incidence of renal scarring is much higher in UTI patients with nephromegaly. When independent predictor variables and their interaction were evaluated to determine their influence on scarring, only nephromegaly was significantly associated with a higher incidence of renal scarring.
**Conclusions:** Our results indicate that ultrasonographic nephromegaly at onset is associated with a very high incidence of renal scarring, and can be a way to pick out these UTI patients at increased risk of renal scarring. And we believe that renal ultrasonography should be done routinely in children with a first febrile UTI.

## Abstract# 315

**Escherichia coli Virulence Factors in Children with Urinary Tract Infections** A. Larakeb,[1] H. Vu Thien,[1] E. Bingen,[2] E. Grimprel,[1] A. Bensman,[1] T. Ulinski.[1] *[1]Pediatric Nephrology, Armand Trousseau Hospital - APHP - UPMC, Paris, France; [2]Microbiology, Robert Debre Hospital, Paris, France.*
**Objectives:** To compare E. coli virulence factors from urine cultures of children with cystitis and pyelonephritis and to analyse the relationship with vesico-ureteral reflux (VUR), sonography and biological inflammation markers.
**Methods:** We have studied prospectively over 22 months, 199 children (age, 1 day to 17 years) with urinary tract infections in one single center. Procalcitonin (PCT), clinical severity, sonography and voiding cystography were analysed. Eleven virulence factors (Sfa/foc, PapC, PapGI, PapGIII, FyuA, Hly, Aer, Cnf1, IroN, ChuA, hra) were analyzed using multiplex PCR.
**Results:** 169 patients had signs compatible with pyelonephritis. 15 had high grade and 30 low grade reflux, 124 patients had no anomaly.
Virulence profile of strains responsible for pyelonephritis without underlying urological anomalies is different from other UTI: Pap C (85% vs. 52.9%, p<=0.001) and Pap GII (82.2% vs. 47.1%, p<=0.001), suggesting stronger adherence capacity.
Strains found in patients with high grade reflux are similar to those with cystitis, and often belong to phylogenetic group A, which is less virulent (23.6% vs 2.0% in patients with high grade VUR; p<0.001).
Serum PCT is lower in patients with high grade (median=0.3 ng/ml) than low grade VUR (3.0 ng/ml) and without urinary tract anomalies (1.2 ng/ml). Absence of Pap GII is associated with major urological anomalies in 53% and has a negative predictive value of 94%.
**Conclusions:** Analysis of E. coli virulence genes by multiplex PCR together with PCT serum levels may help to predict high grade reflux in children with UTI.

## Abstract# 316

**The Postnatal Follow-Up of Infants with Antenatally Detected Hydronephrosis: The Importance of Early Circumcision** O. Yavascan, N. Aksu, O. Turan, A. Bal, F. Kamit, P. Kuyum, C.N. Arslan, M. Anil, A.B. Anil. *Pediatric Nephrology, Izmir Tepecik Teaching and Research Hospital, Izmir, Turkey.*
**Objectives:** The objective of this study was to determine the effect of circumcision on the frequencies UTIs and renal parenchymal damage as well as growth and nutrition of infants with antenatal hydronephrosis.
**Methods:** Infants with a fetal pelvis diameter of ≥5mm identified with antenatal US were followed-up. All patients were evaluated in terms of UTI frequency, pelvic diameter, scarring on DMSA and differential function on DTPA. Growth(heightSDS) and nutrition(relative weight) parameters were recorded as well. These parameters were assessed after circumcision. Statistical evaluation was made by using khi-square test.
**Results:** The study included 178 patients. Of these, 29 were diagnosed by VUR, 87 by obstructive uropathy and 54 by non-obstructive uropathy. Of 133 males, 111 infants were circumcised. The pre-circumcision UTI frequency[(2.97±1.14/year(y)] was significantly higher than post-circumcision period(0.25±0.67/y). Also, pre-circumcision UTI frequency(2.97±1.14/y) was significantly higher than the frequencies observed in female cases(0.85±0.91/y), in males who did not undergo circumcision(0.91±0.99/y) and in overall study group (0.73±0.79/y). In obstructive uropathy patients, the rate of renal damage was increased in uncircumcised patients(28.5%vs35.7%) whereas no change was observed in circumcised patients(11.2%vs12.5%). Overall, growth and nutrition parameters gradually improved after circumcision.
**Conclusions:** The close follow-up and early circumcision of infants with AH will prevent UTIs and renal parenchymal damage, enabling normal growth and nutrition.

## Abstract# 317

**The Fibronectin, beta-2 Microglobulin, and High Sensitive CRP Levels in Urinary Tract Infections with Renal Damage and VUR** H. Poyraz, N. Cetin, B. Yildiz, N. Kural. *Pediatric Nephrology, Eskisehir Osmangazi University, Faculty of Medicine, Eskisehir, Turkey.*
**Objectives:** The aim of this study was to assess the urinary and serum fibronectin and beta-2 microglobulin (B2MG) and high sensitive CRP (hsCRP) levels in urinary tract infections (UTI) with renal damage (RD) and vesico-ureteral reflux (VUR).
**Methods:** The patients were divided into five groups: group I, 20 patients with first UTI and VUR (-); group II, 20 patients with recurrent UTI and VUR (-); group III, 16 patients with recurrent UTI and VUR (+); group IV, 16 patients without UTI and VUR (+) and group V, 16 healthy children as the control group. RD was detected with DMSA in 17 patients.
**Results:** The serum and urinary fibronectin levels were similar in all study groups (p>0.05). The fibronectin levels were not associated with presence of RD (p>0.05). B2MG levels were higher in patients with first UTI than in those recurrent UTI. B2MG levels were higher in UTI(+)/VUR(+) patients than in controls. But B2MG levels were similar between in UTI(+)/VUR(+) patients and UTI(-)/VUR(+) patients (p>0.05). B2MG and hsCRP levels were higher in UTI with RD than in those UTI without RD. The hsCRP levels were higher in patients with RD than in without RD and controls (p<0.001). The hsCRP levels were higher in UTI(-)/VUR(+) patients than in controls.
**Conclusions:** In conclusion, our results suggest that fibronectin seems not to be useful for detection of levels of UTI and/or presence of VUR and RD. B2MG and hsCRP levels were related RD. The hsCRP may be useable for detection of ongoing inflammation UTI (-)/VUR (+) patients with VUR that these patients may be candidate for scar nephropaty.

## Abstract# 318

**Risk Factors of Antimicrobial Resistance in Children with Urinary Tract Infection Caused by Extended Spectrum Beta Lactamase Producing Bacteria** O. Kizilca, R. Siraneci, A. Yilmaz, N. Hatipoglu, E. Ozturk, A. Kiyak, D. Ozkok. *Bakirkoy Maternity and Children Hospital, Istanbul, Turkey.*
**Objectives:** Our aim was to investigate the risk factors of antimicrobial resistance in children with urinary tract infection caused by extended spectrum beta lactamase (ESBL) producing bacteria.
**Methods:** 344 patients, diagnosed as urinary tract infection (UTI) between January 2008 and December 2009, were enrolled in this retrospective study. Causative microorganisms were ESBL (+) in 148 patients and ESBL (-) in 196 patients.
**Results:** The causative microorganism isolated most frequently was Escherichia coli and 41.4% was detected as ESBL (+). Among Klebsiella species, 53.2% was ESBL (+). The resistance rates were 83.1% for TMP-SMX, 18.2% for nitrofurantoin, 47.3% for quinolones, 39.9% for aminoglycosides in ESBL (+) group, whereas 62.2% for TMP-SMX, 4.6% for nitrofurantoin, 9.7% for quinolones, 9.7% for aminoglycosides in ESBL (-) group. Being younger than one year old, high annual reccurence rate of UTI, longer duration of antibiotic prophylaxis, using cephalosporins for prophylaxis, previous hospitalization in last 3 months, having vesicoureteral reflux, using clean intermittent catheterization were found to be more frequent in ESBL (+) UTI group (p<0.05). According to logistic regression analysis age younger than one year old and high annual reccurence rate of UTI were identified as independent risk factors; the risk increased by 1,74 fold and 2,25 fold respectively.
**Conclusions:** Recognition of the risk factors for ESBL (+) bacteria may be helpful to determine new policies in management of UTI.

## Abstract# 319

**Detection and Diagnostic Value of Hepatic and Pulmonary Nodular Lesions in Pediatric Urinary Tract Infections** H.E. Yim, B.M. Choi, K. Yoo, Y.S. Hong, J.W. Lee. *Pediatrics, Korea University Guro Hospital, Seoul, Korea.*
**Objectives:** Clinically significant markers of urinary tract infection (UTI) and vesicoureteral reflux (VUR) can improve their diagnosis and help determine the risk for renal damage. We hypothesized that hepatic and/or pulmonary nodules on the ultrasound and dimercaptosuccinic acid scintigraphy can predict the renal risks in patients with UTI.
**Methods:** The clinical and radiological findings of 387 children with a proven evidence of UTI were retrospectively reviewed. The study sample included 19 pediatric patients with UTI combined with hepatic and/or pulmonary nodular lesions diagnosed by the imaging tests performed for the UTI.
**Results:** Hepatic nodules were detected in five patients, pulmonary nodules in 12, and both hepatic and pulmonary nodules in two patients. The mean age of the children was 24.5 months. VUR was detected in nine out of 17 patients (52.9%), and high grade VUR in 55.6% children with VUR. Acute pyelonephritis was identified in nine out of 18 patients and renal scarring in 57.1% patients with pyelonephritis. On follow-up, the hepatic and/or pulmonary nodules regressed in

all but one case, where the nodule was reduced in size. About 85.7% of patients experienced a recurrence of UTI within 1 year.
**Conclusions:** Our data shows that children with UTI and hepatic and/or pulmonary nodules had a high risk for renal damage, VUR, and recurrent infections. The hepatic and/or pulmonary nodules on the ultrasound and dimercaptosuccinic acid scintigraphy may offer a valuable sign for the proper management of the patients with UTI.

## General Nephrology

### Abstract# 320
**(O-41)**
**Cystatin C Is Superior to Creatinine in Children with Active Malignancy** H.N. Blufpand, J. Tromp, A. Bökenkamp. *Pediatrics, VU University Medical Center, Amsterdam, Netherlands.*
**Objectives:** Monitoring of renal function is crucial in children treated for malignancy. Muscle wasting is common among these patients and hampers the use of serum creatinine. Cystatin C has emerged as an alternative because its metabolism is independent of muscle mass, but data on its use in pediatric oncology are sparse. Hypothesis: estimation of glomerular filtration rate using cystatin C is more accurate than creatinine in children treated for malignancy.
**Methods:** Inulin clearance (Cin), estimated GFR using serum cystatin according to Filler (eGFRcys) and creatinine according to Schwartz (eGFRcrea) were measured in 68 patients with active malignancy and compared with 150 controls. Creatinine was measured using a kinetic Jaffé method, cystatin C by particle-enhanced immunonephelometry. We analyzed the difference between measured and estimated GFRs and the performance of both eGFRs for the detection of mild renal insufficiency.
**Results:** Multiple linear regression analysis showed an overestimation of GFR by eGFRcrea both in females (b=-8.65, p=0.001) and in patients with malignancy (b=-6.05, p=0.069). eGFRcys overestimated GFR in females (b=-7.33, p=0.016) but was independent of malignancy (b=3.90, p=0.335). Agreement between the eGFRs and Cin was comparable in the control group (k-value eGFRcrea 0.367 vs. eGFRcys 0.396), while eGFRcrea performed poorly (k -0.114) in the malignancy group. ROC analysis (cut-off of 90 ml/min/1.73m$^2$) showed that the diagnostic accuracy of eGFRcys was higher than eGFRcrea (AUC 0.823 vs. 0.599) in the malignancy group. Both performed comparably in the control group.
**Conclusions:** Cystatin C is superior to creatinine as a marker of renal function in children with active malignancy.
**DISCLOSURE:** *Bökenkamp, A.*: Other, The immunonephelometric assays were a kind gift from Siemens Healthcare, Marburg, Germany.

### Abstract# 321
**(O-42)**
**Effect of Simvastatin and Erythropoietin on Renal Fibrosis in Obstructive Nephropathy** Y. Acikgoz,[1] B. Can,[2] K. Bek,[1] A. Acikgoz,[3] O. Ozkaya,[1] G. Genc,[1] S. Sarikaya.[3] *[1]Pediatric Nephrology, Ondokuz Mayis University Faculty of Medicine, Samsun, Turkey; [2]Pathology, Ondokuz Mayis University Faculty of Medicine, Samsun, Turkey; [3]Urology, Ondokuz Mayis University Faculty of Medicine, Samsun, Turkey.*
**Objectives:** Progressive interstitial fibrosis is the most dramatic result of obstructive nephropathy (ON). The aim of this study is to show and compare the actions of simvastatin (Simv) and erythropoietin (Epo) in nuclear factor kappa B (NFkB), transforming growth factor-β (TGF β), basic fibroblast growth factor (bFGF), platelet-derived growth factor B (PDGF B), fibronectin expression and development of interstitital fibrosis in rats with experimentally induced unilateral ureteral obstruction (UUO).
**Methods:** Sprague Dawley rats, arranged in four groups as sham, Epo, Simv and control, were used. All rats except group sham underwent UUO. Epo, Simv and control groups were given Epo (1000IU/kg/every other day, intraperitoneal), Simv (2mg/kg/day) and vehicle by gavage respectively, for 14 days. For TGF-β, PDGF B, bFGF, NFkB and fibronectin expressions, immunohistochemical methods were used.
**Results:** TGF-β and fibronectin expressions were higher in control group compared to Epo and Simv groups. In addition, fibronectin expression in Simv group was higher than Epo group. Different than the Simv group, NfkB and bFGF expressions in control group were higher than Epo group.
**Conclusions:** As a result it was seen that Epo and Simv prevented fibrosis in ON. Epo was superior to Simv in this effect by suppressing the expressions of profibrotic factors NFkB, bFGF and fibronectin. Based on this findings, it was thought that Epo might be a better agent than Simv in the prevention of fibrosis in ON.

### Abstract# 322
**(O-44)**
**Prevalence of Impaired Kidney Function in Hospitalized Pediatric Patients** J. Sebestyen, U. Garg, B.A. Warady. *The Children's Mercy Hospital and Clinics, Kansas City, MO, United States.*
**Objectives:** The currently used chronic kidney disease (CKD) classification system characterizes the severity of kidney disease by 5 stages, based on the calculation of an estimated glomerular filtration rate (eGFR). Recently, the Schwartz formula was found to routinely overestimate the eGFR, the difference based on a change in the methodology of serum creatinine determination from Jaffe to enzymatic. A new estimating equation has been generated by the Chronic Kidney Disease in Children (CKiD) study, which more accurately estimates GFR. We recently developed a program at our institution which automatically calculates the eGFR by the CKiD formula based on the patient's serum creatinine, blood urea nitrogen and height recorded after hospital admission.
**Methods:** Collection of eGFR and demographic data from hospitalized children between 1-22 years over a 1 month period of observation.
**Results:** eGFR values were recorded from 262 patients. Mean age: 8.63 years, mean serum creatinine: 0.33 mg/dl and mean eGFR: 102.27ml/min/1.73m2. Prevalence of low eGFR by CKD stage was: stage 4: 0.4%, stage 3:6.4% and stage 2:25.4%. The mean age of patients with low eGFR was 8.6 year. Greater than 50% of patients with low eGFR were male. The subspecialties with the highest prevalence of eGFR<90 were general pediatrics 41.3%, endocrinology 35%, cardiology 50%, critical care 56.2%, nephrology 80%, neurology 60%, surgery 33%, and ENT 50%.
**Conclusions:** Based on this pilot study, the prevalence of impaired kidney function as determined by an accurate assessment of eGFR appears to effect more than one-quarter of hospitalized patients, potentially carrying the risk of previously undetected impaired kidney function in children.

### Abstract# 323
**(O-45)**
**Assessment of Renal Function by Cystatin C Level in Pediatric Patients before and after Induction Phase of Chemotherapy** K. Pirojsakul,[1] K. Tangnararatchakit,[1] A. Kositwattanarerk,[2] P. Chouplywech,[2] U. Anuratthapan,[1] W. Tapaneya-Olarn.[1] *[1]Pediatrics, Ramathibodi Hospital, Mahidol University, Bangkok, Thailand; [2]Radiology, Ramathibodi Hospital, Mahidol University, Bangkok, Thailand.*
**Objectives:** To evaluate glomerular filtration rate (GFR) and renal tubular function in oncologic patients before and after induction of platinum-based chemotherapy and to compare cystatin C with creatinine in determining GFR.
**Methods:** The newly diagnosed oncologic patients were tested for renal tubular functions and GFRs at 0, 1 and 3 months during platinum-based chemotherapy. Gold standard GFRs were measured by $^{99m}$Tc-DTPA plasma disappearance. The estimated GFRs were calculated by creatinine (conventional Schwartz), cystatin C (Bokeq, Zappitelli 1) and both creatinine/cystatin C (Zappitelli 2, new Schwartz). The precisions of estimated GFRs were evaluated by percentages of values were within ± 30% of gold standard GFRs.
**Results:** Fourteen patients were included. At 3 months, GFRs of the patients were normal but fractional excretion of magnesium significantly increased from 2.8 to 4.7% (P=0.017). The precisions of estimated GFRs were as follow: conventional Schwartz (50%), Zappitelli 1 (69%), Zappitelli 2 (73%), Bokeq (76%) and new Schwartz (78%). In malnutrition patients who had weight for height less than 100%, conventional Schwartz GFRs were 37% significantly higher than standard GFRs (P<0.01).
**Conclusions:** At 3 months, GFRs in patients receiving platinum-based chemotherapy were normal but minor tubular injuries were detected. Conventional Schwartz GFRs overestimated standard GFRs in malnutrition patients. Cystatin C may be an alternative marker for GFR estimation in such patients.

### Abstract# 324
**(O-46)**
**Ultrasound Evaluation of Nutcracker Syndrome in Caucasian Children with Orthostatic Proteinuria** D. Swieton, A. Zurowska, W. Kosiak. *Pediatric Nephrology & Hypertension, MUG, Gdansk, Poland.*
**Objectives:** To investigate the prevalence of left renal vein (LRV) entrapment by the aorta and superior mesenteric artery as a cause of orthostatic proteinuria in Caucasian children and to evaluate the value of LRV ultrasonography in identifying the nutcracker syndrome (NCS).
**Methods:** B mode and Doppler ultrasound of left renal vein was performed in 145 children aged 8-18 years (51 with orthostatic proteinuria, 44 with glomerular proteinuria and 50 healthy controls). A-P diameters and peak velocities at hilar (D, VmaxD) and aortomesenteric (N, VmaxN) portions of LRV were performed in both upright and supine positions. Cut off levels for D/N and VmaxD/VmaxN ratios were determined to identify NCS. ROC analysis was performed and AUCs calculated to assess diagnostic performance of sonographic indices.

**Results:** 92% of children with orthostatic proteinuria demonstrated renal vein entrapment on ultrasound examination as compared to 27% of children with glomerular proteinuria and 35% of healthy controls ( p= 0.0001 ). Cut off limits of D/N > 6.4 and VmaxN/VmaxD > 4.3 for upright ultrasound measurements showed a sensitivity of 92% and specificity of 65% for diagnosis of NCS. ROC analysis demonstrated the highest predictive value for orthostatic proteinuria of both the above ratios when measured in the upright position (AUC=0.85).
**Conclusions:** 1. Left renal vein entrapment can be demonstrated in the majority of Caucasian children with orthostatic proteinuria. 2. The best diagnostic performance of LRV sonographic indices are achieved for assessments in the standing position. 3. LRV sonography is a useful, noninvasive tool for confirming NCS in the initial investigation of proteinuria in children.

## Abstract# 325
**(O-47)**

**Melamine-Related Urolithiasis Children: Continuous Attention on the Long-Term Prognosis** X.-Y. Kuang, J. Gao, H. Xu. *Department of Nephrology and Rheumatology, Children's Hospital, Fudan University, Shanghai, China.*
**Objectives:** Melamine is the key factor of the outbreaks of urinary stones in China, 2008. 105 children were diagnosed as urinary system stones of screening 8335 children in our hospital. Continuous attention on their long-term prognosis should be stressed.
**Methods:** Urolithiasis children were followed up after six and eighteen months respectively. We rechecked the patients' urinalysis, urinary system ultrasonography, urinary Ca/Cr, ALB/Cr, IgG/Cr and NAG/Cr.
**Results:** 91.4% and 76.2% children engaged in the first (six months later) and second follow-up (eighteen months later), respectively. Ultrasonography showed 68.9% patients had disappeared stones in the first follow-up, as well as 86.1% patients did in the second, difference was significant (P=0.012). Similar results was found in 61 patients who participated in both follow-ups (54.1%, 86.9%; P<0.001). 74.3% and 76.2% patients were tested urinary ALB/Cr, IgG/Cr and NAG/Cr in both follow-ups. The glomerular injury detection rate in the second one (8.9%) was obviously less than that in the first one (32.1%, P<0.001), but it was no significant difference in the renal tubular injury rate (17.9%, 16.1%; P>0.05). Moreover, all quantitative analysis of IgG/Cr, ALB/Cr and NAG/Cr had statistically significance between them ($P_{IgG/Cr}$=0.038, $P_{ALBU/Cr}$<0.001 and $P_{NAG/Cr}$=0.026). Similar results were obtained from 58 patients who participated in both follow-ups ($P_{IgG/Cr}$=0.015, $P_{ALBU/Cr}$<0.001 and $P_{NAG/Cr}$=0.029).
**Conclusions:** Most Melamine-related urolithiasis children can excrete calculus spontaneously. Renal injury resulted by urolithiasis is reversible in most of them. The renal tubular injury should be paid more attention at further follow-up.

## Abstract# 326
**(O-48)**

**The Long-Term Safety Profile of Oral Desmopressin in Children with PNE** C. Van Herzeele,[1] J. Evans,[2] P. Eggert,[3] H. Lottmann,[4] J. Vande Walle,[1] J.P. Norgaard.[5] *[1]Pediatric Nephrology, UGent, Gent, Belgium; [2]Pediatric Nephrology, Hospital, Nottingham, United Kingdom; [3]Pediatric Nephrology, Hospital, Kiel, Germany; [4]Pediatric Urology, Necker Enfants Malades, Paris, France; [5]Ferring International, Ferring, Copenhagen, Denmark.*
**Objectives:** The recent discussion on the safety-issue of desmopressin as a spray in, has led to the withdrawal of the spray for the indication of enure MNE. The aim of the study is to evaluate the long-term safety profile of oral desmopressin in children with PNE.
**Methods:** Methods: prospective open-label, multinational, phase IV, observational study with up to 6 months follow up. 86 centres in 4 countries (UK, Canada, Germany, and France). Adverse events (AEs) were defined as any symptom during the study-period. Study-population: 936 screened, 744 enrolled, aged 8.7y ± 2.5, 531 M, dose 0.2 – 0.6 mg desmopressin tablet. Duration of therapy 148 ± 77 days.
**Results:** Results: 404 AEs in 222/744 (30%) patients were registered: 89 gastrointestinal symptoms (15 upper abdominal pain, 14 diarrhoea, 13 gastroenteritis, 24 vomiting), 18 general symptoms, including 14/18 pyrexia, 91 episodes of infection, 36 nervous system (35 headache), 12 psychiatric AEs, 85 respiratory AEs and 20 skin AEs. The incidence of AEs was constant in the 4 participating countries. 2% withdrawals for potential drug related side-effects. No deaths, and 8 serious AEs (including appendicitis), but none of them was drug related. Hyponatraemia was not docu-mented.
**Conclusions:** The Drip study is the largest (n=744) prospective long-term follow up study. Long-term desmopressin treatment is well tolerated in children with PNE : No serious AEs were related to the drug.
**DISCLOSURE:** *Van Herzeele, C.*: Grant/Research Support, PhD Grant. *Vande Walle, J.*: Consultant, Safety Board. *Norgaard, J.P.*: Consultant, Medical Director; Other, Medical Director.

## Abstract# 327
**(O-72)**

**Diagnosis, Treatment and Follow-Up of 25 Patients with Melamine-Induced Kidney Stones Complicated by Obstructive Renal Failure in Beijing Children's Hospital** Q. Sun,[1] Y. Shen,[1] G. Zhang,[1] Z. Chen,[1] B. Puschner,[2] J. Fan.[1] *[1]Beijing Children's Hospital, Beijing, China; [2]School of Veterinary Medicine, University of California, Davis, CA, United States.*
**Objectives:** A total of 25 Chinese patients aged 6 to 36 months hospitalised at Beijing Children's Hospital due to melamine-induced kidney stones complicated by acute obstructive renal failure in 2008 were included in a study in order to diagnose and treat these special cases more effectively.
**Methods:** Feeding history, clinical presentation, ultrasound findings, treatments and effects were summarised. Twelve to seventeen months follow-up was reported also.
**Results:** Ultrasound examination showed that calculi were located at the kidney and ureters. Stones were composed of both uric acid and melamine in a molar ratio of 1.2:1 to 2.1:1. Treatments providing liquid plus alkalisation of urine proved to be effective in helping the patients pass the stones. Surgical intervention was needed in severe cases. Renal function returned to normal in all 25 patients after various durations of therapy. Sixty-eight percent of the patients expelled all of the calculi within 3 months, 90% in 6 months and 95% in 9 months, without sequelae till the end of the follow-up.
**Conclusions:** Melamine-contaminated milk formula can cause kidney stones in infants, which should be diagnosed by feeding history, clinical symptoms and ultrasound examination. Composition of the stones was not only of melamine but also uric acid. Providing liquid orally or intravenously plus alkalisation of urine proved to promote the removal of the stones. Follow-up of 12 to 17 months after discharge showed no sequelae.

## Abstract# 328

**Protein Energy Wasting in Children with Chronic Kidney Disease** A.G. Abraham,[1] R.M. Zack,[2] R.H. Mak,[2] M.M. Mitsnefes,[2] R. Kaskel,[2] B.A. Warady,[2] S.L. Furth.[2] *[1]Johns Hopkins University, Baltimore, MD, United States; [2]CKiD Investigator, United States.*
In adults with CKD, protein-energy wasting (PEW), characterized by a panoply of biochemical and nutritional indicators, has been implicated as a risk factor for death and cardiovascular disease. The clinical relevance of these markers in pediatric CKD populations is unknown.
**Methods:** Using cross-sectional data from the Chronic Kidney Disease in Children study (CKiD), a definition of PEW was developed using markers identified from adult CKD populations. We assessed the prevalence of 7 indicators of PEW (anemia, poor weight growth, poor height growth, acidosis, low LDL, low dietary intake and hypoalbuminemia) and investigated the association of GFR and PEW using ordinal logistic regression. The relationship between severe PEW (≥ 4 indicators) and blood pressure was also examined.
Using data from 473 children we found 39 children with severe PEW (≥ 4 indicators) accounting for 8% of the cohort. The most common PEW indicator was anemia, occurring in 38% of the cohort and 92% of those with severe PEW. Acidosis and hypoalbuminemia were present in the cohort population at 18% and 6%, respectively, and they were 4 and 9 times as likely to be found in those with severe PEW (P<0.001). PEW and GFR were also associated (for every 10 ml/min decrease in GFR the odds ratio for being in a higher category of PEW was 1.43 [1.30, 1.57]). Severe PEW was associated with a 10 point higher systolic blood pressure percentile for age, sex and height (P<0.05).
Indicators of PEW were highly prevalent in the CKiD cohort of children with CKD, and PEW severity was associated with lower kidney function and higher blood pressure.

## Abstract# 329

**Sickle Cell Nephropathy in Children Seen in an African Hospital** O.T. Adedoyin, O.P. Fatoye, A.O. Bello, A. Adeniyi. *Paediatrics and Child Health, University of Ilorin Teaching Hospital, Ilorin, Kwara, Nigeria; Paediatrics and Child Health, University of Ilorin Teaching Hospital, Ilorin, Kwara, Nigeria; Paediatrics and Child Health, University of Ilorin Teaching Hospital, Ilorin, Kwara, Nigeria; Paediatrics and Child Health, University of Ilorin Teaching Hospital, Ilorin, Kwara, Nigeria.*
**Objectives:** To review the cases of sickle cell nephropathy seen over the last 14 years (1995-2009) at the Paediatric Nephrology clinic of University of Ilorin Teaching hospital, Ilorin, Nigeria.
**Methods:** The five cases of SCN seen during the period were analyzed for age, sex, and renal manifestations.
**Results:** The age range of the children was 9-15years with a mean of 11years. Four of the five patients were females, with one male. Three of the four females presented with features suggestive of nephrotic syndrome while the other one had gross haematuria which resolved within 24 hours. The only male had enuresis.

The nephrotic syndrome (NS) in one of the patients progressed to end stage renal disease requiring renal replacement therapy.
**Conclusions:** Children with sickle cell disease should be screened for renal complications especially from the late first decade of life. This will help in the early detection of renal disorder that could lead to chronic kidney disease. It is also suspected that the severe forms of Sickle cell nephropathy such as nephrotic syndrome may have a predilection for the female gender. A more extensive study is needed to test the veracity of this observation.

## Abstract# 330

**The Pattern of Kidney Disease among HIV Positive Children Admitted in a Tertiary Hospital in South Western Nigeria** A.D. Ademola,[1] A. Asinobi,[2] R. Oladokun.[2] *[1]Department of Paediatrics, University College Hospital, Ibadan, Oyo State, Nigeria; [2]Department of Paediatrics, College of Medicine, University of Ibadan, Ibadan, Oyo State, Nigeria; [3]Department of Pathology, College of Medicine, University of Ibadan, Ibadan, Nigeria.*
**Objectives:** To describe the pattern of kidney disease among HIV positive children admitted to a tertiary hospital in Nigeria.
**Methods:** We reviewed data on all children who were referred to the Pediatric Nephrology unit of the University College Hospital Ibadan for HIV infection and kidney disease from March 2004 to December 2009.
**Results:** Nine children, five males (55.6%) and 4 females, aged 4-15 years (mean 10.7 ± 3.3 years) were admitted. In three patients (33.9%) kidney disease was the initial manifestation of HIV.
Five children (55.6%) were in renal failure. Three (33.9%) were in acute renal failure while two (22.2%) patients were in chronic renal failure. Two patients had nephrotic syndrome. The eight patient had Kaposi sarcoma, hypoalbuminaemia and mild proteinuria, while the ninth patient developed left renal artery stenosis. Renal biopsies performed in three patients, post-mortem in one, showed focal segmental glomerulosclerosis in two patients, membranous nephropathy the third. Management included use of highly active anti retroviral drugs, ACE inhibitors, other antihypertensives and diuretics, three patients underwent haemodialysis. Mortality was 33.3%.
**Conclusions:** HIV is associated with heterogenous manifestations of kidney disease in our environment with renal failure as the predominant form.

## Abstract# 331

**Atypical Hemolytic Uremic Syndrome (aHUS). A Single Center Experience** M.S. Adragna, A.A. Balestracci, M.G. Caletti. *Nephrology, Hospital de Pediatría Prof.Dr.Juan P. Garrahan, Buenos Aires, Argentina.*
**Objectives:** aHUS is an uncommon disease.Sporadic or familiar,affects children of all ages.Main features:severe hypertension,chronic renal failure (CRF):50%,mortality:10%.Deficiency in complement regulatory proteins(factors H,I and B),MCP,ADMTS 13 or antibodies against factor H are main causes. Plasma therapy is an effective treatment, even though there is no agreement in their use regime. The risk of recurrence in kidney transplant is higher in factor H and I.
To describe the clinical features and outcome of aHUS patients(p) in our Institution.
**Methods:** Retrospective age, gender, familiar history, age at the onset, hypertension, low C3, treatment, follow-up and outcome were recordered. Continued variables expressed as median and categoric as percentages.
**Results:** 11p.:7 male.Two pairs of sibblings and 2 cousins.Median age onset: 0.66 m(0.33-6.8).Hypertension: 9p(82%),low C3:3/9p. and relapse: 8p(73%). Treatment: 7p:plasma infusions.10 needed acute dialysis. Median time follow-up: 9.5y(4.1-15).Outcome: 4 p.normal renal function, 7 developed CRF: 2 in predialysis medical treatment and 5: ESRD. These 5 p. undergo cadaveric kidney transplants: 2 recurrenced with graft lost (one of them received 2nd successful one with plasma exchange prophylaxis), 1 had no recurrence (died due to no renal reason) and two (the cousins) received successful transplants with serial plasma exchange and infusions.
**Conclusions:** aHUS was present in younger children, with more hypertension and higher requirement of dialysis. Outcome was unfavourable, with a high rate of recurrence and ESRD. Our results force us to enhance the efforts to identify the mutations in order to select the best treatment for each type of patient.

## Abstract# 332

**Significance of Protein to Creatinine Ratio Determination in the Initial Work-Up for Pediatric Proteinuria** O.M. Akchurin,[1] J.E. Springate.[2] *[1]SUNY at Buffalo, Buffalo, NY, United States; [2]State University of New York at Buffalo School of Medicine and Biomedical Sciences.*
**Objectives:** To estimate the frequency of urine protein to creatinine ratio testing in children found to have proteinuria on their routine dipstick urinalyses by primary care providers before these patients were referred to a pediatric nephrologist. To assess the impact of protein to creatinine ratio on further diagnostic work-up and outcomes of these patients.
**Methods:** We retrospectively reviewed medical records of patients referred to our pediatric nephrology clinic over the past 4 years. A total of 151 patients were referred with the diagnosis of proteinuria. The ultimate outcomes were analyzed separately for patients with and without urine protein to creatinine ratios ordered by their primary care providers.
**Results:** Of all analysed patients, urine protein to creatinine ratio was tested by primary care providers in 31.2% of cases. Overall, glomerular disease was identified in 8.5% of all referred patients and 4% went on to have a renal biopsy done. 31.3% of patients on their initial visit to nephrologist were found not to have any sufficient reason to be followed in renal clinic based on clinical and laboratory investigation. Among patients with protein to creatinine ratio done by their primary care providers, only 10.0% were not recommended to have follow-up with nephrology, and for those who had not had this study done, 40.9% were cleared after their initial visit ($p<0.05$). Glomerular pathology was found more often in patients referred with a urine protein to creatinine ratio already having been performed (18.1% vs 4.5%).
**Conclusions:** Determining urine protein to creatinine ratio in primary care settings can help practitioners to avoid unnecessary referrals to pediatric nephrologists and thus should be done more universally.

## Abstract# 333

**Ocular Abnormalities in Childhood Chronic Renal Failure** A.J. Al Mosawi. *Pediatrics, University Hospital in Al Kadhimiyia, Baghdad, Iraq.*
**Objectives:** Few literatures reported the incidence of ocular abnormalities in chronic renal failure (CRF).The aim of this paper is to determine the incidence of ocular abnormalities in childhood CRF.
**Methods:** From January 1993 to July 2007, 80 patients with a diagnosis of chronic renal failure (CRF) were observed at the University Hospital in Al Kadhimiyia. They were examined to determine the presence of ocular abnormalities. Fifty one patients were males (63.75%) and 29 (36.25%) were females. The male-female ratio was 1.75, and the age at referral ranged from 2 months to 18 years (mean 9 years).
**Results:** Corneal cystine crystals were the most common ocular abnormalities associated with childhood CRF occurring in 7.5% of the patients in association with Nephropathic cystinosis. Congenital cataract & glaucoma were observed in 3.75% of the patients in association with Oculo-cerebro-renal syndrome (OCRS).

Ocular abnormalities in childhood chronic renal failure

| Ocular abnormality | n (%) | Disorder |
|---|---|---|
| Corneal cystine crystals | 6 (7.5%) | Nephropathic cystinosis |
| Congenital cataract & glaucoma | 3 (3.75%) | Oculo-cerebro-renal syndrome |
| Congenital cataracts & chorioretinal hypoplasia | 1 | Oculo-cerebro-renal syndrome |
| Acquired cataracts | 1 | Hypocalcaemia |
| Retinopathy | 2 | Hypertensive retinopathy |
| Bilateral optic atrophy | 1 | Familial nephropathy associated with club feet |
| Proptosis | 1 | Membranoproliferative Glomerulonephritis |
| Total | 15 (18.75%) | |

**Conclusions:** Ocular abnormalities are relatively common in childhood CRF occurring in approximately 19%.

## Abstract# 334

**The Current Situation of Pediatric Nephrology in Iraq** A.J. Al Mosawi. *Iraqi Society of Pediatric Nephrology, Baghdad, Iraq.*
**Objectives:** The aim of this paper is to describe the current situation of pediatric nephrology (PN) in Iraq and the pattern of childhood renal disorders.
**Methods:** Review the available information available from the most important pediatric nephrology referral center in Iraq and the Iraqi society of pediatric nephrology which is currently located in Baghdad with representives in the north in Mosul and in the south in Basra.
**Results:** PN is not established medical subspecialty in Iraq, and the proper infrastructure and services of a PN unit offering renal diagnostic and therapeutic facilities do not exist. The main form of renal replacement therapy (RRT) available is acute, intermittent peritoneal dialysis. Hemodialysis is available only for children whose weight > 30 kg. Chronic peritoneal dialysis is unavailable for any patients. Renal transplantation (RT) has been performed in only a few older children, with limited success. Acute glomerulonephritis was relatively common during the 1990s, but only a very few cases have been observed during this decade. Idiopathic nephrotic syndrome (INS) remains a common problem. Although the exact incidence of INS is unknown, the clinical features and response to therapy are similar to those observed in North America and Europe. Renal Tubular disorders: The three most common renal tubular disorders are idiopathic hypercalciuria (37%), cystinosis (21%), and RTA (21%). Chronic renal failure: The single most common cause of CRF was chronic glomerulonephritis (GN), accounting for 19%.
**Conclusions:** The pattern for childhood renal disorder differs from patterns in other areas of the world. Optimal use of the available resources and flexibility in the development of a PN health care program in Iraq are essential.

## Abstract# 335

**A New Model for the Management of End-Stage Renal Disease** A.J. Al Mosawi. *Pediatrics, University Hospital in Al Kadhiiyia, Baghdad, Iraq.*

**Objectives:** The aim of this paper is to describe a new model for the management of end-stage renal disease (ESRD) in the developing world and its potential future role in the industrial countries when advancement in stem cell researches makes the need for long-term dialysis obsolete.

**Methods:** During the period from January 1993 to July 2007, 80 patients with a diagnosis of chronic renal failure (CRF) were observed at the University Hospital in Al Kadhimiyia. Fifty one patients were males (63.75%) and 29 (36.25%) were females. The male-female ratio was 1.75, and the age at referral ranged from 2 months to 18 years (mean 9 years). Fourteen (16.5%) patients were treated a new therapeutic approach consisting of acacia gum supplementation plus the traditional conservative measures was used.

**Results:** Amelioration of the uremic symptoms and lowering of blood urea levels delaying the need for dialysis was associated with this new therapy. In this sample of 80 patients the longest survival of 6 years was achieved in 2 patients, both treated initially with IPD. One of them was transplanted and the other was treated with the new therapeutic approach.

**Conclusions:** It's a fact that with appropriate dietary and pharmacologic management, patients with non-terminal CRF can be maintained surprisingly well and the transition from non-terminal CRF to ESRD represents a small decrement of renal function resulting in a large physiologic hurdle for the patient. The addition to these effective traditional measures, an agent enhance fecal nitrogen excretion can possibly bridge this gap resulting from this small decrement of renal function obviating the need for dialysis for some period of time.

## Abstract# 336

**Serum Creatinine as Prognostic Marker in Patients with Hemolytic Uremic Syndrome Who Did Not Require Dialysis** L.F. Alconcher,[1] A.P. Spizzirri,[2] C.J. Cobeñas,[2] R.C. Rahman.[2] *[1]Pediatric Nephrology Unit, Hospital Interzonal General de Agudos Dr José Penna, Bahía Blanca, Buenos Aires, Argentina; [2]Pediatric Nephrology, Hospital de Niños Sor María Ludovica, La Plata, Buenos Aires, Argentina.*

**Objectives:** To asses if serum creatinine (Cr) values during the acute stage of Hemolytic Uremic Syndrome (HUS) could be a useful prognostic marker in the long term follow-up.

**Methods:** Retrospective analysis. Inclusion criteria: patients with postdiarrheal HUS who did not require dialysis with 2 Cr values during acute stage and followed for at least 5 years. The relationship between the peak Cr value (highest value of 2 determinations during acute stage) and outcome at the last follow-up was evaluated. One hundred and forty patients were included and divided into 2 groups: patients with Cr values ≥1.5 mg/dl (63 patients, 45 %) and with values <1.5 mg/dl (77 patients, 55%). At the last visit patients were classified into 4 groups (G): GI, complete recovery; GII had two subgroups: Iia, microalbuminuria, and Iib, proteinuria and/or high blood pressure, both with normal renal function; GIII, chronic renal failure and GIV, end-stage renal disease. Mean follow-up was 8 years 11 months (r: 5-30). Chi square test was applied, a $p < 0.05$ was considered statistically significant.

**Results:** Fourteen out of 63 patients (22 %) with Cr values ≥1.5 mg/dl progressed into groups IIb and III vs. 6 out of 77 patients (8 %) with lower values (p= 0.01).

**Conclusions:** Severity of renal disease and long-term prognosis is not the same in all HUS patients not requiring dialysis during acute stage. Patients with Cr values ≥1.5 mg/dl were more likely to progress into the groups that may develop progressive renal injury.

## Abstract# 337

**Evaluation of 38 Patients with Nutcracker Syndrome** F.S. Altugan,[1] M. Ekim,[1] K. Köse,[2] Z.C. Özcakar,[1] F. Yalcinkaya,[1] A. Kavaz,[1] S. Fitöz.[3] *[1]Department of Pediatric Nephrology, Ankara University School of Medicine, Ankara, Turkey; [2]Department of Biostatistics, Ankara University School of Medicine, Ankara, Turkey; [3]Department of Radiology, Ankara University School of Medicine, Ankara, Turkey.*

**Objectives:** Nutcracker syndrome, presents with gross/microscopic hematuria or orthostatic proteinuria, results from compression of left renal vein between the SMA and the aorta. Doppler sonography is useful in the diagnosis of Nutcracker syndrome. We retrospectively analyzed 38 Nutcracker syndrome patients.

**Methods:** Clinical data from 38 patients with Nutcracker syndrome were reviewed. The diagnosis of the Nutcracker syndrome were done with renal vein Doppler ultrasound.

**Results:** There were 14 male, 24 female patients. The mean age was 11.45±2.8 years. Symptoms included abdominal pain (n = 20), hematuria (n = 4), urinary tract infection ( n=4 ) and others (n = 10). Among these patients 3 had urolithiasis, 11 had recurrent urinary tract infection and 5 had Familial Mediterranean Fever in addition to Nutcracker syndrome. Varicocele was detected in 4 patients. Macroscopic hematuria was seen in four, microscopic hematuria in five and proteinuria in 29 patients. The median body mass index (BMI) and proteinuria at the beginning were 16.5 (range, 13-24) and 8.2 mg/$m^2$/h (range 4-91). The histopathological findings of two patients with FMF, one patient with urolithiasis who underwent biopsy were normal.

**Conclusions:** There was no correlation between BMI and proteinuria at the begining (r=0.153, p=0.359). After a median 26.5 months follow-up time, findings of the urinalysis are still the same. Again we did not find any correlation between changes in BMI and proteinuria (r=0.112,p=0.592).

## Abstract# 338

**Random Serum and Urine Sodium Levels among Children with Pneumonia: A Cross-Sectional Study** M.U. Lumandas, F.E. Anacleto. *Pediatrics, College of Medicine-Philippine General Hospital, University of the Philippine Manila, Manila, Philippines.*

**Objectives:** Intravenous maintenance fluid is widely used in general pediatric practice and more children who come into the hospital receive intravenous fluid than in the past. Recent studies show the potential harm in using hypotonic solutions in children especially in diseased states like pneumonia. The choice of parenteral fluids is usually dictated by the serum sodium and urine sodium. The objective is to determine the random serum sodium and urine sodium levels among children with pneumonia.

**Methods:** Serum sodium, BUN and creatinine concentrations were obtained upon the patients' admission and blood sampling took place before starting any intravenous fluid. Urine samples were also sent within 24 hours upon admission to the Philippine General Hospital laboratory for random urine sodium and creatinine levels. Fractional excrtetion of sodium will be computed.

**Results:** Infants were found to be more affected with pneumonia with males predominating(73%). 60% of the subjects presented with normal serum sodium. The mean serum sodium was 142 mmol/L (SD 5.29 mmol/L) For the urine sodium, 70% had normal values. Mean urine sodium was 90.77 mmol/L (SD 69.51 mmol/L). 57% had low FeNa. Using the Fisher's exact test, the risk for abnormal serum sodium and urine sodium was higher in children than infants.

**Conclusions:** Children with pneumonia have normal serum and urine sodium. Near-isotonic or isotonic fluid is a more physiologic parenteral fluid to use in children with pneumonia.

## Abstract# 339

**Effect of Isotonic Versus Hypotonic Parenteral Fluids on the Serum Sodium Levels of Children Admitted to the Philippine General Hospital: An Open, Randomized, Controlled Trial (A Preliminary Study)** V.C. Ang, F.E. Anacleto. *Pediatrics, College of Medicine-Philippine General Hospital, University of the Philippines Manila, Manila, Philippines.*

**Objectives:** The most widely used parenteral fluid in patients admitted to the hospital is a dextrose-containing hypotonic maintenance fluid. However, this practice has recently been called into question. The objective is to determine whether there will be lower mean serum sodium levels in patients who receive a hypotonic compared with isotonic parenteral fluid.

**Methods:** In an open, randomized controlled trial, patients age 1-15 admitted to the emergency room were placed on nothing per orem and given intravenous fluids for at least 24 hours. Subjects were to receive either D5NM or D5NR. The fluid rate was determined by the attending physician. The serum sodium 24 hours after initiation of fluids was measured. A p-value < 0.05 using the Student's t-test was considered statistically significant.

**Results:** Nineteen patients (11 in the isotonic group and 8 in the hypotonic group) were included. There was no statistical difference in baseline characteristics. Baseline serum sodium was 140.3±9.1 mmol/L in the hypotonic group and 139.9±4.3 in the isotonic group (p=0.43). After 24 hours of intravenous fluid therapy, mean serum sodium was 138.7 ± 9.5 for the hypotonic group and 139.0 ± 3.0 for the isotonic group (p=0.44). One patient in the hypotonic group developed asymptomatic hyponatremia with serum sodium < 128.

**Conclusions:** This preliminary study shows no significant difference in serum sodium 24 hours after initiation of either isotonic or hypotonic fluids. A follow-up study with a larger sample size is recommended.

## Abstract# 340

**Early Age at Debut Is a Predictor of Steroid Dependent and Frequent Relapsing Nephrotic Syndrome** R.F. Andersen,[1] N. Thrane,[2] B. Jespersen,[3] S. Rittig.[1] *[1]Department of Paediatrics, Aarhus University Hospital, Skejby, Aarhus, Denmark; [2]Department of Paediatrics, Regional Hospital Herning, Herning, Denmark; [3]Department of Nephrology, Aarhus University Hospital, Skejby, Aarhus, Denmark.*

**Objectives:** The aim was to identify characteristics of patients with steroid sensitive nephrotic syndrome (SSNS) that point to a high risk of frequent relapsing (FR) or steroid dependent (SD) SSNS.

**Methods:** A retrospective analysis of 54 consecutive patients with SSNS was performed. The incidence was 1.9/100.000 and follow-up 4.0 years. Two different steroid regimens were used and a subanalysis was performed in the group treated with the long steroid course (pred-long) (prednisone 60 mg/m$^2$/24h for 6 weeks and 6 weeks prednisone 40 mg/m$^2$/48h). The short steroid treatment course (pred-short) was 4 weeks with prednisone 60 mg/m$^2$/24h and 4 weeks prednisone 40 mg/m$^2$/48h.
**Results:** In total, 30/54 of the patients were classified with FR/SD. FR/SD patients were younger at debut compared to non-FR/SD patients (3.5 years vs. 8.5 years, $p < 0.002$). Males were overrepresented in the FR/SD group (69% vs. 38%, $p = 0.03$). No differences were found regarding hematuria, hypoalbuminemia, or days to achieve remission. 31 patients were treated with pred-long compared to pred-short in 23 patients reducing the overall number of FR/SD patients from 80% to 38% ($p<0.006$). In the pred-long group, 12 patients were FR/SD and these were younger vs. 19 non FR/SD patients (4.4±3.1 years vs 8.4±4.1 years, $p<0.005$).
**Conclusions:** Low age at debut and male gender is associated with a high risk of FR/SD in this unselected series of patients. This tendency seems consistent despite the prolongation of the steroid course at the debut of SSNS.

## Abstract# 341

**Epidemiology of Renal Failure in Children in Brazil** M.C. Andrade,[1] C. Mangia, J.T. Carvalhaes. *[1]Pediatrics, UNIFESP-EPM, São Paulo, Sao Paulo, Brazil; [2]Pediatrics, UNIFESP-EPM, São Paulo, Sao Paulo, Brazil; [3]Pediatrics, UNIFESP-EPM, São Paulo, Sao Paulo, Brazil.*
**Objectives:** The aim of this study is to evaluate the incidence, costs and outcome of children's renal failure in Brazil.
**Methods:** Observational cohort study based in Brazilian national govern database(SUS). We analyzedall children from neonatal period to 19 years old with diagnosis of renal failure based on the 10th revision of the International Classification of Diseases (ICD-10).
**Results:** Demographic data were collected in eight cohorts from 1998 to 2007. There were 666,725 hospital admissions by kidney disease and 43,174 admissions by renal failure. The mean of mortality rate, 4% in these cohorts. The admissions and mortality rate have been constant with high mortality under the age of 1 year since 1998. The mean of annual costs was $5,075,641 millions and $588 dollars per patient.
**Conclusions:** Renal failure is a prevalent health problem that affects children from neonatal period to adolescence and presents high post natal mortality. Renal failure is associated to a large spectrum of different ethiologies and different levels of morbidity and consequent impact on the outcome.

## Abstract# 342

**5 Year Review of Haemolytic Uraemic Syndrome in Ireland** K. Bruton,[1] M. O'Grady,[2] K. Burns,[3] R. Cunney,[3] M. Waldron,[2] M. Riordan,[12] A. Awan.[1] *[1]Pediatric Nephrology, Children's University Hospital, Dublin, Ireland; [2]Our Lady's Children's Hospital, Crumlin, Dublin, Ireland; [3]Microbiology, Children's University Hospital, Dublin, Ireland.*
**Objectives:** To identify patient characteristics, complications and longterm outcomes of children with both diarrhoea + and atypical HUS in our population.
**Methods:** Review of all children (<16 years) presenting with HUS between January 2005 and December 2009 to two tertiary paediatric nephrology centers serving the entire population of Ireland. Culture or PCR positive VTEC isolates were identified to confirm full case acquisition. Atypical HUS were confirmed by gene analysis or in pneumococcal cases by culture and / or PCR.
**Results:** Preliminary data identified 63 patients (aged 3 months to 14 years). 5 patients were atypical. 60 had diarrhoea at presentation (D+), 33 had bloody diarrhoea. VTEC producing e.coli were identified in 36 cases – O157 (n=26); O26 (n=9); O145 (n=1). 28 patients were anuric for an average of 8.8 days; 10 for >10 days. 32 children were dialysed – peritoneal dialysis was used in 26. 11 underwent plasma exchange (4 atypical cases). Complications included hypertension (25); encephalopathy (8); cardiomyopathy (1), hyperglycaemia (1) and pancreatitis (1). 52 patients recovered fully and 6 had long-term sequelae in the D+ HUS group. 5 patients had atypical HUS – 3 pneumococcal (1 died, 1 long-term sequelae); and 2 with Factor H mutations (1 has renal impairment).
**Conclusions:** 0157 producing e.coli remains the most common cause of D+ HUS. The mortality over 5 years for D+ HUS was zero in our study. 10% D+ patients have long-term renal sequelae. 1 patient with Atypical HUS died and 2 have longterm renal sequelae.

## Abstract# 343

**Pathological Pattern of Renal Diseases in Egyptian Children** A. Bakr,[1] A.M. El-Refaey,[1] A. Hammad,[1] A. Elmougy,[1] A.S. Adediran,[1] F. El-Hoseeny,[3] A. Abdelrahman,[2] A. Sarhan.[1] *[1]Pediatric Nephrology, Mansoura University Children's Hospital, Mansoura, Egypt; [2]Pediatric Radiology, Mansoura University Children's Hospital, Mansoura, Egypt; [3]Pediatric Pathology, Mansoura University Children's Hospital, Mansoura, Egypt.*
**Objectives:** To study the pathological pattern of renal diseases of Egyptian children in a tertiary hospital.
**Methods:** Ultrasound guided biopsies were done using a needle mounted on spring- loaded biopsy gun for 904 cases between 1996 and 2009.
**Results:** The main indications were Steroid Resistant NS (28.3%), SDNS/FRNS(15.1%), atypical NS(14.1%) and suspected Lupus Nephritis(13.3%). The pattern of patients was MCNS and its variants(32.8%), DMP(15.1%), FSGS(10.9%), MPGN(2.7%), and membranous nephropathy(0.7%). In lupus, the commonest lesion was class IV(5.5%), III(3.4%), II(3.2%) then class I(0.1%). Specimens were insufficient in 1.4% of them. Only 3 cases had gross haematuria and one patient died due to uncontrolled hemorrhage.
**Conclusions:** The pathological pattern of kidney diseases in our centre is similar to that of many countries.

## Abstract# 344

**Pronostic Factors of Good Growth in Hypophosphatemic Rickets** L. Bessenay,[1] B. Pereira,[2] E. Merlin,[2] D. Terral,[1] C. Auclair,[3] B. Ranchin,[4] P. Cochat,[4] C. Gay,[5] G. Bourdat-Michel,[6] J.-B. Palcoux.[1] *[1]Pediatry, Clermont Ferrand, France; [2]Cic, Clermont Ferrand, France; [3]Sante Publique, Clermont Ferrand, France; [4]Nephrology and Pediatry, Lyon, France; [5]Pediatry, Saint Etienne, France; [6]Pediatry, Grenoble, France.*
**Objectives:** This study reports on clinical and biological prognostic factors of a good growth in children with hypophosphatemic rickets.
**Methods:** It is based on a retrospective and multicenter collection of 38 children.
**Results:** The mean follow-up was 6.9 years. The patients were treated with 1-α-cholecalciferol and phosphorus supplementation. Twelve patients had nephrocalcinosis with an average time to onset of 117 months with a lower growth than the patients without ($\Delta Z$= -0.75 vs 0.21).
Seventeen patients had a poor growth (difference between final and initial Z score: $\Delta Z < 0$) and 21 had a good growth ($\Delta Z \geq 0$). There was no significant improvement in size over the entire group between the beginning and end of the study (-1.18 SDS vs -1.28 SDS). But patients with a size at diagnosis <-2 SDS tended to have a better final height. Sporadic cases had a significantly better growth than familial cases. Patients with a size ≤ - 2 SDS at 4 years (probably without the Hap1 variant of vitamine D receptor genotype) had a lower size at the end of study but a same growth. Factors predictive of a better $\Delta Z$ score are the 4 following: sporadic cases, initial Z score, female gender, and no nephrocalcinosis. Mean serum phosphate and treatment were not discriminating.
**Conclusions:** The prognostic factors of good growth seem more connected to the initial profile of patients and the spontaneous evolution of the disease than the replacement therapy.

## Abstract# 345

**Clinical Patterns of Steroid Resistant Nephrotic Syndrome (SRNS) in Children: A Single Center Study** R. Bianchi, H. Pinto, A. Teixeira, C. Tavares, A. Caldas Afonso, H. Jardim. *Pediatric Nephrology Unit, Hospital S. João, Porto, Portugal.*
**Objectives:** The aim of this study was to evaluate both epidemiological and clinical features of children with SRNS on current follow-up at the paediatric nephrology clinic of our hospital.
**Methods:** A retrospective analysis was performed on SRNS patients regarding clinical, histopathological, treatment and outcome data.
**Results:** Ten patients were studied (age: 2 -12 years). Follow-up varied between 8 months and 6 years. One patient presented with acute renal failure and 5 had microscopic haematuria. In all cases C3 and ANA had normal values. Eight cases presented as primary SRNS and two cases developed secondary resistance to steroids. Histopathological findings were compatible with FSGS in 4 cases, mesangioproliferative glomerulonephritis (Mp GN) in 5 and C1q nephropathy in one. The two more severe cases had an histology compatible with FSGS and maintain a nephrotic proteinuria despite consecutive treatment with cyclophosphamide, MMF and IECA. The two other cases of FSGS have urine protein-creatinine ratio < 0.3, one is being treated with MMF and the other with ciclosporine. In relation to the cases of Mp GN, two cases achieved total remission and the other three cases had partial remission: one completed triple Mendoza regimen and the other two cases are on MMF therapy. At present, serum creatinine is normal in all cases.

**Conclusions:** The diversity of response to treatment and outcome observed in SRNS, recommends that multicentric studies should be undertaken in order to better identify the potential efficacy, individual response, and side effects of long term therapy with the immunosuppressive drugs presently available.

## Abstract# 346

**Urinary Stone Disease in Turkish Children** S. Al, I. Bilge, S. Emre, A. Sucu, A. Sirin. *Pediatric Nephrology, Istanbul University Istanbul Medical Faculty, Istanbul, Turkey.*

**Objectives:** We aim to assess demographics and clinical data of children with a diagnosis of urinary stone disease.

**Methods:** Four hundred and three patients (218 male, 185 female) with urinary stone disease admitted between years 1999–2009 participated in our study. Their data were assessed retrospectively using patient charts.

**Results:** Of 403 patients (218 male, 185 female) % 91.3 had been residing in Marmara region. Mean presenting age was 52.8 months (1-192 months), presenting symptoms were 49.4% pain, 26.3% urinary tract infection, 22.5% passing out stone, 18.6% macroscopic hematuria.

Metabolic derangements were found in 51.6 % of cases. Structural anomalies were the cause in 21.3 %. Stone analysis were consistent with calcium stones in 60.5 % and metabolic derangements were hypercalciuria in 48.4%, hypocitraturia in 32.3%, hyperuricosuria in 20.1%, hyperoxaluria in 19.7% and cystinuria in 18.7%. All patients had been receiving conservative therapy including diet, increased fluid intake and salt restriction, 131 cases (32.5%) had been treated surgically, 205 cases (50.9%) had been taking medications for underlying metabolic derangements. Of patients receiving conservative therapy, 17.1% had shown no improvement, while 4.7% of patients had developed renal failure. Forty nine patients (12.1%) had recurring urinary stones.

**Conclusions:** Results of our study supports the importance of metabolic evaluation and structural investigations for pediatric urinary stone disease.

## Abstract# 347

**Papillary Stones with Randall's Plaques in Children: Clinicobiological Features and Outcome** K. Bouchireb,[1] C. Pietrement,[2] H. Nivet,[3] H. Martelli,[4] O. Dunand,[5] M. Daudon,[1] R. Salomon.[1] *[1]Necker Enfants Malades Hospital, Paris, France; [2]CHU, Reims, France; [3]CHU, Tours, France; [4]CHU, Kremlin Bicètre, France; [5]CHD, Saint-Denis, French Southern Territories.*

**Objectives:** Randall observed calcium phosphate plaques beneath the renal papillae epithelium that are supposed to be the initial site of overgrowth of urinary calcium stones. The aim of this study was to review the clinical presentation, metabolic screening and management of children producing stones with Randall's plaque.

**Methods:** Among 1687 pediatric stones referred to our institution, 25 were developed from a Randall's plaque. The patient's demographic features, clinical histories, metabolic screening, radiologic investigations and management were all analyzed.

**Results:** Our series included 15 girls and 10 boys aged 18 years or less. Mean age at diagnosis was 13.5 years (5.3-17.5 yrs). Eleven patients had a family history of urolithiasis (44%). All patients presented with renal colic. Hypercalciuria and hyperoxaluria were identified in 5/21 investigated patients (23 %). Ultrasonography found a direct image of stone in 16 patients in kidneys (n=7) and ureters (n=8). Management with NSAI and anti-spasmodic drugs were sufficient in most cases (22/25), stones spontaneously passed within 72 hrs from hospital admission. Stone analysis revealed that calcium oxalate monohydrate (COM) was mostly found (n=23) with a plaque made of carbapatite.

**Conclusions:** Papillary stones in children occur essentially after 10 yrs of age, and the male predominance generally observed in childhood urolithiasis is not found. Stones are mainly composed of COM and medical treatment is sufficient in most cases (88%).

## Abstract# 348

**Pediatric Nephrologists, Chronic Kidney Disease and Tobacco Counseling: Identifying Practice Patterns** K. Bruner,[1] D. Chand,[2] H. Patel,[2] A. Stolfi,[1] A. Omoloja.[2] *[1]Pediatrics, Wright State University, Dayton, OH, United States; [2]Midwest Pediatric Nephrology Consortium, Columbus, OH, United States.*

**Objectives:** Recent studies have shown that cigarette smoking and second hand smoke (SHS) exposure are common in adolescents with chronic kidney disease. The high prevalence of risk factors such as hypertension coupled with use and exposure to tobacco greatly increases the risk for cardiac disease in CKD patients. There are no studies describing the practice patterns of pediatric nephrologists (PN) regarding tobacco counseling.

**Methods:** Three e-mail invitations to participate in a web-based survey were sent in two week intervals to a total of 330 PN in the United States. The survey consisted of twelve questions.

**Results:** Response rate was 25%. Less than half (38%) of respondents directly question adolescents about smoking on a routine basis and only 26.6% ask about exposure to SHS routinely. While the majority were confident about being able to provide guidance to patients on the harmful effects of smoking, 75% were not confident about enrolling or referring patients to a quit line for smoking cessation. Over half (74%) were not confident in prescribing patients tobacco replacement products. Barriers identified were time limitations (81%), lack of professional training (60%) and unfamiliarity with CPT reimbursement codes (57%).

**Conclusions:** Tobacco counseling of children with CKD by pediatric nephrologists is suboptimal due to several barriers. Adequate training of the entire pediatric nephrology team about tobacco counseling is important in attepmts to reduce morbidity and mortality associated with cardiac disease in children with CKD.

## Abstract# 349

**Relation of Voiding Pattern to Symptoms of Urinary Tract Infection (UTI) and Defecation Habits in Girls** M. Chandra. *Dept of Pediatrics, Winthrop University Hospital & Stony Brook Univ School of Medicine, Mineola, NY, United States.*

**Objectives:** Girls with UTI present with a variety of symptoms.This study evaluated the relationship of the patients' usual voiding pattern with their defecation habits and symptoms of UTI.

**Methods:** All 779 girls ages 3-15yrs, referred to the author's nephrology service over the last 22 yrs with history of first documented UTI after age 3 yrs, had prospective data collection and data registry of symptoms of UTI and their usual defecation and voiding pattern.Of them 11 were excluded from analysis because of anatomic or neurologic causes of UTI. Girls were divided into 3 groups: 1.Normal Voiding Pattern (NVP) 2.Overactive Bladder (OAB) i.e. urinary(ur) frequency ≤every 2 hrs, ur urgency, urge incontinence or urine witholding maneuvers to postpone voiding ≥5 times/week and 3. Infrequent Voiding (InfV) i.e. voiding ≤3times /24 hours.

**Results:**

table1

| Symptoms of UTI & defecation habits | OAB | InfV | NVP |
|---|---|---|---|
| N,768 | 600 | 111 | 57 |
| New onset/worsening of ur frequency,urgency,urge incontinence | 48% | 2% | 7% |
| Dysuria | 25% | 55% | 50% |
| Back pain/ lower abdominal pain | 15% | 33% | 49% |
| Fever with chills | 9% | 3% | 19% |
| Abnormal random urinalysis(positive nitrite,bacteria /leukesterase) | 2% | 14% | 2% |
| Stool witholding/leakage >4/month (SW) | 33% | 0 | 4% |
| Infrequent bowel movements <q3days( Infbm) | 11% | 45% | 38% |

**Conclusions:** Symptoms of urinary frequency, urgency and urge incontinence are uncommon with UTI in girls with InfV and NVP ; such symptoms should raise the suspicion for an underlying OAB. The frequent association of Infbm with InfV reflects co-existence of underactive bladder and colon. In contrast, the frequent association of SW with OAB reflects co-existence of overactive bladder and overactive rectosigmoid.

## Abstract# 350

**Is Calcium Oxalate Crystalluria (CaOxC) a Risk Factor for Nephrolithiasis (NL)?** M. Chandra, O. Grossman. *Dept of Pediatrics, Winthrop University Hospital & Stony Brook Univ School of Medicine, Mineola, NY, United States.*

**Objectives:** Calcium oxalate (CaOx) is the most common mineral constituent of renal stones. This study evaluated if calcium oxalate crystalluria is a risk factor for NL.

**Methods:** The charts of 305 patients (pts), ages 0.5-22 years who had microscopic examination of fresh void in the Pediatric Nephrology Clinic over a 19-month period were reviewed for presence of CaOxC, documented passage or imaging evidence of NL and historical risk factors for NL. Of these, 41 pts (19 males) had CaOxC in one or more urines, 23 (13 males) had NL and 12 had both. The frequency of risk factors for NL was compared in pts with and without CaOxC, and pts with & without NL.The latter 2 groups served as community controls for NL risk factors. The relative risk for NL in these 4 groups was analyzed with Chi-square analysis.

**Results:** In our referral population, 29% of pts with CaOxC had NL vs 4% of pts without CaOxC. Conversely, 52% of pts with NL had CaOxC vs 10% of pts without NL. The median age of pts with CaOxC and NL was 14 and 13 yrs respectively and their median urine SG 1.025. The frequency of the following 9 risk factors for NL was significantly higher ($p<0.001$) in pts with CaOxC vs pts without CaoxC : gross hematuria, microhematuria, flank pain, family h/o NL, hypocitraturia, hypercalciuria, supersaturation of urine with CaOx, calcium phosphate or uric acid ($p<0.05$ for uric acid). The frequency of these risk factors and the relative risk of NL from these risk factors was similar in patients with CaOxC and documented NL.

**Conclusions:** Presence of CaOxC in a freshly voided urine is a risk factor for NL since it is a visual evidence of urine supersaturation with CaOx.

## Abstract# 351

**Thrombotic Microangiopathy (TMA) in Childhood Lupus Nephritis: A Clinicopathological and Outcome Study** S.M. Chao, P.H. Tan. *Renal Service, KK Women's & Children's Hospital, Singapore, Singapore; Pathology Dept., Singapore General Hospital, Singapore, Singapore.*

**Objectives:** TMA is rarely reported in childhood SLE. We studied the clinicopathological features of TMA in childhood lupus nephritis and examined how the clinical phenotypes may influence the clinical course and outcome.

**Methods:** We reviewed retrospectively renal biopsy reports of SLE patients done over 12 years. The clinicopathological features and clinical course of those with histological diagnosis of TMA were studied.

**Results:** 7(11%) of 62 renal biopsies showed TMA, 4 patients had pentad of Thrombotic Thrombocytopenic Purpura (TTP), 2 severe SLE flare and 1 antiphospholipid syndrome (APLS). Mean age of onset of SLE was 11.6 years with a female predominance (6:1). TMA lagged behind onset of SLE in all patients except 1 being concurrent. TTP patients had marked neurological and renal impairment, severe anemia and thrombocytopenia. CTscans brain were normal except for 2 MRI findings of Posterior Reversible Encephalopathy. Renal histology was class IV or V nephritis with crescents seen mainly in TTP. TTP patients had stormy clinical course and 2 patients required CVVHD. All 7 patients had IV methylprednisolone and cyclophosphamide pulsing with or without mycophenolate mofetil. Of the 4 TTP patients, 2 underwent plasmaphoresis (PE) with success in one, 2 did not require PE. No mortality was noted. All patients made full recovery.

**Conclusions:** TMA in SLE is associated with TTP, lupus flare and APLS. TTP had the most severe clinicopathological features and most protracted course. PE may be ineffective in treating TTP. Intensive immunosuppression remains the mainstay of treatment in lupus patients with TMA, irrespective of their clinical phenotypes.

## Abstract# 352

**Hemorrhagic Colitis (HC) in Postdiarrheal Hemolytic Uremic Sindrome** C.J. Cobeñas, R.C. Rahman, O.R. Amoreo, A.d.D. Suarez, J.D. Ruscasso, A.P. Spizzirri, J.H. Zalba. *Nephrology, Hospital de Niños "Sup. Sor María Ludovica", La Plata, Buenos Aires, Argentina.*

**Objectives:** 1) To describe the clinical findings of HUS patients with HC; 2) To establish a relationship with renal and CNS involvement; 3) To evaluate mortality rate.

**Methods:** We analysed age, gender, clinical, surgical and histologic findings, renal and CNS disease.

**Results:** We evaluated 54 patients, 28 (51.8 %) boys. Mean age was 36.7 months, 51.8 % were older than 2 years. Clinical findings: abdominal mass in 6 (11.1 %), hematochezia in 24 (44.4 %), abdominal distention in 50 (92.6 %) and abdominal pain in 52 (96.3 %). Thirty five (64.8 %) required surgery and 17 resection. Affected sites: transverse colon 24 (57.1 %), ascending 21 (50 %), descending 17 (40.4 %), distal ileum 11 (26.1 %), sigmoid in 8 (19 %) and rectum in 1 (2.3 %). Macroscopy: hyperemia in 19 (45.2 %), necrosis in 18 (42.8 %), perforation in 12 (28.5 %). Histologic evaluation was available in 29 (53.7 %): 25 (86.2 %) had necrosis of the affected site (transmural in 21). Seven out of 9 evaluated by necropsy (77.7 %) had transmural necrosis. Mean hematocrit was 29.7 % and leukocyte count 26,137/mm3. Eighteen patients (33.3 %) died, all on acute renal failure (ARF). Anuria in the remaining 36 lasted for x: 13.9 days. CNS disease was severe in 38 (70.3 %): 24 had 3 or more seizures, 29 coma, and 19 respiratory assistance.

**Conclusions:** 1) Over half were older than 2 years. Almost 2/3 required surgery. The most affected sites were transverse and ascending colon. The main histologic finding was transmural necrosis. Higher hematocrit and leukocytosis were prominent features. 2) Most patients had severe renal and CNS disease. 3) Mortality rate was high.

## Abstract# 353

**A Case Report of a Teenager with Relative Left Renal Vein Stenosis, Macroscopic Hematuria, Isolated Night Hypertension** M. Cuk,[1] B. Valent-Moric,[1] S. Schmidt,[2] J. Delmiš.[3] *[1]Pediatrics, University Hospital "Sestre Milosrdnice", Zagreb, Croatia; [2]Radiology, University Hospital "Sestre Milosrdnice", Zagreb, Croatia; [3]Pediatrics, Chidren's Hospital Zagreb, Zagreb, Croatia.*

**Objectives:** Macroscopic hematuria may originate from glomeruli, renal tubules, or urinary tract. Here we report a case of macrohematuria probably due to relative renal vein stenosis.

**Methods:** Male 17,5 y born after unremarkable pregnancy, family history negative to renal diseases or hematuria. At the age of 3 was hospitalized because of acute pyelonephritis. Ultrasonography, voiding cystoureterogram and scintigraphy were unremarkable. At the age of 14 he was referred to us because of mycroscopic hematuria of glomerular origine and proteinuria within normal range. Infection, coagulopathy, hypercalciura were excluded, ANCA screening negative C3, C4 within normal range. The audiometry was unremarkable as well as lens examination. The hematuria was intermittent, independent of physical exercise. At the age of 18 he presented with macroscopic hematuria (glomerular), proteinuria within normal range, and flank pain. Ultrasonography was unremarkable. MSCT angiography excluded Nutcracker sy. Transversal view showed left renal vein tension over the front side of the aorta, left renal vein appeared narrower, and the part right to aorta was invisible. Ambulatory blood pressure monitoring /ABPM/ showed isolated night systolic-diastolic hypertension.

Renin in horizontal was higher than in vertical position, noradrenaline, adrenaline,VMA, aldosterone, aldosterone-renine ratio, ACTH, cortisol, TSH, T3 were unremarkable.

Therapy with enalapril was introduced with bp improvement.

## Abstract# 354

**Familial Mediterranean Fever and Henoch – Schönlein Purpura: Similar Symptoms but Different Diagnosis** C.S. Dogan,[1] E. Comak,[1] M. Koyun,[1] A.U. Gökceoglu,[1] I. Keser,[2] S. Akman.[1] *[1]Pediatric Nephrology, Akdeniz University, Antalya, Turkey; [2]Genetics, Akdeniz University, Antalya, Turkey.*

**Objectives:** The aim of this study was to evaluate the prevalance and significance of Familial Mediterranean Fever (FMF) in patients presenting with Henoch Schönlein purpura (HSP).

**Methods:** The patients who were diagnosed as HSP in the last six years were studied retrospectively. Mutation analysis of MEFV gene was studied in half of all patients.

**Results:** 70 children, 44 boys (62.8%), mean age at diagnosis of 8.1±3.4 years (2-15.6 y), were enrolled in the study. The symptoms at admission were skin lesions in 53.1%, gastrointestinal symptoms in 37.6% and symptoms related to joint involvement in 9.3% of the patients. All patients had skin lesions. Joint, gastrointestinal and renal involvement were determined in 58.6%, 54.3% and 28.6% of the patients, respectively. Renal manifestations were observed as the initial symptom in 8 patients (40%), whereas it was within 1 month of admission in 9 (45%) and within 3 months of admission in 3 patients (15%). Median follow-up period in patients with renal involvement was 3 months. Renal biopsy was performed in 8 patients. No chronic renal failure was determined in any patient. Six heterozygote and four homozygote mutations in MEFV gene were identified in 35 patients, whom genetic analysis could be performed. Three of four homozygote mutations were M694V/M694V, one of whom had renal involvement with nephritic–nephrotic syndrome and also gastrointestinal involvement; other patients had no symptoms related to FMF.

**Conclusions:** Children with HSP may display genotypes of FMF; so such children should be considered as occult FMF and treated with colchicine.

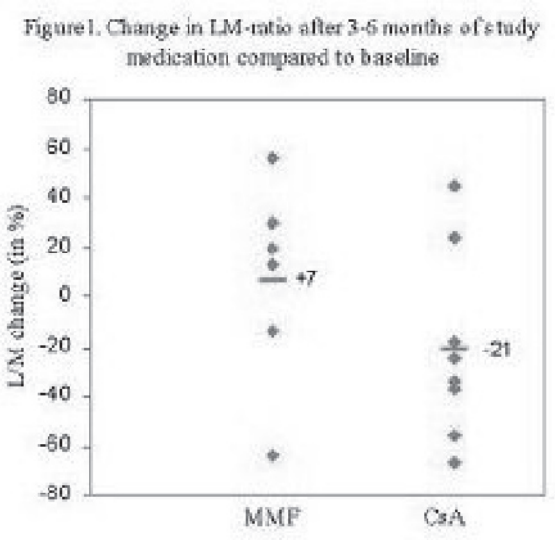
## Abstract# 355

**Intestinal Permeability in Patients Treated with Mycophenolate Mofetil (MMF) for Nephrotic Syndrome (NS)** E. Dorresteijn,[1] R. van Elburg,[2] J. Nauta,[1] B. van der Heijden.[1] *[1]Erasmusmc-Sophia, Rotterdam, Netherlands; [2]VU University Medical Centre, Amsterdam, Netherlands.*

**Objectives:** Chronic diarrhea is the most reported side-effect of MMF. We investigated whether the intestinal permeability, as tested by the sugar absorption test (SAT), is different in children treated with MMF for NS, in comparison to ciclosporine A (CsA).

**Methods:** In a multi-centre trial, children with frequently relapsing minimal change NS were randomized for treatment with MMF 1200 mg/m2/day or CsA 4-5 mg/kg/day. Before start and 3-6 months after start of the study medication, a SAT was performed.

**Results:** Of 15/24 children, two samples were available for analysis (6 MMF, 9 CsA). At start of the study, there was no difference in urinary lactulose-mannitol (LM) ratio between children being treated with CsA or without and all LM ratios at baseline were normal. At the second SAT, again all urinary LM ratios were within normal limits. The change in LM ratio compared to baseline was not different between the 2 groups (p=0.16).




Only one patient with MMF reported temporarily diarrhea. Mean weight gain during the study period of 12 months, -0.03 kg (-0.1%) in the MMF group versus +1.20 kg (3.3%) in the CsA group, was not significantly different (p=0.42).
**Conclusions:** Daily treatment with MMF did not result in abnormal values or significant changes in the SAT and only mild diarrhea in 1/6 patients.

## Abstract# 356

**Ulcerative Colitis and Duodenal Ulcers in a Child with Cystinosis Receiving Cysteamine** E. Dorresteijn, L. de Ridder, K. Cransberg. *Erasmus MC-Sophia, Rotterdam, Netherlands.*
**Objectives:** Since the early 1990s cystinosis is treated with cysteamine to prevent accumulation of cystine. This therapy delays organ damage but may cause gastrointestinal side effects by hypersecretion of gastric acid. In laboratory rats it was found ulcerogenic.
**Methods:** We report two severe gastrointestinal complications in a transplanted cystinosis patient treated with cysteamine.
**Results:** A now 14-year-old girl with cystinosis received cysteamine since the age of 4 years. Doses ranged from 1.0 to 1.3 mg/m$^2$/day; cystine concentrations in leucocytes were below 0.6 nmol/mg protein. Nevertheless she developed ESRD and was transplanted at the age of 9 years. Immunosuppression included prednisolone, tacrolimus, and 1 year MMF. At the age of 14, she was diagnosed with acute severe ulcerative colitis, necessitating subtotal colectomy despite intravenous steroids. Histopathology showed ulcerative lesions without cystine crystals. Cysteamine concentrations were normal. After an episode of acute tubular necrosis she developed duodenal ulcers with massive hematemesis. These were treated with blood transfusions and intervention duodenoscopy. Cysteamine therapy was then interrupted to promote healing of the duodenal ulcers. She was discharged after 2 months with maintenance immunosuppressive therapy for the renal graft consisting of low dose steroids and tacrolimus, and with a half dose of cysteamine and omeprazole.
**Conclusions:** To our knowledge this is the first report of duodenal ulcers in a cystinosis patient with cysteamine therapy (and the second on IBD). Although physical stress, steroids and uremia could all have contributed to the formation of duodenal ulcers, a possible role of cysteamine can-not be excluded.

## Abstract# 357

**The Prevalence of Hypothyroidism in Nephrotic Syndrome** A.R.A. Dy, Z.L. Antonio, A.G. Marbella, M.B. Rosel, O.R. De Leon. *Department of Pediatric Nephrology, National Kidney and Transplant Institute, Quezon City, Metro Manila, Philippines.*
**Objectives:** We determined the prevalence of hypothyroidism in children with Nephrotic Syndrome. We also compared the thyroid function tests (thyroid stimulating hormone, free T3 and free T4) between patients with NS in remission and in relapse. We correlated these with serum albumin, creatinine, total cholesterol, urine protein creatinine ratio (UPCr) and proteinuria.
**Methods:** Children ages 1 to 16 years old diagnosed with NS at the Pediatric Nephrology out-patient department of a tertiary hospital for a period of 6 months were included in the study. Blood samples for thyroid function tests, serum albumin, total cholesterol, and serum creatinine were obtained. Urinalysis and UPCr were also taken.
**Results:** A total of 66 patients were included in the study, 42.42% of subjects were in remission and 57.58% were in relapse. The mean TSH, FT3 and FT4 were statistically significant between the two groups. The parameters taken revealed: albumin had a moderate negative correlation with TSH (r=-0.4152, p <0.001) and a strong positive correlation with FT3 (r=0.7226, p<0.001) and FT4 (r=0.5997, p<0.001); both total cholesterol and UPCr had a positive correlation with TSH and a negative correlation with FT3 and FT4; no significant correlation was observed with serum creatinine. The prevalence rate of hypothyroidism in children with nephrotic syndrome is 16.67%.
**Conclusions:** The prevalence rate of hypothyroidism in nephrotic syndrome is 16.67% in our local setting. This study provides evidence of the presence of hypothyroidism in children with nephrotic syndrome who have hypoalbuminemia, hypercholesterolemia and proteinuria.

## Abstract# 358

**Kidney Function of HIV-Infected Children in Nigeria Using Filler's Serum Cystatin C-Based Formula** C.I. Esezobor, E. Iroha, O.O. Oladipo, E. Onifade, O.O. Soriyan, A.O. Akinsulie, E.O. Temiye, C. Ezeaka. *Paediatrics, College of Medicine, University of Lagos, Lagos, Nigeria.*
**Objectives:** Comparison of kidney function of children with and without HIV infection using serum cystatin C and estimated GFR.
**Methods:** Cystatin C level was measured using ELISA technique while GFR was estimated using Filler's cystatin C formula. Advanced HIV disease was defined as WHO clinical stage 3, 4 disease, CD% <20 or count <350cells/ul. Student t test, Mann Whitney U test, Pearson chi square and Fisher's exact test were used where appropriate to test difference between groups.
**Results:** Compared to the controls the HIV-infected group had higher median (interquartile range) serum cystatin C level {0.77 (0.29) mg/l versus 0.66 (0.20) mg/l; p=0.025} and higher proportion of children with serum cystatin C level >1mg/l {10 (16.7%) versus one (1.7%); p=0.004}. The HIV-infected children had a mean (±SD) eGFR of 96.8 (±36.1) ml/min/1.73m2 compared with 110.5 (±27.8) ml/min/1.73m2 in the controls (p=0.021). After controlling for age, sex and BMI only the study group (HIV infected versus control) remained a significant predictor of serum cystatin C level (β=-0.216, p=0.021). The proportion of HIV-infected children with eGFR <60 ml/min/1.73m2 was 8 (13.3%) versus none (0%) in the control group (p=0.006). However, the serum cystatin C level, eGFR and proportions of children with cystatin C level >1mg/l and eGFR <60 ml/min/1.73m2 were not significantly different between the HIV-infected children with advanced disease and those with milder disease.
**Conclusions:** HIV-infected children in Nigeria have higher serum cystatin C level and lower eGFR compared to age and sex matched controls.

## Abstract# 359

**Number, Location and Size of the Stones in 153 Children with Urolithiasis** M.H. Fallahzadeh, V. Sedighi, M.K. Fallahzadeh, M.A. Fallahzadeh, A. Derakhshan. *Shiraz Nephrourology Research Center, Shiraz, Islamic Republic of Iran.*
**Objectives:** Knowledge of number, location and size of renal stones is very important in predicting underlying causes and management of urolithiasis. The information about these issues in pediatric field is scarce; therefore, we performed this study to evaluate these parameters in children with urolithiasis.
**Methods:** 153 patients (82 boys and 71 girls) ranging in age from 2 months to 18 years (mean age =4.84 years ±4.81) with documented renal stones were enrolled in this prospective cross-sectional study. The number, location and size of the stones were determined by ultrasonography in at least 2 sessions.
**Results:** 62 patients (40.5%) had one, 65 (42.5%) had two and 26 (17%) had three stones or more.The stones were bilateral in 56 patients (36.6%). Out of 244 stones, 127 were located on the right side and 117 on the left side. Of total stones, 241 (98.8%) were located in the kidneys; 103 (42.7%) of these stones were in the middle parts, 97 (40.2%) in lower poles and 41 (17%) in the upper poles of the kidneys. 3 ureteral stones were found while no pelvis or bladder stone was detected. Considering the greatest diameter of the stones, 107 patients (69.9%) had stones less than 5 mm, 7 (4.6%) had stones less than 5 mm and also stones measuring 5-10 mm, 35(22.9%) only had stones measuring 5-10 mm and 4 (2.6%) had stones greater than 10 mm.
**Conclusions:** Compared with the previous reports, large stones are less frequent in this study and bilateral stones, the usual indicators of underlying metabolic disorders, are relatively more common; furthermore, the renal stones are less commonly located in upper poles of the kidneys and the ureters.

## Abstract# 360

**A Report on Metabolic Evaluation of 153 Children with Urolithiasis** M.H. Fallahzadeh, M.K. Fallahzadeh, V. Sedighi, M. Basiratnia, M.A. Fallahzadeh, F. Fallahzadeh, A. Derakhshan, G.H. Hashemi. *Shiraz Nephrourology Research Center, Shiraz University of Medical Sciences, Shiraz, Islamic Republic of Iran.*
**Objectives:** The objective of this study was to evaluate the possible underlying metabolic abnormalities in children with urolithiasis.
**Methods:** 153 patients (82 boys and 71 girls with M/F ratio of 1.15) ranging in age from 2 months to 18 years (mean age =4.84 ±4.81 years) with documented renal stones were enrolled in this prospective cross-sectional study. Full metabolic

evaluations of blood and urine were done for all of the patients.
**Results:** 139 patients (90.8%) had at least one related metabolic abnormality. The frequencies of these abnormalities in decreasing order were as following: hypomagnesuria (n=93, 60.8%), hypocitraturia (n=81, 52.9%), hypercalciuria (n=72, 47.1%), hyperuricosuria( n=57, 37.3%), hypernatriuria ( n=52, 34.4%), hyperoxaluria( n=26, 17%), renal tubular acidosis( n=10, 6.5%), hyperuricemia(n=7, 4.6%) and cystinuria (n=3, 2 %).
Anatomical abnormalities were detected in 6 patients; of these patients, 5 had associated metabolic abnormalities.
**Conclusions:** In contrast to previous reports, higher proportion of our patients have underlying metabolic abnormalities, even in those with anatomical abnormalities. Furthermore, in other reports, hypercalciuria is the most frequent abnormality associated with urolithiasis; however, in our study, hypomagnesuria and hypocitraturia are more common.

## Abstract# 361

**Long Term Renal Function after Diarrheic Hemolytic and Uremic Syndrome (D+HUS)** M. Fila, V. Baudouin, S. AZIB, M.A. Macher, G. Deschenes. *Néphrologie Pédiatrique, CHU Robert Debré, Paris, France.*
**Objectives:** Although complete recovery occurs in 75% of D+HUS patients, renal dysfunction could reappear years later. The aim of the study was to describe renal outcome and to highlight long term prognostic factors for D+HUS patients and ≥10 years follow-up.
**Methods:** Initial data as well as renal function tests at 6 months and at 1 year were collected for 219 patients admitted for D+HUS between 1972 and 1998. Renal function were assessed in these patients between 2007 and 2009 and they were classified in 3 groups according to the level of GFR: CKD0 (no chronic kidney disease), CKD1 (proteinuria/ microalbuminuria/ hypertension, normal GFR); CKD2-5 (GFR 80-0 ml/min/1.73m²).
**Results:** Data from 140 of 210 surviving patients were available with a median follow up of 18.7 years after HUS. Among them, 62 patients (45%) belonged to CKD0, 46 (33%) to CKD1, and 32 (22%) to CKD2-5. Median follow-up was respectively 17.3, 18.2, and 22.7 years*. Multivariate analysis showed that anuria ≥8 days*, need of dialysis*, serum creatinine >60µmol/L 1 month after HUS* were factors of poor long term renal outcome *(p<0,01). In contrast complete recovery of renal function by 1 year after D+HUS was not a reliable marker of favourable outcome.
**Conclusions:** By 20 years of follow up, 55% of patients with a history of D+HUS present CKD. The significant difference in the duration of follow-up between groups suggest that the rate of CKD worsen with time and that some patients currently in CKD1 are at risk to develop CKD2-5 in the next decade. We suggest that all D+HUS patients should benefit of a life long follow-up even in case of complete recovery of renal function in the year following HUS.

## Abstract# 362

**Frequency of Hyponatremia in Hemato-Oncologic Overhydrated Pediatric Patients** d.P. Fonseca L,[1,2] S. Padilla Ch,[1,2] F. Alejo G,[1,2] A. Gordillo M.[2] *[1]Hospital Central Ignacio Morones Prieto, San Luis Potosi, Mexico; [2]University of San Luis Potosi, San Luis Potosi, Mexico.*
**Objectives:** To evaluate the frequency and clinical associations of hyponatremia in overhydrated children with hemato-oncology pathology.
**Methods:** Observational, retrospective study. 138 overhydration events in patients up to 16 years at the Hospital Central "Ignacio Morones Prieto" were reviewed. There were 92 events in patients with acute leukemia and 49 with solid tumors. Demographic and biochemical factors were analyzed. Univariate and multivariate logistic regression analyses were performed with R 2.5.1 software.
**Results:** Hyponatremia was found in 49 (30%) events, 45 (92.0%) in leukemia patients (p<0.001). Sodium content in IV fluids were higher in patients with hyponatremia, 78.5 ± 10 versus 68.5 ± 27 mmol/L (p<0.05). Vincristine and diuretics were prescribed in 85% and 43% of patients respectively, with hyponatremia (p<0.05). No significant associations were found between hyponatremia and age, sex, weight and nutritional status. The initial serum sodium was 138 +/- 7 mmol/L in hyponatremic patients and 141 +/- 3 mmol/L in patients without hyponatremia (p<0.0002).
**Conclusions:** Hypotonic solutions are routinely used in chemotherapy hydration protocols. Hyponatremia is multifactorial and frequent in patients treated with large hypotonic IV fluids. Acute leukemia diagnosis, vincristine and diuretic administration, and initial low serum sodium should be considered as risk factors.

## Abstract# 363

**Effects of Renin-Angiotensin System Blockers in Childhood Chronic Kidney Disease with Hypodysplastic Nephropathies** R. Fujimaru, H. Yamada. *Department of Pediatrics, Osaka City General Hospital, OSaka, Japan.*
**Objectives:** A consensus has emerged that the renin-angiotensin system (RAS) blockers such as angiotensin converting enzyme inhibitors (ACEI) and angiotensin-II receptor blockers (ARB) exert renoprotective effects through anti-hypertensive and anti-proteinuric properties in patients with chronic kidney disease (CKD). It is not clear, however, whether CKD with a lower rate of urinary protein excretion like congenital kidney malformations benefit from the RAS blockers. The aim of this study is to evaluate the renoprotective effects of the RAS blockers to slow renal deterioration in children with hypodysplastic nephropathies.
**Methods:** We studied 8 patients with stage 2 to 4 CKD for bilateral hypodysplastic nephropathies, who ranged from 6.1 to 16.0 years old at the start of the treatment. Hypertension was present in 3 patients, 2 suffered moderate proteinuria and 5 had only mild proteinuria before the treatment. Four patients received ACEI and the rest were treated with ARB.
**Results:** After the mean follow-up of 35 months, all the patients are on treatment. 3 patients with hypertension have shown enough decrements in blood pressure to be within normal range and mild proteinuria decreased in 3 of the 5 patients. The slope of the reciprocal serum creatinine (1/ Cr) curve fit showed slowed deterioration in 3 patients. None developed adverse events such as hyperkalaemia.
**Conclusions:** Though the range of proteinuria in dysplastic kidney disease is usually mild, our results suggest the possible benefits of the RAS blockers to ameliorate the decline of renal function. Further large-scale clinical studies as well as a better understanding of the pathophysiological role of the RAS are expected.

## Abstract# 364

**Childhood Anti-Neutrophil Cytoplasmic Anti-Bodies (ANCA) Positive Vasculitis: A Case Series of Four Patients** P. Gajjar, P. Nourse, P. Komala. *Renal Unit, Red Cross Children's Hospital, Cape Town, South Africa.*
**Objectives:** To document the clinical, histological and serological characteristics of children with ANCA positive vasculitis presenting to our institution.
**Methods:** A retrospective folder review of patients with clinical manifestations of small vessel inflammation, necrotising glomerulonephritis and serological finding of ANCA positivity.
**Results:** Four patients met the criteria. All were girls, ranging from 5 yrs to 12 yrs (mean age of 7.5 yrs). The main clinical findings were: influenza-like symptoms (75%), haematuria/proteinuria (100%), pulmonary-renal syndrome with haemoptysis in 50%, and acute renal failure in 50 %. One patient presented with bilateral lacrimal gland swelling. All were hypertensive. One patient with rapidly progressive nephritis had a pulmonary haemorrhage six years later. In the rest, the diagnosis was confirmed within two months. All had an elevated ESR, with normal complement and antinuclear antibody levels. Three of the four patients had an elevated pANCA; the patient with lacrimal gland swelling was cANCA positive. All underwent a renal biopsy, showing necrotising lesions with fibrous and fibrocellular crescents. Initial treatment included methylprednisone and 6 doses of monthly intravenous Cyclophosphamide followed by maintenance treatment. One patient progressed to end stage renal failure, and subsequently died. The rest are well, in remission.
**Conclusions:** Small vessel vasculitis should be considered early in the differential diagnosis of patients presenting with constitutional symptoms or unusual presentation involving the upper or lower respiratory tract, with abnormal urinalysis. An autoimmune screen should include ANCA serology.

## Abstract# 365

**Blood Pressure Is Not a Reliable Marker of Volume Status in Children with First Episode of Nephrotic Syndrome** P. Geier,[1] S. Skalova,[2] J. Feber.[1] *[1]Division of Nephrology, Children's Hospital of Eastern Ontario, Ottawa, ON, Canada; [2]Department of Pediatrics, Teaching Hospital, Hradec Kralove, Czech Republic.*
**Objectives:** Aim of our study was to evaluate whether systemic blood pressure (BP) can be used to differentiate between intravascular hypo- or hypervolemia.
**Methods:** All newly diagnosed patients with NS from 2004 to 2008 were enrolled. Casual BP was measured in all children at presentation of their NS and converted into Z-scores (SDS) based on age and height related reference values. Urine was tested for sodium (UNa) and potassium (UK). Intravascular volume status was calculated by the formula: UK/(UK+ UNa) > 0.6 = hypovolemia; UK: (UK+ UNa)< 0.6 = normo- or hypervolemia.
**Results:** Thirty seven children were studied, mean (±SD) age at diagnosis was 4.74 ± 0.28 years, 21 children were younger than 4 years and 16 children were older. Hypovolemia was diagnosed in 11 out of 37 (29%) children. In patients <4 years of age, hypovolemia was more frequent (9 out of 21; 43%; RR=1.77, 95% CI 1.075 – 2.923). However, intravascular volume status did not correlate with BP. Mean (±SD) systolic BP SDS was 1.52 ± 0.90 in hypovolemic group and 1.35 ± 1.28 in normo/hypervolemic group (p=0.69). Hypertension (HTN) was diagnosed in 14 children (38%) but the presence of HTN was not significantly different between hypovolemic (5/11) and normo/hypervolemic (9/26) groups (p=0.71, RR=0.83, 95%CI=0.45-1.53).

**Conclusions:** HTN is present in 38% children with the first episode of NS, but does not correlate with the volume status. Therefore, blood pressure is not a reliable marker of intravascular hypovolemia/hypervolemia in children with nephrotic syndrome.

## Abstract# 366

**Multicenter Investigation of Melamine Associated Urolithiasis in Young Children** N. Guan, J. Ding, Q. Fan, Y. Zhao, S. Huang, Q. Fang, D. Zhang, B. Hu, C. Yao, S. Zhu, G. Xu, Y. Ai. *Peking University First Hospital, Beijing, China.*

**Objectives:** By single center study on melamine associated urolithiasis we found potential risk factors. Large scale investigation is needed to further clarify the disease.

**Methods:** Prospective case control research on children less than 36 months old screened for urolithiasis was performed in 6 centers in China. Children with urolithiasis, without congenital abnormality of urinary tract and history of renal diseases were included as cases. Controls were randomly sampled according to the case: control ratio of 1:1. The risk factors, stone status and manifestations were reassessed.

**Results:** There were 1329 cases and 1317 controls with a mean age of 18.4 months. For melamine associated urolithiasis, we confirmed that the factors of high melamine formula and preterm were risk factors, symptoms were lacking and most of urinalysis was normal. We also found children more than 1 year old were 0.538 times unlikely to have stones than younger; children fed together with breast milk have around one half chance to have stones than children fed only with formula. Children with symptom of vomiting or diarrhea were more likely to have stones. Most of stones were small and located at kidney. Children fed with high melamine formula were nearly twice more likely to have large stones.

**Conclusions:** High melamine formula and preterm infant were confirmed to be risk factors for stones. New identified influencing factors were age, breast milk, and vomiting or diarrhea. Children less than 1 year old were at high risk to have stones and were recommended to be fed with breast milk.

## Abstract# 367

**Assessing Worry Domains and Quality of Life in a Population of Adolescents with an Idiopathic Nephrotic Syndrome** A. Meynard,[1] B. Olliac,[1] V. Guigonis,[1,9] F. Bandin,[2,9] E. Berard,[3] M. Caillez,[4] F. Dalla-Valle,[5,9] M. DiMaio,[6] J. Harambat,[7,9] A. Marinier,[8,9] R. Garoux.[1] *[1]Psychiatry and Pediatrics, CHU, Limoges, France; [2]Pediatric Nephrology, CHU, Toulouse, France; [3]Pediatric Nephrology, CHU, Nice, France; [4]Pediatric Nephrology, CHU, Marseille, France; [5]Pediatric Nephrology, CHU, Montpellier, France; [6]Pediatrics, CHU, Poitiers, France; [7]Pediatric Nephrology, CHU, Bordeaux, France; [8]Pediatrics, Hôpital Général, Pau, France; [9]Centre de Référence SORARE, Toulouse, France.*

**Objectives:** Steroid Dependant Nephrotic Syndrome (SDNS) is frequent among chronic pathologies during childhood. Few studies had evaluated its impact on quality of life (QOL) and preoccupations among adolescents.

**Methods:** Fifty patients (mean 14.9y) suffering from SDNS were investigated in a multicentric study. QOL, worries, anxieties and depression were evaluated by, respectively the French version of Children's Health Questionnaire, the French version of the worry domains questionnaire,the French versions of the Goldberg Depression and Anxiety Scales.

**Results:** Our results show that components of QOL were more correlated with anxious or depressive symptoms than with the severity of the SDNS. Degrees of disease severity, age and sex were not associated with QOL. Difficulties in school work or daily life activities were correlated with level of anxiety ($p<0.05$). General health, social limitations, behaviour, mental health, self-esteem were correlated with level of depression($p<0.05$). ll adolescents' population expressed fewer worries than a control population of 350 adolescents ($p<0.05$).

**Conclusions:** Worse QOL was more associated with anxious or depressive symptoms than with the severity of the SDNS in our sample of adolescents.

## Abstract# 368

**What Are the Diagnosis Criteria Necessary To Confirm a Pneumococcal Related Hemolytic and Uremic Syndrome?** A. Loupiac,[1] M. Caillez,[2] A. Adra,[3,6] E. Berard,[4] S. Decramer,[5,6] V. Guigonis.[1,6] *[1]Pediatrics, CHU, Limoges, France; [2]Pediatric Nephrology, CHU, Marseille, France; [3]Pediatric Nephrology, CHU, Montpellier, France; [4]Pediatric Nephrology, CHU, Nice, France; [5]Pediatric Nephrology, CHU, Toulouse, France; [6]Centre de Référence SORARE, Toulouse, France.*

**Objectives:** Hemolytic and Uremic Syndrome related to Pneumococcal infection (HUS-P) is a rare condition. Pediatricians should be more familiar with this disease as early recognition could have important implications for treatment and prognosis. No consensus has been established for the diagnosis criteria of HUS-P and, in some cases, the diagnosis of certitude can be difficult.

**Methods:** We conducted a retrospective study evaluating the diagnosis strategy and criteria used for HUS-P in six nephrology pediatrics centers over the past 10 years. The criteria used for the diagnosis of pneumococcal infection and HUS were recorded as well as all the tests performed during the etiology diagnosis workup.

**Results:** A total of 18 children with HUS-P were studied. The bacteriological diagnosis of pneumococcal infection was confirmed in only 72.2% of cases. Others etiologies of HUS were ruled out in 83.3 % whereas tests to directly confirm HUS-P were performed in only a minority of cases. Indeed, Thomsen Friedenreich antigen testing – using lectin -was only performed in 11.1%.

**Conclusions:** These results confirmed the difficulty to diagnose HUS-P supporting the absence of a consensual diagnosis strategy. An etiological diagnosis workup is proposed based on biological criteria (using Thomsen Friedenreich antigen testing). This should allow faster diagnosis confirmation, avoid unnecessary tests, and lead clinicians more rapidly to the correct therapeutic strategy.

## Abstract# 369

**Lactate Dehydrogenase as a Predictor of Kidney Involvement in Sickle Cell Anemia** S. Gurkan,[1] K.J. Scarponi,[1] H. Hotchkiss,[2] B. Savage,[1] R. Drachtman.[1] *[1]Pediatrics, University of Medicine and Dentistry of New Jersey(UMDNJ)-Robert Wood Johnson Medical School, New Brunswick, NJ, United States; [2]Pediatrics, Mount Sinai School of Medicine, New York, NY, United States.*

**Objectives:** To determine the prevalence of renal involvement in a SCD population and identify potential risk factors for renal involvement.

**Methods:** A retrospective chart review of 40 patients with sickle cell anemia (SCA) between the ages of 5-19 years that were seen within the past year was performed to determine clinical correlates for microalbuminuria and proteinuria. Age, sex, height, body mass index (BMI), serum creatinine (and estimated glomerular filtration rate (eGFR) by Schwartz and MDRD formulas), type of SCA, hemoglobin (Hb) level (total Hb and % HbF), lactate dehydrogenase (LDH) level, reticulocyte count, blood pressure, history of splenectomy, history of hydroxyurea use and history of transfusions were correlated with microalbuminuria and proteinuria by univariate and multivariate regression analysis.

**Results:** The prevalence of microalbuminuria and proteinuria was 15% and 5% respectively. Univariate analyses revealed a significant correlation between LDH level and microalbuminuria (Pearson $r=0.47$, $p=0.04$) and LDH level and proteinuria (Pearson $r=0.48$, $p=0.035$). Multivariate analysis revealed a significant correlation between microalbuminuria and LDH level ($p=0.04$) when controlled for age, sex, eGFR, Hb level, HbF%, type of SCA, BMI, history of transfusions and reticulocyte count.

**Conclusions:** In this pediatric SCA population, LDH is found to correlate with microalbuminuria and proteinuria. Further studies are needed to confirm LDH as an early marker for risk of kidney involvement among SCA patients.

## Abstract# 370

**Side Effects of Initial 4-Week Steroid Therapy in 219 Children with Renal Diseases** R. Hiramoto, H. Fujimaki, S. Matsumoto, H. Eguchi. *Department of Pediatrics, Matsudo City Hospital Children's Medical Centre, Matsudo, Chiba, Japan.*

**Objectives:** To evaluate the side effects of full-dose steroid therapy during the first 4 weeks and to decide the appropriate admission period. In Japan, the introduction of steroid treatment in children with renal diseases is commonly performed as inpatient during the first 4 weeks.

**Methods:** We compiled a retrospective cohort of 219 children with renal diseases (range: 1 – 17 years) admitted for the first time to our hospital from 1986 through 2009. All the patients received either 2 mg/kg/day of prednizolone (PSL) or methylprednisolone pulse therapy followed by PSL (1mg/kg/day) for 4 weeks. Of 219, 130 had nephrotic syndrome, 47 IgA nephropathy, 24 Henoch-Shönlein purpura nephritis and 18 lupus nephritis. The data were collected from medical records regarding hypertension, ophthalmologic complications, psychiatric disturbances, and infections.

**Results:** Within the first 4 weeks, 37(19.2%) showed hypertension and 22(10.0%) developed ophthalmologic complications including cataract, ocular hypertension or glaucoma. This indicated a possibility of much earlier onset of cataract than previous reports. During admission, 19(8.7%) suffered from psychiatric disturbances such as depression and insomnia, and 30(13.7%) needed antibiotics or antiviral drugs against infections. The mean onset of hypertension was 15.4 days, and that of ophthalmologic complications was 21.3 days (range: 5 days – 2 months).

**Conclusions:** These data show that we should be well aware of the above-mentioned side effects, especially during the first 4 weeks of full-dose steroid treatment. From the safety point of view, at least 4 weeks admission is considered to be appropriate in Japan.

## Abstract# 371

**Chronic Kidney Disease in India-Data from a Pediatric Registry** S. Indian Society of Pediatric Nephrology (ISPN), ISPN. *Secretariat Indian Society of Pediatric Nephrology, Room No 3053, Teaching Block, Department of Pediatrics, New Delhi, India.*

**Objectives:** To collate epidemiological information on pediatric chronic kidney disease (CKD) in India.

**Methods:** A summary of data on 435 children with an estimated GFR ≤ 59 ml/min/1.73 $m^2$ (≥ CKD stage 3) from a pediatric CKD registry (13 centers) initiated in 2008. Participation in this is voluntary and mandates involvement of a pediatric nephrologist/ nephrologist in provision of care to patients.

**Results:** Majority of entries are from Northern India (65%). There is a male preponderance (77%), mean age 7.20 ± 4.57 years and symptom onset for a mean period of 3.2 years. Children with CKD stages 3, 4, 5 were 147 (33.7%), 143 (32.8%) and 145 (33.3%) respectively. Commonest presentation was edema (33%); others seen frequently were failure to thrive (15%), acidosis (14 %), hypertension (9%). Common etiologies were obstructive uropathy (25%), vesicoureteric reflux (14%) and renal hypoplasia/dysplasia (11%). Mean serum creatinine was 3.2±2.8 mg/dl, and mean hemoglobin 9 ±2.3 g/dl. Majority of patients were on self finance; 20% receving eryhtropoietin and 0.45% receiving growth hormone. Sixty three patients were receiving renal replacement therapy as CAPD (6%), maintenance HD (5.5%) and 14 (3.2%) had received renal transplant.

**Conclusions:** As a pilot initiative, the data provides important insights: significant delay from onset of symptoms to patient reaching center of care; obstructive uropathy, vesicoureteric reflux, renal hypoplasia/dysplasia account for half the cases; very few receive ideal management viz erythropoietin, growth hormone.

Acknowledgement: Indian Society of Nephrology; Janssen-Cilag India.

## Abstract# 372

**Risk Factors and Timing of Henoch-Schönlein Purpura Nephritis (HSN)** O. Jauhola,[1] J. Ronkainen,[1] O. Koskimies,[2] M. Ala-Houhala,[3] P. Arikoski,[4] T. Hölttä,[2] T. Jahnukainen,[2] J. Rajantie,[2] T. Örmälä,[5] M. Nuutinen.[1] *[1]OUH, Oulu, Finland; [2]HUCH, Helsinki, Finland; [3]TUH, Tampere, Finland; [4]KUH, Kuopio, Finland; [5]Hyvinkää Hospital, Hyvinkää, Finland.*

**Objectives:** To assess the risk factors for developing HSN and the time period when nephritis is unlikely after the initial disease onset.

**Methods:** A prospective study of 223 pediatric pts to examine renal manifestations of Henoch-Schönlein purpura (HSP). The pts' condition was monitored with 5 outpatient visits and urine dipstick tests at home. Of the pts 111 did not receive any medication, 89 were treated with prednisone prophylaxis and 24 either with Cyclosporine-A or MP-pulses due to severe HSN.

**Results:** HSN occurred in 102/223(46%) of the pts consisting of isolated hematuria in 14%, proteinuria in 9%, both hematuria and proteinuria in 56%, nephrotic-range proteinuria in 20% and nephrotic-nephritic syndrome in one case. Nephritis occurred mean 14 days after HSP diagnosis and in 87% within one month. The incidence after 1 and 2 months was 14% and 2%, respectively. The occurrence of HSN increased with age, $p<0.001$ for linear trend. In multivariate analysis the risk factors for nephritis were age > 8yrs (OR 2.7, p=0.002, Cl 1.4-5.1), abdominal pain (OR 2.1, p=0.017, Cl 1.1-3.7) and HSP recurrences (OR 3.1, p=0.002, Cl 1.5-6.3). Pts with 2 or 3 risk factors developed nephritis in 63% and 87% of cases. Laboratory tests or blood pressure at onset did not predict HSN. Early prednisone treatment did not affect the frequency or timing of nephritis.

**Conclusions:** Weekly urine dipstick tests are indicated for 2 months after the HSP diagnosis. Patients > 8yrs, with abdominal pain or HSP recurrences have higher risk for HSN.

## Abstract# 373

**Neutrophil Gelatinase-Associated Lipocalin (NGAL) as Early Marker of Hypertensive and Diabetic Nephropathy in Children** A. Kaczmarek, J. Soltysiak, J. Zachwieja, K. Lipkowska, A. Blumczynski, D. Ostalska-Nowicka, M. Silska. *Pediatric Nephrology, Poznan University of Medical Sciences, Poznan, Poland.*

**Objectives:** In recent years, NGAL has emerged in clinical and experimental nephrology as the most promising tubular biomarkers in the diagnostic field of acute and chronic renal diseases. Clinically detectable hypertensive (HN) or diabetic nephropathy (DN) begins with the development of microalbuminuria (MA). However, early renal dysfunction may be overlooked using that method.

**Methods:** The aim of the present study was to evaluate the level of neutrophil gelatinase-associated lipocalin (NGAL) in children with normal range albuminuria e.g. in those considered as not presenting diabetic nor hypertensive nephropathy. The study group consisted of 40 children with type 1 diabetes mellitus (T1DM) or primary hypertension (PHT). 15 healthy children served as a control group.

**Results:** Children with T1DM showed increased NGAL values with respect to controls; interestingly, both in serum (867,43±341,98 vs. 655,29±196,17 ng/ml; p=0,04) and in urine (433,71±170,99 vs. 305,36±128,85 ng/ml, p=0.02). The positive correlation between sNAGL and UAE was found (UAE mg/kg/d = -0,1042 +, 51E-3 * sNGAL; r=0.58; p=0.005). Children with PHT showed increased NGAL values with respect to controls; interestingly, both in serum (1545,50±1317,19 vs. 655,29±196,17 ng/ml; p=0,02) and in urine (772,75± 658,59 vs. 305,36±128,85 ng/ml, p=0.01).

**Conclusions:** Normal range albuminuria does not exclude hypertensive nor diabetic nephropathy defined as increased sNGAL and uNGAL concentration. NGAL measurement may become a useful tool for the evaluation of renal involvement in diabetic and hypertensive children.

## Abstract# 374

**Pediatric Kidney Biopsy in Orenburg Region, Russian Federation** M.U. Kagan, N.N. Bervina. *Gastroenterology and Nephrology, Orenburg Regional Children's Hospital, Orenburg, Russian Federation.*

**Objectives:** Over a 9 year period from 2001-2009, 58 cases ( 19 girls and 39 boys, mean age 11,2 ± 4,3 years) met criteria for kidney biopsy at our hospital. Clinical presentations included nephrotic syndrome [17], hematuria [28], rapidly progressive glomerulonephritis [8], acute renal failure [3] and proteinuria [2].

**Methods:** Renal biopsies were performed by nephrologists themselves using a real-time ultrasound guidance and a 14-16 G automated needle. The target of the procedure was to obtain two macroscopically adequate specimens from the lower renal pole (usually the left). Only if one of the two specimens appeared too small, we try to obtain a third one.

We obtained two specimens in 23 cases (60,3%), and three specimens in 35 cases (39,7%).

Pathologic evaluation was based chiefly on the combination of the light microscopic observation with immunofluorescence microscopy data. In 35 cases electron microscopy was performed too.

**Results:** Mean number of glomeruli per specimen was 25,3 ± 14,5 [6-65]. Pathological findings included 5 minimal change diseases (8,6%), 7 FSGS (12,1%), 18 IgA- nephropathy (31,0%), 2 membranous glomerulonephritis (3.4%), 8 crescentic glomerulonephritis (13.8%), 4 Alport syndrome (6,9%), 6 thin basement membrane disease (10,4%), 2 trombotic microangiopathy (3,4%) and 6 other lesions (10.4%). Minor complications (gross hematuria and/or perinephric hematoma resolving without the need for transfusion or intervention) occurred in 16 patients (27.6%). No major complications were noted.

**Conclusions:** A renal biopsy in pediatric patients is a safe, reliable and minimally invasive procedure. The results supplemented our knowledge of renal diseases in children in the Orenburg region.

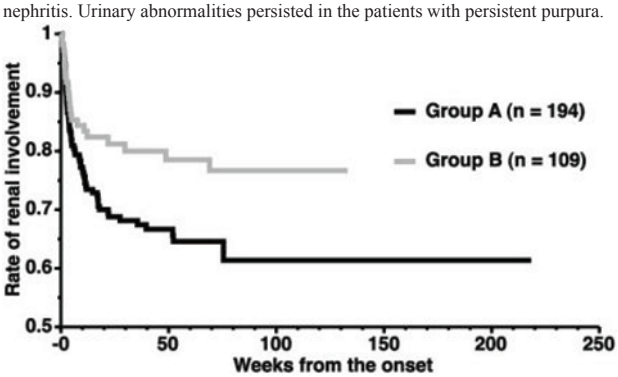
## Abstract# 375

**Corticosteroid Decreases the Risk of Renal Involvement in Henoch-Schönlein Purpura but Not the Course** Y. Kaku. *Nephrology, Fukuoka Children's Hospital, Fukuoka, Japan.*

**Objectives:** It is controversial yet whether steroid can prevent nephritis in Henoch-Schönlein purpura (HSP), although we reported that steroid decreased the risk of nephritis (Kidney Int, 1998). We have treated the HSP patients with these risk factors for nephritis with steroid at the disease onset. The aim of this study is to verify the effect of preventing corticosteroid use against the development and the course of nephritis in HSP.

**Methods:** Group A (historical control group, n=194): Before we were aware of the risk factors of renal involvement, steroids were administered in order to only control the pain. Group B (recent patients group, n=109): From 2001, we administer 1mg/kg/day of predonisolone for 1-2 weeks to the patients with the risk factors. We compared the rate of nephritis by using Kaplan-Meier method and log rank test. The course of nephritis in group B was analyzed similarly.

**Results:** The severity of HSP in the two group was not differ. The rate of nephritis in group B was lower than group A (23.2% vs 38.6%, $p$ <0.0057). 28 patients in group B developed nephritis. Steroid did not affect the course of nephritis. Urinary abnormalities persisted in the patients with persistent purpura.



**Conclusions:** These results elucidate that corticosteroid has the protective effect against nephritis in HSP. However, corticosteroid did not affect the course of nephritis. The therapy for nephritis maybe modified the course.

## Abstract# 376

**Relevance of Microalbuminuria in Screening for HIV-Associated Nephropathy** B.J. Mistry, U.K. Kala. *Paediatrics, University of the Witwatersrand and Chris Hani Baragwanath Hospital, Johannesburg, Gauteng, South Africa.*

**Objectives:** The prevalence and clinical significance of a single screening test for microalbuminuria in a cohort of HIV seropositive children from an inpatient and outpatient setting without any symptoms of renal disease at Chris Hani Baragwanath hospital situated in Johannesburg, South Africa.

**Methods:** A prospective study of HIV seropositive and seronegative patients screened for qualitative proteinuria and microalbuminuria. The exclusion criteria being on antiretroviral therapy, anti tuberculosis treatment, known chronic kidney disease, hypertension, fever, acute illness and urinary tract infection.

**Results:** 180 patients were enrolled into the study. 110 were HIV positive and 70 HIV negative. Majority of the patients were black (98%) with 100 (56%) males and 80 (44%) females. Microalbuminuria was present in 27(25%) of HIV positive patients, p=0.00003. Microalbuminuria was present in 21 (19%) p=0.03 in children with normal renal function and no proteinuria Microalbuminuric patients were moderately immunosuppressed and were WHO clinical stage 2 and 3. Absolute CD4 counts correlate better with microalbuminuria than CD4 percentage as the mean CD4 absolute count in HIV positive patients with microalbuminuria and significantly lower than those without microalbuminuria, p=0.03.

**Conclusions:** Microalbuminuria screening of HIV positive patients is a more sensitive screening test compared to standard urine dipsticks.This may allow for early identification of subclinical renal disease in patients with some evidence of immunosuppression. With an indication for early initiation of antiretroviral therapy thus possibly preventing the deterioration of renal function and severity of HIV disease.

## Abstract# 377

**Albumin as a Surrogate Marker of Oxidative Stress in Idiopathic Nephrotic Syndrome** K. Kaneko, M. Takahashi, S. Yamanouchi, M. Hasui. *Pediatrics, Kansai Medical University, Hirakata-shi, Osaka, Japan.*

**Objectives:** Serious infection, such as spontaneous bacterial peritonitis (SBP), can complicate idiopathic nephrotic syndrome (INS). Though pathogenesis of susceptibility to bacterial infection remains unknown, multiple predisposing factors including defective opsonisation, altered T cell function, altered IgG concentrations have been proposed. Albumin is worthy of note because it plays an important role in the host immunity as one of the important antioxidants.

This study was condcted to investigate whether hypoalbuminemia contributes to declined immunity in INS.

**Methods:** Thirty five heparinised blood samples were obtained from 4 patients with INS aged 10-15 years at the time of relapse or remission. Serum levels of reactive oxygen metabolites (ROM) and biological antioxidant potentials (BAP) were measured by spectrophotometry using a novel analyzer, FRAS4 (Diacron Int., Italy). Serum albumin level (HSA) was measured by colorimetric assay. Relationships between HSA and ROM, BAP, and ROM/BAP ratio were studied.

**Results:** HSA varied from 0.8 to 4.4 g/dL, and correlated with ROM and BAP ($p<0.01$). Correlation coefficient between HSA and BAP ($r_s= 0.74$) was higher than that between HSA and ROM ($r_s= 0.43$). In addition, HSA correlated inversely with ROM/BAP ratio ($r_s= 0.31$, $p<0.05$).

**Conclusions:** Our results suggest that low HSA contributes to a declined immunity, and agree with the finding that children with INS complicated by SBP had lower HSA. It is, therefore, worth studying the efficacy of infusion of albumin in INS with hypoalbuminemia, not to increase osmotic pressure but to prevent SBP by supplying depleted BAP, because the beneficial effect in liver cirrhosis has been confirmed.

## Abstract# 378

**The Role of a Published Algorithm in the Refferal of Pediatric Patients with Asymptomatic Proteinuria (AsP)** S.P. Katz, P. Thomas, R. Jacob. *Pediatrics, Nassau University Medical Center, East Meadow, NY, United States.*

**Objectives:** A previous review of 2 years of our experience (February 2005-February 2007) demonstrated that 9 of 9 patients(pts) would not have been referred if the primary care physician had followed an algorithm published in 2000. We have reviewed more recent experience to determine the nature of referrals and to compare them with our previous data.

**Methods:** A retrospective study of pts referred for AsP to the pediatric nephrology service at Nassau University Medical Center from July 2008 to December 2009. Data was collected for age, sex, prior lab data, random urinalysis (UA), 1st morning UA, urine protein/creatinine ratio, diagnosis and follow-up. The data was compared with our experience from February 2005-February 2007.

**Results:** 22 pts were referred during the 2 study periods. Male /Female ratio was 1:1. Ages ranged from 5-17 years. 6/22pts had no prior labs available at the time of referral. In 19/22pts the proteinuria was transient or resolved & 3/22pts had orthostatic proteinuria. In all of the patients the algorithm was either not applied or not applied properly.

**Conclusions:** 1. None of the pts referred for evaluation for AsP had significant renal disease. 2. It could have been determined if the proteinuria detected was significant prior to referral if the algorithm had been used in evaluating these pts. 3. Use of the algorithm would have led to savings in terms of cost and anxiety. 4. A larger study involving multiple centers would be of value in determining whether our experience is unique to our community or not. 5. The algorithm should be used by primary care physicians to evaluate children & adolescents with asymptomatic proteinuria.

## Abstract# 379

**Atypical Clinical Features of Acute Poststreptococcal Glomerulonephritis (APSGN) in Children** K. Kim,[1] H. Hwang.[2] *[1]Department of Pediatrics, NHIC Ilsan Hospital, Koyang, Korea; [2]Department of Pediatrics, Fatima Hospital, Changwon, Korea.*

**Objectives:** Although APSGN is one of the most common nephritic syndromes, it still continues to be misdiagnosed due to its diverse clinical presentations. Respiratory distress, pulmonary edema and hypertension are most problematic and if unrecognized could lead to a delay in treatment and increased morbidity.

**Methods:** We report 2 cases of APSGN in children with atypical initial symptoms.

**Results:** Case 1. A 10-year-old boy presented acutely with headache, altered mental state, and generalized type of seizure. He had hypertension and microscopic hematuria and the MRI showed lesions suggestive of Posterior Reversible Encephalopathy Syndrome (PRES). Antistreptolysin-O (ASO) titer was increased and complement C3 titer was decreased. He was diagnosed as PRES related with contemporary hypertensive event in APSGN.

Case 2. A 9-year old boy was transferred with dyspnea and alveolar infiltrates with bilateral pleural effusions on plain chest film. Urinalysis and blood pressure measurement showed microscopic hematuria and hypertension. Elevated serum ASO titer and depressed serum complement C3 levels confirmed the diagnosis of APSGN.

**Conclusions:** In children who present with dyspneic respirations and a chest radiograph showing pulmonary edema, proper evaluation including blood pressure recording and urinalysis should be performed immediately.

And blood pressure must be measured promptly in all children presenting with altered mentation, seizures and visual disturbances. If elevated, the diagnosis of PRES should be considered and a workup for renal disease pursued.

Prompt diagnosis and early therapy of APSGN may avoid mortality and unnecessary therapeutic intervention.

## Abstract# 380

**Epidemiology of End-Stage Renal Disease in Slovak Children** G. Kolvek,[1] L. Podracká,[1] S.A. Reijneveld,[2] J.P. van Dijk,[2] J. Rosenberger.[3] *[1]1st Paediatric Department, Košice, Slovakia (Slovak Republic); [2]Department of Social Medicine, Groningen, Netherlands; [3]Nephrology and Dialysis Center Fresenius Košice, Košice, Slovakia (Slovak Republic).*

**Objectives:** The paediatric population suffering from end-stage renal disease (ESRD) is growing, but current reliable epidemiological information on ESRD in children is missing from many countries. Aim of the study was to examine the occurence of ESRD in Slovak children; to compare it with earlier data on Slovakia and with data from neighbouring European countries; and finally to explore aetiology and treatment modes.

**Methods:** Over the years 2005-2008 data on incident and prevalent cases of ESRD in children from all four tertiary paediatric centres in Slovakia were collected. Incidence and prevalence rates were calculated per million age related population (pmarp) and per million total population (pmtp). The data were compared with two earlier Slovak studies and with European data from European Society of Paediatric Nephrology datasets.

**Results:** The mean annual incidence rate of ESRD in Slovak children under 15 years observed in the study period was 5.8 pmarp (0.9 pmtp). The prevalence rate on 31st December 2008 was 23.9 pmarp (3.7 pmtp). Differences between 2008 and 2002 (18.6 pmarp, 3.2 pmtp) were not statistically significant. Incidence and prevalence rates of ESRD in Slovak children are comparable to surrounding European countries. Aetiology mainly concerned congenital anomalies (42.9%) and hereditary nephropaties (21.4%).

**Conclusions:** During the last decade the incidence and prevalence rates of ESRD in Slovak children have remained stable. Aetiology regarding the study period is comparable with neighbouring countries.

## Abstract# 381

**Urinary Levels of Matrix Metalloproteinases (MMP-1 and MMP-2) and Their Tissue Inhibitors (TIMP-1 and TIMP-2) in Proteinuric Children** A. Korzeniecka-Kozerska, A. Wasilewska, K. Taranta-Janusz, W.M. Zoch-Zwierz. *Department of Pediatrics and Nephrology, Medical University of Bialystok, Bialystok, Poland.*

**Objectives:** Recent studies suggest that extracellular matrix metalloproteinases (MMPs) and their tissue inhibitors are involved in many kinds of kidney diseases. We assessed the role of MMPs and TIMPs in patients with minimal changes nephrotic syndrome and with moderate proteinuria due to other glomerulopathies.

**Methods:** The study group consisted of 60 patients subdivided into group IA- n=20 patients with nephrotic syndrome, group IB – patients with nephrotic syndrome after proteinuria subsided; group II (n=20) with moderate proteinuria. The control group (K) contained 20 healthy children in the same age and sex. Urinary levels of estimated MMP-2 and MMP-9 and their tissue inhibitor TIMP-1 and TIMP-2 were determined using **immunoenzymatic method.**

**Results:** Our study revealed increased levels of MMP-2 in patients from I and II group compared with controls, and did not differ between group IA and II. Urinary concentration of TIMP-1 was higher in both groups (I and II) in comparison to healthy controls. TIMP-2 levels were higher only in group II. MMP-2/TIMP2 and MMP-9/TIMP-1 ratios among group IA, IB, II and group K were comparable. Increased MMP-2/TIMP2 ratio was positively correlated with protein level in the urine of children from group I. Positive correlation was found between MMP-2 and proteinuria in group II.

**Conclusions:** We demonstrated that the imbalance between MMP-2, MMP-9, TIMP-1, TIMP-2 may lead to fibrosis. Estimation of MMPs and TIMPs concentrations may be a predictive factor of kidney fibrosis and can be used for monitoring EMC renal accumulation.

## Abstract# 382

**Cost-Effectiveness (CE) of Latent Tuberculosis (LTB) Screening in Nephrotic Syndrome (NS)** B. Laskin,[1] A. Trauernicht,[1] M.H. Eckman,[2] J. Goebel.[1] *[1]Pediatrics, Cincinnati Children's Hospital Medical Center, Cincinnati, OH, United States; [2]General Internal Medicine/Center for Clinical Effectiveness, University of Cincinnati, Cincinnati, OH, United States.*

**Objectives:** Guidelines vary on LTB screening before pediatric immunosuppression (IS), particularly regarding reactivation (TBr) risk and new screening tests (interferon-gamma release assay, IGRA).

**Methods:** CE analysis with Decision Maker® comparing universal (univ) skin testing (PPD), IGRA, and targeted (trgt) PPD with a risk questionnaire (Quest) over 1 year, using a 5 year-old with new-onset NS as base case, model inputs from the literature, Medicare, and published infliximab data, and 2010 US$ and quality-adjusted life years (QALYs) as outcomes metrics (Table 1).

Table 1

| | Probability | US$ | Utility |
|---|---|---|---|
| Sens/Spec Quest | 0.8/0.55 | 2 | |
| Sens/Spec PPD | 0.8/0.97 | 55 | |
| Sens/Spec IGRA | 0.7/0.99 | 39 | |
| Well | | | 1 |
| LTB Prevalence | 0.01 | 772 | 0.95 |
| TBr | 0.0002 | 25000 | 0.8 |
| Case Fatality Rate | 0.1 | 25000 | 0.5 |

Spec:Specificity; Sens:Sensitivity

**Results:** Trgt PPD screening was the least costly and most effective option (Table 2) over a range of parameter values, including TBr rates. For IGRA to be superior, PPD and IGRA sens would have to be <20 and >85%, respectively, and LTB prevalence >60%.

Table 2

| | US$ | QALYs |
|---|---|---|
| Trgt PPD | 34 | 0.995 |
| Univ IGRA | 53 | 0.985 |
| Univ PPD | 86 | 0.989 |

**Conclusions:** This model supports using trgt PPD for American children with new-onset NS before IS. Requiring 1 visit, IGRA may be an option once validated in children. A future Markov CE model will use a longer time horizon to confirm these preliminary results.

## Abstract# 383

**Hemolytic Uremic Syndrome – Experience in 541 Patients** R. Exeni, I. Grimoldi, M.d.C. Laso, J. Baigorri, A. Exeni, G. Falke, C. Ciancaglini, C. Rapetti, G. Montala. *Pediatric Nephrology, Hospital del Niño de San Justo, San Justo, Buenos Aires, Argentina.*

**Objectives:** Hemolytic Uremic Syndrome (HUS) is the most common cause of acute renal failure in children in Argentina, being the highest incidence in the world. The aim of our study was to evaluate the clinical course in the acute period of typical HUS in our center in Argentina.

**Methods:** From November 1974 to December 2007, 541 infants and children with HUS D+ were admitted. Epidemiological aspects, age, gender, clinical manifestations,treatment and mortality,were registered.

**Results:** The age of the patients ranged from 4 -132 months (mean 19.3 months). 48,8 % were males and 51.2 % females.

90% had history of bloody diarrhea, 10 % rectal prolapse, 3.6 % ischemic colitis. All presented haemolytic anemia and thrombocytopenia. Leukocytosis was present in 210/267. 23 % had arterial hypertension. No anuria was found in 10.9 %. Anuria less than 7 days in 54.5 %. Anuria more than 7 days 30.9 %.Anuria without recovery 3.70 % patients. Low levels of Epo were found in 9/10 patients. 23 % had major neurological manifestations.

4 patients with hyperosmolar coma due to pancreatic involvement.

4 patients with severe ocular abnormalities.

Glomerulocystic disease in 2 patients.

Mortality :16 patients (3 %), 6 with Neurological System involvement, 3 due to ischemic colitis,3 with Acute cardiac alterations, 3 Hyperosmolar coma, 1 with Hiperkalemia.

Treatment :99.4 % were transfused, 378 (70 %) received peritoneal dialysis, 4 patients hemodialysis.

**Conclusions:** The study shows the high incidence of SUH in Argentina, the low mortality in the acute period and the predominance of peritoneal dialysis as the treatment.

## Abstract# 384

**Significance of Mass Urinary Screening in School Children: Follow-Up Report of Asymptomatic Urinary Abnormality** H.K. Lee,[1] H.J. Choi,[1] S.J. Park,[1] H.G. Kang,[1] I.S. Ha,[1] H.I. Cheong,[1] Y. Choi.[2] *[1]Pediatrics, Seoul National University Children's Hospital, Seoul, Republic of Korea; [2]Pediatrics, Inje University Haeundae Paik Hospital, Busan, Republic of Korea.*

**Objectives:** The aim of study was to investigate the underlying causes and the prognosis of the children with asymptomatic urinary abnormality detected in mass urinary screening in Korea.

**Methods:** We reviewed 248 children (M:F=108:140, age 10.2±2.6 years) who visited our center between 1998 and 2009 due to the abnormal urinary findings on school screening (microscopic hematuria (mHU): proteinuria (PU): mHU+PU 149: 42: 57).

**Results:** In mHU group (n=149), nutcracker syndrome was detected in 20, hypercalciuria in 19. After 3 years (n=101), mHU disappeared in 10 chidrden and proteinuria developed in 4, whose biopsy findings were IgA nephropathy (IgAN) in 2, thin membranous disease (TMD) in 1 and normal in 1. No patient showed deteriorated renal function or hypertension during follow-up.

In PU group (n=42), half of them had orthostatic PU and 8 (19%) had elevated creatinine level at first visit. The histological findings of 11 cases with persistant PU were IgAN in 4, FSGS in 3, mesangial proliferative glomerulonephritis (mesPGN) in 1, Alport syndrome in 1, lupus nephritis (LN) in 1 and C1q nephropathy in 1.

In mHU+PU group (n=57), the histological findings of 35 cases with persistent PU were IgAN in 18, FSGS in 3, Alport syndrome in 2, LN in 6, mesPGN in 2, MPGN in 3, and TMD in 1.

**Conclusions:** In conclusion, invasive studies including renal biopsy are considered for the patients with persistent proteinuria but not for the patient with mHU, since their clinical findings after more than 3 years were favorable.

## Abstract# 385

**Association of Hyponatremia with Administration of Total Parenteral Nutrition to Normonatremic Hospitalized Children: A Case-Control Study** M. Lemaire,[1] R.S. Parekh,[1] P.P. Pencharz,[2] S.H. Zlotkin,[2] D.F. Geary.[1] *[1]Nephrology, Hospital for Sick Children, Toronto, ON, Canada; [2]Gastroenterology, Hospital for Sick Children, Toronto, ON, Canada.*

**Objectives:** To compare the characteristics of hospitalized children (HC) with normal baseline plasma sodium (PNa) who developed hyponatremia with those who remained hyponatremic following total parenteral nutrition (TPN) prescription.

**Methods:** This is a case-control study of 101 HC with normal baseline PNa and prescribed TPN for >1 day identified by screening 651 consecutive TPN orders from 2007. We excluded children from the ICU, with renal or hepatic failure, chylothorax, or on home TPN. Data gathered during the first 14 days of TPN included biochemical indices, details regarding IVF, medications, demographics and clinical parameters. Mild and moderate are defined as PNa <135 and <130 mmol/L, respectively.

**Results:** The patients with mild (49) or moderate (5) hyponatremia (cases) were compared to those who remained normonatremic (controls). While no between groups difference was found for initial TPN prescriptions characteristics, hyponatremic patients had more frequent ($p<0.001$) and larger ($p<0.0001$) increases in TPN Na during TPN infusion. Pre-TPN Na inputs from IVF (mmol/kg/day) for 36 randomly chosen subjects from both groups were higher for hyponatremic patients. Statistically significant risk factors for hyponatremia

included younger age, longer TPN therapy, and admission to Surgery or Oncology ward.
**Conclusions:** Hyponatremic episodes are common for HC during TPN infusion. Frequent PNa monitoring is advisable when TPN is prescribed to younger patients admitted to Surgery or Oncology who receive large volumes of saline before starting TPN.

## Abstract# 386

**RenalVysion™ Urine Cytologic Analysis (U-Cyto) in the Management of Pediatric Nephrologic Clinical Problems** K. Lieberman,[1] D. Thomas,[2] S. Seshan,[3] L. Ettinger,[1] C. Picarelli.[1] *[1]Pediatr Nephrol, Sanzari Children's Hospital, Hackensack, NJ, United States; [2]Nephrocor, Uniondale, NY, United States; [3]Pathology, Weill Medical College, New York, NY, United States.*
**Objectives:** To investigate U-Cyto in the dx of glomerulonephritis(GN),hematuria (hem) and renal allograft(txp) dysfunction.
**Methods:** Urine was submitted to Nephrocor™ for RenalVysion™ exam in pts with GN and txp dysfunction who were having renal biopsies(bx).Specimens were also submitted for outpts being evaluated for gross & micro hem.Traditional hem investigations were pursued,including bx where indicated.U-Cyto findings were correlated with path.U-Cyto findings were correlated with dx in hem pts.Bx performed on gross hem pts if hem persisted & no dx established.Bx performed on micro hem pts if hem persisted with high RBC count(>10/HPF) & no dx established.
**Results:** U-Cyto performed in 11 pts-gross hem,20-micro hem,4-txp dysfunction,8-GN (1-MPGN,6-SLE,1-HSP).2 pts refused bx,2 pts lost to f/u. Dx established in 9 gross hem pts(3-glom dis by bx,3-nl bx,1-stone,2-exercise). All pts with dysmorphic RBCs,epith cell fragments,>50 PMNs,necrotic tubule cells,RBC casts had glom path.Dx established in 9 micro hem pts(3-thin GBM,1-nl bx,2-hypercalciuria,1-s/p cytoxan,1-exercise,1-Alport carrier).All pts with tubuloepith fragments,>5% dysmorphic RBCs had glom path.2/3 thin GBM pts had >10 necrotic tubular cells,only 1/15 benign micro hem pts had them($p<.05$). GN & txp pts had frequent epith cell fragments,necrotic tubular cells,RBC dysmorphia,lymphs,PMNs,collecting duct cells,RBC casts.
**Conclusions:** U-Cyto correctly identified all hem pts with glom path.U-Cyto may be useful in tracking therapeutic response in GN & txp pts.
**DISCLOSURE:** *Thomas, D.*: Other, Nephrocor - employee.

## Abstract# 387

**Prevalence of Symptoms of Lower Urinary Tract in Children from 6 to 12 Years** E.M. Lima, G.T.B. Vaz, M.M.A. Vasconcelos, P.K.S. Torres, T.M.L. Santos. *Pediatrics, Federal University of Minas Gerais, Belo Horizonte, Minas Gerais, Brazil.*
**Objectives:** To investigate the prevalence of symptoms of lower urinary tract in school children.
**Methods:** Cross-sectional study, including 739 children aged 6 to 12 years of age, enrolled in three public schools of Minas Gerais, with different socioeconomic levels. Symptoms were evaluated by the instrument "Voiding Dysfunction Scoring System" proposed by Farhat et al, which was adapted for children's language, in which the cutoff points by the score as an indicator of urinary tract dysfunction were greater than 6 for girls and greater than 9 for boys. Children with score indicating symptoms received an educational booklet on the function of the lower urinary tract and also a referral for medical evaluation.
**Results:** The symptoms of dysfunction of the lower urinary tract were detected in 161 (21.8%) children. The urinary symptoms more common in children with increased score were holding maneuvers (37.2%), diurnal urinary incontinence (26.7%) and urgency (23.6%). The symptoms were more frequent in girls ($p<0.001$) in children between 6 and 8 years of age ($p<0.028$) and at school with the lowest social level ($p<0.001$). Constipation was the most prevalent symptom (30.7%), regardless of the value of the score. Emotional stress factors were present in 28.4% of children studied.
**Conclusions:** The study showed a high prevalence of lower urinary tract symptoms in children of school age children, higher in younger children and girls belonging to lower social class. These symptoms should be considered and investigated carefully in routine pediatric consultations.

## Abstract# 388

**A Succesfull Model of International Cooperation in Pediatric Nephrology** G. Marra,[1] E. Alberto,[1] M. Sandoval,[2] M. Castellon,[2] F. Sereni.[3] *[1]Pediatric Nephrology Unit, Policlinico Ca'Granda, Milano, Italy; [2]Pediatric Nephrology Unit, HospitalInfantilManuel de Jesus Rivera, Milano, Italy; [3]Pediatric Department, University of Milan, Milano, Italy.*
**Objectives:** Discussing and sharing the methodology of a successful Italian-Nicaraguan cooperative project,started in 2000,for the care of kidney and urinary tract diseases (KUTD).
**Methods:** The project,financed by 2 Italian and Nicaraguan Foundations and 2 Italian Public Institutions and developed with the continuous involvement of local Hospital Authorities and Ministry of Health, included:1-training of medical and nurse team 2-provision to Managua Unit of devices for the diagnosis and treatment of all KUTD 3-implementation of a pediatric clinical-web network between Milan, Managua and 6 Nicaraguan dept hospitals; 4-collection of epidemiological data on KUTD over the territory.
**Results:** The Managua Unit is now able to care for all kind of KUTD, both from the diagnostic(lab-instrumental exams,biopsy) and therapeutic point of view(medications,dialysis,LDR kidney transplant)and is the pediatric nephrology reference centre for all Nicaragua. The local staff progressively increased with three new positions:a pediatric urologist and 2 nephrologists. A network of dept hospitals (covering 61% of pediatric population) has been established for a prompt diagnosis and treatment of all basic KUTD. The KUTD registry contains now 5567 pts. Since 2007 annual costs for medications for pediatrc transplant and hemodialysis are progressively covered by Nicaraguan Health Authority.
**Conclusions:** The cooperation between public and private associations and a strong involvement of Health Institutions and local staff has been the key of success of the project.

## Abstract# 389

**Fractional Excretion of Sodium in Children with Nephrolithiasis and Nephrocalcinosis** A. Mazo,[1] A. Akopyan,[1] S. Zorkin,[1] E. Vasiljeva,[1] N. Mayanskiy,[1] R. Zakalukin,[2] A. Tsygin.[1] *[1]Scientific Center of Children's Health, Russian Academy of Medical Sciences, Moscow, Russian Federation; [2]Shubnikiv Institut of Crystallograhy Russian Academy of Sciences, Moscow, Russian Federation.*
**Objectives:** Recently a correlation between increased excretion of sodium and hypercalciuria in children have been revealed. The value of fractional excretion of sodium (FENa) have not been investigated in children with nephrolithiasis and in children with nephrocalcinosis.
**Methods:** Three groups of children were studied: 40 children with nephrolithiasis (aged 6.6±4.8 years), 30 children without chronic and acute deseases (aged 7.0±5 years), and 6 children with nephrocalcinosis (aged 5.2±3.9 years). Spot urine sample and serum specimen were analyzed. Stone samples (30) were examined by X-ray diffraction method.
**Results:** We found that FENa was significantly increased in children with nephrolithiasis (0.99±1.1%) compared with control group (0.36±0.2%, $p<0.05$). Excretion of calcium was increased in the nephrocalcinosis group (1,5±1,9, $p<0,05$). Fractional excretion of potassium and other solutes didn't differ in all groups. Correlation ($p<0.05$) between FENa and excretion of oxalate, fractional excretion of calcium, and phosphate were revealed in children with nephrolithiasis only. That allowed to suggest tubular sodium transport disturbance in nephrolithiasis. FENa didn't depend from stone composition. There wasn't increased sodium excretion in children with nephrocalcinosis, that could be influenced by therapy.
**Conclusions:** The result indicates that increased FENa is associated with nephrolithiasis, suggesting that a reduction of salt intake may prevent stone recurrence in children.

## Abstract# 390

**The UK Registry of Rare Renal Disease (RaDaR) Enables Research Studies on a National Scale** H. McCarthy,[1] D. Ansell,[2] F. Braddon,[2] M. Taylor,[3] M. Saleem.[1] *[1]University of Bristol, Bristol, United Kingdom; [2]UK Renal Registry, Bristol, United Kingdom; [3]Birmingham Children's Hospital, Birmingham, United Kingdom.*
**Objectives:** In 2009 all member states of the European Union agreed to improve health care in rare disease. Political backing was mirrored by financial support from the Medical Research Council's patient-cohort-initiatives, funding the development of this registry (RaDaR).
**Methods:** Administrative infrastructure has been provided by the UK Renal Registry - a well established registry of all patients in the UK with end stage renal disease.
RaDaR is governed by the Renal Association and backed by the UK renal community. It is operated by a RaDaR committee. Disease Specific Working Groups (DSWG), utilizing collaborations, provide research into multiple aspects of specific diseases including diagnostic, genetic, biomarker development and therapy.
**Results:** Two pilot projects into MembranoProliferative GlomeruloNephritis and Steroid Resistant Nephrotic Syndrome have commenced, with recruitment expanding rapidly. Clinical datasets for the website have been developed by the DSWGs in collaboration with RaDaR. Local teams in the collaborating centres populate those datasets, including histology which will be analysed by a central pathology steering group. The information collected is owned by RaDaR and given to the DSWG in link-anonymised form. Additional data that is generated by the DSWG following analysis will be returned to RaDaR at the end of the study.

**Conclusions:** This project illustrates a comprehensive model for study, management and dissemination of information for rare diseases on a National basis, overcoming governance, data protection and IT hurdles in a coordinated strategy.

## Abstract# 391

**Diagnostics of Infringements of Intrarenal Haemodynamics in Children with Vesicoureteric Reflux and Reflux Nephropathy**

I.V. Zorin,[1] A.A. Vyalkova,[2] A.G. Miroshnichenko,[1] A.V. Zorin,[2] A.I. Karpov.[1] *[1]Pediatric Nephrology, Pediatric Clinic N 6, Orenburg, Russian Federation; [2]Pediatrics, Orenburg Medical academy, Orenburg, Russian Federation.*

**Objectives:** The aim of the study was to estimate informative parameters of infringements of intrarenal haemodynamics in children with vesicoureteric reflux (VUR) and reflux nephropathy (RN).

**Methods:** We examined 150 children with RN and VUR. We established data of color Doppler ultrasound (CDUS) as diastolic velocities (Vd), systolic velocities (Vs), resistive indices (Ri) and pulsatility indices (Pi); DMSA scanning (time of the maximal accumulation (TMA, sec), maximal activity (MA, sob/sec), mean velocities of accumulation (MVA, mm/sec), the contribution to the common accumulation (CCA, %). Patients were divided into 3 groups:

I – children with unilateral RN A according to classification of Smellie J. et all, 1975 (n=50);

II – children with unilateral RN D according to classification of Smellie J. et all, 1975 (n=50);

III – children with VUR without renal damage (n= 50)

**Results:** We established that parameters of CDUS as Vd, Vs, Pi and DMSA scanning as TMA, MA, MVA, CCA in patients with RN and VUR are decreasing proportionally of the degree of renal scarring. With the help of mathematic analyzes we established informative parameters of CDUS and DMSA scanning and revealed that data of CDUS as Vd (8,0), Vs (5,1), DMSA scanning as MVA (9,3), MA (8,8), TMA (6,1) are very informative parameters of diagnostics of infringements of intrarenal haemodynamics in children with VUR and RN.

**Conclusions:** These parameters can be used not only for early diagnostics of scarring in children with VUR but also for diagnostics of progression of renal scarring.

## Glomerular Disease

## Abstract# 392

**(O-43)**

**National Survey of Rituximab Treatment for Childhood Idiopathic Nephrotic Syndrome**

S. Ito,[1] K. Kamei,[1] M. Ogura,[1] T. Udagawa,[1] S. Fujinaga,[2] K. Iijima.[3] *[1]Department of Nephrology, National Center for Child Health and Development, Tokyo, Japan; [2]Department of Nephrology, Saitama Children's Hospital, Saitama, Japan; [3]Department of Pediartrics, Kobe University, Kobe, Japan.*

**Objectives:** Rituximab (RTX), an anti-CD20 monoclonal antibody, is a hopeful option for steroid-dependent (SDNS), frequent-relapsing (FRNS) and steroid-resistant nephrotic syndrome (SRNS). We conducted a national survey of off-label RTX use for refractory NS.

**Methods:** Questionnaires were sent to 130 hospitals in Japan. Sixty-seven hospitals responded and 10 hospitals reported its use.

**Results:** Seventy-one patients (42 males; 29 females; 52 SDNS; 4 FRNS; 15 SRNS) were treated with RTX. Thirty patients with SDNS/FRNS had >10 relapses before RTX. Adverse events of steroid, short stature in 30, hypertension in 15, cataract in 15, glaucoma in 10, spinal fracture in four and bone necrosis in five were observed. Before RTX, cyclosporine (CsA) in 70, cyclophosphamide in 29, MMF in 43, tacrolimus in seven, steroid pulse therapy in 43 and plasma exchange in 12 were tried. In SDNS/FRNS, 45 patients discontinued PSL and 17 discontinued CsA. However, 27 relapsed along with recovery of CD20 lymphocytes. In SRNS, five patients achieved complete remission and four achieved partial remission, but six did not respond. Mild infusion reactions were observed in 35. Two patients develpoed agranulocytosis, and one suffered sepsis.

**Conclusions:** The steroid-sparing effect of RTX in SDNS/FRNS is remarkable. However, recovery of CD20 lymphocytes frequently induces relapse. Clinical trials are required to establish more effective administration modes. In FRNS, RTX may become an alternative treatment.

## Abstract# 393

**(O-49)**

**Insulin Signaling to the Podocyte Is Critical for Glomerular Function**

G. Welsh,[1] L Hale,[1] V. Eremina,[2] M. Jeansson,[2] C. Caunt,[1] C. McArdle,[1] R. Owen,[1] J. Tavare,[1] R. Lennon,[1] C. Kahn,[3] H. Pavenstadt,[4] P. Mathieson,[1] S. Quaggin,[2] M. Saleem,[1] R. Coward.[1] *[1]Bristol University, Bristol, Avon, United Kingdom; [2]University of Toronto, Toronto, ON, Canada; [3]Harvard Medical School, Boston, MA, United States; [4]University Clinics, Muenster, Germany.*

**Objectives:** Diabetic nephropathy (DN) is the leading cause of renal failure in the world and is becoming more prevalent in the young. It has previously been considered a hyperglycaemic complication directed against the renal microvasculature. DN has a natural history dominated by albuminuria and we have previously demonstrated that human podocytes are uniquely insulin sensitive. We therefore set out to examine the biological role of insulin signaling to the podocyte in the kidney.

**Methods:** To determine whether insulin signaling to podocytes affects glomerular function in vivo we generated transgenic mice with specific deletion of insulin receptors from their podocytes.

**Results:** These animals develop significant albuminuria with increased matrix production, mesangial expansion, loss of podocyte cytoskeletal architecture, thickening of the glomerular basement membrane and glomerulosclerosis. Thus, elimination of insulin signaling in podocytes alone is sufficient to recapitulate the major features of DN despite normoglycaemia. Our studies suggest that the mechanism of this effect involves dynamic control of actin cytoskeleton remodeling in the podocyte by insulin receptor signaling.

**Conclusions:** Collectively this shows the critical contribution of altered insulin signaling in the podocyte in the pathogenesis of DN. Thus, therapies that improve the insulin sensitivity of this cell could have great potential in treating this major global health problem.

## Abstract# 394

**(O-50)**

**CD2AP Is a Key Molecule To Maintain the Epithelial Phenotype of the Podocyte**

S. Ramadan,[1] G. Welsh,[1] L. Ni,[1] A. Koziell,[2] M.A. Saleem.[1] *[1]Academic Renal Unit, University of Bristol, Bristol, Avon, United Kingdom; [2]Institute of Child Health, London, United Kingdom.*

**Objectives:** CD2AP is a multifunctional adaptor molecule, and plays a crucial role in the kidney where it is essential for ultrafiltration functions of the slit-diaphragm complex. Knockout of CD2AP in mice leads to nephrotic syndrome. The role of CD2AP in podocytes is not known. We established for the first time CD2AP mutant podocytes from a child with diffuse mesangial sclerosis due to a missense mutation in exon 16 of CD2AP.

**Methods:** Human conditionally immortalised meseangial cells were used as controls.

**Results:** CD2AP mutant podocytes displayed a fibroblast like morphology with spindle shape, WT podocytes grow as groups of cells that preserve cell-cell contacts. Nephrin, CD2AP, Podocin and WT1 were expressed at comparable level in both cell lines. Mesenchymal markers (fibronectin and α-SMA) were significantly overexpressed in mutant podocytes, while in WT they were expressed at low but detectable levels. PAX-2 was expressed in CD2AP mutant cells and not in WT podocytes. Mechanistically, WTIP was shown to translocate to the nucleus in mutant cells.

**Conclusions:** Fibroblast like morphology of the mutant cells, disturbance of the cell sheets and overexpression of EMT markers and re-expression of PAX2 indicate that the CD2AP mutation may have led to podocyte dedifferentiation. We showed that normally CD2AP recruits WTIP to the cell membrane, whereas a mutant involved in human disease shuttles it to the nucleus where it suppresses WT1-dependent gene expression to permit simplification of podocyte phenotype. This is a novel role for CD2AP, with implications for its role in development and progression of glomerular disease.

## Abstract# 395

**(O-51)**

**PI3K and Smad3 Interact To Promote Murine Renal Fibrosis**

G. Finer, T. Hayashida, Y.S. Kanwar, H.W. Schnaper. *Pediatrics, Northwestern University, Chicago, IL, United States.*

**Objectives:** TGF-β plays a central role in renal fibrogenesis. We previously showed in vitro that PI3K and TGF-β stimulated Smad3 activity lead to collagen I expression, and now aimed to extend these findings to an in-vivo model.

**Results:** Balb/c mice treated with ADR (0.15 mg, iv) showed massive albuminuria and advanced global glomerulosclerosis, as described by others. We found increased renal mRNA for TGF-β1, type I collagen, fibronectin and smooth muscle α-actin (αSMA). Immunoblotting showed phosphorylation of Akt, a downstream marker for PI3K activity. A PI3K inhibitor, LY294002 (25mg/kg, i.p, 2x/week), prevented ADR-induced histological changes, albuminuria, and up-regulation of fibrosis markers. The γ isoform of the PI3K p110 catalytic unit was selectively increased in ADR kidneys and in cultured mouse podocytes treated

with TGF-β. 129sj mice treated with ADR showed similar nephrotic changes such as albuminuria and hypoalbuminemia, and azotemia, but histological changes were more consistent with human focal segmental glomerulosclerosis (FSGS) than those in Balb/c. Smad3 phosphorylation, and nuclear localization shown by immunostaining, implicated TGF-β. A TGF-β inhibitor, soluble human TGF-β type II receptor (sTβRII.Fc), protected 129/sj mice from ADR-induced histological changes and diminished collagen I and αSMA mRNA expression, but did not prevent proteinuria. Smad3 activation seen in ADR-treated 129/sj was ameliorated by sTbRII Fc.
**Conclusions:** These results suggest that ADR-induced nephropathy in 129/sj mice shows features similar to human FSGS and suggest that TGF-β and PI3K are promising targets for intervention in kidney fibrosis.

## Abstract# 396
**(O-52)**

**Efficacy of Daily Corticosteroids To Prevent Infection Associated Relapses in Frequently Relapsing Nephrotic Syndrome: A Randomized Controlled Trial** A. Gulati, A. Sinha, P. Hari, A. Bagga. *All India Institute of Medical Sciences, Delhi, India.*
**Objectives:** We examined whether daily administration of maintenance prednisolone during intercurrent infections, reduces relapse rates in children with frequently relapsing nephrotic syndrome (FRNS).
**Methods:** Patients with FRNS (>2 relapses in 6 mo) eligible for therapy with prolonged alternate day (ad) prednisolone (0.5-0.7 mg/kg ad) with/ without levamisole (2 mg/kg ad) were eligible.Randomization done to group I (Intervention) or C (Control). During presumed viral infections,patients in Gr.I were advised to take the same dose of prednisolone every day for 7 consecutive days; patients in Gr. C continued to receive ad prednisolone. Relapse (3+ proteinuria for 3 consecutive days) was considered infection-associated if it occurred within 2-wks of onset of viral illness. Patients were followed up for 12-mo.
**Results:** For 100 patients included, Gr. I (n=50) showed significantly lower relapse rates (mean difference 0.9 episodes/pt/yr; 95% CI 0.4-1.4) at 12-mo.The number of infection-associated relapses was significantly lower in Gr. I (mean difference 0.7; 95% CI 0.3-1.1).The increase in relapses with the number of infections was more apparent in Gr.C. An adjustment for occurrence of infections, showed that daily administration of prednisolone during viral infections independently resulted in 59% reduction in rate of relapses (rate ratio 0.41; 95% CI 0.3-0.6). The intervention is likely to reduce the number of relapses to < 3 in 12 months, for one out of six children treated.
**Conclusions:** Daily administration of maintenance doses of prednisolone, during viral infections, significantly reduces relapse rates and the proportion of children with FRNS.

## Abstract# 397
**(O-53)**

**Inhibition of p38 MAPK/MK-2 Protects Podocytes from Puromycin Aminonucleoside-Induced Injury** R. Pengal,[1] A. Guess,[1] R. Ransom,[1,2] R. Benndorf,[1,2] W. Smoyer.[1,2] *[1]Clinical and Translational Research, The Research Institute at Nationwide Children's Hospital, Columbus, OH, United States; [2]Department of Pediatrics, The Ohio State University, Columbus, OH, United States.*
**Objectives:** Elucidation of the role of the p38 MAPK/MK-2 signaling pathway in mediating nephrotic syndrome-related podocyte injury induced by puromycin aminonucleoside (PAN).
**Methods:** PAN-induced injury of cultured podocytes was monitored by measuring cell viability and integrity of the actin cytoskeleton. Podocytes were treated with specific inhibitors, SB203580 and C23, to block the activity of p38 MAPK and MK-2, respectively. Electrophoretic analyses were used to verify the effects of these drugs in podocytes.
**Results:** PAN-induced loss of podocyte viability was significantly prevented by inhibition of p38 MAPK or MK-2. In addition, inhibition of p38 MAPK or MK-2 almost completely prevented PAN-induced disruption of podocyte actin filaments. At the molecular level, both inhibitors markedly reduced basal phosphorylation of the MK-2 substrate Hsp25, suggesting that prevention of PAN-induced injury indeed was due to inhibition of this signaling cascade. In addition, we found that exposure of podocytes to serum albumin strongly activated p38 MAPK/ MK-2 signaling which suggests that this pathway may contribute to cell damage resulting from protein overload.
**Conclusions:** Collectively, these data suggest a direct role of p38 MAPK/MK-2 signaling in mediating podocyte injury resulting from treatment with PAN or serum albumin. Consequently, specific pharmacological inhibition of p38 MAPK or MK-2 offers a potentially novel therapeutic strategy to prevent or treat podocyte injury in nephrotic syndrome.

## Abstract# 398
**(O-54)**

**HIV Nephropathy in Children in KwaZulu-Natal, South Africa** R. Bhimma,[1] D. Ramsuran,[2] P.K. Ramdial,[3] A. Naicker,[2] E. Naicker,[1] M. Adkikari.[1] *[1]Paediatrics and Child Healtyh, University of KwaZulu-Natal, Durban, KwaZulu-Natal, South Africa; [2]Optics and Image Centre, University of KwaZulu-Natal, Durban, KwaZulu-Natal, South Africa; [3]Department of Anatomical Pathology, University of KwaZulu-Natal, Durban, KwaZulu-Natal, South Africa.*
**Objectives:** To determine the spectrum of HIV related nephropathy in children in KwaZulu-Natal, South Africa.
**Methods:** Fifty patients with persistent proteinuria underwent kidney biopsy for detection of nephropathy.
**Results:** All 50 patients undergoing biopsy had evidence of nephropathy, 49(98%) were black and 1(2%) of mixed ancestry; 33(66%) were males. The average age of presentation was 81 months. Clinical characteristics include oedema 27(54%), proteinuria 45(90%), and 3(6%) hypertension. Nine (18%) had tuberculosis as an opportunistic infection. Laboratory results show hypocomplementaemia in 24(45%) children [low C3 in 9(17%) and low C4 38(73%)], increased liver enzymes in 16(22%) patients [ALT 13 (19%) and GGT 18 (25%)]. Ultrasonography showed increased echogencity in 9(20%) and increased kidney size in 9(23%). Histology showed the following: FSGS 32(64%) with 7(14%) having the collapsing FSGS; tubulointerstitial nephritis 8(16%); HIV immune complex disease 3(6%); membranous glomerulonephritis 3(6%); mesangial proliferative glomerulonephritis 1(2%) and others 4(8%).
**Conclusions:** HIV nephropathy in children is predominantly a non collapsing FSGS as opposed to adults with others showing varied histology. The main presenting feature is asymptomatic proteinuria. Urine analysis should be done as a routine screen in all children who are HIV infected and all having persistent proteinuria should undergo kidney biopsy for the detection of nephropathy.

## Abstract# 399
**(O-55)**

**Puromycin Aminonucleoside Increases Podocyte Permeability by the Modulation of ZO-1 Via Oxidative Stress** T.-S. Ha, E.-M. Ahn, J.-S. Lee. *Chungbuk National University, Cheongju, Chungbuk, Korea.*
**Objectives:** Puromycin aminonucleoside (PAN)-induced nephrosis is a well-described model of human idiopathic nephrotic syndrome because PAN injection into rats results in increased glomerular permeability with the characteristic ultrastructural changes in glomerular epithelial cells (GEpC; podocytes) similar to human nephrosis. We investigated the role of zonula occludens (ZO)-1 and oxidative stress on PAN-induced podocyte phenotypical changes and hyperpermeability *in vitro*.
**Methods:** Rat GEpCs were incubated in media containing various concentrations of PAN. The phenotypical changes of ZO-1were analyzed by confocal imaging, Western blotting, and PCR. We also examined *in vitro* permeability and oxidative stress level.
**Results:** Morphological assessment revealed that *in vitro* PAN not only induced the ultrastructural changes of GEpC, such as shortening and fusion of microvilli, but also separated the intercellular gaps and linear ZO-1 resulting in increased intercellular permeability. Oxidative stress level after PAN treatment was markedly higher than that of basal level. PAN induced the inner cytoplasmic translocation of ZO-1 protein and also reduced ZO-1 protein amount and mRNA expression in a dose-dependent manner. These phenotypical changes of podocyte caused by PAN were augmented by the antioxidative effect of vitamin C.
**Conclusions:** We suggested that the glomerular hyperpermeability caused by intercellular ZO-1 disturbances via oxidative stress would be the mechanism of proteinuria in experimental PAN-induced nephrosis.

## Abstract# 400
**(O-56)**

**Plasminogen/Plasmin in Urine May Be a Mechanism of $Na^+$ Retention in Idiopathic Nephrotic Syndrome** R.F. Andersen,[1] K.B. Buhl,[2] B.L. Jensen,[2] S. Rittig.[1] *[1]Department of Paediatrics, Aarhus University Hospital, Skejby, Aarhus, Denmark; [2]Physiology and Pharmacology, Institute of Medical Biology, University of Southern Denmark, Odense, Denmark.*
**Objectives:** In animal models of nephrotic syndrome (NS), glomerular filtration of plasminogen and activation to plasmin has been reported to activate the epithelial $Na^+$ channel (ENaC) and may cause sodium retention. Our aim was to investigate if plasminogen/plasmin (PLAS) is present in urine from patients with active NS and whether excretion is altered at remission.
**Methods:** Patients with idiopathic NS were included. Urine samples were collected at the day of inclusion and again at remission. Total PLAS concentration was determined with ELISA. Urinary protease activity was examined in 17 of 20 patients by gelatin zymography and western blotting for PLAS in urine was performed in selected patients.

**Results:** A total of 20 patients (7 girls) with mean age 9.1±3.2 yrs were included. Median nephrotic urine PLAS was 1780 ng/ml (range 297-22700) versus 72 ng/ml (range 25-143) in urine at remission (p<0.001). Zymograms showed protease activity at 70-80 kDa in urine in 10 of 17 patients compared to 3 of 17 patients at remission. Western blots showed a 75kDa protein compatible with plasmin in 5 of 10 tested patients and products at 90-100 kDa in 5 patients. At remission, all 10 urine samples were negative.
**Conclusions:** Our results demonstrate significantly elevated excretion of PLAS and proteolytic activity in urine during active NS compared to remission. PLAS may be involved in sodium retention in NS and whether PLAS is indeed causing ENaC activation is currently tested in experiments using an in vitro patch clamp technique.

## Abstract# 401

**Predictors of Renal Involvement in Filipino Children with Henoch-Schonlein Purpura in a Tertiary Hospital: A Five-Year Review** P.J. Galutira,[1] B.B. Canonigo,[1] R.D. Chan,[1] R.F. Cabansag,[1] C.B. Bernal.[2] *[1]Pediatric Nephrology, University of Santo Tomas Hospital, Manila, Philippines; [2]Pediatric Rheumatology, University of Santo Tomas Hospital, Manila, Philippines.*
**Objectives:** General objective: To identify predictors of renal involvement in patients with Henoch-Schonlein purpura.Specific objectives are:(1) to identify clinical features of children with HSP;(2) to enumerate major urinary abnormalities in these patients;(3) to identify the onset of renal involvement.
**Methods:** This is a retrospective study of children diagnosed with HSP. Renal involvement was evaluated by determining the presence of proteinuria and/or hematuria at the onset and on subsequent follow-up. Comparison of the different variables under study was done using the T test, chi-square test and Fisher Exact test. A *P* value of <0.05 was considered to be significant. Logistic regression was performed to identify predictor of an outcome variable.
**Results:** 42 patients (24 male and 18 female) with the mean age of 8.89 ± 3.66 (from 2 to 17 years) were included.64.3% of the patients were >7 years of age, and amongst this, 74.1% had renal involvement. Microscopic hematuria was the most common renal manifestation. Age of onset >7 years old (*P*=< 0.01) and the presence of bloody stools (*P*=0.02) were the independent variables identified to predict renal involvement in HSP. However, on logistic regression, only the age of onset >7 y/o (*P*=0.005) significantly predicts renal involvement in HSP.
**Conclusions:** Microscopic hematuria is the most common major urinary abnormality in HSP and is observed during the first 4 weeks from onset of signs and symptoms. An age of onset >7 y/o significantly predicts renal involvement.

## Abstract# 402

**Candesartan Suppresses Glomerular Renin-Angiotensin System (RAS) Activation, Oxidative Stress and Progressive Glomerular Injury in Rat Anti-GBM GN** Y. Kinoshita, S. Kondo, K. Suga, S. Matsuura, M. Urushihara, S. Kagami. *Department of Pediatrics, University of Tokushima, Tokushima, Japan.*
**Objectives:** We examined the involvement of glomerular RAS activation in the glomerular injury in anti-GBM GN of rats with AT1R antagonist Candesartan.
**Methods:** Progressive anti-GBM GN was induced in rats. Nephritic rats were divided into two groups and given daily oral doses of vehicle or ARB in drinking water until day 28. Age-matched rats without nephritis were used as normal controls.
**Results:** Treatment with ARB significantly improved proteinuria. Vehicle-treated nephritic rats developed crescentic GN accompanied by marked macrophage infiltration and the enhanced expression of glomerular angiotensinogen (AGT), Ang II and Nox2 (NADPH oxidase) on days 7 and 28 of GN. ARB improved glomerular alterations and had a significant inhibitory effect on the levels of these parameters. Western blot using isolated glomeruli also showed that increased AGT and Nox2 expression was significantly attenuated by ARB treatment. Enhanced $O2^-$ production in nephritic glomeruli was also decreased by ARB treatment. Moreover, ELISA using the supernatant of glomerular culture revealed that the level of glomerular Ang II and TGF-β production was significantly increased in vehicle-treated nephritic rats, while ARB treatment significantly inhibited those production on day 28 (p<0.05).
**Conclusions:** This study indicates that increased glomerular Ang II production play an important role in the progressive glomerular injury through the induction of oxidative stress and TGF-β expression, and suggests that ARB treatment provides a novel therapeutic strategy for inhibiting the progression of glomerular injury in GN.

## Abstract# 403

**Neurological Involvement in a Child with Atypical Hemolytic Uremic Syndrome** B. Koehl, O. Boyer, M. Kossorotoff, N. Biebuyck, N. Boddaert, P. Niaudet. *Hôpital Necker Enfants Malades, Paris, France.*
**Objectives:** Central nervous system involvement, the most frequent extrarenal complication in hemolytic uremic syndrome (HUS) includes reversible posterior leukoencephalopathy syndrome (RPLS) and thrombotic microangiopathic (TMA) lesions, difficult to differentiate since presentation is similar.
**Methods:** We report the case of a 4 year old boy, diagnosed with atypical HUS due to an hybrid factor H.
**Results:** He progressed to end-stage renal disease despite plasmatherapy and underwent bilateral nephrectomy because of uncontrolled hypertension. Three days after, he had complex seizures with normal blood pressure, normal blood count and normal magnetic resonance imaging (MRI) which recurred 1 month later. 8 months later, he had a third episode of seizures, with hemoglobin of 10g/dl without schizocyte, low haptoglobin of 0.2g/l, and moderate thrombocytopenia (platelets $98.10^9$/L). He remained deeply confused during 2 days. The third MRI showed bilateral symmetrical hyperintensities of the cerebral pedonculae, caudate nuclei, putamens, thalami, hippocampi and insulae suggesting thrombotic microangiopathy secondary to a relapse of HUS rather than RPLS, usually occipital and asymmetrical. He was treated with daily plasma exchanges (PE) that led to a complete neurological recovery within 2 days although hypertension had remained uncontrolled. After 10 sessions, PE were replaced by weekly plasma infusions. The fourth MRI 10 weeks after was normal and clinical examination remained normal, except for high blood pressure.
**Conclusions:** Brain MRI allows differentiating TMA lesions from RPLS in atypical HUS which is crucial since lesions may be reversible with plasmatherapy.

## Abstract# 404

**Estrogens Reduce Apoptosis in Podocytes** S. Kummer,[1] V. Wegerich,[1] S. Jeruschke,[1] A. Seibt,[1] N. Koleganova,[2] G. Piecha,[2] M.L. Gross,[2] E. Mayatepek,[1] J. Oh.[1] *[1]Department of General Pediatrics, University Hospital, Duesseldorf, Germany; [2]Institute of Pathology, University Hospital, Heidelberg, Germany.*
**Objectives:** Epidemiological data show a significantly better renal prognosis for women with chronic glomerular diseases compared to men. Regarding the critical role of podocytes for pathogenesis, we studied the influences of estrogens on podocyte function.
**Methods:** Estrogen receptor (ER) expression in cultured podocytes and renal biopsies of different diseases was examined on mRNA and protein level. Cultured murine podocytes were treated with Puromycin (PAN) to induce apoptosis, Estradiol (E2) alone or both combined. Apoptotic cells were quantified by FACS analysis and Hoechst nuclear staining.
**Results:** ERα mRNA and protein were detected in podocytes. In vitro, E2 reduced PAN-induced apoptosis by 27% (p=0.01, FACS). Hoechst staining showed similar results (57% reduction of apoptosis, p=0.01). In human biopsies, low ER expression in podocytes was found in 45-55% of controls and MCGN patients (gender-independent). In sclerosed glomeruli of FSGS patients, ER was detected in a significantly higher percentage of women compared to men (89% vs. 33%, p=0.01).
**Conclusions:** The presented data show that podocytes contain specific target structures for estrogenic action. Experimentally induced apoptosis as in-vitro model of chronic glomerular diseases can be significantly reduced by E2. In FSGS, ER positive glomeruli are found to be enriched in women, suggesting a selection of ER positive cells by E2 influence. These findings represent a new model explaining gender differences in glomerular diseases.

## Abstract# 405

**Glomerular Diseases in Macedonian Children: Review of a Renal Biopsy Database** D.B. Kuzmanovska, S. Timovska, E. Sahpazova. *Pediatric Clinic, StCyril&Methodius University, Medical Faculty, Skopje, Macedonia, The Former Yugoslav Republic of.*
**Objectives:** This study aimed to determine glomerular disease frequencies in Macedonian children and it represents the basis for future studies.
**Methods:** All native renal biopsies (January 1996 to December 2008) were reviewed, but only glomerular diseases were analyzed. The diagnosis of each case was based on histological, immunopathological and clinical features. As our Clinic is the only pediatric nephrology centre in the country, the results of this study relate to the whole country.
**Results:** A total of 82 patients ≤ 15 years (mean age 8.59±3.9 %) were included in the study. Primary glomerular diseases were diagnosed in 65 biopsies (79%) and secondary in 17 (21%).The most common primary diseases were minimal change disease (35%), mesangiproliferative glomerulonephritis (17%), immunoglobulin A nephropathy (14%), focal and segmental glomerulosclerosis (12%), membranous glomerulonephritis (8%), crescentic glomerulonephritis (3%). Postinfectious glomerulonephritis (GN) represented 8% of the diagnoses if included as primary GN. In the group of secondary glomerulonephritis, Henoch Shonlein nephritis corresponded to 59% of the entire series and lupus nephritis to 29%. Alport syndrome was found in 12%.
**Conclusions:** The distribution of glomerular diseases in the pediatric age group at R.Macedonia is similar to that described in other countries with some differences. This study illustrates the importance of having a regional register for renal diseases in children.

## Abstract# 406

**A Model of Collaborative Management of Nephrotic Syndrome in Benin, a Developing Country** F.H. Lalya,[1] O. Biaou,[2] P.-C. Hounkpe,[3] M. d'Almeida,[1] B. Ayivi.[1] *[1]Department of Pediatrics, CNHU-HKM, Cotonou, Benin; [2]X-ray Department, CNHU-HKM, Cotonou, Benin; [3]Intensive Care Unit, CNHU-HKM, Cotonou, Benin.*

**Objectives:** To assess management-related difficulties of nephrotic syndrome (NS) in our setting.

**Methods:** This was a prospective observational study of all patients with NS admitted to the pediatric ward of the university hospital CNHU-HKM of Cotonou, Benin from 1 June 2006 to 31 May 2008. All cases were reviewed by an IPNA fellow. Renal biopsy specimens were either analyzed locally or sent by courier to the histopathology unit of Red Cross Children's Hospital in Cape Town, South Africa.

**Results:** NS was present in 15cases (16.5%) of renal patients. Five patients had presumed minimal change/steroid sensitive NS. Ten patients had either steroid resistant of frequently relapsing NS. Eight of these underwent renal biopsy. Only one specimen was analyzed locally and was reported as "showing features of membranous nephropathy and minimal change". The subsequent 7 specimens were analyzed at Red Cross Children's Hospital and showed: minimal change (2 cases), mesangioproliferative glomerulonephritis (2 cases), and focal segmental glomerulosclerosis (1 case). In 2 cases, specimen was not representative of anatomic site. Two patients could not afford the courier cost and we had to pay ourselves. Second line treatment included cyclosporine (1case) and cyclophosphamide (2cases). Drug level monitoring was not possible in the patient who underwent cyclosporine.

**Conclusions:** Management of NS can be challenging in resources-limited countries but a collaborative approach involving hospitals from developed or emerging countries and developing ones can be beneficial to the latter.

## Abstract# 407

**Ophtalmological Evaluation of Children with Nephrotic Syndrome Who Have Used Corticosteroids** F.V. Leão, J.R. Santos, M.A.P. Cançado, N.S. Moraes, Má.A. Fernandes, J.T. Carvalhaes. *Pediatric Nephrology, Federal University of São Paulo, São Paulo, São Paulo, Brazil.*

**Objectives:** Compare ophtalmological evaluation between general ophtalmologists and pediatric ophtalmologists in children and adolescents with nephrotic syndrome (NS) using corticosteroids (CS). CS may cause ocular complications such as, increase in intraocular pressure and posterior subcapsular cataract up to 30% of patients treated.

**Methods:** Patients with NS who have used at least one complete cycle of CS have been selected. They have been referred to Ophtalmology ambulatory and evaluated by three different ophtalmologists, one of them, with large experience in pediatrics. Ophtalmological evaluation included: ocular extrinsical motility, Snellen visual acuity, intraocular pressure, biomicroscopy and direct ophtalmoscopy.

**Results:** Of 107 patients selected, 67 had gone to Ophtalmology ambulatory for evaluation. In general ophtalmologists'evaluation, ophtalmological alterations have not been found in 45 patients (67,2%). Cataract have been diagnosed in 19 patients (28,4%) and signs suggestive of glaucoma appeared in 3 patients (4,4%). In pediatric ophthalmologists' evaluation just 10 patients (10,7%) have presented discrete posterior subcapsular cataract. None of them presented glaucoma or intraocular pressure alterations.

**Conclusions:** Ophtalmological evaluation of children and adolescents with nephrotic syndrome should be performed by professionals specialized in pediatric ophthalmology. Nevertheless, in this group, the prevalence of cataract was lower than that reported in literature for chronic use of steroids(30%).

## Abstract# 408

**Focal Segmental Glomerulosclerosis: Clinical Course and Predictive Factors Related to Treatment in Children and Adolescents Admitted to a Referral Centre from Brazil** R.G. Salum, J.S.S. Diniz, J.M.P. Silva, L.Sé.B. Cardoso, S.V.B. Pinheiro, E.A. Oliveira, E.M. Lima. *Pediatrics, Federal University of Minas Gerais, Belo Horizonte, Brazil.*

**Objectives:** Focal and segmental glomerulosclerosis (FSGS) is one of the most common causes of chronic kidney disease. The objective of this study was to describe the response to treatment in children with FSGS and to correlate it with the clinical an laboratory parameters.

**Methods:** The present work is a retrospective study of 113 children and adolescents between 1 and 18 years with pathological diagnosis of primary FSGS, admitted to the Unit of Pediatric Nephrology at the Federal University of Minas Gerais between 1970 and 2007

**Results:** All patients were initially treated with oral prednisone: 37% achieved complete remission and 22% partial remission. Hematuria (p=0,001) and higher age at the presentation (p=0,034) were associated with poor response to prednisone. Eighty four patients were treated with oral cyclophosphamide (CPM): 50% achieved complete remission and 19% partial remission. Those with nonresponse to initial prednisone had 3.8 times more risk of nonresponse to CPM. Cyclosporin (CsA) was given to 27 patients: 81.5% achieved remission, including 71% of the steroid-resistant group. The level of initial proteinuria was a risk factor associated with nonresponse to CsA in multivariate analysis. Renal survival was better in the group of steroid-sensitive patients (p=0,0002). Response to the CPM or CsA were related to preservation of renal function.

**Conclusions:** Our study showed that initial hematuria is associated with poor response to steroids and CPM represents an alternative for steroid-resistant primary FSGS.

## Abstract# 409

**Epidemiological Profile and Clinical Outcomes of Acute Post-Streptococcal Glomerulonephritis (APSGN) in Children of Brazil** A. Lins, J. Pacheco, K. Freire, C. Sarmento, S. Arruda. *Instituto de Medicina Integral Professor Fernando Figueira, Recife, Pernambuco, Brazil.*

**Objectives:** Analyze the epidemiological profile and clinical outcomes of children diagnosed for APSGN during hospitalization in a hospital in Brazil.

**Methods:** The population of this study included 74 children admitted for APSGN to Instituto de Medicina Integral Professor Fernando Figueira, from March 2009 to February 2010. Diagnostic criteria for APSGN were microscopic or macroscopic hematuria, hypertension, edema and decrease of C3 fraction of complement. Informations were obtained from a questionnaire with caregivers and hospital records data from admission to the end of hospitalization.

**Results:** The children mean ages was 8,2 years. There were more males (60,8%) than females. The skin infection was the most prevalent symptom of recent streptococcal infection (63,5%) followed by upper respiratory tract infection (24,3%), pneumonia (5,4%) and undefined antecedents. Macroscopic hematuria was present in 72,9%, oligoanuria in 62,1%. At urinary analysis, 62,1% had proteinuria. Pyuria was found in 62,1% and cylindruria in 47,2%. The hospitalization due to social and economic factors were 54,1% against clinical indication (45,9%). Renal failure was the most prevalent complication, present at admission in 32,4%. Hypertensive encephalopathy was found in 10,8%, congestive cardiac failure in 8,1%. 24,3% needed to use hypotensive drugs.

**Conclusions:** The APSGN was more frequently founded in males children and after skin infections. Almost a half of hospitalizations were due to social and economic factors, reflecting the brazilian condition of under developing country that needs eradicate the causal agent of APSGN.

## Abstract# 410

**Membranoproliferative Glomerulonephritis (MPGN) Type 1 – A Case for Complement Therapy?** A. Lunn, V. Langlois, C. Licht. *Nephrology, Hospital for Sick Children, Toronto, ON, Canada.*

**Objectives:** Disorders of complement are associated with atypical HUS and MPGN 2. Animal models, C3 deposition GN in humans and reports of complement activation independent of antibody in MPGN 1 led us to the hypothesis that complement dysregulation may be a pathogenic mechanism in MPGN 1.

**Methods:** We present a patient with MPGN 1 who did not respond to conventional therapy and had low C3. Based on our hypothesis we instituted a trial of plasma infusion.

**Results:** A 7 year old male presented with an acute nephritis with low C3. A renal biopsy was consistent with MPGN 1. He was initially treated with steroids and enalapril and his creatinine, albumin and proteinuria normalised. Over the next 5 years he had multiple relapses and only achieved partial remission despite treatment with steroids, enalapril, losartan, mycophenolate mofetil and tacrolimus. Throughout this time his C3 remained low.

In view of his persistent nephrotic state, his low C3 and his poor response to treatment we considered that he may benefit from treatment targetting complement dysregulation. Genetic testing for MCP, Factor H, Factor I and Factor B showed a MCP variant of unknown significance. Further genetic and functional testing is awaited.

A trial of plasma infusions commenced and after one month his C3 normalised. By three months his albumin had also normalised and his proteinuria improved.

**Conclusions:** The clinical and pathological features of our patient and the response to plasma infusion would be consistent with a disorder of complement regulation. We recommend that genetic testing and trials of plasma infusion be considered in patients with MPGN 1 who have persistently low C3 and are unresponsive to conventional therapy.

**DISCLOSURE:** *Langlois, V.*: Grant/Research Support, Astellas Pharma Canada. *Licht, C.*: Grant/Research Support, Optherion; Consultant, Alexion, Optherion.

## Abstract# 411

**Experience of Treating Steroid Resistant Nephrotic Syndrome (SRNS) in a Developing Country** M. Mantan, G.R. Sethi. *Pediatrics, Maulana Azad Medical College, Delhi, Delhi, India; Pediatrics, Maulana Azad Medical College, Delhi, Delhi, India.*

**Objectives:** To look at the short term outcome of patients with SRNS.

**Methods:** Records of all patients with SRNS presenting to the pediatric nephrology outpatient (2004-2009 ) were retrieved. The clinical, biochemical, anthropometric data, details of resistance type, kidney biopsy features and response to different treatment were recorded. Response was defined as complete remission (CR), partial remission (PR) and non response.

**Results:** Forty patients (26 M; 14 F) presented with SRNS. Eighteen children had initial and 22 late resistance. Fourteen (35%) patients had MCD, 12 (30%) FSGS, 5 (12.5%) had mesangioproliferative glomerulonephritis, MPGN in 4 (10%), membranous nephropathy in 3 and lupus nephritis in 2 patients. The median age at onset and presentation was 60 (12-144) and 96 (14-168) months respectively. Of the 31 patients with biopsy changes of MCD, FSGS and MesPGN, 10 received IV cyclophosphamide and oral steroids; 21 received calcineurin inhibitors and oral steroids as the initial therapy. The response rates for CR for patients treated with cyclophosphamide were 40% (4/10) and calcineurin inhibitors were 51.8% (14/27). Almost 85% (23/27) patients achieved CR or PR with calcineurin inhibitors. The median time to follow up of all patients combined was 20 (3-62) months. At the last follow-up 25 patients had steroid sensitive relapses, 7 had partial remission, 5 continued to be SRNS, 2 patient died due to infections and one child was in CKD stage V.

**Conclusions:** Most patients of SRNS respond to calcineurin inhibitors. However in less developed countries intravenous cyclophosphamide is also a reasonable alternative.

## Abstract# 412

**Presence of Congenital Anomalies of Kidney and Urinary Tract (CAKUT): A Predictor of Difficult To Control Nephrotic Syndrome** M. Mantan, G.R. Sethi. *Pediatrics, Maulana Azad Medical College, Delhi, New Delhi, India; Pediatrics, Maulana Azad Medical College, Delhi, New Delhi, India.*

**Objectives:** To look at the course of nephrotic syndrome in children having an underlying CAKUT.

**Methods:** Records of all children who presented (year 2004-2009) with nephrotic syndrome and CAKUT were retrieved. The clinical details, anthropometry, biochemical investigations were recorded. The details of all radionuclear imaging were noted. The course of nephrotic syndrome, details of immunosuppressive therapy and renal biopsy if done was reviewed.

**Results:** A total of 9 patients (8M; 1F) were identified. The median age at onset and presentation of nephrotic syndrome was 48 (16-108) months and 67.3 (17-110) months respectively. The structural abnormalities identified in these patients were horse shoe kidney in one, unilateral PUJO in 2, bilateral grade V VUR in one, bilateral grade II VUR in 1 patient. One child had crossed fused ectopia of left kidney, another had unilateral dysplastic kidney and 2 patients had single kidney. The course of nephrotic syndrome was reviewed. Four children had a FRNS/SDNS course and received oral cyclophosphamide for 3 months. Two children had steroid resistant disease (1 MCD; 1 FSGS on biopsy) and received cyclosporine for attaining remission. Three subjects were infrequent relapsers. The presence of underlying structural abnormality in the above patients was associated with a difficult course of nephrotic syndrome in 67% (6/9) patients. Alternative drugs were required in all these children besides steroids.

**Conclusions:** Presence of CAKUT along with nephrotic syndrome is a predictor of difficult course for such patients. Alternative agents may be required early in the course of the disease.

## Abstract# 413

**Mephedrone-Induced Vasculitis and Segmental Necrotising Glomerulonephritis Mimicking Henoch-Schonlein Purpura and Nephritis** A. Cubero, N.J. Sebire, S.D. Marks. *Department of Paediatric Nephrology, Great Ormond Street Hospital for Children NHS Trust, London, England, United Kingdom.*

**Objectives:** We report a case of vasculitis secondary to mephedrone (amphetamine-like drug) which fulfils the classification criteria of HSP without IgA deposition on skin or renal biopsy.

**Methods:** A 15-year old man presented with a vasculitic rash two hours after taking mephedrone. He developed macroscopic haematuria and lethargy without abdominal pain or joint swelling. He was pale and hypertensive at 144/100mmHg with peripheral and periorbital oedema and purpura on his legs, feet and back. He had no arthritis or joint effusions.

**Results:** He had 3+ proteinuria and 3+ haematuria with urine albumin : creatinine ratio of 237.1mg/mmol. He had haemoglobin of 8.8g/dl with elevated ESR and CRP of 30mm/hour and 68mg/l. His plasma creatinine was 65umol/l (eGFR of 86mls/min/1.73m$^2$) with albumin of 20g/l. He had elevated complement C3 of 2.27g/l with elevated IgA of 3.52g/l which normalised within a month. He had normal C4, IgG and IgM with negative ASOT, ANA, ANCA and autoimmune profile. He had bilateral enlarged echogenic kidneys on renal ultrasound. His skin biopsy showed a leucocytoclastic vasculitis with perivascular deposition of C1q without IgA deposition. His percutaneous renal biopsy showed 61% glomeruli with necrotising segmental lesions and 26% fibrocellular crescents without IgA deposition.

**Conclusions:** He was treated with corticosteroids, enalapril and furosemide, and 3 months later, he has clinically improved with less albuminuria (59.2mg/mmol), normal serum albumin with normal renal function. Although fulfilling the classification criteria for HSP, we present the first case of mephedrone-induced vasculitis.

## Abstract# 414

**Immunosuppressive Treatment Responses in Steroid Resistant Nephrotic Syndrome with Podocine Gene Mutation Positive and Negative Turkish Children** S. Mir, F. Mutlubas, A. Berdeli, O.D. Kara. *Pediatric Nephrology, Ege University Faculty of Medicine, Izmir, Turkey.*

**Objectives:** First line immunosuppressive treatment in steroid resistant nephrotic syndrome (SRNS) in children is still open to discussion. The detection of an NPHS2 mutation affects treatment plan with SRNS. The aim was to compare treatment responses of podocine mutation positive and negative SRNS patients.

**Methods:** 63 SRNS children were included. Mutation analysis was performed in 8exons of NPHS2 gene with direct DNA sequencing. There were 26 children with NPHS2mutations and 37 children with no mutation. Immunosuprresives were various such as cyclosporinA(CsA), cyclophosphamide(CyC), Mendoza protocole(MP), Plasmaphresis(PP), mycophenolate-mophetil(MMF). Responses were as complete(CR), partial remission(PR) and no response(NR).

**Results:** The most prefered drug was CyC and the most long-lasting used drug was CsA. PPand MMF were used (n=2)with NR. The remission rate(RR) of MP was found to be similar in groups (40%-37%) with steroid side effects. While RR with CyC in groups were similar, CsA RR was significantly high in patients with no mutation(75%). The duration ofCsA was longer in group with no mutation. No side-effect detected with both CsA-CyC. The patients,showed CR withCsA, in mutation group were carrying single allele mutation.

**Conclusions:** This study highlights CsA is stil an important drug in children with no mutation in long-lasting usage without side effects. MP may be used as last choice. The efficacy and safety of CyC treatment is not higher thanCsA. The patients who were carrying single allele mutation and no mutation are worth to treat to achieve remission or prolongation of process going to ESRD.

## Abstract# 415

**Pediatric Lupus Nephritis: 12 Year Experience from a Single Centre** A. Mohapatra,[1] G. Basu,[1] V.M. Annapandian,[1] V.G. David,[1] S. Madhivanan,[1] S. Varughese,[1] A. Korula,[2] G.T. John,[1] C.K. Jacob,[1] V. Tamilarasi.[1] *[1]Nephrology, Christian Medical College, Vellore, Tamil Nadu, India; [2]Pathology, Christian Medical College, Vellore, Tamil Nadu, India.*

**Objectives:** The clinicopathological characteristics & outcomes in various classes of treated lupus nephritis in children.

**Methods:** Consecutive patients, age ≤ 18 years, with diagnosis of lupus nephritis by renal biopsy at our centre(1997- 2008) and at least one follow up, were studied retrospectively.

**Results:** 120 (7.5%) (M: F=1:6.9, mean age 15.5±2.3years) children had lupus nephritis.Class IV lupus nephritis was predominant (64.2%).Hypertension, hematuria, nephrotic range proteinuria were seen in 20, 11 & 79.8% respectively. Mean creatinine & serum albumin were 1.3(0.4-15.6)mg % & 2.7(0.4-4.6)gm% respectively. Elevated anti dsDNA titre and low complements were common in class III &IV.75 were followed up 23(0.7 - 106.9) months & median time for remission was 6.9 (0.7-75.2) months. Sustained complete & partial remission were observed in Class II (62.5, 18.7%), III (100, 0%), IV (56.8, 4.5%) & V(66.6, 0%) respectively.Poor response /progression to end stage renal disease were seen in Class II, III, IV & V at 18.7, 0, 31.7& 33.3% respectively. Predictors of poor response were nephrotic range proteinuria (p=0.003, O.R. 5.5(1.4-21.8)), hypoalbuminemia (p=0.030, O.R 3.3(1.1-9.5))& lack of prior steroid therapy (p=0.026, O.R 4.2(1.2-15.1)).

**Conclusions:** Nephrotic range proteinuria, hypertension, renal dysfunction, elevated anti dsDNA & hypocomplementemia were indicative of class IV lupus nephritis.Poorer prognosis, predicted by nephrotic range proteinuria, hypoalbuminemia and absence of prior steroid therapy at presentation.

## Abstract# 416

**Childhood Lupus Nephritis (LN): Mycophenolate Mofetil (MMF) as Induction and Maintenance Therapy** D. Molino,[1] F. Nuzzi,[1] M. D'Armiento,[2] M.M. Balletta,[2] M. Alessio,[2] C. Pecoraro.[1] *[1]NephroUrology, Santobono Hospital, Naples, Italy; [2]University Federico II, Naples, Italy.*

**Objectives:** To evaluate the effectiveness and safety of MMF for inducing and maintaining remission of childhood LN.

**Methods:** 27 children with LN, mean age: 3.3 yrs; M/F: 6:1; Symptoms: proteinuria (89%), hematuria (100%), Nephrotic Syndrome (48%), ARF (22%), Hypertension (33%). Renal Biopsies, before MMF, showed the following classes (Weening): IV in 14 cases, III in 3, II in 8, V in 1, VI in 1. Treatment: i.v. Metilprednisolone pulse for three consecutive days and then MMF (plus oral Prednisone (P)) at mean dose: 29±7.7 mg/Kg/day.

**Results:** At a mean follow up 4.5 yrs (0.9-7.7) proteinuria was absent in 7 pts,< 0.5 gr/day in 9, < 1g/day in 6, < 2g/day in 3, > 2g/day in 2. Renal funcion normalized in 5/6 pts with ARF and worsened in the remaining one. SLEDAI score decreased significantly ($p<0.01$), plasma C3 levels normalized in 66%. In 10 pts a second renal biopsy, after 2 years MMF treatment, was performed: histopathological activity indices reduced significantly (8.76± 2.55 vs 5.3±61.97), chronicity index did not change. P was tapered and, after 5.6±2.5 months, stopped in 15 pts; 12 pts were receiving P at a mean dose 0.3 mg/Kg. 5 pts showed proteinuric flares effectively treated with the increase of oral P. Clinical signs of hypercorticysm dramatically improved. No haematological side effects, in 3 pts gastrointestinal symptoms occurred; just one patient had Herpes Zoster infection.

**Conclusions:** MMF represents a good alternative to traditional therapy of childhood LN allowing, as monotherapy too, a clinical and histopathological control of disease activity without significant side effects.

## Abstract# 417

**A 20-Year Single Centre Experience of Congential and Infantile Nephrotic Syndrome** G. Montini, D. Bockenhauer, L Rees, N. Sebire, K. Tullus, W. van't Hoff, A. Waters, S. Marks. *Pediatric Nephrology, Great Ormond Street Hospital, London, United Kingdom.*

**Objectives:** To determine the clinico-pathological correlations of congenital and infantile nephrotic syndrome (NS).

**Methods:** Retrospective clinical and histological review of patients presenting with NS before the age of two years, from 1990 to 2009.

**Results:** 58 children aged 0.1-23.6 (median 1.8) months at presentation. The histological diagnoses are in the table, with additional four cases, one each of Dense Deposit Disease, Membranous, Collapsing and Mesangial Proliferative GN.

| | n | U albumine:creatinine (mg/mmol) at presentation | Age (months) at presentation |
|---|---|---|---|
| Finnish | 27 | 4301 (460-8460) | 1.4 (0.1-4.8) |
| DMS | 11 | 3840 (1735-7600) | 3.4 (0.2-18.3) |
| FSGS | 11 | 3055 (345-3444) | 13.7 (0.9-23.6) |
| MCD | 5 | 3178 (1225-7850) | 9 (0.1-23.8) |

Age at presentation was younger in the Finnish group; proteinuria was higher in the Finnish and DMS groups ($p$ 0.01 and 0.04 respectively). Extra-renal manifestations were common in the Finnish and DMS groups: 10 cases of major GI disorders, 8 major neurologic complications and 5 major congenital heart disease. The mortality rate was 26% (7) in the Finnish group at median follow-up of 49 months with 4 children going into remission at 2-28 months. These patients had normal renal function 1-15 years later. The mortality rate was 36% (4) and 9% (1) in the DMS and FSGS groups at median follow-up of 55 and 26 months respectively. One patient died from fatal haemorrhagic complications from the renal biopsy.

**Conclusions:** NS during the first two years of life is a rare but serious disease, with a higher mortality and complication rate in the Finnish and DMS groups.

## Abstract# 418

**Specific Podocin Mutations with Clinical Features in Steroid-Resistant Nephrotic Syndrome in Turkish Children** F. Mutlubas, S. Mir, A. Berdeli, N. Dincel. *Pediatric Nephrology, Ege University Faculty of Medicine, Izmir, Turkey.*

**Objectives:** Mutations in podocin(*NPHS2*) cause OR steroid-resistant nephrotic syndrome SRNS). Different populations have different types of NPHS2 mutations and clinical progress. There are also various behavior of the same mutation in different populations. The aim was to display types of NPHS2gene mutation with clinical behaviour in Turkish children.

**Methods:** Steroid resistance was accepted as no achievement with prednisolone (2 mg/kg/d 4 weeks)in primary nephrotic syndrome patients. 63children (38boys and 25girls) with SRNS were included. Mutation analysis was performed in all 8 exons of NPHS2 gene with direct DNAsequencing method.

**Results:** Podocin mutations were present in 41.3% (26/63) of patients and 17//26 were in 2 alleles.Nonsense, frameshift, deletion and missense mutations were in ratios of, 11.5, 11.5, 7.7 and 69.3%, respectively. Nonsense and frameshift mutations were related with early-onset(1,2 years). Patients with R138Q mutations displayed early and late-onset SRNS,developed ESRD. R168H was found to be related early-onset and rapidly ESRD. R229Q mutation were found in 8/26patients and 2 of them reached to ESRD with late-onset. V180M was detected in early-onset SRNS and found to be related remission after CsA.

**Conclusions:** Nonsense and frameshift mutations cause early-onset SRNSand ESRD. Missense mutations such as R138Q and R168H in homozygous associate with early-onset or infantil NS with progressing to ESRD. R229Q seems to be related with late-onset NS and ESRD. Contraversy to literature,V180M may results with early-onset FSGS. These different behaviour of the mutations may originate from additional factors that can modify the phenotype in years.

## Abstract# 419

**Nonischemic Nephrotic Syndrome Associated with Primary Antiphospholipid Syndrome** H.J. Cho,[1] E.G. Soon,[1] M.K. Namgoong.[1] *[1]Pediatrics, Yonsei Universoty, Wonju College of Medicine, Wonju, Korea; [2]Pediatrics, Yonsei Universoty, Wonju College of Medicine, Wonju, Korea; [3]Pediatrics, Yonsei Universoty, Wonju College of Medicine, Wonju, Korea.*

**Objectives:** Antiphospholipid syndrome(APS) nephropathy was the result of vascular obstruction of renal vessles. There was not routinely accepted an immune glomerulonephritis as one of primary antiphospholipid syndrome nephropathy. We present the immune glomerulonephritis with nephrotic syndrome, of primary antiphospholipid syndrome.

**Results:** A 11 year old female child was admitted with generalized edema and severe visual disturbances with nearly blindness for 1 month. BP was 120/80mmHg, On the physical examination, there was eyelid swelling and pitting edema. There's no cranial nerve abnormality. On the fundus examination, both optic disc severe swelling and blunted disc margin with star shaped hard exudate on macula were noted. Serum protein and albumin were 4.4/2.1 g/dL. Serum cholesterol and triglyceride were 342/224g/dL. CBC was as follows: WBC 8,360, Hb 11.7g/dL and platelet 130,000/$mm^3$. PTT was 50.6sec (control 32.3sec). Brain MRI was normal. A diagnosis of primary APS was made on the basis of antiphospholipid syndrome IgG/IgM (+/-), Cardiolipin IgG/IgM (+/+), ANA(-), anti-DNA Ab(-), anti-sm(-), anti-ENA(-), C3/C4 98.6/19.8mg/dL. Kidney tissue showed diffuse proliferative glomerulonephritis with subepithelial electron dense deposits. She has been treated with coumadin, steroid and cyclosphosphamide, but still remained proteinuria without edema. Her vision was much improved.

**Conclusions:** We suggest that there should be a need for concern about the possibility of the development of immune glomerulonephritis in primary antiphospholipid syndrome.

## Abstract# 420

**An Unusual Case of Post Infectious Glomerulonephritis Secondary to *Bartonella quintana* Infection Presenting with Severe Acute Kidney Injury** Y.H. Ng, S.M. Chao. *Department of Paediatrics, KK Women's & Children's Hopsital, Singapore, Singapore.*

**Objectives:** *Bartonella quintana* historically caused Trench Fever. We report an unusual paediatric case with post infectious GN who presented with severe AKI due to Bartonellosis.

**Methods:** M is a 11 years old Malay boy who presented with bilateral massive cervical lymphadenopathy, poor feeding and oliguria. Investigations showed severe AKI: serum creatinine 627umol/L, urea 65.3*, potassium 5.1*, phosphate 3.8* (*mmol/L). Haemodialyis (CVVHD) was commenced for the next 4 days. Serology test for *Barotonella quintana* was positive (Ig M titre ≥ 20). M subsequently developed atypical nephrotic syndrome: hypoalbuminemia, anasarca, hyperlipidemia, nephrotic range proteinuria, hypertension and haematuria. Complement 3 (C3) level was low (C4 normal). Ig A, dsDNA, ANCA, anti GBM and ASOT were negative. Renal biopsy showed diffuse proliferative GN with 38% cellular crescents on LM and isolated subepithelial hump-like electron dense deposits on EM consistent with acute post infectious GN. M was managed conservatively with fluid restriction and antihypertensives with subsequent diuresis, resolution of hypertension and proteinuria. He was discharged well on Day 28 of illness with normalization of C3 and renal function.

**Results:**

**Conclusions:** *Bartonella quintana* is recognized as an emerging pathogen among the homeless and immunocompromised populations but remains rare as a cause of post infectious GN in healthy paediatric patients. The clinical course for most patients with post infectious GN is excellent with full spontaneous recovery. We report a first such case in a healthy 11 year old child who represented with severe AKI with full recovery of renal function.

## Abstract# 421

**Cyclophosphamide Treatment for Diffuse Proliferative Lupus Glomerulonephritis: The Role of Reversal of Renal Histiology** D. Nguyen thi Ngoc, L. Huynh Thoai. *Nephrology, Children's Hospital 1, Ho Chi Minh, Viet Nam.*

**Objectives:** In patients with systemic lupus erythematosus (SLE), renal biopsy not only guides to therapy but also predicts to response and relapse.
**Methods:** Prospective study
**Results:** We described 51 patients with SLE, admitted to the Children Hospital 1 in the period from 1/2003 to 1/2008, whose initial biopsy (Bx1) showed proliferative lupus nephritis class III, IV by ISN/RPS (International Society of Nephrology/ Renal Pathology Society) and who received 6 monthly doses of intravenously administered cyclophosphamide. Repeat renal biopsy (Bx2) was done at sixth dose. Bx2 showed histological improvement by ISN/RPS classification in 35 children (17 complete remission; 15 partial remission, 3 non response), 14 patients were unchanged (6 complete remission; 7 partial remission, 1 non response), and 2 patients were worse (both were partial remission). Activity index (AI) decreased 100% of patients in complete remission group, 62.5% of patients in partial remission (p=0.03). 11 (23.4%) patients relapsed after CYC induction, mean relapse time was 13.7 months. Non-improved AI occurred in 36% of relapse patients and 13.8 % of non relapse patients.
**Conclusions:** There were no correlation between the clinical response and histological improvement after 6 months CYC IV. Improved AI at Bx2 showed response to treatment, those who did not improve are at risk for flares.

## Abstract# 422

**Treatment Strategy and Outcome for Henoch-Schonlein Purpura Nephritis** T. Ninchoji,[1] Y. Hashimura,[1] H. Kaito,[1] K. Nozu,[1] K. Kanda,[2] I. Kamioka,[3] Y. Shima,[4] K. Hamahira,[5] K. Nakanishi,[4] R. Tanaka,[2] K. Iijima,[1] N. Yoshikawa,[4] M. Matsuo.[1] *[1]Kobe University Hospital, Kobe, Japan; [2]Hyogo Prefectural Kobe Children's Hospital, Kobe, Japan; [3]Kakogawa Municipal Hospital, Kakogawa, Japan; [4]Wakayama Medical University, Wakayama, Japan; [5]Himeji Red-Cress Hospital, Himeji, Japan.*

**Objectives:** The management of Henoch-Schonlein purpura nephritis (HSPN) is still controversial. Some studies reported steroids or immunosuppressants were effective, but indication of these drugs remains uncertain. Here we retrospectively examine the efficacy of treatment in HSPN.
**Methods:** Renal biopsy was performed in patients with nephrotic syndrome or persistent proteinuria > 3 months. We divided patients into two groups and treated according to severity. GroupA (31 patients), or the mild case such as ≤ gradeIII(ISKDC) and serum albumin (ALB) > 2.5g/dl, was treated with ACE-I and/or ARB. GroupB (19 patients), or the severe case such as > gradeIII or ALB ≤ 2.5g/dl, received multiple combinated therapy(MCT) consisting of prednisolone, azathioprine/mizoribine, warfarin and dipyridamole. We assessed the effect of therapies by urinary protein/creatinine ratio and estimated GFR(EGFR).
**Results:** After each treatment, all the patients disappeared proteinuria and kept EGFR normal. The mean period of proteinuria after therapeutic intervention was 7.4±1.9 and 6.4±1.2 months, respectively. There were no adverse events of therapy.
**Conclusions:** These findings suggested that the MCT was used effectively in the treatment of severe case. It is necessary to emphasize that the MCT should be used for only severe cases because mild cases will improve enough with ACE-I/ ARB.

## Abstract# 423

**Renal Expression of Response Gene to Complement-32 in Children with IgA Nephropathy** X.-L. Niu,[1] W.-Y. Huang,[1] X.-G. Liu.[2] *[1]Department of Nephrology and Rheumatology, Children's Hospital of Fudan University, Shanghai, China; [2]Department of Pathology, Shanghai Medical College of Fudan University, Shanghai, China.*

**Objectives:** RGC-32 played a critical role in TGF-induced EMT of renal tubular cells. This study aimed to evaluate the expression and clinical significance of RGC-32 in children with IgA nephropathy.
**Methods:** Control group consisted of 6 patients with TBMN. Experimental group consisted of 45 patients with IgAN. The expression of RGC-32 was observed. And the staining of α-SMA and TGF-β1 were observed.
**Results:** 1. The location of RGC-32 in renal tissues: RGC-32 majorly expressed in cytoplasm of proximal tubular and distal renal tubular, while the RGC-32 did not express in glomerular. 2. The degrees of expression of RGC-32 was positive correlation with that of renal lesions and tubulointerstitial injury in children with IgAN (***P***<0.05) in our study. And the obviously positive correlation were also found in the degrees of expression of RGC-32 between the expression of α-SMA, TGF-β1 in children with IgAN. (***r***=0.325, 0.533, 0.294, respectively, ***P***<0.05). 3. The expression of RGC-32 had no relationship with Serum creatinine and urinary NAG, microalbuminuria, micro-globulin, α1-microglobulin in children with IgAN.(r=0.114, -0.116, -0.139, -0.177, -0.029, ***P***>0.05).
**Conclusions:** RGC-32 protein located in cytoplasm of proximal tubular and distal renal tubular in renal tissues firstly. The expression of RGC-32 was positive correlation with the degree of renal lesions and tubulointerstitial injury in children with IgAN, and it is concordance to the expression of α-SMA and TGF-β1. Thus it was hinted that RGC-32 may participate in the course of TGF-induced EMT of renal tubular cells.

## Abstract# 424

**Randomized Controlled Trial of Oral Versus Intravenous Cyclophosphamide in Idiopathic Steroid Resistant Nephrotic Syndrome** A. Ohri, A. Phatarpekar, U. Ali, Y. Tembekar. *Nephrology Division, B.J.Wadia Hospital For Children, Mumbai, Maharashtra, India.*

**Objectives:** To compare efficacy of oral versus IV (CP) in inducing remission in patients with idiopathic SRNS and to compare side effects of 2 regimes.
**Methods:** 35 children with biopsy proven idiopathic SRNS were randomized in to 2 groups to receive the following treatment Group1 Oral CP 2.5mg/kg/daily for 12 weeks.
Group 2 IV CP 750mg/m2 infusion monthly for 6 months.Both groups received alternate day steroids in tapering doses.Patients on immunosuppressive drugs other than steroids in last 6 months were excluded.Remission was assessed at end of therapy.(CR) Complete remission was defined as Up/Uc<0.1(PR)Partial remission as Up/Uc of 0.1-2, serum albumin >2.5gm/dl,no edemaAll other patients were classified as no response.
**Results:** 35 chlidren (24 M),1-12 years, mean 5.12±2.9 years were included. 21(60%) had MCNS, 8(23%)FSGS and 6(17%)mesangioproliferative GN. Gp1had 20 cases and Gp 2 had 15. Baseline age, sex, histological and biochemical parameters were similar in the 2 groups.Total 16 children achieved CR;8/20(40%) in Gp 1,8/15 (53.3%) in Gp 2.PR seen in 5/20(25%) in Gp 1, 2/15(13.5%) in Gp 2.7/20 in Gp 1,5/15 in Gp 2 did not respond. Rates of CR (p=0.6) and PR (p=1)were not different in 2 groups. Time to remit was shorter in Gp1 than Gp 2 (44.6 ± 30/ 87.5± 25 days) t=0.008.Response to CP (Gp 1+Gp 2) in MCNS and Non MCNS was comparable.Side effects were severe infections (1 in Gp 1,5 in Gp 2),transient neutropenia 1 and alopecia 5 which were same in both groups.None died or had hemorrhagic cystitis.
**Conclusions:** Overall two-third of SRNS patients responded to CP. Oral and IV CP were equally efficacious and safe in idiopathic SRNS in children.

## Abstract# 425

**Serum Indoxyl Sulfate as an Early Marker for Detecting Chronic Cyclosporine Nephrotoxicity** D. Umino,[1] Y. Ohtomo,[1] S. Fujinaga,[2] S. Niijima,[1] T. Shimizu.[3] *[1]Department of Pediatrics, Juntendo University Nerima Hospital, Tokyo - Nerima, Japan; [2]Division of Nephrology, Saitama Children's Medical Center, Saitama - Iwatsuki, Japan; [3]Department of Pediatrics, Juntendo University, Faculty of Medicine, Tokyo - Bunkyo, Japan.*

**Objectives:** Cyclosporine A (CsA) is an effective agent for frequently relapsing steroid-dependent nephrotic syndrome (FR-SDNS), but its use can also be complicated by renal toxicity. Because no biochemical markers from urine or blood samples have yet been established for detecting CsA-induced renal injury to date, repeated renal biopsies are therefore required for all patients with long-term CsA treatment. The purpose of the present study was therefore to detect early change of CsA nephropathy (CsAN) using blood samples.
**Methods:** Several biochemical markers were analyzed in an attempt to examine the renal function in 24 patients with FR-SDNS who had been treated with CsA. Those included serum cystatin C and indoxyl sulfate, as well as creatinine and b2-microglobulin.
**Results:** Renal biopsy findings indicated chronic CsAN in 13 of the 24 patients. Among those markers, only serum indoxyl sulfate was significantly elevated in patients with CsAN.
**Conclusions:** It may be possible for measurement of serum indoxyl sulfate level to replace repeated renal biopsies in evaluation of chronic CsAN in pediatric patients with FR-SDNS.

## Abstract# 426

**A Case Report of May-Hegglin Anomaly with Immune Complex-Related Nephropathy** Y. Ohtsuka,[1] T. Sato,[1] K. Taisuke,[2] Y. Sado,[4] H. Kawachi,[5] K. Izuhara,[2] Y. Hamasaki.[1] *[1]Faculty of Medicine, Saga University, Saga, Japan; [2]Shigei Medical Research Institute, Okayama, Japan; [3]Niigata University Graduate School of Medical and Dental Sciences, Niigata, Japan.*

**Objectives:** May-Hegglin anomaly (MHA) is a rare hereditary disease with macrothrombocytopenia and leukocyte inclusions. We showed brothers of MHA, and studied pathological condition using genetic and pathological analysis.
**Methods:** The brothers (8, 5 years old boys) had macrothrombocytopenia. Hematuria and proteinuria was detected in the elder boy only. A genetic analysis

of MYH9 mutation and hematologic evaluation were performed. A renal biopsy was performed on the elder brother. Immunofluorescence microscopy (IF) added anti-alpha1 to 6 antibody of collagen type 4, monoclonal antibody against non-muscle myosin heavy chain IIA (NMMHC-IIA) and antibody of nehrin and podocin.
**Results:** In blood, electron microscopy (EM) of leukocyte inclusion bodies revealed parallel filaments. IF showed NMMHC-IIA antibodies in 1 leukocyte inclusion bodies. These findings consistent with MHA and they were identified to express the MYH9 mutation, D1424H. In the elder boy, the histology revealed mesangial proliferative glomerulonephritis with granular deposits of IgG and C1q. EM showed the dense deposits located in subendothelial cells, mesangial cells and Bowman's capsule. NMMHC-IIA antibodies were localized in podocyte and endothelial cells. The expression of nephrin and podocin was normal. His condition had a temporary effect for steroid therapy, but became refractory proteinuria later.
**Conclusions:** About immune complex-related nephropathy, we concluded that an inflammatory mechanism occur separately from MHA.

## Abstract# 427

**Transient Proteinuria Due to Podocyte Injury under Chemotherapy in a Boy with Wilms Tumor** S. Okamoto,[1] K. Hirai,[1] T. Shimizu,[1] T. Morimoto,[1] F. Niimura,[1] H. Mochizuki,[1] S. Ueno.[2] *[1]Pediatrics, Tokai University School of Medicine, Isehara, Kanagawa, Japan; [2]Pediatric Surgery, Tokai University School of Medicine, Isehara, Kanagawa, Japan.*
**Objectives:** To evaluate the podocyte injury under chemotherapy in a boy, presenting with transient heavy proteinuria during the treatment for bilateral Wilms tumor.
**Methods:** The resected kidney was evaluated in a 1 year-old boy, who presented with gross hematuria and abdominal mass. A diagnosis of Wilms tumor was made, as he had a huge tumor (15 cm) in the right kidney, a smaller one (3 cm) in the left, and multiple metastases in the lungs. Ambiguous genitalia was absent. Prior to surgery, he was treated with a series of chemotherapy with vincristine, actinomycin D and adriamycin. Then, heavy proteinuria and hypertension developed. Serum Alb decreased to 2.1 g/dl as the proteinuria increased to 30 in Pr/Cr (mg/mg) ratio. He underwent right nephrectomy after two month-long chemotherapy. The proteinuria gradually decreased, and the serum Alb increased to 3.6 g/dl by 2 months after surgery. His renal function was stable. His hypertension was controlled by ARB and Ca blocker, and nephrectomy seemed also effective. Genetic analysis of the tumor revealed a homizygous mutation of R390X in WT1 gene.
**Results:** FGS was noted in 3 out of 50 glomeruli. Accumulation of podocytes around the collapsing tuft, and hyaline droplets in podocytes were observed,suggesting the podocyte injury. EM revealed foot process effacement in some capillary walls.
**Conclusions:** The proteinuria in our patient was presumably induced by the chemotherapy and hypertension, which damaged the podocytes, already predisposed to some fragility due to WT1 mutation.

## Abstract# 428

**Mycophenolate Therapy in Children with Idiopathic Steroid-Resistant Nephrotic Syndrome. Valencia, Venezuela** N. Orta, V. Coronel, E. Lara, L. Dominguez, C. Uviedo, P. Zibaoui, Y. Lunar. *Service of Pediatric Nephrology, University of Carabobo/Children´s Hospital "J Lizarraga", Valencia, Venezuela.*
**Objectives:** Idiopathic Nephrotic Syndrome is a frequent glomerular disease in childhood; a number of cases are resistant(SRNS) which is a clinical challenge. Objective: to evaluate the response to mycophenolate mofetil (MMF) in children with SRNS.
**Methods:** We recruited 42 children (26 girls, 16 boys with SRNS), age 3.2 +/- 4.4 years. All patients received at least one course of 8-12 weeks of therapy with oral prednisone; renal biopsy was performed in 37/42 cases prior to the administration of MMF. The dose of MMF was 600 mgs/m2/ day for 6-18 months and prednisone was reduced stepwise. Patients were evaluated clinically periodically, and proteinuria and serum creatinine, among others, were checked every 3 months.
**Results:** 27 patients had complete remission, 9 partial remission and 6 no response. Mean follow up was 9.2 +/- 4.8 months. Of the responders 85% had either minimal change disease or mesangioproliferative nephritis (MGN) (22 cases), 2 focal segmental glomerulosclerosis (FGS); all but one of the partially responders had MGN and the no responders corresponded to 4 FGS, 1 membranous nephropathy and 1 MGN. Evaluation of 26 patients who completed the 18 months treatment showed, after a additional mean 4. 2 months period of follow up, that 18(69%) maintained in remission and 8 had full or partial relapse of proteinuria (31%)
**Conclusions:** This reports shows that MMF is a good alternative for treating children with SRNS, achieving either a complete or partial remission in the majority of cases and stabilization in almost 70% of them.

## Abstract# 429

**Early Corticosteroid Treatment (CT) in Children with Iga Nephropathy (IGAN): A Randomized and Controlled Trial** F. Nuzzi,[1] M. D'Armiento,[2] M.M. Balletta,[2] G. Malgieri,[1] C. Pecoraro.[1] *[1]NephroUrology, Santobono Hospital, Naples, Italy; [2]University Federico II, Naples, Italy.*
**Objectives:** To evaluate if an early CT, with respect to the diagnosis time, can influence the course of childhood IGAN.
**Methods:** Children with IGAN proven by biopsy performed as soon as possible after the disease onset. CT protocol: Metilprednisolone 1g/1.73 m2 BA for 3 consecutive days, followed by oral prednisone (P) 0.5mg/Kg/day for a month and then on alternate days for the following 5 months.
**Results:** Treated group (P): 15 children; Control group (C) 12 children age matched. Kidney biopsy was performed after 1.7 (group P) and 1.6 years (group C) from the presumed IGAN onset. Pathological classes (Lee) in group P and C, respectively: class I: 1 and 3 pts, class II: 9 and 8 pts and class III (3 and 3 pts). Mean follow up: 26.8 and 29.8 months in IgAN and C groups, respectively. GFR, A.P. and serum C3/IgA ratio were not different between the groups both at start and at the end of followup and inside each group at the start compared with the end of follow up. Mean basal proteinuria: 0.64 g/day in group P and 0.16 g/day in group C; at last followup mean proteinuria was significantly decreased in group P (0.21 g/day) and unchanged in group C (0.26g/day). During followup 2 pts of group P and 5 pts of group C showed some episodes of gross hematuria. At last follow up, microscopic hematuria was absent in 5 pts of group P and in 1 pts of group C; in the remaining ones it, estimated as +1 to +5, was, as mean value, + 2.5 in group P and +3.75 in group C.
**Conclusions:** Our study indicates that CT administered as soon as possible with respect to the IgAN onset can influence positevely the course of the disease.

## Abstract# 430

**Renal Survival in Pediatric Patients with Iga Nephropathy: A 20 Year Follow-Up** M.-G. Penido, M.-V. Freitas, M. Tavares, E. Soares, F. Trindade, M. Ferreira, C. Bernardes. *Pediatric Nephrology Unity, Medicine Faculty Federal University of Minas Gerais, Belo Horizonte, Brazil.*
**Objectives:** To evaluate the evolution of renal function in children and adolescents with IgAN followed between 1987 and 2007, and to identify unusual risk factors for progressive loss of renal function.
**Methods:** It was a retrospective cohort study. Variables of interest were: end stage renal disease (ESRD), evidences of progressive chronic renal disease (CKD), protein/creatinine ratio>0.5 (P1), protein/creatinine ratio>2. (P2), high blood pressure (HBP), gross hematuria (GH), microhematuria (MH), familial history of renal replacement therapy (FHRRT), age, gender. Tests: Kaplan-Meyer survival curve and Cox-regression model.
**Results:** The study evaluated 61 children, 51% male, median age of 8yr and median time of follow-up of 12yr. At the time of last evaluation, 4(7%) had ESRD, 10(16%) CKD, 25(41%) P1, 23(38%) P2, 13(21%) HBP. Cox regression model identified the age of onset of clinical symptoms as predictive of ESRD (RR 1.6; CI 95% 1.1-2.4; p=0.016). The risk of P1 in children aged 8 yr or older at onset of clinical manifestation was 2.4 times higher than those younger than 8 (RR 2.4; CI 95% 1.0-5.8; p=0.05). For those with GH as initial clinical presentation, the risk of developing P1 was 5-fold higher than those with MH (RR 5.1; CI 95% 1.5-17.5; p=0.009). FHDRRT increased the risk of P2 by 3.4 times (RR 3.4; 95% CI 1.4-8.3; p=0.006).
**Conclusions:** 29(48%) patients developed CKD and 4(4%) developed ESRD; age of clinical onset of the disease, hematuria and familial history of renal replacement therapy were significantly associated with risk of progressive loss of renal function.

## Abstract# 431

**Impaired Ability of GCs To Induce IL-10 and/or Inhibit TNF-alfa Secretion by LPS-Stimulated PMBC Might Underlie Modifications in Steroid Sensitivity of Patients with INS** K. Szilagyi, L. Podracka, J. Mojziš, L. Mirossay. *1Department of Pharmacology, Faculty of Medicine, UPJS, Kosice, Slovakia (Slovak Republic); 1.Department of Paediatrics, Faculty of Medicine, UPJS, Košice, Slovakia (Slovak Republic).*
**Objectives:** To investigate wheather IL-10 and TNF-alfa might determine a response to glucocorticoids (GCs) in children with idiopathic nephrotic syndrome (INS).
**Methods:** 43 patients with INS (mean age 9.8 y) and 13 healthy children (mean age 14.5 y) were enrolled into the study. Patients were classified according to GCs response as responders RE (16), partial responders PR (19) and non-responders NR (8) and also subdivided based on relapse/remission of NS. IL-10 and TNF-alfa was measured and IL-10/TNF-alfa ratio was determined after LPS stimulation and/or dexamethasone. Percentage of IL-10/TNF-alfa increase by dexamethasone was calculated.
**Results:** Significantly higher IL-10/TNF-alfa after steroids was observed in relapsing RE than in PR and/or NR. Dexamethasone concentration of $10^{-8}$M

significantly increased IL-10/TNF-alfa ratio in relapsing RE in comparison to relapsing NR (p=0.049). Higher dexamethasone concentration of $10^{-7}$ M increased this ratio in relapsing RE in comparison to both PR and NR (p<0.05). These changes were not observed in the remission. Stimulated PMBC of PR or NR patients in relapse failed to increase IL-10/TNF-alfa after dexamethasone treatment in contrast to the RE group.
**Conclusions:** We hypothesized that impaired ability of GCs to induce IL-10 and/or inhibit TNF-alfa secretion by LPS-stimulated PMBC might underlie modifications in steroid sensitivity of patients with INS.
Supported by Norway grant through the EEA Financial Mechanism.

## Abstract# 432

**Hyperhomocysteinaemia and MTHFR C677T Gene Polymorphism in Children with Steroid-Resistant Nephrotic Syndrome (SRNS)**
L. Prikhodina,[1] T. Vinogradova,[1] N. Poltavets,[2] A. Polykov,[2] V. Dlin.[1]
*[1]Pediatric Nephrology, Research Institute of Pediatrics & Children Surgery, Moscow, Russian Federation; [2]DNA Laboratory, Research Center for Medical Genetics, Moscow, Russian Federation.*
**Objectives:** The study was conducted to investigate the relationship between total homocysteine (tHcy) level and MTHFR C677T gene polymorphism in children with SRNS.
**Methods:** 44 children (25M/19F) median age 13.5 (9.5-16.0) years with biopsy-proven SRNS (mesangial proliferative glomerulonephritis (n=17), FSGS (n=15), MPGN (n=11), membranous nephropathy (n=1)) were studied. tHcy level (>10 µmol/L) investigated by immunoassay analysis was considered as hyperhomocysteinaemia (HHcy). MTHFR C677T genotypes were investigated in all pts and 50 healthy subjects as controls by PCR and RFLP analysis.
**Results:** The allele frequency and genotype distribution of the MTHFR gene in SRNS pts and controls were in Hardy-Weinberg equilibrium (p>0.05). SRNS pts with MTHFR genotypes had no differences in tHcy level: C/C:C/T:T/T=11.1 (7.4-14.7):12.9 (7.8-15.3):10.3 (8.1-16.1) µmol/L and in frequency of Hhcy: 47:60:50(%) (p>0.05). Pts with stage II CKD (n=21) compare with stage I (n=22) had significantly higher tHcy level: 14.0 (11.2-17.8) vs. 8.0 (7.0-12.7) µmol/L (p=0.01) and Hhcy: 81% vs. 27% (p=0.0005). There was an inverse correlation between tHcy level and eGFR (r=-0.49; p=0.00085). Hhcy was associated with declining of eGFR in children with SRNS: HR 2.97 (95% CI: 1.46-6.1) (p=0.004).
**Conclusions:** Renal function was the strongest determinant for tHcy level in children with SRNS. There were no apparent association between tHcy level and MTHFR C677T gene polymorphism in childhood SRNS.

## Abstract# 433

**Tacrolimus Therapy in Children with Steroid-Resistant Nephrotic Syndrome (SRNS)** L. Prikhodina, O. Turpitko, V. Dlin, M. Ignatova. *Pediatric Nephrology, Research Institute of Pediatrics & Children Surgery, Moscow, Russian Federation.*
**Objectives:** The study was conducted to evaluate the efficacy and safety of tacrolimus (TAC) in children with idiopathic SRNS.
**Methods:** Seven children (6 girls/1 boy) median age 12.5 (range: 9.5-13.0) years with initial SRNS were treated with a 12 months course of TAC. The hystological assessment revealed FSGS (n=4), mesangial proliferative glomerulonephritis (MsPGN) (n=2) and MPGN (n=1). Initial TAC dose was 0.1 mg/kg/24h (target through level 5.0-10.0 ng/mL) in combination with alternate-day prednisolone dose 0.5-1.0 mg/kg/24h for 3 months. The median time from the diagnosis to TAC initiation was 28.0 (range: 6.0-96.0) months. Four patients had failed Cyclosporin A, MMF or cyclophosphamide therapy prior to the treatment with TAC. None of the patients had mutations of the NPHS2 or NPHS1 genes.
**Results:** Complete remission was achieved in 3 children (42.9%) (1 FSGS, 1 MPGN, 1 MsPGN) within a median time of 2.5 (range: 2.0-4.5) months. All 3 patients remained in complete remission after withdrawal of TAC within 13.0 (10.0-14.0) months. Partial remission was reached in one child with FSGS (14.2%). There was no response in 3 patients (42.9%) (2 FSGS, 1 MPGN). SRNS progressed to ESRD in one child with advanced FSGS. The side effects observed were worsening of hypertension (n=4), hyperkalemia (n=4) and acute nephrotoxicity (n=1).
**Conclusions:** Tacrolimus can be considered as effective immunosuppressive therapy for SRNS in children allowing for 57.1% rate of combined complete and partial remission. Further randomized control trials are required to determine potential therapeutic value of tacrolimus in the management of SRNS in children.

## Abstract# 434

**IgA Nephropathy and Paracoccidioidomycosis brasiliensis** S.Z.P Rigatto, V.M.S. Belangero, A.T. Tresoldi, R.M. Pereira, A. Billis. *State University of Campinas, Campinas, Brazil.*
**Objectives:** To present the first description, based on the literature, the association of IgA Nephropathy and Paracoccidioidomycosis brasiliensis.
**Methods:** Girl 6 years old had abdominal pain for 1 year, worsening in last 6 months with vomits and painful nodules in the neck. In last month, daily high fever, weight loss (4 kg), and progressive pallor for 15 days, urine color of cola. From the urban area of metropolitan city, had no family history relevant. Physical examination: pallor, lymphadenopathy and hepatosplenomegaly.
**Results:** Urinalysis :> 100 red blood cells / field with codócitos and acanthocytes, hyaline casts, granular, hemoglobin and leukocyte and 24-hour proteinuria of 420mg. Creatinine clearance of 28 ml/min/1, 73m2.
Serum complement: C3 and C4 normal. Ac anti-DNA and ANA negative
Lymph node biopsy: Presence of sporulation compatible with Paracoccidioides brasilienisis
Renal biopsy: renal tubular atrophy, interstitial fibrosis and increase in mesangial cellularity. Immune complexes of IgA paramesangial
There was resolution of renal disease after 5 months of trimethoprim-sulfamethoxazole. She is now 4 years without medication with normal urinalysis and renal function, no proteinuria.
**Conclusions:** The temporal association between the IgA nephropathy and paracoccidioidomycosis suggests a direct relation for the etiology. the mechanism could be similar than the related with Tuberculosis and IgA nephropathy. Both are antigens presented precociously in the respiratory mucosal, stimulating cellular and humoral immunity for prolonged time. This report reinforces the possibility of different etiological mechanisms of and immunopathogenicity in N. IgA and therefore different prognosis.

## Abstract# 435

**Is Cyclophosphamide Effective in Patients with IgM Positive Minimal Change Disease?** A. Roushdi, P. Geier, J. Vethamuthu, J. Feber. *Pediatrics, Children's Hospital of Eastern Ontario, Ottawa, ON, Canada.*
**Objectives:** The aim of the study was: 1) To assess the efficacy of cyclophosphamide (CYC) in patients with steroid dependent nephrotic syndrome (SDNS) and minimal change disease (MCD). 2) To compare the response to CYC between IgM positive and IgM negative patients with MCD.
**Methods:** We performed a retrospective chart review of all children who received CYC for SDNS/MCD on kidney biopsies from 1998 to 2009. Patients were divided into two groups based on IgM positivity on biopsy. Time to the first relapse post CYC was calculated and used for relapse-free survival analysis.
**Results:** Out of 17 children included in the study, 11 were MCD/IgM positive and 6 were MCD/IgM negative. The mean±SD cumulative CYC dose was 173±33 mg/kg. The percentages of patients without relapse post CYC were: 82% at 12 months, 69% at 24 months, 60% at 36 months and 40% at 48 months. There was no statistical difference in relapse free survival time between MCD/IgM negative and MCD/IgM positive patients; hazard ratio = 1.77 (95% CI = 0.4 to 8.4), p=0.47.
**Conclusions:** Almost 70% of patients with SDNS/MCD remained relapse-free for two years post CYC therapy without exceeding the recommended cumulative dose. CYC can therefore be recommended as a first line steroid sparing agent for patients with SDNS and MCD regardless of IgM positivity on kidney biopsy.

## Abstract# 436

**Development of a Global Registry of Children with Complicated Idiopathic Nephrotic Syndrome** N. Rutjes, A. Bouts, J.-C. Davin. *Pediatric Nephrology, Emma Children's Hospital, Amsterdam, Netherlands.*
**Objectives:** Idiopathic nephrotic syndrome (INS) in childhood is a rare disease. As a result, randomized controlled trials evaluating medical therapies are scarce and often of poor quality. Consequently there is no consensus regarding treatment of patients with INS, which leads to discrepancies in the management of INS. A global web-based registry (www.nsregistry.org) was developed to study children with complicated INS, and provide a framework for the organization of clinical trials. The registry is free for clinical use.
**Methods:** Consensus was reached among pediatric nephrologists from The Netherlands, Belgium, Germany, Italy, the United Kingdom and France regarding the sets of data to be registered about patients with INS complicated by steroid resistance / dependence or frequent relapses. These data sets were synchronized with various other nephrotic syndrome registries to allow for future comparison/ assimilation of data.
**Results:** A web-based registry was developed with domains for pediatric nephrologists / patients. Disease specific data can be entered regarding all aspects of disease e.g. first manifestation of disease, therapy, genetic analysis and outcome. In addition overviews and various statistics can be generated.

**Conclusions:** The use of this multi-purpose disease registry could enhance our knowledge about demographics, treatment and prognosis of complicated INS and aid in the development of treatment guidelines for these patients. Its use could facilitate the management of patients with complicated INS. In addition international multi-center randomized controlled trials can be implemented within the framework of this registry. (Presented on behalf of the EURO-WINS working group)

## Abstract# 437

**Development of a Global Registry of Children with Complicated Idiopathic Nephrotic Syndrome: Patient Domain for Parents** N. Rutjes,[1] M. van Meel,[2] J.-C. Davin.[1] *[1]Pediatric Nephrology, Emma Children's Hospital, Amsterdam, Netherlands; [2]NephcEurope, Bodegraven, Netherlands.*

**Objectives:** Parents of children with idiopathic nephrotic syndrome (INS), organized in NephcEurope, have expressed their desire for the development of a system that allows them to register data regarding their child's disease. For this purpose, we now have developed a dedicated patient domain as part of the web-based Nephrotic Syndrome Registry (NSR, www.nsregistry.org).

**Methods:** In close collaboration with the European patient organization NephcEurope, data sets were defined to meet the needs and situation of parents of children with INS complicated by steroid resistance / dependence or frequent relapses. In addition data forms were designed to complement the registry data collected by their pediatric nephrologist.

**Results:** As part of the web-based NSR registry a patient domain was developed. After registration of a patient by their pediatric nephrologist in the registry, parents can access the patient domain and enter data regarding demographics, patient surveys, relapses, urine dipstick measurements, hospital visits, medication, dialysis, transplantation and receive reminders of upcoming hospital visits by email or SMS. Statistics can be easily generated to provide insight in the child's past medical / treatment history.

**Conclusions:** A dedicated patient domain was developed within the web-based idiopathic nephrotic syndrome registry, allowing parents to register data regarding their child's disease. This registry could help parents manage their child's disease, complement registration by their nephrologist and further improve the care of children with complicated INS.

## Abstract# 438

**Study of the Factor Which Affects to the Concentration of Mizoribine with Idiopathic Nephrotic Syndrome Patients** A. Saito,[1] M. Ikoma.[2] *[1]Pediatrics, St. Marianna Univercity School of Medicine, Kawasaki, Kanagawa, Japan; [2]Pediatrics, St. Marianna Univercity School of Medicine, Kawasaki, Kanagawa, Japan.*

**Objectives:** To evaluate the factor which affect to the concentration of mizoribine (MZR) with idiopathic nephrotic syndrome patients. MZR is the inhibitor of inosine monophosphate dehydrogenase, which was made in Japan. The treatment of steroid-dependent nephrotic syndrome (SDNS) and frequently relapsing nephrotic syndrome (FRNS) has been considered. In Japan, we usually use cyclophosphamide, cyclosporin A and MZR with or without predonisolone. It was reported that the concentration of MZR was necessary to control proteinuria above 3.0 μg/ml.

**Methods:** We retrospectively analyzed 17 cases of childhood-onset idiopathic nephrotic syndrome (9 males and 8 females, FRNS: 1, SDNS: 14, steroid resistant nephrotic syndrome: 1, FSGS: 1), and studied 60 samples. The concentrations of MZR were analyzed 2 to 3 hours after administration.

**Results:** Mean dose of MZR was 4.9±1.7mg/kg/day, mean concentration of MZR was 2.3±1.0μg/ml,mean dose of predonisolone (PSL) was 0.3±0.2mg/kg/day. The correlation of the dose of MZR and the concentration of MZR was not significantly. The inversely correlation of the dose of PSL and the concentration of MZR was significantly ($p<0.01$).

**Conclusions:** The concentration of MZR was affected by the high dose of PSL.

## Abstract# 439

**Mycophenolate Mofetil Therapy for Steroid-Resistant Nephrotic Syndrome in Children** J.R. Santos, L.C. Aguiar, M.A. Barbosa, F.M. Del Caro, F.V. Leão, M.A.P. Cançado, J.T. Carvalhaes. *Pediatric Nephrology, Federal University of São Paulo, São Paulo, São Paulo, Brazil.*

**Objectives:** Evaluation of clinical and laboratory response of children with steroid-resistant nephrotic syndrome (SRNS) using mycophenolate mofetil (MMF), as well as its efficacy and safety. MMF is a selective reversible inhibitor of the inosine monophosphate dehydrogenase, responsible for the inhibition of the de novo synthesis of purines. Preliminary studies suggest beneficial effects of MMF as adjunctive therapy in the nephrotic syndrome (NS). However, there are limited data on the efficacy of MMF in pediatric patients with SRNS.

**Methods:** Retrospective evaluation of clinical and laboratory data of 13 pediatric patients with SRNS, previously treated with an 8-week course of prednisone 2 mg/kg/day, followed by a 3-month course of cyclophosphamide and one year of cyclosporine (5 mg/kg/day for 6 months and 2,5 mg/kg/day for more 6 months), with no response. These patients were submited to 250 to 500 mg/m2 of body surface per day of MMF. After a period of one year, data was acessed for laboratory results, clinical response to treatment and adverse side effects.

**Results:** 10 patients showed no edema and remained asymptomatic. Two patients presented clinical descompensation, but only one experienced adverse events (nausea and vomiting) and had the medication discontinued. However, all patients remained proteinuric (average 44 mg/kg/day).

**Conclusions:** Effective clinical response was observed with the use of MMF in children with SRNS. Poor or none laboratory response was observed. Additional studies are necessary to confirm MMF effectiveness and evaluate alternative treatments to SRNS in children.

## Abstract# 440

**Cyclosporine Use in Children with Steroid-Resistant Nephrotic Syndrome** J.R. Santos, F.V. Leão, M.A.P. Cançado, J.T. Carvalhaes. *Pediatric Nephrology, Federal University of São Paulo, São Paulo, São Paulo, Brazil.*

**Objectives:** Evaluation of clinical and laboratory response to cyclosporine in pediatric patients with SRNS.

**Methods:** The study evaluated retrospectively clinical and laboratory data of 36 pediatric patients with SRNS, previously treated with an 8-week course of prednisone 2 mg/kg/day with no response, followed by a 3-month course of cyclophosphamide 2 mg/kg/day. After this period these patients were treated with cyclosporine 5 mg/kg/day for 6 months, followed by 2,5 mg/kg/day for more 6 months, completing a one-year course of cyclosporine.
10 patients were excluded from this study (9 have had adverse effects and 1 was still under treatment).
Patients were submitted to clinical and laboratory evaluations once-a-month while they were using cyclosporine.

**Results:** All 26 patients have had disappearance of edema. 16 demonstrated persistent nephrotic range of proteinuria ; non nephrotic range of proteinuria in 7 patients and lack of proteinuria in 3 patients. Maintenance of hypercholesterolemia in 21 patients. Normal cholesterol levels in 5 patients. Adverse effects: gastric intolerance (n=1), creatinine levels increase (n=3), hepatic enzymes levels increase (n=1), gengival hyperplasia (n=1), systemic arterial hypertension (n=1), infection (sepsis/ spontaneous bacterial peritonitis) (n=2).

**Conclusions:** Effective clinical response was observed with the use of cyclosporine for a period of one year in children with SRNS. Poor or non laboratory response was observed. Additional studies are necessary to confirm cyclosporine effectiveness and evaluate alternative treatments to SRNS in children.

## Abstract# 441

**Acute Postinfectious Glomerulonephritis (APGN) Versus Membrano-Proliferative GN Type I (MPGN-I), a Diagnostic Challenge** A. Sarkissian,[1] H. Nazaryan,[1] A. Sanamyan,[2] A. Gaspert,[3] E. Leumann,[4] G. Sparta,[4] G. Laube.[4] *[1]Nephrology, Arabkir Joint Medical Center, Yerevan, Armenia; [2]Clin. Pathology, State Medical University, Yerevan, Armenia; [3]Inst. of Surgical Pathology, Univ. Hospital, Zurich, Switzerland; [4]University Children's Hospital, Zurich, Switzerland.*

**Objectives:** Differentiation may be difficult in cases of endocapillary proliferative GN with subepithelial deposits (humps, typical for APGN) and concurrent subendothelial deposits (typical for MPGN-I). We studied 7 patients (Yerevan 4, Zurich 3) with overlapping histology.

**Methods:** Biopsies of native kidneys were performed during 2001-2009 in 265 pediatric patients (Yerevan 130, Zurich 135). Additional evaluation of Armenian patients was done in Zurich by LM, EM and immunohistochemistry (for patients in Zurich immunofluorescence).

**Results:** Patients (5 m, 2 f; 7-17 y/o) had biopsies 2-6 months after initial presentation. All seven showed both humps and subendothelial deposits. Based on the main distinguishing finding (diffuse double contours of the GBM with subendothelial deposits, diagnostic for MPGN-I) definitive diagnosis was made in 4 (2 APGN, 2 MPGN-I), but diagnosis remained presumptive in 2 APGN and 1 MPGN-I. Repeat biopsy in one of them considered to have APGN now showed unequivocal MPGN-I.
Therapy consisted of ACE-inhibitors (7) and prednisone (6). After 1/2 to 4 years four had proteinuria and 1 each ESRF, haematuria and remission.

**Conclusions:** Histological differentiation between APGN and MPGN-I may be difficult in biopsies showing endocapillary proliferative GN. Finding of diffuse double contours of the GBM with subendothelial deposits, suggesting MPGN-I, is helpful. Rarely a second biopsy may be required.

## Abstract# 442

**Role of Zinc Supplements in Reducing Relapses in Nephrotic Syndrome** A.R. Sherali, K.N. Moorani, S.I. Khan, S.H. Chishty. *Pediatric Nephrology, The Kidney Center & NICH, Karachi, Sindh, Pakistan.*

**Objectives:** To determine whether zinc suplements reduces infection related relapses in children with steroid sensitive nephrotic syndrome(SSNS).

**Methods:** A double blind randomized controlled trial was conducted from December, 2007 to June, 2009 at National Institute of Child Health(NICH) and The Kidney Centre, Karachi. Patients of 2-16 years with SSNS were included. Age at onset of NS and present age,sex, number of relapses and infections associated with relapses during the study period and in the preceding 1 year were noted. Zinc level < 70 µg/dl was taken as low. Steroid resistant NS, asthma and tuberculosis were excluded. Patients received either Zinc (10mg/day) or placebo and followed for one year. Side effects were noted. Data was analyzed using SPSS ver 11.0. Quantitative variables were expressed as percentage, mean ± SD, and qualitative as 95% CI and a P-Value <0.05 was significant.

**Results:** Among 55 cases, 26 were in Zinc group(one lost follow up), 29 in placebo.Baseline characteristics in the 2 groups were similar. Most common infection was acute respiratory infection. Zinc deficiency was seen in 23 cases. In Zinc group, infection related relapses occurred in 7 (28%) and 10 in placebo (34%). No significant difference was seen in the relapses in 2 groups (P>0.335). However, infection associated relapses in Zinc group was significantly lower(46% )compared to the pre-study period(P-value 0.01). While in placebo group, this difference was 27% (P-value 0.335).

**Conclusions:** Zinc supplements is useful in reducing the relapse rate in children with NS compared to the preceeding one year with no supplements. Further evidence and larger studies are required to support this finding.

## Abstract# 443

**SOCS1 Over-Expression in Peripheral Blood Lymphocyte May Predict Resistance to Steroids in Childhood Nephrotic Syndrome** M. Silska, D. Ostalska-Nowicka, M. Smiech, J. Zachwieja, W. Szaflarski, M. Jaroniec, M. Nowicki. *Pediatric Cardiology and Nephrology, Uniwersity of Medical Sciences, Poznan, Poland; Pediatric Cardiology and Nephrology, Uniwersity of Medical Sciences, Poznan, Poland; Pediatric Cardiology and Nephrology, Uniwersity of Medical Sciences, Poznan, Poland; Pediatric Cardiology and Nephrology, Uniwersity of Medical Sciences, Poznan, Poland; Histology and Embryology, Uniwersity of Medical Sciences, Poznan, Poland; Pediatric Cardiology and Nephrology, Uniwersity of Medical Sciences, Poznan, Poland; Histology and Embryology, Uniwersity of Medical Sciences, Poznan, Poland.*

**Objectives:** Cytokines induces a JAK/STATs, which is controlled by suppressors of cytokine signaling (SOCS).

It becomes expressed in the cells secondary to different factors including steroids. In the present report we studied expression of JaK/STATs pathway in peripheral blood lymphocytes at mRNA level in children with steroid-sensitive nephritic syndrome (SSNS, n=8) and SRNS (n=10).

**Methods:** We used the fluorescently labeled primer and analyze gene expression by using cDNA as a template in RealTime PCR reaction.

**Results:** IN 6/10 of SRNS subjects the SOCS1 was 4x over-expressed as compared to SSNS patients and controls. In SRNS mRNA of pro-inflammatory factors was within the control limits, while expression of pro-inflammatory mediators in all SSNS was decreased.

**Conclusions:** We speculate SRNS patients respond to re-modulate the immune system (SOCS1 over-expression) but, probably secondary to kidney genetic alterations, do not eradicate proteinuria. In that way SOCS1 over-expression may predict resistance to steroids in children with nephritic syndrome.

## Abstract# 444

**Steroid-Dependent Nephrotic Syndrome: Outcome after Use of Steroid-Sparing Drugs** G. Silva, S. Branco, A. Teixeira, H. Pinto, C. Tavares, C. Afonso, H. Jardim. *Pediatric Nephrology Unit, Hospital de São João, Porto, Portugal.*

**Objectives:** At least 80% of patients (pts) with idiopathic nephrotic syndrome(NS) are sensitive to steroid treatment.Of these, up to 60% will have frequent relapses or become steroid dependent requiring steroid-sparing agents to avoid toxicity. The aim of the study was to review the options and response to alternate regimes used in children with steroid-dependent NS (SDNS) under current follow-up at our Unit.

**Methods:** A retrospective analysis was performed regarding clinical and biological data at onset and at latest follow-up in relation to treatment options undertaken. In all cases remission was achieved with steroids in a tapering regime with subsequent introduction of other agents.

**Results:** Nine children were included. Median age at onset was 4.2 (3-8.5) years. Follow-up period:15-153 months. Total relapses number was 6.3± 4.1(CI 95%). Renal biopsy was done in 3 pts (2 mesangioproliferative glomerulonephritis;1 minimal change disease). Levamisole (LS) was used in 2 pts (no response-1), cyclophosphamide (CyA) in 3 (remission-2), and 4 received mycophenolate mofetil (MMF).The non responders to LS and CyA responded to subsequent MMF. At present 5 pts remain on MMF (in association with low dose alternate days prednisolone). Relapses number decreased with the options undertaken (5.8±3.2 *vs* 1.6 ±1.6; p=0.017; median follow-up of 24 *vs* 26 months), with no influence in renal function (mean GFR 138±34 *vs* 133±17ml/min/1.73m$^2$).

**Conclusions:** In our experience, the options used to avoid long term steroid exposure in SDNS showed, to date, a benefit with no adverse effects. However, some cases needed to maintain association with low dose steroids or a second agent trial in order to achieve the best efficacy, with an overall duration which is difficult to establish.

## Abstract# 445

**Children with IgA Nephropathy and Henoch – Schoenlein Purpura Nephritis in Czech Registry of Renal Biopsies** S. Skalova,[1,2] A. Kolsky,[1] E. Jancova,[1] J. Dusek,[1] M. Hladik,[1] K. Vondrak,[1] P. Geier,[1] V. Smrcka,[1] J. Starha,[1] J. Skibova,[1] J. Stejskal,[1] J. Janda,[1] I. Rychlik,[1] V. Tesar.[1] *[1]Czech Registry of Renal Biopsies, Prague, Czech Republic; [2]Department of Pediatrics, Faculty of Medicine and University Hospital, Hradec Kralove, Czech Republic.*

**Objectives:** This study aimed to determine the occurence of IgA nephropathy (IgAN) and Henoch-Schoenlein purpura nephritis (HSPN) in Czech children.

**Methods:** Clinical and histopathological data were obtained from Czech Registry of Renal Biopsies (CRRB).We evaluated data relating to 1903 native renal biopsies performed in children and adolescents < 18 years of age by 10 pediatric centers in the years 1994–2008.

**Results:** IgAN was detected in 440 (23.5%) children and HSPN was diagnosed in 66 (3.2%) children. The mean age was 13.92 years in IgAN and 11.68 years in HSPN patients. Male to female ratio was 1.9 : 1.0 and 0.9 : 1.0, respectively. Mean serum creatinine levels were 77.2 umol/L (range 20-600) vs 70.98 umol/L (range 28 –280). Proteinuria > 3g/day at the time of biopsy was seen in 13.7% of IgAN and 25% of HSPN patiens (p=0.001). Microscopic haematuria was present in 61.3% of IgAN and in 80% of HSPN patients, gross haematuria was observed in 34.4% and 16.7%, respectively. Arterial hypertension at the time of biopsy was present in 0.9% of children with IgAN and 26.7% of patiens with HSPN.

**Conclusions:** IgA nephropathy is the most frequently encountered primary glomerulonephritis while HSPN is the third most common secondary nephropathy in children in CRRB. Compared to IgA nephropathy, patients with HSPN are younger with higher incidence of proteinuria and hypertension at the time of diagnosis.

## Abstract# 446

**The Prognostic Effect of the Long Term Prednisone Treatment in Idiopatic Nephrotic Syndrome, a Single Center Study** B. Sozeri, N. Dincel, O.D. Kara, E. Toroslu, S. Mir. *Pediatric Nephology, Ege University Faculty of Medicine, Izmir, Turkey.*

**Objectives:** The prognosis of the idiopathic nephrotic syndrome is dependent on treatment modalities.

**Methods:** We assessed 120 cases controlled in Ege University Nephrology Department, Izmir, between1987-2009.

Initially, all patients given prednisone 2mg/kg/ day per 4 weeks, followed by 8 weeks of the same dose given every other day. Doses were decreased with a rate of 0.5mg/kg per 15 daily intervals until the end of 18th week. Steroid resistant, > 8 years and < 2 years patients at the initiation of disease underwent renal biopsy. Also, NPHS2 gene analysis was revealed in the steroid resistant patients.

**Results:** The mean age at onset of disease was 5.49 ± 3,8 years. At the end of the four week of treatment,there was steroid responsive patients(n=106) and steroid resistant patients(n=14). 35.8 %(n=43) of patients underwent biopsy. Cytoctoxic therapy was given to depended and resistants.

Four patients had NPHS2 mutation. At the end of the follow up period, 84(70%) patients were steroid responsive without any adverse effect of treatment and all of them had full remission (85% in 5 years, 100% in 10 years).18/22 patients in steroid dependance had remission(82%) and 4 progressed to resistance.In steroid resistant patients, 14/ 18 achieved remission with a rate of 70% in 10 years.

**Conclusions:** Most of patients had remission end of follow up time. When we compared with our results to the literature, the long term use of steroid treatment has significantly better prognosis. We suggest that cytotoxic therapy should be started earlier in steroid dependent and resistant groups with NPHS2 negative.

## Abstract# 447

**Analysis of Factors Influencing the Prognosis of Children with Lupus Nephritis in Shanghai** L. Sun, H. Xu, H.-M. Liu, L.-J. Zhou. *Department of Nephrology and Rheumatology, Children's Hospital of Fudan University, Shanghai, China.*

**Objectives:** To explore the factors affecting the long-term survival of children with lupus nephritis.

**Methods:** The data of 101 patients who diagnosed as lupus nephritis from Jan. 1996 to Jun. 2007 in our hospital were investigated retrospectively. According to

different recipes in induction therapy, divided into CTX group, MMF group and other drugs group. According to whether receive renal biopsy and pathological classification, divided into group A (type I, II), group B (type III, IV), group C (type V) and group D (no biopsy). According to different outcomes, patients were divided into remission group (complete and partial remission) and ineffective group (non-response).
**Results:** Average follow up time is 19.98 months, maximum 11.5 years, 20 cases follow up more than 3 years. 2, 5 and 10 years renal survival rate is 88.9%, 84.0% and 60.0% respectively. Univariate analysis showed following 4 variables were correlated to prognosis: different pathological types (P=0.005), heavy proteinuria at onset (P=0.003), different recipes (P=0.001), and irregular treatment (P=0.001). No biopsy group and type V group have a relative worse prognosis. The prognosis of MMF group is better than the other groups. Heavy proteinuria at onset and irregular treatment are two influence prognosis factors. In multivariate analysis, it was confirmed that only irregular treatment (OR=9.955) was significantly associated with outcome.
**Conclusions:** For SLE patients with evidence of nephritis, renal biopsy should be suggested, in order to guide treatment and predict prognosis. Regular treatment plays a more important role to prognosis than different therapies, especially in developing country.

## Abstract# 448

**Steroid Sensible Nephrotic Syndrome - A Rare Disease in the New Era?** L.C. Sylvestre, A.P. Smaniotto, J.F. Sousa, K.P. Olandoski, M.F. Munhoz da Cunha, E.T. Vargas, E.I. Wladika, R.P. Meneses, D.D. Giamberardino Filho. *Pediatric Nephrology, Hospital Pequeno Principe, Curitiba, Parana, Brazil.*
**Objectives:** To analyze if steroid sensitivity is still the leading pattern of response in patients with Primary Nephrotic Syndrome (PNS), followed in our center.
**Methods:** Retrospective study,reviewing the charts from all patients with PNS treated in the same Hospital for at least one year, from January 2000 until December 2006. Analysis of the age of the beginning of PNS, sex, response to steroids, histology of the biopsy and follow-up.
**Results:** 129 patients were suitable for the analysis. Seventy-nine were male, mean age at diagnosis was 5.4 ± 3.2 years and mean follow-up was 49 ± 24 months (2 to 96 months). Forty-nine (38%) were steroid resistants and 80 (62%) were sensible. Eighty-two patients needed a second immunossupression. Renal BIOPSY: 47 (36%) pts did not perform any, 50 (61%)of the performed had Minimal changes(MCG),22% FSGS and in 14 (17%) there were other diagnosis. Twenty-three MCG patients were steroid sensible and 27 were resistant; 13 FSGS patients were steroid resistant and 5 were steroid sensible. Follow up: 2 deaths, 6 progressed to End Stage Renal Disease, 18 lost follow up, 3 were transferred, 74 were still being followed, 26 on remission for more than 1 year.
**Conclusions:** We have a predominance of male sex, the most frequent biopsy result was Minimal Changes and the majority had a good response to steroids. Therefore, we can conclude that in our population it is still true to say that PNS is a benign disease with most of the patients responding to the usual therapy.

## Abstract# 449

**Variation in the Course of Membranous Nephropathy** S. Tanaka,[1] Y. Yoshimoto,[1] K. Ushijima,[1] Y. Ito,[1] S. Hisano.[2] *[1]Pediatrics, Kurume University Medical Center, Kurume, Fukuoka, Japan; [2]Pathology, Fukuoka University, Fukuoka, Fukuoka, Japan.*
**Objectives:** The incidence of membranous nephropathy is low in children. There are many underlying causes of secondary membranous nephropathy. However, in many cases, the underlying disease cannot be clearly determined. This situation has made determination of the clinical course, prognosis, and treatment of this disease more complicated. We report four cases of secondary membranous nephropathy with various causes and prognosis.
**Results:** Case 1: Mild proteinuria and decline in renal function were observed within a year after the onset of membranous nephropathy. Histopathologic examination revealed stage II disease, which was treated with Angiotensi n II Receptor Blocker. However, renal function deteriorated.
Case 2: Nephrosis occurred following graft versus host disease, and histopathologic examination revealed stage II disease. Treatment was performed with CsA, and two years later renal function impairment was not observed, with confirmation of stage IV disease and subsequent remission.
Case 3: SLE occurred during the course of ITP. Although urinalysis revealed no abnormalities, histopathologic examination revealed class V findings.
Case 4: Proteinuria 2g/day persisted for several months, and the patient was followed without drugs because of stage I histopathological findings. Remission was achieved within a year.
**Conclusions:** The severity of secondary membranous nephropathy is not correlated with histopathological and urinary findings. The causes and age at presentation of membranous nephropathy change prognosis. It is important to accumulate the cases of secondary membranous nephropathy.

## Abstract# 450

**Optimal Treatment of Steroid Sensitive Nephrotic Syndrome in Children: A Multicenter Randomised Controlled Trial** N. Teeninga,[1,2] J.E. Kist-van Holthe,[1] N.I. de Mos,[1] N. Peters,[2] W.C.J. Hop,[3] A.J. van der Heijden,[2] J. Nauta.[2] *[1]Pediatrics, Leiden University Medical Center, Leiden, Netherlands; [2]Pediatrics, ErasmusMC/Sophia Children's Hospital, Rotterdam, Netherlands; [3]Epidemiology and Biostatistics, ErasmusMC, Rotterdam, Netherlands.*
**Objectives:** In a multicenter, randomised placebo controlled trial we compare the efficacy of three months to six months corticosteroid therapy for a first presentation of nephrotic syndrome (NS).
**Methods:** Children 1-16 years old with a first episode of NS are randomised into 2 treatment regimens with equal cumulative dosage of prednisolone. Regimen A: 6 weeks prednisolone 60 mg/m$^2$ daily, then 6 weeks prednisolone 40 mg/m$^2$ on alternate days, followed by 12 weeks placebo. Regimen B: 24 weeks prednisolone starting at 60 mg/m$^2$ daily in a gradually tapering scheme. Primary outcome: percentage frequently relapsing NS after 2 years. Patients are monitored for adverse effects of prednisolone. In addition we study glucocorticoid sensitivity (DNA and partial dexamethason suppression test) and pharmacokinetics of prednisolone.
**Results:** In a 5 year period 150 patients are enrolled in the study (2005 to 2010). Median (range) age of the patients with a first episode NS is 4.0 (1.6.-16.9) years, 102 (68%) are boys. After 2 years of follow up, 31/38 (82%) of the patients had one or more relapses. In addition nearly half of all patients (17/38) developed a frequently relapsing NS.
**Conclusions:** The relapse rate is relatively high in our patient population with (minimal change) NS. This multicenter randomised placebo controlled trial will reveal in 2012 whether (frequent) relapses can be prevented by prolongation of the initial corticosteroid treatment.

## Abstract# 451

**Health Related Quality of Life and Disease Activity in Children with SLE and Lupus Nephritis: A Multicenter Study in Thailand** P. Thirakhupt, A. Pattaragarn, K. Tangnararatchakit, P. Dissaneewate, W. Chartapisak, A. Jirawuttipong, N. Chongchate, K. Wathanapenpaiboon, R. Phuaksungnern, S. Khorprasert, J. Jungthirapanich. *Thai Pediatric Lupus Study Group, Bangkok, Thailand.*
**Objectives:** To determine the correlation of health related quality of life (HRQOL) and disease activity as well as damage in Thai children with SLE and Lupus nephritis.
**Methods:** A cross-sectional survey was conducted in eleven hospitals in all parts of Thailand.The data of 264 patients with SLE, age at diagnosis less than 15 years, diagnosed during 1 January 2002 to 31 May 2009 were reviewed. Severity and complication of the disease were assessed using European Consensus Lupus Activity Measurement index ( ECLAM ) and SLICC/ACR damage index (SDI) respectively. PedsQL™4.0 Generic Core Scale (Thai version) was used to assess HRQOL of the patients.
**Results:** There were 41 (15.5%) male and 223 (84.5%) female patients, the mean age was 11.09 ± 2.5 years.The patients had SLE mean duration of 1.98 ± 1.7 ( range 0-7) years. Among 203 patients who underwent kidney biopsy 152 patients (74.8 %) had Lupus nephritis class III - V. The mean ECLAM index was 1.07 ± 1.7. Mean SDI was 0.3 ± 0.5. The mean HRQOL of the total summary score was 22.6 ± 13.6. The means score for physical health, emotion, social and school function were 22.2 ± 16, 23.5 ± 19.1, 15.6 ± 16.7 and 29.4 ± 16.9 respectively. Using Spearman's correlation test, no significant correlation of HRQOL, ECLAM and SDI was found.
**Conclusions:** Significant correlation of HRQOL, using PedsQL™4.0 Generic Core Scale (Thai version), and SLE activity as well as damage are not demonstrated in Thai children.

## Abstract# 452

**Relationship between Urine Endothelin-1 and Albumin Excretion in Healthy Adolescents** P.N. To, L.A. Ortiz, O. Nwobi, G.A. Harshfield. *Pediatric Nephrology/Georgia Prevention Institute, Medical College of Georgia, Augusta, GA, United States.*
**Objectives:** Studies have shown that microalbumin excretion is related to increased risk for progression of hypertension and cardiovascular diseases in adults. Furthermore, in healthy children, it has been previously demonstrated that albumin excretion rate (AER) is correlated with higher blood pressure and is higher in blacks than in whites. However, there are limited data available looking at the relationship between AER and urine endothelin-1 (ET-1). ET-1 is known for being a potent vasoconstrictor and plays a major role in blood pressure regulation. In the kidneys, we know that urine ET-1 acts on the microvessels, but it is still not clear as to its role in renal function.
**Methods:** We evaluated AER and urine ET-1 in healthy adolescents participating in a study of stress-induced change in urine ET-1. A total of 86 healthy, normotensive adolescents, 42 blacks and 44 whites (50% males, 50% females),

aged 15-18 years, participated in a 5-hour testing protocol. Subjects started with 2 hours of rest prior to a 1-hour period of mental stress and followed by a 2-hour rest period. Urine microalbumin and ET-1 levels were measured immediately before and after stress.
**Results:** In blacks, the stress induced change in ET-1 correlates with lower AER ($P = 0.008$, $R^2 = 0.16$). This relationship is not as significant in whites ($P = 0.375$, $R^2 = 0.018$). This means ET-1 regulation contributes to 16% of AER in blacks and only 1.8% of AER in whites.
**Conclusions:** These findings suggest that renal ET-1 is a major player in renal function and may be used as marker for renal injury in blacks.

## Abstract# 453

**NPHS2 Mutations as a Cause of Childhood Steroid-Resistant Focal and Segmentary Glomerulosclerosis/Minimal Change Disease (FSGS/MCD) in Central Russia** A. Tsygin,[1] E. Tikhomirov,[2] L. Leonova,[3] T. Voznesenskaya,[1] T. Margieva.[1] *[1]Nephrology, Institute of Pediatrics NCZD RAMS, Moscow, Russian Federation; [2]Ntl. Genetic Centre, Moscow, Russian Federation; [3]Pathology, Russian State Medical University, Moscow, Russian Federation.*
**Objectives:** The aim of this study was to evaluate the prevalence of NPHS2 mutations in Russian pediatric patients with biopsy proven steroid-resistant FSGS/MCD.
**Methods:** During the years 2003-2009 sixty nine children were diagnosed with steroid resistant NS in the reference pediatric centre with the wide geographic representativeness across Russia. Afterwards Cyclosporine A (CsA) treatment with or without methylprednisolone pulses and oral prednisolone was introduced. If no response was seen after 8-10 months, patients were investigated for NPHS2 by direct sequencing.
**Results:** Finally, 24 patients were classified as having CsA resistance. After direct sequencing, in eight of them mutations of NPHS2 were determined. So, the total prevalence of podocin mutations in steroid-resistant FSGS/MCD is 11,6% where as in CsA-resistant it was 33,3%.
Children with a confirmed mutations had early (1,6±0,34 years old) onset without any clinical differences from non-genetic forms of the disease. Half of these patients had an early hypertension, two had normochromic anemia. Two children were from the same family carrying same mutation. The most often mutations were R139Q (3 children), R229Q and G87A (2 children each).
**Conclusions:** We conclude, that podocin mutations may be the cause of steroid-resistant FSGS/MCD in at least 11,6% of cases. All patients, especially with early onset of the disease need direct sequencing before aggressive treatment.

## Abstract# 454

**Method of Quantitative Assessment of IHC Staining in Kidney Glomeruli** I.V. Sakharau,[1] N.I. Tur.[2] *[1]Department of Pathology, Belarusian State Medical University, Minsk, Belarus; [2]Republic Center of Pediatric Nephrology and Renal Replacement Therapy, Minsk, Belarus.*
**Objectives:** The purpose of our study was to work out a method of quantitative assessment of IHC staining in kidney glomeruli taking only cell volume into account.
**Methods:** 31 needle renal biopsy specimens from children with nephrotic syndrome were studied. Histologically minimal change disease, mesangioproliferative nephropathy, and focal segmental glomerulosclerosis were diagnosed. IHC staining of specimens for podocalyxin was performed. Images of 5 glomeruli in the section were taken at a x400 magnification and further analyzed using WCIF ImageJ 1.42q software. The glomerular area (GA) was calculated by drawing a region of interest around the glomerulus. Capillary lumen area (CLA) of glomerulus was calculated using color based thresholding. Cellular area (CA) was determined as a difference between GA and CLA. The number of positive pixels was calculated using color based thresholding with a spectrum specific for the diaminobenzidine staining. Staining density was stated as a number of positive pixels per square micrometer of CA.
**Results:** Podocalyxin immunostaining was found in glomeruli along the membranes of podocytes and endotheliocytes. Staining density was calculated for each specimen and the mean was 29.68±17.78 pixels per square micrometer. Mean staining density calculated per GA was 20.09±12.74. This data shows that staining density per CA is more variable than per GA. This may have influence on the results of investigation of IHC markers expression in glomeruli.
**Conclusions:** The described method may be useful for quantitative assessment of IHC staining in glomeruli and for its comparison in different glomerular lesions.

## Abstract# 455

**Long-Term Outcomes of Steroid-Resistant Nephrotic Syndrome in Children** T. Udagawa, M. Ogura, K. Kamei, S. Ito. *Department of Nephrology, National Center for Child Health and Development, Okura, Setayaga-ku, Japan.*
**Objectives:** The prognosis of childhood steroid-resistant nephrotic syndrome (SRNS) is poor. Approximately 40% of patients progress to end-stage renal failure (ESRD).Since there have been few reports of SRNS in Japan,we examined its prognosis.
**Methods:** Forty five children (29 males, 16 females) with SRNS, who were treated in our institute between 1989 and 2008, were retrospectively studied. Median follow-up period was 93.3 months (15-250 months). Congenital and secondary nephrotic syndrome were excluded.
**Results:** The mean onset age was 6.2 years. Primary steroid non-responders were 29 of the 45 patients and 16 were late. Renal biopsies showed minimal change disease in 26 focal segmental glomerulosclerosis in 14 and diffuse mesangial proliferation in 5. Nine patients never remitted, among whom 6 progressed to ESRD and 3 still presented proteinuria. Thirty-six patients became steroid-sensitive nephrotic syndrome through cyclosporine (CsA), and only one of these 36 progressed to ESRD. Primary non-responders had more risk of ESRD than late non-responders(odds ratio, 3.61). Seven become free from medication, but 20 suffered steroid-dependent or frequent-relapsing nephrotic syndrome (SDNS/FRNS). Owing to severe side effects of steroid and/or CsA,16 of the 20 SDNS/FRNS patients were unavoidably treated with off-label rituximab.
**Conclusions:** This prognosis seems better than those in previous reports. However, long-term and complete remission were rare, and approximately 40% of SRNS patients suffered SDNS/FRNS. For such refractory patients, rituximab is a hopeful option for achieving long-term remission.

## Abstract# 456

**Demographic, Clinical Features, Initial Management and 12 Months Follow up of Children with Idiopathic Nephrotic Syndrome in a Teaching Hospital at Dhaka, Bangladesh** G.M. Uddin, S. Jahan, C.A. Kawser, H. Rahman, M.M. Hossain. *Paediatric Nephrology, Bangabandhu Sheikh Mujib Medical University(BSMMU), Dhaka, Dhdka, Bangladesh.*
**Objectives:** To evaluate the demographic, clinical features, treatment response and 12 months outcome of all children with Idiopathic Nephrotic Syndrome (INS) over a period of 3 years.
**Methods:** Type of study:Prospective observational done in the department of paediatric nephrology. Children under 15 years attending the department who fulfilled the criteria of initial attack of nephrotic syndrome used as per ISKDC criteria were enrolled for study. Duration of study was from January'06 to January'09. The regimen of prednisolon used for 1st attack 60 mg/m2/day for 28 days followed by 40 mg/m2, alternate day for 4 to 12 weeks. For relapse children were treated with 40 mg/m2, alternate day for 4 week after achieving remission with daily dose of prednisolon 60 mg/m2/day.
**Results:** Total 108 children were included in this study. Mean age were 4.7±3.1 and male: female were 1.5:1. Atopy were identified in 47% cases, 39% had hematuria and 18.5% had elevated blood pressure. 98% of them had normal renal function. Among them 101 (94%) were steroid responsive and 7 (6%) were steroid resistance. Meantime of response was 11.5 days and meantime to relapse was 15.3 weeks. Among the initial steroid sensitive group 45% were infrequent relapser, 18% frequent relapser and 10% were steroid dependent. 22% were non relapser. Amongst 6 renal biopsy in steroid resistance cases, 3 had minimal changes, to had FSGS and 1 had DMPG on histology.
**Conclusions:** ISKDC recommendation is as effective in our study population as that of other.

## Abstract# 457

**Is Intravenous Cyclophosphamide a Worthwhile Treatment in Pediatric Onset Lupus Nephritis? A 108 Case Experience in a Single Center** P. Vachvanichsanong,[1] P. Dissaneewate,[1] E. McNeil.[2] *[1]Pediatrics, Prince of Songkla University, Hat Yai, Thailand; [2]Epidemiology Unit, Faculty of Medicine, Prince of Songkla University, Hat Yai, Thailand.*
**Objectives:** To evaluate the long-term results of intravenous cyclophosphamide (IVCY) therapy in pediatric onset severe lupus nephritis (LN).
**Methods:** The medical records of patients aged < 18 years who attended the Department of Pediatrics, Prince of Songkla University from October 1993 to December 2008, and were diagnosed with SLE and severe LN, defined as either clinical nephritis or renal histopathology classification, and who were selected for a 17-treatment IVCY course, were reviewed.
**Results:** There were 19 boys and 89 girls with a mean age of 12.6±2.7 years and mean follow up time of 5.7±4.3 years. 48 patients completed the 36-month IVCY course of therapy, 29 did not complete the course for various reasons, and 31 were ongoing at the time of analysis.

Of the 48 who completed the course of IVCY therapy, 8 later died, and 26 of the 40 survivors were available for evaluation for this study. Of the 26, only 9 patients were free of medication; the other 17 patients still required medication to control their disease.
Overall 23 patients died, and patient survival rates at 2, 5 and 10 years were 93%, 78% and 66%, respectively.
**Conclusions:** Three years of IVCY treatment in severe lupus nephritis provides initial good remission, unfortunately, it is not permanent, since if long term outcomes are considered, some patients died and some patients required a return to medication.

## Abstract# 458

**Pharmacokinetics of Cyclosporin (Neoral) in Children with Idiopathic Nephrotic Syndrome** L.S. Henriques, M.H. Vaisbich, A. Ferraro, F. Matos, V. Koch. *Pediatric Nephrology, Instituto da Criança, São Paulo, Sao Paulo, Brazil.*
**Objectives:** To verify possible differences in the pharmacokinetic parameters of Cyclosporin (Neoral) between nephrotic children on remission and relapse.
**Methods:** This study evaluates the pharmacokinetic parameters of CSA during remission and relapse of the nephrotic syndrome with the same dose of CSA, with normal renal function and C0 between 50 and 150 ng/ml who achieved complete remission. We evaluated the 12-hour area under the curve ($AUC_{0-12}$). The patients were hospitalized to measure CSA trough level (C0) and after 1, 2, 4, 6, 8 and 12 h of the drug administration. We calculated the $AUC_{0-12}$ for each patient.
**Results:** We studied 10 children (mean age at presentation, 3.0 ± 1.6 years). There was no significant difference between creatinine clearance during remission and relapse (p=0.2). We detected no significant differences at any points of the curve during remission vs. relapse; $AUC_{0-4}$ (on remission r= 0.95 and on relapse r= 0.93) and C2 (on remission r=0.86 and on relapse r=0,80) were the parameters that correlated best with $AUC_{0-12}$. We were not able to detec a significant correlation between cholesterol, albumin and 24 hour proteinuria with $AUC_{0-12}$.
**Conclusions:** We found no significant differences at any points of the curve during remission vs. relapse. Our study suggests C2 and $AUC_{0-4}$ could be the more adequate parameters to fit CSA dosage in nephrotic children. We observed no significant influence of the serum levels of cholesterol, albumin or hematocrit or proteinuria on the pharmacokinetics of CSA. More controlled studies should be done to verify the target value of C2 to maintain remission with minimal toxicity.

## Abstract# 459

**Enteric Coated Mycophenolate Sodium in Patients with CDNS** G.G. Vallejo, M.M. Liern, V.V. De Reyes. *Nephrology Unit, Hospital de Niños Ricardo Gutierrez, Buenos Aires, Argentina.*
**Objectives:** To asses the effect of EC-MPS in treatment of patients with SDNS.
**Methods:** We traeted 13 patients during 12 months ( age Mean 8 y.)
**Inclusion criteria :** glomerular filtration rate normal.
**Exclusion criteria:** Leukopenia, tumoral or infective active disease
**Treatment**: EC-MPS: 450-700 mg/m2/day, methyl prednisone was progressively decreased until complete withdrawal.
**Results:** 8 patients remained in remission.
**relapsing episodes patient/year: Pre-** EC-MPS 3 **Post-**EC-MPS 0.

EVOLUTION

| PATIENT | URINARY PROTEIN mg/m2/h | CREATININE (mg/dl) | CHOLESTEROL (mg/dl) | ALBUMIN (g/dl) | TIME UNTIL STEROID WITHDRAWAL (months) |
|---|---|---|---|---|---|
| | Pre / Postt | Pre / Post | Pre / Post | Pre / Post | |
| 1 | 75 / 4 | 0.5 / 0.4 | 656 / 221 | 1.1 / 3.7 | 1.5 |
| 2 | 54 / 3 | 0.8 / 0.8 | 488 / 208 | 1.2 / 4 | 2 |
| 3 | 48 / 2 | 0.3 / 0.5 | 772 / 171 | 2.4 / 4.2 | 2 |
| 4 | 46 / 1 | 0.4 / 0.6 | 512 / 198 | 1.6 / 3.9 | 1.8 |
| 5 | 54 / 3 | 0.8 / 0.9 | 389 / 189 | 1.5 / 4.0 | 3.3 |
| 6 | 78 / 4 | 0.7 / 0.6 | 459 / 148 | 1.1 / 3.9 | 3.8 |
| 7 | 44 / 4 | 0.9 / 1.1 | 605 / 196 | 2.2 / 4.4 | 2.1 |
| 8 | 87 / 2 | 0.5 / 0.7 | 443 / 176 | 2.3 / 3.7 | 3.3 |
| 9* | 66 /18 | 1.1 / 1.0 | 390 / 255 | 2.1 / 4.1 | 3.0 |
| 10* | 71 / 14 | 0.9 / 0.9 | 652 / 248 | 0.9 / 3.8 | 2.5 |
| 11 | 85 / 46 | 1.0 / 1.1 | 790 / 778 | 0.8 / 1.0 | 3.8 |
| 12 | 62 / 55 | 0.7 / 0.5 | 894 / 890 | 2.0 / 0.8 | 3.2 |

**Conclusions:** the number of relapsing episodes decreased significatly.

## Abstract# 460

**Glomerular Diseases in the Gambia/Africa** U. Vester,[1] M. Hoelscher,[1] A. Fombah,[2] M. Tapgun,[2] U. Helmchen,[3] T. Corrah,[2] P.F. Hoyer.[1] *[1]Pediatric Nephrology, University Hospital, Essen, Germany; [2]Medical Research Council Laboratories, Banjul, Gambia; [3]Dept of Pathology, University Hospital, Hamburg, Germany.*
**Objectives:** Nephritic or nephrotic syndrome is a formerly underestimated cause of hospital admission in The Gambia/West-Africa. However, the underlying diseases are mostly unknown and diagnostic or therapeutic options poorly defined. The aim of the study was to evaluate the nature of glomerular diseases by analyzing clinical and laboratory data and including renal biopsy.
**Methods:** Renal function, blood pressure, proteinuria and hematuria were assessed in 100 children and adults (mean age 15.4±11.2 yrs, range 1.8 – 52 yrs) admitted with edema and proteinuria at the hospital of the Medical Research Laboratories in Fajara / The Gambia. Percutaneous renal biopsy was performed locally after informed consent.
**Results:** The main diagnosis were post-infectious glomerulonephritis (PIGN, n=42, age 8.3±6.7 yrs), FSGS (n=27, age 22±11.8 yrs), MCNS (n=15, age 19.1±10.5 yrs) and membranous nephritis (n=7, age 18.9±9.9 yrs).
Hypertension and hematuria were more prevalent in cases with PIGN whereas serum albumin was higher (34.1±10.2 g/l) compared to patients with FSGS (19.8±7.1 g/l), MCNS (17.3±8.5 g/l) or membranous nephropathy (21.7±6.7 g/l).
Prednisolone treatment resulted in remission in all 9 treated patients with MCNS while 10 out of 11 treated patients with FSGS were steroid-resistant.
**Conclusions:** The spectrum and age distribution of glomerular diseases in The Gambia is quite different from that reported in the northern hemisphere. Our data may serve to guide diagnosis and treatment for glomerular diseases in West-Africa.

## Abstract# 461

**High Incidence of Hemolytic Uremic Sndrome in Switzerland Is Associated with Indicators of Livestock Farming Intensity** M. Fontana,[1] H. Schmid,[2] E. Girardin,[3] T.J. Neuhaus,[1] M.G. Bianchetti,[4] C. Rudin,[5] R.O. von Vigier.[6] *[1]Children's Hospital, Lucerne, Switzerland; [2]Federal Office of Public Health, Bern, Switzerland; [3]University Children's Hospital, Lausanne & Geneva, Switzerland; [4]Department of Pediatrics, Bellinzona & Mendrisio, Switzerland; [5]University Children's Hospital, Basel, Switzerland; [6]University Children's Hospital, Bern, Switzerland.*
**Objectives:** Survey of age-specific incidence rate of childhood hemolytic-uremic syndrome (HUS) and association of Shiga-toxin associated HUS (Stx-HUS) with indicators of livestock farming intensity.
**Methods:** Epidemiological-ecological analysis based on nationwide data through the Swiss Pediatric Surveillance Unit (1997-2003) and the national census (Swiss Federal Statistical Office).
**Results:** One hundred-fourteen cases were registered, 88% were ≤5 years old. The annual incidence rate was 1.42 (0.60–1.91) and 4.23 (1.76–6.19) per $10^5$ children ≤16 and ≤5 years, respectively ($P$<.01). Stx-HUS was more frequent compared to non-Stx-HUS and more frequent in younger (≤5 years) patients ($P$<.01). The present incidence rate in Switzerland is higher compared to data from most other national studies. Strong association was found between incidence rate of Stx-HUS and indicators of rural density (livestock breeding/ population, cattle/cultivated area), $P$<.05 and $P$<.01 in children ≤16 and ≤5 years, respectively.
**Conclusions:** HUS is frequent in young Swiss children and is mostly associated with Shiga-toxin producing *Escherichia coli* (STEC). The incidence rate significantly correlates with livestock farming intensity, supporting the impact of contact with animals or fecal contaminants in transmission of STEC to humans.
**DISCLOSURE:** *Neuhaus, T.J.*: Stockholder, Spouse has stocks from Novartis. *von Vigier, R.O.*: Stockholder, Myself; stocks from GSK, Novartis & Roche.

## Abstract# 462

**Long Term Follow up of Severe IgA Nephropathy Treated with Prednisone and Azathioprine** A. Wilhelm-Bals, P. Parvex, E. Girardin. *Pediatric Nephrology, University Hospital, Geneva, Switzerland.*
**Objectives:** IgA nepropathy (IgAN) is the most common nephritis in children. It was first considered a benign condition. It is now clear that a large number progress to renal failure. The appropriate treatment is still a matter of controversy.
We report 8 cases of severe IgAN treated with prednisone and azathioprine.
**Methods:** 8 patients (pts) with severe IgAN presenting with heavy proteinuria (PU) or renal failure, underwent renal biopsy and were treated with solumedrol (3pushs1g/1.73m2/d 4/8 pts), prednisone (start 2mg/kg/d 8/8 pts), azathioprine (AZA 1.5-2 mg/kg/d 8/8 pts) and ACE ± ARI medication.
PU and creatinine clearance (creatCl ml/mn/1.73m2) were assessed at 0, 6, 12 months (m) and at the end of follow-up.
A control biopsy, was done after 1 year (y) in 6/8 pts.
Remission was considered in absence of PU (prot/creat < 20g/mol) and normal creatCl.
**Results:** Patients: 6 M, 2 F, mean (M) age 10.5y
Prednisone treatment length: M 18 m (11-36m), AZA: M 13.5 m (12-24m). At last follow up M 7y (3-10y), 1/8 nephrotic PU and chronic renal disease (CRD, stage IV).

| | T0 | 12 m | last follow up |
|---|---|---|---|
| macrohematuria | 8/8 | 3/8 | 4/8 |
| Oedema | 3/8 | 0/8 | 1/8 |
| PU g/mol | 8/8 M 411 (130-1209) | 1/7 70 | 4/8 3/8 M 70 1/8 818 |
| creatCl ml/mn/1.73m2 | N 4/8 M 148 (106-202) RF 4/8 M 44 (24-89) | N 8/8 M 128 (93-244) | N 7/8 M 138 (90-225) RF 1/7 24 |
| Mesangioproliferation | 7/7 | 6/6 | |
| Glomerulosclerosis | 5/7 | 1/6 | |
| Tubular atrophy/fibrosis | 1/7 | 2/6 | |
| Crescent % | 7/7 M 32% (5-59) | 4/6 M 5.5% (4-15) | |

N: normal; RF: renal failure

**Conclusions:** The combination of AZA-prednisone showed good efficacy in inducing remission (6/7 at 1y) and regression of histologic lesion. At last follow up, 4pts are still in remission, 3 have mild PU and 1 has CRD.

## Abstract# 463

**Congenital Nephrotic Syndrome in New Zealand Children, Prolonged Renal Survival without Renal Replacement Therapy** W. Wong, M.C. Morris, T. Kara. *Nephrology, Starship Children's Hospital, Auckland, New Zealand.*

**Objectives:**
Congenital nephrotic syndrome of the Finnish type (CNF) leads to an inexorable progression to end stage renal failure within the first decade of life. Congenital nephrotic syndrome (CNS) infants in New Zealand appear to have a different course to those with the typical Finnish mutation. The demography, clinical features and outcome of all with CNS children born in New Zealand is reviewed.

**Methods:**
All cases of CNS diagnosed between 1975 and 2009 were reviewed. Demographic data, clinical features, treatment and outcome were extracted from clinical records and contact with nephrologists. Mutation analysis was undertaken in patients born after 1995.

**Results:**
Thirty one patients with CNS were identified, 21 children of Maori descent, 10 Caucasians. Age at follow up ranged from 2.5 months to 29 years. Twelve had died (6/10 Caucasians, 6/21 Maori) at 4 days to 7 months. None of the 21 Maori children received bilateral nephrectomies. Maori children with CNS had displayed a highly variable and protracted timeline to end stage renal failure with some patients developing end stage renal failure as early as 3yrs of age while other not developing chronic renal failure until their third decade of life. Mutation analysis of NPHS1 showed mutations different to classical Fin Major or Minor.

**Conclusions:**
Congenital nephrotic syndrome in New Zealand Maori children exhibit a different clinical course to Caucasian children and have mutations which are unique to this racial group. Conservative medical therapy is often successful in controlling nephrotic symptoms and promoting acceptable growth in Maori children with CNS.

## Abstract# 464

**Clinical-Pathological Study of 212 Primary Focal Segmental Sclerosis (FSGS) in Children** J.H. Xiao, C.J. Liu, Y.J. Yang, X.S. Wang, P.J. Huang. *Department of Pediatrics, Peking University First Hospital, Beijing, China; Department of Pediatrics, Peking University First Hospital, Beijing, China; Department of Pediatrics, Peking University First Hospital, Beijing, China; Department of Pediatrics, Peking University First Hospital, Beijing, China; Department of Pediatrics, Peking University First Hospital, Beijing, China.*

**Objectives:** The correlation of clinical;pathological features of primary focal segmental sclerosis (FSGS) in children.

**Methods:** 212 pedictric patients with biopsy-proven primary FSGS were included Five pathologic variants of primary FSGS are collapsing (COLL), cellular (CELL), glomerular tip lesion (GTL), perihilar, and not otherwise specified (NOS). Retrospective analysis were made.

**Results:** Nephrotic syndrome is the main cinical manifestation (N=178;84%), GTL variants were mostly appeared to be nephrotic syndrome (N=28). NOS (N=86(40.6%)) were the most common types. GTL and NOS variants initially appeared to be responsive to medication, than became resistent to steroid. Perihilar;CELLand COLL were appeared to be primary steroid resistent. COLL FSGS had the highest rate of ESRD.

**Conclusions:** FSGS is defined as a clinical-pathologic syndrome. Cortico-resistant nephrotic syndrome is the main manifestation. Collapsing glomerulopathy is the most aggressive variant of fsgs. GTL variant may be the best types. Different histologic variants of FSGS have substantial differences in clinical features and substantial differences in renal outcomes.

## Abstract# 465

**Influence of Steroid Medication on Bone Mineral Density in Children with Nephrotic Syndrome** M.A.B. Zahrane, A. Esmael. *Pediatric Nephrology, Aboul-Rich-Hospital, Cairo-University, Egypt; Radiology, Kasr el enni, Cairo-University, Egypt.*

**Objectives:** Glucocorticoid induced osteoporosis is caused by decreased bone formation and increased bone resorption. So, in our study we evaluated the effects of long and high dose steroid on bones in children with nephrotic syndrome.

**Methods:** We performed dual-energy X-ray absorptiometry of the spine in 37 children with nephrotic syndrome and correlate the result of dexa with duration of steroid, age of onset, type of NS, laboratory data.

**Results:** Bone mineral density(BMD) at lumbar spine (L2-L4) was measured by dual energy X-ray absorptiometry in 37 children with nephrotic syndrome with mean age (8.6 ± 3.3) years, according to their response they were classified as steroid responsive wh (67.6%) and the steroid resistant (32.4%), the steroid responsive group is subdivided into frequent relapsers (FR) (21.6%) and steroid dependent (46%). The mean age of onset were (4.9 ± 2.8) years, mean duration of steroids (3.6 ± 2) years, (89.2%) were on steroid and (10.8%) were off steroid, as regards laboratory data all were within normal ranges. As regard BMD patients were divided into 3 groups: Mild osteopenic group (48.7%), severe osteopenic group (18.9%) and the remaining group are normal (32.4%). By comparison the severe osteopenic group have older age of onset (P = 0.003) and 85.7% of this group are steroid resistant., Z-score shows –ve correlation with age of the patients (r = -0.501) and +ve correlation with serum albumin and alkaline phosphatase, r = 0.408, r = 0.427 respectively.

**Conclusions:** So, we can conclude that patients with nephrotic syndrome, on long and high dose of steroid have decrease bone density, especially the steroid resistant group.

## Abstract# 466

**IgA Nephropathy in Chinese Children, Severe or Not?** Y.-H. Zhai, Q. Cao, J. Chen, X.-Y. Fang, Q. Shen, H. Xu. *Department of Nephrology and Rheumatology, Children's Hospital of Fudan University, Shanghai, China.*

**Objectives:** To analyse the pathology of IgA nephropathy (IgAN) with various clinical features in 158 Chinese children retrospectively.

**Methods:** Children with primary IgAN were divided clinically into two groups, asymptomatic (aIgAN) and symptomatic IgAN (sIgAN). Prognosis was graded by urine protein excretion and renal function.

**Results:** The 125 children in sIgAN were younger than that in aIgAN (8.9±0.3 yr. vs. 10.5 ± 0.6 yr, $P$<0.05). Most children (50%) clinically presented macroscopic hematuria. The urine protein excretion of children in sIgAN was more than that in aIgAN (1.8±0.2. vs. 0.4±0.1g/d) and the average course before renal biopsy was longer than that of sIgAN (466±115 .vs. 159±40d)( $P$<0.01). More children in aIgAN had minor histologic lesions than those in sIgAN, manifested by lower mesangial score and less presentation of other pathologic variables ($P$<0.05), however, histologically severe cases in 2 groups were similar (~10%). By logistic analysis, hypertension was related to the severity of histological lesions (OR:9.77, 95%CI:1.78~53.62, $P$ <0.01). Most children in both groups had good clinical outcome at >1yr of follow up and complete clinical data from 68 cases indicated hypertension related to short-term prognosis (OR:15.27, 95%CI:1.08~21.84, $P$ <0.05).

**Conclusions:** Most in aIgAN have minor histologically lesions, but a few of them may have severe lesions. The short-term prognosis is good and hypertension is related to the severity of histologic lesions and prognosis. So monitoring blood pressure, and developing appropriate indication of renal biopsy in children with suspected IgAN may have potent significance in retarding disease progress.

# Transplantation

## Abstract# 467

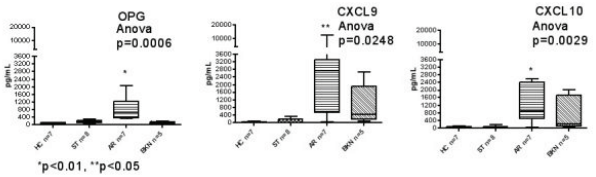
**(O-57)**

**Non-Invasive Test for Renal Transplant Rejection in Children** J. Jackson, J. Rossio, L Kean, A. Kirk. *Emory, Atl, GA, United States; Life Tech., MKE, WI, United States.*

**Objectives:** Noninvasive testing for renal dysfunction is critical for pediatric transplant recipients, who have a long-term need for transplant monitoring, and for whom invasive testing is problematic. We have investigated a urine-based assay for CXCL9, CXCL10, and osteoprotegerin (OPG), proteins associated with T cell based inflammatory processes.

**Methods:** Urine collected from pediatric healthy, non-transplanted controls and transplant patients (n=27) was analyzed. Each sample was verified to be representative of a single condition: healthy control (HC, n=7), stable transplant with normal renal function (ST, n=8), acute rejection (AR, n=7), or BK nephritis (BKN, n=5).

**Results:** Elevated urinary OPG clearly distinguished AR from all other analysis groups. Using an ROC curve, 236.8 pg/mL was chosen as an optimal cutoff, which resulted in perfect sensitivity (1.0) and high specificity (0.77). The PPV using this cutoff was 0.7 and NPV was 1. While OPG identified patients with AR, elevations in CXCL9 and CXCL10 identified patients experiencing either AR or BKN.




**Conclusions:** These noninvasive assays identified patients experiencing post-transplant graft inflammation due to either AR or BKV. The ability to non-invasively screen for either AR or BK nephritis provides ease and flexibility for ongoing pediatric transplant monitoring, identifying patients in need of attention and possible intervention by a transplant nephrologist.
**DISCLOSURE:** *Rossio, J.*: Other, Employee.

## Abstract# 468
**(O-58)**

**Transplantation in Atypical Hemolytic Uremic Syndrome** [#§] M. Riedl, J. Hofer, A. Rosales, Y. Yeutukhova, T.C. Jungraithmayr, L.B. Zimmerhackl. *Pediatrics, Medical University, Innsbruck, Austria.*
**Objectives:** Transplantation in atypical hemolytic uremic syndrome (aHUS) is characterized by a high rate of disease recurrence and subsequent graft loss.
**Methods:** Since 2001 the European Paediatric Research Group for HUS and related disorders *(HUSnet)* (www.hus-online.at) has been collecting data and performing complement analysis in patients with atypical STEC negative HUS. In 25 out of 135 patients of the registry 47 renal transplantations (n=1-4) were performed, including 5 patients with heterozygous factor H (fH), 1 homozygous MCP, 2 heterozygous factor I (fI), 1 combined mutation including fH and fI and 1 patient with fH antibodies and CFHR1 deletion.
**Results:** Graft loss occurred in 33/46 transplants (72%), in median after 7 months (1 day – 11.4 years). The main reasons for graft loss were HUS recurrence in 76% (25/33), followed by rejection in 5 cases (15%). Loss of function was reported in 8 of 28 grafts (29%) in the first month after transplantation, in 6 cases due to disease recurrence. 16 of 28 (57%) grafts were lost within the first year after renal transplantation.
The follow-up time of the 13 grafts with preserved renal function is about 15 months in median (6 days – 8 years).
**Conclusions:** The data of our registry shows that graft failure due to disease recurrence occured in more than 50% in the first year of transplantation. New treatment options like Eculizumab give hope for the future to perform renal transplantations in aHUS.
[#] Supported by ÖNB grant
[§] cooperation partners: Toenshoff, Heidelberg; Montoya, Munich; Konrad, Muenster; Reusz, Budapest; Pape, Hannover; Wygoda, Leipzig; Mueller-Wiefel, Hamburg; Fiedler, St. Poelten; Simonetti, Bern.

## Abstract# 469
**(O-59)**

**Chronic Respiratory Symptoms and Bronchiectasis in Paediatric Renal Transplant Recipients on Mycophenolate Mofetil** N.M. Dolan, R. Suri, C. Owens, S.D. Marks. *Great Ormond Street Hospital for Children NHS Trust, London, United Kingdom.*
**Objectives:** Evaluate incidence of respiratory complications,in particular bronchiectasis, in paediatric renal transplant recipients(RTR) receiving mycophenolate mofetil(MMF).
**Methods:** Review of renal transplant database and RTR with history of chronic cough and chest signs were identified.Immunosuppression used,interval from transplantation to symptom onset and associations between therapy and onset of symptoms were documented.
**Results:** Of 143 transplant recipients,93(65%) received MMF as part of immunosuppression.11(12%) of these patients developed chronic respiratory symptoms,compared to none(0/50) on other immunosuppression($p<0.05$).50 %(5/10) symptomatic patients who underwent CT scanning had radiographic evidence of bronchiectasis.Time to onset of symptoms in patients receiving MMF ranged from 9- 96(mean 31;median 24)months.In asymptomatic patients,duration of therapy ranged from 1-98(mean 32;median 29) months($p>0.05$).Duration of immunosuppression among patients who received non-MMF regimens ranged from 10 - 170(mean 55;median 49.5)months($p<0.001$).Mean 12-hour trough MPA levels in symptomatic and asymptomatic patients were 6.85 and 3.78 mcg/ml respectively($p<0.001$).
**Conclusions:** High incidence(12%) of significant chronic respiratory symptoms related to the use of MMF.Significant association between high trough MPA levels and development of symptoms.No association between duration of MMF-based,or other immunotherapy and onset of respiratory symptoms.Bronchiectasis should be considered in all patients who develop chronic respiratory symptoms post renal transplantation.

## Abstract# 470
**(O-60)**

**Allograft Survival in Patients with Henoch-Schonlein Purpura: Analysis of the United Network for Organ Sharing (UNOS) Database**
J.P. Samuel,[1] C.S. Bell,[1] D.A. Molony,[2] M.C. Braun.[1] *[1]Division of Pediatric Nephrology and Hypertension, UTHSC-H, Houston, TX, United States; [2]Department of Internal Medicine, UTHSC-H, Houston, TX, United States.*
**Objectives:** Henoch-Schonlein Purpura (HSP) is the most common form of renal vasculitis in childhood, however there is little data on outcomes of renal transplantation in patients with HSP. This study was designed to examine the impact of HSP on allograft survival in a well-defined transplant population.
**Methods:** The UNOS database (1987-2005) was analyzed to determine the outcomes of primary renal transplants in patients with HSP; each case was matched 1:3 with non-HSP patients and controlled for age, gender, donor source, ethnicity, and year of transplantation.
**Results:** Of the 189,217 primary renal allografts in the UNOS database, 339 (0.18%) were in individuals with a diagnosis of HSP. Compared to the remainder of the database, the HSP group was younger (mean age at transplantation of 28.1 +/- 14.9 years vs. 44.2 +/- 15.4 years), had a higher proportion of females (47% female vs. 40%), live donors (50% vs. 35%), and Caucasians (77% vs. 60%). Only 97 HSP patients (28.6%) were <18 years of age at the time of transplant. Graft survival for patients with HSP was 77.0% at 5 yrs and 55.4% at 10 yrs compared to 74.6% and 47.9% in the control population ($p> 0.05$). Graft failure from recurrent disease was reported in 13.5% of patients with HSP, compared to 5.2% in the database as a whole.
**Conclusions:** This study represents the largest cohort to date evaluating the outcomes of renal transplantation in HSP. While there was an increased risk of graft failure from recurrent disease, overall graft survival in patients with HSP was comparable to matched controls.

## Abstract# 471
**(O-61)**

**Succesful Use of Eculizumab after Kidney Transplant for Atypical Hemolytic Uremic Syndrome (aHUS) Associated with a *CFH* Mutation** J.-C. Davin, V. Gracchi, A.H. Bouts, J.W. Groothoff. *Pediatric Nephrology Department, Emma Children's Hospital-Academic Medical Centre, Amsterdam Z-O, Netherlands.*
**Objectives:** Kidney transplant in patients with aHUS is associated with a poor outcome because of recurrent disease, especially in patients known to have a factor H mutation. Long-term prophylactic plasma exchange (PE) and combined liver-kidney transplant have prevented graft loss caused by recurrence. However, the mortality associated with liver transplant is not negligible, and prophylactic PE requires permanent vascular access and regular hospitalization and exposes the patient to allergic reactions to plasma. Eculizumab is a high-affinity humanized monoclonal anti-C5 antibody that prevents C5a and MAC formation. We report the use of eculizumab instead of PE to prevent aHUS recurrence after transplantation in a patient with highly plasma-dependent aHUS related with a CFH mutation.
**Methods:** A 17 years old girl with aHUS associated with CFH mutation and ESRF since the age of 3 y presenting with multiple cerebral vascular stenosis, PE dependency and severe allergic reactions to plasma after a third kidney transplantation received 900 mg of eculizumab intravenously 2 h after the last PE.The same dose of eculizumab was given weekly for 4 weeks. Thereafter, 1,200 mg was given every 2 weeks indefinitely. Anti-meningococcus vaccination was performed prior to eculizumab.
**Results:** Twelve months after the start of eculizumab and PE discontinuation, no relapse had occurred and plasma creatinine level is stable at 128 micromol/L. No side-effect was observed.
**Conclusions:** This and other reports suggest that the promise of complement inhibitors in the management of aHUS is going to be fulfilled.
**DISCLOSURE:** *Davin, J.-C.*: Other, Novartis Transplantation Advisory Board for Netherlands. *Groothoff, J.W.*: Grant/Research Support, Oxthera.

## Abstract# 472
**(O-62)**

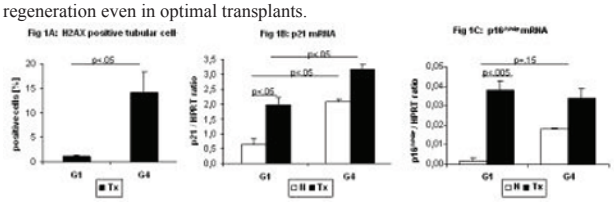
**Telomerase Knockout Mice – A Model for Renal Transplants with Limited Regenerative Capacity** C. Jacobi,[1] A. Weißbrodt,[1] S. Rong,[2] V. Bröcker,[3] C. Wang,[4] F. Güler,[2] A. Melk.[1] *[1]Pediatric Nephrology, Medical School, Hannover, Germany; [2]Nephrology, Medical School, Hannover, Germany; [3]Pathology, Medical School, Hannover, Germany; [4]Ageing and Health, Univ., Newcastle, United Kingdom.*

**Objectives:** Cellular senescence results in reduced regenerative ability. Telomerase (Terc) KO mice show critically short telomeres in late generations. The outcome of Terc KO kidneys in a syngeneic life-supporting transplantation (tx) model is evaluated.

**Methods:** Early (G1,n=9) and late (G4,n=5) generation male Terc KO kidneys were transplanted into female WT recipients and sacrificed after 6 wks.

**Results:** G4 transplants showed significantly more interstitial fibrosis (IF) and a tendency to more glomerulosclerosis. Telomere-dependent senescence markers were significantly higher in G4: more H2AX pos. tubular cells (fig.1A), higher p21 expression (fig.1B). p16 reflecting telomere-independent senescence, even though higher in G4 prior to tx, was not different after tx (fig.1C).

**Conclusions:** Mild transplantation stress induced telomere-dependent and -independent senescence. This was more pronounced leading to increased IF in G4 mice that reflect the situation of an old donor. But p16 increases in G1 suggest that senescence can be triggered even in ideal donors. As this tx model enables us to mimic the human situation, it should be used to explore factors that reduce regeneration even in optimal transplants.




## Abstract# 473
**(O-63)**

**Changes of Left Ventricular Mass in Children on Hemodialysis and Post Transplant – A Prospective Follow-Up Study** B. Letavernier,[1] F. Lorton,[1] B. Aoun,[1] I. Tillous-Borde,[2] T. Ulinski.[1] *[1]Pediatric Nephrology, Armand Trousseau Hospital - APHP - UPMC, Paris, France; [2]Pediatric Cardiology, Armand Trousseau Hospital - APHP - UPMC, Paris, France.*

**Objectives:** To analyse changes of left ventricular hypertrophy (LVH) in a cohort of ESRD children from the beginning of hemodialysis (HD) until four years post renal transplant (RT).

**Methods:** We studied prospectively the changes of left ventricular mass (LVM) during the whole hemodialysis (HD) period and during four years after RT and analysed contributing factors and possible predictability for LVH. 24 children (10.3; 2.1 17.5 years) were included.

**Results:** LVMi at HD onset was 101±5.7 g/m$^2$. After one year on HD there was a significant decrease (89 4.5 g/m$^2$) and stabilization until renal transplantation (86.3±4.7 g/m$^2$). Six months post transplant a dramatic re-increase was noticed (104±9.0 g/m$^2$) followed by a decrease until two year post RT (83.5 g/m$^2$). After the second year post transplant LVM developed very heterogeneously (99±10.0 g/m$^2$), and a subset of patients had moderate to severe LVH.

In the HD period LVM was positively correlated with mean, systolic and diastolic blood pressure (MBP, SBP, and DBP), but not with Hb levels. However, in the post RT period only correlation with SBP was identified, but surprisingly not with DBP, MBP, or renal function. There was no difference in patients with CyA or FK based immunosuppression. The risk to have a LVH one year post RT was partially predictable by LVM at the beginning of the HD period ($p<0.01$).

**Conclusions:** LVH decreases during HD and re-appears after the second year post transplant in about 50% of patients in particular those with severe LVH at the beginning of the HD period.

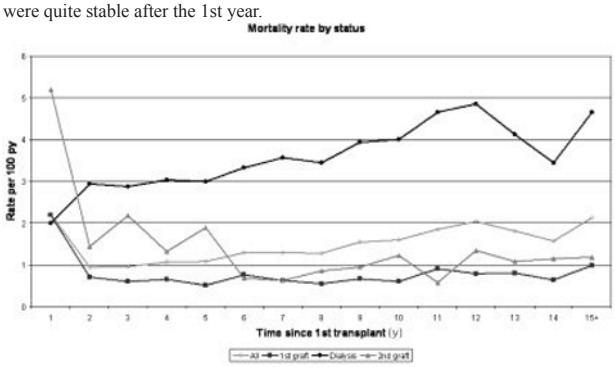
## Abstract# 474
**(O-64)**

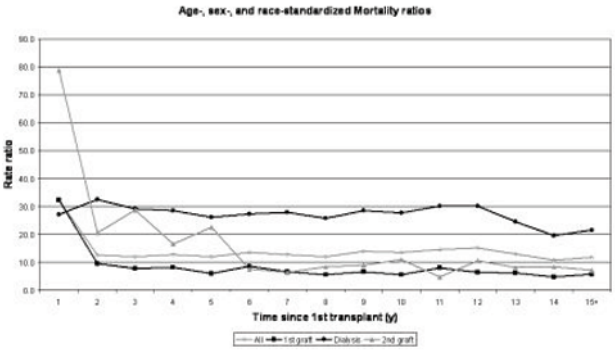
**Change in Mortality Risk over Time in Young Renal Transplant Recipients** B. Foster,[1] X. Zhang,[2] M. Dahhou,[2] R. Platt,[1] J. Hanley.[1] *[1]McGill University, Montreal, QC, Canada; [2]Montreal Children's Hospital Research Institute, Montreal, QC, Canada.*

**Objectives:** To determine the impact of increasing accumulated time since 1st transplant on mortality risk in young renal transplant recipients.

**Methods:** Patients who received a 1st transplant at <21 years of age (1983-2006), whose data were recorded in the USRDS were included. Mortality rates were calculated for each 1-year interval after the 1st transplant for patients observed in each of the following statuses: 1st transplant functioning, dialysis, 2nd transplant functioning. Patients changed status upon return to dialysis or retransplant. Age-, sex-, and race-standardized mortality ratios (SMR) relative to the general American population were calculated for each 1-year interval.

**Results:** Of 18,911 patients with median 10.8 y follow-up, 2713 died. Mortality rates were highest in the 1st post-transplant year and after return to dialysis. SMR were quite stable after the 1st year.




**Conclusions:** The relative risk of mortality does not increase substantially with increasing accumulated time since 1st transplant.

## Abstract# 475
**Protocol Biopsies 6 Months after Pediatric Kidney Transplantation: Influence of Different Factors** N.K. Kanzelmeyer,[1] V. Broecker,[2] L. Pape.[1] *[1]Hanover Medical School, Pediatric Nephrology, Hannover, Germany; [2]Hanover Medical School, Pathology, Hannover, Germany.*

**Objectives:** Protocol biopsies (PB) are seldom performed in pediatric kidney transplantation. It is not known what factors influence the results.

**Methods:** We performed PBs in 79 children 6 months after kidney transplantation (mean age at renal transplantation 10 ± 5 years). PBs were evaluated using BANFF 2007-criteria by two local pathologists. The influence of different factors on the results was tested by co-variance analysis.

**Results:** PB results were influenced by the immunosuppressive regimen in co-variance analysis (p=0.045): children treated with mTOR-Inhibitor and low dose cyclosporine A (CsA) showed less borderline findings (18% vs. 29%, p=0.000 ANOVA) and no subclinical rejection (0% vs. 19%) compared to patients treated with regular-dose CsA and mycofenolat mofetile. There was a tendency for less interstitial fibrosis and tubular atrophy (IF/TA) in younger children (8 ± 5 years vs. 11 ± 5, covariance analysis: p= 0.095, ANOVA: p=0.072). There was no influence of other factors on PBs (table 1).

Covariance analysis (dependent variable: result of biopsy)

| | |
|---|---|
| Pred., CNI, MMF. vs. Pred., low dose-CNI, Everolimus | 0.000 * |
| Mismatches | 0.578 |
| Deceased donors vs. living donor | 0.406 |
| Preemptive vs. post dialysis | 0.412 |
| Donor age | 0.910 |
| Recipient age | 0.095 (*) |
| Cold ischemia time | 0.780 |
| GFR at Tx | 0.530 |

**Conclusions:** Immunosuppression with mTOR-inhibitor and low dose CsA leads to a decreased number of borderline findings and subclincal rejection. In younger recipients the amount of IF/TA is slightly decreased. The influence on long term outcome has to be evaluated.

## Abstract# 476

**Protocol Biopsy-Driven Interventions after Pediatric Renal Transplantation** N.K. Kanzelmeyer,[1] T. Ahlenstiel,[1] V. Broecker,[2] L. Pape.[1] [1]*Pediatric Nephrology, Hannover, Germany;* [2]*Pathology, Hannover, Germany.*

**Objectives:** The therapeutical value of protocol biopsies in renal transplant recipients remains unclear.

**Methods:** We performed protocol biopsies in 57 children 6 months after renal transplantation. Based on our local treatment algorithm, we increased CNI dose in patients with borderline findings. In cases of Banff grade Ia, 6 prednisolone-iv-pulses were given and CNI dose was increased. CNI toxicity and polyomavirus nephropathy led to a reduction of CNI dose. GFR was compared to a control group of 51 children without protocol biopsies transplanted in the same period.

**Results:** 42% of protocol biopsies had no pathological changes, 24% interstitial fibrosis and tubular atrophy (IF/TA). Borderline findings were detected in 11%, Banff grade Ia in 15%, calcineurin inhibitor (CNI) toxicity in 8% and one case showed polyomavirus nephropathy. GFR measured at 1.5, 2.5 and 3.5 years after transplantation revealed no differences between the groups. Only GFR 3.5 years after transplantation was significantly higher than in the control group (57 ± 17 vs. 46 ± 20, p=0.036, ANOVA).

**Conclusions:** The performance of protocol biopsies followed by a standardized treatment algorithm did not lead to better long term graft function. Prospective randomized studies comparing the GFR after renal transplantation with and without protocol biopsy driven interventions are needed.

## Abstract# 477

**Long-Term Results of Combined Liver and Kidney Transplantation (LKTx) in Children with Autosomal Recessive Polycystic Kidney Disease (ARPKD)** M.J. Kemper,[1] I. Klaassen,[1] M. van Husen,[1] A. Briem-Richter,[2] J. Pollock,[3] R. Ganschow,[2] F. Brinkert.[2] [1]*Pediatric Nephrology, University Medical Center Hamburg-Eppendorf, Hamburg, Germany;* [2]*Pediatric Hepatology, University Medical Center Hamburg-Eppendorf, Hamburg, Germany;* [3]*Transplantation Surgery, University Medical Center Hamburg-Eppendorf, Hamburg, Germany.*

**Objectives:** ARPKD often results in end-stage-renal disease but also congenital liver fibrosis is a feature and thus LKTx may be necessary. Aim of the study was to analyse the long-term course of children with ARPKD after LKTx.

**Methods:** We conducted a retrospective chart analysis of seven patients who underwent a combined Tx between the years 2003 and 2009. Clinical and laboratory as well as longitudinal growth data were collected.

**Results:** Seven children were transplanted at a median age of 10 years (1.7-16 years) and with a median weight of 18.2 kg (10.5–55 kg). After a median follow-up of 2.7 years (0.6-6.3 years) all patients are alive. One patient required re-transplantation of the liver only. The first transplanted patient had to be retransplanted due to kidney artery thrombosis. Recently a futher kidney transplantation was necessary due to graft loss 2º to non adherence. Currently liver and kidney function of all patients are stable. Median serum creatinine is 0.69 mg/dl (0.36-1.57mg/dl) resulting in a eGFR of 120 ml/min/1.73m² (range 68-133 ml/min/1.73m²). Mean height SDS improved form -2.1±0.66 to -1.18±0.65 after 18 months of follow-up.

**Conclusions:** The results of LKTx for patients with ARPKD are encouraging. Both the lack of mortality and the catch-up growth shows that LKTx is an important therapy for patients with ARPKD affecting liver and kidney.

## Abstract# 478

**Comparison of Hyperglycemic Effects of Cyclosporine and Tacrolimus in 105 Cases with Renal Transplantation** M. Koyun,[1] C.S. Dogan,[1] E. Comak,[1] A.U. Gökceoglu,[1] A. Gürkan,[2] A. Dinckan,[2] S. Akman.[1] [1]*Pediatric Nephrology, Akdeniz University, Antalya, Turkey;* [2]*General Surgery, Akdeniz University, Antalya, Turkey.*

**Objectives:** We aimed to analyze to compare the adverse effect of hyperglycemia of tacrolimus and cyclosporine in pediatric renal transplant recipients.

**Methods:** The records of children who were transplanted at our center were evaluated retrospectively. Serum glucose levels were determined on post-transplant days 7, 30, 90 and 180. The patients in both groups received the same glucocorticoid therapy protocol. The target plasma levels of tacrolimus was 10-20 ng/ml for the first 6 months and 5-10 ng/ml thereafter; for cyclosporine 300-350 ng/ml for the first 3 months, 250-300 ng/ml between 3-6 months, 150-250 ng/ml for 6-12 months and 100-150 ng/ml thereafter.

**Results:** 105 children, 58 boys and 47 girls, were enrolled in the study. Mean age at the time of transplantation was 11.8±3.8 years and median follow-up period was 20 months. 78 of the patients were receiving tacrolimus and 27 were CsA. Serum glucose levels were higher in tacrolimus group than CsA at days 7 and 180 (84.0±15.7 g/dL vs 75.3±8.1, p=0.001; 86.2 ± 8.4 g/dL vs 80.5±7.8, p=0.003, respectively); whereas it was similar in both groups at days 30 and 90. Hyperglycemia was observed in 9 patients receiving tacrolimus (11.5%) within post-transplant 1-9 months, whereas no patient receiving CsA had hyperglycemia. Hyperglycemia was persistent in 6 cases, who needed to switch to cyclosporine. Moreover, diabetes mellitus was developed in one of them.

**Conclusions:** Hyperglycemia is more prominent in tacrolimus compared to cyclosporine in pediatric renal transplant recipients.

## Abstract# 479

**Frequency and Risk Factors of Acute Rejection Episodes in 115 Pediatric Renal Transplant Recipients** A.U. Gökceoglu,[1] M. Koyun,[1] C.S. Dogan,[1] E. Comak,[1] A. Gürkan,[2] A. Dinckan,[2] S. Akman.[1] [1]*Pediatric Nephrology, Akdeniz University, Antalya, Turkey;* [2]*General Surgery, Akdeniz University, Antalya, Turkey.*

**Objectives:** The aim of this study was to analyze the prevalence and risk factors for acute rejection episodes in children who underwent renal transplantation.

**Methods:** The records of children who were transplanted at our center were evaluated retrospectively. Acute rejection was defined as an increase of 0.3 mg/dl in serum creatinine.

**Results:** 115 children, 69 boys, were enrolled in the study. Mean age at the time of transplantation was 11.7±3.8 years and median follow-up period was 18 months. 100 of 115 transplants (87%) were from living-related donors. Two patients received kidneys from ABO-incompatible donors and three children from HLA full-mismatch donors. A total of 43 acute rejection episodes were observed in 26 patients (22.6%). One episode was seen in 18 children and more than one episode in the others. Renal biopsy was performed in ten patients. Hypertension was detected in six patients; BK virus infection was accompanying in four. No statistically significant difference was found between acute rejection episodes of patients receiving tacrolimus and cyclosporine (24% vs 16.7%, p>0.05) as well as between living-related and cadavaric donors (24.8% vs 21.4%, p>0.05). Also, we found no effect of HLA mismatches on acute rejection episodes. Five patients who had one or more acute rejection episodes developed graft failure.

**Conclusions:** Acute rejection episodes has still been a reason of significant morbidity in pediatric renal transplant recipients despite novel immunosuppressive agents.

## Abstract# 480

**Acute Humoral Rejection in Pediatric Renal Transplant Recipients: Therapy and Outcome in Three Cases** B. Kranz,[1] E. Kuwertz-Broeking,[1] A. Schulze-Everding,[1] R. Kelsch,[2] H. Wolters,[3] M. Konrad.[1] [1]*General Pediatrics/Pediatric Nephrology, University Children's Hospital Muenster, Muenster, Germany;* [2]*Institute of Transplantimmunology, University Hospital Muenster, Muenster, Germany;* [3]*Clinic of Generqal Surgery, University Hospital Muenster, Muenster, Germany.*

**Objectives:** Acute humoral rejections(AHR) after renal transplantation are associated with acute deterioration of graft function combined with the detection of donor specific antibodies (DSA) and the positive staining of C4d in renal biopsy. Despite a multimodal therapy consisting of steroids, immunoglobulin, Rituximab and plasmapheresis AHR lead to 40% graft failure in adults.A therapy regimen for children has not been validated so far.

**Methods:** We report 3children (9,10 +12y) suffering from AHR after living related renal transplantation.Immunological monitoring prior to transplantation excluded pre-existing DSA and the repetition of forbidden antigens.

**Results:** Two children developped an AHR in their 2nd renal graft 5 days post renal transplant.C4d staining was positive but DSA were detected in one girl only.The second child showed a booster of the DSA of the first renal transplant.A third girl suffered from AHR without prior renal transplant.In addition to DSA AT1-receptor antibodies were detected associated with severe hypertension.All children were treated with immunoglobulin, 10 sessions of plasmapheresis and a single dose of Rituximab.They all recovered from acute renal failure with a GFR of 58-99ml/min/1.73m² (follow up 8,9 and 16 months post AHR).

**Conclusions:** A therapy of immunoglobulin, plasmapheresis and Rituximab seem to be effective in the treatment of AHR in children.

## Abstract# 481

**Post-Transplant Anaemia in Paediatric Renal Allograft Recipients. A Comparison between Mycophenolate Mofetil (MMF) and Azathioprine (AZA) Based Immunosuppression** A. Lamb,[1] J. Boyle,[2] L. Krischock,[1] I.J. Ramage.[1] [1]*Renal Unit, Yorkhill Hospital, Glasgow, United Kingdom;* [2]*Faculty of Medicine, University of Dundee, Dundee, United Kingdom.*

**Objectives:** To compare the prevalence of early post transplant anaemia in MMF or AZA treated paediatric renal allograft recipients.

**Methods:** A retrospective observational study was undertaken of paediatric renal allograft recipients between January 2002 and March 2009. Prior to 2006 standard immunosuppression consisted of AZA,, Tacrolimus and Prednisolone, thereafter MMF, Tacrolimus, Daclizumab and short course prednisolone. Data

collected included haemoglobin (Hb), creatinine erythropoietin stimulating agent (ESA) usage and concomitant immunosuppression. Anaemia was defined as Hb <10.5 g/dL.

**Results:** Sixty three patients {36 male (57%)} aged 2-18 years were identified. 9 were excluded due to switch in therapy. 54 patients were therefore included. 23 (43%) received AZA and 31 (57%) MMF. 2 patients in the AZA group had Day 90 follow-up elsewhere.

Prevalence of Anaemia

| MMF | | AZA | |
|---|---|---|---|
| Baseline | 5/31 (16%) | Baseline | 4/23 (17%) |
| Day 20 | 22/31 (74%) | Day 20 | 11/23 (48%) |
| Day 90 | 9/31 (29%) | Day 90 | 5/21 (24%) |

ESA use was documented in 9 patients all of whom were treated with MMF (9/31 - 29%).

**Conclusions:** Early post transplant anaemia is common in paediatric renal allograft recipients. There appears to be an increased prevalence of anemia at Day 20 in those receiving MMF compared to AZA. The increased use of ESA's in this patient population may reflect an alteration in prescribing practice over time. Given the prevalence of anemia in both groups we advocate the continued use of ESA's in the peri and postoperative transplant period.

## Abstract# 482

**Pharmacodynamic Monitoring of Immunosuppressive Drugs in Pediatric Kidney, Heart and Liver Transplant Patients** A.L. Lapeyraque,[1,2,3] R.M. Brito,[3] V. Durrieu,[3] M.J. Clermont,[2] Y. Théorêt,[1,2,3] F. Le Deist.[1,2,3] *[1]Unité de Pharmacologie Clinique, CHU Sainte Justine, Montréal, QC, Canada; [2]Département de Pédiatrie, CHU Sainte Justine, Montréal, QC, Canada; [3]Centre de Recherche, CHU Sainte Justine, Montréal, QC, Canada.*

**Objectives:** Variability of suceptibility to tacrolimus (Tac) and to mycophenolate mofetil (MMF) are at least in part due to a variability of T cell sensibility to these drugs. Residual T-lymphocyte (TL) function measurements may be useful in individualizing Tac and MMF dosage to improve efficacy and tolerability.

**Methods:** 22 pediatric kidney (18), liver (1) and heart (3) transplant recipients treated with Tac, MMF (22/22) and prednisone (18/22) were studied. TL function was measured following stimulation with concavalin A (ConA) or monoclonal anti-CD3 antibody (OKT3). CD4 and CD8 TL activation was quantified by CD25 expression and TL proliferation was measured after propidium iodide incorporation.

**Results:** Percentage (%) of CD25 expression in CD4 TL showed a bimodal distribution, with 2 (low and high) TL activation levels and thus 2 groups of patients (low/high responder). Time variability in TL activation levels was observed in 3/7 patients.

Significant differences in % of CD25 expression in CD8 TL as well as CD4 and CD8 TL proliferation were also seen between the 2 groups. Except for age which was lower in the low responder group (mean age of 12.13 (± 3.6) years vs. 16.35 years (±2.29)), clinical parameters did not differ between the two groups.

**Conclusions:** This study confirms and quantifies a wide inter and intra-individual variability in TL response to tac and MMF in children. Correlations between TL function values and clinical, pharmacokinetic and pharmacogenetic parameters need to be performed.

## Abstract# 483

**Clinical Course of Pediatric Focal Segmental Glomerulosclerosis after Renal Transplant; Korean Experience** S.E. Lee,[1] K.H. Han,[1] Y.H. Jung,[1] H.K. Lee,[1] H.G. Kang,[1] I.S. Ha,[1] Y. Choi,[2] H.I. Cheong.[1] *[1]Department of Pediatrics, Seoul National University College of Medicine, Seoul, Korea; [2]Department of Pediatrics, Inje University Haeundae Paik Hospital, Busan, Korea.*

**Objectives:** Focal segmental glomerulosclerosis(FSGS) is common but often recurs after renal transplant, so we designed this study to investigate the risk factors of recurrence in childhood FSGS and its optimal management.

**Methods:** We reviewed the medical records of 50 renal transplants between Jan.1990 and Jan.2010 at out center in 46 children with primary FSGS.

**Results:** The median age of onset was 5.8 years and the presentation was nephrotic syndrome(NS) (n=37) or asymptomatic urinary abnormality. They received kidney transplants from living donors (n=33) or cadaver donors (n=17). In 18 allografts (38%), excepting 2 patients recurred after 3 years, FSGS recurred at a median of post-transplant 2 days. When compared the grafts with recurrence with those without recurrence, the recurrence group had more NS as the initial presentation ($P$=0.09). Recurrence did not significantly affect the graft survival. The statistically significant risk factors of recurrence were not found in our study, although recurrence group had more NS as initial presentation. Plasmapheresis, methylprednisolone pulse therapy and cyclophosphamide were tried, and the remission was achieved in 14 of 18 patients.

**Conclusions:** In Korean children with FSGS, recurrence occurred in 38% and the graft survival was poorer than total population of kidney transplant. Initial presentation of FSGS with NS was the important risk factor of recurrence, but the donor type or the duration of dialysis was not significant in contrast to the previous studies.

## Abstract# 484

**Identity Development and Quality of Life (QOL) in Youth with Renal Transplants (RTx) or Type 1 Diabetes (DM)** T. Lugasi,[1] M. Achille,[1] M.-J. Clermont,[2] V. Phan,[2] T. Blydt-Hansen,[3] S. McMurrich,[3] L. Geoffroy,[2] L. Legault,[4] L. Bell.[4] *[1]Psychology, Université de Montréal, Montreal, QC, Canada; [2]Nephrology and Endocrinology, CHU Sainte-Justine, Montreal, QC, Canada; [3]Nephrology, Manitoba Institute of Child Health, Winnipeg, MB, Canada; [4]Nephrology and Endocrinology, Montreal Children's Hospital, Montreal, QC, Canada.*

**Objectives:** Identity development is a central task of adolescence: identity achievement, in contrast to identity diffusion, can help individuals navigate the challenges of adulthood. As part of a prospective longitudinal study on transition to adult care, differences in identity and quality of life (QOL) between patients with RTx or DM and healthy controls were investigated.

**Methods:** Patients with RTx ($n$=26, *Mean*=18.95, *SD*=3,8) or *DM* ($n$=49, *M*=17,66, *SD*=2,58) were recruited from 3 Canadian pediatric hospitals and compared to 89 controls (*M*= 17,64, *SD*= 3,46). Participants completed the QOL Profile: Adolescent Version and the Objective Measure of Ego Identity Status II.

**Results:** Analyses revealed a main effect of group yielding F ratios of $F(2, 156)$= 4,457, $p$= .013 for interpersonal diffusion and $F(5,563, 2)$= 5,563, $p$= .005 for interpersonal foreclosure, with significantly higher means for RTx patients (*M*= 27,65, *SD*= 7,64 and *M*= 24.20, *SD*= 10,71). No significant differences in QOL were found between the 3 groups. Higher identity achievement scores were associated with higher QOL ($r$= .198, $p$= .017).

**Conclusions:** Youth with RTx showed less exploration and commitment in interpersonal aspects of their lives (e.g. dating, friendship) compared to their healthy or DM peers, but no differences in ideological measures (e.g. occupation, religion).

## Abstract# 485

**The Incidence, Outcome and Risk Factors for Recurrent FSGS Following Renal Transplantation** A. Lunn, V. Langlois, D. Hebert. *Nephrology, Hospital for Sick Children, Toronto, ON, Canada.*

**Objectives:** Recurrent FSGS is reported in 30% of renal transplant recipients. The aim of our study was to define the incidence of recurrent FSGS in our institution and describe the treatment, outcomes and risk factors for recurrence.

**Methods:** We retrospectively reviewed our clinical database to identify all patients with FSGS attending our clinic from May 2000 to Oct 2009. Data on ethnicity, genetic testing, recurrence, treatment and outcomes were collected by chart review.

**Results:** Of 260 patients followed 18(6.9%) had FSGS(11 male, median age at transplant 11.2 yrs, range 4 to 16) and received 21 transplants. One transplant was excluded(primary non-function). Disease recurrence occurred in 13(65%) of 20 transplants included in the study.

All patients with recurrence received plasmapheresis(TPE) (median 44 sessions - range 8 to 153). In 3(23%) of 13 grafts treatment achieved a normal serum albumin and non-nephrotic proteinuria.

At most recent follow-up 7(35%) grafts failed secondary to FSGS, 3(15%) grafts were functioning but with proteinuria(1 patient was receiving TPE) and 10(50%) grafts were functioning without FSGS.

Risk factors for recurrence were identified as time from diagnosis to transplantation less than 3 years(100%) and white race(69%).

4 patients were tested for ACTN4, TRPC6, CD2AP, and NPHS2 mutations. Two patients (1 with a NPHS2 mutation) did not have recurrence. Two patients with no disease causing mutations had recurrence.

**Conclusions:** The incidence of recurrent FSGS is high in our population, response to treatment is poor and graft loss is frequent. Risk factors for recurrence were white race, time from diagnosis to transplantation less than 3 yrs and the absence of genetic mutations.

**DISCLOSURE:** *Langlois, V.*: Grant/Research Support, Astella Pharma Canada.

## Abstract# 486

**Outcome of Kidney Transplantation in Adolescents and Children below Twelve – A 10-Year Review in Hong Kong** A.L.T. Ma, A.W.F. Hui, P.-c. Tong, W.-m. Lai, N.K.C. Tse, M.-c. Chiu. *Paediatrics & Adolescent Medicine, Paediatric Nephrology Centre, Princess Margaret Hospital, HK, Hong Kong.*

**Objectives:** Adolescent kidney transplant recipients are noted to have poorer graft survival as compared to younger children, which could be related to drug non-adherence. We compared the graft survival and function between adolescents and children in our centre.

**Methods:** Retrospective review was performed on data of patients who underwent kidney transplant between Jan 1999 to Jan 2010. Demographics, clinical data, estimated glomerular filtration rate (eGFR) and graft survival rates were compared between adolescents (12-22 years) and children ( <12 years).
**Results:** A total of 46 patients underwent kidney transplant in the study, with a mean age of 14.7 years. 36 patients (78.3%) were adolescents. Congenital kidney disease was the commonest primary diagnosis. No statistical significant difference was noted between the overall 1, 3 and 5- year survival rates between adolescents( 91.7%, 85.3% and 81%) and children (100% at all times). The baseline eGFR and serial eGFR were comparable between them. Although all patients with graft loss were adolescents, their outcomes were explained by specific reasons. 2 patients suffered from recurrence of focal segmental glomerulosclerosis, while other patients suffered from renal artery stenosis, renal vein thrombosis and chronic rejection respectively.
**Conclusions:** Reduced graft survival rate was not observed in adolescent recipients in Hong Kong. This may be related to the different parenting style in Chinese, where parents are more engaged in supervision of patients on drug compliance. Counselling and monitoring by case nurses also help preventing non-adherence.

## Abstract# 487

**Factors Affecting Long-Term Graft Function in Children with Kidney Transplant in a Paediatric Renal Centre in Hong Kong** A.L.T. Ma, A.W.F. Hui, P.-c. Tong, W.-m. Lai, N.K.C. Tse, M.-c. Chiu. *Paediatric and adolescent Medcicine, Paediatric Nephrology Centre, Princess Margaret Hospital, Hong Kong, Hong Kong.*
**Objectives:** Kidney transplant has been the treatment of choice for end stage kidney disease in children. We aim to identify factors which may affect graft function with estimated glomerular filtration rate (eGFR).
**Methods:** Retrospective review was carried out on data of patients who underwent kidney transplant in our centre between Jan 1999 to Jan 2010. Demographics, pre and post transplant clinical data were collected for analysis. GFR was estimated by the Schwartz's formula and documented every 2 years post transplant.
**Results:** Among the 46 patients who underwent kidney transplan in the past 10 years, 5 patients (13.9%) developed graft failure. Of all the factors analyzed, pre-emptive transplant (p=0.027) was associated with better graft survival. Acute tubular necrosis (ATN) was noted to be a significant independent factor affecting the baseline eGFR (p=0.033). Multivariate repeated measure analysis showed that low baseline eGFR, the presence of ATN, significant infections and focal segmental glomeruloscelorosis (FSGS) were associated with lower subsequent eGFR (p= 0.001, 0.043, 0.006 and 0.049). Patients with FSGS showed more significant deterioration of eGFR over time (p=0.004).
**Conclusions:** Despite the retrospective nature of the study and a small sample size, ATN and significant infection were identified to be significant factors affecting long term graft function. Patients with FSGS should be monitored closely and aggressive treatment should be instituted for recurrence.

## Abstract# 488

**Delayed Graft Function Is Reduced with ATG Induction in Cadaveric Paediatric Kidney Transplantation** A.D. Madrid, C. Herrero, E. Lara, R. Vilalta, S. Chocron, N.L. Jose. *Pediatric Nephrology Department, Hospital Vall d'Hebron, Barcelona, Spain.*
**Objectives:** This is a report of graft outcomes with ATG induction compared with interleukin-2 receptor antagonists.
**Methods:** 52 paediatric kidney cadaveric transplanted patients
**Group A**, 26 patients received antithymocyte globulin ATG ( 3 mg/kg/dose) at days 0 and 1,3,5, (dose 12mg/kg).
**Group B**: 26 patients received Basiliximab at days 0 and 4.
Maintenance therapy tacrolimus, target 6-10 ng/ml, mycophenolate mofetil (MMF), target 2-4 ng/ml, and tappered steroids until 6-9 months. Delayed graft function (DGF), acute rejection, and renal function the 1, 3, 6 and 12 months were examined.
**Results:** 24 over 26 patients of groupA presented immediate diuresis and 8 over 26 in Gorup B. CMV or EB disese were not registered in both groups. BK polyomavirus were positive in 3 patients in Group A and in 2 patient in Group B and were controlled reducing immunosuppression.
One year patient survival was 100% in both groups. One-year graft survival was 100% (group A) and 95 % (group B). Two patients in group B presented acute humoral rejection and 1 patient loosed the graft. At the 1,3, 6 and 12 months of follow-up no differences in serum creatinine or proteinuria were appreciated between both groups.
**Conclusions:** ATG induction reduces delayed graft failure and acute rejection rate.
DGF could be related to innate immunity activity, enhanced in brain-death donors and diminished under ATG treatment.
ATG treatment could also induce T regulatory cells and modulate the immune response related to the ischemic-reperfusion phenomena due to brain-death kidney donor procurement.

## Abstract# 489

**Growth with mTOR and CNI in the Maintenance of Kidney Transplanted Children** R. Vilalta, Á. Madrid, C. Herrero, E. Lara, S. Chocron, J. Nieto. *Hospital Vall d'Hebron, Barcelona, Spain.*
**Objectives:** It seems rational to inhibit first and third signals in the interaction of antigen presenting cells and T lymphocites, using CNI and mTOR in the maintenance of children with a kidney transplant. Some concern related to toxicity and interference with growth has been raised.
**Methods:** 29 children transplanted with a cadaveric donor between 2003 and 2009 received induction with basiliximab (2003-2007) ATG (2007-2009), and maintenance in the first year with tacrolimus, sodium mycophenolate and tapered steroids. When steroids were withdrawn, mTOR (18 patients everolimus, 11 patients sirolimus), were introduced in a coexistence regime with CNI to obtain mTOR and CNI concentration of 4 and 5 ng/ml around respectively. Co-treatment with sodium mycophenolate was maintained (2-4 ngml). Growth and renal function were evaluated.
**Results:** Patient and graft survival was 100 and 92 % respectively.
Mean creatinine level was 1.4 mg/100 ml (range 0.4-2.1).
Mean proteinuria was 20 mg/m2/h ( range 5-30 ) and did not increase after mTOR introduction (12+/-8 to 14+/- 10 mg/m$^2$/h).
Mean height before transplant was between -2 and -1 SD. Height growth velocity was same after mTOR introduction in 70 % of patients (-2, -1 SD) or even improved in 30 % of patients (-1 SD, M), possibly due to the achievement of a normal renal function.
**Conclusions:** Maintenance immunosuppressive regime with mTOR and CNI to obtain relative low blood levels (4-5 ng/ml) added to sodium mycophenolate appears to be safe combination in children with a kidney transplant.
When mTOR are introduced with urine proteins in the normal range, proteinuria do not appear at all.
Height velocity is preserved under this treatment.

## Abstract# 490

**Paediatric Renal Transplantation: 20-Year Experience in Singapore** S. Mahmud,[1] K.H. Ng,[1] Y. Lau,[1] W.S. Yeo,[1] K. Prabhakaran,[2] H.K. Yap.[1] *[1]Shaw-NKF Children's Kidney Centre, University Children's Medical Institute, National University Hospital, Singapore, Singapore; [2]Department of Paediatric Surgery, National University Hospital, Singapore, Singapore.*
**Objectives:** Paediatric renal transplantations were initiated in Singapore in 1989. We aimed to examine outcomes over 20 years from 1989 to 2009.
**Methods:** The renal registry database at the Shaw-NKF-NUH Children's Kidney Centre was examined retrospectively. Crude patient and graft survival rates were calculated using Kaplan-Meier survival analysis and log-rank test used to determine survival differences.
**Results:** Forty-four renal transplants were performed at our centre. Another four local patients with overseas transplants were included. The proportion of living donor (LD) transplants was 64.6%. LD transplant recipients were younger (12.7yrs vs 15.8yrs) and had a shorter waiting time (1.8yrs vs 5.8yrs) than deceased donor (DD) recipients. Overall patient survival rates were 95.7%, 92.7%, 85.6%, and 74.9% at one, five, ten, and fifteen years respectively. There were four deaths, of which three were infection-related. Graft survival rates at one, five, ten, and fifteen years for LD and DD transplants were 100%, 89.7%, 75.9%, 75.9% and 87.8%, 70.3%, 56.2%, 37.5% respectively, and were significantly higher in LD transplants. Multivariate analysis showed male gender, acute tubular necrosis and late acute rejections as predictors of graft failure.
**Conclusions:** Graft survival rates for LD transplants in Singapore were comparable to North American rates, although DD transplant rates were slightly worse, probably a reflection of the prevailing transplant policies.

## Abstract# 491

**Successful Paediatric ABO Incompatible Renal Transplantation (ABOiRT) with Quadruple Immunosuppression and B Lymphocyte Depletion** S.D. Marks, N.J. Sebire, S. Bradley, E. Wright, N. Mamode. *Department of Paediatric Nephrology, Great Ormond Street Hospital for Children NHS Trust, London, England, United Kingdom.*
**Objectives:** ABOiRT has been reported as a successful form of renal replacement therapy in children and adults with antibody removal.
**Methods:** To report the successful outcome of ABOiRT using quadruple immunosuppression (basiliximab, mycophenolate mofetil, tacrolimus and corticosteroids) with only B lymphocyte depletion (intravenous rituximab 375mg/m$^2$ at one month pre-transplantation). A 14-year old young man received his

second living related renal transplantation (first from father, second from mother) for end-stage renal failure (ESRF) on haemodialysis for steroid resistant nephrotic syndrome due to focal and segmental glomerulosclerosis (FSGS).
**Results:** The recipient and donor (mother) blood groups were A and B respectively with anti-B titres of 1 in 8 pre-transplantation. His renal transplant was performed successfully without further antibody removal or surgical complications. Six months post-transplant his renal function has stabilised with eGFR of 62mls/min/1.73m$^2$. He does not have clinical evidence of recurrent FSGS post-transplantation with negative anti-B titres and donor specific antibodies, low but detectable CD19 count and no evidence of viraemia (having required oral valganciclovir as CMV mismatch). He had a normal transplant renal biopsy at ten days post-transplant without evidence of acute rejection and one focal segmental sclerotic lesion seen on protocol biopsy at three months.
**Conclusions:** In view of the reported success of ABOiRT, clinicians must consider this option in ESRF patients whose parents have previously been excluded as living donors.

## Abstract# 492

**Transfusion-Dependent Pure Red Cell Aplasia (PRCA) Secondary to Anti-Erythropoietin Antibodies Successfully Treated with Renal Transplantation** A. Demetriou, S.E. Ledermann, N.J. Sebire, N. Casadevall, S.D. Marks. *Department of Paediatric Nephrology, Great Ormond Street Hospital for Children NHS Trust, London, England, United Kingdom.*
**Objectives:** PRCA is an isolated arrest of erythropoiesis which has been reported to occur in patients with chronic kidney disease (CKD) secondary to the development of antibodies against erythropoiesis-stimulating agents, usually erythropoietin alpha.
**Methods:** To report the successful outcome of pre-emptive living related renal transplantation in a 6.5 year old boy with CKD due to renal dysplasia who developed anti-erythropoietin antibodies (anti-epo Abs) resulting in transfusion-dependent PRCA.
**Results:** He was on subcutaneous erythropoietin beta for two years but two months prior to transplantation, his haemoglobin was 4.6g/dl with reticulocytes of 0.1% requiring his first blood transfusion. His bone marrow aspirate and trephine biopsy one month before renal transplantation confirmed PRCA (other causes excluded). His erythropoietin beta was discontinued and he had high titres of anti-epo Abs. He received his second blood transfusion before his renal transplant. He underwent a living related renal transplantation from his maternal uncle (mismatch 011) using triple immunosuppression. His anti-epo Abs reduced further two weeks post-transplantation but required his third blood transfusion. He is six months post-transplant with haemoglobin of 11.6g/dl, ferritin of 363mcg/l, negative anti-epo Abs and good renal allograft function (eGFR of 74mls/min/1.73m$^2$).
**Conclusions:** This is the first reported paediatric case where renal transplantation has successfully treated transfusion-dependent PRCA secondary to neutralising anti-epo Abs against erythropoietin beta.

## Abstract# 493

**Maintaining an Adequate Intra-Operative and Immediate Post-Operative Systolic Blood Pressure during Renal Transplantation in Children under Five Influences Outcome – A Single Centre Experience** H.J. McCarthy, J. Dudley. *Nephrology, Bristol Royal Children's Hospital, Bristol, United Kingdom.*
**Objectives:** Young children are at increased risk of graft loss due to thrombosis. A likely major contributing factor relates to peri-operative perfusion of the donor kidney. At our unit every effort is made to maintain peri-operative systolic blood pressure above 100mg Hg for all patients regardless of age and size of patients.
**Methods:** An audit of peri-operative BP monitoring and fluid management in children transplanted between 1/1/06 and 31/12/08 was undertaken.
**Results:** 32 children received a transplant, 11 of whom were under 5 years of age. Graft survival at 1year was 94% in patients under five and 95% in patients over five. Patient survival at 1year was 100% in both groups. One graft was lost in each cohort due to early thrombosis. Patients under the age of 5 were more likely to require blood products and inotropes to maintain systolic BP above 100mg Hg compared with those older than 5 years.

Comparison by age of the support required to maintain a systolic blood pressure >100mmHg peri-operatively in renal transplantation

| | No. | Age (mean in years) | Cyrstalloid (%) | Colloid (%) | Blood (%) | Inotropes (%) | Admission to PICU (%) |
|---|---|---|---|---|---|---|---|
| <5 years | 11 | 2.7 | 100 | 67 | 56 | 100 | 94 |
| >5 years | 21 | 12.3 | 100 | 22 | 11 | 11 | 5 |

**Conclusions:** Our data suggest that optimisation of systolic BP in younger children requires close attention to the need for blood products and inotropes compared with older children.

## Abstract# 494

**Timing for Removal of Peritoneal Dialysis Catheters in Post-Transplant Patients** E. Melek,[1] E. Baskin,[1] U. Bayrakçi,[1] K. Gülleroglu,[1] S. Sevmis,[2] H. Karakayali,[2] M. Haberal.[2] *[1]Pediatric Nephrology, Baskent University, Ankara, Turkey; [2]General Surgery, Baskent University, Ankara, Turkey.*
**Objectives:** The peritoneal dialysis (PD) catheter is a potential source of infection for immunosuppressed renal transplant patients. Although catheter related complications can be prevented by removel of catheter, the catheter removal time (CRT) is a contraversial issue. Our objective was to evaluate the complications related to peritoneal dialysis (PD) catheter at the post-transplant period and to determine the optimum time for removal of PD catheter.
**Methods:** We retrospectively analysed 33 renal transplants patients received PD before transplantation. Mean ages at transplantation and mean CRT of patients were 12.77±4.02 (range:3.5-18.0) years and 81.12±36.19 (22-152) days respectively. The catheters are electively removed approximately at 3rd month of post-transplant period in our center.
**Results:** Peritonitis (one patient) and exit-site infection (one patient) developed in only 2 patients (6%) at 2nd month and at 3rd month of post-transplant period respectively. PD catheter was used for 6 of 33 (12.1%) patients (2 patients for acute tubular necrosis, 1 patient for acute graft lost, 3 patients for post-transplant ascitis) at first 15 days of post-transplant period.
**Conclusions:** The risk of catheter related complications in our study is low and occur after 1st month of post-transplantation period. The need for catheter in our study was in the first 2 weeks of post-transplantation period. Therefore, PD catheter can be removed after the first 2 weeks of post-transplantation period and should not be in place more than 2 months.

## Abstract# 495

**Familial Mediterranean Fever in the Differential Diagnosis of Recurrent Fever in Renal Transplant Patients** E. Melek,[1] U. Bayrakçi,[1] E. Baskin,[1] K. Gülleroglu,[1] S. Sevmis,[2] H. Karakayali,[2] M. Haberal.[2] *[1]Pediatric Nephrology, Baskent University, Ankara, Turkey; [2]General Surgery, Baskent University, Ankara, Turkey.*
**Objectives:** Familial Mediterranean Fever (FMF) is an autosomal recessive disorder, which is common in Mediterranean countries. One of the major complications of FMF is systemic amyloidosis which can result in end-stage renal disease. We retrospectively reviewed our renal transplant patients to evaluate clinical course and prognosis of patients with diagnosis FMF after renal transplantation.
**Methods:** We retrospectively analyzed 72 renal transplant patients. Nine of them was found to have the diagnosis of FMF. Amyloidosis was the cause of chronic renal failure in 3 of them. 2 patients had the diagnosis of FMF after renal transplantation.
**Results:** The mean follow-up time of renal transplanted patients with FMF was 23.0±18.1 months. There were no statistically significant difference between patients with and without FMF regarding graft survival, immunosuppressive treatment and creatinine levels. Amyloidosis in renal allograft was observed in only 1 patient which was thought to be due to drug withdrowel.
**Conclusions:** All FMF patients with or without amyloidosis should receive colchicine to prevent development or recurrence of amyloidosis. FMF patients with or without amyloidosis may undergo kidney transplantation safely expecting outcomes similar to those patients whitout FMF. Also FMF should be thought in the differential diagnosis of recurrent fever in post-transplant period especially in medeterranean countries.

## Abstract# 496

**Outcomes of 2-Drug Maintenance Immunosuppression for Pediatric Renal Transplantation: 10 Year Follow-Up in a Single Center** M. Michael, A.S. Kale, E.D. Brewer. *Department Pediatrics, Renal Section, Baylor College of Medicine/Texas Children's Hospital, Houston, TX, United States.*
**Objectives:** Little data is available for long-term outcome for 2-drug maintenance therapy (2DT) for pediatric renal transplantation (RTx). Less immunosuppression (IS) may lead to less complications, but worse graft survival (GS). We reviewed 10y long-term outcomes of 2DT at a large urban pediatric center.
**Methods:** Retrospective review of all pts receiving RTx Aug 1988-Jul 2008 included 139 pts treated with prednisone (P) & cyclosporine (CyA) without induction (group 1, 1988-98) or with daclizumab induction+P+CyA (group 2, 1999-2008). Primary outcome was GS at 1y,3y,5y&10y as determined by Kaplan-Meier (KM) survival curves. Differences in GS were compared using log rank test. Secondary outcomes were delayed graft function (DGF+), acute rejection <6m (AR) & malignancy (MX).
**Results:** 139 pts (84M/55F; 36%Cauc/34%Hisp/25%Black/5%other) had RTx mean age 11.8 ±6.2y, & ESRD cause 38% congenital anomalies (obstr/dysplasia), 18% FSGS, 8% reflux nephropathy. 67% had deceased donor (DD); 33% living

donor (LD); 22% preemptive. Demographics did not differ for groups 1 (86 pts) & 2 (53 pts). GS rates for all pts at 1y, 3y, 5y were 97.1, 85.4, & 76.2%, respectively. GS was significantly better in LD vs DD (p=0.009).

Table: Outcomes by IS group

| | %GS1y | %GS3y | %GS5y | %GS10y | %DGF+ | %AR | %MX |
|---|---|---|---|---|---|---|---|
| Group1: 86pt | 90 | 83 | 67 | 45 | 19 | 27* | 2.3 |
| Group2: 53pt | 96 | 84 | 76 | 58 | 26 | 22 | 1.9 |

%GS=KM survival, p=NS by log rank; other variables groups 1&2 compared by Chi-2, *p=0.001

**Conclusions:** In this study, RTx 1y, 3y & 5y GS and AR rates with 2DT were comparable to those of NAPRTCS for pts maintained mostly with 3DT, and 10y MX rate was very low with 2DT.

## Abstract# 497

**Development of De Novo Donor Specific Antibodies after Post-Transplant UTI** B. Morgenstern, M. Joseph, S. Hsieh, K. Papez, I. Dvorchik. *Division of Nephrology, Phoenix Children's Hospital, Phoenix, AZ, United States.*

**Objectives:** The development of donor-specific antibodies (DSA) is associated with humoral rejection, interstitial fibrosis/tubular atrophy, and ultimate graft loss in kidney transplantation. Urinary tract infection (UTI) can be associated with the up-regulation of HLA antigens on renal tubular epithelial cells. We sought to determine if there is an association between post-transplant UTI and the development of DSA.
**Methods:** We reviewed the kidney transplant experience at Phoenix Children's Hospital since 2002. DSA have been drawn by protocol at 6 months post-transplant since 2008, and are always obtained when there is evidence of antibody-mediated rejection diagnosed on for-cause allograft biopsies. Kaplan-Meier survival and Fisher exact tests were performed. Urine cultures for bacteria were performed when there were localizing symptoms or in febrile patients.
**Results:** Sixty-nine kidney transplants were analyzed. Predominantly, immunosupression has been with rabbit anti-thymocyte globulin induction and tacrolimus, mycophenolate and prednisone as maintenance therapy. Overall 1, 3 and 5 year graft survival rates are 98%, 93% and 88% respectively. 13 children (18%) have had documented post transplant UTI. 11 children (16%) have developed DSA. Of these 11, 8 had a UTI post transplant. There was an association between UTI and DSA (p<0.01). The odds ratio of developing DSA after UTI was 5.96 (1.62 - 21.89).
**Conclusions:** Pediatric renal transplant recipients are more likely than adult recipients to have congenital urological abnormalities. This can predispose them to UTI. UTI is clearly a risk factor for the development of DSA, which puts graft survival at risk.

## Abstract# 498

**Nineteen Years Experience with Antithymocyte Globulin Induction in Kidney Pediatric Transplantation** C. Mota,[1] L. Martins,[2] T. Costa,[1] M.d.S. Faria,[1] A. Castro Henriques.[1] *[1]Department of Pediatric Nephrology, Centro Hospitalar do Porto, Porto, Portugal; [2]Department of Nephrology/ Transplantation, Centro Hospitalar do Porto, Porto, Portugal.*

**Objectives:** The optimal immunosuppressive therapy in kidney transplantation remains controversial. Since 1990, we included, in our department, antithymocyte globulin Fresenius® (ATG-F) in sequential immunossupressive therapy in pediatric kidney transplantation.We analysed retrospectively the long-term outcomes.
**Methods:** Ninety eight kidney transplants were performed in 91 children and adolescents using deceased donor source grafts, between November 1990 and October 2009. In 86.8 % recipients ATG-F was used as antibody induction, and in 12.2 % recipients no ATG-F induction was used.
**Results:** Overall graft survival rates at 1, 5, 10 and 15 years were 91.8%, 86.1%, 75.9% and 61.9% respectively. In the group ATG-F the graft survival at 1, 5, 10 and 15 years was 93%, 89.1%, 79%, 62.4% and in group without ATG-F it was 83.3%, 66.7%, 55,6% at 1, 5, 10 years respectively(p=0.27). A papillary thyroid carcinoma was diagnosed and no lymphoid malignancies were observed during the observational period. All the patients are alive at the end of follow-up, except one who died of cardiovascular disease, 7 months after graft loss.
**Conclusions:** These results indicated that ATG-F induction in pediatric kidney transplantation was associated with good graft and patient survival rates using deceased donor kidneys, with low levels of complications.

## Abstract# 499

**Clinical and Histologic Efficacy of Interleukin-2-Receptor Antagonists in Pediatric Kidney Transplantation** G. Ghirardo, M. Della Vella, B. Andreetta, E. Vidal, E. Benetti, L. Murer. *Pediatric Nephrology, Dialysis and Transplant Unit, Department of Pediatrics, University of Padua, Padua, Italy.*

**Objectives:** Basiliximab is a non-depleting recombinant chimeric murine/human monoclonal anti-interleukin-2-receptor antibody used for prevention of acute organ rejection in renal transplant recipients. We analysed 2 groups of pediatric kidney recipients (R): one including patients treated with Basiliximab and corticosteroids, the other recipients receiving exclusively corticosteroid pulse induction. Immunosuppression in both groups was based on CsA or FK506 ± MMF ± corticosteroids.
**Methods:** We retrospectively analysed 130 paediatric kidney recipients: 78 (60%) treated with Basiliximab (R/D age 12±7/15±8 yrs, HLA mismatches 3.6±0.9) and 52 (40%) with only corticosteroid pulses (R/D age 15±5/13±10 yrs, HLA mismatches 3.1±1.06).
**Results:** Patients who received IL-2 receptor-antagonist induction had lower incidence of acute clinical rejections (p=0.0049). Steroid withdrawal at 12 and 24 months post-transplant was possible in a larger number of patients receiving Basiliximab then corticosteroid pulses (p=0.0015 and p=0.0001). No differences in terms of organ survival, creatinine clearance, acute/chronic subclinical lesions at protocol biopsies, mean corticosteroids dosage, CsA/FK506 blood through levels, and growth at 6, 12 and 24 months post-transplant.
**Conclusions:** Our results highlight that the use of IL-2-receptor antagonists allows an early corticosteroid withdrawal and confirm an higher efficacy of Basiliximab in preventing clinical acute rejections. We found no differences regarding the prevalence of post-transplant short or long term acute and chronic histological lesions.

## Abstract# 500

**Outcome of Focal Segmental Glomerulosclerosis after Renal Transplantation with/without Podocine Gene Mutations in Turkish Children** F. Mutlubas, B. Sozeri, A. Berdeli, C. Hoscoskun, S. Mir. *Pediatric Nephrology, Ege University Faculty of Medicine, Izmir, Turkey.*

**Objectives:** Focal segmental glomerulosclerosis (FSGS) is the most frequent acquired disease resulting in ESRD in children. The risk of recurrence in the allograft is reported to be ~30%. Podocine gene mutations are well-known to be a protective factor for FSGS recurrence. The aim of the study was to compare the clinical features of FSGS in renal recipients with or without podocine gene mutations.
**Methods:** We sequenced all eight exons of NPHS2 in 18 Turkish pediatric renal recipients. All patients had biopsy-proven primary steroid-resistant FSGS. 3 of 18 children received living related donor. All patients were evaluated with clinical, laboratory and demographic features.
**Results:** 11 of 18 patients had NPHS2 gene mutations while 7 had no mutation. The mean age at onset was significantly younger in mutation positive group. The mean age at renal transplantation was 10.53±3,64 yr. Biopsy proven FSGS recurred only in 2 patients with homozygote mutation in the early and late period. Late FSGS recurrence resulted with graft failure. The patients without podocine gene mutation had no recurrence for 2,3–6,6 yr. The age onset of FSGS was the only difference between groups.
**Conclusions: In Conclusion**, we detected FSGS recurrence in 2 children who had causative homozygote NPHS2 gene mutations.Podocine gene mutation positive group had early onset FSGS. Other clinical features are similar for patients with or without NPHS2 mutations. Contraversy to literature patients carrying homozygote mutation may display early or late onset FSGS recurrence. FSGS patients must be followed-up with urine protein in transplant clinics.

## Abstract# 501

**Systematic Anticoagulation Following Renal Transplantation: Benefits and Risks** P. Niaudet, E. Balzamo, G. Guest, M. Charbit, O. Boyer, R. Salomon. *Pediatric Nephrology, Hôpital Necker-Enfants Malades, Paris, France.*

**Objectives:** Vascular thrombosis is responsible for at least 10% of early graft failures. Factors that increase the risk include a thrombophylic state, a recipient less than 5 years or less than 15 kgBW, a donor aged less than 5 years, several arteries, history of prior transplantation or vascular rejection. The aim of this study was to retrospectively analyze the benefits and risks of systematic anticoagulation in renal transplant recipients.
**Methods:** From March 2001 to January 2007, 116 renal transplants were performed in 112 patients aged 1.2 to 21.5 years (mean 10.3). Two patients with early graft loss were excluded as well as 4 pts who did not receive anticoagulants. Exaparine was started between 12 and 24 hours after surgery and continued during 3 weeks in 110 children.
**Results:** One graft was lost from artery thrombosis in a 18 year old patient following a cadaveric transplantation (0.89%). The graft had two arteries. No other risk factor was present and antiX a activity was 0.41. Ten patients had bleeding complications (perirenal or intra renal hematoma, wound hematoma, digestive bleeding) necessitating blood transfusion (antiXa mean/ maximal : 0.47/0.75 ; 0.28/0.38 ; 0.26/0.4 ; <0.1/<0.1 ; 0.51/1.76 ; 0.62/0.87 ; 0.59/1 ; 0.4/0.72 ; ND ; 0.4/0.8). Two other patients (1.8%) had a polar hematoma without fall in hematocrit.
**Conclusions:** In conclusion, prophylactic anticoagulation decreases the risk of early graft thrombosis after renal transplantation. AntiXa activity shoud be between 0.3 and 0.5. Carefull monitoring of ultrasound and hematocrit allows to detect bleeding complications and stop anticoagulation.

## Abstract# 502

**Everolimus with Low-Dose Cyclosporine A after Pediatric Kidney Transplantation: No Acute Rejections, Less Infections** L. Pape, N. Kanzelmeyer, J. Drube, M. Kreuzer, T. Ahlenstiel. *Pediarric Nephrology, Medical School of Hannover, Hannover, Germany.*

**Objectives:** The number of acute rejections and infections after pediatric kidney transplantation could not be reduced in the last years. In order to improve these parameters, we investigated a new immunosuppressive protocol in a prospective, controlled trial.

**Methods:** Twenty children were treated with Basilximab, CsA (trough-level=C0 200-250 ng/ml) and Prednisolone at the time of Tx. After 2 weeks, CsA dose was reduced to 50% (C0 50-100 ng/ml, after 6 months: 50-75 ng/ml) and Everolimus (1.6 mg/m²/d) was started (C0 3-6 ng/ml). Six months after Tx, Prednisolone was stopped.

**Results:** All 20 protocol biopsies showed no acute rejection or borderline findings. had ATN was found in 4 of 8 indication biopsies, two had borderline findings, one 5% interstitial fibrosis, one was normal. Mean GFR 1 year after Tx was 71 ± 25 ml/min/1.73m². The number of serious adverse events, infections and gastrointestinal problems was significantly lower as compared to a historic age matched cohort that was treated with normal-dose CsA, MMF and steroids. Without CMV-prophylaxis, only 2 primary CMV infections were seen despite a donor-CMV+, recipient-CMV– constellation in 10/20 children.

**Conclusions:** In pediatric kidney transplantation primary immunosuppression with low dose CsA, Everolimus and steroid withdrawal after 6 months leads an absence of acute rejections and a reduction in infections, especially CMV.

## Abstract# 503

**Timing of Ureteric Stent Removal in Paediatric Renal Transplant Recipients (RTR)** P. Patel, J. Olsburgh, S.D. Marks. *Nephrology, Urology and Transplantation, Guy's and St Thomas' NHS Foundation Trust, London, United Kingdom; Nephrology, Urology and Transplantation, Guy's and St Thomas' NHS Foundation Trust, London, United Kingdom; Paediatric Nephrology, Great Ormond Street Hospital for Children NHS Trust, London, United Kingdom.*

**Objectives:** Transplant ureteric stents (TUS) reduce the incidence of urological complications post-operatively. However, there can be complications with TUS (UTI, migration, haematuria and pain) which may be related to how long the TUS remains in situ. We assessed two techniques for TUS removal in an aim to determine optimal timing for TUS removal.

**Methods:** Retrospective study of all RTR identified from our database in a single centre from 2002 to 2008. Medical records were reviewed and data collected on the method and timing of TUS removal; TUS related and ureteric complications.

**Results:** Of 180 RTR, 157 (64 [41%] female, 79 [50%] living donor transplants) were included. Cystoscopic TUS removal was performed in 151 patients at 8-90 (median 42) days with 30(20%) TUS complications: 16 UTI, 9 macroscopic haematuria, 4 migration, 1 blocked stent. These resulted in early TUS removal in 18 (64%) patients. Ureteric complications were found in 7(5 %) patients in this group (2 leak and 5 stenosis). Six patients had the TUS sutured to catheter, with stent removal on day 5 (4), day 10 (1) or day 21(1 stent attached to SPC). No patients in this group developed stent or ureteric complications.

**Conclusions:** 20% of RTR had TUS complications, often resulting in the need for early removal. Suturing the TUS to the catheter to facilitate early removal appeared to be safe and may reduce the incidence of stent complications.

## Abstract# 504

**BK Virus Associated Nephropathy in Pediatric Kidney Transplant Recipients** V.M. Pinto, P.C. Salas, J. Grandy, P. Zambrano, B.C. Corta. *University of Chile, School of Medicine,Pediatric Department, Dr. Exequiel González Cortés Children's Hospital, Santiago, Chile.*

**Objectives:** BK Virus associated nephropathy (BKVAN) causes allograft dysfunction and graft loss.Treatment of BKVAN is not well defined.We report our experience with kidney transplant recipients, who developed BKVAN, the diagnosis, study and treatment.

**Methods:** Retrospective case report.

**Results:** Two female, aged 8, 11 and 9 yr respectively developed BKVAN at 24, 12 and 3 months after kidney transplant.All received, polyclonal antibody therapy, MMF, tacrolimus and prednisone. Both female have insidious rise of creatinine, at 12 and 24 months.The male had an abrupt rise in creatinine at the third month. All treated initially with intravenous methylprednisolone.BK viral DNA measure from plasma by quantitative PCR assay and allograft biopsy consistent with polyoma virus infection.Imunosuppression reduction and IVIg was administered. The first patient also received ciprofloxacin 4 weeks, but had progressive deterioration of renal function and BK PCR failed to decrease. Second patient received 6 course of cidofovir (0.25-0.50 mg/kg/dose), every 2 wk, and plasma viral load decreased to undetectable levels. Serum creatinine remained on average close to 2 mg/dl. Tthird patient BK viremia cleared with the immunnosuppression reduction and IVIg administration, but a month after had a rise in plasma BK viral DNA, initiating cidofovir.The plasma BK PCR began to decrease.

**Conclusions:** BKVAN should be suspected in patients with graft dysfunction, especially those who receive aggressive induction. Cidofovir is effective in reducing BK viral load. Randomized, controlled trials regarding the most appropriate antiviral therapy are needed.

## Abstract# 505

**Efficacy of Sirolimus in Children with Chronic Allograft Nephropathy** T. Seeman,[1] L. Podracka.[2] *[1]Dept. Pediat., Medical Faculty, Charles Univ., Praha, Czech Republic; [2]Dept. Pediat., Medical Faculty, UPJS, Kosice, Slovakia (Slovak Republic).*

**Objectives:** Sirolimus (SRL) is a potent immunosuppressant acting on the mTOR receptor with no adverse effects on renal function.

**Methods:** We retrospectively evaluated the efficacy and safety of SRL in 8 pediatric renal transplant recipients (6 girls/ 2 boys),who were 10.90 ± 4.01 years at time of transplantation and received a CNI as the core immunosuppression.

**Results:** The patients were converted to SRL at 3.8 ± 2.5 years posttransplantation. The indication for SRL therapy was chronic allograft nephropathy, in 3 of them due to calcineurin toxicity (37.5 %). Sirolimus was initiated at 1 to 2 mg/m² given orally once daily. Mean follow-up after the switch to SRL was 1 year. Patients and graft survival was 100%. No children experienced acute rejection episodes. The mean serum creatinin (Cr) at the time of switch to sirolimus was 179.6±63 umol/l and decreased slowly during receiving SRL to 147.6 ±53.3 umol/l after 12 months of treatment (n.s.). We observed slight improvement in glomerular filtration rate assessed using the Schwartz formula at 1 month (42.4 ±12.6 v.s. 54.9 ±23.8 ml/ml/1.73m2), which was sustained thereafter. No serious adverse events were observed. Proteinuria increased significantly ($p<0.05$), however, no significant changes in serum cholesterol and trombocytes have been found. Although the prevalence of hypertensive patients decreased during follow-up, it was not significant.

**Conclusions:** Sirolimus as a concomitant immunosuppressant is effective in chronic allograft nephropathy. Further data are needed in pediatric transplantation recipients to best assess the role of TOR inhibition in corticosteroid and/or calcineurin inhibitor-sparing regimens.

## Abstract# 506

**Is the Cylex Immunoknow® Assay Clinically Useful or Cost Effective in Pediatric Kidney Transplantation?** W. Primack, Z. Smith, M.A. Amamoo, C. Wang, M. Brundette, N. Bitner, K. Gibson, T. Kozlowski, M. Ferris. *UNC Kidney Center, University of North Carolina, Chapel Hill, NC, United States.*

**Objectives:** The Cylex Immunoknow® Assay (IK) measures cell mediated immunity in transplant recipients by determining ATP generation in PHA stimulated CD4 cells. We report a single center study of the utility of IK to predict rejection or viral infection and to identify predictors of IK levels.

**Methods:** Retrospective analysis of 67 renal transplant patients (pt) ages 1-20 from 11/06 to 12/09.

**Results:** 494 IK were performed on 67 pt ( average 5±3.6 tests/pt). The mean IK level was 362±151 ng/ml comparable to the adult normal range of 226-525 ng/ml. In pt < 12 years IK was 350±149 and in pt >12 years was 369±152 ($p>0.5$). African American (AA) pt had lower IK compared to all other races ($p=0.004$). IK correlated with wbc ($r=.38$, $p<0.0001$), and with tacro dose ($r= -0.186$, $p=0.0002$), but did not correlate with MMF dose ($r= -0.12$, $P=0.8$), or CyA dose ($r= -0.009$, $p=.9$)

Careful chart review identified 14 rejection events (R) in 11 pts. 7 R had IK within 30 days prior to R. 1/7 had IK>nl, 1/7 had IK< nl. There were 12 EBV (IK 310±166), 8 BK(IK 328±115), 8 CMV (IK 298±123) infections in pts. Most were subclinical and diagnosed only by positive PCR.

The UNC Hospital Lab cost to perform IK is $298 and the charge is $364 per test. The total 494 IK tests cost $147,212.

**Conclusions:** The Cylex Immunoknow® test in our hands is not useful to predict rejection or viral infection. It correlates best with the total wbc. In contrast to other studies, age was not significant. The IK level was significantly lower in AA pt. This finding needs further exploration.

## Abstract# 507

**Kidney Transplantation in Children, 25 Years Experience in Transitional Country** Z. Puretic,[1] J. Slavicek,[1] I. Humar,[2] L. Bubic Filipi,[1] N. Basic-Jukic,[1] M. Subat-Dezulovic,[4] Z. Kastelan,[3] R. Zunec,[2] Z. Mustapic,[1] J. Pasini.[3] *[1]Department of Dialysis, University Hospital Centre Zagreb, Zagreb, Croatia; [2]Tissue Typing Center, University Hospital Centre Zagreb, Zagreb, Croatia; [3]Department of Urology, University Hospital Centre Zagreb, Zagreb, Croatia; [4]Department of Pediatric Nephrology, Universiity Hospital Center Rijeka, Rijeka, Croatia.*

**Objectives:** The single specialized Pediatric Dialysis Unit includes follow up after kidney transplantation (KTx).It operates within adult Center and children are followed up to 21 yrs,or longer.Croatia has total inhabitans of 4,4 and children <18ys 0,9 Mill.

**Methods:** Epidemiological,demographical and descriptive analysis of pts in whom KTx was performed1985-2009.Retrospectively reviewed data on 153 pts aged 1 mo-8 ys,treated with renal replacement therapy (RRT) caused by terminal renal failure (TRF).Inclusion:start dialysis or preemptive KTx <18 ys.

**Results:** During 25 ys in 98 children KTx was performed.Males 71%.Preemptive KTx underwent 9 pts.From living related donors were 29 %.With functioning graft died 2, and graft loss was in 4 during the 1st yr and in 24 from 2-12 ys after KTx. Croatia entered Eurotransplant (ET) as regular member in August 15th 2007. Until than 84 children were transplanted.Average waiting time (WT) on dialysis prior joining to ET was 4.5 yrs or on average 3-4 pts were transplanted/yr and after is 2,5 ys or 6 pts/yr.

**Conclusions:** In Croatia organized KTx program in TRF was established before 25 ys. Last 2,5 years Croatia is member of ET.Benefits of these are: more KTx/yr,and resolving long-waiting pts.Problems:longer cold ischemia time,and higher miss match.

## Abstract# 508

**Development of a Screening Assay for Actions of Traditional Chinese Medicine on Human Alloreactive T Cells** J. Reid-Adam,[1] P. Heeger,[3] L. Satlin,[1] X.-M. Li.[2] *[1]Pediatric Nephrology, Mount Sinai School of Medicine, New York, NY, United States; [2]Pediatric Allergy & Immunology, Mount Sinai School of Medicine, New York, NY, United States; [3]Adult Nephrology, Mount Sinai School of Medicine, New York, NY, United States.*

**Objectives:** We hypothesize that Traditional Chinese Medicines (TCMs), through their immunomodulatory and immunosuppressive properties, can serve as an effective primary or adjunct pharmacotherapy for prevention of transplant rejection. We developed a screening assay for measuring effects of TCMs on alloreactive T cell immunity in vitro.

**Methods:** Peripheral blood mononuclear cells (PBMCs) are cultured with allogeneic HLA-typed, B cell lines in ELISPOT plates coated with capture antibodies for IFN-g and IL-17. Readouts are IFN-g and IL-17 production as markers of primed T cells. These mixed lymphocyte reactions (MLRs) are performed in the absence or presence of various Chinese herbs. The resulting spots are counted on a computer-assisted Immunospot image analyzer.

**Results:** Of 30 herbs screened to date, 5 candidates have been identified as having decreased INF-g, IL-17 or both cytokines in a 24 hour MLR by ELISPOT. At concentrations of 0.5 mg/ml: Compounds 0182 and 0190 reduced IFN-g production by ≥ 80%; Compound 0038 reduced IL-17 production by 74%; Compounds 0126 and 0130 reduced IFN-g and IL-17 production by ≥ 50%.

**Conclusions:** Our work identifies a novel screening approach for testing immunomodulatory compounds relevant to transplantation. Identification of those compounds capable of inhibiting alloreactive T cells in vitro will become candidate immunosuppressants for further testing in animal models and ultimately human transplant recipients.

## Abstract# 509

**Chylous Ascites Complicating Pediatric Renal Transplant** S. Riar, S. Amaral, B. Warshaw. *Dept. of Pediatrics, Division of Nephrology, Emory Univ. School of Medicine, Atlanta, GA, United States.*

**Objectives:** Chylous ascites (CA), an unusual complication of pediatric abdominal surgery has scarce data on management. We describe approach & outcome in 2 children with CA post renal transplant.

**Methods:** We performed a retrospective chart review of 2 patients who developed CA after renal transplant to evaluate clinical course & management.

**Results:** A 7 years old boy underwent a transperitoneal deceased donor renal transplant (DDRT) with venous anastomosis to portal vein. CA was diagnosed 2 months later on paracentesis. Feeds were changed to Medium Chain Triglyceride (MCT) formula. Ascites re-accumulated so feeds were held & total parenteral nutrition (TPN) was provided with some benefit. CA recurred after introduction of low fat diet & paracentesis was done 4 times over 3 months. At 5 months post-DDRT a peritoneo-venous shunt (Denver) was placed. Shunt malfunction led to revision at 3 months & replacement after 4 months. To prevent malfunction, shunt was pumped frequently. A right sided chylothorax was managed symptomatically. At his last follow up at 16 months post-DDRT, CA was still present; otherwise he was doing well.

A 2 years old girl had a transperitoneal DDRT & developed abdominal distention 2 weeks later. Paracentesis showed CA. An external abdominal drain was placed, feeds were held & TPN was started. Gradual introduction of MCT formula allowed weaning of TPN. CA improved & the drain was removed on day 29 without further intervention.

**Conclusions:** CA should be considered in renal transplant patients with post-operative abdominal distension. Initial approach should be non-operative with TPN support & p.r.n. paracentesis. Placement of peritoneo-venous shunt can be complicated by malfunction.

## Abstract# 510

**Successful Renal Transplantation in a Child with Factor H Associated Atypical Hemolytic Uremic Syndrome (aHUS) with Prophylactic Eculizumab Treatment**[#] M. Riedl,[1] J. Hofer,[1] A. Rosales,[1] T. Giner,[1] T.C. Jungraithmayr,[1] W. Mark,[1] R. Wuerzner,[1] K.O. Kliche,[2] L.B. Zimmerhackl.[1] *[1]Pediatrics, Medical University, Innsbruck, Austria; [2]Alexion Pharma Germany GmbH, Munich, Germany.*

**Objectives:** Renal transplantation in atypical hemolytic uremic syndrome (aHUS) associated with factor H mutation is characterized by a high rate of disease recurrence and subsequent graft loss. Here we present a case of prophylactic use of the humanized monoclonal anti C5 antibody Eculizumab in renal transplantation.

**Methods:** Eculizumab monitoring was performed with the measurement of the terminal complement complex (TCC, C5b-9) in serum, plasma and after zymosan activation (ELISA).

**Results:** In November 2008 a ten year old boy with aHUS associated with a heterozygous factor H mutation received a deceased donor kidney. Immunosuppression included prednisone, mycophenolate mofetil and tacrolimus. Before and after transplantation daily plasma exchange was performed until Eculizumab was available on day 10 after renal transplantation. 600mg i.v. was administered every second or every third week until present. Eculizumab managed to normalize platelets and complement immediately. For the treatment period of 16 months an excellent renal function, no recurrence and no need for plasma therapy can be reported.

**Conclusions:** The prophylactic use of Eculizumab enabled us to perform a successful renal transplantation in this high risk patient. Eculizumab is a promising new drug in aHUS and a controlled trial for the prophylactic use of Eculizumab in renal transplantation in HUS is urgently needed.

[#] Supported by ÖNB grant & Alexion

**DISCLOSURE:** *Kliche, K.O.*: Speaker's Bureau, Alexion Germany.

## Abstract# 511

**Inosine 5'-Monophosphate Dehydrogenase (IMPDH) Activity in Children: Physiological Regulation and Response to Mycophenolic Acid (MPA) Therapy** A. Rother,[1] P. Glander,[3] E. Vitt,[2] N. von Ahsen,[4] V. Armstrong,[4] M. Oellerich,[4] K. Budde,[3] B. Tönshoff,[1] L.T. Weber.[2] *[1]University Children's Hospital, Heidelberg, Germany; [2]University Children's Hospital, Munich, Germany; [3]University Hospital, Berlin, Germany; [4]Clinical Chemistry, Göttingen, Germany.*

**Objectives:** MPA acts as a specific inhibitor of human lymphocyte proliferation by inhibiting IMPDH. Outcome after renal transplantation (RTx) has been shown to depend on pretransplant IMPDH activity in adults. We investigated a potential developmental regulation of IMPDH during childhood.

**Methods:** We analyzed IMPDH activity in peripheral blood mononuclear cells (PBMC) by HPLC (normalized to the adenosine monophosphate (AMP) content of the cells) in 27 healthy infants (2-5.9 yrs), 31 school-age children (6-11.9 yrs), 22 adolescents (12-17.9 yrs) and 106 adults. Pretransplant IMPDH activity was obtained from 31 children and 81 adults with end-stage renal disease (ESRD). 12 hours PK/PD profiles of MPA and IMPDH after RTx were obtained in 17 children and 21 adults under MPA therapy.

**Results:** IMPDH activity displayed high interindividual variability (coefficient of variation 40.5%). Median IMPDH did not differ significantly in healthy infants (80.5 micro-mol/s/mol AMP), school-age children (60.6), adolescents (82.7) and adults (83.1). Pretransplant IMPDH was comparable in children (median 83.8) and adults with ESRD (92.0). IMPDH activity was inversely correlated with MPA plasma concentration in both children and adults.

**Conclusions:** There is no pronounced developmental regulation of IMPDH activity in PBMCs in children. MPA inhibits IMPDH in children and adults to a comparable extent. The analysis of IMPDH activity prior to RTx may optimize MPA therapy.

## Abstract# 512

**Influence of Corticosteroid in Renal Transplantation** G. Roussey, Y. Foucher, G. Guest, B. Ranchin, A. Maisin, R. Novo, J.L. André, S. Cloarec, C. Guyot. *Pediatric Department, CHU Nantes, Nantes, France.*

**Objectives:** To evaluate the relationship between Corticosteroid (CS) and renal function and growth in pediatric renal transplant recipients.

**Methods:** Data were extracted from the French pediatric database for renal transplantation (DIVAT). Children with a functional graft one year after a first transplantation from Jan 1995 to Oct 2007 were included. CS was calculated as a Mean CS Dose per day (MCSD), and was divided into 3 classes: low (<0.1mg/kg/d), intermediate ([0.1-0.2[mg/kg/d) and high (>0.2mg/kg/d). A decrease >30% of the initial eGFR was considered as a significant degradation of renal function. A cox model with time-dependent variables was used to identify significant factors associated with renal survival and dysfunction. Linear mixed model was used to study the growth evolution with time.

**Results:** 535 children had a functional graft at 1 year. Only eGFR (>50ml/min) at 1 year (HR=3.7, p<0.05) and intermediate MCSD (HR= 2.6, p=0.005) were significantly associated with a better graft survival. A low MCSD was associated with a higher risk of renal function degradation. Growth was significantly improved with a low MCSD and eGFR >50ml/min. Interestingly, growth was improved during the first 3 years after renal transplantation for recipients younger than 6 years or older than 12 years, regardless of the MCSD. After this period, growth was improved only for low MCSD.

**Conclusions:** This observational retrospective study should be interpreted with caution. However, long-term administration of CS seemed to be positively associated with a better graft function, but with a negative association on growth. CS continuation or withdrawal has to be discussed for children after renal transplantation.

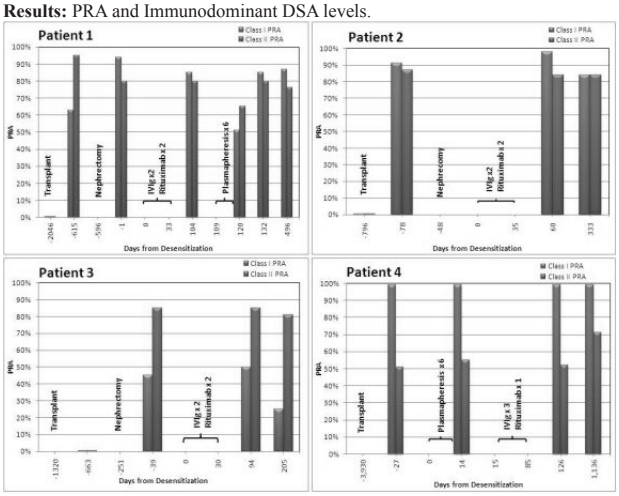
## Abstract# 513

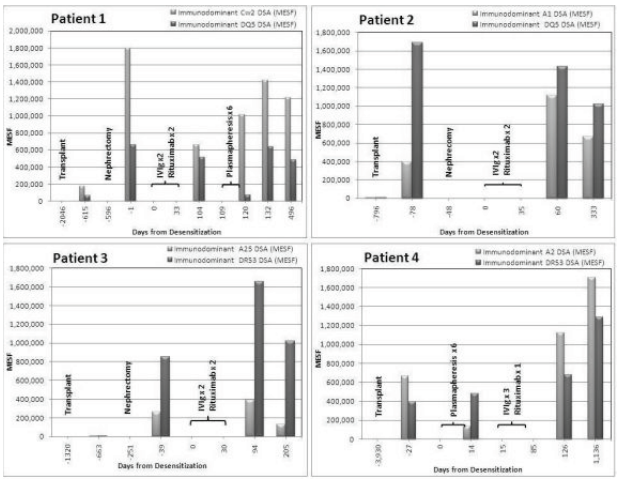
**Effect of IVIG/Rituximab & Allograft Nephrectomy on PRA/DSA in Highly Sensitized Renal Transplant Recipients** R. Ruebner, C. Lind, D. Monos, M. Pradhan. *Pediatrics, The Children's Hospital of Philadelphia, Philadelphia, PA, United States.*

**Objectives:** To describe the effect of IVIG/Rituximab & allograft nephrectomy on panel reactive antibodies (PRA) and donor specific antibodies (DSA) in highly sensitized pediatric renal transplant recipients.

**Methods:** Four highly sensitized patients with renal allograft failure were treated with IVIG & Rituximab. 2/4 patients underwent plasmapheresis and 3/4 had allograft nephrectomy. PRA and DSA were measured by FlowPRA™ single antigen beads. Graft loss occurred 2-8yrs post tx secondary to acute rejection and BK nephropathy in 2/4 patients. Patient 1-3 were treated with 2 doses each of IVIG 2g/kg and Rituximab 375mg/m$^2$ over 30 days. Patient 4 received 3 monthly doses of IVIG 1g/kg and 1 dose of Rituximab 375mg/m$^2$. Patients 1&4 underwent 6 plasmapheresis treatments over 2 weeks.

**Results:** PRA and Immunodominant DSA levels.




**Conclusions:** 1. IVIG & Rituximab did not decrease PRA or DSA in highly sensitized children with renal transplant.
2. No consistent change seen in PRA/DSA following allograft nephrectomy.

## Abstract# 514

**Renal Transplantation in Immunologic High Risk Recipients** P. Salas, V. Pinto, P. Zambrano, J. Grandy, B. Corta. *Pediatrics Nephrology, Exequiel Gonzalez Cortes Hospital, Santiago, Chile.*

**Objectives:** Sensitized kidney recipients have increased significantly. New therapies modulating antiHLA antibody like intravenous immune globulin (IVIG), plasmapheresis (PP) and antiCD20 antibody (ab) have improved transplantation's chances. We report our experience with sensitized children. Pretransplant patients in waiting list for cadaveric transplantation received desensitization with IVIG and living donors received PP until negative crossmatch was achieved. After transplantation they received prophylactic PP and donor specific alloantibody (DSA) monitoring.

**Methods:** Between May 1992 and February 2010, 126 patients underwent kidney transplantation. 9 had PRA> 40%. Median age 12.4 years, 3 females. 8 received graft from diceased donor and one from living donor; median cold ischemic time was16 hrs. Patients missmatch (MM) varied from 1 through 4MM. Patients pretransplant PRA ranges from 0-85%.

**Results:** 2/9 had desensitization with IGEV; 5/9 treated with prophylactic PP after transplantation. All had induction therapy with antithymocite globulin and maintained on prednisone, MMF or azatioprine, and tacrolimus. In 3 an acute humoral rejection (AHR) was diagnosed. Two with graft dysfunction, 3 had DSA and 2 biopsyes showed positive C4D. They were treated with IVIG, PP and AntiCD20 ab. Patients have a mean follow up of 18 months. 100% of graft survival and mean creatinine of 0.94 mg/dl.

**Conclusions:** New protocols with IVIG, PP and antiCD-20 antibodies have become sucessfull in lower anti HLA antibodies allowing renal transplantation. The same therapies can be used after transplantation for AHR. HLA sensitized patients should be monitored for increase risk of antibody mediated rejection.

## Abstract# 515

**Conversion from Twice-Daily Tacrolimus to Once-Daily Tacrolimus in Pediatric Renal Transplant** P. Salas, V. Pinto, H. Corbalan. *Nephrology, Exequiel Gonzalez Cortes Hospital, Santiago, Chile.*

**Objectives:** Non compliance induced from twice-daily tacrolimus (TAC) may adversely influence long term renal allograft outcome. We report 6 months follow-up of patients converted from TAC twice daily to once-daily (OD).

**Methods:** In our department 129 patient have received a renal transplant. 70 are on prednisone, MMF and TAC. 9 patients receiving TAC twice-daily with stable graft function in the last 6 months were converted to OD TAC on a one to one (mg for mg) total daily-dose basis. Through levels of TAC were monitorized on day 7, 14, 28 after converted and then once a month. Target through level range between 4.5-6,5 ng/ml. Renal function and adverse events were analyzed.

**Results:** Patients enrolled had a mean age of 14 years old, 7 female, non hypertension, mean creatinine 0.9 mg/dl, mean baseline TAC through level 5,7 ng/ml. Day 7 after conversion mean through level was 4.5ng/ml, 6 patients increased TAC daily dose. Day 14 after conversion mean trough level was 4.8 ng/ml and 3 patients increased daily dose. One month after conversion mean through level was 4,8 ng/ml and daily dose remained stable in the 9 patients. Mean creatinine at month 2, 4 and 6 post conversion was 1.1 mg/dl. Mean through level at month 2, 4 and 6 were 6.36, 5.5, and 5.4 ng/ml respectively. During the follow up patients didn't showed hypertension, diabetes or hyperlipidemia. 2 patients had urinary infection and one patient had renal disfunction.

**Conclusions:** Children converted to OD TAC may improve compliance to medications. It seems that conversion should be done 1:1.1 (mg for mg) basis for the total daily dose. The OD TAC formulation did not suggest and increased risk of adverse events or greater incidence of acute rejection.

## Abstract# 516

**Varicella Zoster Virus (VZV) Infection in Immunocompromised Patients: Treatment with Specific Human VZV Polyclonal Immunoglobulins** A.-L. Sellier-Leclerc,[1] B. Aoun,[2] V. Baudouin,[1] T. Kwon,[1] G. Deschênes,[1] T. Ulinski.[2] *[1]Department of Pediatric Nephrology, Robert Debré Hospital - APHP, Paris, France; [2]Department of Pediatric Nephrology, Armand Trousseau Hospital - APHP, Paris, France.*

**Objectives:** Infection with the varicella-zoster virus (VZV) is more dangerous in immunocompromised patients than it is in the general population. High doses of acyclovir and immediate reduction of immunosuppression, may improve the prognosis of severe forms, but multiorgan failure and death may occur despite early antiviral treatment.

**Methods:** Here, we report four cases of VZV infection in children receiving steroid therapy and immunosuppressive drugs for renal allograft, idiopathic nephrotic syndrome, and systemic lupus.

**Results:** The only clinical manifestation in three patients was general malaise, fever, and disseminated vesicular rash, whereas one patient also showed severe and diffuse visceral involvement with multiorgan failure. Adjuvant treatment with specific human varicella zoster polyclonal immunoglobulins (Varitect®) led to a dramatic improvement of VZV infection in all four patients.

**Conclusions:** In conclusion, Varitect® is probably a useful adjuvant therapy to acyclovir in declared and severe varicella in immunocompromised children.

## Abstract# 517

**Evaluation of Calcium, Phosphor, Parathoromone, and Alkaline Phosphates before and Early after Transplantation** F. Sharifipour, M.J. Mojahedi, A. Nazeri. *Internal Medicine, Mashhad University of Medical Sciences, Mashhad, Khorasan Razavi, Islamic Republic of Iran; Internal Medicine, Mashhad University of Medical Sciences, Mashhad, Khorasan Razavi, Islamic Republic of Iran; Mashhad University of Medical Sciences, Mashhad, Khorasan Razavi, Islamic Republic of Iran.*

**Objectives:** Renal transplant is the treatment of choice in ESRD patients. Unfortunately complications are common due to previous metabolic disorders and side effects of immunosuppressive therapies.A great portion of these complications can be prevented if diagnosed at early stages.In the present study we try to evaluate the prevalence of hyperparathyroidism in kidney recipients.

**Methods:** This study was conducted on 20 ESRD children receiving successful renal transplant.Serum Calcium, Phosphor,Alkaline phosphates,and Parathyroid hormone were assessed before and after transplantation,one and six months later.

**Results:** Calcium levels were not significantly different but PTH, Phosphor,Creatinine and ALP levels were significantly different before and one month after transplantion :(p=0.005),(p=0.000),(p=0.000)and(p=0.001) respectively.

Sera levels of Calcium, PTH, Phosphor,Creatinine and ALP were also significantly different between the first and sixth mo nth after operation with P values as followed: (p=0.014),(p=0.005), (p=0.000)and (p=0.005).

**Conclusions:** These results showed screening sera of calcium, phosphor, PTH and ALP is helpful in hyperparathyroidism diagnosis. Data analysis showed the longerthe history of dialysis before transplant the higher prevalence of persistent hyperparathyroidism (p=0.0265).

Renal transplant should be considered as soon as possible in ESRD patients to reduce the risk of persistent hyperthyroidism.

## Abstract# 518

**Evaluation of Bone Densitometry in Children with ESRD before and after Kidney Transplantation** F. Sharifipour, M.J. Mojahedi, B. Dadpoor. *Mashhad University of Medical Sciences, Mashad, Islamic Republic of Iran; Mashhad University of Medical Scinces, Mashhad, Islamic Republic of Iran; Mashhad University of Medical Sciences, Mashhad, Islamic Republic of Iran.*

**Objectives:** Decreased bone density is one of the most common complications in renal failure cases who underwent renal transplantation.Early diagnosis and treatment of osteoporosis and osteopenia may prevent these complication.

**Methods:** Bone densitometry was done by DEXA method for 20 ESRD children and who had underwent renal transplantation.

Bone densitometry was performed before, 6 and 12 month after renal transplantation. Z-score in children were used to compare the changes in bone density early after transplantation.

**Results:** All children were completed three stages, among them: 44.8% were female and 45.2% were male, mean age was 9.6 $\pm$ 2.5 years.

Decreased bone density (osteoporosis and osteopenia) in femur and spines was 42% and 29% respectively.Six months after renal transplantation, decreased bone density in femur and spines was 84% and 65% respectively that was significantly different compare to pretransplantation time.

12 months after transplantation, decreased bone density in femur and spines, was seen in almost 68% for both that wasn't significantly changed compare to the first 6 month after transplantation.

**Conclusions:** According to the prevalence of osteoporosis and osteopenia during the first six months after renal transplantation we suggest prophylactic and curative interventions before and early transplantation.

## Abstract# 519

**Anti-Thymocyte Globulin in the Treatment of Early and Late Steroid Resistant Acute Allograft Rejection in Paediatric Renal Transplants** M. Shenoy, M.A. Lewis, N.J. Webb. *Paediatric Nephrology, Royal Manchester Children's Hospital, Manchester, United Kingdom.*

**Objectives:** To evaluate the outcome of early (<3months) and late (>3 months) episodes of corticosteroid resistant rejection (CRR) treated with anti-thymocyte globulin (ATG) in pediatric renal allograft recipients.

**Methods:** Single centre retrospective study of 15 children (10 male), mean age 13.2 years (SD 5.16 y) who received 7-14 days of ATG for treatment of biopsy proven graft rejection which was unresponsive to methylprednisolone from 1994 to 2009.

**Results:** Seven children received ATG for ER (median 26 days post transplantation, range 6-86 days) and 8 for LR (median 763, days, range 390-4223). All the children who developed ER were on a ciclosporin based immunosuppression (IS) at the time of the rejection episode. Following ATG, there was a significant improvement in the 3 month (76umol/l, SD 21, p 0.03) and 6 month serum creatinine (74umol/l, SD 19, p 0.03) when compared with the value prior to ATG treatment (295umol/l, SD 224) in the ER group. In the LR group there was no significant improvement in the creatinine at 3 (147umol/l, SD 45, p 0.17) or 6 months (158umol/l, SD 74, p 0.64) when compared to value prior to ATG (164umol/l, SD 59). At the latest review, the eGFR in the ER group was 72.3 ml/min/1.73m$^2$ (SD 33) (mean follow up 10.4 y, range 3-15y) compared to 37.7ml/min/1.73m$^2$ (SD 17.9) in the LR group (mean follow up of 1.2y, range 0.52-3.2y). During ATG therapy, three children developed septic shock like illness. There was one graft loss in the LR group.

**Conclusions:** ATG therapy in CRR is associated with reversal of rejection and excellent graft outcome in children with ER while the benefits remain uncertain in those with LR.

## Abstract# 520

**Bartter-Like Syndrome Induced by Tacrolimus in a Renal Transplanted Boy: Case Report** A.C. Simoes e Silva, A.C.Q. Mendonça, B.P. Froes, M. Raminho, K. Cartaxo, T.N. Fernandes, J.M. Penido Silva, E.M. Lima. *Pediatrics, Federal University of Minas Gerais, Belo Horizonte, Minas Gerais, Brazil.*

**Objectives:** The aim of this report was to show the development of Bartter-like syndrome in a renal transplanted boy treated with tacrolimus, mycophenolate and prednisone.

**Methods:** CFC, male, 10 years old, previously diagnosed as endoextracapilar glomerulonephritis, was submitted to renal transplant in December 2009 from a deceased donor. Few days after transplant, serum creatinine normalized. Imunossupression included tacrolimus, mycophenolate and prednisone. At the second month after transplant, the patient presented vomits and dehydration accompanied by hypocalemia (1.3 mEq/L), hiponatremia (125 mEq/L), hypochloremia (84 mmol/L) and hypophosphatemia (1.8 mg/dL). He was hospitalized to investigate the etiology of these disturbances.

**Results:** During hospitalization, the patient evolved with polyuria, polydipsia and hydroelectrolyte disorders that needed chronic replacement therapy with potassium salts (8mEq/Kg/day) and sodium chloride (3g/day). Laboratorial evaluation revealed elevation of urinary potassium excretion (excreted fraction, EF=36-86%), hyperphosphaturia (EF=23-36%), hypercalciuria (10-12mg/Kg/day), metabolic alcalosis, hyperfiltration (500ml/min/1.73m$^2$) and proteinuria (1.5g/day). Due to tubular function alterations, tacrolimus dose was reduced, determining a better metabolic profile. However, the patient developed graft rejection (Banff IIb) that needed pulsotherapy and elevation of tacrolimus dose. Despite the recovery of renal function, metabolic disorders worsened.

**Conclusions:** This is the first report of Bartter-like syndrome due to tacrolimus.

## Abstract# 521

**Can Renal Transplant Be Successfully Performed in Children under 6 Years of Age from Adult Donors?** I. Singh,[1] S. Marks,[2] M. McCulloch,[1] J. Taylor,[1] G. Koffman.[1] *[1]Renal Unit, Evelina Childrens Hospital, London, United Kingdom; [2]Renal Unit, Great Ormond Street Hospital for Children NHS Trust, London, United Kingdom.*

**Objectives:** We report our experience of renal transplantation in children under 6 years of age over a 5 year period.

**Methods:** Retrospective review of all children under the age of 6 years who had a renal transplant between 2004 and 2009.

**Results:** Forty five children (78%) male, aged 15-71 (median 37) months received a renal transplant with a median weight and height of 15 (9-25) kg and 87 (74.5-112) cm respectively. 53% had a kidney from a deceased donor. 92% were adult donors. 91% of the transplants were intra-peritoneal with 100% primary wound closure rate.

Median hospital stay was 14.5 (7-60) days. 56% were admitted to PICU for a median duration of 24 (3-504) hours, for ventilation. 28% had a post-operative complication and 38% had an episode of acute rejection. Primary function was seen in 91% of the children, 7% had delayed graft function and 2% had primary non-function. The median creatinine at discharge and 5 years was 33 (12-126) and 94 (62-137) umoml/l respectively.

The 2 year graft and patient survival was 98% and 100% respectively and the 5 year graft and patient survival were both 98%.

1 year graft and patient survival in 1986 compared to 2009.

| | 1986 | 2009 |
|---|---|---|
| 1 Year Graft Survival | 70% | 98% |
| 1 Year Patient Survival | 80% | 100% |

**Conclusions:** We report a large series of children receiving renal transplants under the age of 6 years, predominantly from adult donors, using an intra-peritoneal approach with 100% primary wound closure rate.

Renal transplant can be successfully performed in children under 6 years of age from adult donors.

## Abstract# 522

**Monotherapy Maintenance Immunosuppression in Paediatric Renal Transplantation** R. Sinha, Y. Tse, S. Marks. *Paediatric Nephrology, Great Ormond Street Hospital, London, United Kingdom.*

**Objectives:** To describe a cohort of paediatric renal transplant recipients (RTR) who were converted to monotherapy maintenance immunosuppression (IS).

**Methods:** Single centre retrospective review.

**Results:** 16 RTR's aged 7.9 - 17.2 (median 12.4) years, of whom 88% Caucasian and 63% male were identified who had monotherapy maintenance IS instituted at 2.5 - 13.5 (median 6.5) years post-transplant. All were initially on triple IS except one RTR with atypical haemolytic uraemic syndrome. Nine RTR were switched to MMF monotherapy for chronic allograft nephropathy and seven were switched to CNI (tacrolimus 5, ciclosporin 2) for persistent Epstein Barr (EB) viraemia. Post conversion follow up was for 1 - 4.5 (median 2.1) years. Mean eGFR droppped significantly at two years. This fall was statistically significant only in the MMF (p=0.03) group. Donor specific antibodies (DSA) were negative in all patients at the time of instituting monotherapy, but subsequently became positive in 56% (5/9) among MMF group and 14% (1/7) among CNI group (p=0.09) at 0.8 - 1.6 (median 1.3) years. A strong correlation was observed between appearance of new DSA and fall in eGFR.

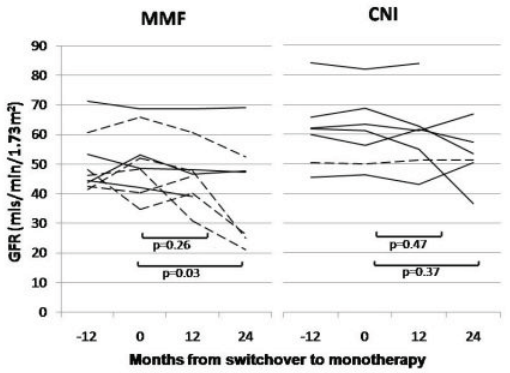



**Figure 1: Estimated glomerular filtration rates of renal transplant recipients among MMF and CNI monotherapy groups. Donor specific antibodies +ve (- - -) and Donor specific antibody -ve (——)**

**Conclusions:** Switch to CNI mono-therapy yielded stable eGFR and was successful in reducing EB viraemia. Larger prospective studies are needed to corroborate our findings as well as determine the role of DSA as a monitoring tool.

## Abstract# 523

**Chronic Kidney Disease Parameters among Paediatric Pre-Emptive and Non Pre-Emptive Renal Transplants** R. Sinha, S. Marks. *Paediatric Nephrology, Great Ormond Street Hospital, London, United Kingdom.*

**Objectives:** To compare chronic kidney disease (CKD) parameters between pre emptive renal transplant (PRT) and non-PRT (NPRT) recipients.

**Methods:** This was a single centre cross-sectional study of renal transplant recipients with at least one year post-transplant follow-up. CKD parameters as per K/DOQI were compared between PRT and NPRT.

**Results:** Of 129 children (39 PRT and 90 NPRT); 54% (70) had Stage 3, 33% (43) Stage 2, 12% (15) Stage 4 CKD and only one child Stage 1 CKD (NPRT). Post-transplant follow-up was 1.3 to 14.2 (median 4.2) years in PRT and 1.2 to 17.7 (median 4.7) years in NPRT (p=0.5) with 36% and 51% living-related renal transplants in PRT and NPRT respectively (p=0.1). Despite similar baseline characteristics a significantly lower proportion of PRT (1, 2%) were in Stage 4 CKD in contrast to NPRT (14, 16%; p=0.03). CKD parameters ((hypertension, anaemia, hypocalcemia, hyperphosphatemia, hyperparathyroidism, hypoalbuminemeia, albuminuria and acidosis) were better among PRT with the incidence of hypertension and acidosis achieving statistically significant difference (p=0.02). Similarly uses of CKD medications (anti hypertensive, iron supplements, erythropoietin, 1 alfacalcidol, phosphate binders and bicarbonate supplement) were more common among NPRT. Among these anti hypertensive, iron supplements, erythropoietin and 1 alfacalcidol showed statistically significant difference (p= 0.03, 0.02, 0.01 and 0.04 respectively).

**Conclusions:** We did demonstrate better CKD parameters and lower use of CKD medications among PRT when compared with NPRT. This finding should act as an an added impetus to paediatric PRT programs.

## Abstract# 524

**Long-Term Outcome of Kidney Transplantation in Children with FSGS** J. Slavicek, L. Puretic, Z. Mustapic, L. Bubic-Filipi. *Dialysis Department, UHC Zagreb, Zagreb, Croatia.*

**Objectives:** Reccurence of focal segmental glomerulosclerosis( FSGS) after renal transplantation ( Tx) has impact on graft survival. Treatment of reccurent FSGS still remain problematic.

We report our experience to access outcome of the renal Tx in children with FSGS.

**Methods:** During period from 1982-2010 in 12 of 98 ( 12.2%) transplanted children primary renal disease were FSGS. Medical records of this 12 pts (4F,8M) were analysed. In all patients FSGS as original disease and as reccurences was biopsy proven. Imunsupression was CyA, Aza, Steroids earlier and CyA/ or Tac, MMF, Steroids recently. Plasmaexchange (PE) was performed in one patient with FSGS reccurence and rituximab with PE in one.

**Results:** The mean age was 11.46+-7.2yrs. The mean follow up after Tx was 6,8+-14,2 yrs. All except two received cadaveric kidney. 2 pts received second graft. Post Tx nephrotic proteinuria was recorded in 3 pts. Reccurence of FSGS was confirmed in 2 patients. Onset of proteinuria was 2 /5days, and 7 months postTx. Proteinuria ranged from 1,5- 35,8 g. Acute cellular rejection episodes were 4 (1,3 per patient). Despite of therapy with PE / rituximab none of the patient achieved remission. Patient with the later onset of proteinuria treated with PE and rituximab had graft loss 3,5 years postTx. Impared graft function was recorded in one patient.

**Conclusions:** Reccurence of FSGS after kidney transplantation is associated with impared graft function and graft loss. Current therapy with PE and rituximab is not allways effective.

## Abstract# 525

**The Factors Effecting Graft Survey in Pediatric Renal Recipients: A Single Center Study** B. Sozeri,[1] O.D. Kara,[1] N. Dincel,[1] A. Keskinoglu,[1] C. Kabasakal,[1] C. Hoscoskun,[2] S. Mir.[1] *[1]Pediatric Nephology, Ege University Faculty of Medicine, Izmir, Turkey; [2]General Surgery, Ege University Faculty of Medicine, Izmir, Turkey.*

**Objectives:** Renal transplantation remains the treatment of choice for end-stage renal failure (ESRF) in regard to patient survival. We report our experience with 96 renal transplantions (Rtx) performed between 1994-2008.Our aim was to evaluate the factors effecting graft survival in pediatric renal transplantation.

**Methods:** We retrospectively reviewed the pediatric renal transplant database at our institution. We evaluated recipient demographics, treatment indications, graft characteristics, graft outcomes, number of HLA mismatches and ischemia time.

**Results:** Ninety-six patients (50 girls/ 46 boys) were involved.Transplantation from 38 living related donors (LRDs) and 58 cadaverics donors was performed. In cadaveric kidney the mean cold ischemia time was 13,2±8,52 hours. The mean number of human leukocyte antigen (HLA) A/B/DR allele mismatches were in patients(2,59±1,7 and 3,25 ±1,25,respectively). 18(20.9%) patients had acute rejection, which were more detected in cadaveric group. In patients with acute rejection had higher mismatches in HLA allele than other patients (2.88±1,23 vs 2.85±1.43, p=0.006).

Ten (10.4%) patients died in follow up time. One-year graft survival was 100% in the LRD group while 98% in cadaver group. Ten year graft survival was %76 in the LDR and %72 in the cadaver group. The graft survey was better in LRD than cadaveric group.
**Conclusions:** The survival rate of LRDs was better than cadaveric grafts. We suggest that the best prognosis in graft survey will be obtaining by transplantation from young healthy donors.

## Abstract# 526

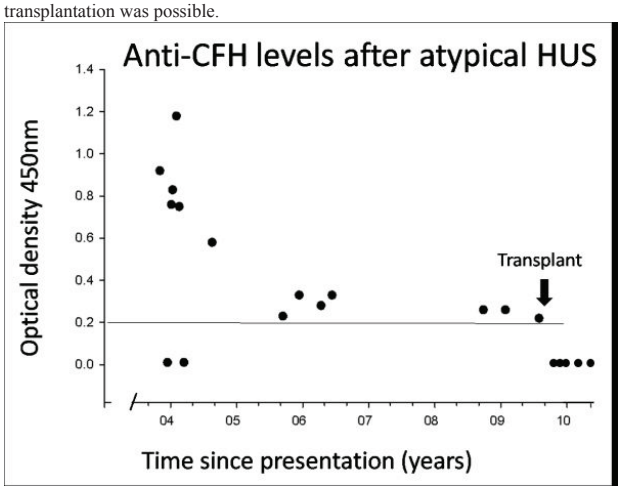
**The Adaptor Protein TSAd Plays a Crucial Role in Alloimmune T Regulatory Cell Function In Vivo** M.P. Stack,[1,2] T. Seto,[1,2] F. D'Addio,[1,2] E. Flynn,[1,2] M. Sayegh,[2] D. Briscoe.[1,2] *[1]Nephrology, Children's Hospital Boston, Boston, MA, United States; [2]Harvard Medical School, Boston, MA, United States.*
**Objectives:** T-cell-Specific Adaptor Protein (TSAd) mediates TCR-dependent activation of T effector cells (Teff), but little is known about its function in immunoregulation.
**Methods:** B6.C-H-2bm12 donor hearts were transplanted into TSAd$^{-/-}$ or C57BL/6 wild type (WT) recipients (H-2b) and the knockout demonstrates accelerated rejection ($p<0.05$) which we investigate.
**Results:** At 16 days evaluation of CD62L$^{lo}$CD44$^{hi}$ Teff cell subsets revealed that the increase in CD8$^{+}$ Teff cells was marked (P=0.08). There was a significant reduction in the numbers of CD4$^{+}$CD25$^{hi}$FoxP3$^{+}$ Tregs in TSAd$^{-/-}$ recipients. Using Luminex, we assessed Teff cytokine responses in TSAd$^{-/-}$ vs. WT recipients splenocytes. We observe increases in Th1 and Th2 cytokines from TSAd$^{-/-}$ recipients. To evaluate Treg function, we harvested spleens on day 16 post-transplant, and isolated CD4$^{+}$CD25$^{hi}$ TRegs. Responders were cocultured with donor APCs, alone, or following the addition of Tregs. The suppression of IFNγ production following addition of WT Tregs into cultures was markedly greater than TSAd$^{-/-}$ Tregs (>30% difference). We evaluated Teff's and regulation in C57BL/6 or TSAd$^{-/-}$ recipients of fully MHC mismatched BALB/c hearts treated with anti-CD40L. We note a significant increase in CD8$^{+}$ Teffs in treated TSAd$^{-/-}$'s, but similar numbers of iTregs in both groups.
**Conclusions:** Collectively, these studies demonstrate for the first time that TSAd is a key molecule in the development of alloimmune regulation following transplantation.Our observations suggest that TSAd-inducible Tregs function to control CD8+ Teff responses in vivo.
**DISCLOSURE:** *Briscoe, D.*: Grant/Research Support, Support for Different Projects in Lab, not related to this project.

## Abstract# 527

**Encrusted Cystitis Caused by MRSA in a Pediatric Renal Transplant Recipient** R. Tanaka,[1] Y. Teraoka,[1] T. Shimooka,[1] E. Hisamatsu,[2] S. Takagi,[2] Y. Sugita,[2] K. Kamei,[3] K. Iijima.[4] *[1]Pediatric Nephrology, Hyogo Prefectural Kobe Children's Hospital, Kobe, Hyogo, Japan; [2]Urology, Hyogo Prefectural Kobe Children's Hospital, Kobe, Hyogo, Japan; [3]Nephrology, National Center for Child Health and Development, Setagaya, Tokyo, Japan; [4]Pediatrics, Kobe University Graduate School of Medicine, Kobe, Hyogo, Japan.*
**Objectives:** The most frequent causative agent of Encrusted cystitis (EC) is *Corynebacterium urealyticum* (CU) that splits urea, resulting in alkaline urine. Here, we describe a girl who developed EC caused by MRSA under aciduria after renal transplantation.
**Methods:** A case report
**Results:** Pyuria and aciduria appeared soon after transplantation and urine cultures were positive for MRSA. She was treated with VCM and EC was diagnosed by CT. VCUG revealed VUR, and we considered that she was at high risk of advancing from EC to encrusted pyelitis (EP). The plaque was removed by cystoscopy, and the dose of MMF was reduced. A histopathological examination of the vesical mucosa revealed severe inflammatory cell infiltration and calcification. Oral ST was prescribed for 5 months to prevent EC recurrence after VCM administration. She remains free of EC at 3 years after discontinuing ST.
**Conclusions:** Recipients seem to be at particularly high risk for EC and require specialized management to avoid the risk of EC progression to EP and graft loss. The most frequent causative agent of EC is CU that splits urea, resulting in alkaline urine. However, our patient did not have alkalinuria. We considered that EC can develop due to etiologies other than alkalinization. This report describes the first indication of MRSA being the primary bacterial cause of EC.

## Abstract# 528

**Natural History of Factor H Autoantibody in Atypical HUS and Successful Living Related Renal Transplant** Y. Tse,[1] L. Kerecuk,[1] K. Marchbank,[2] H. Lambert,[1] M. Ognjanovic.[1] *[1]Royal Victoria Infirmary, Newcastle Upon Tyne, United Kingdom; [2]Institute of Cellular Medicine, Newcastle University, Newcastle Upon Tyne, United Kingdom.*
**Objectives:** Complement factor H autoantibodies (anti CFH) are described in 9% of aHUS patients, often concurrent with additional susceptibility factors or mutations in genes encoding complement regulator/activators. The natural history of anti CFH has not been well described as most reported cases had been pre-exposed to plasma.
**Methods:** We report a girl presenting aged 9 in established renal failure with aHUS. She was not treated with plasma as biopsy showed end stage histology. No mutations were identified in complement regulator genes. Stored serum samples from 4 years after presentation were assayed for anti CFH.
**Results:** Anti CFH were elevated 4 years after presentation and declined to borderline at 6 years. Living related transplant was successful aged 18 after pre-transplant plasma exchange and.rituximab. Immunosuppression regime was basiliximab induction then tacrolimus, steroids and azathioprine. Post transplant plasma exchange was discontinued after 3 cycles as anti CFH became undetectable and remains so at follow up 10 months later.
**Conclusions:** Anti CFH can naturally decline but may take many years. In this patient without other identified complement regulator gene mutations, successful transplantation was possible.




## Abstract# 529

**Protocol Biopsies in Pediatric Renal Transplant Recipients on Cyclosporine *Versus* Tacrolimus** T. Ulinski,[1] B. Aoun,[1] F. Bandin,[2] R. Vitkevic,[1] B. Letavernier,[1] S. Decramer.[2] *[1]Pediatric Nephrology, Armand Trousseau Hospital, Paris, France; [2]Pediatric Nephrology, CHU-Toulouse, Toulouse, France.*
**Objectives:** Protocol biopsies have been useful for the detection of subclinical rejection and early signs of anti-calcineurin toxicity. We evaluated protocol biopsies 3 and 12 moths post transplant in CyA and in FK based immunosuppression.
**Methods:** We included in our prospective study all transplanted children from two centers (A and B) from 2006 to 2008 and studied the results of protocol biopsies 3 and 12 months post transplant in patients on CyA (N=24, center A) *versus* patients on FK (N=12, center B). All patients received basiliximab, MMF and prednisone.
**Results:** In center A, 24 kidney biopsies were performed during the six months after transplantation. 16 protocol biopsies were performed at three months post Tx, 12 were normal and the other four showed 2 borderline rejection and 2 Banff II. 8 biopsies were motivated by increase of serum creatinine, four were normal and four reveled signs of acute rejection (2 borderline and 2 Banff II). 12 protocol biopsies were performed after 12 months; all were normal.
In center B, 12 protocol transplant biopsies were performed at three months post Tx. None showed signs of rejection. All patients underwent a new protocol biopsy at 12 months post Tx and one borderline rejection was detected. No biopsy was performed for increase of serum creatinine in center B.
There were no differences in patient age, number of HLA incompatibilities or other patient characteristics.
**Conclusions:** Patients on FK had less acute rejection episodes detected on protocol biopsies three months post transplant than those on CyA.

### Abstract# 530

**Successful Liver-Kidney Transplantation in a Patient with ESRD Due to Homozygous Factor H Mutation** T. Ulinski, C. Grapin, V. Fremeaux-Bacchi, P. Tissieres, D. Debray, G. Deschenes, A. Bensman. *Pediatric Nephrology, Armand Trousseau Hospital - APHP - UPMC, Paris, France.*

**Objectives:** End-stage renal disease (ESRD) in patients with aHUS due to factor H deficiency is a challenge because of very high risk of disease relapse in an isolated kidney graft, requiring preventive high dose plasma therapy. Liver-kidney transplantation (LKT) is the only strategy to cure definetely Factor H deficiency. Here, we report LKT in a patient with homozygous factor H deficiency.

**Methods:** Atypical HUS was diagnosed at the age of 5 months. $C_3$ was undetectable and factor H activity was <5%. A homozygous factor H (FH) mutation was detected (both parents are heterozygote and healthy). Despite plasma therapy he reached ESRD at the age of 10 years.

As high dose plasma therapy with 2 weekly plasma exchanges (PE) was continued to avoid further disease manifestation, he was put on a liver-kidney transplant waiting list. Therefore, his FH serum levels were 25-55%. Immediately before LKT, the patient received a PE (30 ml/kg) and 30 ml/kg during surgery as a compensation of liquid losses.

**Results:** Both grafts worked immediately, resulting in FH activity of 50%, 72%, and 100% on day 1, 2, and 3 respectively and normal C3 level. Three months post transplant, with a standard FK-MMF based immunosuppression, serum creatinine was 75 micromol/l, proteinuria negative and immunosuppression well tolerated. All hepatic markers are normal.

**Conclusions:** LKT seems to be a possible strategy for patients with aHUS due to homozygous FH mutations. Individual clinical and biological evaluation is mandatory before LKT in patients with FH anomalies.

### Abstract# 531

**Switch to Sirolimus in Pediatric Kidney Transplantation: One Center Experience** A. Vogel, M. Aglony, V. Pérez. *Pontificia Universidad Católica de Chile, Santiago, Chile.*

**Objectives:** In adults, sirolimus has shown to prevent chronic allograft nephropathy. The aim of this study is to determine the evolution of renal function in pediatric kidney receptors after switching the immunosuppressive therapy to sirolimus.

**Methods:** Retrospective analysis of 10 children with kidney allograft switched to sirolimus.

**Results:** The cause of end stage renal disease was: renal dysplasia 2, unspecified 2, ARPKD 1, HUS 1, reflux nephropathy 1,obstructive nephropathy 1, cystinosis 1, FSGS 1. Mean age at transplantation was 8 years. All patients had induction therapy; maintenance treatment was prednisone, cyclosporin and mycophenolate mofetil in 9 and prednisone, tacrolimus and mycophenolate in 1. Patients were switched to sirolimus because of anticalcineurinic toxicity, CAN, hirsutism and PTLD. The switch was made in average 2 years after transplantation (range 3 months- 9 years, 10 months). 6 patients were switched in the first year of transplantation. Trough levels of sirolimus ranged between 4,6 and 11,8 ng/ml. The follow up is in average 2 years 6 months (range 2 months-5 years 8 months). No acute rejection episodes have been documented. After the switch all patients showed an improvement of kidney function; in average the GFR rose from 64,8 to 83 ml/min/1,73m2. Patients switched after the first year had a smaller improvement (average GFR from 40.9 to 46.3 ml/min/1,73m2). No significant variations in leucocyte count, hemoglobin or cholesterol levels were seen.

**Conclusions:** Kidney transplanted children switched to sirolimus had an improvement in renal function. This effect was bigger in children switched in the first year after transplantation suggesting that an early switch may have greater benefits.

### Abstract# 532

**Does Monitoring the Cylex ImmuKnow Assay Help Managing Children after Renal Transplantation?** S. Vyas, J. Orsini, I. Roberti. *Pediatric Nephrology and Transplantation, Saint Barnabas Medical Center, Livingston, NJ, United States.*

**Objectives:** The ultimate goal for an allograft is a balanced immunosuppression. In this study we correlate the immunosuppresion level as determined by Cylex Immuknow assay (Cylex) with clinical events.

**Methods:** A retrospective analysis was done of **children with kidney txp** with Cylex levels.

Demographic data as **age at the time of txp, gender, ethnicity, time posttxp, tacrolimus level and induction therapy** were correlated with **cylex levels**. Cylex (ng/mL ATP) obtained within 6 mos of a significant event (infections requiring hospitalizations or acute rejections) were compared to those from stable patients. All children received induction with basiliximab or thymoglobulin followed by standard regimen with tacrolimus, steroids and mycophenolate mofetil.

**Results:** We had 59 Cylex results in 44 pediatric renal txp recipients done between 1 mo to 9 yrs posttxp (median = 10 mos). Cylex values ranged from 20-728 ng/mL (median 283). **We did not find significant correlation between any of the demographic characteristics studied, tacrolimus level, drug used for induction and cylex levels.**

We had **4 rejections** (3 cellular - all with cylex levels above 250 ng/mL and 1 humoral - cylex = 31ng/mL).

Fifteen patients had cylex levels within 6 months of hospitalization for severe infections: 11/15 (73%) had cylex < 130ng/mL; These levels differed significantly from those obtained in patients without infections (mean levels:140 vs 360 ng/mL) ($p<0.002$).

**Conclusions:** We found that clinical utility of Cylex is limited in children with kidney transplants as it did not correlate with prescribed dosage of medications. However, low Cylex levels were were highly correlated with serious infections.

## Nutrition, Growth, and Development

### Abstract# 533
**(O-65)**

**Factors Affecting Growth in Children Younger Than 2 Years on Peritoneal Dialysis: A Study from the International Pediatric Peritoneal Dialysis Network (IPPN)** L. Rees,[1] D. Borzych,[2] B. Warady,[3] F. Schaefer and coinvestigators.[2] *[1]Nephrology, Gt Ormond St Hospital for Children, London, United Kingdom; [2]Center for Pediatric Medicine, Heidelberg, Germany; [3]Children's Mercy Hospital, Kansas, MO, United States.*

**Objectives:** To determine factors influencing growth in children who commenced chronic peritoneal dialysis(CPD) in the first 2 years of life.

**Methods:** 153 children aged <2 years at entry to the IPPN registry (49 centers in 18 countries), including 84 children with ≥6 months of follow-up.

**Results:** 153 children entered the study at age 1.0±0.6 years. 69(46%) were fed on demand, 51(33%) by nasogastric(NG) tube and 33(22%) by gastrostomy. BMI SDS and HtSDS were higher in gastrostomy fed children: BMI SDS demand fed -0.75±1.70, NG -0.26±1.26, gastrostomy 0.15±1.38; HtSDS demand fed -2.83±1.58, NG -2.57±2.06, gastrostomy -1.88±1.77 ($p<0.05$). Of 84 infants studied prospectively for 1.3±0.6 years, only those fed by gastrostomy had an increase in mean HtSDS: change in demand fed -0.06±1.03(ns), NG -0.01±1.37(ns), gastrostomy +0.75±1.18($p<0.05$). Also children dialyzed with 'biocompatible' PD solutions had a better HtSDS at study entry (-2.09±1.69 vs -2.88±1.86, $p=0.008$) and significant catch-up growth (+0.42±1.45 SDS/year) compared to those using conventional PD fluids(-0.13±0.98,$p<0.05$). According to stepwise multiple regression, a positive age and height-adjusted change in HtSDS was independently predicted by gastrostomy feeding or the mean BMI SDS achieved, use of biocompatible fluids and higher total PD fluid turnover.

**Conclusions:** Gastrostomy feeding, biocompatible PD fluid and high fluid turnover may improve absolute height and longitudinal growth in very young children receiving CPD.

### Abstract# 534
**(O-66)**

**IGF Resistance in Growth Plate Chondrocytes: Inhibition of IGF-I-Related Intracellular Signalling Pathways by Pro-Inflammatory Cytokines** D. Kiepe, U. Huegel, B. Toenshoff. *Pediatric Nephrology, University Children's Hospital, Heidelberg, Germany.*

**Objectives:** Chronic uremia is associated with a resistance towards the anabolic action of IGF-I in growth plate chondrocytes. Because the malnutrition-inflammation-atherosclerosis syndrome (MIA) is a hallmark of uremia, we hypothesized that pro-inflammatory cytokines might interfere with IGF signalling in IGF target tissues.

**Methods:** We used the mesenchymal chondrogenic cell line RCJ3.1C5 as a cell culture model of the growth plate. Cell proliferation was measured by [$^3$H]-thymidine-uptake, cell differentiation was assessed by gene expression (quantitative RT-PCR) of the differentiation markers Indian hedgehog (Ihh) and IGFBP-5. Key signalling molecules of the respective IGF-I-related intracellular pathways were determined by Western immunoblotting.

**Results:** Coincubation with IL-1ß (10 ng/ml), IL-6 (100 ng/ml) or TNF-α (50 ng/ml) inhibited IGF-I-stimulated cell proliferation by 50%, while baseline cell proliferation was not altered by these cytokines. IL-1ß, IL-6 or TNF-α inhibited the IGF-I-activated phosphatidylinositol-3 (PI-3) kinase pathway by 30-50% and the MAPK/ERK1/2 pathway by 50-75%. Also IGF-I-enhanced cell differentiation was reduced in the presence of IL-1ß, IL-6, or TNF-α by 50-80%. In parallel, we observed a 50-80% inhibition of the PI-3-kinase pathway. IL-1ß, IL-6 or TNF-α did not alter basal or IGF-I-modulated gene expression of the inhibitory IGF-binding proteins IGFBP-2, -4, or -6.

**Conclusions:** The resistance towards the anabolic action of IGF-I in the growth plate in conditions of chronic inflammation such as uremia is partially due to the inhibition of IGF-I-specific signalling pathways by pro-inflammatory cytokines.

## Abstract# 535
**(O-67)**

**Changes in Vitamin D Status in Children and Adolescents with Chronic Kidney Disease** M. Denburg,[1] H. Kalkwarf,[2] J. Shults,[1] B. Zemel,[1] M. Leonard.[1] *[1]The Children's Hospital of Philadelphia, Philadelphia, PA, United States; [2]Cincinnati Children's Hospital Medical Center, Cincinnati, OH, United States.*

**Objectives:** To identify determinants of changes in 25-hydroxyvitamin D [25(OH) D] levels in children and adolescents with chronic kidney disease (CKD).

**Methods:** Prospective cohort study of 103 subjects (ages 5-21 yr) with CKD stages 2-5D. Serum 25(OH)D levels were measured at baseline and 12 ( ± 4) months later. Estimated glomerular filtration rate (eGFR) was calculated using the CKiD equation based on sex, height, and creatinine, cystatin C and BUN levels. Multivariate linear regression analysis was used to assess determinants of change in 25(OH)D levels.

**Results:** The median change in 25(OH)D was 2.2 ng/ml (range -22.6 to 35.8). Baseline 25(OH)D level, season, and concurrent changes in serum albumin and eGFR were independent correlates of the change in 25(OH)D. The baseline 25(OH)D was inversely associated (p=0.001) with the subsequent change in 25(OH)D, and decreases in levels were greater (p<0.01) when the second measure was obtained during winter months. A 1 g/dl change in serum albumin was positively associated with a 4.1 ng/ml change in 25(OH)D (p=0.02). A fall in eGFR of >10 ml/min/1.73m2 and being on dialysis were associated with a greater decrease in 25(OH)D of 12.6 and 7.2 ng/ml respectively (p=0.001 and 0.015) compared with pre-dialysis subjects with no decline in eGFR over the study interval. Age, underlying renal disease, and race were not associated with changes in 25(OH)D in this multivariate model.

**Conclusions:** In children and adolescents with CKD, decreases in serum albumin and eGFR and dialysis therapy were independent determinants of worsening vitamin D status.

## Abstract# 536
**PICA: An Important and Unrecognized Problem in Pediatric Dialysis Patients** C. Katsoufis,[1] M. Kertis,[2] J. McCullough,[3] W. Seeherunvong,[1] J. Chandar,[1] G. Zilleruelo,[1] C.L. Abitbol.[1] *[1]Pediatric Nephrology, University of Miami/Holtz Children's Hospital, Miami, FL, United States; [2]Nutritional Services/Pediatric Dialysis, Holtz Children's Hospital, Miami, FL, United States; [3]Pediatric Psychology/Mental Health, Holtz Children's Hospital, Miami, FL, United States.*

**Objectives:** Little is known regarding the metabolic complications of PICA in children. We sought to determine the prevalence of PICA in our pediatric dialysis population.

**Methods:** Eighty-five patients followed on chronic dialysis were surveyed to determine the prevalence of PICA. Dialysis efficiency was estimated by calculating urea clearance per patient volume (Kt/V). Sixty-five (76%) patients were receiving hemodialysis and 20 were maintained on peritoneal dialysis.

**Results:** Mean age was 19.1 ± 8 years. Dialysis efficiency reflected by Kt/V was 1.3±0.4. Seventy per cent of the patients experienced some form of PICA divided into simple "ice" versus "hard" PICA. Ice pica accounted for 51% of the patients. Hard pica occurred in 18.6% and included compulsive consumption of chalk, starch, soap, sand, clay, ajax cleaner, sponge and potting soil. Once PICA was initiated, an "addictive" nature to the consumption became apparent. Greater than 5 years on dialysis was associated with a 6.4 relative risk (RR) of having pica. Anemia resistant to erythropoietin therapy was the most significant morbidity, occurring at a RR of 2. Other consequences included weight loss, hypercalcemia, hypo- and hyperphosphatemia, and hyperparathyroidism.

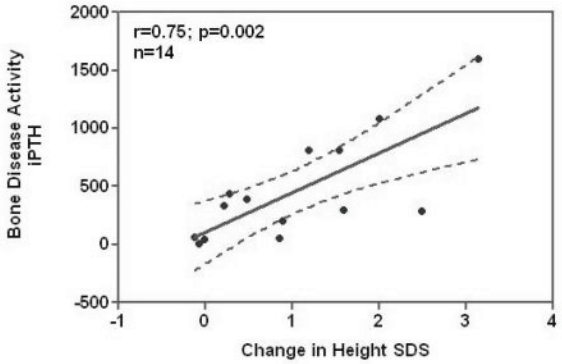
**Conclusions:** PICA is a prevalent and potentially harmful affliction that needs further attention in the nutritional management of dialysis patients.

## Abstract# 537
**Nitrogen Balance and Bone Disease Activity in Pediatric Dialysis Patients Treated with Growth Hormone** C.L. Abitbol,[1] W. Seeherunvong,[1] M. Kertis,[2] D. Francouer,[1] J. Chandar,[1] G. Zilleruelo.[1] *[1]Pediatric Nephrology, University of Miami/Holtz Children's Hospital, Miami, FL, United States; [2]Nuritional Services/Pediatric Nephrology, Holtz Children's Hospital, Miami, FL, United States.*

**Objectives:** To measure growth and nitrogen balance in 14 prepubertal children (8 males) on chronic dialysis during treatment with rhGH.

**Methods:** Growth was correlated to dialysis efficiency (Kt/V) and bone disease as estimated by intact parathyroid hormone activity (iPTH). Anthropometric measurements were expressed as the standard deviation score (SDS). Dialysis efficiency was calculated from the clearance per patient volume (Kt/V). Nitrogen Balance (NB) was calculated by urea kinetics. Growth was the change in HT-SDS after one year.

**Results:** Baseline anthropometric measurements revealed markedly short stature with normal Wt/Ht ratios. A single NB correlated significantly with Kt/V (r=0.74; p=0.005). Average dialysis efficiency correlated significantly with growth after one year(r=0.6; p=0.04). Moreover, growth response correlated positively and significantly with bone disease activity as measured by iPTH (r=0.75 p=0.002).




**Conclusions:** In pediatric dialysis patients, positive nitrogen balance and increased dialysis efficiency appear to improve growth response during treatment with rhGH.

## Abstract# 538
**Validation of Anthropometric Indices Using Composite Scoring of Obesity-Related Morbidity as Diagnostic Criterion** O.F. Bamgbola. *Pediatric Nephrology, Louisiana State University HSC, New Orleans, LA, United States.*

**Objectives:** BMI is widely used for obesity screening. Due to a delayed onset of obesity complications in children, unlike adults there is no clinical validity for anthropometric indices [AI].

Since obesity metabolic profiles are predictive of adult cardiovascular events; relationship between its composite score (OMP) and AI will be examined.

OMP will be validated using MRI-derived visceral fat mass (VFM) as criterion.

**Methods:** 15 subjects [BMI 30-42 kg/m$^2$] & Controls [BMI 18-25 kg/m$^2$] between ages of 5 and 21 yrs will be selected by systematic randomization. Waist-height ratio (WHtR), BMI, & MRI-derived VFM will be obtained.

Abbreviated Composite Score of Obesity Morbid Profiles [OMP]

| Scores* | 0 | 1 | 2 |
|---|---|---|---|
| Systolic BP (percentile) | < 90 | 90-99 + 5 mm Hg | > 99 + 5 mm Hg |
| ECHO: LVH | None | Mild-mod | Severe |
| Triglyceride (mg/dl) | < 100 | 101-104 | > 104 |
| FEV1 | > 95% | 90-95% | < 90% |
| Urine Alb/Cr | < 0.5 | 0.5 - 1.0 | > 1.0 |

*Other indices scored: insulin resistance, non-alcoholic fatty liver, total cholesterol, HDL-C, LDL-C, serum uric acid and CRP**Validation:** MRI-derived VFM will be correlated with OMP, BMI & WHtR while the most predictive model of OMP [indices] for VFM is determined by regression analysis.

**Results: Outcome measure:**

A strong correlation between i) MRI-derived VFM & OMP and ii) OMP for AI will suggest OMP as valid measure of obesity.

**Conclusions:** Composite score of morbidity profiles is a potential diagnostic tool for obesity.

Because obesity score measures morbidity, it may be useful for validation of AI in children

Unlike AI, obesity score is less likely confounded by body morphology (that may vary with ethnicity, puberty & athleticism).

## Abstract# 539
**Double-Blinded Placebo Controlled Crossover Trial To Determine Impact of Folic Acid Use on EPO Resistance in Pediatric Subjects on Chronic PD** O.F. Bamgbola. *Pediatric Nephrology, LSUHSC, New Orleans, LA, United States.*

**Objectives:** Although risk factors are prevalent in dialysis subjects, studies with methodological flaws often suggest absence of folate deficiency (FD).

A study of HD subjects, using therapeutic response to folic acid (FA) as diagnosis, found a high prevalence of FD. Using same criterion, study will examine i) prevalence of FD & ii) impact of FA intervention on EPO resistance in PD patients.

**Methods:** 1-21 yr old PD cohort on EPO, stratified by age, will be assigned to 1-2 mg daily-FA or placebo [PL] for 6 mos. Cross-over performed by 2nd 6 mos. Baseline & monthly lab for CBC, RBC FA, serum homocysteine & vit $B_{12}$ will be assessed. Quantitative score of i) FD (FDS) and ii) EPO requirement per Hb g/dl (ERI) will be obtained.

Indices for the composite score of folate deficiency (FD)

| Folate indices | FD Score |
|---|---|
| MCV ≥ 1SD for age | 2 |
| RDW ≥ 1SD | 1 |
| Hb < 11 g/dl | 1 |
| MCH/ MCHC < 1SD | 1 |
| Serum folate < 2SD | 2 |

**Statistics:** FD diagnosis is assessed by comparing monthly sequential changes (Δ) in Hb, MCV, ERI before and after FA use [1st vs. 2nd 6 mo; Paired t-test]. Impact of FA on EPO use [mean ERI] is examined by analysis of variance [ANOVA] for the FA and PL arms during the 1st vs. 2nd 6 mo of study.
**Results: Outcome:**
i) Decrease in FD score during the 6 mo of FA in a subject; and increase in score in the absence of FA will suggest FD
i) Reduced dose of EPO (ERI) during 6 mo of FA; and an increase in dose in the absence of FA measure impact of FA intervention.
**Conclusions:** Use of therapeutic response to FA as diagnostic criterion is more likely to detect FD in dialysis subjects
Unlike Fe, routine use of FA is not a standard care. Untreated FD may increase EPO requirement and its associated morbidity.

## Abstract# 540

**Prevalence of Metabolic Syndrome in Children and Adolescents with Steroid-Dependent Nephrotic Syndrome** V.M.S. Belangero, L.C. Prates, I.C. Oliveira. *Pediatrics (Pediatric Nephrology Unit), State University of Campinas - UNICAMP, Campinas, São Paulo, Brazil.*
**Objectives:** To evaluate the prevalence of metabolic syndrome (MS) in children and adolescents with steroid dependent (SDNS).
**Methods:** All children followed at the Pediatric Nephrology Unit, aged from five to eighteen with SDNS. The following data were taken from the medical records: age of the onset of NS, time of treatment, total doses of steroids (mg/kg/dial) and previous use of cyclosporine. Weight, stature, waist circumference, blood glucose and insulinemia were determined at the beginning of this study. MS was defined using four previous published definitions by: Cook et al (2003); Weiss et al (2004); Silva et al (2005) and Jollife et al (2007).
**Results:** Twenty-one patients, twelve male, 12.4±3.4 years old were studied. The prevalence of MS varied from 14.28% (3/21) to 23.8% (5/21) according to MS definition. The frequency of overweight was 19.0% (4/21) and obesity was 23.5% (5/21). In the evaluation of predisposing factors associated to MS (time of treatment, doses of steroids, ethnicity, family antecedents of obesity) only the previous use of cyclosporine (p=0.042) and the presence of NS due to systemic disease (p=0.041) were statiscally significant.
**Conclusions:** The high prevalence of overweight and obesity could be related to the high prevalence of MS. Besides that, the use of cyclosporine and the presence of systemic disease are also predisposing factors to MS.

## Abstract# 541

**Effect of Postnatal High Protein Diet on Renal Damages of Rats with Intrauterine Growth Retardation** J. Chen, H. Xu, Q. Shen, W. Guo. *Department of Nephrology and Rheumatology, Children's Hospital of Fudan University, Shanghai, China.*
**Objectives:** Our previous results have shown that intrauterine growth retardation (IUGR) rats had a reduced number of glomeruli and developed hypertension and proteinuria in adulthood. Here we designed the disparity between the prenatal and postnatal enviorment, and investigated the influence of high protein diet on renal development and renal function in IUGR rats.
**Methods:** A model of IUGR was built by maternal low-protein (6%) diet. Male pups were divided into 2 groups, fed either a control diet containing 22% protein (IUGR group) or high protein diet (30% protein, HP group).
**Results:** At the 12$^{th}$ week, the blood pressure and proteinuria in HP group were more server than those of IUGR group(138.6±2.8mmHg vs. 132.1±2.9 mmHg, $P$<0.01; 202.61±62.55 mg•kg$^{-1}$•d$^{-1}$ vs. 118.46±21.85, $P$<0.001), but total number of glomeruli per kidney in HP group was not significantly different from IUGR group(23043±595 glomeruli vs. 22900±926 glomeruli, $P$>0.05). The extracellular matrix area was significantly inceased in HP group compared with that in controls (31.6±2.3% vs. 21.3±2.1%, $P$<0.01). Electron microscopy showed fusion of foot process was scarcely observed in IUGR rats, otherwise there was partial fusion of foot process and hyperplasia of mesangial cells in HP rats. Renal desmin mRNA levels was greatly increased in both IUGR and HP groups, (IUGR:3.33±0.41, HP:4.06±0.27 vs. Con:2.77±0.37, $P$<0.05), with more significant in HP group.
**Conclusions:** Feeding postnatal high protein diet to IUGR rats leads to more severe renal damages. Podocyte injury may be one of the mechanisms in which IUGR and postnatal high protein diet lead to proteinuria.

## Abstract# 542

**Renal Growth in Children with Growth Hormone Deficiency** A. Ece,[1] S. Çetinkaya,[2] S. Eksioglu,[3] S. Senel,[4] S. Özkasap,[4] T. Ginis,[4] V. Sen.[1] *[1]Pediatric Nephrology, Dicle University Medical School, Diyarbakir, Turkey; [2]Pediatric Endocrinology, Dr. Sami Ulus Children's Hospital, Ankara, Turkey; [3]Pediatric Radiology, Dr. Sami Ulus Children's Hospital, Ankara, Turkey; [4]Pediatrics, Dr. Sami Ulus Children's Hospital, Ankara, Turkey.*
**Objectives:** The growth hormone (GH)/insulin-like growth factor system plays important roles in the development and growth of kidney. Despite experimental studies on the role of growth GH, and IGF-1 on renal growth, no clinical data has been reported about the renal growth in children with GHD, that is the aim of this study.
**Methods:** A total of 21 children with IGHD and 15 with MPHD and 68 healthy controls were included. Kidney sizes were measured by ultrasonography and hormone levels were measured by standard methods. The GH treatment durations were 20 and 19.6 months in IGHD and MPHD groups, respectively.
**Results:** Body height, weight and body surface area (BSA) and kidney sizes (kidney length, parenchymal width and kidney volume) were lower in GH-deficient children compared with their controls (p<0.05). However, IGHD/MPHD groups and their control groups had similar kidney volume/body weight and kidney volume/BSA ratios (p>0.05). Significant positive correlations were found between kidney length, kidney volume and anthropometric measurements (p<0.05). Multiple regression analysis revealed that body height was the most significant predictor of kidney volume in children with GHD (p<0.001).
**Conclusions:** Children with IGHD and MPHD had smaller kidneys compared with their healthy controls despite 20 months of rhGH treatment. However relative kidney volumes in respect of body weight and BSA were similar to controls.

## Abstract# 543

**Growth in Patients with Chronic Renal Disease (CRD) during Puberty** C. Fernandez, M. Navarro, M. Carmen, M. Marta, E. Laura. *Pediatric Nephrology, Hospital Infantil "la Paz", Madrid, Madrid, Spain.*
**Objectives:** Analyze the evolution of height and its relationship with other parameters during puberty in patients with chronic renal disease (Stages 1-4).
**Methods:** Retrospective study of 114 patients (87♂ 27♀) with a structural disease in 71% of boys. Various parameters are recorded 2 years prior and after Tanner III stage onset. Data of final height are also recorded.
**Results:** Mean age of onset of Tanner III stage was 13.6±1.2 years for boys and 12.4± 1.4 years for girls. Mean height standard deviation (SD) at -2 years, Tanner III and + 2 years was: -0.69SD (♂)/-0.84SD (♀); -0.6SD(♂)/-1.1SD(♀); -0.5SD(♂)/-1.35SD(♀).SD height remain stable during puberty for boys and girls.
Boys kept a significantly better (p=0,012) standard deviation in height than girls during puberty
The percentage of patients treated with growth hormone at -2 years, Tanner III and + 2 years was: 14% (male)/17% (female), 28% (male)/33% (female), 9% (male)/29% (female) (NS)
There was no significant difference in body mass index (BMI) between boys and girls.
Mean final height was 170.6 ±8.8 cm for boys and 155.5± 6.8 cm for girls. 85.3% of the boys and 80.7% of the girls reached a final height higher than -2SD, with the boys with a significantly better final height (p<0.05).
Final height correlated with the initial height difference (p=0.008) and was independent of GFR, BMI or age of onset of puberty.
**Conclusions:** Althought standard deviation in height remains stable during puberty for boys and girls with CRD, boys reach a significantly better final height.

## Abstract# 544

**Improvement of Nutritional State by Intradialytical Parenteral Nutrition vs. Enteral Nutrition in Hemodialysis Children** Y. Fuentes, G. Toussaint, S. Valverde, A.M. Hernandez, P. Garcia-Roca, L. Ortiz, M.E. Camara, M. Medeiros. *Hospital Infantil de México Federico Gómez, Mexico, Mexico.*
**Objectives:** To examine the effectiveness of intradialytic parenteral nutrition compared to oral supplementation to improve the nutritional state in hemodialysis children.
**Methods:** Prospective, crossover, randomized design in children (6-17 years) on hemodialysis program, with an ABN (anthropometry and bioelectric impedance analysis) score ≤ 10.33 at baseline. During dialysis procedure they received three months the intervention A (oral supplementation) or B (IDPN), designed to provide a third of the required daily caloric intake, protein RDA for age plus 0.5mg/kg/day. There is no washout period and patients switched to another three months of either Treatment A or B in order to complete the other arm of the crossover design.
Nutritional status was evaluated monthly by anthropometry and the ABN score.
**Results:** Nine patients completed six months of treatment. Five started with IDPN and four with oral supplementation. The ABN score improves after six months of nutritional intervention from 8.0 ± 1.5 to 9.5 ± 1.9 p= 0.02, there is also an increase in serum albumin from 3.2 ± 0.4 to 3.4 ± 0.30, p=0.07. Serum cholesterol decreased from 166 ± 42 to 132 ± 22, p= 0.02. There is no statistical difference between groups of nutritional intervention in the z scores of weight, height, body mass index, ABN score and serum albumin after six months of treatment.

**Conclusions:** Nutritional intervention (oral and IDPN) is safe and well tolerated in children with ESRD in hemodialysis. After six months of treatment the ABN and serum albumin improves in both groups.
**DISCLOSURE:** *Medeiros, M.*: Consultant, Novartis, Mexico.

## Abstract# 545

**Vitamin A Deficiency Results in Dysregulation of Lipid Efflux Pathway in Rat Kidney** Q. Li, H. Yang. *Children's Hospital of Chongqing Medical Universtiy, Chongqing, China; Centre for Lipid Research, Chongqing Medical University, Chongqing, China.*
**Objectives:** The aim was to investigate the mechanisms of action of vitamin A deficiency (VAD) on the lipid metabolism in rat kidney.
**Methods:** Adult female rats and their offspring were randomized into three groups: Control: the mother and offspring received a normal diet (4000 retinol IU/kg diet) till 8 weeks; VAD: the mothers and offspring received a VAD diet (400 retinol IU/kg diet) till 8 weeks; Vitamin A–refed: one group of 8 weeks' VAD pups received a complete diet (6500 retinol IU/kg diet) 15 days. The lipid metabolism and immune status of offspring kidney and its relation to the expression of apolipoprotein B100 (Apo-B100), liver X receptor α (LXRα), retinoid X receptor-α/β (RXRα/β), peroxisome proliferator-activated receptorα(PPARα) mRNA expression and protein levels of transforming growth factor-β1 (TGF-β1), interleukin-1β(IL-1β) was analyzed.
**Results:** VAD alters renal lipid metabolism and its immune environment due to expression of apo-B100. Transforming growth factor-B1 (TGF-B1) significantly increased, while ABCA1, a key gene involved in cholesterol efflux and tissue lipid homeostasis, decreased compared with control.Moreover, expression of LXRα, RXRα/β mRNA decreased in VAD rat kidney. Vitamin A refeeding reverted all the changes. And there were no obvious changes of PPARα and IL-1β expression in VAD rats' kidney compared with control or vitamin A -refed groups.
**Conclusions:** Lipid metabolism involved in renal reverse cholesterol transport (RCT) might be mediated by decreasing signaling through the ABCA1 cholesterol efflux pathway, which is significantly modified in kidneys of vitamin A–deficient rats.

## Abstract# 546

**Renal Injury of Methylmalonic Aciduria in 7 Paediatric Cases** X. Liu. *Nephrology, Beijing Children's Hospital Affiliated to Capital Medical University, Beijing, China.*
**Objectives:** We describe 7 patients with methylmalonic aciduria who developed renal dysfunction in our department of nephrology between December 2008 and November 2009 in order to improve the prognosis of the patients.
**Methods:** Seven children with MMA include 3 with hyperhomocysteinemia. The clinical course, laboratory examination, treatment and outcome of the patients detected by GC/MS were analyzed.
**Results:**

nephrotic impairment of the patients

| sex | age | Proteinuria | Haematuria | Hypertension | Edema | Renal failure | cysteine |
|---|---|---|---|---|---|---|---|
| M | 9.8 | + | + | - | + | - | - |
| F | 7.5 | + | - | - | + | - | + |
| F | 3.6 | + | - | + | + | - | + |
| M | 2.6 | + | + | + | + | + | + |
| M | 2 | - | - | + | - | - | + |
| M | 9.5 | + | + | - | - | - | - |
| F | 4.1 | + | + | - | + | + | - |

The nephrotic impairment of the patients are dicribed in table1. After diagnosis 7 patients were treated with high dose vitamin B12 and long term management was include vitamin B12 intramuscular injection and Calomide-Me oral administration. After 3-14 days MMA was decreased expressly, also the renal function of the patients' was improved except No.4 patient was dead of MODS. We followed up all the 6 patients with 1-6 months. The urine analyze, renal function test and mental development were all normal.
**Conclusions:** Nephrotic impairment of the patients with methylmalonic aciduria is not common, most patients presented with tubulointerstitial injury. But in our patients, impairment of glomeruli was principally. Clinical presentations were include proteinuria, haematuria, edema, hypertension and renal failure. We consider we should screen metabolic disease and analyze urine organic acid as early as possible if the patient not only with proteinuria, haematuria and renal failure in nubibus but also with neurological impairment.

## Abstract# 547

**Plants from Sundarbans to the Diet of Lactating Mothers during Puerperium of Barguna District of Bangladesh** A.H. Mollik. *Epidemiology, Biostatistics, Community Nutrition and Noncommunicable Diseases, Peoples Integrated Alliance, Dhaka, Bangladesh.*
**Objectives:** In Bangladesh, it is the custom to breast feed infants for prolonged periods ranging from 06 months up to even 03 years. According to ICMH, studies carried out on nursing mothers have revealed that when they were given extra amounts of body building foods, they produced a large amount of breast milk for their infants. At the same time their health also showed improvement. Inspiring from it, on the same line, a study was undertaken to record the nutritional plants intervention from Sundarbans given to lactating mother immediately after delivery for 42 days and why? Puerperium period is considered to be of 42 days. This paper is presented from a project which is on going to prepare a database on the diet of lactating mothers, nutritional intervention, and special care given to her among different communities of Barguna district, Bangladesh during puerperium.
**Methods:** All the ladies who experienced at least one pregnancy were amongst the sample, or the elderly ladies of the family. The interview schedule was prepared, tested on a small sample, and then data collection was done. All plants were identified and vouchers were stored at the Bangladesh National Herbarium.
**Results:** The plant names obtained in this study included *Hemidesmus indicus*, *Carica papaya*, *Aegle marmelos*, *Punica granatum*, *Nigella sativa*, *Emblica officinalis*, *Asparagus racemosus*, *Swertia chirata*, *Aloe vera*, *Cocos nucifera*, *Syzygium cumini*, *Euphorbia hirta*, *Sesamum indicum*, and *Abrus precatorius*.
**Conclusions:** Taken together; the above plants provide effective remedies for the rural population such that they do not have to visit modern medicinal practitioners.

## Abstract# 548

**Height Impairment and Pubertal Delay in Children with Chronic Kidney Disease (CKD): A Report from the Chronic Kidney Disease in Children (CKiD) Cohort Study** N.M. Rodig,[1] M.F. Schneider,[2] H.M. Hotchkiss,[2] O. Yadin,[2] M.G. Seikaly,[2] B.A. Warady,[2] S.L. Furth.[2] *[1]Pediatrics, Children's Hospital Boston, Boston, MA, United States; [2]Members of the CKiD Study Group, NIDDK, Bethesda, MD, United States.*
**Objectives:** Pubertal delay occurs in children with CKD, but its relationship with growth and measured glomerular filtration rate (GFR) are poorly characterized.
**Methods:** We examined the association of Tanner stage with growth parameters (using age-sex specific SDS) and GFR using repeated measures on 368 children aged > 11 yrs at a total of 893 CKiD study visits.
**Results:** Participants' mean age was 14.9 ± 2.2 yrs. They were Tanner Stage I, II, III, IV, and V in 15%, 16%, 16%, 26%, and 28%, of study visits respectively. Mean SDS for height (ht), weight, and BMI were -0.68, -0.02 and 0.31, respectively. Mean iohexol GFR was 42.6 ± 17.3 ml/min/1.73m2. Participants with ht SDS <-1.88 (n=132 person-visits) or >-1.88 to <0 (n=512 person-visits) had similar GFR levels; both groups had lower GFR compared to those with ht SDS >0 (n=249 person-visits). In groups defined by ht SDS, those who were tallest were more likely to be in Tanner Stage IV or V (P<0.001). After adjusting for age (P<0.001), sex (P<0.001), GFR level (P=0.127), and type of CKD diagnosis (P=0.226), compared to those with a ht SDS <-1.88 those with a ht SDS score of >-1.88 to <0 or >0 had a 1.88 (95% CI: 1.17 to 3.01) and 6.81 (95% CI: 3.84 to 12.05) higher odds, respectively of being in a higher Tanner Stage.
**Conclusions:** Severe height deficits and to a lesser extent lower GFR are associated with delayed onset of puberty.

## Abstract# 549

**Vitamin D Deficiency in Children after Kidney Transplantation: Prevalence, Risk Factors and Response to Therapy in a Predominantly African-American Population** K. Sgambat, L. Ryan, R. Wood, L. Midgley, A. Moudgil. *Children's National, Washington, DC, United States.*
**Objectives:** Pediatric renal transplant(Tx) recipients are at increased risk for bone disease, but there is little data on the prevalence of 25OH Vitamin D(VitD) deficiency in this population.
**Methods:** Records of 46 Tx children were reviewed.VitD was measured in all and classified as deficient(<15), insufficient(16-30),or sufficient(>30 ng/ml).Tx patients with low VitD were treated with VitD2 for 12 weeks.Pretreatment VitD levels were analyzed for associations with risk factors:Pearson's correlation for age and BMIz;Student's t test for gender, season and time post-Tx (early vs.late); ANOVA for ethnicity.VitD of AA Tx patients was compared to healthy AA controls using Chi Square.
**Results:** Of 46 Tx children,34.8% were VitD deficient,28.3% insufficient and 37% sufficient.Of 28 AA Tx,42.9% were deficient and 39.3% insufficient.Of 7 Caucasian,1 was insufficient and 6 sufficient. AA(p=.002) and early(< 6 mo) time post-Tx (p=.019) were significant risk factors for low VitD.Caucasian(p=.002) and summer season (p=.033) were associated with higher VitD.Though mean VitD levels of AA Tx children did not differ from controls(19.9 + 9.2 vs 22.9+ 7.9 ng/dl),AA Tx were more likely to be deficient(p=.019).Of 22 Tx (deficient and insufficient), 5(22.7%) achieved sufficiency after therapy.Of 14 who were deficient,only 2(14.3%) became sufficient post-treatment.
**Conclusions:** VitD deficiency is prevalent after renal Tx, particularly in AA children. VitD should be measured routinely and treated after Tx. Duration of therapy to achieve sufficient VitD is perhaps longer than current guidelines recommend and should be futher investigated.

## Abstract# 550

**Is Abdominal Volumetric Estimate a Better Marker of Visceral Adiposity in Pediatric Obesity?** C.E. Straatmann, O.F. Bamgbola. *Pediatrics, Louisiana State University Health Sciences Center and Children's Hospital, New Orleans, LA, United States.*

**Objectives:** Visceral adiposity has emerged as a better predictor of cardiovascular risk in obese patients. Accurate evaluation of childhood obesity must delineate visceral from subcutaneous adiposity in order to identify patients at high risk for future metabolic consequences. Waist circumference (WC) is a more accurate measure of truncal obesity than body mass index (BMI), but WC is a combined measure of subcutaneous and visceral fat masses. Abdominal volumetric estimate (AVE) may be a novel anthropometric marker of visceral adiposity.

**Methods:** Given the cylindrical morphology of the abdominal cavity, abdominal volume ($cm^3$)= (height)(length)(width)= (2)(abdominal length)(WC). Abdominal length will be measured as the distance from xiphoid process to pubic symphysis and WC will be measured at the superior aspect of the iliac crest. The AVE will be corrected for musculoskeletal mass and will be correlated with MRI-derived visceral fat measurements.

**Results:** MRI-derived visceral fat measurements will be correlated with anthropometric indices (BMI, WC, waist-height ratio, skin fold thickness) and with each corrected AVE formula. The best predictive model will be selected using multiple regression analysis.

**Conclusions:** An anthropometric index that best correlates with visceral fat mass will be most predictive of cardiovascular morbidity. An improved method of estimating visceral fat mass will minimize the confounding effect of gender, ethnicity, and pubertal growth on adiposty. Abdominal volume estimate may be a superior marker of visceral fat mass and may be a better screening tool to target high-risk obese children for specific intervention.

## Abstract# 551

**Nutritional Supplementation in Children on Peritoneal Dialysis** S. Testa, C. Felice-Civitillo, F. Paglialonga, M.R. Grassi, S. Salera, A. Edefonti. *Pediatric Nephrology and Dialysis, Fondazione IRCCS Ca' Granda, Osp. Maggiore Policlinico, Milan, Italy.*

**Objectives:** Protein-energy (PE) malnutrition is a common problem in children on chronic peritoneal dialysis (PD). Nutritional supplementation (NS) has proved to be useful in increasing nutritional intake, but there is a lack of consensus about efficacy, indications, methods of NS and timing.We retrospectively studied the prevalence and outcome of NS in children aged <10 years who underwent PD between 2001 and 2009.

**Methods:** We selected 25 pts (18 M), median age 1.6 yrs (0.4-9). 17 of them underwent NS (Group A) because of PE wasting and 8 were not supplemented (Group B) because of family not compliance (4 pts) or normal nutritional status (4). Methods of NS in Group A were oral supplements (11 pts), nasogastric tube (5) or gastrostomy (1). Daily energy intake (DEI, %RDA), weight (W) SDS, height (H) SDS, sAlbumin, Hb, $HCO_3$ were assessed for each case at the beginning and at the end of the follow-up (FU).

**Results:** Median FU was 20.2 mo (5.8-58) in Group A and 25.8 mo (4-69) in Group B. Median age at the start of FU were 1.4 yrs (0.4-8.9) in Group A and 5.0 yrs (0.7-9) in Group B ($p<0.005$); no differences for the other parameters were present at the start of FU. During FU, Group A showed a significant improvement in W SDS (-1.93±1.14 vs -1.49±1.03, $p<0.05$) but no change in H SDS (-2.0±1.39 vs -1.97±0.95, ns), whereas in Group B W SDS remained stable (-1.75±1.04 vs -1.63±1.07, ns) and H SDS slightly decreased (-1.84±1.27 vs -2.07±1.07, ns).

**Conclusions:** In conclusion, our experience suggests that nutritional supplementation, irrespective of the way of NS, can be useful in increasing energy intake and improving protein energy wasting in children on PD.

## Abstract# 552

**Early Life Overnutrition Impacts on Subsequent Renal Risks in Male Rats: Experimental Evidence of Developmental Programming** K. Yoo, H.E. Yim, K. Ha, I.S. Bae, Y.S. Hong, J.W. Lee. *Pediatrics, Korea University Guro Hospital, Seoul, Republic of Korea.*

**Objectives:** Overnutrition during critical perinatal periods is associated with determining susceptibility to obesity and well known comorbidities. We aimed to investigate the effects of overnutrition during neonatal periods on the development of renal pathophysiological changes.

**Methods:** Rat pups were adjusted to either 3 or 10 pups per mother (overnurition and control, respectively) from day 1 to days 21 of life. We measured the effects of early postnatal nutrition excess on potential renal changes related to obesity by 28 days.

**Results:** Smaller litter male pups weighed heavier than controls on days 4 and between days 10 and 28 after birth ($P < 0.05$). By 28 days of age, overnutrition had no significant effects on renal cell proliferation, apoptosis, numbers of macrophages and glomerulosclerosis. In immunoblots and immnohistochemistry, renin and angiotensin II type (AT) 2 receptor protein expressions significantly increased in overnutrition rats ($P < 0.05$). In contrast, plasminogen activator inhibitor (PAI)-1 and matrix metalloproteinase (MMP)-9 protein expressions decreased in the overnutrition group ($P < 0.05$). AT 1 receptor, monocyte chemoattractant protein-1, tissue inhibitor of MMP-1, tumor necrosis factor-α, osteopontin and adiponectin expressions were not different between the two groups.

**Conclusions:** Our data demonstrate that postnatal overfeeding leads to unexpected and deleterious renal alterations. Increased renin and AT2 may decrease renal PAI-1 and MMP-9, suggesting an inhibitory effect on accumulation of extracellular matrix proteins or the impairment of matrix turnover in neonatally overfed male rats.

## Abstract# 553

**Platelet Aggregation in Children with End Stage Kidney Disease in Relation to Level of L- Arginine as a Risk Factor for Vascular Thrombosis** D.M. Youssef,[1] M.M. Atfy,[1] M.M. Atfy,[2] A.M. Ghareeb.[3] *[1]Pediatrics, Zagazig University, Zagazig, Zagazig, Egypt; [2]Clinical Pathology, Zagazig University, Zagazig, Zagazig, Egypt; [3]Biochemistry, Zagazig University, Zagazig, Zagazig, Egypt.*

**Objectives:** to evaluate the level of L- arginine in patients of ESKD under hemodialysis, and to detect its relation to platelet aggregation as a risk factor for vascular thrombosis, we tried to find the relationship between L-arginine, platelet aggregation and nutritional state of those patients.

**Methods:** comparing data between 30 patients with ESKD under treatment with regular hemodialysis group A; group A1;16 patients with malnutrition, group A2 ;14 patients with normal nutrition state, and group B ;10 subjects as healthy control group, each patient is evaluated for serum arginine level by HPLC, platelet aggregation, and other routine investigation.

**Results:** our study shows significant higher level of platelet aggregation to ADP in group A than group B as it was119±11.9% and 81.1±6.5 %, $p<0.05$, a lower level of serum arginine in group A (71±45.3 µg\l )than in group B (120.5±14.6 µg\l), $p<0.05$, also our study shows a lower level of serum arginine in group A1 (60.5±12.3 µg\l) than group A2 (83±27 µg\l) and a lower level in group A2 than group B (120.5±14.6 µg\l), and we found a negative correlation between serum arginine level and platelet aggregation.

**Conclusions:** we concluded that patients with ESKD has a low level of L-arginine specially in those with malnutrition and that leads to increase in platelet aggregation and this may increase the possibility of thrombus formation, and we suggest the use of nutritional supplementation of L-arginine to suppress this process of atherothrombosis.

## Bone Disease and Mineral Metabolism

## Abstract# 554

**(O-68)**

**Spectrum of ROD in Pediatric CKD** K. Wesseling-Perry,[1] R.C. Pereira,[1] O. Yadin,[1] B. Gales,[1] H. Juppner,[2] I.B. Salusky.[1] *[1]Pediatric Nephrology, UCLA, Los Angeles, CA, United States; [2]Endocrine Unit, Mass General Hospital, Boston, MA, United States.*

**Objectives:** The current study was performed in order to assess the prevalence of lesions of bone turnover (T), mineralization (M) and volume (V) in pediatric patients with CKD.

**Methods:** A cross-section of 59 pediatric pts (age 17.5±0.2 yrs) with CKD stages 2-5 underwent BBx. Prior to BBx, pts received $CaCO_3$ and calcitriol as deemed necessary by their treating physician. S-Ca, P, Alk P'tase, PTH, and FGF-23 (2nd generation C-term, Immutopics®) were obtained at BBx.

**Results:**

| Biochemical Data | | | | | | |
|---|---|---|---|---|---|---|
| | CKD 2 (n=7) | CKD 3 (n=17) | CKD 4 (n=5) | PD (n=10) | HD (n=20) | Normal range |
| Ca (mg/dl) | 9.4 ± 0.1 | 9.2 ± 0.1 | 9.1 ± 0.2 | 8.7 ± 0.3 | 8.9 ± 0.3 | 8.2-10.4 |
| P (mg/dl) | 4.8 ± 0.3 | 4.7 ± 0.2 | 6.2 ± 0.4* | 7.7 ± 0.4* | 6.7 ± 0.5* | 2.2-4.7 |
| Alk Phos (IU/l) | 240 ± 27 | 236 ± 33 | 286 ± 44 | 212 ± 58 | 308 ± 78 | 31-103 |
| PTH (pg/ml) (median (IQ range)) | 64 (40,99) | 102 (68,142)* | 269 (168,708)* | 974 (378,1255)* | 339 (237,878)* | 10-65 |
| FGF-23 (RU/ml) (median (IQ range)) | 162 (105,227)* | 220 (122,456)* | 285 (168,708)* | 4932 (1027, 11216)* | 2410 (834,16652)* | 1-48 |

| Bone Histomorphometric Parameters | | | | | | |
|---|---|---|---|---|---|---|
| BV/TV (%) | 23. 1 ± 2.7 | 27.5 ± 1.5 | 25.9 ± 2.9 | 36.0 ± 4.8 | 27.2 ± 2.6 | 8.9-34.4 |
| OV/BV (%)<br>Pts w/ abnl parameter (%) | 5.1 ± 1.6<br>50% | 8.8 ± 1.8*<br>52% | 10.9 ± 1.6*<br>90% | 4.0 ± 1.2<br>33% | 11.1 ± 2.8*<br>37% | 0.2-5.8 |
| O.Th<br>Pts w/ abnl parameter (%) | 9.0 ± 1.1<br>0% | 14.1 ± 1.2*<br>52% | 14.6 ± 0.8*<br>80% | 10.3 ± 1.1<br>11% | 14.4 ± 2.0*<br>24% | 2-13.2 |
| OMT<br>Pts w/ abnl parameter (%) | 10 ± 1<br>40% | 16 ± 1*<br>77% | 17 ± 2*<br>90% | 27 ± 11*<br>71% | 55 ± 36*<br>53% | 1-12 |
| MLT<br>Pts w/ abnl parameter (%) | 58 ± 14<br>40% | 70 ± 17<br>23% | 61 ± 25<br>10% | 402 ± 272*<br>29% | 467 ± 375*<br>28% | 2-64 |
| BFR/BS<br>Pts w/ abnl parameter (%) | 27 ± 5<br>0% | 42 ± 7<br>13% | 83 ± 18<br>40% | 23 ± 10<br>0% | 36 ± 12<br>11% | 8-73 |

*p<0.05 from normal range

**Conclusions:** Ca, P, and PTH were in the normal range in early CKD, while FGF-23 values were increased in all pts. Skeletal mineralization was impaired in early CKD and worsened as CKD progressed while BFR was normal until late CKD. Thus, increases in FGF-23 and defective skeletal mineralization occur early in the course of CKD and progress as renal function declines, suggesting a key role of the osteocyte in the regulation of skeletal mineralization in all stages of CKD.

**DISCLOSURE:** *Wesseling-Perry, K.*: Other, Honorarium - Genzyme. *Salusky, I.B.*: Other, Honoraria - Genzyme, Johnson&Johnson.

## Abstract# 555
**(O-69)**

**Self-Adjustment of Phosphate Binder Dose to Meal Phosphorus Content Improves Management of Hyperphosphatemia in Children with Chronic Kidney Disease (CKD)** T. Ahlenstiel,[1] L. Pape,[1] J.H.H. Ehrich,[1] M.K. Kuhlmann.[2] *[1]Pediaric Nephrology, Medical School of Hannover, Hannover, Germany; [2]Nephrology, Vivantes Klikum in Friedrichshain, Berlin, Germany.*

**Objectives:** Hyperphosphatemia in CKD is associated with bone disorder and increased cardiovascular mortality. Despite phosphate binders (PB), the prevalence of hyperphosphatemia remains high. An inadequate relation of PB dose to meal inorganic phosphorus (iP) content may be an important factor for failure of phosphate management.

**Methods:** The innovative "Phosphate Education Program" (PEP) bases on patient empowerment to eye-estimate meal iP content by newly defined "Phosphate Units" (PU; 1 PU per 100 mg iP) and self-adjust PB dosage to dietary iP intake by an individually prescribed PB/PU ratio (PB pills per PU). In a prospective study 16 children with CKD and their parents were trained with the PEP-concept and followed for 24 weeks.

**Results:** Within 6 weeks after PEP-training the percentage of children with serum phosphate (PO) >1.78 mmol/l dropped from 63% to 31%. Mean serum PO level decreased from 1.94±0.23 at baseline to 1.68±0.30 mmol/l in weeks 7-12 ($p<0.05$) and to 1.78±0.36 mmol/l in weeks 19-24, whereas serum calcium (2.66±0.3 vs. 2.60±0.23 mmol/l in weeks 7-12) and serum potassium (4.69±0.48 vs. 4.58±0.68 mmol/l in weeks 7-12) remained unchanged. The mean daily PB dose rose from 6.3±2.9 to 8.2±5.4 pills during observation period with increased meal-to-meal-variability ($p<0.05$). Dietary iP intake was not affected by PEP-concept.

**Conclusions:** The empowerment of children with CKD and their parents to self-adjust PB dose to eye-estimated meal iP content significantly improved management of hyperphosphatemia without reducing dietary iP intake.

## Abstract# 556
**(O-70)**

**Accelerated Progression of Vascular Calcification in Paediatric CKD and Dialysis Patients Is Associated with Baseline Vessel Changes** R.C. Shroff,[1] A. Gullett,[1] M. Hiorns,[3] C. Shanahan,[4] L. Rees.[1] *[1]Renal Unit, Great Ormond Street Hospital for Children, London, United Kingdom; [2]Vascular Physiology Unit, Institute of Child Health, London, United Kingdom; [3]Radiology Unit, Great Ormond Street Hospital for Children, London, United Kingdom; [4]Cardiovascular Division, King's College London, London, United Kingdom.*

**Objectives:** Vascular calcification begins early in CKD and progress on dialysis. We compared vascular changes from imaging and arterial biopsy samples to study progression of vascular changes.

**Methods:** 39 children (13 pre-dialysis CKD 4-5 and 26 on dialysis) had vascular imaging (intima-media thickness [IMT], pulse wave velocity [PWV] and coronary artery calcification [CAC] on CT) as well as an arterial biopsy (at the time of renal transplantation or PD catheter insertion). 32 children had a second scans after 10.8 months. The change in vascular measures and correlations with arterial biopsies was noted.

**Results:** At initial measurement the vessel Ca load showed a positive correlation with IMT in dialysis patients ($p<0.003$) whereas 11/13 pre-dialysis patients had normal IMT. PWV and CAC were abnormal in 4 children with highest vessel Ca loads. At follow-up scans, 19 were on dialysis (2 after failed transplants) and 13 had transplants. Dialysis patients had an increase in IMT (0.54 to 0.67mm), PWV (6.4 to 6.9 m/sec) and CAC. IMT progression correlated with baseline IMT and particularly with vessel Ca load ($p=0.03$, $r=0.23$ vs $p = 0.004$, $r=0.58$). Patients with IMT progression had the highest apoptotic index. Transplants showed a deterioration in PWV only.

**Conclusions:** Calcification is rapidly progressive on dialysis and strongly correlates with baseline vessel wall changes.

## Abstract# 557
**(O-71)**

**Bone Assessment in Children with Chronic Kidney Disease: Data from Two New Bone Imaging Techniques in a Single-Centre Pilot Study** J. Bacchetta,[1,2,3] S. Boutroy,[2,3] N. Vilayphiou,[2,3] B. Ranchin,[1] P. Cochat.[1,3] *[1]Hospices Civils de Lyon, Lyon, France; [2]Université de Lyon, Lyon, France; [3]INSERM 831, Lyon, France.*

**Objectives:** Bone damage in children with chronic kidney disease (CKD) is a challenge for pediatric nephrologists. Recent international guidelines have concluded that areal measurement of bone mineral density (BMD) by DXA was of less value in CKD. The aim of this study was to evaluate bone status in CKD children using new bone imaging techniques in a pilot cross-sectional single-centre study.

**Methods:** We performed bone imaging (High Resolution peripheral Quantitative Computed Tomography, HR-pQCT), to assess compartmental volumetric BMD and trabecular microarchitecture in 22 CKD children, 5 children with nephrotic syndrome and 19 controls. In 7 younger patients (i.e., under 10 years of age), we performed bone texture analysis (BMA) in comparison to 15 healthy prepubertal controls.

**Results:** In older children, CKD patients had significant lower height and body weight without significant impairment of BMD and microarchitecture. In univariate analysis, there were significant correlations between cortical BMD and glomerular filtration rate ($r= -0.46$), age ($r= 0.60$) and body mass index ($r= 0.67$). In younger children bone texture parameters were not different between patients and controls.

**Conclusions:** In spite of the small study population, our results seem quite reassuring regarding bone status in CKD children. Novel bone imaging techniques seem feasible in children, but further longitudinal studies are required to thoroughly explore long-term cardiovascular and bone consequences of phosphate-calcium metabolism deregulation during pediatric CKD.

## Abstract# 558
**Open Randomized Clinical Study To Evaluate Efficacy and Safety of Deflazacort Versus Prednisolone in Idiopathic Nephrotic Syndrome** I. Agarwal, J.A. Gemson, P.D. Moses, L. Mathew, P. Prashanth. *Child Health and Pediatric Nephrology, Christian Medical College, Vellore, Tamil Nadu, India.*

**Objectives:** To compare the efficacy and tolerability of Prednisolone versus Deflazacort in children with Idiopathic nephrotic syndrome.

**Methods:** Open labeled, randomized study was conducted in children with first presentation of Nephrotic syndrome.After informed consent, block randomization into Prednisolone or Deflazacort groups was done. Clinical and biochemical examination,response and compliance was evaluated with minimum of 80% being taken for efficacy analysis. For safety profile BMD lumbar spine (DEXA scan) and biochemical tests were done. Statistical analysis was done using SPSS for Windows Version 10.0.

**Results:** 42 children (22 in Prednisolone and 20 in Deflazacort) were recruited and followed up. By 2 weeks 92.8% in Prednisolone group and 78.5% in Deflazacort group were in remission; by 6 weeks both groups had 92.8% in remission. Mean decrease of hemoglobin in Prednisolone group and mean drop in total leukocyte count in Deflazacort group was noted with no significant change in mean fasting blood glucose or serum creatinine in either group. Net decrease in BMD from baseline value was 73.3% in Prednisolone group versus 70.59% in Deflazacort group at 3 months. Between 3-6 months, 20% of Prednisolone had further decrease in BMD compared to 11.6% with Deflazacort. Overall Bone Mineral Content (BMC) reduction was more with Prednisolone (40%vs 23.3%);none were statistically significant.

**Conclusions:** Remission at the end of 6 weeks was comparable in both groups. Decrease in Bone Mineral Density (BMD) was more in Prednisolone group and net effect was more persistent.

## Abstract# 559

**Cinacalcet; a Novel Adjuvant Treatment for XLH** U.S. Alon, C. Haney, W.V. Moore. *Pediatric Nephrology and Endocrinology, Children's Mercy Hospital, University of Missori at Kansas City, Kansas City, MO, United States.*

**Objectives:** We evaluated the long term effect of calcimimetic in treatment of XLH, and report on the first 5 patients who completed at least 120 days of treatment.
**Methods:** The subjects, ages 10, 14.5, 17, 17.5 and 21 years (3 males) were in stable condition on oral calcitriol and K-Phos. After obtaining baseline data, Cinacalcet was added to their medications as a single daily oral dose, 30 mg for those <30kg and 60mg ≥30kg. Every 30 days, doses of calcitriol, Cinacalcet and K-Phos were adjusted based on serum creatinine, Ca, $PO_4$, intact PTH, alk. phosph. and 24-h urine Ca. Pharmacy monitored compliance.
**Results:** Treatment was well tolerated, adherence complete and no adverse effects recorded. At day 120, Cinacalcet dose was 30 mg in the 3 younger and 60 mg in 2 older patients. Calcitriol dose decreased from 16.2±3.5 to 8.1±3.0 ng/kg ($p<0.001$) and K-Phos from 31.0±22.9 to 18.8±17.9 mg/kg ($p<0.05$). Serum creatinine and albumin stayed normal and stable in all. PTH decreased from 44.4±20.6 to 15.6±12.8 pg/mL, Ca 9.82±0.48 to 8.60±0.40 mg/dL, alk. phosph. 237±145 to 220±135 U/L (in all $p<0.05$); $PO_4$ remained within the normal range 2.90±0.28 to 2.74±0.34 mg/dL. In 4 patients, 7 asymptomatic episodes of serum Ca <8.4 mg/dL were recorded. 2 children developed hypercalciuria (>4.0 mg Ca/kg/24h) and responded well to the addition of a thiazide/amiloride diuretic.
**Conclusions:** Cinacalcet was: well tolerated, it lowered PTH level enabling use of lower doses of calcitriol and K-Phos while sustaining serum $PO_4$ level in its therapeutic range. Expected hypocalcemia and hypercalciuria were treatable. Future reports will assess the effect of Cinacalcet on growth, X-rays and kidney ultrasound.

## Abstract# 560

**Vitamin D Deficiency Is Independent of GFR in French Children** J. Bacchetta,[1,2] B. Ranchin,[1] L. Dubourg,[1,2] P. Cochat.[1,2] *[1]Hospices Civils de Lyon, Lyon, France; [2]Université de Lyon, Lyon, France.*

**Objectives:** Vitamin D deficiency is more and more prevalent worldwide whereas current data show that optimal levels should be > 30 ng/mL. The aim of this study was to assess vitamin D status in children issued from the INU23 study.
**Methods:** Biomarkers of phosphate/calcium metabolism, among them 25 OH and 1-25 OH2 vitamins D, were measured in a prospective cohort of 227 children (119 boys, age 11±4 yrs) undergoing GFR assessment (inulin clearance).
A 25OH deficiency was defined by a 25OH level below 20 ng/mL and an insufficiency by a level between 20-30 ng/mL.
**Results:** When GFR decreased, PTH increased and 1-25$OH_2$ decreased. By contrast, 25OH was not modified. A wide majority of children had a low 25OH serum level, independently of their GFR.

| Range of GFR (mL/min/1.73 m²)° | > 140 | 90-139 | 60-89 | 30-59 |
|---|---|---|---|---|
| N | 20 | 116 | 61 | 28 |
| 25OH mean±SD (ng/mL) | 22±11 | 24±10 | 26±10 | 23±7 |
| 25OH median (ng/mL) | 24 | 22 | 26 | 23 |
| 25OH insufficiency (%) | 30 | 37 | 39 | 50 |
| 25OH deficiency (%) | 45 | 41 | 34 | 39 |
| 25OH abnormality (%) | 75 | 78 | 73 | 89 |

Multivariate analysis in KDOQI 1 children showed a significant association between 25OH and PTH / calcium (β parameter -0.36 and 0.21, respectively) whereas in KDOQI 2 children, 25 OH was significantly associated with calcium and urine calcium/creatinine ratio (β parameter 0.56 and 0.24, respectively).
**Conclusions:** Low vitamin D levels are highly prevalent in French children independently of their GFR. New roles of vitamin D in global health have recently been highlighted: vitamin D could be a protective factor against infections, cancers, auto-immune and cardiovascular diseases. All these data reinforce the need of regular vitamin D supplementation in children.

## Abstract# 561

**Management and Diagnostics of Kidney Stones in Bosnian Children** A. Bajraktarevic,[1] A. Hadzimuratovic,[2] A. Hadzimuratovic,[3] J. Ceman Saric,[4] I. Suljevic.[4] *[1]Pediatrics Department, Public Health Institution of Canton Sarajevo, Sarajevo, Bosnia, Bosnia and Herzegowina; [2]Pediatrics Nephrology Department, Pediatrics Clinic Sarajevo, Sarajevo, Bosnia, Bosnia and Herzegowina; [3]Children's Surgery, Clinical Medical Center Sarajevo, Sarajevo, Bosnia, Bosnia and Herzegowina; [4]Biochemical Department, Clinical Medical Center Sarajevo, Sarajevo, Bosnia, Bosnia and Herzegowina; [5]Biochemical Department, Clinical Medical Center Sarajevo, Sarajevo, Bosnia, Bosnia and Herzegowina.*

**Objectives:** Kidney stones are one of the most painful conditions a child can have, and they are one of the most common disorders of the urinary tract. In this article we report our experience with the diagnostic screening and management of children with kidney stones.
**Methods:** This study is a retrospective study of all children who were referred to the Pediatric Nephrology outpatient clinic at the Children's Surgery Hospital of Sarajevo from 2005 to 2007 for the evaluation of kidney stones in children until age of sixteen. The best approach uses spiral computed tomography scans. Ultrasound can detect translucent uric acid stones and obstruction in the urinary tract.
**Results:** Most kidney stones pass out of the body as fluids are increased. Twenty seven children had kidney stones and eleven had hypercalciuria. Twelve kids patients had no hematuria among the presenting symptoms, more than one third had normal urinalysis at our first examination, one half had microcalculi.
**Conclusions:** Treatment options that may be recommended if an increase in fluid doesn't work include extracorporeal shock wave lithotripsy, ureteroscopy, tunnel surgery, or major surgery. The lack of hematuria is not predictive of absence of urolithiasis.

## Abstract# 562

**Facing Neonatal Hypercalcemia, Hypercalciuria Must Not Exclude Inactivating Mutation in the Gene Encoding the Calcium Sensing Recepteur (CASR)** K.H. Braun,[1] A.B. Lienhardt,[2] L.M. Geslin,[1] C.I. Magdelaine,[2] A.F. Armougon,[1] B.R. Boudailliez.[1] *[1]Paediatric Nephrology, CHU University Jules Verne of Picardie, Amiens, France; [2]Molecular Genetic Unit, CHU Limoges, Limoges, France.*

**Objectives:** Phenotype of patient suffering from FFH due to inactivating mutation of CASR is mild hypercalcemia associated with mildly elevated circulating PTH levels and inadapted low urine calcium excretion.
**Methods:** We report a newborn (39 weeks)girl presenting at D7 with asymptomatic hypercalcemia (serum ionized calcium :1.6 to 1.8 mmol/l, N: 1.22-1.40),first kid of non consanguineous parents. Urinary calcium to creatinine ratios were elevated, ranging 1.75-3.7 mmol/mmol [N < 1], serum PTH were low 2.3 pg/ml [5-55].
**Results:** Investigations ruled out Williams Beurren syndrome, cystosteatonecrosis and vitamin D intoxication. Pamidronate acid did not correct hypercalcemia. Follow-up showed a serum calcium decrease (SCa : 1.45 to 1.55) associated with a dramatic decrease in UCa (U Ca/Creat 0.15 to 0.3) at D 120.Her mother also had asymptomatic hypercalcemia (SCa $^{++}$ 1.51 mmol/l) and a low urinary calcium excretion (U Ca/Creat:0;07) whereas father's dosages were normal. Molecular genetic analysis of CASR identified a heterezygotous mutation (c1868G>A) for the girl and her mother. This mutation has been previously reported in FFH.High serum neonatal calcium levels can be explained by really high fetal levels, higher than mother's level,leading to neonatal hypercalciuria in spite of CASR mutation.
**Conclusions:** In neonatal period, hypercalciuria must not exclude the hypothesis of a putative CASR mutation: molecular genetic analysis offers a valuable supplement to the clinical diagnosis of FFH in hypercalcemic neonate.

## Abstract# 563

**Osteoprotective Effect of Calcium and Vitamin D Supplementation in Children with New Onset Nephrotic Syndrome on Steroids** S. Choudhary,[1] I. Agarwal,[1] M.S. Seshadri.[2] *[1]Child Health, Christian Medical College, Vellore, Tamil Nadu, India; [2]Endocrine and Metabolism, Christian Medical College, Vellore, Tamil Nadu, India.*

**Objectives:** To evaluate the effects and prophylactic role of Calcium and Vitamin D on bone health in children with new onset Nephrotic Syndrome (NS) on short term high dose corticosteroid therapy.
**Methods:** Randomized controlled interventional prospective study conducted on 41 steroid naive prepubertal children (29 boys and 12 girls) with new onset NS. All children received prednisolone (60 mg/$m^2$/day for 6 weeks followed by 40mg/$m^2$/day on alternate days for 6 weeks). Recruited children were randomized into Intervention (Vitamin D 1000 IU and 500 mg elemental Calcium daily) and Control groups. Bone Mineral Content (BMC) and Bone Mineral Density (BMD) at lumbar spine were estimated at the beginning and 12 weeks after treatment. The means of percentage change in BMC and BMD over pretreatment values over the 12 weeks study period were compared.
**Results:** Children receiving Calcium and Vitamin D showed a marked increase of 11.2% in BMC of lumbar spine in contrast to controls who showed 8.9% fall over the same time period. The difference between the 2 groups was highly significant ($p<0.0001$). The net intervention attributable difference in BMC was 20.1%. There was a small gain in BMD in both Intervention (2.8%) and Control (0.74%) groups which was not significant ($p=0.27$).
**Conclusions:** In growing children, BMC more accurately reflects changes in the skeleton than BMD. Short term high dose steroid therapy decreases BMC of lumbar spine by 9% in children with new onset NS. Vitamin D and Calcium co-administration not only prevents the decrease in BMC but also enhances BMC of lumbar spine.

## Abstract# 564

**Paricalcitol in Hyperparatiroidism Refractary to Habitual Treatment in Mexican Children** I.E. Del Moral,[1,2] M.F. Rangel,[1] Y. Fuentes.[2] *[1]Pediatric Nephrology, Centro Medico Issemym Ecatepec, Ecatepec, Edo de Mex, Mexico; [2]Pediatric Nephrology, Hospital Infantil de México Federico Gómez, Mexico City, DF, Mexico.*

**Objectives:** Analyze the response of paricalcitol in patients with secondary hyperparathyrodism refractory to calcitriol.

**Methods:** 18 patients with CKD in calcitriol treatment for a period of at least one year on dialysis, with iPTH levels in 1273 pg/ml SD±405.95, average age of 14.6 years SD±2.4;13 in peritoneal dialysis(PD) and 5 in hemodialysis(H). 4 have bone disorders, all with osteopenia in x ray of long bones. After a wash out period of 4 weeks, we calculate the dose of paricalcitol ( 0.1mcg/kg if initial intact parathyroid hormone (iPTH) > 1000pg/ml, 0.08 mcg/kg iPTH level > 500 pg/mL, 0.04 mcg/kg iPTH < 500 pg/mL, 0.02mcg/kg <300 ). We assure levels of P < 6mg/dl and Ca x P product < 75mg/dl. Administration in all patients was intravenous, PD patients once a week and H patients 3 times per week. After iPTH levels lower than 600 pg/dl in PD, we switched to oral dose 3 days a week.

**Results:** After 8 months of follow up iPTH levels 182pg/ml±71.18, rate reduction at 3 months; 66.25±SD 16.89 at 8 months 75.42±15.47, with z -2.896 (0.004) Wilcoxon Signed Ranks Test, calcium 8.6mg/dl±0.95, phosphorus 5mg/dl±1.12, albumin 3.7gr/dl±0.38, CaxP product 44.78mg/dl±.11.2.Have stadistic significancy by Friedman test. Long bone x ray, shows increase in bone density.

**Conclusions:** Paricalcitol decreased iPTH levels in children with CKD, stop the metabolic bone disease, with no significant changes in serum calcium, phosphorus, or CaxP product values during the course of the study, with improve of albumin levels, it can be a useful treatment for hyperparathyroidism that doesn't respond to calcitriol.

## Abstract# 565

**Bone Mineral Disorders in Pediatric Renal Transplantion** A. Derakhshan, A. Ghalehgolab Behbahan, M.-H. Fallahzadeh, M. Basiratnia, M. Lotfi. *Shiraz-Nephro Urology Research Center, Shiraz, Islamic Republic of Iran; Pediatrics, Tabriz University of Medical Sciences, Tabriz, Islamic Republic of Iran; Shiraz-Nephro Urology Research Center, Shiraz, Islamic Republic of Iran.*

**Objectives:** Bone mineral metabolism disorders associated with chronic kidney disease persists following renal transplantation.

**Methods:** In 57 children demographic data, height, weight, serum calcium (Ca), phosphorus (P), alkaline phosphatase (Alk-P), parathyroid hormone (PTH), 25(OH)-Vitamin $D_3$, BUN and creatinine were obtained.Left hand-wrist radiography and "bone mineral densitometry"by DXA technique were carried out. Appropriate softwares were used for interpretation of Z-score of bone mineral density (BMD)and statistical analyses.

**Results:** Fifty seven children all with well-functioning allograft,with a mean age of 18.7 ±4.25, age at transplantation 13.1±3.46 (4.5-20) years with a mean follow up of 67.1±33.8 months were studied. The patients' height-age and bone-age of 11.9±1.8 and15.6±3.3 years respectively, revealed a mean height-age and bone-age retardation of 5.7±2.3 and 1.22±1.47 years; respectively.
Hyperparathyroidism, hyperphosphatemia, hypercalcemia and hypophosphatemia was found in 27(47.3%),9 (15.8%), 9 (15.8%) and 5 (8.8%) of the patients respectively.
The mean BMD Z-score was -1.77±1.13 (-4.2 -1.1) for lumbar spine and -1.64±0.89 (-3.9-1.9) for femoral neck. The BMD Z-scores showed meaningful correlations with the serum Alk-P level independent of serum Ca, P and PTH (p=0.002). Inverse correlation was found between serum level of PTH and GFR (p = 0.011).

**Conclusions:** Relatively high prevalence of bone-mineral disorder in pediatric renal recipients warants a periodic bone densitometry.

## Abstract# 566

**Simple Renal Cysts Are a Sign of Prelithiasis?** M.J. Hernández-González,[1] M.I. Luis-Yanes,[1] M. Monge,[1] S. González-Cerrato,[1] F.J. González-Paredes,[2] V. García-Nieto.[1] *[1]Pediatric Nephrology Uniy, Hospital Nuestra Señora de Candelaria, Santa Cruz de Tenerife, Canary Islands, Spain; [2]Unidad de Investigación, Hospital Nuestra Señora de Candelaria, Santa Cruz de Tenerife, Canary Islands, Spain.*

**Objectives:** Simple renal cysts are rare lesions in pediatric patients. It was considered that the association of simple renal cysts with certain symptoms is just coincidence. There are no studies in children to establish a possible relationship between the presence of simple renal cysts and their association with metabolic abnormalities that cause kidney stones.

**Methods:** We reviewed 22 patients (12M, 10F) diagnosed with simple renal cysts by renal ultrasound. We analyzed renal function, urinary excretion of calcium citrate and investigate about family history of kidney stones.

**Results:** The mean age was 6.04 ± 2.9 years. The symptoms presented at the time of diagnosis were urinary tract infection (40.9%), abdominal pain (27.3%), hematuria (9.1%) and other (18.2%). One patient was asymptomatic (4.5%). The cyst presentation was more common on the left side (56.5%). Nine patients had hypercalciuria (40.9%), three hypocitraturia (13.6%) and two associated hypercalciuria hypocitraturia (9.1%). Thirteen families had history of renal stones. Overall metabolic abnormalities in patients and/or history of stones in their families were present in 19 of the 22 families (86.3%).

**Conclusions:** We found a high proportion of metabolic abnormalities and family history of urolithiasis in children with simple renal cysts. Our hypothesis is that both entities may be related. Further studies are needed to demonstrate the relationship described in this work and the pathological mechanisms involved.

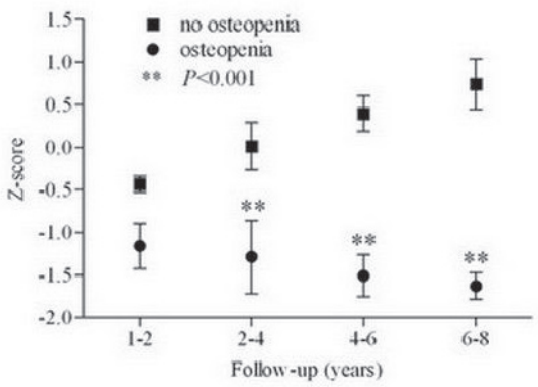
## Abstract# 567

**Follow-Up of Bone Mineral Density in Children with Severe Idiopathic Nephrotic Syndrome (NS)** V. Leroy, P. Parvex, E. Girardin. *Ped Nephrol, HUG, Geneva, Switzerland.*

**Objectives:** To assess the effect of prolonged steroid treatment on bone mineral density in children with NS.

**Methods:** We retrospectively studied patients treated by steroids and MMF, ciclosporine and/or cyclophosphamide. Bone mineral density at the spine were longitudinally evaluated on dual energy X-linked absorptiometry (DXA) and expressed as Z-score.

**Results:** Sixty-three DXA were performed in 17 patients. Initial and final Z-scores, obtained after 2.4±1.7 and 8.1±2.6y of follow-up, were -0.66±0.94 and -0.75±1.08. Eight children experienced osteopenia (Z-score<-1). There was no difference in age and height SDS at diagnosis, yearly number of relapses or immunosuppressors use in children with or without osteopenia. Early DXA did not show any significant difference in Z-scores between the 2 groups. Whereas Z-score significantly increased over time in children without osteopenia, along with the decrease of steroids dosage, it did not improve in children with osteopenia (Fig), despite similar yearly cumulative steroids dosage. At the same time, height SDS remained stable in the first group, but significantly decreased in the second.




**Conclusions:** Moderate bone demineralisation, observed in 50 % of children with severe NS, was not related to steroids dosage. Patients with osteopenia also experienced growth retardation, suggesting that individual factors modulate the effect of steroid on both bone mineralisation and growth.

## Abstract# 568

**Secondary Hyperparathyroidism in Children after Kidney Transplantation** M. Molchanova,[1] E. Molchanova,[2] E. Petrosyan,[1] A. Valov.[2] *[1]Russian State Medical University, Moscow, Russian Federation; [2]Russian Children's Clinical Hospital, Moscow, Russian Federation.*

**Objectives:** Secondary hyperparathyroidism (sHPT) in patients with chronic kidney disease (CKD) is a strong contributor to the development of mineral and bone disorder (MBD). The aim of the research was to investigate the dynamics of the serum parathyroid hormone (PTH) level and growth inhibition in children with a transplanted kidney.

**Methods:** We examined 12 children at the age of 9-17 (14.3 ± 2.9) who had gone through renal transplantation. The serum PTH level and the children's height were measured before the transplantation and a year after it.

**Results:** The serum PTH level was increased in all children before the transplantation and varied: in 2 patients it was less than 300 pg/ml (178 ±5.1), in 4 patients - less than 1000 pg/ml (518.11±90.11) and in the others ranged from 1800 to 2500 pg/ml (2077.33 ± 329.4). The serum PTH level did not depend on the dialysis duration (R=0.5; p=0.39). 10 patients showed definite growth inhibition: 5 patients had height on the 3rd centile, 7 on the 10th centile. A year after the transplantation the serum PTH level remained increased 267.48±188.28

pg/ml in all patients. 3 patients showed considerable longitudinal growth, their height was on the 25th centile. The others remained on the 10th centile despite a certain increase in the height. The research did not reveal any correlations between the height increase and the serum PTH level before and after the transplantation (R=-0.6, p=0.28).
**Conclusions:** Therefore, we concluded that kidney transplantation in patients with CKD leaded to decrease in serum PTH level and improvement of longitudinal growth but further sHPT treatment was still required.

## Abstract# 569

**Vitamin D Metabolites in Children with Nephrotic Syndrome** M. Panczyk-Tomaszewska,[1] D. Adamczuk,[1] A. Mazur,[1] M. Mizerska-Wasiak,[1] M. Roszkowska-Blaim,[1] E. Gorska,[2] A. Stelmaszczyk-Emmel.[2]
*[1]Department of Pediatrics and Nephrology, Warsaw Medical University, Warsaw, Poland; [2]Department of Laboratory Medicine and Clinical Immunology of Developmental Age, Warsaw Medical University, Warsaw, Poland.*
**Objectives:** Glucocorticoid therapy in children with nephrotic syndrome (NS) may lead to bone metabolism disorders. Vitamin D has a number of pleotropic effects in a variety of tissues. The aim of the study was to assess stores of vitamin D in children with nephrotic syndrome during one year observation.
**Methods:** We retrospectively studied 25 children aged 5 to17, mean 10 years; 21 with idiopathic NS and 4 with IgA and Schönlein-Henoch nephropathy. At the start of the study 24 children were given 10-60 mg/48h of prednisone, 23 patients were supplemented with 400-800 IU of vitamin D. After 12 months 11 patients were given 2.5-47.5 mg/48h of prednison, vitamin D was supplemented in 14 children. In all children serum levels of 25-hydroxyvitamin$D_3$ ($25OHD_3$), $1,25(OH)_2D_3$, parathormone (PTH), calcium, phosphorus, alkaline phosphatase (AP), osteocalcin (OC), bone AP were mesured.
**Results:** The defficiency of $25OHD_3$ (<20ng/mL) was found in 9 (36%) children before study period and in 10 (40%) after 12 months (5 of them without glucocorticoid therapy). Positive significant correlation between $25OHD_3$ and OC (R=0.47, p<0.05) and negative correlation between $25OHD_3$ and PTH (R=-0.59, p<0.05) were found. There were no correlation between $25OHD_3$ and prednisone dose and other tested parameters.
**Conclusions:** 1. About 1/3 of children with NS have vitamin D deficiency despite supplementation.
2. The dose of vitamin D used in prevention of osteoporosis in children with NS should be adjusted individually, based on the serum $25OHD_3$.

## Abstract# 570

**Impact of Pharmacological Treatment on Bone Mass and Stature of Children and Adolescents with Primary Hypercalciuria** M.-G. Penido, M. Tavares, M. Cunha, M. Linhares, C. Freitas, A. Barbosa, M. Penido.
*Pediatric Nephrology Unity, Medicine Faculty Federal University of Minas Gerais, Belo Horizonte, Brazil.*
**Objectives:** To evaluate the impact of potassium citrate and/or thiazides on bone mass density (BMD) and growth parameters of children and adolescents with primary hypercalciuria (PH).
**Methods:** Historical cohort of 84 patients with PH. Median follow-up of 12years. All patients were submitted to bone mass densitometry with DEXA-Lunar DPX-IQ. Lumbar-spine BMD (L1-L4), Z-score before (BMD1 and ZS1) and after (BMD2 and ZS2) treatment with potassium citrate and/or thiazides (at least 24m) were performed. Patients were classified in 2 groups: G1 those who followed the prescription and G2 who did not. Height, body weight and respective percentiles according to NCHS were obtained. Height percentiles were categorized (<P5 as 1, P5 as 2, P5-P10 as 3 and so on) and compared before and after treatment. Tests: T-test and Wilcoxon signed-rank test.
**Results:** Forty-six (56%) boys and 38 (45%) girls were regularly followed for 154±40 months. Mean initial height percentile score (MIHPS) was 7.2±2.7 (P25-P50) compared to 7.8±2.6 (P25-P50)p<0.0001. Mean interval between BMD was 27.14±17m. Sixty-four (76%) patients were classified as G1. MIHPS in G1 increased from 7.21±2.7(P25-50) to 8.12±2.69 (P50-75) (p<0.001). No difference was observed in G2. BMD changes significantly from 0.745±0.175 to 0.858±0.184g/cm³(p<0.001).
**Conclusions:** Improvement of BMD with treatment was observed as well as of height percentile scores suggesting a beneficial effect and potential need of treatment. Regular medication use seems to contribute effectively to this effect in contrast to irregular use.

## Abstract# 571

**Bone Evaluation by Quantitative Ultrasonography and Dual Energy X-Ray Absorptiometry in Children with Chronic Kidney Disease and with Renal Transplantation** N. Printza,[1] A. Christoforidis,[1] C. Goga,[1] K. Kollios,[2] F. Papachristou.[1] *[1]1st Pediatric Department, Aristotle University, Thessaloniki, Greece; [2]3rd Pediatric Department, Aristotle University, Thessaloniki, Greece.*
**Objectives:** Aim of this study was to assess bone parameters in children with advanced chronic kidney disease (CKD) and with renal transplantation by dual energy X-ray absorptiometry (DXA), quantitative ultrasonography (QUS) and biochemical markers.
**Methods:** Twenty children with CKD stages III to V (mean age:9.47) and 11 transplanted patients (mean age:11.57) were included. Bone Mineral Density (BMD) at lumbar spine by QUS and Speed of Sound (SOS) at radius and at tibia by QUS were evaluated. Additionally, intact parathormone (iPTH), 25-hydroxycholecalciferol ($25OHD_3$), serum bone alkaline phosphatase (BAP) Ca, P and creatinine were measured.
**Results:** Results showed that 30% of patients with CKD had SOS Z-score at tibia <-1 and 52.7% at radius <-1, whereas only 16.67% had BMD Z-score <-1. Regarding transplanted patients 27% had SOS Z-score at tibia <-1, 70% at radius <-1 and 81% BMD Z-score <-1. Serum iPTH and BAP were higher in patients with CKD compared to transplanted patients (p<0.001, p<0.001 respectively). In patients with CKD multivariate regression analysis showed that SOS Z-score measured at tibia was the major predictor of increased levels of serum iPTH. No correlation between biochemical markers and DXA was found.
**Conclusions:** In patients with CKD, QUS is better than DXA correlated to biochemical indices of osteodystrophy such as PTH and seems to effectively detect bone disease. Transplaned patients presented high incidence of abnormal DXA, probably due to steroids induced osteoporosis.

## Abstract# 572

**Ultrasound and Scintigraphy of the Parathyroid Glands in Children at a Specialist Paediatric Nephrology Centre: 5 Year Experience** V. Shah,[1] M. Easty,[2] S. Ledermann,[2] L. Rees,[2] R. de Bruyn,[2] R. Shroff.[2] *[1]Imperial College Hospitals NHS Trust, London, United Kingdom; [2]Great Ormond Street Hospital for Children, London, United Kingdom.*
**Objectives:** Secondary and tertiary hyperparathyroidism are commonly encountered in children with CKD. This may be due to hyperplastic (Hyp) or adenomatous (Ad) change in the parathyroid glands (PTG). We performed ultrasound (US) and scintigraphic (Tc99m-MIBI) imaging of the PTG at a specialist paediatric nephrology centre over a 5 year period, to evaluate the relationship between biochemical parameters and imaging findings.
**Methods:** Of 23 children, 6 were in CKD stage IV-V, 16 on dialysis and 1 post-transplant. The median age was 11.8 (range 1-18) years. All 23 underwent US and 9 had MIBI; results were categorized as normal, Hyp or Ad. Biochemical data and relevant medication history over the preceding 12 months was recorded.
**Results:** 12/23 had a normal US. US suggested Ad in 5; all 5 had MIBI, with 2/5 positive for Ad. US suggested Hyp in 6 of whom 1 had MIBI which was normal. 1 MIBI identified an ectopic PT Ad in the mediastinum. Patients with Ad had a significantly longer time in CKD stage IV-V and on dialysis (p<0.001). The mean serum Ca and percentage of hypercalcaemic episodes was significantly higher in patients with Ad compared to other groups. There was no difference in PTH levels based on imaging findings, but patients with Hyp and Ad had lower 25-hydroxyvitamin-D and 1,25-dihydroxyvitamin-D levels (p<0.001).
**Conclusions:** Children with PT Ad have longer duration of CKD and dialysis as compared to those with Hyp or normal PTG. Vitamin D supplementation and careful monitoring of levels may prevent the development of PT Hyp and Ad.

## Abstract# 573

**Bilateral Slipped Capital Femoral Epiphysis (SCFE) in a Boy with Familial Hypomagnesemia with Hypercalciuria and Nephrocalcinosis (FHHNC), Renal Failure and Severe Hyperparathyroidism** P. Sikora,[1] M. Zajaczkowska,[1] T. Raganowicz,[2] H. Borzecka,[1] A. Gregosiewicz.[2]
*[1]Pediatric Nephrology, Medical University of Lublin, Lublin, Poland; [2]Pediatric Orthopedics, Medical University of Lublin, Lublin, Poland.*
**Objectives:** SCFE is a common orthopedic disorder affecting otherwise healthy adolescents in which the femoral head is displaced from its normal position. The most cases are classified as idiopathic but SCFE may be also secondary to different endocrinopathies. Exceptionally, SCFE was also described as a consequence of chronic renal failure (CRF).
**Methods:** Therefore, we present a patient with FHHNC and bilateral SCFE as a consequence of untreated osteodystrophy in a course of CRF.
**Results:** The 15-years old boy with diagnosis of FHHNC and CRF was admitted to our clinic due to severe knees pain. During last 2 years, he discontinued the adequate treatment. Initial laboratory tests revealed uremia, hypocalcemia, hyperphosphatemia, extreme hyperparathyroidism (PTH- 3977 pg/ml), metabolic acidosis and anemia. Therefore hemodialysis accompanied by typical

conservative treatment was started. Radiographs of hips showed bilateral SCFE of severe grade. After 2 ½ months of treatment and lowering serum PTH level below 300 pg/ml, successful surgical stabilization in situ of both epiphyses was performed.
**Conclusions:** There is a first reported case of FHHNC complicated with SCFE. Although this association is rather accidental, we believe that severe secondary hyperparathyroidism due to untreated renal osteodystrophy, additionally aggrevated by FHHNC per se, was a causative factor. Therefore proper management of these conditions is essential to prevent SCFE.

## Abstract# 574

**Parathyroid Hormone-Related Peptide (PTHrP) Mediated Hypercalcemia in Benign Congenital Mesoblastic Nephroma (CMN)** A. Kats, U.S. Alon, T. Srivastava. *Children's Mercy Hospital, Kansas City, MO, United States.*
**Objectives:** PTHrP-mediated hypercalcemia of malignancy is rare in children, nor is hypercalcemia in the setting of a benign tumor common. We report 2 infants with the rare condition of PTHrP-mediated hypercalcemia secondary to a benign tumor.
**Methods:** We describe our experience with 2 infants who presented with severe hypercalcemia secondary to secretion of PTHrP from benign CMN, and their outcome after removal of the tumor.
**Results:** Boy #1- presented as 29 wk preterm 1.4 kg with known left renal mass. On day 2 serum Ca 13.7 mg/dl, iCa 1.99 mmol/L, P 7 mg/dl, PTH <3 pg/ml, PTHrP 17 pg/ml (Mayo Clinic, Normal <2), normal vitamin D levels. The 8x6x4 cm mass was excised on 16d showing Stage 2 CMN.
Boy#2- A term baby presented at 34d, weight 2.96 kg with emesis, lethargy and abdominal mass. Serum Ca 16.7 mg/dl, iCa 2.12 mmol/L, P 4 mg/dl, PTH <3 pg/ml, PTHrP 378 pg/ml (Quest Diagnostics, Normal <27), normal vitamin D levels. The 7x6x5 cm mass was excised on 41d showing Stage 3 CMN.
Hypercalcemia was corrected with saline hydration, lasix, calcitonin and pamidronate. A week after surgery PTHrP normalized. Post-op patients had high serum PTH and alkaline phosphatase, with low (or normal) serum Ca, P, and undetectable urinary Ca and P due to their movement into bone. Children were treated for 6-8 weeks with calcitriol, Ca and P until urinary Ca and P re-appeared, suggesting bone replenishment.
**Conclusions:** The pediatric nephrology community should be aware that benign tumors such as CMN can secrete PTHrP, leading to hypercalcemia. Following the excision of the PTHrP generating tumor one should anticipate a transient "hungry bone"-like condition requiring Ca, P and calcitriol therapy.

## Abstract# 575

**Hypervitaminosis A and Elevated Parathyroid Hormone Related Protein (PTHrP) in a Toddler with End Stage Renal Disease (ESRD) and Hypercalcemia** L.K. Steinmetz, D.M. Okamura, J.T. Flynn. *Pediatric Nephrology, Seattle Children's Hospital, Seattle, WA, United States.*
**Objectives:** To review hypervitaminosis A in patients with ESRD on dialysis and suggest a possible interaction between retinol and PTHrP in the development of hypercalcemia.
**Methods:** We describe a 2 yr old female with ESRD secondary to a pauciimmune necrotizing glomerulonephritis who developed hypercalcemia approximately 4-6 weeks after progressing to ESRD and being started on chronic dialysis. Her ionized calcium ranged from 1.3-1.76 mmol/L and total calcium ranged from 10.7-13.3 mg/dL for several months despite stopping both her activated and inactivated vitamin D. Her PTH level was <30 pg/mL. Vitamin D 25-OH was 47mg/ml (range: 30-100). Her alkaline phosphatase rose in conjunction with calcium levels. A further workup for possible causes of hypercalcemia was obtained.
**Results:** Vitamin A level was elevated at 2036 mcg/L (range: 200-430). PTHrP was also elevated at 359 pg/ml (range: 14-27). She has no signs or symptoms of malignancy and a bone scan was normal. After decreasing her vitamin A intake, her vitamin A level has decreased to 1403 mcg/L and her and calcium levels have trended down.
**Conclusions:** Studies suggest that hypervitaminosis A adversely affects bone metabolism and increases bone turnover but the mechanism remains unclear. PTHrP also modulates endochondral bone formation and has been shown to interact with retinol binding protein in chondrocytes suggesting a possible relationship between vitamin A and PTHrP effects on bone metabolism. This relationship may explain the elevated PTHrP in the context of hypervitaminosis A. Further animal studies are needed explore this possible link.

## Abstract# 576

**Bone Metabolism and Fibroblast Growth Factor 23 (FGF23) after Pediatric Renal Transplantation (PRT)** M. van Husen, A.-K. Fischer, I. Klaassen, A. Lehnhardt, K. Möller, M.J. Kemper. *Pediatric Nephrology, University Medical Center, Hamburg, Germany.*
**Objectives:** FGF23 is a circulating factor that regulates renal reabsorption of phosphate and inhibits renal 1α-hydroxylase activity. Data on FGF23 in children, especially after PRT are scarce.
**Methods:** Intact FGF23 and parameters of bone metabolism were analyzed in 62 children after PRT and 11 controls. Estimated GFR (eGFR) after PRT ranged from 15-174 ml/min*/1.73qm. Median time after PRT was 40 months (range 5-135).
**Results:** Mean (±SE) serum FGF23 and PTH levels after PRT were significantly increased compared to controls (142±28 vs. 43±3 pg/ml, p=0,001 and 177±39 vs. 74±18 ng/l, p=0,003, respectively). FGF23 was significantly higher in children with an eGFR <60 ml/min*1.73qm compared to those with eGFR >60 ml/min*1.73qm (268±64 vs. 62±4 pg/ml, p<0.001). PRT patients showed a significant inverse correlation between serum FGF23 and eGFR (r=-0.45, p<0.0001). FGF23 closely correlated with PTH levels (r=0.69, p<0.0001), but not with serum phosphate levels. 9 patients (14.5%) showed a persistent hypophosphatemia after PRT (14±7 months) according to KDOQI-criteria. A correlation of phosphate levels and tubular phosphate reabsorption (TPR) was found (r=0.78, p=0.012). In contrast to PTH, FGF23 showed a negative correlation with TPR (r=-0.9, p=0.001).
**Conclusions:** These first data in children after PRT indicate a significant increase of FGF23. Further studies on the impact of FGF23 on hypophosphatemia in the early post operative period are necessary. Taken together, FGF23 seems to be a key regulator with dual effect on calcium-phosphate homeostasis after renal transplantation, both early after PRT and in chronic graft dysfunction.

## Abstract# 577

**A MC, Efficacy, Safety and PK Study of Cinacalcet in Dialyzed Children with SHPT: Protocol Outline** K. Perri,[1] O. Della Casa Alberighi,[1] A. Pistorio,[1] G. Gianoglio,[2] S. Maringhini,[2] G. Montini,[2] C. Pecoraro,[2] S. Picca,[2] P. Sorino,[2] R. Chimenz,[2] G. Lavoratti,[2] F. Paglialonga,[2] I. Ratsch,[2] E. Vidal,[2] E. Verrina.[2] *[1]IRCCS Gaslini, Genoa, Italy; [2]Italian Pediatric Dialysis Registry, Genoa, Italy.*
**Objectives:** This MC, intra-subject controlled, open-label, active-treatment study will assess in dialyzed children aged 2-18 yrs with SHPT not responsive to standard of care (SoC) therapy, the response after 6-mo cinacalcet (Ci) compared intra-subject to SoC alone at screening visit 6 mos before Ci start. Secondary objectives: effect on growth over 18 mos and PK profile.
**Methods:** At baseline children have PTH levels≥300 pg/mL, plasma P<6 and Ca>9.4 mg/dL, or CaxP product>60. Initial Ci dosing will be 0.5-0.75 mg/Kg *per os* OD to be adjusted up to a max of 180mg for target PTH values<180 pg/mL. In 12 centres of a national pediatric dialysis registry, 30 children will be enrolled, corresponding to an α=0.05 and a power of 80% using McNemar test, with an expected % of responders to Ci or SoC of 40% or 5% respectively, and a 15% drop-out rate.
**Results:** Primary endpoint (EP) will be % of children who will have a reduction from baseline ≥25% in mean PTH levels during the 6-mo efficacy-assessment period. Secondary EPs over 18 mos will be: % of patients with mean PTH levels<300 pg/mL; % change in PTH, Ca, P, and CaxP product; PK profile (or population profile by age) and its correlation with PTH and testosterone levels; auxological indices and growth velocity; % of children with treatment-emergent AEs and lab abnormalities; retention on treatment and reasons of treatment withdrawal.
**Conclusions:** The study will evaluate whether Ci represents a safe and effective therapeutic option for SHPT children.

## Abstract# 578

**FGF-23 Levels Predict Response to Treatment of Secondary Hyperparathyroidism** K. Wesseling-Perry,[1] R.C. Pereira,[1] B. Gales,[1] H. Wang,[2] R. Elashoff,[2] H. Juppner,[3] I.B. Salusky.[1] *[1]Pediatrics, UCLA, Los Angeles, CA, United States; [2]Biomathematics, UCLA, Los Angeles, CA, United States; [3]Endocrine Division, Mass General Hospital, Boston, MA, United States.*
**Objectives:** To evaluate whether levels of FGF-23 predict biochemical and skeletal resistance to treatment of 2°HPT.
**Methods:** 60 pts on CCPD (age 13.9 ± 0.5 yrs) with BBx proven 2°HPT received either sevelamer or $CaCO_3$ in conjunction with either calcitriol or doxercalciferol for 8 mos. S- Ca, P, Alk P'tase, 1st PTH-IMA, and FGF-23 were obtained monthly. Vitamin D sterols and P binders were adjusted to maintain serum PTH levels between 300-500 pg/ml, Ca between 8.4 and 10.2 mg/dl, and P between 4-6 mg/dl. BBx was repeated after 8 mos of therapy. Predictors of response to therapy (final PTH: 300-500 pg/ml) were assessed by ROC.

**Results:**

| Parameter | Group 1 (Doxercalciferol + $CaCO_3$) (n= 16) | | Group 2 (Doxercalciferol + Sevelamer) (n= 14) | | Group 3 (Calcitriol + $CaCO_3$) (n=16) | | Group 4 (Calcitriol+ Sevelamer) (n=14) | |
|---|---|---|---|---|---|---|---|---|
| | Initial | Final | Initial | Final | Initial | Final | Initial | Final |
| Calcium (mg/dl) | 8.9±0.2 | 9.7±0.3* | 9.0±0.2 | 9.1±0.2 | 8.9±0.2 | 9.9±0.3* | 8.7±0.2 | 9.0±0.2 |
| Phosphorus (mg/dl) | 6.4±0.3 | 6.8±0.8 | 6.6±0.5 | 5.9±0.5 | 6.3±0.4 | 5.6±0.3 | 5.9±0.3 | 6.0±0.3 |
| Alk P'tase (IU/L) | 317±48 | 233±58 | 437±92 | 260±64* | 429±57 | 236±49* | 410±93 | 307±87* |
| PTH (pg/ml) | 840±109 | 357±75* | 853±137 | 610±179* | 1051±84 | 505±76* | 1013±115 | 605±136* |
| FGF-23 (RU/ml) | 1469 (264,5280) | 4684 (1840,11003) | 437 (169,4347) | 1024 (282,8310) | 936 (247,2603) | 1345 (714,2078) | 262 (179,1073) | 571 (369,6159) |
| FGF-23 (% baseline) | | 732±434* | | 592±191* | | 676±423* | | 328±85* |
| BFR/BS ($um^2/mm^2/d$) | 124±41 | 45±22* | 103±41 | 53±29* | 152±77 | 62±52* | 101±70 | 60±27* |

**p<0.05 from baseline, †median (IQ range)*

Pts treated with sevelamer received higher vitamin D sterol doses than those treated with $CaCO_3$ ($p<0.05$). FGF-23 levels increased and BFR/BS decreased similarly in all groups throughout the study. The AUC for baseline FGF-23 < 1000 RU/ml and baseline PTH were 0.76 and 0.58, respectively. FGF-23 levels increase similarly with different D analgues and P binders and baseline values did not predict response of BFR to therapy.

**Conclusions:** Baseline FGF-23 values predict the biochemical, but not skeletal, response of 2oHPT to therapy, suggesting a role for FGF-23 in skeletal resistance of bone to PTH in ESRD.

**DISCLOSURE:** *Wesseling-Perry, K.*: Other, Honorarium - Genzyme. *Salusky, I.B.*: Other, Honoraria - Genzyme and Johnson & Johnson.

## Genetics

### Abstract# 579
**(O-73)**

**Rare Copy Number Variants in Congenital Anomalies of the Kidney and Urinary Tract** S. Sanna-Cherchi,[1] K. Burgess,[1] M. Bodria,[2] K. Kiryluk,[1] R. Sterken,[1] P.L. Weng,[1] N. Kacak,[1] L Allegri,[3] F. Scolari,[4] R.P. Lifton,[5] A. Latos-Bielenska,[6] V. Tasic,[7] G.M. Ghiggeri,[2] A. Gharavi.[1]
*[1]Columbia Univ, NY, NY, United States; [2]G.Gaslini Inst, Genoa, Italy; [3]Children's Hosp, Skopje, Macedonia, The Former Yugoslav Republic of; [4]PRCM, Poznan, Poland; [5]Univ Hosp, Parma, Italy; [6]Hosp, Montichiari, Italy; [7]Yale Univ, New Haven, CT, United States.*

**Objectives:** Congenital anomalies of the kidney and urinary tract (CAKUT) are the leading cause of pediatric ESRD. An innovative strategy to discover genes relies on genome-wide search for copy number variants (CNVs). Significant evidence indicates that CAKUT can be caused by rare CNVs.

**Methods:** We performed a genome-wide search for CNVs using the ILMN 610-Quad chips in 165 CAKUT patients and 3,060 controls. We developed an analytic pipeline for identification of rare pathogenic CNVs: absence in public databases and controls; disruption of genes; recurrence in multiple patients; prioritization based on linkage loci and gene expression.

**Results:** The analysis yielded 2,532 CNVs. Comparison with 3,060 controls and with public databases resulted in 319 rare genic CNVs. 38 patients had high-priority CNVs: 9 carried CNVs diagnostic of known syndromes, such as 17q12 (TCF2: renal cyst-diabetes syndrome; 4 cases), indicating that our approach is robust for identification of pathogenic CNVs; 29 carried novel high-priority CNVs. We validated 13 of these and 3 were *de novo*: chrs. 3q13-22 (novel), 13q12 (novel) and 17q12 (TCF2).

**Conclusions:** Our study suggests a major role of genic CNVs in CAKUT pathogenesis. Using a rigorous pipeline, we identified high-priority CNVs that we are testing for replication in an independent cohort of patients to identify underlying genes.

### Abstract# 580
**(O-74)**

**Complement Component C3 Aberrations in Patients with Atypical Haemolytic Uraemic Syndrome (aHUS)** E. Volokhina,[1] D. Westra,[1] M. HendriksFranssen,[2] E. van Loon,[2] M. Huigen,[2] P. Gros,[3] N. van de Kar,[1] L. van den Heuvel.[1,2,4] *[1]Department of Paediatric Nephrology, Radboud University Nijmegen Medical Centre, Nijmegen, Netherlands; [2]Department of Laboratory Medicine, Radboud University Nijmegen Medical Centre, Nijmegen, Netherlands; [3]Department of Crystal and Structural Chemistry, Utrecht University, Utrecht, Netherlands; [4]Department of Paediatrics, University Hospital Leuven, Leuven, Belgium.*

**Objectives:** Atypical haemolytic uraemic syndrome (aHUS) is associated with alterations in complement (regulatory) genes and with the presence of autoantibodies against complement factor H (CFH). Around half of the aHUS cases result in ESRD and aHUS etiology is of important prognostic value for renal transplantation. Here we performed mutational screening of the C3 gene encoding central complement component.

**Methods:** The C3 gene was analysed by means of PCR and DNA sequencing in 70 aHUS patients. The possible pathogenic effects of the missense alterations was assessed using structural data available for C3.

**Results:** Two missense and one splice site mutation were found in five aHUS patients. Furthermore, one strongly predisposing non-synonymous SNP was found in 12 patients and only in three healthy individuals. None of these changes were described before. Analysis of the available structural data indicates that the amino acids altered by the two missense mutations and a predisposing SNP are in the proximity of the C3b interface with CFH.

**Conclusions:** Novel aHUS predisposing alterations in C3 are described. In total, we encountered four potentially pathogenic alterations in 22.9% (16/70) of patients. Missense mutations and predisposing SNP might influence C3b interaction with CFH.

### Abstract# 581
**(O-75)**

**The Collaborative European Cohort of Primary Hyperoxalurias: Clinical and Genetic Characterization with Prediction of Outcome** C. van Woerden, G. Mandrile, S. Hulton, S. Fargue, B. Beck, J. Harambat. *AMC, Amsterdam, Netherlands; UNITO, Torino, Italy; UK, Cologne, Germany; BCH, Birmingham, United Kingdom; UCL, London, United Kingdom; CHU, Bordeaux, France.*

**Objectives:** Primary hyperoxalurias (PH) are characterized by excessive endogenous oxalate production and a heterogeneous phenotype with respect to end-stage renal disease (ESRD). We document the genetic spectrum and clinical outcome of PH in a large multicenter European study.

**Methods:** Patient data collected from eight European medical centers (retrospectively and prospectively), with documentation of genotype and clinical outcome. Participating countries included The Netherlands, Italy, Germany, United Kingdom, Spain, France, Poland and Sweden.

**Results:** We identified 579 patients (501 PH type 1, 53 PH type 2, 25 non type 1, non type 2). Genotype and outcome were known in 408 of them (median follow-up of 17.7; range 0.3 – 74.7 years). ESRD occurred in 36% at diagnosis. At last review 50.5% developed ESRD and 12% died. More than 80 *AGXT* mutations were identified, of which the most common (allele frequency) were p.Gly170Arg (25%), c.33dupC (10%), p.Ile244Thr (7%), p.Phe152Ile (3%). In homozygous p.Gly170Arg patients, 50% developed ESRD occurring at a median age of 34 years, compared to 14.6 years in those with other mutations ($p<0.001$).

**Conclusions:** The heterogeneous clinical presentation in PH1 is related to a wide mutational spectrum. Whereas progression to ESRD occurs in all genotypes, renal function in PH1 is preserved longest in patients carrying the common p.Gly170Arg mutation. Ongoing data collection as part of an international registry will clarify disease outcomes further.

### Abstract# 582
**(O-76)**

**Novel Insertion-Deletion Mutation in Uromodulin in a Large Kindred with Familial CKD and Nephrotic Range Proteinuria** R.A. Gbadegesin, M.P. Winn. *Pediatrics, Medicine and Center for Human Genetics, Duke University, Durham, NC, United States.*

**Objectives:** Focal and segmental glomerulosclerosis (FSGS) remains an important cause of nephrotic syndrome (NS) and chronic kidney disease (CKD). The objective of this study is to positionally cloned the gene mutated in a large kindred with familial CKD and proteinuria.

**Methods:** We identified a large family with eight affected individuals spanning three generations. We performed genome-wide linkage analysis (GWLA) using the Illumina Infinum II HumanLinkage-12 beadchip genotyping assay and fine mapping with informative microsatellites.

**Results:** Two of the affected individuals had 3g and 5g of proteinuria. Renal biopsy in four affected individuals showed foci of interstitial infiltrates and focal global glomerulosclerosis. GWLA and fine mapping yielded a multipoint parametric LOD score of 2.9 on chromosome 16p. Positional cloning of the genes within the locus yielded a novel in-frame insertion deletion mutation 278_289delins T**CTGCCCCGAAG>CCGCCTCCT** in exon 3 of *uromodulin.* The in-frame change leads to loss of a highly conserved cysteine residue in position 94. The mutation segregates in affected individuals in the family.
**Conclusions:** Phenotype previously associated with uromodulin defects include chronic interstitial nephritis, JFHS and glomerulocystic disease. This finding in combination with the recent GWAS reveal that uromodulin is a common disease locus in subjects with CKD and suggests that defective uromodulin may have wider deleterious effects in the kidney. In conclusion, a novel insertion-deletion mutation in uromodulin in a large family with CKD and proteinuria expands the clinical spectrum of disease associated with defective uromodulin.

## Abstract# 583
**(O-77)**

**Heritability of Gal-Deficient IgA1 in Pediatric IgA Nephropathy and Henoch-Schönlein Nephritis** K. Kiryluk,[1] Z. Moldoveanu,[2] J.T. Sanders,[3] H. Suzuki,[2] B.A. Julian,[2] J. Mestecky,[2] J. Novak,[2] A.G. Gharavi,[1] R.J. Wyatt.[3] *[1]Columbia Univ., New York, NY, United States; [2]Univ. of Alabama at Birmingham, Birmingham, AL, United States; [3]Univ. of Tennessee Health Sciences Center, Memphis, TN, United States.*
**Objectives:** Serum galactose-deficient IgA1 (Gd-IgA1) is an inherited risk factor for adult IgA nephropathy (IgAN). The goal of this study is to determine the heritability of this trait in pediatric IgAN and Henoch-Schönlein Nephritis (HSPN).
**Methods:** We obtained serum from 34 Caucasian families (20 cases of HSPN, 14 cases of IgAN, and 54 of their relatives), as well as from 51 age- and ethnicity-matched pediatric controls and 141 healthy adult controls. Serum Gd-IgA1 levels were quantified using the HAA-lectin based ELISA assay. Polygenic modeling and heritability estimates were performed using a variance components method (SOLAR).
**Results:** Children with either IgAN or HSPN had significantly higher Gd-IgA1 levels compared to pediatric controls (p=1x10-4 and p=8x10-7, respectively). Serum levels of Gd-IgA1 were also elevated in a large fraction of the relatives of pediatric IgAN and HSPN compared to adult controls (p=5x10-4 and p=0.002, respectively). The unilineal transmission of the trait was observed in 75% of families, bilineal transmission in 5%, and sporadic occurrence in 20% of cases. The age-, gender-, BMI-, and household-adjusted heritability of Gd-IgA1 was estimated at 75.9% (p=0.02) in pediatric IgAN and at 68.9% (p=0.008) in HSPN. The household effects were responsible for 5%, while age for 2.8% of the trait's variance.
**Conclusions:** Serum Gd-IgA1 levels are highly inherited in both, pediatric IgAN and HSPN, providing support for a common pathogenic link between these disorders.

## Abstract# 584
**(O-78)**

**Evidence for a Chromosome 16p11.2 Microdeletion Associated with a Syndrome of Congenital Anomalies of the Kidney and Urinary Tract and Hirschsprung Disease** M. Sampson, C. Coughlin, M. Kevin, E. Zackai, P. Kaplan, N. Spinner, C. Lawrence. *Pediatric Nephrology, Children's Hospital of Philadelphia, Philadelphia, PA, United States.*
**Objectives:** We investigated a recurrent microdeletion at 16p11.2 in three children with a spectrum of clinical anomalies consisting of Congenital Anomalies of the Kidney and Urinary Tract (CAKUT) and Hirschsprung disease (HSCR).
**Methods:** Single nucleotide polymorphism (SNP) analysis performed using the Illumina HumanQuad610 BeadChip on DNA from peripheral blood on the index patient demonstrated a microdeletion at 16p11.2. Two additional patients with similar CAKUT-HSCR phenotypes were identified with a recurrent microdeletion at 16p11.2.
**Results:** Case 1 was a 2 day old male with left renal agenesis, Grade IV vesicoureteral reflux, and HSCR. A 217-kb microdeletion was identified (UCSC Human Genome Version Hg18, NCBI Build 36, Chr16: 28733550-28950951).
Case 2 was a 17 year old male with left renal agenesis, Stage II CKD, chronic constipation, seizure disorder, and developmental delay.
Case 3 was a 12 year old male with HSCR, Beals' Sydrome (as a result of a pathogenic Fibrillin-2 mutation), and normal kidneys.
Both Case 2 and Case 3 had a recurrent 1.69Mb microdeletion; (Chr16:28396413-30085308).
The smallest region of overlap among the three patients contained 9 described genes (OMIM). We identified one gene, *SH2B1*, as having biologic plausibility in the molecular pathogenesis of the CAKUT-HSCR phenotype, based on its role in the RET-GDNF signaling pathway.
**Conclusions:** Microdeletion of 16p11.2 appears to be associated with a CAKUT-HSCR syndrome. Deleted within this region is *SH2B1*, a candidate gene for further investigation.

## Abstract# 585
**(O-79)**

**mRNA Splicing Defects Caused by *PKD1* and *PKD2* Exonic Mutations in Autosomal Dominant Polycystic Kidney Disease** F.J. Gonzalez-Paredes,[1] E. Ramos-Trujillo,[1] V.M. Garcia-Nieto,[2] F. Claverie-Martin.[1] *[1]Unidad de Investigacion, Hospital Universitario N. S. de Candelaria, Santa Cruz de Tenerife, Spain; [2]Nefrologia Pediatrica, Hospital Universitario N. S. de Candelaria, Santa Cruz de Tenerife, Spain.*
**Objectives:** Autosomal dominant polycystic kidney disease (ADPKD) is the most common inherited kidney disorder. ADPKD is caused by mutations either in *PKD1* or *PKD2* genes. Recent studies have shown that some mutations classified as missense and synonymous can induce pre-mRNA splicing defects. Our goal is to determine the functional consequences of this kind of *PKD1* and *PKD2* mutations in pre-RNA splicing.
**Methods:** *PKD1* and *PKD2* minigenes were constructed with exons 2-3, 11, 23, 24-25-26 or 37-38-39, and with exons 4, 5 or 13, respectively. Mutations were introduced by site-directed mutagenesis. Minigenes were transfected into HEK293T cells and mRNA was analysed by RT-PCR.
**Results:** Five missense and two synonymous *PKD1* mutations and six *PKD2* missense mutations were analysed. G109G creates a donor splice site, leading to an aberrant mRNA lacking the 3' end of exon 3. R3753R activates a cryptic donor splice site and generates an altered mRNA missing the 3´ end of exon 39. Analysis of *PKD2* missense mutations R306Q, R322W, R325Q and A356P showed increased incorporation of exon 4 in the mRNA. The rest of mutations showed no effect in pre-mRNA splicing.
**Conclusions:** *PKD1* synonymous variants G109G and R3753R act as splicing mutations. These represent the first *PKD1* synonymous mutations that affect pre-mRNA splicing. *PKD2* missense mutations seem to alter the regulation of the mRNA splicing process. Our results emphasize the importance of performing mRNA analysis when evaluating the effect of exonic mutations.

## Abstract# 586
**(O-80)**

**Fine Mapping of Chr.12p11-q13 VUR Locus with Interval-Specific Association** P. Weng,[1] S. Sanna-Cherchi,[2] H. Snyder,[2] N. Kacak,[2] R. Schlussel,[2] L. Clark,[2] F. Scolari,[3] G.M. Ghiggeri,[4] K. Glassberg,[2] A. Gharavi.[2] *[1]Mt Sinai School of Med, NY, NY, United States; [2]Columbia Univ, NY, NY, United States; [3]Spedali Civili, Brescia, Italy; [4]Gaslini Institute, Genoa, Italy.*
**Objectives:** Primary vesicoureteral reflux (pVUR) is a major cause of pediatric ESRD. We localized a gene for pVUR on chr. 12p11-q13 under a recessive model. Evidence for linkage was strongest among the Hasidic Jewish, which has a founder effect. To narrow down the linkage interval we performed a case control association study in our Hasidic Jewish cohort.
**Methods:** Cases were ascertained with a positive VCUG. Controls were of the same ethnicity and did not have a family history of any urologic/kidney disease. We genotyped 18 familial Jewish cases and 437 controls with the Illumina 660W arrays. 3,489 SNPs in the LOD-1 interval (29.5 Mb) were used for the association study. Standard QC filters were performed.
**Results:** Our analysis with PLINK revealed a genomic inflation factor $\lambda$ =1.03, indicating almost no population stratification. Within the LOD-1 interval on chr. 12 we found 9 suggestive SNPs (p-value<$10^{-4}$). The most promising signal was an intronic SNP in a gene highly expressed in the metanephric mesenchyme and ureteric bud of the developing kidney (p-value=$2.62 \times 10^{-5}$, OR= 3.9). This association is significant after Bonferroni correction.
**Conclusions:** pVUR is a complex disorder with a recessive gene on chr. 12p11-q13. Initial association results in familial Jewish cases and controls indicate several potential candidate genes for pVUR in this interval. We are now performing a replication study in Jewish and non-Jewish cohorts to identify the gene underlying our locus.

## Abstract# 587
**Familial Nephrotic Syndrome in Children in Port Harcourt, Nigeria** I.C. Anochie,[1] F.U. Eke.[1] *[1]Paediatrics, University of Port Harcourt Teaching Hospital, Port Harcourt, Rivers State, Nigeria; [2]Paediatrics, University of Port Harcourt Teaching Hospital, Port Harcourt, Rivers State, Nigeria.*
**Objectives:** Nephrotic syndrome (NS) is the commonest renal disease seen in children in Nigeria. We have reported sporadic cases, with minimal change being the commonest pathology seen. Familial NS has rarely been reported in Nigeria.
**Methods:** We report a 4year old boy and his 16year old sister from a non-consanguineous marriage who presented with features of nephrotic syndrome. Renal biopsies done in both showed Focal segmental glomerulosclerosis.
**Results:** The diagnosis of Familial Nephrotic syndrome was therefore made and they were placed on prednisolone and cyclophosphamide. A genetic study was not done due to unavailability of laboratory facilities.

**Conclusions:** This is the first report of familial nephrotic syndrome in Nigeria, and genetic study would be important to identify either the less severe autosomal dominant or a more severe recessive form.
Keywords- Familial Nephrotic syndrome, focal segmental glomerulosclerosis, Children, Nigeria

## Abstract# 588

**Micosatellite Analysis of Steroid Resistant Nephrotic Syndrome in Indian and Black Children in South Africa** K. Asharam, R. Bhimma, M. Adhikari. *Deapartment of Paediatrics and Child Health, University of KwaZulu-Natal, Durban, KwaZulu-Natal, South Africa.*
**Objectives:** 1. To determine the existence of mutations in the NPHS2 gene in Indian and Black children in South Africa with steroid resistant nephrotic syndrome (SRNS).
2. To determine differences in expression of the NPHS2 gene between Indian and Black children with SRNS and steroid sensitive nephrotic syndrome (SSNS).
**Methods:** 60 children with idiopathic SRNS were recruited into the study during the period January 2005 to June 2008. Blood and kidney biopsy samples collected from patients were subjected to microsatellite analysis to scan for mutations in the 1q25-1q35 region and the raw data was analysed using the Fragment Manager software. Microsatellite analysis was based on microsatellite instability (MSI) and allelic imbalance and/or loss of heterozygosity (AI/LOH). Mutations in SRNS were compared to 20 controls with steroid sensitive NS.
**Results:** 55% were male and 45% were female. 37% were Indian and 63% were black. 75% showed focal segmental glomerulosclerosis on histology, (7%) minimal change disease and 18% had other histological forms of NS. 25 cases showed mutations on microsatellite analysis: 32% had MSI only, 56% AI/LOH only, and 12% had both. No significant associations for sex, age or race were detected using all markers. No mutations were detected in the controls with steroid sensitive NS. Comparison of SRNS patients to SSNS patients showed significant increase in mutations in the NPHS2 gene using microsatellite analysis in SRNS patients.
**Conclusions:** Mutations of the NPHS2 gene may predispose to the development of SRNS on microsatellite analysis. Other candidate genes not tested for may also play a role.

## Abstract# 589

**Detection of Genomic Damage in Lymphocytes of Children with Chronic Kidney Disease by Comet Assay** B. Aykanat,[1] G. Çakmak,[1] K. Fidan,[3] K. Gülleroglu,[4] A. Sepici,[5] U. Bayrakçi,[4] N. Buyan,[3] E. Baskin,[4] H. Karakayali,[2] M. Haberal,[2] S. Burgaz.[1] *[1]Toxicology, Gazi Univ, Ankara, Turkey; [2]General Surgery, Baskent Univ, Ankara, Turkey; [3]Pediatric Nephrology, Gazi Univ, Ankara, Turkey; [4]Pediatric Nephrology, Baskent Univ, Ankara, Turkey; [5]Biochemistry, Gazi Univ, Ankara, Turkey.*
**Objectives:** Chronic kidney disease (CKD) is a serious illness that causes severe and irreversible reduction in kidney function. One consequence of CKD is an elevated cancer risk. However, the underlying mechanism is unclear. There is no genomic data which has been demonstrated by some cytogenetic markers in children with CKD.
**Methods:** In this study, DNA/oxidative damage in lymphocytes from 17 children in pre-dialysis stage (PreD), 15 children on regular hemodialysis (HD), and 17 renal transplanted (TX) children has been compared to 20 healthy children by the strand breaks and endonuclease III (Endo III), formamidopyrimidine glycosylase (FPG) sensitive sites in Comet Assay. Blood chemistry parameters, TAC, CRP, IL-6 and homocysteine have also been measured.
**Results:** Our results reveal that strand breaks (tail intensity) [median(IQR)] are significantly increased in overall CKD patients [5.23(2.38)] and individual Tx, HD, PreD groups [6.53(3.10), 4.67(2.16), 5.08(1.51) respectively] vs. control group [2.29(1.88)] ($p<0.001$).Significant increases have only been found in the FPG sites for Tx and PreD groups vs.HD and control groups($p<0.05$).
**Conclusions:** In conclusion, CKD is related with higher basal DNA damage in overall patients and its sub-groups against the controls.However,data on Endo III and FPG sensitive sites and IL-6 partly explain for the oxidative damage in the patients.

## Abstract# 590

**Phenylketonuria as a Metabolic Disease – Connection with Urine and Plasma Sample Diagnostics** A. Bajraktarevic,[1] A. Hadzimuratovic,[2] A. Hadzimuratovic,[3] J. Ceman Saric,[4] I. Suljevic.[4] *[1]Pediatrics Department, Public Health Institution of Canton Sarajevo, Sarajevo, Bosnia, Bosnia and Herzegowina; [2]Pediatrics Nephrology Department, Pediatrics Clinic Sarajevo, Sarajevo, Bosnia, Bosnia and Herzegowina; [3]Children's Surgery, Clinical Medical Center Sarajevo, Sarajevo, Bosnia, Bosnia and Herzegowina; [4]Biochemical Department, Clinical Medical Center Sarajevo, Sarajevo, Bosnia, Bosnia and Herzegowina; [5]Biochemical Department, Clinical Medical Center Sarajevo, Sarajevo, Bosnia, Bosnia and Herzegowina.*
**Objectives:** Phenylketonuria is an inherited metabolic disorder. Newborns with phenylketonuria initially don't have any symptoms.
**Methods:** Phenylketonuria is diagnosed by blood test, as a routine screening tests given to a newborn. In Bosnia and Herzegovina, the heel prick test-Guthrie test, is carried out on all newborn babies shortly after birth. The level of phenylalanine in the plasma and phenylalanine metabolites in freshly collected urine samples were tested.
**Results:** Classic PKU and the other causes of hyperphenylalaninemia affect about one of every 19,000 to 20,000 Bosnian births. Significant variations in phenylalanine transport parameters in untreated, normal intelligent patients indicated that blood-brain barrier transport or intracerebral phenylalanine consumption are causative factors for the individual vulnerability to phenylketonuria.
**Conclusions:** The enzyme phenylalanine hydroxylase normally converts the amino acid phenylalanine into the amino acid tyrosine. The main phenylketonuria treatment is a strict diet with very limited intake of phenylalanine, which is mostly found in protein-rich foods. These pathologic compounds in phenylketonuria are usually not detected in normal urine.
**DISCLOSURE:** *Bajraktarevic, A.*: Consultant, No interest. *Hadzimuratovic, A.*: Consultant, No interest.

## Abstract# 591

**Urinary UPIb mRNA Levels in Vesicoureteral Reflux and Urinary Tract Infection** I. Kaplan Bulut, S. Mir, A. Berdeli, B. Sozeri. *Paediatric Nephrology, Ege University Medical Faculty, Izmir, Bornova, Turkey; Paediatric Nephrology, Ege University Medical Faculty, Izmir, Bornova, Turkey; Moleculer Medicine, Ege University Medical Faculty, Izmir, Bornova, Turkey; Paediatric Nephrology, Ege University Medical Faculty, Izmir, Bornova, Turkey.*
**Objectives:** Uroplakin is an integral nembran protein and found in the structure of urotelium. Recently, there have been a number of studies describing an association between the UP and Escherichia coli related UTI and VUR.
We investigated the urine UPIb mRNA levels of the patients of whom are suffering from the UTI for the first episode or recurrent UTI or UTI with VUR.
**Methods:** Eighty-nine patients who have the diagnosis of UTI and 26 healthy children were enrolled to the study. Twenty-eight of the patients were diagnosed as UTI for the first time, 31 patients were diagnosed as recurrent UTI, and 30 patients were diagnosed as UTI with VUR. Quantitative real-time RT-PCR for UPKIb mRNA was performed on exfoliated urothelial cells from patients. Housekeeping gene GPADH was used.
**Results:** The mean value of UPIb were found 33.88±4.16 in the patient group who were diagnosed as UTI for the first time, 32.86±4.5 in the recurrent UTI group, 33.64±3.7 were in the patient group diagnosed as UTI with VUR, and 35.52±2.2 in the control group.
UPIb mRNA was found to be lower in VUR+UTI and recurrent UTI than in control exfoliated urinary cells (mean±SE: 33.64±3.7, 32.86±4.5 and 35.52±2.2 copies, respectively, $P < 0.05$).
**Conclusions:** Urine UPIb levels may be useful in predicting the risk of recurrence UTI in cases diagnosed as UTI for the first time. These levels may be considered as a noninvasive screening test for the patients.

## Abstract# 592

**Secondary Inherited NDI: A Diagnostic Pitfall** D. Bockenhauer,[1] W. van't Hoff,[1] A. Lehnhardt,[2] M. Subtirelu,[3] F. Hildebrandt,[4] D.G. Bichet.[5] *[1]Nephrology, Great Ormond Street Hospital for Children, London, United Kingdom; [2]University Childrens Hospital, Hamburg, Germany; [3]T.C. Thompson Children's Hospital, UT College of Medicine Chattanooga, Chattanooga, TN, United States; [4]Pediatrics and Human Genetics, University of Michigan, Ann Arbor, MI, United States; [5]Medicine and Physiology, Université de Montréal, Montreal, QC, Canada.*
**Objectives:** Nephrogenic diabetes insipidus (NDI) is a serious condition, which unrecognised can lead to permanent brain damage. Primary inherited NDI is due to mutations in either AVPR2 or AQP2, encoding the vasopressin type 2 receptor

and aquaporin 2, respectively. Yet NDI can also occur as a secondary complication and we aimed to define NDI in selected inherited renal diseases.
**Methods:** Review of clinical features and analysis of AVPR2 and AQP2 genes of four patients with NDI and molecularly proven diagnosis of nephropathic cystinosis, Bartter syndrome, nephronophthisis and apparent mineralocorticoid excess (AME), respectively.
**Results:** All patients excreted hypotonic urine during hypernatremic dehydration and had no further increase in urine osmolality after DDAVP, consistent with NDI, yet no mutation in AVPR2 and AQP2 genes could be identified. In two patients the diagnosis of NDI was missed, leading to repeated hypernatremia. In the other 2 patients, NDI was mistakenly assumed to be the primary diagnosis, delaying identification of the underlying disorder. In one patient, NDI was reversible with treatment of her AME.
**Conclusions:** The recognition of this potential complication is important as it has direct implications for the clinical management. The occurrence of NDI in these conditions provides clues for the etiology of aquaporin deficiency.

## Abstract# 593

**A Novel Mutation of the COL4A5 Gene in a Family with Alport Syndrome – (Case Report)** A. Bosakova,[1] T. Sulakova,[1] P. Plevova,[2] J. Dvorackova.[3] *[1]Dpt. of Pediatrics, University Hospital, Ostrava, Czech Republic; [2]Dpt. of Medical Genetics, University Hospital, Ostrava, Czech Republic; [3]Dpt. of Pathology, University Hospital, Ostrava, Czech Republic.*
**Objectives:** Alport syndrome is a clinically and genetically heterogeneous nephropathy. The majority of cases are transmitted as an X-linked condition due to COL4A5 mutations.
**Methods: Case report:** We report on an 11-year-old girl who has been followed since the age of 4-years for repeated urinary tract infections (cystitis), microscopic hematuria and later also proteinuria (< 0.5g/day) and hypertension. The ultrasound examination and the audiogram have been normal. She was treated with ramipril with a good effect on hypertension (ABPM). Renal biopsy revealed changes typical for Alport syndrome.
Father of the girl has been suffering from perceptive hearing loss and glomerulonephritis since childhood. He underwent two renal transplantations. There are also 3 relatives with a similar symptomatology in the family. Molecular genetic analysis of the COL4A5 gene was performed and revealed a germline nonsense mutation c.372-373del.CA(p.C124X) in exon 6 of the gene.
**Results:** This mutation results in the synthesis of a shortened protein (123 amino-acid versus normal 1691 amino-acid chain). Based on the biopsy findings and molecular genetic analysis the diagnosis of X-linked Alport syndrome has been proved.
**Conclusions:** The mutation found in the observed family is a new one and has not been previously described. The mutation has deleterious effect on the protein level. It may be considered to be the cause of Alport syndrome due to the segregation of the mutation with the disease in the presented family.

## Abstract# 594

**Genetic Heterogeneity of Familial Congenital Anomalies of the Kidney and Urinary Tract** K. Burgess,[1] S. Sanna-Cherchi,[1] P.L. Weng,[1] G. Caridi,[2] M. Bodria,[2] S. Testa,[3] L Kerecuk,[4] G. Ardissino,[3] A.S. Woolf,[4] F. Scolari,[5] G.M. Ghiggeri,[2] A. Gharavi.[1] *[1]Columbia Univ, NY, NY, United States; [2]G.Gaslini Inst, Genoa, Italy; [3]Osp Maggiore, Milan, Italy; [4]Inst of Child Health, London, United Kingdom; [5]Hosp, Montichiari, Italy.*
**Objectives:** Congenital malformations of the kidney and urinary tract (CAKUT) are the leading cause of pediatric ESRD. Using positional cloning approaches we identified significant loci for CAKUT on chrs. 1p32-33 and 12p11-q13. We report a genome-wide scan for linkage on 16 CAKUT families.
**Methods:** Genome-wide genotyping was performed using Affymetrix 10K 2.0 arrays. Parametric analysis of linkage (disease gene frequency 0.001, penetrance 0.8, phenocopy rate 0.001) was performed using Allegro 2.0. To avoid misspecification of the disease model, we performed non-parametric, model-free analysis.
**Results:** The 16 families included 69 affected and 35 unaffected individuals. Multipoint analysis of linkage resulted in 3 novel suggestive loci: 3p13-q11 (HLOD=1.85; alpha=0.43; NPL=1.3), 11q23-24 (HLOD=1.7; alpha=0.43; NPL=3; $p=2x10^{-3}$) and 18q12-21 (HLOD=1.9; alpha=0.50; NPL=1.7; p=0.05). These data indicate substantial genetic heterogeneity for familial CAKUT.
**Conclusions:** Our genome-wide scan for linkage identified 3 novel suggestive signals on chrs. 3p13-q11, 11q23-24 and 18q12-21. High genetic heterogeneity complicates positional cloning studies, requiring alternative strategies to identify disease-causing genes in each family. The use of novel high-throughput technologies, such as high-resolution search for copy number variations and massive parallel sequencing, can result in successful identification of novel genes when combined with traditional linkage approaches.

## Abstract# 595

**Macrophage Migration Inhibitory Factor-173 Polymorphism in Turkish Children with Nephrotic Syndrome: A Preliminary Study** M. Buyukcelik,[1] S. Pehlivan,[2] B. Demircioglu Kilic,[1] T. Sever,[2] S. Balci,[2] A. Balat.[1] *[1]Pediatric Nephrology, Gaziantep University, Gaziantep, Turkey; [2]Medical Biology and Genetics, Gaziantep University, Gaziantep, Turkey.*
**Objectives:** Macrophage migration inhibitory factor (MIF) is a proinflammatory cytokine and is produced by mesangial cells, glomerular epithelial cells and tubular epithelial cells. Recent studies have shown that MIF may play a pathogenic role in the kidney diseases such as idiopathic nephrotic syndrome (INS). The MIF promoter contains a single nucleotide G to C polymorphism at position -173. It has been shown that MIF-173 C allele is associated with higher MIF production and linked to susceptibility to inflammatory diseases.
**Methods:** In this study we genotyped the MIF-173 polymorphism in 108 children with INS and 122 controls.
**Results:** Differently from the other studies in literature, MIF C allele was more frequent in controls (23%) compared with INS (12. 5%) (p=0.003). Frequency of carriers of the MIF-173 G allele was also similar in both groups (87.5% vs 77%, respectively). Frequencies of GG and GC genotypes of MIF- 173 were not statistically different in INS and controls (75% vs 58.1%, and 25% vs 37.7%, respectively).
**Conclusions:** We suggest that MIF-173 polymorphism may not be associated with INS in children, but further studies in a larger population are needed to confirm the results.

## Abstract# 596

**Polymorphism of the NR3C1, a Glucocorticoid Receptor Gene, in Turkish Children with Steroid-Sensitive and Resistant Nephrotic Syndrome** M. Gundogdu,[1] M. Buyukcelik,[1] S. Pehlivan,[2] N. Aydin,[3] T. Sever,[2] B. Demircioglu Kilic,[1] A. Balat.[1] *[1]Pediatric Nephrology, Gaziantep University, Gaziantep, Turkey; [2]Medical Biology and Genetics, Gaziantep University, Gaziantep, Turkey; [3]Public Health and Biostatistic, Gaziantep University, Gaziantep, Turkey.*
**Objectives:** Idiopathic nephrotic syndrome is one of the most commonly primary glomerular diseases in children, and glucocorticoid receptors may have an important role in responsiveness to steroid therapy.
**Methods:** We investigated Bcl1 nucleotide polymorphism of the glucocorticoid receptor gene (NR3C1 gene) in 84 children with nephrotic syndrome and 66 healthy controls using polymerase chain reaction-restriction fragment length polymorphism, and analyzed the correlation between genotypes and clinicopathological features of the patients.
**Results:** Forty nine children (58.3 %) were initial steroid responder and thirty five (41.7 %) were steroid resistant. C/C and G/C genotype frequencies of Bcl1 were similar in all groups (steroid-responsive, steroid resistant and control group), while G/G genotype frequencies were similar in steroid-responsive and control groups. We could not find G/G genotype in steroid-resistant group.
**Conclusions:** These data suggested that C/C and G/C genotype of Bcl1 in the NR3C1 may not affect steroid-responsiveness in Turkish children with NS. On the other hand, absence of the GG genotype and G allele of Bcl1 may play a role in the steroid- resistance in these children. However, further studies in a larger population are needed to test this hypothesis.

## Abstract# 597

**X-Linked Nephrogenic DI in Female Siblings Due to Skewed X-Chromosome Inactivation** V. Chadha. *Pediatric Nephrology, VCU Medical Center, Richmond, VA, United States.*
**Objectives:** X-linked nephrogenic DI (X-NDI) is a rare disease caused by mutations in the *AVPR2* gene. These patients are overwhelmingly males. Symptomatic presentation in females is rare and can occur due to skewed X chromosome inactivation.
**Methods:** Three siblings (2 girls 3 yrs and 2 yrs, and a boy 4 mo) presented with severe polyuria and polydipsia. DDAVP challenge test confirmed the diagnosis of NDI. Their mother was asymptomatic and all had different fathers (also asymptomatic) thus making the diagnosis of autosomal recessive or dominant form of NDI due to aquaporin 2 (AQP2) gene mutations unlikely.
**Results:** Genetic testing was initially performed on the eldest sibling. There were no mutations in the AQP2 gene. However, she was heterozygous positive for a complex allele in the AVPR2 gene with three mutations (R64Q, A147V, and A163P). Of these mutations, A163P has been associated with X-NDI. Genetic testing of mother and other two siblings revealed the presence of identical mutations. The only other male family members (maternal grandfather and maternal cousin) were asymptomatic and genetic testing in both of them was negative for mutations in the AVPR2 gene. Over the last 2 years, the severity of DI has decreased in both sisters (urine osmolality improved from < 100 to slightly over 200 mOsm/kg).

**Conclusions:** *Conclusion:* Although rare in females, X-NDI should be included in the differential diagnosis of symptomatic females presenting with features of DI in early infancy. Severe phenotype in females can result from extremely skewed inactivation of X chromosome. X chromosome inactivation occurs early in embryogenesis and is irreversible in descendents of that cell. The reason for decrease in severity in the two sisters is thus unclear.

## Abstract# 598

**Linkage and Association Study of Neurotrophins and Their Receptors as Novel Susceptibility Genes for Childhood IgA Nephropathy** W.-H. Hahn, J.-S. Suh, B.-S. Cho. *Pediatrics, East West Kidney Diseases Research Institute, School of Medicine, Kyung Hee University, Seoul, Korea.*

**Objectives:** Neurotrophins (NTs) and their receptors (NTRs) are known to be important for pathogenesis of various inflammatory diseases that occur in not only neuronal but also non-neuronal tissues, including kidney. Thus, we performed genetic association study between childhood IgA nephropathy (IgAN) and NTs and NTRs.

**Methods:** We genotyped and analyzed single nucleotide polymorphisms (SNPs) of genes encoding NTs (nerve growth factor (NGF) and brain-derived neurotrophic factor (BDNF)) and NTRs (nerve growth factor receptor (NGFR) and neurotrophic tyrosine kinase receptor 1-3 (NTRK1-3)) in 197 IgAN patients and 289 healthy controls.

**Results:** The genotyping data of IgAN patients and healthy control subjects revealed significant association between NGF SNP rs11102930 and presence of IgAN. Patient subgroup analysis revealed that development of proteinuria (> 4 and ≤ 4 mg/m2/hr) was associated with rs1187321 and rs1187323 of NTRK2, and that presence of nephrotic range proteinuria (> 40 mg/m2/hr) was associated with rs6334 of NTRK1, and rs11030104, rs7103411, rs7103873, and rs6484320 of BDNF. Significant differences were observed in podocyte foot process effacement for rs1187321 and rs1187323 of NTRK2. Furthermore, some SNP frequencies were significantly different in patient subgroups with either pathologically mild or advanced disease, specifically in rs6334 of NTRK1.

**Conclusions:** Our results suggest that SNPs of NTs and NTRs are associated with susceptibility, pathological advancement, podocyte foot process effacement, and development of proteinuria in childhood IgAN.

## Abstract# 599

**Polymorphisms of CXCL8 and Its Receptor CXCR2 Contribute to the Development and Progression of Childhood IgA Nephropathy (IgAN)** B.-S. Cho, J.-S. Suh, W.-H. Hahn. *Department of Pediatrics, School of Medicine, Kyung Hee University, Seoul, Korea.*

**Objectives:** Many studies have suggested that CXCL8 and CXCR2 play an important role in the pathogenesis of several types of renal diseases. However, there is no prior study on the association between polymorphisms of these genes and IgA nephropathy (IgAN), especially in children.

**Methods:** The present study was conducted to investigate the association between three single nucleotide polymorphisms (SNPs) of the CXCL8 gene and two SNPs of the CXCR2 gene and childhood IgAN.

**Results:** Genotyping of 192 patients with childhood IgAN and 397 controls showed significant differences in the allele frequencies of the CXCL8 gene with rs2227306 (dominant, *P*=0.019; overdominant, P=0.009), rs2227543 (dominant, *P*=0.01; overdominant, *P*=0.0057), and rs4073 (codominant, *P*=0.034; dominant, *P*=0.011; overdominant, *P*=0.022). In addition, two SNP frequencies of the CXCR2 gene (rs4674257 and rs4674259) significantly differed between the patients with pathologically mild and patients with advanced disease. Furthermore, five SNPs of the CXCL8 and CXCR2 genes significantly differed in the patients with infiltration of inflammatory cells on the renal biopsy samples.

**Conclusions:** The results of this study suggest that polymorphisms of CXCL8 are associated with increased susceptibility to IgAN, and polymorphisms of CXCR2 with the pathological progression of childhood IgAN. These genetic variations might provide insight into novel individualized antichemokine regimens for treatment.

## Abstract# 600

**Neuroimaging Findings of Children with Autosomal Dominant Polycystic Kidney Disease** J.F.S. Crocker, A.O. Skotnicki, P.A. O'Regan, P.D. Acott. *Pediatrics, Dalhousie University and IWK Health Center, Halifax, NS, Canada.*

**Objectives:** This study evaluates the neuroimaging findings of a subset of ADPKD children presenting with headaches and/or family history of subarachnoid hemorrhage (SAH) in first-degree relatives.

**Methods:** We follow 63 children (age 1 – 20 years; 25% < 1 year) with ADPKD. Twenty (31.7%), age 3-20 years, who presented with headaches or a family history of SAH had central nervous system (CNS) investigation.

**Results:** Twenty ADPKD children who were imaged with CAT (n = 3) or MRI angiography (n = 17) had no cerebral vascular aneurysms. Five (25%) children (age 16-18 years) showed CNS abnormalities including areas of white matter demyelination (n = 2); cerebral/cerebellar atrophy and partial atrophy of posterior corpus callosum (n = 1); a 3 mm periventricular cyst (n = 1); and CNS lesions typical of known spina bifida with VP shunt placement (n=1).

**Conclusions:** This database documents neuroimaging anomalies before adulthood in 4 children (exclusive of expected findings of VP shunted child), 2 of which had demyelinative changes, compared to an incidence of approximately 3% in non-ADPKD children with headaches. These findings are likely developmental and are not located in a vascular pattern. There is a need for longer studies and an understanding of the relevance of these to the patients' complaints. ADPKD during early development has abnormal cilary function and primary cilia are critical to neural stem cell development as demonstrated by mice with Kif3A or Smo protein anomalies, which demonstrate failure of radial astrocytes responsible for adult neurogenesis. CNS neuroimaging findings may predict a cohort of ADPKD children at higher risk of developing cerebral aneurysms.

## Abstract# 601

**Clinical and Genetic Features of X-Linked Alport Syndrome in 202 Chinese Families** D. Zhao, J. Ding, F. Wang, W.H. Zhang. *Department of Pediatrics, Peking University First Hospital, Beijing, Beijing, China.*

**Objectives:** We aimed to elucidate featuress of phenotype and genotype in Chinese patients with XLAS.

**Methods:** The study included 202 unrelated patients with XLAS. Clinical characteristics were collected. COL4A5 gene was analyzed by RT-PCR, PCR and direct sequencing, SSCP, and southern blot.

**Results:** Nearly all patients ( 99%) had microscopic hematuria. Proteinuria was found in 91.7% male patients and in 76.2% female patients. About 42% male patients reached ESRD before 30 years old, and 24.7% female patients reached ESRD before 40 years old. Hearing loss occurred in 58.8% male patients and in 14.7% female patients. Ocular lesions were detected in 29.4% male patients and in 18.2% female patients. Thicking of the GBM was observed in 67.6% patients with a median age of 10.5 years. Abnormal staining of a5 (IV) was found in GBM of 71.1% patients, and in epidermal basement membranein of 86.9% patients. COL4A5 gene was analyzed in 111 families with a detection rate of 91%. The rate was 10.8% in large rearrangements, and 41.6% in missense.

**Conclusions:** XLAS was a high-penetrance disease in Chinese patients. Proteinuria occurred early and was severe. Chinese female patients seemed to have more severe renal damage. Disaccordance staining of a5 (IV) in GBM and epidermal basement membrane in male patients were first revealed. Analysis of COL4A5 cDNA fragments had significantly improved the mutation detected rate. Missense was the predominant mutation type in Chinese patients. Splice site mutation was more common in Chinese patients with variable splicing effects. Mutation located at exon 41A was first detected. No significant genotype-phenotype correlations revealed in this study.

## Abstract# 602

**FISH Analysis of TCF2 Microdeletion** S. Bourthoumieu,[1] V. Guigonis,[2] C. Bellané-Chantelot,[3] P. Brosset,[2] F. Esclaire,[1] C. Laroche,[2] F. Terro,[1] C. Yardin.[1] *[1]Cytogenetics, CHU, Limoges, France; [2]Pediatrics, CHU, Limoges, France; [3]Molecular Biology, Hôpital Pitié Salpétrière. AP-HP, Paris, France.*

**Objectives:** Hepatocyte nuclear factor-1 beta, coded by TCF2 gene plays an important role in the kidney, liver, gut and pancreatic development. Studies have shown that TCF2 abnormalities are associated with MODY 5 diabetes in adults and predominant kidney abnormalities in children. Studies have shown that these abnormalities could be secondary to point mutations or large deletions (1.5 Mb) of TCF2 gene. This latter abnormality is more frequent in children.These abnormalities are studied by molecular biology: QMPSF and direct sequencing. Our study aim was to demonstrate the presence of these deletions using FISH (Fluorescence in situ Hybridisation) technique.

**Results:** Two clones RP11-115K3 and RP11-697E22 (BAC-PAC chori), located on TCF2 gene (17q12) – and validated by QMPSF®-, allowed to produce probes FISH in order to study deletion of this region. Results of FISH analysis of deleted patients (previous QMPSF® results) confirmed the abnormality and allowed us to observe TCF2 deletion on metaphasic and interphasic cells.

**Conclusions:** These results confirm that TCF2 deletion could be detected using FISH method on interphasic cells. This analysis can be performed on a buccal cells smear which is more easy to obtain than a blood sample. Moreover, FISH on interphasic cells is a rapid method allowing the results to be obtained within 24 hours. This approach could as well be proposed antenatally when implications of such an antenatal diagnosis are determined. Nevertheless, if FISH results are normal and the phenotype suggestive of TCF2 disorder, point mutations have to be searched for using direct sequencing.

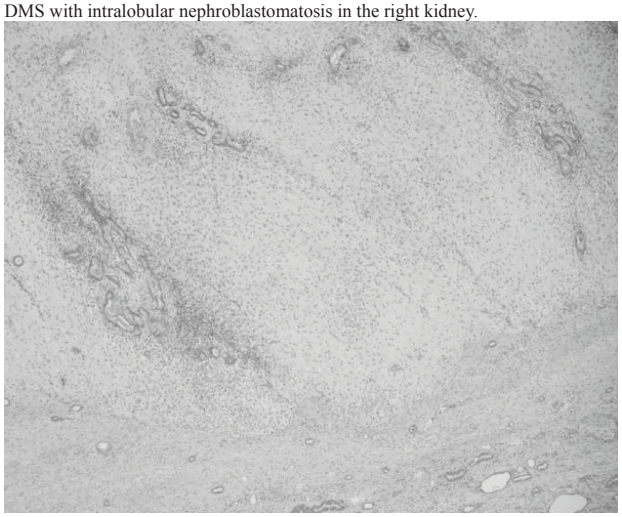
## Abstract# 603

**Homozygous Mutation of WT1 in a Wilms Tumor of a Patient with Denys-Drash Syndrome** S. Habbig,[1] M. Bönsch,[1] M. Ortmann,[2] M. Fischer,[1] B. Hoppe,[1] B.B. Beck.[1] *[1]University Childrens´ Hospital, Cologne, Germany; [2]Pathology, University Hospital, Cologne, Germany.*

**Objectives:** Denys-Drash syndrome is defined as early onset nephrotic syndrome with diffuse mesangial sclerosis (DMS), genital anomalities and bilateral Wilms´ tumor. It is characterized by mutations in the WT1-gene affecting the zinc finger regions of exon 8 and 9 which are responsible for DNA-binding.

**Methods:** We describe an 11-months old girl presenting with microhematuria, proteinuria of nephrotic range, hypertension and abdominal tumor. Renal function was severely reduced. Genitalia were female, 46XX. Imaging revealed a huge tumor in the left kidney and a cystic structure in the right kidney.

**Results:** Histologic analysis showed a stroma-rich nephroblastoma in the left and DMS with intralobular nephroblastomatosis in the right kidney.



Mutational analysis of the WT1 gene identified a heterozygous germ line mutation c.1187A>G in exon 9 (p.D396G). In WT cells, DNA sequencing disclosed a homozygous/hemizygous state of the mutation, while the DMS cells were heterozygous as observed in blood cells. Array-CGH as well as quantitative copy number analyses revealed a disomic status of the WT1 locus in WT cells.

**Conclusions:** The genetic data in our patient indicate that the tumor cells harbored a homozygous WT1 mutation suggesting that the second hit was the same missense mutation as observed in germ line cells.

## Abstract# 604

**Clinical Presentation, Diagnosis and Outcome of Adenine Phosphoribosyltransferase (APRT) Deficiency in Children** J. Harambat,[1] I. Ceballos,[2] G. Bollée,[2] M. Daudon,[2] M. Almeida,[7] G. Champion,[3] C. Dheu,[4] S. Ferrando,[5] G. Guest,[2] B. Horen,[6] S. Taque,[8] A. Bensman.[9] *[1]University Hospital, Bordeaux, France; [2]Necker Hospital, Paris, France; [3]University Hospital, Angers, France; [4]University Hospital, Strasbourg, France; [5]University Hospital, Valencia, Spain; [6]University Hospital, Toulouse, France; [7]University Hospital, Lisbon, Portugal; [8]University Hospital, Rennes, France; [9]Trousseau Hospital, Paris, France.*

**Objectives:** APRT deficiency is a rare autosomal recessive disorder characterized by 2,8-dihydroxyadenine (2,8-DHA) crystalluria which can cause urolithiasis and chronic kidney disease. We aim to document the clinical presentation, diagnosis, and outcome of APRT deficiency in a pediatric cohort.

**Methods:** All pediatric cases of APRT deficiency confirmed at Necker Hospital, between 1978 and 2009 were reviewed.

**Results:** We identified 19 cases from 18 families. Phenotype was known in 17 patients. Median age at diagnosis was 3 years after a mean number of urolithiasis of 3.5 (median 1) in 15 children. In one patient 2,8-DHA crystals were found fortuitously on urinalysis and one additional patient was diagnosed by family screening. All children were found with null APRT enzyme activity. At diagnosis, 13 children had normal renal function, GFR was decreased in 3, and 1child presented with acute renal failure. Allopurinol was given to 16 patients at a median dose of 9 mg/kg/day. After a median follow-up of 6 years, 4 patients exhibited recurrence of urolithiasis, all patients had normal renal function, and all except one had normal height.

**Conclusions:** Whereas urolithiasis can occur in early childhood, renal function is preserved in children with APRT deficiency treated by allopurinol.

## Abstract# 605

**T16M Mutation Causing Autoimmune Polyendocrine Syndrome Type 1 (APS-1) in a Greek Child** A. Tsolaki,[1] C. Antachopoulos,[1] N.A. Karantaglis,[1] M.A. Morris,[2] E. Roilides,[1] K. Kollios.[1] *[1]Third Pediatric Department, Hippokration Hospital, Aristotle University of Thessaloniki, Thessaloniki, Greece; [2]Molecular Diagnostic Laboratory, Geneva University Hospitals, Geneva, Switzerland.*

**Objectives:** Autoimmune Polyendocrine Syndrome Type 1 (APS-1 or Autoimmune Polyendocrinopathy-Candidiasis-Ectodermal Dystrophy Syndrome- APECED) is an autosomal recessive disease caused by mutations of the AutoImmune REgulator (AIRE) gene. We present a case of APS-1 in a 10 -year old girl caused by a rare mutation of the AIRE gene.

**Methods:** The child presented at the age of three years with recurrent episodes of mucocutaneous candidiasis and onychomycosis. Candidiasis subsided at the age of six, following intermittent treatment with itraconazole or fluconazole. Hypoparathyroidism was diagnosed at the age of eight years.

**Results:** Genetic analysis revealed the rare mutation T16M (exon 1) in homogeneity, which is different from the most common mutations R257X and 3-bp deletion. Autoantibodies to thyroid and adrenal gland were not detected and all other endocrine functions were normal. The hypoparathyroidism was treated with calcitriol and calcium carbonate, by which serum levels of plasma calcium and phosphate were normalized during the first two years of treatment. However, a persistent increase of serum phosphate levels has been noted over the last six months for which therapy with the phosphate binder sevelamer has been instituted.

**Conclusions:** T16M mutation in this child caused APS-1 complicated by hypoparathyroidism at the age of eight; normal renal function is still maintained and therapy with sevelamer is required to control the abnormal serum phosphate levels.

## Abstract# 606

**Inflammatory Stress Exacerbates Lipid Mediated Renal Injury in Triple-Knockout (CD36$^{-/-}$SRA$^{-/-}$ApoE$^{-/-}$) Mice** Q. Li, Z. Xu. *Children's Hospital of Chongqing Medical Universtiy, Chongqing, China.*

**Objectives:** To investigate whether inflammatory stress exacerbates lipid accumulation and, thereby exacerbating the progression of renal injury by non–scavenger receptor-mediated pathways.

**Methods:** Male CD36–/–SRA–/–ApoE–/–mice were fed a western diet and were randomly assigned to receive either subcutaneous injections of 10% casein (to induce inflammation) or vehicle (control). Animals were sacrificed after 14 weeks, terminal blood samples were taken for plasma creatinine, BUN, serum cholesterol, triglycerides, LDL, HDL, amyloid A (SAA) and IL-6 assays. Renal sections were used for histological assessments. The lipid accumulation in kidney was evaluated by Oil Red O staining. The mRNA and protein expression of TGF-β, Fibronectin, SREBP2, SCAP, LDLr were analysed by Real-Time Polymerase Chain Reaction and western-blot analysis.

**Results:** Blood levels of SAA was higher (n=8) compared to controls. ORO staining showed lipid accumulation in the kidney was more extensive in casein-injected mice despite lower blood lipid levels. PAS, Masson and HE staining demonstrated casein injection induced renal injury in the kidney. Meanwhile, casein injection increased TGF-β, fibronectin mRNA and protein expression in the kidney, exacerbated renal function failure compared to controls. Casein injection increased LDL receptor gene and protein expression in the kidney, probably by disrupting the intracellular cholesterol sensor, regulatory function of SREBP cleavage activating protein (SCAP), suggesting that SCAP/SREBP/LDL receptor pathway plays an important role in the inflammation-induced glomerular injury.

**Conclusions:** Inflammation lowers serum cholesterol levels and exacerbates lipid mediated renal injury.

## Abstract# 607

**Pseudohypoaldosteronism, Type 1 – A Case Report** V.P. Despotova,[1] M.V. Kalajdgieva,[2] M.I. Lilova,[1] A.S. Paunov,[2] R.Z. Maslarska,[2] S.Z. Deneva,[2] V.L. Konstantinova,[2] K. Grozdeva.[1] *[1]Pediatrics, Tokuda Hospital, Sofia, Bulgaria; [2]Neonatology, Tokuda Hopsital, Sofia, Bulgaria.*

**Objectives:** Pseudohypoaldosteronism ( PHA) is a rare syndrome characterized by defective sodium transport in the distal nephron and renal salt wasting despite high serum aldosterone concentrations. Two clinically distinct forms of PHA have been described. The renal form is characterized by salt loss from the kidneys. It is inherited as an autosomal dominant trait and is caused by mutations in the mineralocorticoid-receptor gene.

**Methods:** We report a case of PHA type 1, renal form that presented as severe salt wasting on the 20th day of life, initially appearing like congenital adrenal hyperplasia (CAH).

**Results:** The baby was with severe dehydration, hyponatremia (104-106 mmol/l), metabolic acidosis and normokalemia. Poor response of the dehydration and electrolyte abnormalities to steroid therapy should make one suspect PHA.

Diagnosis was established by demonstrating the greatly increased values of plasma renin activity >25 ng/ml/h and plasma aldosterone concentration 3518 pmol/l (N 471). The child was treated with sodium chloride and alkali therapy with sodium bicarbonate supplements. This patient presented as life threatening condition, required intensive care. After adequate supplementation the child was clinically stabilized.
**Conclusions:** PHA might be life threatening condition required intensive care and adequate supplementation for clinical control.

## Abstract# 608

**NPHS2 Promoter Polymorphisms Predict Clinical Outcome in Singaporean Chinese Children with Nephrotic Syndrome** I.D. Liu, J.-L. Ng, E. Ng, K.-H. Ng, C.-K. Heng, S.S.C. Chong, H.K. Yap. *Pediatrics, National University Health System, Singapore, Singapore.*
**Objectives:** NPHS2 mutations have been reported in familial and sporadic nephrotic syndrome (NS). We investigated the prevalence of NPHS2 mutations among Singaporean Chinese children and their association with clinical outcomes.
**Methods:** Genomic DNA from 80 patients with primary sporadic NS (mean age at onset 5.4±4.0 years, range 0.58-12.0 years) and 76 cord blood controls was screened using direct sequencing.
**Results:** Eight NPHS2 single nucleotide polymorphisms (SNPs) were identified - four were located in the 5' promoter region (-670 C/T, -213G/A, -116 C/T and -51 G/T); with four in exons 2,5, and 8 (285 C/T, 685 C/A, 954 T/C and 1038 A/G). The genotypic frequencies were consistent with Hardy-Weinberg expectations. There was no significant difference in allele frequencies between patients and controls. Using binary logistic regression analysis, homozygotes for -670 C were significantly associated with the finding of focal and segmental glomerulosclerosis (FSGS) on biopsy (p=0.019, OR=5.9, 95%CI:1.34-25.9) and with poor response to prednisolone and cyclosporine alone, necessitating the use of mycophenolate (p=0.047, OR=3.73, 95%CI:1.02-13.7). Furthermore, this group was associated with eventual inability to wean off all medications with sustained (five year) remission (p=0.027, OR=10.8, 95%CI:1.32-89.9). The presence of at least one copy of T at promoter position -51 was also associated with poor response to steroid therapy alone (p=0.024, OR=4.0, 95%CI:1.21-13.73).
**Conclusions:** NPHS2 promoter polymorphisms may influence clinical outcomes in childhood NS. This effect may be mediated by regulation of NPHS2 transcription.

## Abstract# 609

**Frequency and Functional Relevance of C3 Gene Mutations in Children with Atypical HUS** M. Malina, L. Roumenina, C.P. Schmitt, L.B. Zimmerhackl, M. Pohl, A. Jankauskiene, L. Halbwachs-Mecarelli, V. Fremeaux-Bacchi, F. Schaefer. *Center for Children and Adolescent Medicine, University of Heidelberg, Heidelbereg, Germany.*
**Objectives:** Atypical hemolytic uremic syndrome (aHUS) is a complement dysregulation disorder with a major impact of genetic background. C3 gene mutations have been proposed as a novel susceptibility factor. We screened a large population of children with aHUS from Europe for mutations in the C3 gene.
**Methods:** An unselected cohort of 67 children with aHUS was examined for C3 gene mutations. To investigate the pathogenic relevance of the identified mutations, complement deposition on endothelial cell (EC) surfaces after incubation with sera of patient and their parents was studied.
**Results:** Sequencing revealed 3 novel heterozygous mutations (4%) in 3 unrelated patients and one uncommon C3 polymorphism. The mutations were not found in 100 healthy donors. One mutation is located within the Factor H (SCR3) binding site of C3b. EC exposed to serum containing this mutant deposited increased amounts of C3 compared to normal, at a level similar to Factor H depleted serum. The second mutation did not caused increased C3 deposition in any of the tested conditions. The third one is located in proximity to the Factor B binding site. Carriers of this mutation present with low C3 levels, precluding the use of their sera in EC assays.
**Conclusions:** We identified 3 novel mutations in the C3 gene in our aHUS cohort. One mutation leads to markedly increased C3 deposition on EC surfaces. Another mutation possibly results in a hyperfunctional C3 convertase, leading to C3 consumption. Functional tests with recombinant C3 are in progress.

## Abstract# 610

**Nephrosis, Growth Hormone Deficiency, and Mental Retardation in Three Brothers – Association or New X-Linked Syndrome?** R. Mallmann. *Dept. of Pediatrics and Adolescent Medicine, Elisabeth Hospital, Essen, Germany.*
**Objectives:** Three brothers and half-brothers respectively present with large proteinuria, intermittent microscopic hematuria, mental retardation and short stature. There is one completely healthy (half-) sister.
Renal function as well as blood pressure have been normal over time. Renal biopsy, performed in the oldest boy, shows minimal changes with only a few glomeruli presenting thin membranes. No mutation is found in either NPHS2 or WT1. Magnetic resonance imaging of the brain shows so far non-diagnostic white matter lesions in the two older boys. Growth hormone deficiency is diagnosed in all three.
Growth hormone deficieny is effectively treated by daily hormone injections, proteinuria well contolled by ACE inhibitor.
In summary in this family an inherited X-linked disorder more than mere association is assumed.

## Abstract# 611

**Celiac Disease HLA Aplotypes in Children with Glomerular Diseases** S.S. Maringhini,[1] V.V. Azzolina,[2] D.D. Rallo,[2] S.S. Teresi,[3] E.E. Gucciardino,[3] M.C.M.C. Sapia,[1] C.C. Corrado,[1] M.M. Li Vecchi.[2] *[1]Pediatric Nephrology, Ospedale "G Di Cristina", Palermo, Italy; [2]University of Palermo, School of Nephrology, Palermo, Italy; [3]Laboratory Hospital, Ospedale "G Di Cristina", Palermo, Italy.*
**Objectives:** Celiac disease (CD) is emerging as a common disorder. HLA class II aplotypes DQ2 and/or DQ8 are present in 99% of CD patients and in 30% of the normal population. Sporadic studies have reported an increased prevalence of these aplotypes in patients affected by glomerular diseases (GN) In all children with nephrotic syndrome (NS) or (GN) admitted to our Unit in the last year we determined DQ2/DR3, DQ2/DR7, DQ2/DR4 e DQ8/DR4 aplotypes.
**Methods:** HLA typing was done by DNA extraction and PCR amplification and electrophoresis in agarose. As control groups we examined 27 children with CD and 70 first degree relatives of theirs (CDR)
We studied 79 children with NS; 72 were steroid sensitive (SSNS), 7 steroid resistant (SRNS), 55 males and 24 females (1 -13 years; median 4.5 years). A renal biopsy was done in 17 Thirty children had GN, 14 males and 16 (3 – 16 years, median 9 years. A renal biopsy was done in 20.
**Results:** DQ2 and/or DQ8 aplotypes were present in 69 of 79 patients with NS (87.3%) and in 63 of 72 SSNS (87.5%), in 14 of 30 patients with GN (47%), in 43 of 70 CD relatives (61%) and in all CD patients. DQ2/DR3 combination was present in a smaller percentage of NS and GN compared to CD.
**Conclusions:** In our population a high percentage of children with NS has HLA alleles which predispose to a CD, the same wasn't observed in other glomerular diseases.

## Abstract# 612

**Long-Term Follow-Up of Adolescent (Intermediate) Cystinosis Untreated with Cysteamine** J.P. Midgley,[1] R. El Kares,[2] F. Mathieu,[2] P.R. Goodyer.[2] *[1]Paediatrics, Alberta Children's Hospital, Calgary, AB, Canada; [2]Pediatrics, Montreal Children's Hospital, Montreal, QC, Canada.*
**Objectives:** Document the long-term survival of an individual with untreated nephropathic cystinosis.
**Methods:** Information was abstracted from health records and reviewed with the patient. Exons 3-12 of the CTNS gene were amplified from fibroblast DNA and sequenced.
**Results:** When his older brother was diagnosed with cystinosis, this individual (9 years old) was found to have urinary frequency, moderate Fanconi tubulopathy and corneal crystals. Leukocyte cystine was 4.2 µmol half cystine/g protein. At age 16 he had a pre-emptive kidney transplant from his father that functioned for 28 years, although he developed insulin dependence, hypothyroidism and eye disease in this period. A second kidney transplant was done at age 52.
Systemic and topical cysteamine was started at 53 years of age. We identified a homozygous deletion (c.198_218del) in the 5th exon of *CTNS* that deletes 7 amino acids (in-frame) from the N-terminal domain of cystinosin; this deletion has been described in one other homozygote and six compound heterozygotes with milder or juvenile onset nephropathic cystinosis and was estimated by Kalatzis to have about 20% residual cystine channel activity.
**Conclusions:** Although the cystinosis phenotype has been divided into three forms, there is likely a continuum of disease severity due to residual lysosomal cystine carrier activity with certain CTNS mutations. Our patient had nephropathic cystinosis requiring transplantation at age 16, but has had relatively mild extra-renal disease despite lack of early cysteamine therapy. He attended university, pursued a professional career and survives into the 6th decade.

## Abstract# 613

**Functional Analysis of NPHS1 Gene Mutations with Japanese Patients Suffering from Congenital Nephrotic Syndrome (CNS)** T. Miyai,[1] K. Aya,[1] M. Takaiwa,[1] K. Yan,[2] Y. Sado,[3] H. Tanaka,[4] T. Morishima.[1] *[1]Pediatrics, Okayama University Medical School, Okayama, Japan; [2]Pediatrics, Kyorin University School of Medicine, Mitaka, Japan; [3]Shigei Medical Research Institute, Okayama, Japan; [4]Pediatrics, Okayama Saiseikai General Hospital, Okayama, Japan.*
**Objectives:** Some Japanese patients with CNS have homozygous or compound heterozygous mutations in NPHS1 gene (Aya et al. NDT 2009). Among the gene

mutations we identified, 736G>T(E246X) and nt2515del(C) were unique to Japanese and more frequently detected than other mutations. In that paper, we speculated as to whether the mutation is polymorphism or etiological cause using just the mode of inheritance, the preservation in species of amino acid, and a comparison with the mutations in healthy individuals. We examined functional analysis for the three NPHS1 mutations in vitro.
**Methods:** We generated the mutant nephrin expression vectors using a pcDNA3 vector containing human nephrin cDNA and a QuikChange II XL Site-Directed Mutagenesis Kit (Stratagene). Transfections of plasmids into HEK293 cells were performed using lipofectamine (Invitrogen). These cells were examined by an immunohitochemical study using an anti-nephrin antibody and a secondary antibody conjugated to FITC without a preincubation with triton-X.
**Results:** The cells transfected plasmids carrying either 736G>T(E246X, nt2515del(C) were less stained by the anti nephrin antibody than the cells transfected with the wild type nephrin vector.
**Conclusions:** This functional analysis can help to judge whether the mutation is the etiological cause of CNS. We concluded both mutations are playing a crucial role in the etiology of CNS.

## Abstract# 614

**A Case of Renal-Coloboma Syndrome Associated with Mental Developmental Delay Exhibiting a Novel PAX2 Gene Mutation** T. Miyazawa, K. Sugimoto, H. Yanagida, M. Okada, T. Takemura. *Department of Pediatrics, Kinki University School of Medicine, Osaka-Sayama, Osaka, Japan.*
**Objectives:** Renal coloboma syndrome (RCS) is an extremely rare disorder caused by the PAX2 gene (*PAX2)* abnormality. RCS is clinically characterized by renal hypoplasia and optic disk coloboma, and mainly exhibits an autosomal dominant pattern of inheritance, although sporadic occurrences have also been reported. A case of an adolescent male with RCS showing developmental delay is described.
**Methods:** The patient was a 19-year-old male. Proteinuria was initially observed at the age of 7 years during an annual mass screening program for school children. His urine was checked periodically at a local hospital. Because of an increase in proteinuria, he was referred to our hospital for further clinical evaluation. Proteinuria was moderate, ranging from 1.0 to 1.5 g/day, and was coupled with mild renal dysfunction. At that time, he was found to have myopia associated with astigmatism. He exhibited mild developmental delay, assessed by a WISC-III test.
**Results:** A renal biopsy sample showed marked glomerular enlargement, collapse of glomerular capillaries, mesangial matrix expansion, and tubulointerstitial change, demonstrating typical histologic features of RCS. Approximately five years after starting follow-up, the patient had severe renal dysfunction. Furthermore, optic nerve coloboma was also evident. Previous reports of *PAX2* abnormalities in RCS included five types of frame shift mutation in exon 2, 5 or 8, two types of missense mutation in exon 3, and chromosomal translocation of *PAX2* intron.
**Conclusions:** Genetic analysis of the patient revealed a novel heterozygous mutation in exon 3 of the PAX2 gene (P130H).

## Abstract# 615

**Spry1 Molecular Analysis in Subjects with Ureteral Duplicity** E. Benetti,[1] S. Centi,[2] S. Negrisolo,[2] G. Caridi,[3] L. Murer,[1,2] L. Artifoni.[2] *[1]Pediatric Nephrology, Dialysis and Transplant Unit, Department of Pediatrics, University of Padua, Padua, Italy; [2]Laboratory of Immunopathology and Molecular Biology of the Kidney, Department of Pediatrics, University of Padua, Padua, Italy; [3]Laboratory of Physiopathology of Uremia, Gaslini Institute, Genoa, Italy.*
**Objectives:** Studies on murine models demonstrated that Sprouty1 protein, encoded by Spry1 gene, modulates Gdnf/Ret signal, which activates a crucial gene network in urinary tract development. Spry1-knockout mice develop supernumerary ureteric buds, which result in multiple ureters and kidneys. In the literature, there is only one report about mutational analysis of human SPRY1 gene, the homologue of murine Spry1, even if the gene is known to be expressed in human fetal renal tissue.
**Methods:** We carried out mutational analysis of SPRY1 gene in patients with duplex ureter: 23 isolated and 4 familial cases. On each DNA sample, the coding region and 5'UTR were analysed by SSCP and all PCR products were directly sequenced (ABI PRISM 3100 Applied Biosystem). DNA from 6 subjects without kidney and urinary tract anomalies was used as control.
**Results:** We detected 5 polymorphisms (SNPs), previously reported in databases, and 1 nucleotide substitution, which has never been reported. The frequency of this substitution was estimated in 127 umbilical cord blood DNA samples and was 0.094.
**Conclusions:** In order to understand SPRY1 role in renal development, mutational analysis will be extended to a population of subjects with different malformative nephrouropathies and association studies with the detected polymorphisms will be performed.

## Abstract# 616

**PAX2 Mutation Analysis in Children with Isolated Congenital Anomalies of Kidney and Urinary Tract** E. Benetti,[1] S. Negrisolo,[2] S. Centi,[2] M. Della Vella,[2] L. Murer,[1,2] L. Artifoni.[2] *[1]Pediatric Nephrology, Dialysis and Transplant Unit, Department of Pediatrics, University of Padua, Padua, Italy; [2]Laboratory of Immunopathology and Molecular Biology of the Kidney, Department of Pediatrics, University of Padua, Padua, Italy.*
**Objectives:** PAX2 gene encodes a transcription factor expressed during ureteric bud induction and branching, and in metanephric mesenchyme condensation that leads to nephron formation. PAX2 mutations are associated with human Renal-Coloboma Syndrome (RCS), a rare autosomal dominant disease, whose hallmarks are optic coloboma and renal developmental anomalies. PAX2 mutations in subjects with renal but not ocular clinical features have also been reported.
**Methods:** We carried out PAX2 gene molecular analysis in 20 children with renal hypoplasia/oligomeganephronia with or without associated vesico-ureteric reflux (VUR), and renal agenesia. None of the subjects had optic coloboma.
**Results:** A novel PAX2 truncating mutation (*c.619delC* NM_003987) in exon 2 was detected in a patient with bilateral renal hypoplasia and VUR. Furthermore, a novel intronic sequence variant (*c.960+5g/a* NM_003987) was identified in a subject with oligomeganephronia. This nucleotide substitution, not detected in 100 control DNA samples, may affect PAX2 splicing.
**Conclusions:** Our results confirm the role of PAX2 gene in kidney and urinary tract developmental diseases and highlight the importance of performing PAX2 molecular analysis in patients with isolated congenital kidney and urinary tract anomalies.

## Abstract# 617

**Dent Disease in Two Croatian Patients and a Novel H731P Mutation** I. Palcic,[1] G. Petkovic,[1] A. Cvitkovic Roic,[2] I. Barišic,[1] G. Roic,[1] M. Bastic,[1] J. Delmiš,[1] F. Anglani,[3] M. Coel,[3] F. Mezzabotta.[3] *[1]Children´s University Hospital Zagreb, Zagreb, Croatia; [2]Helena Polyclinic, Zagreb, Croatia; [3]Laboratory of Molecular Biology of the Kidney, University of Padova, Padova, Italy.*
**Objectives:** We present two Croatian patients with Dent disease.
**Methods:** First patient is a 12 years old boy diagnosed with persistent asymptomatic proteinuria and normal renal function at the age of 3 years. First renal ultrasonography (US) was normal. Kidney biopsy showed FSGS. 6 years later low-molecular-weight proteinuria, hypercalciuria and nephrocalcinosis grade II were found. Second patient is a 12 years old boy referred for nocturnal enuresis at the age of 6 years. Low-molecular-weight proteinuria and hypercalciuria with normal renal function were found. US showed nephrocalcinosis grade II. Kidney biopsy showed total sclerosis of 9% of investigated glomeruli and calcifications in interstitium and medular tubule.
**Results:** DNA analysis of CLCN5 gene in first patient revealed the 637delT mutation in exon 4 leading to a frame-shift mutation introducing a premature stop codon in position 137 of the protein chain (C116VfsX137). CLCN5 gene analysis in second patient showed a complex allele (IVS2-17T>G; H731P) constituted of nucleotide substitution in intron 2 and exon 12 leading to a missense mutation changing histidin with prolin in protein chain. This allele was inherited from the mother who is heterozygous for the complex mutation.
**Conclusions:** These patients are the first two patients with Dent disease confirmed by DNA analysis in Croatian population. To our knowledge the mutation H731P is a novel mutation not reported before.

## Abstract# 618

**Renal Features of Pediatric Tuberous Sclerosis Complex** E. Paul,[1] S.E. Camposano,[2] E.A. Thiele.[2] *[1]Pediatric Nephrology, MGH, Boston, MA, United States; [2]Neurology, MGH, Boston, MA, United States.*
**Objectives:** Tuberous sclerosis complex (TSC) is a genetic disease whose renal lesions can cause morbidity and mortality. In predominantly adult populations, angiomyolipomas, cysts and carcinomas are seen in up to 75%, 35% and 5% of patients, respectively. This study determines the prevalence and nature of renal lesions in a single center pediatric TSC population and measures rates of lesion growth.
**Methods:** Retrospective chart review at the MGH Herscot Center for TSC identified nearly 200 individuals with renal imaging before age 21. Renal lesions are evaluated for radiologic appearance and growth in patients who are characterized by gender, age, size and genotype.
**Results:** Renal lesions are seen in over 70% of children with TSC. Lesions were detected in 5% of patients in the first year of life, 65% in the first decade and 35% between ages 10 and 21. Renal masses are seen in approximately two thirds of these patients, of whom 80% also have cysts. Cysts alone are seen in the remaining 30%. Over 80% of these renal patients have TSC2 gene mutations in contrast to 35% of pediatric TSC patients without renal lesions. Initial lesion size and the subsequent rates of growth vary tremendously. One child was diagnosed with papillary renal cell carcinoma at age 7.
**Conclusions:** TSC renal lesions are more prevalent in pediatric patients than

previously appreciated. While most of these lesions have no morbidity before the third decade, many display steady growth and some warrant embolization and rarely even resection. Regular follow up of these lesions is required to identify those children for whom medical or mechanical intervention may eliminate renal-associated mortality and preserve renal longevity.

## Abstract# 619

**Prophylactic Embolization of Renal Angiomyolipoma in Pediatric Tuberous Sclerosis Complex** E. Paul,[1] T.J. Norton,[2] E.A. Thiele,[3] S. Wicky,[2] G.M. Salazar.[2] *[1]Pedi Nephrology, MGH, Boston, MA, United States; [2]Vascular Interventional Radiology, MGH, Boston, MA, United States; [3]Neurology, MGH, Boston, MA, United States.*

**Objectives:** Renal lesions are seen in a majority of patients with Tuberous Sclerosis Complex (TSC), most of whom have renal angiomyolipomas (AMLs). Since hemorrhage from these vascular hamartomas is a leading cause of mortality in TSC, prophylactic embolization is recommended for AMLs that exceed 4 cm in their longest diameter (LD). This project reports a single center experience with renal AML embolization in a series of 10 children with TSC.

**Methods:** Retrospective chart review at the MGH Herscot Center for TSC identified 10 patients assessed by angiography for prophylactic embolization of renal AMLs before age 21. Embolic methods, peri-procedure complications and long term changes are described.

**Results:** Thirteen of 16 renal AMLs (mean LD 6.7 cm; range 2.8-12.7) evaluated by renal angiography were selectively embolized in 10 children with TSC (mean age 12.6; range 5.8-19.5) in a total of 12 separate procedures. The only AML embolized to arrest bleeding was the smallest one in the series. Some children required antibiotics and glucocorticoids to treat post-embolization syndrome. None exhibited increases in baseline blood pressure or creatinine in a mean follow up of 26 months (range 6-60).

**Conclusions:** The AML size best suited for prophylactic embolization in children with TSC has yet to be determined. Most in our series were considerably larger than 4 cm and the only one to bleed was smaller. Nonetheless, the procedure is apparently safe in children with large renal AMLs, with no induction of hypertension or renal insufficiency and no post-procedure hemorrhage to date.

## Abstract# 620

**CD48 Deficiency Precipitates Autoimmune Nephritis in Lupus Prone Mice** E. Paul,[1] Y. Latchman,[2] A. Kirby,[1] A.H. Sharpe,[3] M.J. Daly,[1] R.B. Colvin.[1] *[1]MGH, Boston, MA, United States; [2]U Washington, Seattle, WA, United States; [3]BWH, Boston, MA, United States.*

**Objectives:** Sle1b is a lupus susceptibility locus that is associated with humoral autoimmunity. This region contains members of the SLAM/CD2 gene family including CD48. In the context of a permissive Sle1b haplotype, CD48 deficiency promotes fatal glomerulonephritis (GN) prompting the hypothesis that CD48 protects lupus prone individuals from progressive autoimmune disease.

**Methods:** C57BL/6 and BALB/c mice deficient for CD48 were characterized by serology and histology for features of lupus.

**Results:** B6.CD48-/- develop proliferative GN with immune complex deposition, mesangial proliferation, leukocyte infiltration, crescent formation and progressive fibrosis that culminates in renal failure. B6.CD48-/- mice also have autoantibodies. In contrast, CD48-/-BALB/c mice and CD48-/-F1 [B6 x BALB] progeny are disease free. One backcross of F1 animals to the B6.CD48-/- parental strain restores autoantibody production in enough N2 progeny to implicate a mendelian modifier of this humoral trait. However, renal disease is recovered only after additional backcrosses to the B6.CD48-/- strain, demonstrating that GN pathogenesis is polygenic. Genome wide SNP and QTL analyses of N2 animals have linked autoantibody production to the MHC region of chr 17. Mapping of the nephritic phenotype is underway.

**Conclusions:** CD48-deficient mice develop fatal lupus-like GN. Presumably in combination with 129 alleles of the Sle1b locus, permissive genetic modifiers of disease lie within the C57BL/6 (versus BALB/c) genome and include linkage to the murine MHC. These data suggest that CD48 is a crucial participant in multigenic interactions that can precipitate lethal autoimmune disease.

## Abstract# 621

**A Point-Mutation in ROMK Gene Leads to Bartter Syndrome in Two Sisters** P.C.B. Pereira,[1] D.M. Miranda,[1] H. Sarubi,[2] L.A.C. de Marco,[2] E.A. Oliveira,[1] A.C. Simoes e Silva.[1] *[1]Pediatrics, Federal University of Minas Gerais, Belo Horizonte, Minas Gerais, Brazil; [2]Nacional Institute of Science and Technology of Molecular Medicine - INCTMM, Belo Horizonte, Minas Gerais, Brazil.*

**Objectives:** The aim of this study was to identify the molecular defect in two sisters with Bartter syndrome.

**Methods:** DNA was isolated from peripheral blood samples by high salt method of Lahiri and Nurnberger. PCR-amplified DNA of the hot spot region of the ROMK gene from the two girls and their parents was performed. The primers used were KCNJ1, KCNJ2 and KCNJ3. PCR protocol was performed in a final volume of 25µl: 1 X PCR buffer (10mM Tris-HCl pH 8.4, 40mM NaCl, Triton X-100 0,1%, 1.5mM MgCl2), 500µM dNTPs, 1.0U of Taq DNA polymerase, 20ng of genomic DNA, 0.4µM of each forward and reverse primer. A direct sequencing was performed twice with different PCR products in automated DNA sequencer (ABI 3130 Applied Biosystems, Foster City, CA). Sequences were analyzed by Sequencer *4.9* software.

**Results:** Genomic DNA sequencing revealed a homozigous mutation in exon 5 of KCNJ1 gene that changed nucleotide 641C into T. This point-mutation resulted in the substitution of an alanine residue by a valine residue at codon 214 (A214V) in both daughters, but not in their parents. The parents are heterozigous for this mutation. Genomic control confirmed that parents are not related.

**Conclusions:** *KCNJ1* gene codifies the kirk1.1 (ROMK) protein, which is a member of inwardly rectifying family of potassium channels, the major responsible for potassium homeostasis. Molecular evaluation of Bartter syndrome patients will certainly enable better understanding of the disease and of renal tubular physiology.

## Abstract# 622

**Renal and Extra-Renal Phenotype Associated with *PAX2* Mutations in 17 Families** L. Heidet,[1] V. Morinière,[1] A. Pawtowski,[2] A. Vandewalle,[1] A. Corinne,[2] S. Rémi,[1] The French Society of Pediatric Nephrology.[3] *[1]AP-HP, Centre de Référence des Maladies Rénales Héréditaires de l'enfant et de l'adulte (MARHEA), Paris, France; [2]AP-HP département de Génétique, Hôpital Necker, Paris, France; [3]The French Society of Pediatric Nephrology, France, France.*

**Objectives:** To characterize the renal and extra-renal phenotype associated with *PAX2* mutation in 17 families.

**Methods:** We retrospectively analyzed the medical records of 28 patients from 17 families for whom a *PAX2* mutation was identified.

**Results:** Prenatal US showed bilateral hypoplasiain 3/7 cases, unilateral agenesia in 1/7 case and multicystic dysplasia in 1/7 case. After birth, 3 patients had single kidney, 1 had multicystic dysplasia and 17 a bilateral hypo-dysplasia. In one kidneys looked normal. In 5 patients, 4 having reached ESRD, we had no information on the renal phenotype. Nine patients had vesico-ureteral reflux. Two pregnancies were terminated. Most patient developed renal failure, but the progression was very variable from one patient to another, even in a same family. For patient who have reached ESRF, age at ESRF was on average 17,9 years (4 to 60). Ophthalmologic examination showed anomalies in 17/24 cases. One patient had retinal detachment, 2 had sensorineural deafness, 1had epiphyseal dysplasia, 1ligamentous laxity, and 1pulmonary valva stenosis.Thirteen mutations were identified in 17 probands. The mutation was *de novo* 4/9 times and inherited 5/9 times.

**Conclusions:** The presence and severity of the renal and ocular phenotype associated with *PAX2* mutations is very variable. Ocular involvement, which is frequently mild, can be complicated of retinal detachment. *De novo* mutations are frequent.

## Abstract# 623

**Tricho-Rhino-Phalangeal Syndrome in Girl with Chronic Renal Failure and Severe Growth Deficiency** N. Ristoska- Bojkovska,[1] V. Tasic,[1] Z. Gucev,[1] V.J. Lozanovski,[1] D. Wieczorek,[2] H.-J. Lüdecke.[2] *[1]Nephrology, University Children's Hospital, Skopje, Macedonia, The Former Yugoslav Republic of; [2]Institut für Humangenetik, Universitätsklinikum, Essen, Germany.*

**Objectives:** Tricho-rhino-phalangeal syndrome (TRPS) is a very rare autosomal dominant disease characterized by a triad of hair, craniofacial and skeletal abnormalities. In this work we present a young female with typical clinical features of TRPS, severe growth retardation and chronic renal failure.

**Methods:** The index patient and her first degree relatives underwent detailed dysmorphologic, anthropometric and skeletal examination, imaging of the urinary tract and mutational analysis of the TRPS1 gene.

**Results:** A 13 year old female presented with typical facial dysmorphy, sparse and slowly growing hair and brachyphalangia. She had severe staturoponderal deficit [height 109 cm (-5.7 SD) and weight 16.2 kg (-3.6 SD)], bone deformities of both proximal radii resulted in marked limitation of extension in elbows and chronic renal failure. Kidney ultrasound demonstrated small and hyperchogenic kidneys in favor of bilateral hypoplasia/dysplasia. Sequencing of coding regions of the TRPS1 gene in the index patient and his father revealed heterozygous missense mutation in the exon 6 A919V [c.2756C>T].

**Conclusions:** Our patient with TRPS had two new features: bone deformities of both proximal radii and bilateral VUR with hypodysplasia leading to chronic renal failure. Ultrasound screening should be offered to all patients with TRPS and vice versa – one should look for typical hair, facial and skeletal features in patients with congenital anomalies of the kidney and urinary tract.

## Abstract# 624

***CLCN5* and *OCRL1* Negative Dent Disease** S.K. Sethi,[1] M. Ludwig,[2] A. Sinha,[1] P. Hari,[1] A. Bagga.[1] *[1]Division of Pediatric Nephrology, All India Institute of Medical Sciences, New Delhi, India; [2]Department of Clinical Chemistry & Pharmacology, University of Bonn, Bonn, Germany.*

**Objectives:** Dent disease is characterized by low molecular weight proteinuria (LMW), hypercalciuria and nephrocalcinosis/lithiasis. Mutations in the *CLCN5* gene (Dent disease 1, OMIM #300009) or the *OCRL1* gene (Dent disease 2, OMIM #300555) are responsible. We report 5 patients with Dent phenotype without mutations in these 2 genes.

**Methods:** Clinical and laboratory findings of 5 patients with Dent disease are presented. All patients were analyzed for LMW proteinuria (particle-enhanced immunonephelometry) and *CLCN5* and *OCRL1* mutations (by automated gene sequencing). DNA samples from 100 unrelated healthy subjects served as controls.

**Results:** Out of 5 patients, there were 3 boys. Age ranged from 7 months to 8 years. All patients presented with polyuria, polydipsia and resistant rickets. Three patients also had features of recurrent vitamin A responsive night blindness. All patients showed hypophosphatemia, proximal renal tubular acidosis with generalized aminoaciduria and marked hypercalciuria. One patient had glucosuria. Two patients had nephrocalcinosis. All patients showed marked LMW proteinuria with increased excretion of beta-2 microglobulin, alpha-1 microglobulin, retinol binding protein and transferrin. All lacked defects in the *OCRL1* and *CLCN5* genes.

**Conclusions:** Our findings emphasize the genetic heterogeneity of Dent phenotype in children. Further studies are needed to determine the spectrum of genetic defects underlying this condition.

## Abstract# 625

**Primary Hyperoxaluria Type2: A Case Report from India** J. Sharma,[1] C.G. Monico,[2] S.K. Lalwani.[1] *[1]Dept. of Pediatrics, Bharati Vidyapeeth University Medical College, Pune, Maharashtra, India; [2]Division of Nephrology, Mayo Clinic Hyperoxaluria Center, Rochester, MN, United States.*

**Objectives:** Primary hyperoxaluria type 2 (PH2) is a rare, autosomal recessive disease due to mutations in hepatic GRHPR, characterized by elevated urine oxalate, L-glycerate, recurrent urolithiasis/nephrocalcinosis & ESRD. Of 203 patients in the International Primary Hyperoxaluria Registry (IPHR), only 19 have PH2, 5 are children; ~ 40 cases are reported worldwide. We report a case from India homozygous for the c.494G>A (G165D) *GRHPR* mutation.

**Methods:** *Case Report*: 11 month, male, 3rd product of $2^0$ consanguineous marriage, presented with UTI & bilateral renal calculi. He was normotensive, Screatinine was 0.6 mg% & urine oxalate was elevated (0.86 mmol/1.73m2/24h). A sibling died due to ESRD & his X-ray showed bilateral nephrocalcinosis; screening for *GRHPR* mutations revealed homozygosity for c.494G>A(G165D). Therapy to reduce stone formation (elemental phosphorous) was instituted; and he was advised to avoid dehydration & oxalate rich foods. Review after 2 years: has grown well, no increase in nephrocalcinosis, renal function is preserved. Segregation analysis in his parents and a healthy sibling confirmed carrier (G494A heterozygosity) status in all.

**Conclusions:** This 1st PH2 case of Indian descent in the IPHR highlights intra-familial phenotypic heterogeneity & benefits of molecular genetic testing for early diagnosis. PH should be suspected in any pediatric stone former, with significant family history, including parental consanguinity; & measures to delay/prevent progression instituted early.

We acknowledge: IPHR DK073354, Rare Kidney Stone Consortium U54KD083908, Oxalosis and Hyperoxaluria Foundation.

## Abstract# 626

**Mutations in NPHS2 & WT1 Genes in Indian Children with Steroid Resistant Nephrotic Syndrome (SRNS)** S. Sharma, M. Kabra, A. Dinda, P. Hari, A. Bagga. *Pediatrics, All India Institute of Medical Sciences, New Delhi, India.*

**Objectives:** Mutations in NPHS2 & WT1 genes are reported in 10-28% patients with SRNS. Since the frequency of mutations in these genes is unknown for Indian children, we examined for their prevalence & association with clinical outcome.

**Methods:** Patients showing initial resistance to therapy with prednisone (2 mg/kg/d for 4 wk) from across the country were studied. Controls were 80 patients with steroid sensitive nephrotic syndrome. Conformation sensitive gel electrophoresis was done for all exons of NPHS2 & WT1. If an aberrant band was found, sequencing was done. Their response to therapy with calcineurin inhibitors (CNI) and long-term outcome was noted.

**Results:** 120 patients [111 sporadic, 9 familial] (82 boys) with SRNS were studied; age at onset of disease was 4.4 yr. Renal biopsies showed FSGS (36), MCD (54) & MesPGN (21). 16 patients (13%) showed 10 different mutations in NPHS2 gene. Homozygous & compound heterozygous mutations were seen in 8 patients (1 familial, 7 sporadic). Three, who were compound heterozygous for R229Q, showed remission following CNI therapy. Of 14 patients with infantile onset, one had a homozygous NPHS2 mutation. All but one patient with heterozygous variants (n=8) showed remission with CNI therapy; none progressed to ESRD. We detected six novel mutations in the NPHS2 gene; none were seen in controls. Three girls had mutations in WT1 gene [Frasier syndrome: IVS9+4C>T, IVS9+5G>A; Denys Drash syndrome: R394W]. Two of 3 showed partial remission following CNI therapy.

**Conclusions:** Mutations in the NPHS2 & WT1 gene are present in 9% children with SRNS in India. Further studies are required to determine the significance of benefit with CNI therapy.

## Abstract# 627

**Screening for Fabry Disease in a Pediatric Population – Preliminary Data** L.C. Sylvestre, M.S. Santos, M. Bandeira, E.C. Pellissari, L.R. Xavier, R.A. Torres, N. Almeida, P.G. Iankilevich. *Hospital Pequeno Principe, Curitiba, Parana, Brazil.*

**Objectives:** To identify the occurrence of Fabry Disease in a Pediatric Population followed in a Tertiary Hospital in South Brazil.

**Methods:** Patients from 0 to 18 years old referred from the Departments of Nephrology, Rheumatology and Neurology of a Tertiary Pediatric Hospital were eligible according to specific signs and symptoms. All of them were submitted to a screening dosage of alfa Galactosidade A (alfa GAL) activity with enzymatic assay for boys and girls and DNA analysis for girls; they were also evaluated by a Dermatologist in order to detect specific skin lesions and Cardiologist, to perform an Electrocardiogram and Echocardiography. The study was approved by the hospital's Ethics Committee.

**Results:** We analyzed a subgroup of 47 out of 66 patients included in the complete study. These patients had already performed the majority of the screening exams. 19 patients (40%) were female and 28 (60%) were male. Mean age was 10 ± 4 years old. LVH was found in 5 of 43 patients and Electrocardiogram had changes in 11 out of 46 patients. No specific skin lesion was detected. The most frequent symptoms included generalized pain, burning sensations and hyperhidrosis.In 34 patients the results of alfa GAL activity were available, in 3 patients it was in the lower limit and they have repeated the tests and 1 girl presented a mutation despite of having a normal level of alfa GAL activity.

**Conclusions:** Fabry disease is a rare condition but early symptoms may occur in childhood and adolescence. Screening programs can help on making early diagnosis, leading to early treatment and preventing late life-threatening conditions.

**DISCLOSURE:** *Sylvestre, L.C.*: Grant/Research Support, Shire. *Santos, M.S.*: Grant/Research Support, Shire. *Bandeira, M.*: Grant/Research Support, Shire.

## Abstract# 628

**Interleukin 8 Gene 2767 A/G Polymorphism Is Associated with Increased Risk of Nephritis in Children with Henoch Schönlein Purpura** Y. Tabel,[1] S. Mir,[2] A. Berdeli.[3] *[1]Pediatric Nephrology, Inonu University, Malatya, Turkey; [2]Pediatric Nephrology, Ege University, Izmir, Turkey; [3]Molecular Medicine Research Laboratory, Ege University, Izmir, Turkey.*

**Objectives:** The objective of this study is to investigate the association between IL-8 gene 2767 G/A polymorphism and clinical features, kidney involvement and prognosis in childhood Henoch Schnölein purpura (HSP).

**Methods:** A total of 115 patients with HSP (59 male, 56 female) were included in the study with age at diagnosis between 2 and 17 (8.0±3.0 years). Hundred and eight healthy adults were included in the study as controls. The patients had been followed up for kidney involvement for at least 6 months, and in average 8.2±7.5 months. Interleukin 8 (IL–8) gene 2767 G/A polymorphism was studied by PCR-RFLP method.

**Results:** Frequency of the "A" allele was 0.37 in the patient group, whereas it was 0.36 in the control group. The difference was not statistically significant (p=0.696). No association was detected between the IL-8 gene G/A polymorphism and the clinical, laboratory and demographic data related to the patients with HSP. Kidney involvement was more common in those with the G/A polymorphism of the IL-8 gene. While a 0.44 frequency of the "A" allele was detected in those with kidney involvement, this rate was 0.29 in those with no kidney involvement (p=0.046). Follow-up of those with the "A" allele revealed higher proteinuria (p=0.023, odds ratio 0.176, %95CI 0.034–0.917), and higher creatinine levels (p=0.049, odd's ratio 0.024, %95CI 0.036–0.094).

**Conclusions:** These results suggest that the kidney involvement to be more common in patients with the "A" allele, and higher proteinuria and creatinine levels in these patients at follow-up.

## Abstract# 629

**Genetics of Renal Hypodysplasia in the Chronic Kidney Disease in Children Cohort Study (CKiD)** R. Thomas,[1] S. Sanna-Cherchi,[2] P.L. Weng,[2] B. Warady,[3] S.L. Furth,[4] F.J. Kaskel,[1] A.G. Gharavi.[2] *[1]Ped Nephrology, Children's Hospital at Montefiore, Bronx, NY, United States; [2]Nephrology, Columbia University, New York, NY, United States; [3]Ped Nephrology, Children's Mercy Hospital, Kansas City, MO, United States; [4]Ped Nephrology, Children's Hospital of Philadelphia, Philadelphia, PA, United States.*

**Objectives:** 1) Determine the prevalence of mutations in *TCF2* and *PAX2* genes in the North American CKiD population with RHD.
2) In patients with no mutations in *TCF2* or *PAX2*, explore the contribution of rare copy number variations using high-density SNP arrays.

**Methods:** Genomic DNA was obtained from the NIH biorepository. We performed direct sequencing of *TCF2* and *PAX2* exons and intron boundaries. Sequence traces were analyzed using Sequencer 4.8 software. Pathogenic relevance was inferred by publicly available prediction programs.

**Results:** We received 73 DNA samples of children with RHD. To date, we have confirmed 5 likely pathogenic variants (7% of cases). Two are previously noted pathogenic mutations in *TCF2*: a premature termination signal (p.R181X) and a missense mutation (p.S148L). In *PAX2*, we found one missense mutation (p.G24E), one coding frame shift mutation (G24fsX28), and one splice site mutation (IVS4-1G>T) which are all likely pathogenic and have not been noted previously. These five cases are Caucasian.

**Conclusions:** A proportion of patients with non-syndromic RHD carry mutations in either *TCF2* or *PAX2* genes. These patients should be evaluated for complications (e.g. diabetes for *TCF2* mutations, colobomas for *PAX2*) and referred for genetic counseling.
A genome-wide screen for novel pathogenic structural variants in patients without mutations in these genes is ongoing.

## Abstract# 630

**Additional Molecular Findings in Turkish Cystinosis Patients** R. Topaloglu,[1] T. Vilboux,[2] B. Tinloy,[2] T. Coskun,[3] M. Gunay-Aygun,[2] A. Jeong,[2] A. Bakkaloglu,[1] N. Besbas,[1] S. Ozen,[1] S. Sivri,[3] R. Kleta,[4] W.A. Gahl.[2] *[1]Pediatric Nephrology, Hacettepe University Faculty of Medicine, Ankara, Turkey; [2]National Human Genome Research Institute, NIH, Bethesda, MD, United States; [3]Pediatric Metabolic Diseases, Hacettepe University Faculty of Medicine, Ankara, Turkey; [4]Department of Medicine, University College London, London, United Kingdom.*

**Objectives:** We reported the additional molecular findings in Turkish cystinosis patients.

**Methods:** Molecular analyses involve an initial multiplex PCR, checking for 57 kb deletion and sequencing of the 10 coding exons of *CTNS*.

**Results:** None of the 12 patients carried the 57kb deletion. We found a new 10kb deletion of exons 4 to 8 and 2 new missense mutations in exons 7 and 8, c.451G>A and c.518A>G, respectively. The most common variant was a new possible exonic splice site mutation in exon 9, c.681G>A. Of 24 alleles studied, 38% carried this variant which is expected to disrupt proper splicing. Interestingly we have two affected siblings while one is homozygote, the other one is heterozygote for this variant. This may imply that this variant is not disease causing but linked with the real cause of the disease. This hypothesis is comforted by the patient in whom we found three different variants, homozygote for c.140+1G>T and heterozygote for c.681G>A. The homozygote variant is most likely disease causing since it affects the canonical site for splicing. In one patient no possible variant was found.

**Conclusions:** More molecular studies may be needed, not only to identify new possible mutation but also to confirm if c.681G>A is a polymorphism. Our data confirm that the mutational spectrum of the *CTNS* is wide.

## Abstract# 631

**Linkage Analysis and Mutational Screening in a Family with Atypical HUS** D. Westra,[1] E. Volokhina,[1] T. van der Velden,[1] N. van de Kar,[1] L. van den Heuvel.[1,2] *[1]Pediatric Nephrology, Radboud University Nijmegen Medical Centre, Nijmegen, Netherlands; [2]Pediatrics, University Hospital Leuven, Leuven, Belgium.*

**Objectives:** Atypical hemolytic uremic syndrome (aHUS) is associated with predisposing mutations in genes encoding complement (regulating) proteins and with the presence of autoantibodies against complement factor H (αFH). About 50% of the patients progress to ESRD. In this study, we performed linkage analysis and mutational screening in a family with aHUS. Six family members have low C3 plasma values; three of them are diagnosed with aHUS with an age at onset of the disease of 33, 14, and 2, respectively.

**Methods:** Linkage analysis was performed; in case of a positive LOD score, the function of the proteins encoded by genes located between these markers was checked in databases. In addition, mutational screening was performed for complement (regulating) genes: *CFH*, *IF*, *MCP*, *C3*, and *FB*. Two patients were tested for the presence of αFH.

**Results:** Linkage was found on chr. 1 (LOD score: 1.73) and chr. 15 (LOD score: 1.46). No complement candidate genes were identified within these regions. A heterozygous mutation (K323E) was found in *FB*, located on chr. 6, in the family members with low C3 values, including the aHUS patients. This change has been identified as a gain of function mutation resulting in a C3bBb complex that is more resistant to decay by DAF and CFH. No genetic changes were found in the other screened genes and no αFH were identified.

**Conclusions:** A pathogenic mutation (K323E) was found in *FB* in three aHUS patients and three family members. An incomplete penetrance of the disease is found, indicating that, next to genetic predispositions, other factors are involved in the development of aHUS.

## Abstract# 632

**The Collaborative European Cohort of Primary Hyperoxalurias: Clinical and Genetic Characterization with Prediction of Outcome in PH Type 2** C. van Woerden,[1] B. Beck,[2] S. Hulton,[3] G. Mandrile.[4] *[1]AMC, Amsterdam, Netherlands; [2]UK, Cologne, Germany; [3]CHB, Birmingham, United Kingdom; [4]UNITO, Torino, Italy.*

**Objectives:** Primary hyperoxalurias (PH) are characterized by excessive endogenous oxalate production and a heterogeneous phenotype with respect to end-stage renal disease (ESRD). We document the genetic spectrum and clinical outcome of PH type 2 in a large multicenter European study.

**Methods:** Patient data collected from eight European medical centers (retrospectively and prospectively), with documentation of genotype and clinical outcome. Participating countries included The Netherlands, Italy, Germany, United Kingdom, Spain, France, Poland and Sweden.

**Results:** We identified 53 PH2 patients. Genotype and outcome were known in 24 patients (median follow-up of 16.0; range 5.0 – 43.1 years). ESRD occurred in none of the patients at presentation. At last review 4% of PH2 patients had developed ESRD and 6% had died. Of fourteen identified GRHPR gene mutations the most common were c.103delG (15 patients), c.403_405+2delAAGT (13 patients), c.494G>A (8 patients).

**Conclusions:** PH2 is heterogeneous in clinical presentation and mutational spectrum. The vast majority of patients preserves renal function. Metabolic screening in patients with urolithiasis without renal insufficiency may uncover new PH2 patients. Ongoing data collection as part of an international registry will clarify disease presentation and outcomes further.

## Abstract# 633

**Mutations in NPHS1 in Russian Patients with Finnish-Type Congenital Nephrotic Syndrome** T.S. Voznesenskaya, E.E. Tikhomirov, N.S. Averyanova, A.N. Tsygin. *Scientific Centre of Children Health, Moscow, Russian Federation.*

**Objectives:** Congenital nephrotic syndrome is defined as nephrotic syndrome with onset within first 3 month of life. Congenital nephrotic syndrome of the Finnish type (CNF) is an autosomal recessive disorder mainly caused by mutations in the nephrin gene. CNF incidence is highest in Finland, but it may occur all over the world.

**Methods:** Genomic DNA was extracted from leukocytes, and all exons and exon-intron boundaries were analyses for NPHS1 using polymerase chain reaction and sequence analyses in 5 CNF Russian patients.

**Results:** Compound heterozygous mutations of NPHS1, 6 different mutations totality, were found in four patients and homozygous common polymorphism G349A in one. Two among five patients had the same mutation: 1020del T in exon 9, one of them had it geterozygously in compound with G349A. No one had typical Finnish mutations. All mutations detected except for one have never been described before.

**Conclusions:** Nephrin gene mutations are the cause of Finnish-type congenital nephrotic syndrome in Russian patients.

## Abstract# 634

**Improvement of Tubular Function in Infantile Nephropathic Cystinosis after Delayed Onset of Cysteamine Treatment** A.W. Wade, J.P. Midgley. *Paediatrics, Alberta Children's Hospital, Calgary, AB, Canada.*

**Objectives:** In infantile cystinosis document proteinuria and electrolyte supplementation following delayed initiation of cysteamine.

**Methods:** Protein to creatinine ratio (PCR), albumin to creatinine ratio (ACR), alpha 1 microglobulin to creatinine ratio (α1MCR) and electrolyte supplementation was analysed in relation to the initiation of cysteamine and for the last 2 years.

**Results:** At 7 months of age this infant was diagnosed with cystinosis with an elevated WBC cystine level of 0.7 nmol ½ cystine/mg protein (NR <0.1). Despite advice to start immediate treatment, cysteamine was only initiated at 17 months of age and reached a therapeutic dose (1.33 g/m$^2$) at 22 months of age with WBC cystine levels thereafter between <0.1 and 0.3 nmol ½ cystine/mg protein.

Proteinuria improved after the initiation of cysteamine with a log plot of α1MCR vs ACR showing a linear progression towards the normal range suggesting an improvement in tubular proteinuria. The table shows a 75% reduction in PCR, a 79% in ACR, and a 64% decrease in α1MCR when comparing the pre-treatment with the late time period. There was also a reduction in the requirement for electrolyte supplementation with this effect being greatest for bicarbonate.

Proteinuria & Electrolyte Supplementation by Time Period

| | Duration | PCR | ACR | α1MCR | K | HCO3 | PO4 |
|---|---|---|---|---|---|---|---|
| | months | mg/mmol | | | mmol/kg/day | | |
| Pre-Cysteamine | 9.7 | 545 | 137 | 123 | 11.0 | 13.0 | 2.8 |
| Initiating Cysteamine | 4.6 | 345 | 109 | 125 | 12.3 | 9.9 | 2.1 |
| Post Cysteamine | 10.1 | 266 | 55 | 94 | 9.1 | 7.9 | 2.8 |
| Late | 25.3 | 133 | 29 | 44 | 8.7 | 5.4 | 2.1 |

**Conclusions:** These data suggest that there is an improvement in tubular function as measured by proteinuria and electrolyte supplementation after the start of cysteamine in cystinosis.

## Abstract# 635

**Study on Gene Polymorphism of Renin Angiotensin System in Chinese Children with Nephrotic Syndrome** F. Yang, W. Liu, P. Liu, G. He, Z. Guo, P. Liu. *Department of Pediatrics, The First Affiliated Hospital of Jinan University, Guangzhou, Guangdong, China.*

**Objectives:** In order to determine whether nephrotic syndrome (NS) in Chinese children associated with difference genotype of renin angiotensin system (RAS).

**Methods:** The $M^{235}T$ angiotensinogen( $M^{235}T$ AGT )gene, angiotensin II receptor type I(ATR1)gene, and angiotensin converting enzyme (ACE) gene were examined by PCR in 40 children with NS ( 29 patients were steroid sensitive, 11 patients were steroid resistant ) and in 50 children of health control group.

**Results:** In the NS group, the MM, MT and TT genotypes of $M^{235}T$ AGT were 37.5%, 27.5% and 35.0% respectively; the AA, AC and CC genotypes of ATR1 gene were 45.0%, 27.5% and 27.5% respectively. the DD, DI and II genotypes of ACE gene were 12.5%, 42.5% and 45.0% respectively;And these genotypes in the control group (in the same order) were 66.0%, 18.0% and 16.0%; 68.0%, 20.0% and 12.0%; 10.0%, 44.0% and 46.0% respectively. There were significant difference of AGT genotypes between the two groups ($\chi^2$=7.569, P<0.05), whereas there was no difference with respect to the ACE and ATR1 between the two groups ($\chi^2$=0.141, P>0.05;$\chi^2$=5.397, P>0.05). Additionally, there were no differences of the genotype distributions of AGT, ATR1 and ACE genes between steroid sensitive and resistant groups ($\chi^2$=2.600, P>0.05;$\chi^2$=2.610, P>0.05; $\chi^2$=2.235, P>0.05).

**Conclusions:** TT genotype of AGT gene may be associated with the pathogenesis of NS in Chinese children. The frequency of the gene polymorphisms of ATR1 and ACE gene are not associated with the pathogenesis of NS. The difference genotype of RAS related with the response to steroid therapy in the children with NS can not be demostrated.

## Abstract# 636

**Mutations in *WT1* and *PLCE1* in Familial Steroid-Resistant Nephrotic Syndrome in Chinese** Z. Yu, R. Fu, X. Chen, J. Wang. *Department of Pediatrics, Fuzhou Dongfang Hospital, Xiamen University, Fuzhou, Fujian, China.*

**Objectives:** Idiopathic nephrotic syndrome (INS) is the most common glomerular disease in childhood. Approximately 10-20% of children with INS are resistance to standard steroid treatment (this situation is called steroid-resistant nephrotic syndrome, SRNS) and may progress to end-stage renal failure. Recent studies have demonstrated that mutations in several genes cause SRNS in humans. This study aims to examine mutations in *WT1* and *PLCE1* in three Chinese families with SRNS once mutations in *NPHS2* had been excluded.

**Methods:** Blood samples were collected for genetic analysis from 3 probands from Chinese families with autosomal recessive SRNS, and their siblings and parents, and 50 adults with normal urinalysis. Genomic DNA was isolated from blood leucocytes. Ten exons, exon-intron boundaries of *WT1* and 31 exons, exon-intron boundaries of *PLCE1* were amplified by polymerase chain reaction. Mutational analysis was performed by DNA sequencing directly.

**Results:** No mutation of *WT1* and *PLCE1* in all exons and exon-intron boundaries was found in the 3 probands. However, three variants of *WT1* and 13 variants of *PLCE1* were identified in all 3 probands and some controls, indicating that these variants are polymorphisms. One novel variant of *WT1*, IVS5-64A>G, and one novel variant of *PLCE1*, IVS22-26T>A, were also identified in two probands as well as some controls. There was no significant difference in the allelic frequencies of the two polymorphisms of *WT1* and *PLCE1* between the patients and controls.

**Conclusions:** The results suggest that *WT1* and *PLCE1* mutations are not major causes of familial SRNS in Chinese in the study.

## Abstract# 637

**Mutations in *NPHS2* and *WT1* in Southern Chinese Children with Steroid-Resistant Nephrotic Syndrome** Z. Yu, J. Wang, R. Fu, L. Ye, X. Chen. *Department of Pediatrics, Fuzhou Dongfang Hospital, Xiamen University, Fuzhou, Fujian, China.*

**Objectives:** Idiopathic nephrotic syndrome is one of the most common glomerular diseases in Chinese children, of which 10-20% of cases show steroid resistance. To our knowledge, there were several studies regarding the incidence of mutations in *NPHS2* and *WT1* genes in childhood steroid-resistant nephrotic syndrome (SRNS). This study aims to examine mutations in both *NPHS2* and *WT1* in children with sporadic SRNS in Southern Chinese.

**Methods:** Peripheral blood samples were collected for genetic analysis from 20 children with sporadic SRNS in Southern Chinese, and 50 unrelated adult volunteers with normal urianalysis studied as controls. Genomic DNA was isolated from peripheral blood leucocytes. The mutational analysis of *NPHS2* and *WT1* gene was performed by polymerase chain reaction and DNA sequencing directly.

**Results:** No mutation in both *NPHS2* and *WT1* in their all exons and exon-intron boundaries was detected in the 20 patients with sporadic SRNS, whereas two polymorphisms of *NPHS2*, 102G>A and 954T>C, and 6 variants of *WT1*, 5'-UTR-7G>T, 126C>T, IVS3+16G>A, IVS5-64 A>G, 903A>G and IVS7-32C>A, were identified in some of the patients and the controls. There was no significant difference in the genotypic and allelic frequencies of 102G>A of *NPHS2*, and the 6 variants of *WT1* between the 20 patients and 50 controls, respectively.

**Conclusions:** Our results suggest that mutations in both the *NPHS2* gene and the *WT1* gene are not major causes of the children with sporadic SRNS in Southern Chinese in the study.

## Abstract# 638

**Variations of *NPHS2* and *WT1* in Six Children with Chronic Renal Failure in Chinese Han Ethnic Group** Z. Yu, J. Wang, R. Fu, L. Ye, X. Chen. *Department of Pediatrics, Fuzhou Dongfang Hospital, Xiamen University, Fuzhou, Fujian, China.*

**Objectives:** To examine genetic variations of *NPHS2* and *WT1* in children with chronic renal failure (CRF) in Chinese Han ethnic group.

**Methods:** Peripheral blood samples were collected for genetic analysis from 6 children with CRF in Chinese Han ethnic group, and 50 unrelated adult volunteers in Chinese Han ethnic group whose urinalysis was normal studied as controls. Genomic DNA was isolated from peripheral blood leucocytes. Eight exons and exon-intron boundaries of *NPHS2*, and 10 exons and exon-intron boundaries of *WT1* were amplified by polymerase chain reaction (PCR). Mutational analysis was performed by DNA sequencing directly and RFLP/PCR (RFLP, restriction fragment length polymorphism).

**Results:** No mutation in both *NPHS2* and *WT1* was detected in the 6 patients with CRF, whereas two already reported polymorphisms of *NPHS2*, 102G>A and 954T>C, and 4 variants of *WT1*, 5'-UTR-7G>T, 126C>T, IVS5-9T>C and 903A>G, were identified in some of the patients. Three already reported variants of *WT1* (5'-UTR-7G>T, 126C>T and 903A>G) were also identified in some controls, indicating that these variants are polymorphisms. There was no significant difference in the genotypic and allelic frequencies of 102G>A of *NPHS2*, and 5'-UTR-7G>T, 126C>T and 903A>G of *WT1* between the 6 patients and 50 controls, respectively. In addition, a novel variant, IVS5-9T>C of *WT1*, was identified in one patient and his father whose urinalysis was normal, whereas it was not found in 50 controls.

**Conclusions:** Mutations in both the *NPHS2* gene and the *WT1* gene are not major causes of the six children with CRF in the study.

## Abstract# 639

**A Heterzygous Mutation of *NPHS2*Identified in Chineses Child with Steroid-Dependent Nephrotic Syndrome** Z. Yu, R. Fu, X. Chen. *Department of Pediatrics, Fuzhou Dongfang Hospital, Xiamen University, Fuzhou, Fujian, China.*

**Objectives:** Idiopathic nephrotic syndrome is a common glomerular disease in children, of which 80% of cases show steroid sensitivity (SSNS). Most children with SSNS will have a frequently relapsing or steroid-dependent course (FRNS/SDNS) associated with therapy-related morbidity. Homozygous/compound heterozygous mutations in *NPHS2* are frequent cause of steroid-resistant nephrotic syndrome (SRNS), while an *NPHS2* mutation is absent from children with uncomplicated SSNS. Significant heterozygous mutations were identified in Italian children with FRNS/SDNS, whereas no heterozygous mutation was found in American children with the syndrome. However, whether or not heterozygous mutations of *NPHS2* may be causing SDNS in Chinese children has not been established. We examined a Chinese child with SDNS and focal segmental glomerulosclerosis for mutations in *NPHS2*.

**Methods:** The mutational analysis of *NPHS2* was performed by polymerase chain reaction and direct sequencing.

**Results:** A heterozygous missense mutation in exon 5 of *NPHS2*, 596A>T, which leads to an asparagine to isoleucine substitution (N199I) and is novel, was detected in the child, whereas it was not found in 100 chromosomes from 50 controls. A heterozygous silent mutation of 954T>C in exon 8 of *NPHS2* was also detected in the case.
**Conclusions:** A Chinese child with SDNS was identified with a heterozygous mutation of *NPHS2*, suggesting that heterozygous mutation of *NPHS2* may be associated with SDNS in the Chinese child.

## General Nephrology

### Abstract# 640
**(O-81)**
**Montelukast Sodium Reduces the Risk of Relapse in Nephrotic Syndrome** A.S. Abeyagunawardena. *Department Paediatrics, University of Peradeniya, Peradeniya, Sri Lanka.*
**Objectives:** Children with nephrotic syndrome the relapses are frequently triggered by viral upper respiratory tract infections, possibly mediated by cytokine release. Montelukast sodium has been successfully used to reduce the exacerbations of bronchial asthma due to viral infections in children. The hypothesis that montelukast sodium would reduce the incidence of viral infections thereby reduce the risk of relapse was tested by this study.
**Methods:** Sequential patients receiving low dose (<0.6mg/kg) prednisolone on alternate day with or without adjuvant therapy as maintenance were recruited. Montelukast sodium was prescribed for a period of one year at a dose of 4mg for children below 5 years and 8mg for above 5 years. The number of relapse episodes was compared for the twelve months before and after commencing on montelukast therapy. A freshly voided urine sample was tested each morning and the presence of 3 + proteinuria 3+ proteinuria for three consecutive days was diagnostic of relapse.
**Results:** 118 patients were recruited and 112 completed the study. 79 male ; 33 female. Age at entry ranged from 2.5 to 13.2 (median 6.2) years. The rates of relapse episodes before and after montelukast therapy were analysed by using a Poisson regression model. There was a significant reduction in the rate of relapse during the 12 months of montelukast therapy when compared to the 12 months before ($p<0.01$). The average yearly relapse episode rate was 1.42 during montelukast therapy compared with 2.51 for the 12 months before.
**Conclusions:** The results suggest that prescribing montelukast sodium can reduce the risk of relapse in frequently relapsing nephrotic syndrome. The results however need to be confirmed by randomised controlled studies.

### Abstract# 641
**(O-82)**
**Hemolytic Uremic Syndrome – Long Term Follow Up** R. Exeni, M.d.C Laso, I. Grimoldi, C. Exeni, A. Vazquez, A. Santiago, G. Neiro, K. Temperato, G. Perez y Gutierrez, K. Alvarez, S. Diaz, V. Zalabarria, A. Fernandez Couso. *Pediatric Nephrology, Hospital del Niño de San Justo, San Justo, Buenos Aires, Argentina.*
**Objectives:** Define the rate of sequealae in a longitudinal follow up study of patients with typical Hemolytic Uremic Syndrome (HUS).
**Methods:** 231 patients who survived diarrhoea + HUS were evaluated. The examinations were performed in patients whose HUS occurred at least 10 years before our study ( 10,8-.31 years)
**Results:** Patients were allocated into four groups defined by renal function. Group A (n=137, 59.3%) had normal kidney function, no proteinuria or hypertension. Group B (n=48, 20.8%) had normal kidney function, proteinuria, with or without hypertension. Group C (n=26, 11.2%) had creatinine clearance from 50 to70 ml/min/1.73, and proteinuria with or without hypertension. Group D (n=20, 8.7%) creatinine cleareance below 50 ml/min/1.73 m2). In the acute period 124 (23 %) had neurological manifestations Only 5 of the 231 patients had learning disorders, 1 had hemiparesis, and 1 chronic seizures.We found no correlation between the severity of the CNS involvement in the acute period and permanent neurological damage. Twelve (5.2%) of the 231 patients had hyperglycemia.
**Conclusions:** HUS is the most common cause of acute renal failure in our paediatric population and is a cause of acute mortality and chronic morbidity.41.7 % of our group of patients registered abnormal creatinine cleareance after 10.8 to 31 years of follow up. There is a low frequency of residual neurological, haematological, pancreatic, and ocular abnormalities.

### Abstract# 642
**(O-83)**
**Calculation of the Glomerular Filtration Rate (GFR) in Children: Cystatin Based Formula Versus Creatinine Based Formulae** J. Hoefele, M. Alberer, M.R. Benz, B. Lange-Sperandio, L.T. Weber. *Pediatric Nephrology, Children's University Hospital, Munich, Germany.*
**Objectives:** The inulin clearance is regarded as the gold standard measurement for the calculation of the kidney function in children, but it is time consuming and expensive. Simpler methods like the "old" serum creatinine based Schwartz-formula (OS) are available. GFR can also be calculated by cystatin-C based formulae e.g. the "new" Schwartz- (NS), LeBricon- (LB), Larsson- (L), and Rule-formula (R). The goal of this study was to compare the GFR calculated by using these four formulae with the OS-formula.
**Methods:** The GFR was calculated in 281 children with renal diseases using the above mentioned formulae (1003 measurements). Correlation analyses and analyses by the Bland-Altman-plot were performed.
**Results:** Although, there was a significant correlation between the GFR calculation according to NS, LB, L, R and OS, the Bland-Altman-plot showed a mean negative difference of GFR between these formulae and the OS-formula. A subgroup analysis of 266 patients with calculated creatinine clearance showed an overestimation of the GFR by the OS-formula and an underestimation by NS, LB, L and R.
**Conclusions:** The Bland-Altman-plot points to a definite overestimation of GFR-values by the OS-formula. GFR calculated by the NS-formula fits with the other cystatin-C based formulae, although there seems to be an underestimation of the GFR-values compared to the creatinine clearance. This study contributes to the discussion about the best surrogate marker of renal function in childhood and has validated the NS-formula in a large cohort of pediatric patients in comparison with the other existing formulae.

### Abstract# 643
**(O-84)**
**Prevalence of Microalbuminuria among Secondary School Children in Port Harcourt, Rivers State, Nigeria** A.N. Okpere, I.C. Anochie, F.U. Eke. *Pediatrics, University of Port Harcourt Teaching Hospital, Port Harcourt, Rivers, Nigeria.*
**Objectives:** To determine the prevalence and risk factors of microalbuminuria among secondary school children.
**Methods:** A cross-sectional study of 820 students aged 10-19 years was carried out between September 29 and November 10, 2008. The biodata, symptoms, medical and family history and social class of each subject were obtained. Physical examination was performed on all subjects. The 205 (25.0%) subjects with overt proteinuria and haematuria detected on dipstick urinalysis were excluded from analysis. The remaining 615 (75.0%) subjects formed the study population. Their random spot urine samples were tested for microalbuminuria using micral test strips. Data was analysed using computer softwares.
**Results:** There were 299 (48.6%) male and 316 (51.4%) female subjects. The prevalence of microalbuminuria was 33.2%. This was significantly ($p=0.000$) higher in females (45.3%). The prevalence increased with increasing age. It was high in obese subjects (35.4%) and higher (57.1%) when hypertension co-existed with obesity. Subjects with hypertension had a significantly ($p=0.001$) higher prevalence (70.6%) compared to those with normal blood pressure (32.1%). Subjects with family history of hypertension and diabetes mellitus had significantly ($p=0.000$ and 0.027 respectively) higher prevalence of 59.5% and 46.4% respectively compared to those without such family history. Microalbuminuria was present in 1 (50.0%) of 2 subjects with sickle cell anemia.
**Conclusions:** The prevalence of microalbuminuria in adolescents aged 10-19 years is high. The risk factors identified were obesity, hypertension, sickle cell anemia and family history of hypertension and diabetes mellitus.

### Abstract# 644
**(O-85)**
**Rituximab (RTX)-Associated Lung Injury (RALI)** M. Bitzan,[1] L. Carpineta,[1] J. Ouahed,[1] M. Anselmo,[1] F. Ghane Sharbaf.[1,2] *[1]Montreal Children's Hospital, Montreal, QC, Canada; [2]Dr. Sheikh Pediatric Hospital, Mashhad, Islamic Republic of Iran.*
**Objectives:** Unexpected infectious and pulmonary complications after B-cell depleting therapy (BCDT) are emerging medical concerns. Previously limited to older patients with B-cell malignancies, we report the first pediatric cases of RALI.
**Methods:** Case presentations & systematic literature review. Data reporting as median & interquartile range.
**Results:** Case 1. 14 yr-old boy with glucocorticoid-dependent nephrotic syndrome presented with fever and respiratory distress, associated with diffuse ground-glass alveolar infiltrates 18 days after the last RTX infusion. Patient recovered after 12 days and tolerated subsequent RTX treatment without pulmonary complications.

Case 2. 12 year-old girl with pulmonary nodules, histologically identified as cryptogenic organizing pneumonia, detected 8 wks after the last RTX infusion for post kidney transplant lymphoproliferative disease.
Systematic review of reported cases (93% with B-cell-related malignancies), diagnosed at age 64 (57-68) yrs, 75% male, revealed time to onset of 14 (11-22) days after last RTX treatment. 17 % had gradual onset, which worsened with continued rituximab infusions. 30% required mechanical ventilation, of whom >50% died.
**Conclusions:** RALI is an emergent complication of BCDT with increasing importance among (pediatric) renal patients. Diagnosis is confirmed by transthoracic lung biopsy. Current treatment recommendations are high-dose prednisone, although RALI may develop while patients receive glucocorticoids. Better outcomes in recent reports may be due to recognition of milder cases and/or earlier treatment. Recurrence risk with repeat courses of BCDT is unknown.

## Abstract# 645
**(O-86)**

**Risk Factors for Progressive Chronic Kidney Disease: Longitudinal Study of 5,351 Children with Heavy Proteinuria in Taiwan** C.-Y. Lin, C.-Y. Lee. *Division of Pediatric Nephrology, China Medical University and Hospital, Taichung, Taiwan.*
**Objectives:** We investigate those with heavy proteinuria and risk factors of renal progression in chronic kidney disease (CKD).
**Methods:** A prospective cohort of 5,351 children with heavy proteinuria detected by school urine screenings from 1992 to 1996 and followed to 2009. Main outcome measures including urinalysis, height, weight, blood pressure at baseline and 14 year follow-up; CKD risk factors including albumin, cholesterol, BMI, and systolic and diastolic pressure.
**Results:** There were 328 (6.13%) children in CKD Stage I; 2,931(54.77%) in Stage 2; 1,868 (34.91%) in Stage 3; 66(1.23%) in Stage 4 and 39 (2.22%) in Stage 5. Cumulative rate of end-stage renal disease (ESRD) and mortality increased, especially in CKD Stage 3. Comparing CKD group and non-CKD group children revealed no significant difference in biomarkers below 6 years old; at 7-12 years of age, the CKD group had markedly higher diastolic blood pressure, lower body length, low serum albumin and higher serum cholesterol levels. Among 13-to-17-year-olds, lower body length, lower serum albumin, lower total protein and higher cholesterol levels appeared in the CKD group; beyond 18 years old, besides the above-cited risk factors, higher fasting blood sugar was also noted.
**Conclusions:** At 7-17 years of age, glomerulonephritis (GN) with renal progression was the main cause of CKD. Above 17 years, metabolic syndrome became predominant. Under six years, mainly due to decreased renal mass with renal progression such as congenital urinary tract abnormalities. Early transfer to pediatric nephrologist, and individualized health care before CKD stage 3, may prevent or delay renal progression.

## Abstract# 646
**(O-87)**

**Early Detection and Quantification of Liver Fibrosis in Pediatric ARPKD Patients Using Transient Elastography (Fibroscan®)** S. Kummer,[1] S. Habbig,[2] G. Kircheis,[3] M. Feldkötter,[2] A. Sagir,[3] S. Pandey,[1] D. Ney,[1] B. Hoppe,[2] E. Mayatepek,[1] D. Haeussinger,[3] J. Oh.[1] *[1]General Pediatrics, University Hospital, Duesseldorf, Germany; [2]Pediatric Nephrology, University Hospital, Cologne, Germany; [3]Clinic for Gastroenterology, Hepatology and Infectiology, University Hospital, Duesseldorf, Germany.*
**Objectives:** Autosomal-recessive polycystic kidney disease (ARPKD) is an inherited progressive disease leading to ESRD. Almost all patients develop liver fibrosis that may require liver-kidney-transplantation. Early detection and quantification of fibrosis can provide useful information for management of patients with ARPKD.
**Methods:** We report the use of a new sonography based method (transient elastography, Fibroscan®) for early and noninvasive detection of liver fibrosis in patients suffering from ARPKD. This method is already well established in adult patients with hepatic disease, measuring ultrasound wave spreading as an indicator for tissue stiffness.
**Results:** ARPKD patients (n=7, mean age 9.5 years) showed a significantly increased liver stiffness compared to matched healthy controls (11.2 vs. 4.1 kPa; p=0.018). All patients had results above the upper reference value (7.5 kPa), while healthy controls and three patients with renal cysts of other reasons and no liver fibrosis were within normal reference values. Conventional sonography in ARPKD patients gave evidence for hepatic fibrosis in 4 patients, while 3 had still normal liver ultrasound.
**Conclusions:** The presented method provides an early sensitive non-invasive detection of liver fibrosis in children with ARPKD. This method could provide diagnostic information for unclear cystic renal diseases by identification of liver involvement.

## Abstract# 647
**(O-88)**

**Serum Cystatin C Based Prediction Equations for Glomerular Filtration Rate in Children with Early Chronic Kidney Disease** P. Hari,[1] A. Bagga,[1] R. Lakshmy,[2] Y. Anil.[1] *[1]Pediatrics, All India Institute of Medical Sciences (AIIMS), Delhi, India; [2]Cardiac Biochemistry, AIIMS, Delhi, India.*
**Objectives:** To estimate glomerular filtration rate using cystatin C based prediction equations and compare them with serum creatinine derived GFR for detection of early chronic kidney disease (CKD).
**Methods:** 111 children (age 2-17 yr) with GFR between 60-90 ml/min/1.73m$^2$ were studied. Serum cystatin C (CysC) and creatinine (Cr) were determined by immunonephelometry using DAKO PET kit and kinetic Jaffe method respectively. Creatinine derived GFR was estimated using 2 equations: (1) Schwartz (0.55 x height/Cr), (2) our creatinine formula (0.43 x height/Cr). GFR was derived from cystatin using 3 equations: (1) Larsson (99.43 x CysC $^{-1.5837}$), (2) Grubb (84.69 x CysC $^{-1.680}$ x 1.384), (3) Hojs (90.6 x Cys C$^{-1.192}$). $^{99}$Tc-DTPA clearance was used as reference GFR.
**Results:** The mean $^{99}$Tc-DTPA GFR was 76.5 ml/min/1.73m$^2$; mean serum Cys C level was 0.86 (range 0.5–1.9 mg/L)

Comparison of Cys C and creatinine based formulae with 99Tc-DTPA GFR

| Formula | Bias ml/min/ 1.73m2 | Precision ml/min/ 1.73m2 | Accuracy within 30% of DTPA GFR | P * |
|---|---|---|---|---|
| Schwartz | 22.9 | 34.5 | 52.9 | <0.0001 |
| Schwartz | -3.1 | 23.3 | 79.3 | |
| Larsson | 70.7 | 56.8 | 17.7 | <0.0001 |
| Grubb | 101.9 | 72.9 | 11.6 | <0.0001 |
| Hojs | 43.7 | 35.6 | 24.8 | <0.0001 |

*Compared to our creatinine formula All equations overestimated the measured GFR except our creatinine formula which had the least bias, best precision and highest accuracy. Cys C based equations correctly predicted CKD stage II in 6-15% patients as compared to 87% by our formula.
**Conclusions:** Cystatin C based prediction equations do not reliably predict early CKD.

## Abstract# 648
**(O-119)**

**Cytoskeleton Rearrangement Induced by EGF Via Activation of RhoA Signal Pathway in Podocytes** Q. Feng, J. Ding, J. Miao, L. Jiang. *Department of Pediatrics, Peking University First Hospital, Beijing, China.*
**Objectives:** Effacement of podocyte foot processes occurs in many proteinuric nephropathies and is accompanied with rearrangement of the actin cytoskeleton. In this study the intracellular pathways of EGF in podocytes, which induced abnormality of cytoskeletal architecture and dysfunction of podocytes, was investigated.
**Methods:** EGF expression was detected both in nephrotic rat model and podocyte injury induced by puromycin. F-actin distribution, nephrin expression, and RhoA activation as well, were detected in mouse podocytes after EGF treatment.
**Results:** Both in nephrotic rat model and podocyte injury induced by puromycin, an increased EGF expression was revealed. In addition, redistribution of F-actin fibers and down-regulation of nephrin expression in mouse podocytes were also detected with the treatment of EGF. Meanwhile the increased RhoA activity was revealed in podocytes exposed to EGF. The rearrangement of F-actin as well as the decreased nephrin expression could be rescued by the addition of Y27632, a reagent to inhibit Rho kinase-dependent stress fiber formation.
**Conclusions:** The results from this study suggested a role of EGF in the regulation of the cytoskeleton arrangement, nephrin expression and podocyte dysfunction, which might involve the activation of RhoA.

## Abstract# 649

**Acute Kidney Injury in Non-Renal Cases** K.P. Mehta,[1] A. Deokar,[2] A.A. Bagde,[1] N.P. Shenvi,[1] B. Dalvi.[2] *[1]Dept. of Pediatrics, Jaslok Hospital & Research Centre, Mumbai, Maharashtra, India; [2]Dept. of Cardiology, Nanavati Hospital, Mumbai, India.*
**Objectives:** Kidney is extremely vulnerable to insults by co-morbid conditions.In this paper 2 unusual non renal cases with
baseline normal urine UO[40] and normal s.creatinine values,are presented.
**Methods:** Case 1
6 month old girl diagnosed as patent ductus arteriosus[PDA] on2 -D-Echo underwent transcatheter PDA closure. On D- 5 murmur reappeared due to displacement of the device in descending arota which was removed via rt.Femoral artery.The procedure lasted for 8-10 hours during which S.Cr.went up from 0.5 to 2.9mg/dl and UO went down to 0.5ml/kg/hr over 32 hours.Peritoneal dialysis was given for 36 hours.UO increased to 2.2ml/kg/hr and S.Cr went down to 0.4mg/dl at 72 hrs,child was discharged on oral feeds.
Case 2

2 year old boy of Budd Chiari Syndrome under went transjugular intrahepatic portosystemic shunt[TI PASS] during which 200ml of radio contrast [ osmolality 600 mosm/kg was used.On day -2, S Cr.went up from 0.5mg to2.9mg/dl, but UO put was normal.S.Potassium[K+] went up to 7.4meq/L with cardiac arrythmias. PD was started.6 cycles with short dwelve[ 10-15 mins] time given to normalise S.K+[4.2meq/l] PD was stopped due to bleeding with sudden low HB and hyotension. A large hematoma in the capsule of liver was evacuted after stopping heparin.On D-10 postoperative renal parameters were normal and baby was discharged with resolving ascities and weight gain on D-25.
**Conclusions:** Both non renal cases with AKI were managed in PICU with simple monitoring of UO on hourly basis and S.Cr every 6- 12 hourly to detect AKI. ASAP timely intervention with PD in both the cases.

## Abstract# 650

**Changes of Urinary Enzymes as Test of Nephropathy in Children with Obstructive Megaureter** G. Nikulina, L. Migal, I. Serbina. *Laboratory Biochemistry, State Institution "Institute of Urology, Academy of Medical Sciences of Ukraine", Kyiv, Ukraine.*
**Objectives:** The aim was to compare the enzyme activities of renal specific brush edge gamma-glutamiltranspeptidase (GGT), neutral alpha-glucosidase (NGL) and lysosomal N-acetyl-beta-D-glucosaminidase (NAG) in urine of 25 healthy children and 44 little patients with obstructive megaureter (OMU).
**Methods:** In our work we used the biochemical methods of investigation of urine for diagnistics kidney pathology at OMU in children.
**Results:** It was estimated that in healthy children urine levels of GGT, NGL and NAG were accordantly 22.9±2.0, 84.9±6.4 and 11.6±0.7 mcmol/h/mmol Creatinine. In group of patients with OMU the enzyme activities increased: GGT – up to 91.2±6.7, NGL – up to 200.7±13.2 and NAG – up to 31.1±1.4 against to control group ($p < 0.001$).
**Conclusions:** So the detected changes of enzyme findings under study showed on the tubular nephrothelium damage and development of nephropathy at OMU in children.

## Abstract# 651

**Renal Vein Obstruction and Postural Proteinuria: A Systematic Review of the Literature** G.P. Milani,[1] M.C. Mazzoni,[1] M.G. Bianchetti,[2] E.F. Fossali.[1] *[1]Pediatric Emergency Department, Fondazione IRCCS Cà Granda, Ospedale Maggiore Policlinico, Milan, Italy; [2]Division of Pediatrics, Ospedale San Giovanni, Bellinzona, Switzerland.*
**Objectives:** The cause of postural proteinuria is not clear but may often relate to obstruction of the left renal vein in the fork between the aorta and the superior mesenteric artery (= renal nutcracker). However, reports dealing with proteinuria only marginally refer to this cause of postural proteinuria. We performed a formal analysis of the literature.
**Methods:** All reports published in peer-reviewed journals through 1992 were considered. For the final analysis, we retained 13 reports providing details on renal nutcracker in postural proteinuria.
**Results:** Five reports addressed the frequency of renal nutcracker in a total of 229 subjects with postural proteinuria. Their aged ranged between 5 and 17 years (female-to-male ratio: 1.05:1.00. Imaging studies demonstrated renal nutcracker in 156 (68 percent) subjects. Renal nutcracker was also demonstrated in 10 anecdotal reports for a total of 51 subjects with postural proteinuria. Very recently, 13 Italian subjects with postural proteinuria associated with renal nutcracker were reassessed ≥6 years after the initial diagnosis: in 9 subjects both postural proteinuria and renal nutcracker had disappeared, in 3 subjects both postural proteinuria and renal nutcracker had persisted, and in one case postural proteinuria had persisted unassociated with renal nutcracker.
**Conclusions:** This review of the literature provides substantial support for renal nutcracker as the most common and benign cause of postural proteinuria.

## Abstract# 652

**Hemolytic Uremic Syndrome (HUS) in Children – Follow-Up and Prognosis** A. Moczulska, K. Zachwieja, E. Slowiaczek, D. Drozdz, I. Ogarek, K. Wilkosz, Z. Stec, J. Kwinta-Rybicka, M. Miklaszewska, J.A. Pietrzyk. *Dpt of Pediatric Nephrology and Dialysis Unit, Collegium Medicum Jagiellonian University, Cracow, Poland.*
**Objectives:** HUS is a common cause of acute kidney injury in children. The aim of the study was to analyze the clinical course and prognosis in children with HUS.
**Methods:** The study group consisted of 23 children with HUS treated 1996-2009, 11 girls and 12 boys aged 3 months - 9 yrs ( 3 yrs 3 m). 14 pts had typical D+HUS (preceded by diarrhea), in 9 atypical D-HUS was diagnosed. The clinical course was analyzed after mean 4,2±4 yrs of follow-up (3 m - 13 yrs).
**Results:** The general condition of patient at admission was severe in 17/23 (74%) cases (7 neurological signs, 2 pancreatitis). In 5/14 D+ pts enterohemorrhagic E.coli was diagnosed. In D-HUS pts 1 tonsillitis, 1 streptococcal sepsis, 1 varicella, 4 respiratory tract infection were diagnosed. The renal replacement therapy (RRT) was introduced 1,8±1,5 days after admission in 16/23 pts (4 HD a. 12 PD). Anuria occured in 12/23 patients for 5-49 days (mean 16±12). 3/14 D+ and 4/9 D-HUS patients improved without RRT. Plasmapheresis was performed in 3/14 D+ and in 6/9 D- pts (with dialysis 2 and 4 respectively). another 10 pts received fresh frozen plasma (8D+, 2D-).
**Conclusions:** Chronic kidney disfunction is diagnosed in 7/23 patients (30,4%): - 5 pts (3D+ and 2 D-HUS) stage 2-3; 2 pts (D-) end-stage (8,7%). The anuria time was important, but not significant (p=0,07) predictor for CKD. One patient (D-) died during plasmapheresis (TRALI reaction). Two patients after recovery stay hypertensive. Patients who did not require RRT, had no long-term complications (3D+, 2D-).

## Abstract# 653

**Evaluation of Childhood Urinary Stone in Gorgan, Iran** M. Moghtaderi, H. Pasha Soltani. *Pediatrics, Gorgan University of Medical Science, Gorgan, Islamic Republic of Iran.*
**Objectives:** Evaluating presenting features, predisposing factors, and medical treatment strategies used in paediatric renal stones in Gorgan,Iran.
**Methods:** A total of 71 children presented with a urinary tract renal stone, to the center over one year period. All children were reviewed in nephrology clinic and fully investigated. Treatment was assessed by retrospective hospital note review.
**Results:** Metabolic abnormality was found in 35.6% of children, 30% were classified as infective, and46% idiopathic.
Bilateral stones on presentation occurred in 49.1% of the metabolic group compared to 12% in the
infective/idiopathic group Coexisting urinary tract infection was common (23%) in the metabolic group.
Surgically, minimally invasive techniques (lithotripsy, percutaneous nephrolithotomy, and endoscopy) were used in 1 of patients. Urinary stones were more common in girls than boys (57.6 versus 42.4). Mean age of patients were 37.6 months and the most important complaint was abdominal pain and/or hematuria. Mean size of stones was 35.5+/- 2.7 mm. 87.7 percent of patients responded to medical therapy. Conclusions: Underlying metabolic causes are now the most common but can be masked by coexisting urinary tract infection. All children with renal stones should have a metabolic screen. Early detection of renal stone and finding of predisposing factor can prevent serious complications.
**Conclusions:** Underlying metabolic causes are now the most common but can be masked by coexisting urinary tract infection. All children with renal stones should have a metabolic screen. Early detection of renal stone and finding of predisposing factor can prevent serious complications.

## Abstract# 654

**Incidence of Microscopic Haematuria in School Age Children in Gorgan, Iran** M. Moghtaderi, A. Noohi. *Pediatrics, Gorgan University of Medical Science, Gorgan, Islamic Republic of Iran; Pediatrics, Gorgan University of Medical Science, Gorgan, Islamic Republic of Iran.*
**Objectives:** Persistent microscopic haematuria in children and may be a clue of a serious illness.
**Methods:** In cross section study we evaluated 3000 children 6-14 year old from October 2008 to June 2009. A fresh morning urine sample was examined in hospital laboratory with dipstick. Positive cases were asked to repeat it twice with 2 weeks interval. Urine was examined for pr, dysmorphic Rbc, Ca, uric acid, Cr, oxalates, and cultured.kidney sonography was applied to positive cases.
**Results:** This study was done for 3000 school age children in Gorgan city, north east of Iran. Age range was 6-14 year. with mean age 9.7 +/_ 1.97 year. 1200 of them were male. In the first stage 208 (6.9%) cases had microscopic haematuria, after repeating the dipstick exam 2 times later we had 35 (1.16%) positive cases. There was 27 female (77.1%) and 8 males (22.9%). 9 cases did not follow the study. 13 of 26 positive students had nephrolithiasis 12 of them had normal sonography and1 case had a large cyst that evaluated with CT scan and then removed surgically. Pathologic diagnosis was oncocytoma. Hypercalciuria was seen in 8 cases (22.9%). Hyperuricosuria was reported in 5 cases (14.3%) and there was pyuria in 2 cases (5.7%) and documented UTI was seen in 1 student. In 51.4% of patients had familial history of nephrolithiasis and 11.4% had family history of renal disease. One student had hearing defect. 14.3% of cases had symptoms the most common of them was abdominal pain.
**Conclusions:** Routine screening of microscopic hematuria in school age children can help us to early diagnose and manage most of renal disease early, such as our study.

## Abstract# 655

**Spectrum of Biopsy Proven Renal Diseases in the Paediatric Age Group – A Single Center Experience** A. Mohapatra,[1] G. Basu,[1] V.M. Annapandian,[1] V.G. David,[1] S. Madhivanan,[1] S. Varughese,[1] A. Korula,[2] G.T. John,[1] C.K. Jacob,[1] V. Tamilarasi.[1] *[1]Nephrology, Christian Medical College, Vellore, Tamil Nadu, India; [2]Pathology, Christian Medical College, Vellore, Tamil Nadu, India.*

**Objectives:** To study the distribution of various renal diseases in pediatric age group & to analyze patient characteristics, clinical and biochemical parameters among common biopsy proven renal diseases.

**Methods:** The study includes children ≤18 years over a period of 12years(1997 - 2008) who underwent renal biopsy for various indications. Out of a total of 1649 children who underwent renal biopsy, complete data were available for 1049 children which were analyzed for this study.

**Results:** Among 1049 children who underwent renal biopsy, the mean age was 12.0±5.6 years, M:F=1.7:1,The mean creatinine & serum albumin were 1.2 ±1.9 mg% &2.7
±1.1gm%. Distribution of various common diseases among children were as follows:Minimal change disease in 329 (31.3%), Mesangialproliferative GN in 193 (18.3%), Lupus nephritis in 120 (8.0%), Proliferative GN in 103(9.8%), IgA nephropathy in 72 (6.8%), Focal segmental glomerulosclerosis in71(6.7%), Diffuse mesangial hypercellularity in 53 (3.5%),Membranous glomerulonephropathy in 33(3.1%), Crescentic glomerulonephritis in 15(1.69%), MPGN in 12(1.2 %), end stage disease of unknown etiology in 22(2.1%) of case.

**Conclusions:** Minimal change disease was the most common entity among the children under 8 years of age, whereas FSGS predominated in children with age greater than 8 years. The age at onset of nephrotic syndrome was significantly higher in the non-MCD group than the MCD group.MPGN showed decreasing trend in the study.

## Abstract# 656

**Infliximab (Anti-TNFalfa) as Rescue Therapy in Parvovirus B19-Associated Vasculitis** E. Benetti, F. Scozzola, E. Vidal, D. Meneghesso, G. Ghirardo, M. Della Vella, L. Murer. *1Pediatric Nephrology, Dialysis and Transplant Unit, Department of Pediatrics, University of Padua, Padua, Italy.*

**Objectives:** Parvovirus B19 (ParvoB19) infection has been reported in some types of vasculitis such as Henoch-Schonlein purpura (HSP). Renal involvement may be induced by both an indirect (immunocomplex) and direct (TNF-alfa-mediated) mechanism.

**Methods:** In a 6-months period in 2009, 5 children with HSP and persistent proteinuria or nephritic/nephrotic syndrome underwent renal biopsy in our Unit. We screened blood and renal biopsy samples of the patients for ParvoB19 and other viruses (EBV, CMV, Adenovirus, HHV6 and 8).

**Results:** ParvoB19-DNA was positive in both blood and renal tissue in 2 children. In both boys, an upper respiratory tract infection had preceded the onset of symptoms. The first boy (4 years) had a histological picture of mesangio-proliferative glomerulonephritis. After two i.v. Ig courses, viraemia disappeared in 2 weeks and proteinuria in 2 months.
The second boy (5 years), had a histological picture of mesangial glomerulonephritis with crescents in 80% of glomeruli. Viraemia and proteinuria failed to decrease after i.v. Ig injection, prednisone, plasmapheresis with oral cyclophosphamide, and mofetil-micofenolate therapy. Anti-TNF-alfa-antibody (Infliximab) was then administered. Proteinuria desappeared in 6 months and viraemia after 9 months. No crescents were detected in the control biopsy.

**Conclusions:** We suggest a screen for viral infections in patients with acute vasculitis-associated renal involvement. In ParvoB19 associated forms, Infliximab may be used as rescue therapy in patients with glomerulonephritis who do not respond to conventional therapy.

## Abstract# 657

**Identification of Risk Factors for Chronic Renal Disease among Schoolchildren** A.V. Naghettini,[1] C.M. Salgado,[2] L.M.R. Salgado,[3] J. Freitas.[4] *[1]Pediatric, Universidade Federal de Goiás, Goiânia, Goias, Brazil; [2]Pediatric, Universidade Federal de Goiás, Goiânia, Goias, Brazil; [3]Pediatric, Universidade Federal de Goiás, Goiânia, Goias, Brazil; [4]Pediatric, Universidade Federal de Goiás, Goiânia, Goias, Brazil.*

**Objectives:** Assess the frequency of risk factors–systemic arterial hypertension, obesity, low birth weight, voiding dysfunction, diabetes, or renal disease in the family–for developing renal disease and their correlations.

**Methods:** A cross-sectional population study was carried out in six municipal schools of the eastern region of Goiânia, Brazil. We calculated a sample of 274 schoolchildren, with margin of error ranging from 4% to 4.5%. Factors investigated: history of urinary tract infection (UTI), enuresis, voiding dysfunction, family disease, arterial blood pressure, waist and hip circumference, hematuria and proteinuria, body mass index (BMI).Statistical analyses included calculating means, standard deviation, and frequency,Fisher's exact test and Pearson's correlation coefficient($P \leq 0.05$).

**Results:** We evaluated 274 children, with 8.06 ± 1.33 years, among which 139 (50.7%) were female and detected: history of UTI in 45 (16.4%); enuresis in 50 (18.2%); voiding dysfunction 83 (30.3%); BMI above P85 in 18.8% (45); waist circumference above P90 in 17 (6.9%); arterial blood pressure above P95 in 17 (7.1%).Also, we observed correlation between arterial blood pressure, obesity, and increased waist circumference.Enuresis and voiding dysfunction were factors associated to history of UTI($P< 0.05$).

**Conclusions:** This study demonstrated high frequency of risk factors for chronic renal disease in a schoolchildren population, with special attention to voiding dysfunction and increased BMI.

## Abstract# 658

**The Effect of ACEI and ARB on Serum Potassium Concentrations in a Pediatric Nephrology Outpatient Population** A.B. Nayak,[1] S.J. Wassner,[1] D. Kees-Folts,[1] M.L. Shaffer.[2] *[1]Pediatrics, Penn State Hershey Children's Hospital, Hershey, PA, United States; [2]Health & Information Science, Penn State Hershey Children's Hospital, Hershey, PA, United States.*

**Objectives:** There is little published data on whether the administration of angiotensin converting enzyme inhibitors (ACEI) or angiotensin receptor blockers (ARB) affect serum potassium concentrations [K] in pediatric patients. Our objective was to retrospectively assess the effects of ACEI or ARBs on [K] in a pediatric population within a single pediatric renal outpatient clinic.

**Methods:** 110/469 pts had serum [K]drawn 0-3m before and after starting ACEI/ARB therapy. All ACEI/ARB doses were converted to "enalapril or losartan equivalents" as a ratio of the highest doses noted in the PDR for the respective ACEI/ARB. To adjust for those who started/restarted ACEI/ARB therapy multiple times, linear mixed-effects models were utilized.

**Results:** The age range was 0.1-19.8 yrs; 53% were male. Most patients were treated with enalapril (63), lisinopril (23) or valsartan (13). The median number of days [K] was measured before and after starting therapy were 0 & 27 respectively. For all patients, pre-therapy [K] averaged 4.4 $\pm$ 0.7 and post-therapy [K] averaged 4.4 $\pm$ 0.9 mEq/L (p= 0.65). There was no significant difference noted when the ACEI or ARB groups were evaluated separately and no relation between delta [K] and mg/kg dosing. Neither eGFR nor furosemide use were significant. For children with eGFR values < 90 mL/min/1.73m$^2$ (81 records/59 pts) the delta [K] was 0.05 mmol/L (p =0.53).

**Conclusions:** In our patients there was no relationship between ACEI/ARB administration, either as total dose or as mg/kg, and change in [K].

## Abstract# 659

**Prevalence of Proteinuria and Microalbuminuria in HIV-Infected Children Admitted to Hospital** P. Nourse,[1,3] P. Gajjar,[1] C. Esezobor,[2] M. McCulloch.[1] *[1]Pediatrics, School of Child and Adolescent Health, University of Cape Town, Cape Town, South Africa; [2]Pediatrics, College of Medicine, University of Lagos, Lagos, Nigeria; [3]Pediatrics, University of Stellenbosch, Cape Town, South Africa.*

**Objectives:** To document the prevalence of proteinuria and microalbuminuria in HIV infected children.

**Methods:** Consecutive HIV-infected children aged zero to 16 years admitted to our institution whose parents/guardian gave written informed consent were studied. From each child demographics and HIV clinical and immunological parameters were documented. Urine was tested with dipstick for protein and if present was sent for protein-creatinine ratio; if negative it was tested with dipstick for microalbuminuria. Urine samples testing positive for microalbuminuria were sent for microalbumin-creatinine ratio. Immunological staging of HIV disease was done using the CDC criteria. Continuous and dichotonuous data were summarized as mean ±SD and proportions respectively. Univariate analysis was performed were appropriate.

**Results:** Thirty-six children aged 0.2 to 10.9 years (2.8 ±3.1) with 58.3% males were studied. Fourteen (38.9%) had advanced immunosuppression and 17 (47.2%) were on HAART. The frequency of proteinuria, nephrotic range proteinuria and microalbuminuria were 38.9%, 11.1% and 11.1% respectively. The frequency of proteinuria was not statistically different across gender, immunological stage of HIV disease, use of HAART or otherwise and age ≤2 years or older.

**Conclusions:** Proteinuria is common among HIV-infected children admitted to Hospital. Microalbuminuria is also more common than the general population and may be an early marker of subclinical renal disease.

## Abstract# 660

**A Patient with Marked Hyper-Immunoglobulinemia E Showing Minimal Change Nephrotic Syndrome and a STAT3 Gene Mutation**
M. Okada, K. Miyazaki, T. Miyazawa, K. Sugimoto, S. Fujita, H. Yanagida, T. Takemura. *Department of Pediatrics, Kinki University School of Medicine, Osaka-sayama, Osaka, Japan.*
**Objectives:** We encountered a patient with marked hyper-immunoglobulinemia E who had a mutation of the signal transducer and activator of transcription 3 gene (*STAT3*) and developed minimal change nephrotic syndrome (MCNS).
**Methods:** From early infancy, the patient showed repeated episodes of refractory chronic eczema accompanied by impetigo vulgaris with cicatrisation, as well as otitis media. Serum IgE was markedly increased (from 4000 to 25000 IU/mL). The nephrotic syndrome (NS) frequently relapsed, and was alternately responsive and resistant to corticosteroids.
**Results:** The *STAT3* mutation was heterozygous, located in exon 23 of the transactivation domain and causing A744V substitution. Presently treated with mycophenolate mofetil, the patient has less frequent MCNS recurrences. Increases in circulating Th2 cytokines and IgE combined with suppression of the Th1 cytokine interferon-g caused by the *STAT3* abnormality, presumably caused MCNS by altering the Th1/Th2 balance among T-lymphocytes.
**Conclusions:** To our knowledge, this is the first report of type I hyper-IgE syndrome (HIES) showing a STAT3 gene mutation and MCNS.

## Abstract# 661

**Urine Cotinine, Second Hand Smoking [SHS] Exposure and Adolescent Chronic Kidney Disease [CKD]**
A. Omoloja,[1] M. Ferris,[1] L. Greenbaum,[1] A. Wilson,[1] V. Bastian,[1] D. Chand,[1] A. Stolfi,[2] H. Patel.[1] *[1]Midwest Pediatric Nephrology Consortium, Columbus, OH, United States; [2]Pediatrics, Wright State University, Dayton, OH, United States.*
**Objectives:** To evaluate exposure to SHS in adolescents with CKD using urine cotinine levels.
**Methods:** Subjects aged 13-18 years with CKD were recruited from pediatric nephrology clinics. Subjects anonymously answered a 12-item survey about smoking habits and exposure. They also provided a freshly voided urine sample that was evaluated for urine cotinine. Log transformed urine cotinine levels (ng/ml) for self-identified non-smokers were compared among subjects grouped by SHS based on if they lived with a smoker (LY) or not (LN) and/or had friends who smoke (FY) or not (FN). Group comparisons were made with two-sample t tests or analysis of variance.
**Results:** Cotinine levels from 102 non-smokers were analyzed, 46% lived with a smoker and 50% had friends that smoked. Median urine cotinine levels were significantly higher in the LY group vs. LN group [3.7 vs. 0.5, P<.001]. Levels were also higher in FY vs. FN subjects [2.0 vs. 0.9, P=.03]. There was a significant incremental increase in urine cotinine levels with increasing SHS exposure: levels were 0.3 for LNFN subjects, 0.6 for LNFY, 1.6 for LYFN, and 5.5 for LYFY (P<.001).
**Conclusions:** In adolescents with CKD urine cotinine levels confirm that SHS may be another important variable to consider when evaluating CKD progression and cardiovascular outcomes.

## Abstract# 662

**Case Report – Cerebral Venous Thromboses in a Child with Nephrotic Syndrome**
O. Oniyangi,[1] D. Hahn,[2] K. Petersen.[2] *[1]Deaprtment of Paediatrics, National Hospital, Garki, Abuja, Nigeria; [2]Department of Paediatric Nephrology, Johannesburg Hospital, Johannesburg Private Bag X39, South Africa.*
**Objectives:** Nephrotic syndrome is associated with several complications among which are thrombo embolic phenomena. These are uncommon in children.
This report describes an 8 year old male child with relapse of steroid resistant minimal change nephrotic syndrome who developed cerebral sagittal and transverse sinus thromboses. He presented with headaches, vomiting and photophobia; and developed VI cranial nerve palsy during the course of the illness. Diagnosis was made by Computed Tomographic Scan and Magnetic Resonance Angiography of the brain. He was treated with low molecular weight heparin initially and then, oral warfarin with close monitoring of the INR and Factor Xa; and mannitol. His prednisolone and cyclosporine A treatments for NS were continued. He recovered without neurological deficits from the venous thromboses, and with marked improvements in his radiological features.

## Abstract# 663

**Renal Functional Reserve in Children with Single Functioning Kidney**
A. Peco-Antic,[1] D. Paripovic,[1] G. Scekic,[2] D. Kruscic.[1] *[1]Nephrology Department, University Children's Hospital, Belgrade, Serbia; [2]Pediatrics, KBC, Zemun, Serbia.*
**Objectives:** To evaluate functional response of kidney to oral protein load (OPL) in children with single functioning kidney (SFK).
**Methods:** RFR was measured during 45 studies in 37 children (25 boys) median age of 10 years (range 2,5-16) as the difference between the baseline clearance of creatinine (CsCr) and CsCr following a meat-free protein meal (1 g/kg body weight). Most patients had congenital SFK due to renal agenesis (54%) or multicystic dysplastic kidney (22%), while remaining patients had nephrectomy. Cimetidine was given 48h prior to the study at daily dose of 12 ±0.75 mg/kg, while they were on a diet free of meat, fish and fowl. Ambulatory blood pressure (ABP) monitoring was performed with Spacelab 90207. Serum Cystatin C was examined before and 2 hours after OPL and urinary protein/creatinine (mg/mg) was determined from accurately timed urine collection of approximately 2 h duration pre and post OPL. Data were expressed as mean±SEM.
**Results:** After OPL CsCr (92.81± 4.95 vs 81.04±4.72) and proteinuria/creatinuria (0.32±0.05 vs 0.23± 0.02) significantly increased while serum cystatin C remained unchanged. There was no difference in RFR between patients with congenital and acquired SFK(4.27±7.55 vs 6.72±11.96). RFR was inversely correlated with stimulated proteinuria, but not with ABP.
**Conclusions:** The defective renal response to OPL manifested by reduced RFR and increased proteinuria may be markers of glomerular hyperfiltration in children with single functioning kidney. No change in serum cystatin C after protein load was observed. The lack of correlation between RFR and ABP suggest that RFR is not directly modified by blood pressure.

## Abstract# 664

**Renal Involvement in Obese Children and Adolescents**
A. Savino, P. Pelliccia, C. Cecamore, C. Giannini, T. de Giorgis, A. Mohn, F. Chiarelli. *Department of Pediatrics, University of Chieti, Chieti, Italy.*
**Objectives:** The present study aimed to investigate whether renal function indexes (GFR estimated by the Schwartz formula -eGFR-, cystatin C -CysC-, albumin excretion rate -AER-), nitric oxide (-NO- an important modulator of renal function and morphology), urinary isoprostanes (a marker of oxidative stress) and blood pressure (BP) modifications can be detected in obese children and adolescents when compared to normal weight controls.
**Methods:** 107 obese children and adolescents and 50 controls were evaluated. Blood and urinary samples were collected to evaluate markers of renal function, serum and urinary NO and urinary isoprostanes. Ambulatory BP monitoring was performed in all. Obesity and insulin resistance (IR) degree were expressed by SDS-BMI and HOMA-IR.
**Results:** CysC and e-GFR did not significant differ between the 2 groups. AER was increased in obese children. CysC and GFR were related to HOMA-IR, AER was related to SDS-BMI and HOMA-IR. Significantly reduced NO levels and increased urinary isoprostanes and BP measurement were observed in obese children. All were significantly related to adiposity and IR indexes.
**Conclusions:** Difference seen in AER supported a role of obesity and IR in the genesis of glomerular leaking of albumin; we also observed differences in NO and urinary isoprostanes, but they were probably related to adiposity and IR rather than represent a clear risk factor for renal disease.
Renal involvement is not an early manifestation of obesity in childhood, since no relevant differences can be detected when comparing renal function indexes of obese and non-obese children. A longer exposure to obesity and IR is probably needed.

## Abstract# 665

**Effects of Prematurity on Blood Pressure and Renal Functions in Medium- and Long-Term**
I.E. Özden,[1] H.M. Poyrazoglu,[1] A. Öztürk,[2] F. Bastug.[1] *[1]Pediatric Nephrology, Erciyes University Medical Faculty, Kayseri, Turkey; [2]Neonatology, Erciyes University Medical Faculty, Kayseri, Turkey.*
**Objectives:** We aimed to asses effects of prematurity and on blood pressure and renal functions in medium- and long-term by evaluating the children older than 5 years who were previously monitered for prematurity.
**Methods:** One hundred and four children who were born as premature between 2000 and 2004 were enrolled in this study. All children were examined, blood pressures were measured and renal function tests were calculated by analysing urine and blood chemistry.
**Results:** The mean age of 104 children (56 boys, 48 girls) included the study was 8.1±0.9 years. Growth retardation was found in three and obesity in five of the children. Blood urine nitrogen (BUN) and serum creatinine levels were in normal ranges in all children. Six children had mild reduction in GFR. Increased phosphorus excreation in urine was found in two children, natriuria in four, kaliuria in eight, mild proteinuria in 11. In the ultrasound evaluation of 208 kidneys, renal heights in 59 kidneys were found smaller than expected values in same age group. At causal blood pressure measurement, all children were normotensive. However, at ambulatory blood pressure monitoring, mean systolic blood pressure was higher than 95 percentile in 10 children, mean diastolic blood pressure in 14 children and mean arterial pressure in nine children.

**Conclusions:** Children who were born as premature may have increased blood pressure and impaired renal functions in advancing years. Periodic monitoring of blood pressure and renal functions may be helpful to early detection of possible hypertension and renal dysfunction.

## Abstract# 666

**Compliance with H1N1 Immunization in Children with Chronic Renal Disease** N. Printza, E. Farmaki, J. Bosdou, C. Goga, F. Koukourgianni, F. Papachristou. *1st Pediatric Department, Aristotle University, Thessaloniki, Greece.*

**Objectives:** WHO recommended the immunization of children with chronic renal disease (CKD) and immunosuppressed patients as the most effective way to reduce H1N1 morbidity and mortality. Aim of our study was to evaluate the compliance of our patients with H1N1 vaccination.

**Methods:** A total of 64 eligible children/parents were approached to fill in a standardised questionnaire on influenza immunization profile and kind of influence on being or not vaccinated with H1N1 vaccine.

**Results:** Patients aged 1.5-18 years were categorized in four groups as follows: transplant recepients (TR) 21.9%, patients on peritoneal dialysis (PD) 20.3%, patients with CKD 34.4% and with glumerulonephritis on immunosuppressive drugs 23.4%. The vaccination rates of each group were 57.1%, 61.5%, 36.4%, 26.7% respectively. TR and children on PD had a fourfold higher rate of being vaccinated (odds ratio 4.0, 95% CI 1.008 to 15.868). H1N1 infection presented 19.4% of all children and 7.8% of family members. Most common causes of denying vaccination included fear of adverse effects (48.9%), insufficient information (31.9%) and others (19.2%). Patients being vaccinated were all urged by their pediatrician (100%), while patients not vaccinated were influenced by media (41.4%), friends (24.1%), pediatrician (20.7%) and others (13.8%). Regarding parents education, higher level was associated with increased rate of vaccination (odds ratio 1.44, 95% CI 1.024 to 2.026).

**Conclusions:** It seems that patients with severe renal disease had better compliance with H1N1 vaccination. The pediatrician had positive influence in contrast to the media which had the most negative influence.

## Abstract# 667

**Migration Background and Patient Satisfaction in a Large City Pediatric Nephrology Outpatient Clinic** D. Züllich,[1] M. Zimmering,[1] T. Keil,[2] U. Querfeld.[1] *[1]Pediatric Nephrology, Charité, Berlin, Germany; [2]Epidemiology, Charité, Berlin, Germany.*

**Objectives:** Data on health-related issues in children and adolescents with a migration background are yet scarce but needed for future public health planning. The present study was conducted to obtain data on migration background, family social and financial status, education, and patient satisfaction of children and their families treated by our institution.

**Methods:** In a cross sectional survey of patients seen during 2008 in the Pediatric Nephrology Outpatient Department at the Charité University Children's Hospital in Berlin, a total of 348 families answered a prepared questionaire. The final data set contained basic information, a standardized patient satisfaction score, a subjective categorical rating of disease severity and satisfaction with treatment, and a subjective categorical external evaluation (by doctors and nurses) of patient compliance.

**Results:** A migration background was present in 131 patients (38%). Patient satisfaction (on a scale from 8-40) was significantly higher in families without (32.9 + 4.6) than in those with a migration backgroud (30.8 + 4.7; $p<0.0001$). In contrast, patient satisfaction was not significantly associated with income, education (school level achieved by parents), and religious background. Patient satisfaction was not significantly associated with subjective ratings of disease severity or friendliness of nurses or doctors, but showed a significant correlation with trust in doctors ($p<0.0001$). There was no apparent correlation of patient satisfaction with compliance.

**Conclusions:** Measurement of patient satisfaction by the ZUF-8 score is a promising tool for evaluating quality of care in pediatric populations.

## Abstract# 668

**Macro-Autophagy and Chaperone Mediated Autophagy in Kidneys of Aging Mice** A. Quiroga,[1] R. Kiffin,[2,3] A.M. Cuervo.[2,3] *[1]Pediatric Nephrology, Montefiore Childrens Hospital, Bronx, NY, United States; [2]Developmental and Molecular Biology, Albert Einstein College of Medicine, Bronx, NY, United States.*

**Objectives:** Background: Age related functional decline has been associated with macro-autophagy and chaperone mediated autophagy. This association has been characterized in many organs. Since the kidney suffers a functional deterioration with age and the molecular mechanisms responsible are not fully identified, we have studied the expression of autophagy related molecules in kidneys of aging mice.

**Methods:** Methods: Immunoblot of mouse renal tissue from three different age groups (young, middle aged and old) was performed for essential proteins of the two major autophagic pathways in mammalian cells: macro-autophagy and chaperone mediated autophagy. Functional studies were carried out using lysosomes isolated from kidneys of the same mice groups.

**Results:** Results: 1) Levels of both effector and regulator proteins of macroautophagy and chaperone mediated autophagy increase considerably with age in mice. 2) The ability of lysosomes from old mice kidney to take up cytosolic proteins for degradation is impaired.

**Conclusions:** Conclusion: Expression of proteins related to macro-autophagy and chaperone mediated autophagy is decreased in a time dependent fashion in kidney and this may contribute to the observed functional decline in autophagic activity in this organ with age.

## Abstract# 669

**Nocturnal Enuresis: What Do Enuretic Children Think? What Do Their Parents Think?** E. Taborga,[1] L.M. Rodríguez,[2] C. Cebrián,[3] A. Cobo,[1] F. Santos,[4] V. Martínez.[1] *[1]Pediatría, Atención Primaria, Asturias, Spain; [2]Nefrología Pediátrica, CALE, León, Spain; [3]Cirugía Pediátrica, HUCA, Oviedo, Spain; [4]Nefrología Pediátrica, HUCA, Oviedo, Spain.*

**Objectives:** The study attempts to determine the effect that the condition has on enuretic children and their parents as well as their opinion of it.

**Methods:** 104 enuretic children (7 - 14 years) and their parents were surveyed on the effects of enuresis on their lives. Both groups were asked the following questions:

Do you consider enuresis a normal condition?
Do you consider enuresis to be a disease?
Do you feel indifferent, angry, or embarrassed about enuresis?
Does enuresis limit your ability to socialize?
Parents were asked if their children felt guilty about their enuresis
Children were asked if their parents worried about their problem.

**Results:** 77% of parents, but only **18%** of children consider enuresis a normal and frequent occurrence

22% of parents and children **consider enuresis a disease**

Regarding their children's enuresis, parents feel: indifferent (77%), angry (8%) and embarrassed (4%). Enuretic children feel: indifferent (33%), angry (13%) and embarrassed (**53%**)

25% of the parents and 17% of the children believe that enuresis affects the children's ability to socialize

28% of the parents believe that enuresis elicits feelings of guilt in their children and 21% of the children believe that their parents are not worried about their enuresis.

**Conclusions:** Enuresis is not seen as a normal condition and it causes embarrassment for the children who suffer it. However, most parents consider enuresis a normal condition and do not experience any negative effects of their children's circumstances. 22% of the parents and enuretic children still consider enuresis a disease.

## Abstract# 670

**Urinary Levels of TGF-β1 in Patients with Vesicoureteral Reflux and Renal Parenchymal Scar** N.M. Zaicova,[1] V.G. Rotaru,[1] P.M. Stratulat.[1] *[1]Padiatric Nephrology, Institution of Scientific Research, Mother and Child Care, Chisinau, Moldova, Republic of Moldova; [2]Pediatric Nephrology, Institution of Scientific Research, Mother and Child Care, Chisinau, Moldova, Republic of Moldova; [3]General Pediatric, Institution of Scientific Research, Mother and Child Care, Chisinau, Moldova, Republic of Moldova.*

**Objectives:** to evaluate the urine levels of TGF-β1 as a noninvasive marker in the pathogenesis of tubulointerstitial damage in children with different grade of vesicoureteral reflux (VUR).

**Methods:** 33 patients aged between 1 and 14 years (78,8% female) with VUR and 10 normal children (control group) were enrolled in the study. Recently, a more accurate test for renal parenchymal damage such as the DMSA scan has been used. Children were divided into 2 groups according of renal parenchymal scars: gr. I - 24 patients (1-2 scars) and gr. II – 9 patients (more than 3-4 scars). Urinary excretion of TGF-β1/ creatinine was measured by ELISA method.

**Results:** all the patients demonstrated significantly elevated urine level of TGF-β1 in comparison with controls. Children with high degree of nerosclerosis showed higher urine concentration of TGF-β1/creatinine than patients with RN gr.1-2 ($p<0.05$).

**Conclusions:** high urine level of TGF-β1/creatinine is closely associated with renal parenchimal scars in children with different degree of VUR. The results of this study have showed the important role of TGF-β1 in progression of interstitial fibrosis and maybe used as diagnostic marker for renal parenchymal scarring.

## Abstract# 671

**Tubular Function in Patient with Thalassemia Minor** S. Sadeghi-bojd,[1] M. Hashemi.[2] *[1]Pediatric, Child and Adolescences Health Center, Zahedan Medical Sciences of University, Zahedan, Sistan&baloochestan, Islamic Republic of Iran; [2]Biochemistry, Zahedan Medical Sciences of University, Zahedan, Sistan7baloochestan, Islamic Republic of Iran.*

**Objectives:** β – Thalassemia minor is a common heterozygous hemoglobinopathy that is characterized by both microcytosis and hypochromia. It has been postulated that low grade hemolysis, tubular iron deposition and toxins derived from erythrocytes might cause renal tubular damage in adult patients with β – thalassemia minor. The aim of this study is to investigate the renal tulbar function in children with β – thalassemia minor and to determine its possible harmful effects.

**Methods:** The study was conducted on 50 children (22 male and 28 female) at the age of years (range 4-19 years) with β – Thalassemia minor. A control group was formed with 50 healthy children whose ages and sexes match those in the first group.

Blood and 24-hour urine samples were obtained for hematologic and biochemical analysis.

**Results:** There was statistically significant difference among the two groups in terms of the results of $FE_{UA}$ (%), TPR (%), $FE_k$(%), GFR, Urine uric Acid, serum Mg (P < 0/05). But other significant signs of renal tubulopathy such as hypercalciuria, and tubular proteinuria ($\beta_2$ – microglobolinuria) were not seen.

**Conclusions:** On the contrary of children with β – Thalassemia major, renal tubular dysfunction is not common in children with β – Thalassemia minor. For this cause following up the patients with β – Thalassemia minor after 20 years is reasonable.

Alsow it suggested that in future studies to confirm renal tubular disfunction should be considered specific test such as NAG measurement.

## Abstract# 672

**Determining Risk Factors for Developing Hypothyroidism in Cystinosis** M.C.M. Scholten, H. de Jong, E.A.M. Cornelissen. *Pediatric Nephrology, Radboud University Medical Centre, Nijmegen, Netherlands.*

**Objectives:** Cystinosis is caused by intralysosomal accumulation of cystine and characterized by renal failure as well as deterioration of other organs. One of the known complications is thyroid failure. We analyzed risk factors for hypothyroidism.

**Methods:** Medical histories of 36 cystinosis patients were analyzed. Data were collected on last known measurement in non hypothyroidism group (TSH<4.5 mU/L, n=23) and on time of diagnose of hypothyroidism in hypothyroidism group (TSH>4.5 mU/L, n=13). Mean age was not significantly different (median 21.4 yrs (range 4.2-28.7) vs 13.9 (3.3-41.7), p=0.62). Mann-Whitney U test and chi-square test were used to analyze the data.

**Results:** TSH was higher in hypothyroidism group (5.5 mU/L (2.8-36.7) vs 1.7 (0.4-4.2), p<0.01), while fT4 was similar in both groups (12.0 pmol/L (9.0-19.3) vs 12.6 (7.8-20.9), p=0.74). GFR was definitely lower in patients with hypothyroidism (43.0 ml/min/1.73m$^2$ (21.5-95.1) vs 91.8 (3.5-131.8), p<0.01). Thereby more patients with hypothyroidism received a renal transplant (10/13 vs 9/23, p<0.05), of whom 8 developed hypothyroidism only after transplantation. Age at starting Cysteamine therapy was higher in hypothyroidism (2.8 yrs (0.6-25.3) vs 1.3 (0.3-16.3), p<0.05). Also dose of Cysteamine was lower in hypothyroidism group (14.0 mg/kg/day (0-118.5) vs 51.5 (15.2-73.0), p<0.05). There was no significant difference in urinary protein/kreat ratio and mean blood cystine-levels of the last 5 years.

**Conclusions:** Renal failure and not fanconi syndrome seems to be an indicating factor for thyroid deterioration. A renal transplant does not protect. Indeed, early starting with an adequate dose of cysteamine prevents or delays thyroid failure.

## Abstract# 673

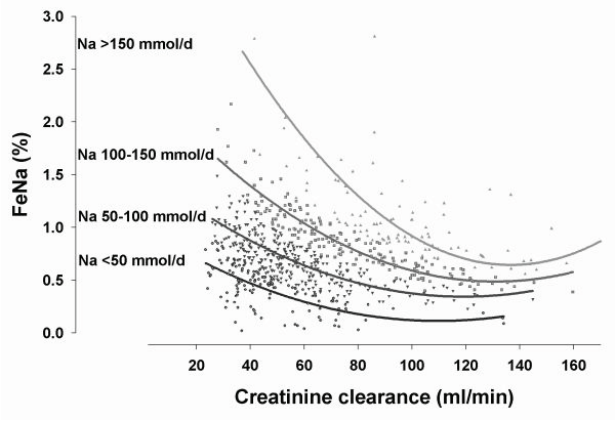
**Interpretation of the Fractional Excretion of Sodium in Absence of Renal Failure** M.F. Schreuder,[1] A. Bökenkamp,[2] J.A.E. van Wijk.[2] *[1]Department of Pediatric Nephrology, Radboud University Nijmegen Medical Centre, Nijmegen, Netherlands; [2]Department of Pediatric Nephrology, VU University Medical Center, Amsterdam, Netherlands.*

**Objectives:** The fractional excretion of sodium (FeNa) may be helpful in establishing the cause of acute renal failure. This study was performed to determine the influence of the glomerular filtration rate (GFR), sodium intake and tubular function on FeNa in children without renal failure.

**Methods:** Reliable 24h-urine collections from patients (4-18years of age, GFR >60 ml/min/1.73m$^2$) were used to determine sodium excretion, GFR and FeNa. The influence of tubular function was studied in 5 patients with generalized tubular dysfunction.

**Results:** Based on data from 761 patients, a multiple regression formula was designed based on GFR and sodium excretion that predicted over 80% of the variation in FeNa (R square=0.824, p<0.001). Using this formula, the predicted FeNa was significantly lower than the measured FeNa in the children with tubular dysfunction.

**Conclusions:** FeNa depends on GFR, sodium intake, and tubular function, and interpretation is impossible without knowledge on all factors.




Therefore, no normal range for FeNa can be given. However, a large difference between measured and predicted FeNa may indicate tubular dysfunction. The clinical usefulness of our formula should be tested in other cohorts of children both with normal and abnormal tubular function.

## Abstract# 674

**E-Learning for Patients and Physicians in Pediatric Nephrology – A Success Story!** S.K. Sethi,[1] A. Bagga.[2] *[1]Department of Pediatrics, PGIMER and associated RML Hospital, New Delhi, India; [2]Department of Pediatrics, All India Institute of Medical Sciences, New Delhi, India.*

**Objectives:** With rapid increase in the availability of new web based applications, there is a need for Pediatric Nephrology community to adopt these applications into professional and educational services. Till date, there is only one mailing list (pedneph) and Blog (Golden Thoughts).

**Methods:** Website Pediatric-Nephrology (https://www.pediatric-nephrology.com) was designed in November 2009, integrating technologies like Blog with daily updates, discussion forums, polls, RSS, powerpoint presentations, you-tube videos, social-tagging like twitter, facebook and newsletter services. The website features all the latest content moderated by Dr SK Sethi. 'Pediatric Renal Grand Rounds' is a fortnightly newsletter on recent developments in the field.

**Results:** The website met with unprecedented success, with 50,000 hits in 2009 and 2,50,000 hits in 2010. The website has more than 10,000 unique visitors from all parts of the world (majority USA, Russia and India).The website opens on Page 1 of Google with the search keywords "pediatric nephrology" and Number 1 with search keywords "pediatric nephrology blog". Pediatric Renal Grand Rounds (6 volumes till date) is now viewed, appreciated with contributions from all over the world.

**Conclusions:** Integration of the new technologies could offer a way to enhance clinician's and patient's learning experiences, and deepen levels of learner's engagement and collaboration within digital learning environments. All the readers are invited to access the website and contribute to the Pediatric-Nephrology virtual world (https://www.pediatric-nephrology.com).

## Abstract# 675

**Follow-Up and Experience in Preschool and School-Age Children Screened by Urinalysis in Shanghai** Q. Shen,[1] H. Xu,[1] M.-J. Wei,[2] G.-H. Zhu.[3] *[1]Department of Nephrology and Rheumatology, Children's Hospital of Fudan University, Shanghai, China; [2]Department of Pediatrics, Xinhua Hospital of Jiaotong University, Shanghai, China; [3]Department of Nephrology, Children's Hospital of Jiaotong University, Shanghai, China.*

**Objectives:** The aim of this study was to analyze follow-up data of these children who show abnormal urinary findings in the preschool and school screening to investigate the distribution of diagnoses and to preliminarily evaluate the effect of preschool urine screening programme.

**Methods:** Between 2003 and 2007, a total of 419 children identified with abnormal urinary screening, aged 3 to 18 years, received regular follow-up in designated hospitals.

**Results:** Children with hematuria accounted for the majority in the preschool and elementary school group. However, in the high school group, proteinuria was noted in 36% of referred children. Out of the 419 children, 39 cases received renal biopsies. Results showed 20 cases of minor glomerular abnormalities, 7 cases of IgA nephropathy and thin basement membrane disease, respectively. Besides 39 cases of renal biopsies, 27 cases were diagnosed with left renal vein

entrapment syndrome, 6 cases with idiopathic hypercalciuria and urinary tract infection, respectively. Finally, we analyzed the follow-up data of preschool children and found quite some patients were diagnosed with IgA nephropathy, UTI, nephrotic syndrome, hypercalciuria, respectively.
**Conclusions:** Our results indicate urine screening program and regular follow-up are effective for early detection of kidney diseases. Our pilot preschool screening program suggests urine screening is also quite important for preschool children to receive early diagnosis and timely treatment.

## Abstract# 676

**Ultrasound Assessment of Kidney Size by Using the Ratio of Kidney Length to the Distance between the Spinous Processes of the Fourth and Fifth Lumbar Vertebrae** M. Shimoda, E. Kikuchi. *Department of Pediatrics, Musashino Red Cross Hospital, Musashino Tokyo, Japan.*
**Objectives:** In recent years, ultrasonography (US) has replaced intravenous pyelography (IVP) as the primary imaging study of kidneys. In the case of IVP, the ratio of the length of the kidney to that of the vertebral bone is used for the assessment of kidney size. However, there have been no reports on the US measurement of this ratio. In this study, we determined whether the ratio of the length of the kidney to the distance between the spinous processes of the fourth and fifth lumbar (L4-5) vertebrae is constant and could then be used to assess kidney size.
**Methods:** The length of the kidney and the distance between the spinous processes of L4-5 in the normal prone position were measured by US using a 5-MHz convex probe in 111 children (maximum age, 16 years ). These children were referred to our hospital for urinary abnormalities detected during a urine screening program in school or for nonurinary complaints. Children with a history of renal disease other than asymptomatic hematuria were excluded.
**Results:** A positive correlation was observed between the L4-5 distance and subject age ($r = 0.67$, $P < 0.01$) and height ($r = 0.87$, $P < 0.01$). The kidney length progressively increased with subject age and height. In contrast, the ratio of the kidney length to the distance between the spinous processes of L4-5 was found to be almost constant over all age and height ranges. The average ratio was 4.8 (0.7) (mean (SD)) for the right kidney and 4.9 (0.8) for the left kidney.
**Conclusions:** The ratio of kidney length to the distance between the spinous processes of L4-5, calculated using US, is valuable for assessing the kidney size in children.

## Abstract# 677

**Prevalence of Nutcracker Syndrome and Its Correlation with Urinary Protein Excretion Using Renal Doppler Ultrasound in Children with Orthostatic Proteinuria** J.I. Shin,[1] J. Kim,[1] J.S. Lee,[1] M. Kim.[2] *[1]Department of Pediatrics, The Institute of Kidney Disease, Yonsei University College of Medicine, Seoul, Korea; [2]Department of Diagnostic Radiology, Yonsei University College of Medicine, Seoul, Korea.*
**Objectives:** To evaluate the prevalence of nutcracker syndrome and its correlation with urinary protein excretion in children with orthostatic proteinuria.
**Methods:** We measured renal Doppler sonographic indices serially in 17 children (M:F=6:11, age 7-15 years) with orthostatic proteinuria and 17 age and sex-matched controls. The protein/creatinine ratio (PCR) during day and night (daytime PCR/nighttime PCR = PCR ratio) were also calculated.
**Results:** The PV ratios of the LRV were $6.32 \pm 3.05$ in the patient group and $2.77 \pm 0.72$ in the control group ($p < 0.0001$). When the cut-off value of 4.21 (mean ± 2 SD of normal controls) was applied, nutcracker syndrome was detected in 13 (76.5 %) of the 17 patients with orthostatic proteinuria and in none of the normal controls. The prevalence of nutcracker syndrome decreased from 76.5 % to 47.6 % at follow-up despite persistent orthostatic proteinuria. The PV ratio of the LRV was correlated positively with the nighttime PCR ($r = 0.464$, $p = 0.039$) and the age was correlated negatively with the PCR ratio with a borderline significance ($r = - 0.399$, $p = 0.082$).
**Conclusions:** Nutcracker syndrome may be an important cause of orthostatic proteinuria in children. Renal Doppler ultrasound may be a useful method in screening nutcracker syndrome among patients with orthostatic proteinuria. Nutcracker syndrome and PCR ratio may improve during adolescence, but orthostatic proteinuria itself may persist for a relatively long time.

## Abstract# 678

**Complement Dysregulation in Hemolytic Uremic Syndrome (HUS)** A. Sinha,[1] M.A. Dragon Durey,[2] C. Blanc,[2] A. Gulati,[1] S. Sethi,[1] A. Dinda,[1] P. Hari,[1] A. Bagga.[1] *[1]All India Institute of Medical Sciences, New Delhi, India; [2]APHP, Hôpital Européen Georges Pompidou, Laboratoire d'Immunologie, Université Paris Descartes, Paris, France.*
**Objectives:** To screen for abnormalities in complement regulatory proteins & anti-factor H antibodies (AFHA) in patients with HUS.
**Methods:** Levels of C3, C4, CD46, factors H, I & B, & AFHA were analyzed in consecutive children with HUS from Jan 2007-Dec 2009. Patients with AFHA were tested for deletion of factor H-related genes *CFHR1/CFHR3*. Beside supportive therapy, frequent plasma exchanges were performed. Patients with AFHA also received IV immunoglobulin (IVIG) (2g/kg), IV cyclophosphamide (500 mg/m$^2$ q monthly x 6) & prednisolone (1 mg/kg/d X 4 weeks, then tapered). Levels of creatinine, complement & AFHA were monitored. Outcome was assessed by eGFR and dialysis dependence.
**Results:** Of 25 patients (19 boys), age 8.5±4.3 (0.6-15.8) yr, 7 had low C3. Factors H, B & I were low in 4, 7 & 2 cases respectively. A patient with homozygous deletion in factor H gene progressed to ESRD. Anti-factor H antibodies (187- >20,000 AU/ml) were present in 16 (64%), with high titers in dialysis dependent patients. All patients with AFHA were homozygous for *CFHR1/CFHR3* gene deletion. Immunosuppression decreased AFHA titres in dialysis dependent patients. Six patients had CKD 1-3; 4 patients with AFHA had relapses. A patient with factor I deficiency died in the acute stage. Of 7 patients with no complement abnormalities, 4 had CKD 1-3 at 1-y.
**Conclusions:** Patients with HUS show a spectrum of complement abnormalities, with variable outcome. Patients with antifactor H antibodies are an important cause of HUS in Indian children and might benefit from immunosuppression, but have disease relapses.

## Abstract# 679

**Clinical & Pathological Correlates of Nephrotoxicity in Children Receiving Prolonged Calcineurin Inhibitor (CNI) Therapy for Steroid Resistant Nephrotic Syndrome** A. Sinha,[1] A. Mehta,[1] A. Sharma,[2] R. Gupta,[2] P. Hari,[1] A.K. Dinda,[2] A. Bagga.[1] *[1]Division of Pediatric Nephrology, All India Institute of Medical Sciences (AIIMS), New Delhi, India; [2]Department of Pathology, AIIMS, New Delhi, India.*
**Objectives:** To assess the clinical, biochemical & histological parameters associated with calcineurin inhibitor toxicity (CNIT) in renal biopsies following >2-yr of CNI therapy for steroid resistant nephrotic syndrome (SRNS).
**Methods:** Records of patients with SRNS who had received therapy with CNI [cyclosporine (CsA) or tacrolimus (Tac)] for >2 yr were reviewed. CNIT was defined as presence of striped interstitial fibrosis or peripheral nodular arteriolar hyalinosis.
**Results:** Forty patients (24 boys) with SRNS (initial resistance 24), who received therapy with CsA (28) or Tac (12) were included. The ages at onset of nephrotic syndrome & CNI therapy were 3.5±3 &5.7±3.6 yr. Patients received CNI therapy for 3.5±1.3 yr. Renal biopsy, performed after 2.8±0.9 yr, revealed CNIT in 10 (25.5%) biopsies. Risk factors for CNIT were type of resistance (P=0.03), hypertension (0.03), serum creatinine (0.07), duration of heavy proteinuria (0.06), global sclerosis (0.07), interstitial fibrosis (0.02), arteriolar hyalinosis (0.003) & arteriolar smooth muscle vacuolization (0.01). On multivariate regression, initial resistance (0.02), global sclerosis (0.05) & arteriolar hyalinosis (0.02) were significantly associated ($R^2$ 0.60).
**Conclusions:** Following prolonged therapy, almost 1 in 4 biopsies show features of CNI toxicity. The presence of initial resistance, uncontrolled hypertension & persistent nephrotic proteinuria may increase the risk of CNIT; subtle histological features may alert to increased risk of CNIT.

## Abstract# 680

**Familial Mediterranean Fever and Pras Scoring System** E. Yilmaz, B. Sozeri, A. Berdeli, S. Mir. *Pediatric Nephology, Ege University Faculty of Medicine, Izmir, Turkey.*
**Objectives:** The Aim was to demonstrate the phenotype and genotype relationship of FMF and demonstrate effects of MEFV gene mutations on Pras Score.
**Methods:** 191 patients had 1 or 2 mutations by FMF strip assay. Patients were investigated regarding the mutation type, clinical characteristics at the time of inflammatory attacks such as fever, abdominal pain, arthritis, chest pain, erysipelas- like erythema, epidemiological data, consanguinity, severity score and family history of FMF and amyloidosis. Pras Scoring was made all.
**Results:** Of the 191 patients ( 99 male, 92 female), and median age of FMF diagnosis was 8±2 years. Most common symptom was abdominal pain. The most frequent seen mutation was the M694V/M694V.
Pras score was found high in patients with arthritis, arthraljia, myaljia and familial history of amiloidozis. The patients carrying M694V homozygous mutation had high Pras scores and need high colchicine dosage in order to control disease activity. Severe Pras score seen in 5 patients (M694V homozigot, M694V heterozygote, E148Q, M694V/M680I heterozygote).
**Conclusions:** M694V mutation presence increase Pras score and it is associated with severe disease activity.

## Abstract# 681

**MEFV Gene Single Nucleotid Polymorphisms and Their Clinical Relevance** E. Yilmaz, B. Sozeri, S. Mir, A. Berdeli. *Pediatric Nephology, Ege University Faculty of Medicine, Izmir, Turkey.*
**Objectives:** There is a lack of information concerning single nucleotide polymorphisms (SNP) over MEFV in patients and healthy individuals. Aim was to describe the MEFV SNP mutational data and demonstrate its phenotypic expression.

**Methods:** Of the 261 unrelated patients investigated. 191 patients had 1 or 2 mutations by FMF strip assay. DNA sequence analysis was made to 70 patients who had no mutation with stripe MEFV gene analysis method.
**Results:** Of the 261 patients (136 girls, 125 boys) and the age ranged from 104,8±49 months. There were common seen mutations in 191 patients, 70 patients had MEFV single nucleotide polymorphisms. The most frequent mutation was the M694V/M694V. R202Q mutation was the common expressed polymorphism in 70 patients without any identified mutation by strip assay.We demonstrate coexpression of R314R single nucleotide polymorphism present on 3th exon and D102D, G138G, A165A variants and R314R single nucleotide polymorphism and E474E, Q476Q, D510D variants. We thought with all these findings that R314R is the hot spot point for occurrence of different haplotypes. Analysis of clinical symptoms for R202Q, R314R single nucleotide polymorphisms showed no statistical significance.
**Conclusions:** It was thought that the patients diagnosed as having FMF according to Tel Hashomer's criteria and carrying MEFV gene polymorphisms expressing phenotyping effects of these polymorphisms. DNA sequence analysis should be used in patients presented no mutation by traditional stripe MEFV gene analysis, by the way we could understand the association of MEFV gene polymorphism and phenotype relationship.

## Abstract# 682

**Nasopharyngeal Carriage (NC) of Streptococcus pneumoniae (SP) in Polish Children with Nephrotic Syndrome (NS)** A. Szmigielska, M. Roszkowska-Blaim, A. Wasilewska, J. Tyl, I. Ogarek, H. Borzecka, M. Tkaczyk, T. Krynicki, A. Cieslak-Puchalska, A. Medynska, A. Morawiec-Knysak. *Ped. Neph., Med. Univ, Warszawa, Poland; Ped. Neph., Med. Univ, Warszawa, Poland; Ped. Neph., Med. Univ, Bialystok, Poland; Ped. Neph., Med. Univ, Gdansk, Poland; Ped. Neph., Med. Univ, Kraków, Poland; Ped. Neph., Med. Univ, Lublin, Poland; Ped. Neph., Med. Univ, Lódz, Poland; Ped. Neph., Med. Univ, Poznan, Poland; Ped Neph., Ped. H ospital, Szczecin, Poland; Ped. Neph., Med. Univ, Wroclaw, Poland; Ped. Neph., Med. Univ, Zabrze, Poland.*
**Objectives:** The prevalence of NC of SP, antibiotic susceptibility (AS) of serotypes, risk factors for NC in pts with NS.
**Methods:** 95 children with NS (I), 32 healthly pts (II) aged 2-5 years. SP was detected with microbiological tests, serotypes with Quellung reaction. AS was studied with E-tests and D-tests. Risk factors for NC of SP were identified in a questionnaire.
**Results:** NC of SP was 13.7% in pts with NS, 21.8% healthy pts. The most common serotypes of SP were: I - 6B, 9N, 11A, II-6B, 9V. In pts with NS, 80% isolates of SP were susceptible to penicillin, 80% to chloramphenicol, tetracycline, erythromycin, clindamycin. In group II 87.5% serotypes were susceptible to penicillin, 75% to chloramphenicol, 62.5% to erythromycin, clindamycin, 50% to tetracyclines. There was no resistance to rifampicin and levofloxacin. The risk factors for NC were: male gender and urban residence.
**Conclusions:** The frequency of NC of SP in the children with NS <5 years was 13,7%. The most common isolated serotype was 6B. The AS for SP was higher in children with NS. The risk factors for NC were: male gender and urban residence.

## Abstract# 683

**Neutrophil Gelatinase-Associated Lipocalin (NGAL): A New Marker of Cyclosporine Nephrotoxicity?** K. Taranta-Janusz, A. Wasilewska, W. Zoch-Zwierz, J. Michaluk – Skutnik. *Department of Paediatrics and Nephrology, Medical University of Bialystok, Bialystok, Poland.*
**Objectives:** Serum and urinary neutrophil gelatinase - associated lipocalin (sNGAL; uNGAL) may be useful as an early indicator of cyclosporine A (CsA) nephrotoxicity in steroid - dependent nephrotic children (SDNS).
**Methods:** The study group (I) consisted of 19 children with SDNS treated with CsA. The children were examined four times: A – at proteinuria relapse, before CsA treatment, then after 3, 6 and 12 months of CsA treatment. Control group (II) consisted of 18 healthy children. Immunoenzymatic ELISA commercial method was used to measure NGAL concentration.
**Results:** Serum NGAL levels in patients from group IA did not differ from healthy controls ($p<0.05$). After 3 and 6 months of CsA treatment, the sNGAL levels were three- and as much as sixfold higher than those of controls ($p<0.01$). After 12 months of CsA therapy, the sNGAL had decreased. We found a positive correlation between the serum levels of NGAL and CsA at examinations B and C. The uNGAL/cr ratio in NS children increased during CsA treatment ($p<0.01$). The urinary NGAL/cr ratio positively correlated with CsA serum concentration in examination IB, IC, and ID. The level of sNGAL, but not the uNGAL/cr ratio was found to be positively correlated with serum creatinine values ($r=0.33$, $p<0.001$) and, inversely, with eGFR ($r=-0.22$, $p<0.05$). The analysis indicated that the sNGAL assay did not appear to be predictive of CN, because the receiver operating curve (ROC) was quite close to the diagonal line.
**Conclusions:** We found that both serum and urinary NGAL concentration increases in our NS pediatric patients during the course of CsA treatment.

## Abstract# 684

**Bilateral Cystic Kidneys in an Infant with WAGR Syndrome** V. Tasic,[1] O. Muratovska,[1] L. Misevska,[2] J. Crolla,[3] Z. Gucev.[1] *[1]University Children's Hospital, Skopje, Macedonia, The Former Yugoslav Republic of; [2]Pediatric Surgery Clinic, Skopje, Macedonia, The Former Yugoslav Republic of; [3]Wessex Regional Genetics Laboratory, Wiltshire, United Kingdom.*
**Objectives:** The WAGR contiguous gene deletion syndrome is a combination of Wilms tumor, aniridia, genito-urinary abnormalities, and mental retardation. Herein we report a 1.5 year old female infant with WAGR syndrome and bilateral cystic kidneys.
**Methods:** The index patient underwent detailed clinical examination, ultrasound of the kidneys and FISH study.
**Results:** A 1.5 year old female infant was investigated for congenital bilateral aniridia, cataracts, glaucoma and epicantus. The ultrasound (US) scan showed several cysts in both kidneys with the overall appearance mimicking polycystic kidney disease. A small atrial septal defect and a dysplastic pulmonary valve without haemodynamic effect were also diagnosed. FISH study revealed a deletion of the WT-1 and PAX6 gene in the 11p13 WAGR region. Forty days after the first kidney US, the second US revealed a 3 cm tumor in the right kidney. After nephrectomy histological analysis revealed Wilms tumour, treated successfully with the Wilm's tumor protocol.
**Conclusions:** Identification of deletion in the WAGR region in patients with aniridia should be mandatory. In addition, Wilms tumor can have a very rapid growth, which, per se requires frequent and careful ultrasound kidney controls. Cystic kidneys may be part of the WAGR presentation and precede development of Wilms tumor.

## Abstract# 685

**Long Term Follow up of the Patients with Cystinosis-Hacettepe Experience** R. Topaloglu, A. Gultekingil, Y. Bilginer, F. Ozaltin, A. Duzova, S. Ozen, N. Besbas, A. Bakkaloglu. *Pediatric Nephrology, Hacettepe University Faculty of Medicine, Ankara, Turkey.*
**Objectives:** Charts of twenty-four cystinosis patients were evaluated.
**Methods:** Nineteen of these patients were investigated for extrarenal complications. Swallowing studies, electromyography, abdominal ultrasonograpy, respiratory function test and ophthalmic examination were performed.
**Results:** Mean age of diagnosis was 36.7 months (range 1month-14 years). Most of the patients were suffering from polyuria, polydipsia and growth retardation at the time of diagnosis. At mean age 10.6 year, 40% of patients progressed to renal failure. Five patients (20%) had renal transplantation. 84% of patients had cystine crystals in cornea and 42% had photophobia at time of diagnosis. One patient had elevated creatin kinase levels and electromyography revealed myopathy. 31% of patients were found to have gastroesophageal reflux, and 21 % of them had remnants in esophagus while swallowing. Two out of 4 patients had restrictive pattern of respiratory functions. While 72% of patients had hepatomegaly, 38% of them had fine granular echo pattern and two had minimal heterogeneity of liver at ultrasonography, 55% also had increased minimum and maximum velocities of blood flow in portal vein.
**Conclusions:** Renal complications are early manifestations of cystinosis, some of patients need renal replacement at first decade even under cysteamine therapy, but many could survive until adulthood without great loss of renal functions. Thus extra renal complications are more common in adults, cystinotic children also have the risk of developing many extrarenal complications and deserve close follow-up and management.

## Abstract# 686

**Long Term Endocrinologic Complications of Cystinosis-Hacettepe Experience** R. Topaloglu,[1] A. Gultekingil Keser,[1] O. Engiz,[2] Y. Bilginer,[2] N. Kandemir.[2] *[1]Pediatric Nephrology, Hacettepe University Faculty of Medicine, Ankara, Turkey; [2]Pediatric Endocrinology, Hacettepe University Faculty of Medicine, Ankara, Turkey.*
**Objectives:** We aimed to evaluate endocrine involvement in 19 patients with cystinosis.
**Methods:** Anthropometric measurements, pubertal stage were noted. IGF1, IGFBP3, thyroid functions, gonadotropins, fasting and postprandial glucose, insulin, c-peptid levels, bone mineral density were measured and oral glucose tolerance test was performed.
**Results:** While mean age of patients (12 male, 7 female) was 11.8 years, mean age at diagnosis was 35.2 months. Mean duration of follow up was 9.7 years. 57% of patients had low body weight (≤3p), 84% had short stature (≤3p). 29% of patients have decreased bone mineral density (z score <-2), one of these patients had renal transplantation and 2 of them have end stage renal disease. 42% of the patients were found to have primary hypothyroidism, one of these

patients had renal transplantation and other 3 patients had end stage renal failure. 10% had central hypothyroidism, one of them was on CAPD. Out of 9 patients who reached pubertal age, 33% of them had pubertal delay and one had hypogonadotrophic hypogonadism, all of these 4 were boys. OGTT was performed in 13 patients, 10% were found to have diabetes, both of them are in end stage renal failure and 26% had glucose intolerance 2 of them had renal transplantation and 2 of them were at end stage renal failure.
**Conclusions:** Endocrinologic complications are seen in patients with poor renal functions as well as in patients with no deterioration of renal functions and under cysteamin therapy. Therefore endocrinologic functions should be meticulously evaluated to prevent future problems.

## Abstract# 687

**Screening for Renal Diseases in Asymptommatic Children** V.H. Tru, T.T. Phong. *Pediatric Nephrology, University of Medicine, Hochiminh City, Viet Nam; Pediatrics, Children Hospital N 2, Hochiminh City, Viet Nam.*
**Objectives:** Screening for renal diseases in asymptommatic children.
**Methods:** Using dipstick to detect blood, protein, le, and nitrit
First urine test for : ph, density, protein, blood, LE, nitrit
The positive children receive the second urinary dipstick 7 days later. After that they were tested for 24 h proteinuria, urine culture, ultrasound.
They were divided into 4 groups :
Group 1: Microscopic hematuria without proteinuria.
Group 2: Proteinuria without hematuria
Group 3: Microscopic hematuria with proteinuria
Group 4 : LE (+) and nitrit (+).
**Results:** 762 children aged 1 to 15 years old were examined.
The prevalences of grade 1,2,3,4 for the first screening were 4,6 %; 0,9 %; 0,2%; 4,6 %.
The prevalences of grade 1,2,3,4 for the second screening were 1,4 % 0,4 % ; 0,1% ; 1 %.
In the group 4 there are 53,3 % abnormalities in ultrasound : cystitis, hydronephosis.
**Conclusions:** Urine screening tests should be mandatoty to detect asymatomatic renal diseases in children.

## Abstract# 688

**Biopsy Native Kidneys with Child Lying in Lateral Position Rather Than Prone** Y. Tse, P. Yadav, I. Herrema, L. Kerecuk, H. Lambert, M. Ognjanovic, M. Coulthard. *Royal Victoria Infirmary, Newcastle Upon Tyne, United Kingdom.*
**Objectives:** Traditionally, renal biopsies are performed in the prone position. Many children require general anaesthesia to avoid distress. Prone positioning under GA has higher risks requiring paralysis, endotracheal intubation, ventilation and protection of face. Lateral position is safer for GA. We compared the efficacy and safety of USS guided renal biopsies between the two positions.
**Methods:** In our department physician preference dictates positioning. We audited retrospective case notes and biopsy quality of consecutive biopsies in the two groups.
**Results:** 44 lateral and 47 prone position biopsies were reviewed. Patient characteristics were similar in both groups for age, sex and indication for biopsy. Endotracheal intubation was carried out in all prone and 18% lateral patients. Diagnostic yield and complications were similar in both groups. No patients changed positions due to failed sampling.
**Conclusions:** Lateral positioning result in no worse outcome for children requiring GA for renal biopsy. This position is recommended to reduce anaesthetic risk without compromising diagnostic yield.

| | | Prone | Lateral | p value |
|---|---|---|---|---|
| n= | | 47 | 44 | |
| Age (years) | med (range) | 11.4 (1.5-15.9) | 9.1 (2.3-16.4) | ns |
| Endotracheal intubated | | 100% | 18% | <0.001 |
| No of passes | med (range) | 2 (1-4) | 2 (1-4) | ns |
| | mean | 2.4 | 2.1 | |
| Cores obtained | med (range) | 2 (1-4) | 2 (1-4) | ns |
| | mean | 2.2 | 1.9 | |
| Cores per pass | med (range) | 1 (0.25-1) | 1 (0.25-1) | ns |
| | mean | 0.93 | 0.94 | |
| No of glomeruli | med (range) | 30 (8-102) | 25 (4-98) | ns |
| | mean | 33 | 31 | |
| Complications | | Pain & haematuria 2 | Pain & haematuria 2 | |
| | | Pain & haematoma 1 | Pain & haematoma 2 | |

ns - not significant

## Abstract# 689

**Clinical Characteristics, Histological Subtypes and Outcome of Children with Nephrotic Syndrome at Steve Biko Academic Hospital, South Africa** G. Van Biljon. *Department of Paediatrics, Faculty of Health Sciences, University of Pretoria, Pretoria, Gauteng, South Africa.*
**Objectives:** To determine the clinical characteristics, histological subtypes and outcome of children with nephrotic syndrome (NS) treated at Steve Biko Academic Hospital.
**Methods:** A retrospective study was conducted. NS was defined according to standard criteria. Patient demographics, anthropometry, presence of hypertension, macroscopic haematuria and renal function were analysed. Kidney biopsies were performed as clinically indicated. Treatment with corticosteroids and cyclophosphamide was individualised and not according to standard protocol.
**Results:** Over 23 years (1986 – 2009) 358 children were studied of whom 73% were black, 19% white, 4% of mixed race and 3% Indian. The male/female ratio was 1.3:1. Relevant special investigations identified secondary NS in 7%. Hereditary NS was suspected in 2%. Of the group as a whole macroscopic haematuria was present in 9% and 63% had hypertension. Kidney biopsies were performed in 89% of patients. The main histological diagnoses were minimal change nephrotic syndrome (MCNS) in 43% and focal segmental glomerulosclerosis (FSG) in 31%. The incidence of MCNS was lower in blacks (35% vs 68%, $p<0.001$) while the incidence of FSG was higher in blacks (37% vs 14%, $p<0.001$). Remission with corticosteroids was achieved in 80% of white vs 56% of black children with MCNS ($p=0.01$). More black children developed chronic renal failure (52/263 vs 3/68) ($p<0.01$) and black children had a higher mortality (40/263 vs 3/68) ($p<0.001$).
**Conclusions:** Significant racial differences in clinical presentation, histological subtypes and response to treatment are present in South African children with NS.

## Abstract# 690

**Thrombotic Complications in Nephrotic Syndrome (NS)** A.S. Vasudev, V.K. Agarwal, R.N. Srivastava. *Pediatric Nephrology, Apollo Indraprastha Hospital, New Delhi, India.*
**Objectives:** To study the pattern of major thrombotic complications in NS.
**Methods:** 5 children with NS with thrombosis of major vessels were seen and treated with anticoagulants.
**Results:** Case 1. 1.6- year-old-girl with steroid responsive (SR) NS was on prednisolone (P) 1.5 mg/kg on every other day (EOD). She developed generalised seizures and hemiparesis. Her serum albumin was 1.6 gm/dl and cholesterol 264 mg/dl. MRI:thrombosis of superior sagittal and transverse sinuses.
Case 2. 2-year-old boy with NS was on P 2mg/kg/day. He developed irritability and drowsiness. His serum albumin was 1.9 g/dl and cholesterol 352 mg/dl. MRI:thrombosis of superior sagittal, left transverse and straight sinuses.
Case 3. 5-year-old boy with SRNS was in remission on P 20 mg EOD. He developed severe frontal headache. MRI showed thrombosis of sagittal sinus.
Case 4. 7- year-old boy had SRNS with frequent relapses. He was in remission on P 15mg EOD. He had pain in left leg followed by bluish discoloration of foot. Leg was cold and arterial pulsations feeble. Blood albumin and cholesterol were normal. Ultrasound Doppler study showed narrowing of anterior and posterior tibial arteries.
Case 5. 12-year- old boy with NS was in remission on P20 mg EOD. He developed increasing swelling of his right shoulder and arm. Ultrasound Doppler study showed thrombosis in brachial and subclavian veins. Serum albumin and cholesterol were normal
Coagulation profile was normal in each case. All had a prolonged course but made a complete recovery.
**Conclusions:** Major thrombotic complications in NS are uncommon and may occur without a clear precipitating cause. Prompt detection and treatment with anticoagulants and supportive care usually leads to full recovery.

## Abstract# 691

**Urinary Transforming Growth Factor beta1 in Children and Adolescents with Congenital Solitary Kidney** A. Wasilewska, W. Zochzwierz, K. Taranta-Janusz. *Department of Paediatrics and Nephrology, Medical University of Bialystok, Bialystok, Poland.*
**Objectives:** The aim of this work was to check if, in children with a single functioning kidney from birth, the urinary excretion of TGF beta1 differed from that of healthy controls and whether it was correlated with age, compensatory overgrowth of the remnant kidney and the parameters of renal function.
**Methods:** The study group (I) consisted of 65 children and young adults with CSK and no other urinary defects. The control group (C) contained 44 healthy children and adolescents. Urine levels of TGF beta1 were determined by enzyme-linked immunosorbent assay (ELISA), using commercially available R&D Quantikine kits and expressed in picogrammes per milligram creatinine (pg/mg Cr).
**Results:** Children with a solitary kidney revealed compensatory overgrowth of the kidney (median 19.44%). The average kidney length was approximately 1.2-times longer than in healthy controls. The left kidney was more commonly

absent than the right one. Of the 65 patients with CSK, 97% (63) had normal blood pressure. Microalbuminuria was present in three patients in the CSK group, and, in 12 patients, albumin excretion was high normal. The concentration of urine TGF beta1 in children with CSK was more than twice as high. Urine TGF beta1 concentration was positively correlated with eGFR, uric acid concentration, and percentage of overgrowth and body mass index (BMI) centile.
**Conclusions:** We concluded that, although proteinuria and progressive renal insufficiency is not observed in patients with CSK during childhood, the renal haemodynamic changes are present and may be a risk factor for impairment of renal function and hypertension in future life.

## Abstract# 692

**High-Sensitivity CRP and Mean Platelet Volume (MPV) in Pediatric Hypertension** A. Wasilewska, E. Tenderenda, K. Taranta-Janusz, W. Zoch-Zwierz. *Department of Paediatrics and Nephrology, Medical University of Bialystok, Bialystok, Poland.*
**Objectives:** Investigation of ambulatory blood pressure parameters (ABPM) along with serum uric acid (SUA), hs – CRP and MPV in prehypertensive (PH) and hypertensive (HT) children and checking if SUA along with hs - CRP and MPV may be a predictors of hypertension in prehypertensives.
**Methods:** The study group consisted of 80 children (10 - 19 years) subdivided into prehypertension (PH) and hypertension (HT) groups according to mean systolic or diastolic daytime or nighttime BP levels. The control group (C) contained 25 normotensive children. Serum hs - CRP was determined using nephelometric method (Behring).
**Results:** The median SUA in PH and HT subjects was significantly higher compared to controls ($p< 0.01$)). Serum uric acid exceeded 5.5 mg/dl in 56% PH and 72% HT patients. Similarly serum hs - CRP was significantly higher in both examined groups of patients when compared to controls ($p< 0.01$). The difference between PH and HT group was statistically significant ($p< 0.05$). MPV in PH group did not differ from controls ($p> 0.05$), but was significantly higher in HT patients ($p< 0.01$).
Positive correlation between SUA and hs - CRP ($r= 0.415$, $p< 0.01$), SUA and MPV ($r= 0.436$, $p< 0.01$), and hs - CRP and MPV ($r= 0.352$, $p< 0.01$) was found. Only SUA was positively correlated with all the ABPM parameters. Linear regression analysis indicated that each 1 mg/dl increase in SUA was associated with an average increase of 6.7% in SBP load and 7% in DBP load.
**Conclusions:** We demonstrated that in HT children increased SUA with parallel increase in hs – CRP and PLT with MPV is observed. In PH children we found higher SUA and hs – CRP level with normal MPV.

## Abstract# 693

**Concentration of sRANKL and OPG in Nephrotic Children Treated with Glucocorticosteroids** A. Rybi-Szuminska, A. Wasilewska, W. Zoch-Zwierz, A. Korzeniecka-Kozerska. *Department of Paediatrics and Nephrology, Medical University of Bialystok, Bialystok, Poland.*
**Objectives:** RANK/RANKL/OPG seems to play the key role in the pathogenesis of glucocorticoid-induced osteoporosis.
The aim of the study was to compare the concentration of sRANKL, OPG and RANKL/OPG ratio in the serum of children with INS treated with GCs depending on the total dose of GCs.
**Methods:** The study group consisted of 90 children with INS, treated with GCs This group was subdivided according to the total dose of used GCs: A >1 g/kg m.c., B ≤ 1 g/kg m.c. The control group (C) consisted of 70 healthy children at a similar age. Both OPG and sRANKL serum concentrations were determined using ELISA method, according to the instruction given by the manufacturer (Biomedica Medizinprodukte GMBH&Co KG, A-1210 Wien).
**Results:** RANKL concentration was significantly higher in the group of children with INS in comparison to the group C ($p<0.05$), while OPG concentration in the serum of the ill children was lower in comparison to controls ($p<0.05$). RANKL/OPG ratio was significantly higher in the group with INS ($p<0.01$). The concentration of RANKL was higher in the group of children treated with high doses of GCs in comparison to children with small exposition on GCs ( $p<0.01$). The concentrations of OPG were similar in the both groups. RANKL/OPG ratio was almost three times higher in the group A in comparison to group B ($p<0.01$). There was a positive correlation between the total dose of GCs and the concentration of sRANKL as well as with the RANKL/OPG ratio ($r=0.33$, $p<0.05$).
**Conclusions:** In the long-term observation GCS treatment leads to increase of the sRANKL concentration but has no influence on the OPG serum concentration.

## Abstract# 694

**Urinary MCP-1 Excretion in Children with Glomerular Proteinuria** A. Wasilewska, W. Zoch-Zwierz, K. Taranta-Janusz, Z. Kolodziejczyk. *Department of Paediatrics and Nephrology, Medical University of Bialystok, Bialystok, Poland.*
**Objectives:** Proteinuria is an independent predictor of renal function decline. Proximal tubules overloaded with protein stimulates the production of chemokines, especially monocyte chemotactic protein 1 (MCP-1).
The aim of the study was to examine the urinary levels of monocyte chemoattractant protein - 1 (uMCP-1) in children according to histological diagnosis and degree of proteinuria.
**Methods:** The study group consisted of 49 children with proteinuria: I – 20 children with idiopathic nephrotic syndrome (INS), examined twice: A - during INS relapse; B - after proteinuria subsided, group II - 17 children, with persistent proteinuria due to focal segmental glomerulosclerosis (FSGS) and 12 children with IgA nephropathy (IgAN). The control group (C) contained 22 healthy children. UMCP-1 was determined using ELISA method by Quantikine kit.
**Results:** The median uMCP-1/ cr. in group IA was significantly higher compared to C ($p< 0.05$). Children from groups II and III revealed higher uMCP-1/ cr. levels when compared to MCD children and healthy controls ($p< 0.01$).
We found a strong positive correlation between the uMCP-1/ cr. and serum total cholesterol and LDL.
Assessment of the uMCP-1/ cr. according to the histological diagnosis revealed that patients with higher uMCP-1/ cr. were more likely to have FSGS or IgAN (as compared to MCD) (Odds ratio 9.27; 95% CI 2.30 – 36.35, RR 4.138; 95% CI 1.64 – 12.13).
**Conclusions:** Urinary MCP-1 was increased in both examined groups of children, however it was much higher in the course of FSGS and IgAN and correlated with cholesterol level and proteinuria.

## Abstract# 695

**Integrating Clinical and Psychosocial Knowledge through Knowledge Brokering (KB) in a Paediatric Nephrology Multiprofessional Team (MPT)** A.R. Watson,[1] G. Currie,[2] T. Starr.[2] *[1]Children's Renal Unit, NUH, Nottingham, United Kingdom; [2]University of Nottingham, Nottingham, United Kingdom.*
**Objectives:** The management of chronic kidney disease requires clinical and psychosocial knowledge to be integrated. These domains can be rather disparate and require KB across professional boundaries. We examined patterns of KB amongst the MPT to identify the barriers and facilitators to brokering clinical and psychosocial knowledge.
**Methods:** A mixed methods research design was undertaken over 3 yrs with 2 rounds of social network analysis and interviews with all involved complimented by quarterly observations at team meetings and ward rounds.
**Results:** KB is facilitated through development of social capital (SC). Organisational structures and human resources practices link knowledge domains and members laterally; i.e. structural dimension to SC. Co-location of team members and frequent interaction builds high trust relationships and are a prerequisite for effective KB at a healthcare system level; i.e. relational dimension of SC. Effective knowledge brokers in healthcare possess expertise related to the knowledge domains between which they are brokering; i.e. cognitive dimension to SC.
**Conclusions:** Clinical managers can help build necessary social capital for effective knowledge exchange through the ways in which staff are socialised into a new way of working that integrates clinical and psychosocial knowledge for patient benefit; eg ensuring the provision of psychosocial as well as clinical expertise; recruiting staff already exposed to the new way of working (eg underwent their training in the department); ensuring that situated learning across clinical and psychosocial domains ensues in meetings or ward rounds.

## Abstract# 696

**The Relative Function of Damaged Kidneys in Children Does Not Deteriorate Further at 3 Year Follow-Up; a Retrospective Analysis** A. Whittaker,[1] M.S. Moon,[1] M. Rossleigh,[1,2] A. Rosenberg.[1,2] *[1]Prince of Wales and Sydney Children's Hospitals, Sydney, NSW, Australia; [2]Medicine, University of New South Wales, Sydney, NSW, Australia.*
**Objectives:** To examine Split Renal Function (SRF) over time in children with vesicoureteric reflux (VUR) and/or urinary tract infection (UTI) where one kidney was found to contribute ≥30% to overall function on initial DMSA scan and the contralateral kidney was unscarred.
**Methods:** We identified 30 children (27 boys, 3 girls) who met the inclusion criteria and who had multiple renal radionuclide studies between January 1997 and September 2008. Information on the pathology, interventions, UTIs and scarring was recorded and SRF at 1st and last scan was compared.
**Results:** Twenty-seven boys and 3 girls were first scanned at a median of 0.8 years (0.08-13.05). Eight patients had unilateral reflux, 21 patients had bilateral reflux, and 1 patient had UTIs without reflux. High grade VUR (IV-V) was

present in 70% of poorly functioning kidneys and in 38% of contralateral kidneys. Twenty-one patients underwent reimplantation surgery and 9 were managed conservatively. SRF was a mean of 19% (11-28%) at the time of the 1st scan. Patients were followed up for a mean of 2.64 years (0.26-6.77). Follow-up SRF was 18% (9-29%) resulting in a non significant mean decrease of 1% (+4 to -13%). The mean change in renal function was not affected by the severity of the initial SRF. Scarring was focal in 19 patients and progressed to global in 2.
**Conclusions:** In the medium term there is no deterioration in SRF in children with severely malfunctioning kidneys associated with either VUR or UTI managed either surgically or conservatively. Boys are at a much greater risk of severe reflux nephropathy.

## Abstract# 697

**Risk Factors of Cardiovascular Disorders in Children with Nephrotic Syndrome** I. Wikiera-Magott,[1] M. Hurkacz,[2] D. Kubicki,[1] D. Zwolinska.[1] *[1]Department of Pediatric Nephrology, Medical University, Wroclaw, Lower Silesia, Poland; [2]Department of Clinical Pharmacology, Medical University, Wroclaw, Lower Silesia, Poland.*
**Objectives:** Nephrotic syndrome (NS) is connected with metabolic disorders and plasma volume changes that create the risk factors of early cardiovascular disturbations.
The aim is to assess the selected markers of epithelium condition in children with NS and to estimate the potential role of this factors in atherosclerosis.
**Methods:** 76 children ( 45 boys and 31 girls ) with NS in the course of primary glomerulonephritis, age range 2 -17 years were examined. The time of disease duration 1 – 109 months (average time 23,5 months).We estimate the concentration of endothelin, homocysteine, von Willebrand factor (vWF), CRP, ADMA in every child with NS at the time of remission and relapse. Further more we assess the kidney function, lipid and protein metabolism and blood pressure. 20 children with nocturnal enuresis served as the control group.
**Results:** The concentration of endothelin, homocysteine, vWF in children with NS at the time of remission and relapse were statistic higher ($p<0,05$) than in the controls. The highest values of homocysteine and endothelin were found in children with hypertension. We didn't observe significant difference of the ADMA and CRP levels in both groups. Our study showed 2 positive correlations between the time of the disease and the concentration of endothelin, vWF and homocysteine and between frequency of relapses and endothelin concentration.
**Conclusions:** 1 In children with NS there is higher risk of cardio-vascular disorders.
2.The risk of cardio-vascular disorders depends on time of disease duration and frequency of relapses.

## Abstract# 698

**Haemolytic Uremic Syndrome in New Zealand Children. A Nationwide Surveillance Study from 1998-2009** W. Wong, M.C.M. Morris, T. Kara, J. Ronaldson. *Nephrology, Starship Children's Hospital, Auckland, New Zealand.*
**Objectives:** Diarrhoea associated haemolytic uremic syndrome (D+HUS) is one of the most common causes of acute renal failure requiring acute dialysis. In New Zealand, HUS has been a disorder under national surveillance since 1998.Results of 12 years of national surveillance are presented.
**Methods:** All paediatricians were requested to notify cases of children 0 – 15 years of age satisfying the case definition of HUS to the New Zealand Paediatric Surveillance Unit. An initial questionnaire was sent to reporting paediatricians requesting relevant information and 12 month follow up information was sought. Shiga toxin producing Escherishia coli (STEC) infections were monitored through a mandatory notification system.
**Results:** The incidence of STEC infections rose from 2.6 per 100,000 in 1998 to 15 per 100,000 in 2009 in children age < 5 years. There were 94 (D+HUS) cases. Mean age at diagnosis was 3.5 years (95% CI 2.9-4.1). E coli 0157 H7 was identified in 46 (48.9%). Mean time from onset of symptoms to diagnosis was 6.8 days (CI 6.0-7.6). Acute dialysis was required in 57 children (60.6%) for a mean of 5.8 days.(95% CI 4.4-7.2) Multivariate model showed delay to diagnosis, serum sodium < 130mmol/L during illness, neutrophil count at presentation of greater 20.0 x 109 predicted severe renal failure. Twelve month follow up in 82 patient,(87 %) and showed that 24 (29.7%) had persistent urinary abnormalities, hypertension and or impaired GFR.
**Conclusions:** Conclusion.
The incidence of STEC in infections has been steadily rising in young children resulting in HUS which is a significant cause of acute morbidity and a cause of long term renal morbidity in New Zealand children.

## Abstract# 699

**Case Report: Microscopic Polyangiitis with Finger Amputation and Bowel Perforation** N. Cetin, B. Yildiz, N. Kural. *Pediatric Nephrology, Eskisehir Osmangazi University, Faculty of Medicine, Eskisehir, Turkey.*
**Objectives:** Microscopic Polyangiitis (MPA) is an autoimmune disease characterized by pauci-immune, necrotizing and small-vessel vasculitis without necrotizing-granulomatous inflammation.
**Methods:** We describe a patient with ANCA (-) MPA with bowel perforation and multiple vasculitic skin lesions that resulted in fingers amputation.
**Results:** A four months old girl with no significant medical history was admitted to the hospital because of purpuric lesions of the left fifth finger and fever. Her blood pressure was 130/90 mmHg. Laboratory examinations were revealed coombs positive anemia, proteinuria, narrowing ulnar artery with blood flow deceleration in doppler ultrasound, a neutrophilic vasculitis involving capillaries, venules, and arterioles in skin biopsy. The complements, protein S and C and anticardiolipin antibodies levels were normal. Factor 5 Leiden and prothrombin gene mutation were negative. The patient was treated with ilioprost, pulse methyl prednizolone and cyclosporin. Despite this treatment, gangrene occurred on the second, third, fourth and fifty left fingers. New purpuric lesions occurred on the third and fifty right fingers, also. Gangrene was treated with finger amputation and purpuric lesions on right fingers resolved with this treatment. Later diagnosis of our patient was bowel perforation with abdominal distension and free air on x-ray. Therefore, patient was operated but bowel biopsy was non- specific for MPA.
**Conclusions:** In conclusion, ANCA negative MPA is rare in children but should be promptly diagnosed because it may cause mortality and morbidity if left untreated.

## Abstract# 700

**Long-Range Follow up of Pediatric Patients Diagnosed as Steroid Resistant Nephrotic Syndrome Treated with Low Dose Ciclosporin** T. Yokoyama, Y. Onodera. *Department of Nephrology, Sapporo Tokushukai Hospital, Sapporo, Hokkaido, Japan.*
**Objectives:** A long-range follow up study was performed to clarify the clinicopathological changes in long-term and low dose ciclosporin monotherapy in four patients with steroid resistant nephrotic syndrome (SRNS).
**Methods:** Histological diagnosis showed minimal change (MC) in two patients, another had IgA nephropathy (IgAN) and the fourth patient had membranoproliferative glomerulonephritis (MPGN). Patients' age at initial onset ranged from 7 to 16 years old. Ciclosporin monotherapy for at least three months was started at 3-7mg/kg/day at relapses of nephrotic syndrome that occurred after using large amounts of steroids. When proteinuria decreased from 4.2-15.4g/day to below 0.3g/day, ciclosporin dosage was reduced to below 1.0mg/kg/day. Renal biopsies were performed before steroid therapy and at 3 to 17 years after initiation of the therapy.
**Results:** 1) A marked reduction of proteinuria (0.1-0.3g/day) was observed in all patients. 2) Post-therapy biopsies after 3-17 years of ciclosporin treatment showed mild tubular atrophy accompanying interstitial fibrosis in only one case, the patient with IgAN. In the two cases with MC, no nephrotoxicity was detected. In the patient with MPGN, gradual decreases in endocapillary proliferation and in double contours were observed. 3) The complete remission could be obtained despite ciclosporin monotherapy being terminated 5 to 13 years after the onset of nephrotic syndrome in all cases.
**Conclusions:** Long-term and low dose ciclosporin monotherapy proved to be quite effective for patients with SRNS because of the low incidence of adverse effects such as nephrotoxicity and a better quality of adult life could be achieved.

## Abstract# 701

**Value of Renal Resistive Index as an Early Marker of Diabetic Nephropathy in Children with Type 1 Diabetes** D.M. Youssef,[1] F.M. Fawzy.[2] *[1]Pediatrics, Zagazig University, Zagazig, Zagazig, Egypt; [2]Radiology, Zagazig University, Zagazig, Zagazig, Egypt.*
**Objectives:** Is to evaluate the renal resistive index in children with type 1 diabetes versus normal children to predict early DN.
**Methods:** we studied 25 patients with T1DM as group A (15 females/10 males, mean age 10.8±2.2) years old, and duration of diabetes 5±1.1 years, versus Group B 20 healthy children (12 females/8 males, mean age 11.6±2 years old), comparing both groups as regard there age, serum Creatinine, albumin excretion rate (AER), GFR, HbA1c and mean renal RI of both kidneys.
**Results:** we found increase in mean RI in diabetic patients versus healthy children as the mean RI in group A was 0.64 while it was 0.58 in group B with ($p<0.000$).

[table1] Comparison between patients group and healthy group

| | Group A n=25 | Group B n=20 | p |
|---|---|---|---|
| HbA1c % | 8.9±0.8 | 5.2±0.57 | 0.000* |
| AER mg/24h | 13.6±3.6 | 11.8±3.9 | 0.113 |
| Mean renal RI | 0.64±0.55 | 0.58±0.0.28 | 0.000* |
| GFR ml/min/1.73m2 | 140.6±9.3 | 113.9±15.6 | 0.000* |

*p less than 0.05 significant This increase in RI had a positive correlation with duration of diseases, HbA1c level and GFR, but there was no correlation with serum Creatinine or with AER.

[table 2]Correlation between different parameters and mean renal RI

| | r | p |
|---|---|---|
| Duration of disease in years | 0.52 | 0.000* |
| HbA1c% | 0.70 | 0.000* |
| AER mg/24h | 0.01 | 0.939 |
| GFR ml/min/1.73m2 | 0.41 | 0.005* |

**Conclusions:** we concluded that RI is increased in TIDM as early as it can be a predictor marker for DN and that it may reflect increased GFR, it is increased with poor glycemic control, and it is independent to microalbuminuria.

## Abstract# 702

**How Should GFR Really Be Estimated in Children?** K. Zachwieja,[1] P. Korohoda,[2] J. Kwinta-Rybicka,[1] M. Miklaszewska,[1] J. Berska,[1] J. Bugajska,[1] J.A. Pietrzyk.[1] *[1]JUMC, Cracow, Poland; [2]AGH, Cracow, Poland.*

**Objectives:** The aim of the study was to assess the various methods of estimating GFR in children.

**Methods:** The study group consisted of 123 patients aged 2-19 (mean 12.5±4.2). GFR was calculated according to: 1. the classical Schwartz method (S1); 2. the Schwartz equation with a new coefficient (S2); 3. the new Schwartz equation based on three markers (S3); 4. creatinine clearance based on daily urine collection (DUC); 5. the Filler equation (F). The results were compared to the reference method of serum iohexol elimination technique (I).

**Results:** The best agreement in GFR was achieved between S3 and I (r=0.84, p<0.0001), as corroborated by Bland-Altman analysis. The lowest accordance was observed for DUC and I (r= 0.68, p<0.00001).The differences between the methods of Schwartz, new Schwartz, Filler, DUC and the reference iohexol method were greater for values of GFR > 60 than for GFR < 60 ml/min/1.73m$^2$. It was shown that Filler and and the classical Schwartz equation overestimated GFR by a mean of 21 and 15.5 ml/min/1.73m$^2$, respectively, in comparison to the iohexol method, and the S2 and the S3 formula underestimated GFR by 15.1 and 17.6 ml/min/1.73m$^2$, respectively. GFR according to DUC had the lowest precision and an unpredictable error. The same tendency was noted in children with BSA < 1m$^2$ and BSA> 1 m$^2$.

**Conclusions:** Estimating GFR by means of a single formula might be imprecise, especially at high GFR. The three markers Schwartz formula yields the lowest error in comparison with the reference iohexol method.

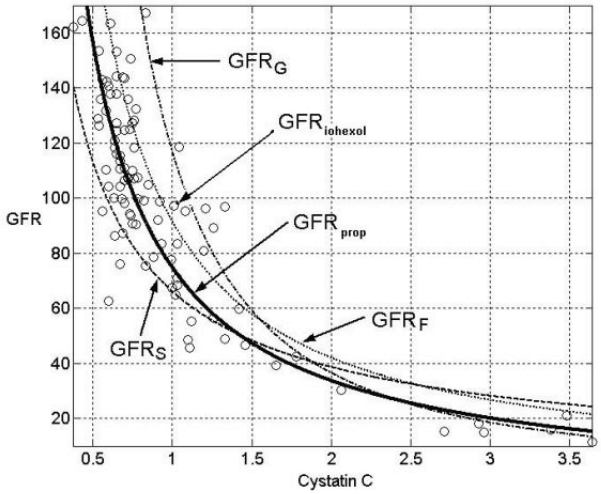
## Abstract# 703

**A New Equation Proposal for eGFR Estimation in Children Based upon Serum Cystatin C Concentrations** P. Korohoda,[1] K. Zachwieja,[2] J. Kwinta-Rybicka,[2] J.A. Pietrzyk.[2] *[1]AGH, Cracow, Poland; [2]JUMC, Cracow, Poland.*

**Objectives:** The aim of the study was to evaluate the differences the published pediatric eGFR based on cystatin C concentration (sCys) formulas results in calculations and the proposal for a new equation.

**Methods:** The iohexol elimination technique was used as reference method in the study group of 93 patients.sCys was measured by nephelometric method. The results of the following equations were analysed: 1. Filler's equation ($GFR_F = C^{-1,123} \times 91,622$); 2. Grubb's pediatric ($GFR_G = 84,69 \times C^{-1,68} \times 1,384$); 3. Schwartz's et al. 2009 ($GFR_s = 66,22 \times C^{-0,777}$). All values were in ml/min/1,73m$^2$.

**Results:** The detailed mathematical comparison was performed and the statistically significant differences among the results from equations (1), (2), (3) and the reference method were observed.




The concordance of the results was noted only in lower values of eGFR. The sCys data allowed to create a new equation : $eGFR = -7.28+82.29 \times C^{-1}$. The high correlation was proven between eGFR ac.to the new formula and to the reference method (r=0,952; p<0,00001) as corroborated by Bland-Altman figure.

**Conclusions:** Filler's, Grubb's and Schwartz's et al. (2009) formulas provide significant differences in eGFR calculations in comparison to the iohexol method. The newly proposed by us formula: $eGFR = -7.28+82.29 \times C^{-1}$ seems to be more accurate for this purpose.

## Abstract# 704

**Hypernatriuria as a Cause of Hematuria in Children** M.B. Zanolli, L.M.S. Borges, T.C. Reis, A.A.P. Silva, I.M. Araujo. *Nephrology, Marilia Medical School, Marilia, SP, Brazil.*

**Objectives:** It is known that excessive intake of sodium leading to hypernatriuria (> 3 mEq/kg/day) is associated with hypercalciuria (> 4 mg/kg/day) and/or hyperuricosuria (10 - 15 mg/kg/day). The objective was to evaluate this association in children and adolescents referred to an university clinic because of hematuria.

**Methods:** Observational and retrospective study of medical records of 177 children and adolescents presenting complaint and/or laboratory evidence of macro (76,3%) or microscopic (23,7%) hematuria, treated at the Pediatric Nefrology Clinic from May 1999 to January 2009. Of the patients studied, 117 (66.1%) were male and the mean age at initial consultation was 8.0 years (9 months to 15 years). Thrity patients were excluded from the analysis for having hematuria of glomerular origin, and 31 who did not complete the metabolic investigation. The investigation was carried out from the interview data and laboratory tests.

**Results:** Of 109 patients studied, the etiology was not detected in 19 (17%); 27 had nephrolithiasis and 63 had metabolic disorders. Of the patients with nephrolithiasis, in 7 was the cause not found and among the remaining 20 with metabolic disorders, 17 (85%) had hypernatriurua and in 6 of them (30%) it was the only finding. Among patients with metabolic disorders, 39 (62%) had hypernatriuria, which was isolated in 8 (13%).

**Conclusions:** Among the causes of non-glomerular hematuria in childhood there was a predominance of metabolic disorders in agreement with the description in literature, and the presence of hypernatriuria, probably due to high sodium ingest, was quite high. Therefore the prescription of low sodium diet for these patients should always be recommended.

## Abstract# 705

**Effectiveness and Impact of World Kidney Day 2010 Campaign: Assessment of Kids Knowledge about Kidney Disease** M.B. Zanolli, G.R. Oliveira, A.A. Negrini, V.P. Pretti, V.T.R. Ferreira, D.M. Tan. *Nephrology, Marilia Medical School, Marilia, SP, Brazil.*

**Objectives:** Raise awareness and warn children about the importance of kidney care and evaluate the level of knowledge about the prevention of kidney disease identifying the risk factors as an important strategy for health promotion.

**Methods:** The campaign theme was about diabetes and kidney in a private school in Brazil. All the kids were informed about diabetes risks and complications and kidney disease, as well as general guidance on changes in lifestyle. At the approach, 240 male and female individuals aged between 9 and 13 years of age answered a two-choice questionnaire with the options YES (quantified) or NO.

**Results:** In our sample, 97% have the habit of eating fast food or snacks at least once a week, 26% intake more than 2 liters of water per day and 90% pratice physical exercise at least twice a week. None of the children knew what creatinine was, 44% had already had urinalysis previously, 88% have a history of high blood pressure or diabetes in the family and 34% have a history of renal

disease in one of the family members. Most children evaluated the approach as very important for the prevention of diseases that affect the kidney (98%), and the initiative of the researchers (94%).
**Conclusions:** Health education and prevention for kidney disease in children is feasible and have great epidemiological significance. The children are overweight, with a high sodium intake, low fluid intake, and most are unaware of behaviors that harm kidney function. There is a deficiency in advising children in establishing rules and habits, as well as warning about the importance the family history has for risk factors.

## Abstract# 706

**Measurement of Glomerular Filtration Rate Estimated by Schwartz Formula in Children with Chronic Kidney Disease and Comparison with $^{99m}$Tc-DTPA Clearance** J. Zhang, Q. Cao, H. Xu. *Department of Nephrology and Rheumatology, Children's Hospital of Fudan University, Shanghai, China.*
**Objectives:** Radionuclide has offered an alternative method of estimating GFR that avoids some of the practical disadvantages of inulin clearance. But radioisotopic method has the disadvantage of precautions being required in handling and disposal of radioactive materials. To avoid the practical difficulties of formal measurement of clearance, several prediction formulas have been published. The most commonly used in children is the Schwartz formula. The aim of this study is to identify an adequate measurement of GFR in pediatric clinical practice.
**Methods:** $^{99m}$Tc-DTPA clearance (mGFR) was measured as GFR marker in 170 patients with different chronic kidney diseases. Estimated GFR (eGFR) was calculated via Schwartz formula. The agreement between eGFRs and mGFRs was evaluated by *Kappa* statistics. The precision was evaluated by $R^2$ from linear regression and the accuracy was measured by the percentage of GFR estimates within 30% of the mGFR ($P_{30}$).
**Results:** The mean mGFR was (73.26±22.58) ml/min/1.73m$^2$ and the eGFR was (80.35±18.79) ml/min/1.73m$^2$. The median bias was 5.28 ml/min/1.73m$^2$ ($P<$ 0.01). Pearson correlation analysis showed good correlation between mGFR and eGFR. Both of them correlated well with age and serum creatinine ($P<$ 0.05). For Schwartz formula, the *Kappa* value, $R^2$ and $P_{30}$ was 0.362, 0.573 and 69.4% respectively. The overestimation of the Schwartz formula increased while GFR decreased.
**Conclusions:** Schwartz equation shows a certain degree of accuracy and it is suitable in estimating GFR in children without radioisotopic equipment. The equation remains useful for follow-up of renal function in patients.

## Abstract# 707

**Time To Rethink about Our Renal Biopsy Indications: A Thirty-Year Review of Pathology from Renal Biopsy** Y.-B. Zheng, H. Xu. *Department of Nephrology and Rheumatology, Children's Hospital of Fudan University, Shanghai, China.*
**Objectives:** To identify the patterns of childhood renal biopsy pathology and the change over recent 30 years and to study how to improve the renal biopsy indication.
**Methods:** A retrospective study analysis of the paediatric renal biopsies performed from Jan 1979 to Jan 2010. All the cases were from Children's Hospital and were analyzed in 10-year intervals: period A (1979 to 1990), period B (1991 to 2000) and period C (2001 to 2010).
**Results:** A total of 1934 renal biopsies in 1925 patients were performed in 30 years. Mean age was 7.9±3.7 years (6m-18y). Primary GN (PGN) accounted for 68.1%, secondary GN (SGN) 16.8% and hereditary GN (HGN) 12.0%. Common causes of PGN were IgAN (16.6%) (Most were diffuse proliferative type 37.3%) and MCD (14.6%); FSGS only accounted for 1.4%. The percentage of IgAN was relatively high and that of FSGS was low. In SGN, HSPN (9.8%) (Most were focal segmental proliferative type 45.5%) ranked first and TBMN (10.5%) was the most in HGN. During the 30 years, the incidence of PGN decreased while SGN increased. In PGN, MsPGN was on the top in period A and decreased in B and C (p<0.001). MCD and IgAN had a rise in prevalence (p<0.001). In SGN, HBV-GN was sharply dropped in period C compared with B (p<0.001). HSPN and LN had no obvious change. In different age groups, the percentage of MCD declined with the age and that of the IgAN was opposite (p<0.001).
**Conclusions:** It's time for us to rethink the renal biopsy indication. For MCD with steroid dependent and frequent relapse, the renal biopsy should be performed for those who starting to use immunosuppressive drugs as CyA. The indication of renal biopsy for isolated hematuria children should be more strict.

## Abstract# 708

**Reversible Ceftriaxone-Associated Biliary Pseudolithiasis in Three Children with Renal Diseases** X. Zhong,[1] Y. Yao,[1] H. Chen,[2] X.-Y. Zhuo,[1] J. Huang,[1] H. Xiao,[1] Y. AI.[1] *[1]Department of Pediatrics, Peking University First Hospital, Beijing, China; [2]Department of Pediatrics, Second Affiliated Hospital of Zhongshan University, Ghuangzhou, China.*
**Objectives:** To study the clinical characteristics of ceftriaxone-associated biliary pseudolithiasis in children with renal diseases.
**Methods:** We retrospectively reviewed three children with renal diseases who developed biliary pseudolithiasis when treated with ceftriaxone.
**Results:** Case one was an 11-year-old boy. The initial diagnosis was primary nephrotic syndrome. Ceftriaxone was administered intravenously at a dose of 50 mg/kg/d for gastroenteritis. After that he complained of nausea and loss of appetite. Abdominal sonogram obtained on day 3 of ceftriaxone therapy revealed gallbladder sludge. Case two was a 10-year-old boy. The primary diagnosis was post-streptococcal glomerulonephritis with acute renal failure. He was treated with 30 mg/kg/d intravenous ceftriaxone for gastroenteritis. Then he complained of abdominal pain with positive Murphy's sign. Gallstone was detected on day 6 of ceftriaxone therapy. Case three was an 12-year-old boy. The primary diagnosis was nephrotic syndrome. He was treated with 40 mg/kg·d ceftriaxone for gastroenteritis. Gallbladder lithiasis was detected on day 17 of ceftriaxone therapy. After cessation of ceftriaxone treatment, the symptoms gradually disappeared, with complete sonographic resolution in all of the three children. None of them showed abnormal gallbladder ultrasonography before ceftriaxone was administered.
**Conclusions:** The risk of developing ceftriaxone-associated biliary pseudolithiasis might increase in patients with renal diseases who are treated with ceftriaxone.

## Abstract# 709

**Continuous Hemofiltration in the Treatment of Acute Renal Failure in Children** F. Zhong, Y. Gao. *Nephrology, Guangzhou Women and Children's Medical Center, Guangzhou, Guangdong, China; Nephrology, Guangzhou Women and Children's Medical Center, Guangzhou, Guangdong, China.*
**Objectives:** Observation on the effect and characteristic for the treatment of children with acute renal failure by continuous hemofiltration.
**Methods:** Clinical data of 42 children patients with acute renal failure treated by continuous venovenous hemofiltration(CVVH)and 51 children patients treated by HD(hemodialysis) in Guangzhou Children's Hospital were analyzed retrospectively.
**Results:** Original disease of the HD group mainly made by glomerulonephritis, while the CVVH group appeared serious infection. After blood purification, the Bun, Scr and the serum levels of $Na^+$ in CVVH group and HD group were significantly improved than before it($P$<0.05). After blood purification, the comparison on Bun, Scr, the serum levels of $Na^+$ between CVVH group and HD group shows no difference($P$>0.05). In CVVH group, metabolic acidosis, hyperkalemia and hypokalemia all be corrected, however 9 patients with metabolic acidosis and 1 patient with hyperkalemia in group HD are not be corrected. Capacity of ultrafiltration in CVVH group is more than HD group($P$<0.05), but less in appearing these complication of hypotension and imbalance syndrome.
**Conclusions:** Continuous hemofiltration is an effective and safe renal replacement therapy to children with acute renal failure.

## Abstract# 710

**The Multi-Center Cross-Sectional Study of Urine Screening in Urban and Rural Young Children in Beijing** X. Zhong,[1] J. Ding,[1] P. Zhang,[2] X. Chen,[3] Y. Shen,[4] H. Song,[5] S. ZHU,[1] C. Yao,[1] G. Xu,[1] X. LIU,[1] H. Wang,[4] W. Wang,[5] Y. Xu.[1] *[1]Peking University First Hospital, Beijing, China; [2]Chinese Center for Disease Control and Prevention, Beijing, China; [3]Beijing Maternal and Child Health Care Hospital, Beijing, China; [4]Beijing Children's Hospital, Beijing, China; [5]Peking Union Medical College Hospital, Beijing, China.*
**Objectives:** To study the epidemiological characteristics of the screening urinalysis among children younger than 3 years old from urban and rural districts in Beijing.
**Methods:** The children were selected by stratified, cluster and random sampling. If the first urinalysis was abnormal, a second screening would be performed to confirm the result. The guardians of the children were asked to fill out the uniform questionnaire.
**Results:** Approximate 12,105 children (from 4 urban districts and 4 rural districts) have been studied. The analysis of the questionnaire showed that 27.1% of the guardians knew nothing about the impairment caused by kidney diseases (urban, 20.4% vs rural, 33.3%; P<0.05). In the first screening, the prevalence of abnormal urinalysis was 12.2% (urban, 11.1% vs rural, 13.2%; $P$<0.05). During the second screening, abnormal urinalysis was identified in 10.0% of the subjects (urban, 14.2% vs rural, 7.0%; $P$<0.05). The overall prevalence of confirmed

abnormal urinalysis was 0.98% (urban, 1.17% vs rural, 0.79%; P<0.05). Female and children younger than 12-month-old were independently associated with abnormal urinalysis.
**Conclusions:** The abnormal urinalysis in urban children was more common than that from rural districts. The rural guardians had less knowledge about the importance of kidney diseases.

## Abstract# 711

**Results of DMSA Scanning in Children with Vesicoureteric Reflux and Reflux Nephropathy** I.V. Zorin,[1] A.A. Vyalkova,[2] A.G. Miroshnichenko,[1] A.V. Zorin,[2] A.R. Zabirova.[2] *[1]Pediatric Nephrology, Orenburg Pediatric Clinic N 6, Orenburg, Russian Federation; [2]Pediatrics, Orenburg Medical academy, Orenburg, Russian Federation.*
**Objectives:** The aim of the study was to determine parameters of DMSA scanning of patients with vesicoureteric reflux (VUR), reflux nephropathy (RN).
**Methods:** We examined 150 children with RN and VUR. All patients underwent ultrasound, X-ray and DMSA scanning. They were divided into 3 groups:
I – children with unilateral RN A according to classification of Smellie J. et all, 1975 (n=50);
II – children with unilateral RN D according to classification of Smellie J. et all, 1975 (n=50);
III – children with VUR without renal damage (n= 50)
**Results:** We established that data of DMSA scanning (time of the maximal accumulation 11,8±0,91 sec, maximal activity 189,3±20,4 sob/sec, mean velocities of accumulation 29,1±1,9 mm/sec, the contribution to the common accumulation 45,3±3,5 %) are characteristic for patients with VUR without renal scars. Data of DMSA scanning (time of the maximal accumulation 7,5±0,86 sec, maximal activity 81,5±11,9 sob/sec, mean velocities of accumulation 11,66±2,35 mm/sec, the contribution to the common accumulation 34,27±3,03%) are characteristic for patients with RN A. Data of DMSA scanning (time of the maximal accumulation 6,43±0,84 sec, maximal activity 65,8±12,04 sob/sec, mean velocities of accumulation 7,72±1,07 mm/sec, the contribution to the common accumulation 26,86±2,63 %) are characteristic for patients with RN D. The ranges of DMSA scanning were significant different between children from comparing groups (p<0,05).
**Conclusions:** Data of DMSA scanning can be used for early diagnostics of scarring and for diagnostics of progression of renal scarring.

## Renal Replacement Therapy

## Abstract# 712
**(O-89)**

**Cardiac Geometry in Children Receiving Chronic PD: A Study of the International Pediatric Peritoneal Dialysis Network (IPPN)** S.A. Bakkaloglu, D. Borzych, B.A. Warady, F. Schaefer, IPPN Registry Investigators. *Gazi University, Ankara, Turkey.*
**Objectives:** Left ventricular hypertrophy (LVH) is a risk factor and intermediate endpoint of cardiovascular (CV) morbidity. We aimed to assess the prevalence, incidence and predictors of LVH in pediatric PD patients.
**Methods:** LVH was defined by the 95$^{th}$ percentile of the Khoury LVMI percentiles, using height age to account for growth retardation.
**Results:** 507 children followed in 25 countries were analyzed. 39% of the patients had office hypertension. The overall LVH prevalence was 48.1% (30.8 concentric and 17.4% eccentric); 21.3% had concentric remodeling. Longitudinal analysis of 128 pts revealed 33 % incidence of de novo LVH and a 45 % incidence of LVH regression per year on dialysis. Transformation to and reversion from concentric geometry were equally common (37%).
By logistic regression analysis, hypertension, obesity, the use of the CAPD, renal disease other than hypodysplasia, and hyperparathyroidism were risk factors for the presence of LVH. The use of RAS antagonists and a high total fluid output were protective from concentricity. The risk of LVH (persistent/de novo) at one year was increased by systolic BP SDS and reduced in children with renal hypodysplasia.
**Conclusions:** LVH was highly prevalent among pediatric PD patients. Dynamic changes were observed during follow up in favour of LVH regression with better BP control. Hypodysplasia appears to be protective against LVH, likely due to lower BP and polyuria even on dialysis. Obesity, hyperparathyroidism and fluid overload are additional, potentially modifiable determinants of LVH. The use of APD modalities and RAS antagonists may be associated with better CV outcomes.

## Abstract# 713
**(O-90)**

**Risk Factors of Losing Residual Renal Function (RRF) in Children on Peritoneal Dialysis (PD)** K.H. Han,[1] H.J. Choi,[1] H.K. Lee,[1] Y.H. Jung,[1] S.E. Lee,[1] H.G. Kang,[1] H.I. Cheong,[1] Y. Choi,[2] I.S. Ha.[1] *[1]Pediatrics, Seoul National University College of Medicine, Seoul, Korea; [2]Pediatrics, Inje University Haeundae Paik Hospital, Pusan, Korea.*
**Objectives:** Maintaining RRF is known to reduce the cardiovascular complications and mortality in patients on dialysis. However, only a few studies on RRF have been performed in children. Thus we investigated the risk factors of deterioration of RRF in children on PD.
**Methods:** Fifty-five children (M:F 36:19, median age 13 years) with RRF and on chronic PD were retrospectively studied. RRF deterioration rates in 136 interval periods were measured by dividing Δ renal Kt/V, Δ renal Ccr and Δ urine volume per body surface area (UV/BSA) by the length of the period. Clinical and biochemical parameters of the periods were analyzed to determine the risk factors of losing RRF.
**Results:** Presence of hypertension by the definition of NHBPEP correlated with more rapid decline of renal Kt/V (P=0.017), renal Ccr (P=0.010) and UV/BSA (P=0.007); high systolic BP correlated with all the three indices while high diastolic BP correlated only with UV/BSA. Low hemoglobin level was associated with loss of renal Kt/V (P=0.023) and Ccr (P=0.002), but not with UV/BSA. However, gender, age, underlying disease, baseline RRF, BMI standard deviation score, PD modalities and duration, PD solutions, peritoneal permeability, and echocardiographic indices were not associated with the decline rate of RRF.
**Conclusions:** Hypertension and anemia are associated with more rapid loss of RRF in children on PD. Prospective studies are necessary to prove the causal relationship between these factors and RRF loss.

## Abstract# 714
**(O-91)**

**A Hospital-Based Nocturnal Hemodialysis Program for Children and Adolescents: Prospective 4-Year Data** A. Hoppe, C. Puttkamer, M. Zimmering, I. Hirte, S. Schley, J. Gellermann, B. Utsch, S. Briese, J. Thumfart, U. Querfeld, D. Müller. *Pediatric Nephrology, Berlin, Germany.*
**Objectives:** To evaluate the advantages and disadvantages of a nocturnal hemodialysis regimen in children and adolescents.
**Methods:** A hospital-based NHD program was started as a regular treatment option for maintenance HD patients in 2006. Over a period of 48 months, 15 patients (12 to 17 years old) were prospectively enrolled in this program. NHD was performed for 8 hours, 3 nights per week and treatment was supervised at all times by a pediatric nephrologist and dialysis nurses. Central venous lines or fistulae were used for blood access**.** Uremia-associated parameters, medication dosage as well as dietary parameters were regularly monitored. The SF36 test was used to evaluate the quality of life. Results were compared to data from age-matched patients treated with conventional HD (3x4 hours/week).
**Results:** Compared with controls, NHD patients had a significant decrease in predialytic mean arterial blood pressure, as well as in phosphate levels, calcium-phosphate product and parathyroid hormone levels. The median Kt / V value was 2.15 in NHD compared to 1.47 in HD patients. Dietary and fluid restrictions could be lifted in all NHD patients and the dosage of phosphate binders, antihypertensive medication, and erythropoietin could be reduced. Quality of life and school attendance improved in all children treated with NHD.
**Conclusions:** NHD allows a better control of uremia - associated parameters as well as free dietary and fluid intake, a reduction of medication, greater individual developmental opportunities (education, training) and an improved quality of life.

## Abstract# 715
**(O-92)**

**Peritoneal Protein Loss in Nephrotic Syndrome on Peritoneal Dialysis** H.G. Kang, I.S. Ha, H.I. Cheong, Y. Choi. *Pediatrics, Seoul National University College of Medicine, Seoul, Korea.*
**Objectives:** The pathophysiologic mechanism of nephrotic syndrome is not yet known clearly. At least in some cases, certain 'circulating factors' are thought to increase the glomerular protein permeability, which may have systemic effect on peritoneal membrane. We evaluated the loss of protein through peritoneal membrane in patients on peritoneal dialysis due to the end stage renal disease (ESRD) caused by steroid resistant nephrotic syndrome (SRNS).
**Methods:** We reviewed retrospectively the medical records of 26 pediatric patients on peritoneal dialysis ensued during the period from 2001 to 2007 at our clinic. Twelve patients had SRNS, while 14 patients had ESRD caused by the congenital anomalies of urinary system.
**Results:** While the other parameters including nPNA indicating the adequacy of protein intake were similar between the two groups, serum albumin was lower in SRNS patients than the non-SRNS patients (3.7 ± 0.3 g/dL vs. 4.0 ± 0.4 g/dL, *P*= 0.021) and peritoneal protein loss was higher (3044.4 ± 837.6 mg/m$^2$/day vs.

1791.6 ± 1244.0 mg/m²/day, *P*= 0.007). The protein permeability of the peritoneal membrane measured by the ratio of total protein concentration in dialysate to plasma was twice as high in SRNS patients as the non-SRNS (1.06 ± 0.46 % vs. 0.58 ± 0.43 %, *P*= 0.010). After 1 year, peritoneal protein loss increased in both patient groups, but to a significantly greater degree in non-SRNS patient (*P*= 0.023).
**Conclusions:** The results of our study support the notion that in nephrotic syndrome there are some 'circulating factors' with the systemic effect. In these patients more meticulous nutritional support and close monitoring on the nutrition are required.

## Abstract# 716
**(O-93)**
**Global Outcomes of Anemia Management in Dialyzed Children: Results from the IPPN Registry** D. Borzych,[1] Y. Bilginer,[2] B.A. Warady,[3] F. Schaefer and Coinvestigators.[1] *[1]Center for Pediatric Medicine, Heidelberg, Germany; [2]Hacettepe University, Ankara, Turkey; [3]Children's Mercy Hospital, Kansas City, MO, United States.*
**Objectives:** The IPPN collects clinical, biochemical and pharmacological information from children on CPD. This analysis assessed practices and outcomes of anemia management.
**Methods:** Data from 1,138 pts. from 26 countries followed for 845 pts-yrs were analyzed. Data was time-averaged for statistical analysis.
**Results:** Median hemoglobin (Hgb) was 11 g/dl (IQR 10.1-12); 45% of pts had Hgb below recommended targets (>10 for age<2y, >11 for age>2y), varying regionally from 38% in Europe to 62% in Turkey (p<0.0001). Age, PTH>500 ng/ml, low serum albumin, the use of bio-incompatible PD fluids and high ESA dose were independent predictors of sub-target Hgb.
ESAs were administered in 92% of pts (EPO-alfa 40%; EPO-beta 39%; darbepoetin 13%), without differences between drugs in achieved Hgb. The weekly ESA dose was inversely correlated with age (r=-0.26, p<0.0001).
84% of pts received iron supplements (71% oral, 13% iv). Iron administration mode was not predictive of Hgb or EPO sensitivity. Serum ferritin was <100 in 29%, 100-500 in 56% and >500 in 15% of pts. Ferritin levels increased with age (r=0.19, p<0.0001) and decreasing urine output (r= -0.12, p=0.008). Serum ferritin was inversely correlated with Hgb levels (-0.13, p<0.005), suggesting that high serum ferritin indicates hyporegeneratory anemia.
**Conclusions:** While anemia management in pediatric PD patients has improved in recent years, sub-target Hgb levels are still found in a large fraction of patients. Age, hyperparathyroidism, and malnutrition/inflammation are critical determinants of ESA responsiveness.
**DISCLOSURE:** *Warady, B.A.*: Consultant, Amgen, Takeda and Amag. *Schaefer and Coinvestigators, F.*: Consultant, Amgen.

## Abstract# 717
**(O-94)**
**Buffer Dependent Regulation of Aquaporin-1 Expression and Function in Human Peritoneal Mesothelial Cells** Y. Zhai,[1,2] J. Bloch,[1] M. Hoemme,[1] G. Eich,[1] T. Hackert,[3] H. Xu,[2] F. Schaefer,[1] C.P. Schmitt.[1] *[1]Division of Pediatric Nephrology, Center for Pediatrics and Adolescent Medicine, Heidelberg, Germany; [2]Division of Nephrology and Rheumatology, Children's Hospital of Fudan University, Shanghai, China; [3]Division of Surgery, Heidelberg University Medical Center, Heidelberg, Germany.*
**Objectives:** PH neutral peritoneal dialysis fluids (PDF) with low glucose degradation product (GDP) contain lactate or bicarbonate buffer, the specific impact on peritoneal membrane is unknown. We therefore investigated buffer dependent regulation of aquaporin-1 (AQP1), a protein essential for transcellular water transport and cell migration in human primary peritoneal mesothelial cells (HPMC).
**Methods:** We incubated HPMC and human umbilical vein endothelial cells with low GDP, pH neutral PDF containing bicarbonate and lactate buffer.
**Results:** BPDF increased AQP1 mRNA and protein (209±80 and 193±27% vs. medium), whereas LPDF reduced it (48±18 and 25±12 %, p<0.05). Half life of mRNA was not altered. Immunofluorescence revealed buffer dependent regulation of AQP1 in the cell membrane. Addition of bicarbonate to LPDF reversed the inhibitory effect on AQP1 expression. BPDF increased the number of migrating HPMC, migration velocity, and increased wound healing (p<0.05). Gene silencing by siRNA demonstrated AQP1, but not AQP3 dependency of HPMC migration and of dialysate induced changes in migration capacity. PET in rats revealed increased sodium sieving with BPDF.
**Conclusions:** Bicarbonate buffer enhances AQP1 expression in HPMC and AQP1-dependent cell migration. The buffer composition of PDF may thus have important implications with respect to long term peritoneal membrane integrity and function.

## Abstract# 718
**(O-95)**
**Renal Replacement Therapy in Infants with Chronic Renal Failure in the First Year of Life** M. Wedekin, J.H.H. Ehrich, L. Pape. *Pediaric Nephrology, Medical School of Hannover, Hannover, Germany.*
**Objectives:** Although results of renal replacement therapy (RRT) in small children have improved during recent years, data about RRT in neonates are scarce.
**Methods:** In a retrospective study, we analyzed the outcome of infants who had chronic kidney disease and started RRT within their first year of life. Between 1997 and 2008, all 29 infants who were younger than1 yr, had end-stage renal failure, and underwent RRT (dialysis or transplantation) at Hannover Medical School were analyzed for up to 12 yr.
**Results:** Twenty-seven of 29 infants with chronic kidney disease received peritoneal dialysis, starting at a mean age of 112 d; two children received preemptive renal transplantation (RTx). During follow-up, 21 of 29 children survived with RTx. The 5-yr patient and graft survival rate after RTx was 95.5%. Six of 29 children died, one with a functioning graft and five while on peritoneal dialysis. The main causes of death were severe cardiovascular and cerebral comorbidities. The mean GFR at last follow-up of patients who underwent RTx (mean time after RTx 5.1 yr) was 63.2 ml/min per 1.73 m2.
**Conclusions:** RRT in infants who are younger than 1 year offers excellent chances of survival and should be offered to all infants who do not have severe, life-limiting extrarenal comorbidity. Contrary to previous observations, the long-term outcome of infants may be comparable to that of older children who undergo RRT.

## Abstract# 719
**(O-96)**
**Continuous Erythropoietyn Receptor Activator Use for the Treatment of Anemia in Pediatric Patients under Chronic Peritoneal Dialysis. A Preliminary Report** F. Cano, C. Alarcon, C. Lizama, M. Gonzalez, A.M. Lillo, P. Arellano, M. Azocar. *Pediatric Nephrology, Luis Calvo Mackenna Childrens Hospital,University of Chile, Santiago, Chile.*
**Objectives:** to evaluate the efficacy and safety of CERA in the management of anemia in PD children.
**Methods:** Prospective protocol in stable PD children. Patients under twice-a-week erythropoietyn (EPO) were converted to Mircera®, subcutaneous doses scheduled each 2 weeks for 6 months. The main goal was to achieve and maintain haemoglobin (Hb) levels between 11-13 g/dl. Exclusion criteria: ferritin<100 ng/ml, Hb saturation <20%, paratohormone >500 pg/ml, concomitant acute illness. Initial dose was 0.5 mcg/kg. An hemogram was evaluated each 15 days to adjust doses. Each month were evaluated ferritin, Hb saturation, paratohormone, and dialysis dose (KtV). Descriptive and correlation statistics were used, p<0.05 was considered.
**Results:** 16 children, 9.7±3.6 y.o, 11 males, were included. Fourteen children completed 4 months of follow-up. Pretreatment haemoglobin level was 10.5±1.52 g/dl. Pretreatment EPO dose was 100-160 U/kg/dose twice a week. At day 30, haemoglobin decreased to 10.4±1.2 g/dl, CERA dose was 1.23±0.4 mcg/kg. Target haemoglobin level was reached at day 90, 11.3±1.28 g/dl, final dose was 1.71±0.49 mcg/kg. By 4 months of therapy, 5 pts have not reached target haemoglobin. CERA dose was holded and administered once a month in 2 patients because an increase in Hb >13 g/dl. Adverse effects were not observed.
**Conclusions:** CERA therapy is an effective and safe treatment in maintaining hemoglobin levels when administered twice up to once a month in PD children. Doses required are higher than published adult experiences.

## Abstract# 720
**Peritoneal Dialysis Using Improvised PD Catheter and Self-Constituted Dialysis Solution** S. Antwi. *Paediatric Nephrology Unit, Dept of Child Health, School of Medical Sciences, Kwame Nkrumah University of Science and Technology/Komfo Anokye Teaching Hospital, Kumasi, Ashanti, Ghana.*
**Objectives:** In developing countries, many children die from ARF largely due to lack of facilities for dialysis. But peritoneal dialysis (PD) is a simple procedure that only requires the instillation into the peritoneum of solution of appropriate constituents. In this report, I share my experience of performing acute PD using improvised PD catheter and self-constituted PD solution.
**Methods:** 1-year old girl was referred on account of anuria despite fluid and diuretic challenge. On arrival, an in-situ urinary catheter only had scanty, concentrated urine limited to catheter lumen. Acute PD was performed using size 12 thoracic trocar catheter and self-constituted PD fluid.

Table 1: 2.5% PD solution formula (modified from Red Cross hospital's protocol, S/Africa)

| Ingredient | 5% dextrose | 0.9% NaCl | 8.4% NaHCO3 | 10% Ca gluconate | 20% NaCl | Heparin | Cefotaxime |
|---|---|---|---|---|---|---|---|
| Amount in 1-litre dialysis solution | 500ml | 500ml | 40mls | 7.5mls | 8mls | 1000 U | 125mg |

**Results:** For 31 days, urine output remained < 0.15ml/kg/hr. The PD functioned effectively with significant improvement in the biochemical parameters.

Table 2: Serum chemistry flow chart.

| Day on PD | Urea mmol/l | Creatinine µmol/l | Na+ mmol/l | K+ mmol/l | PO4- mmol/l | Blood gas | Urine output/ 24hrs |
|---|---|---|---|---|---|---|---|
| 0 (pre-dialysis) | >40 | 895 | 132 | 6.4 | 2.9 | PH 7.2 SBC 8.1BE -19 | 0 ml |
| 31 | 14.6 | 133 | 124 | 5.1 | 1.9 | pH 7.4 SBC 19.4 BE -4 | 10 mls |

**Conclusions:** Acute PD can still be performed in areas where traditional PD catheters and solutions are not available.

## Abstract# 721

**Procalcitonin Serum Levels in Hemodialyzed Children; a Useful Marker for Bacterial Infection?** L. Tostivint, B. Aoun, C. Azema, T. Ulinski. *Pediatric Nephrology, Armand Trousseau Hospital - APHP - UPMC, Paris, France.*

**Objectives:** Infectious complications are a major problem in hemodialyzed (HD) children. Differentiation between bacterial and viral infections is often difficult. In order to treat patients rapidly a sensitive and specific biological marker would be of interest. Procalcitonin (PCT) has been recognized as an early, sensitive and specific marker for bacterial infections in children with normal renal function.

**Methods:** We have retrospectively analyzed PCT serum levels in end-stage renal disease (ESRD) children on hemodialysis who presented with febrile episodes attributed to bacterial infections or presumed viral infection.

**Results:** Twenty-three patients (age 12.3±6.2 years) were included and 35 febrile episodes were identified in 16 out of 23 patients between 2006 and 2009.

The mean interval of a febrile episode after the onset of HD was 3.9±4.2 months. Mean interval between HD onsetand a first bacterial infection was 1.9 months (8 days - 2 months)

PCT serum levels during bacterial infections were higher (101.5 ng/ml; 1.18-976.4) than those detected during seven non-bacterial (presumably viral) infections with negative blood culture and absence of infectious focus (35; 5.27- 510 ng/ml; p<0.0002). Serum PCT and CRP levels were highly correlated (p<0.001).

PCT has a small molecular weight and is eliminated partially by glomerular filtration.

**Conclusions:** Reduction of renal PCT clearance in children on HD is probably the reason for increased PCT levels even in presumably viral infections. A smaller stimulation of PCT synthesis during viral infections produces a dramatic increase of PCT serum levels. Therefore, PCT levels have to be interpreted with caution in ESRD children.

## Abstract# 722

**Haemodialysis in Children Weighing Less Than 15 kg in past 10 Years: Challenges and Outcomes** M. Bates, M. Kinlough, M. Riordan, A. Awan. *Nephrology, Children's University Hospital', Temple Street, Dublin, Ireland.*

**Objectives:** To review demographics, complications and outcomes of children less than 15 kg dialysed at National Haemodialysis center.

**Methods:** Retrospective and prospective analysis of 11 children who required Haemodialysis (HD) from 2000 onwards.

**Results:** Mean age of these 11 children commencing Haemodialysis (HD) was 23.9 months (range 5-64months). Mean duration on HD was 23.7months (range 5-50months). Mean weight was 9.8 kg at the start of HD (range 7.5-14.2 kg). 7 patients had renal dysplasia, 2 bilateral wilm's tumor, 1 congenital anapheric, 1 infantile oxalosis. 8 patients were switched from peritoneal dialysis (PD) to HD. Central Venous Catheters (CVL) used were10 Kimal and1Quinton permcath, placed in right internal jugular in all. 6 CVLs were changed, 3 due to breakages, 1 pulled by patient, 2 revised due to malposition. 3 CVLs had confirmed thrombus and were salvaged with Tinzaparin subcutaneous therapy daily for 4 weeks. One episode of CVL infection due to strep was successfully treated. Poor growth remains a major challenge for this particular cohort, 10 patients are/were on Nasogastric or Peg feeds. 1 had renal osteodystrophy and required osteotomies. 2 patients had hypertension and required oral medications. 7 patients have been transplanted, 1 switched backed to PD after 50 months on HD.

**Conclusions:** Haemodialysis is an erroneous but suitable option for children less than 15 kg. CVL breakages were the main reason for changing lines in our patients. Very low CVL infection in our unit is a result of strict adherence to our CVL access protocol. CVL thrombus can be safely and adequately resolved with Tinzaparin in renal failure children receiving HD.

## Abstract# 723

**Renal Replacement Therapy in Children with Hemolytic Uremic Syndrome in Belarus: First 5 Years Experience** S.V. Baiko, A.V. Sukalo, N.I. Tur. *National Center for Pediatric Nephrology and Renal Replacement Therapy, Minsk, Belarus.*

**Objectives:** The objectives of this study were to evaluate the incidence, renal replacement therapy (RRT), mortality of hemolytic uremic syndrome (HUS) in children in Belarus from Jan 2005 to Jan 2010. The medical records of 103 children (48 m, 55 f) at average age of 2,08±2,47 years at the time of admission to hospital were analyzed.

**Methods:** Retrospective, folder review.

**Results:** The incidence of HUS in Belarus was 4,24 cases/100 000 in children < 5 y and 1,37 cases/100 000 when all children < 15 y were taken into account. 100 typical and 3 atypical HUS was diagnosed. 74 children (71,8%) required dialysis (59 PD and 15 HD). The duration of PD was 12,09±5,13d and HD 10,08±4,25d. The mean time of anuria duration was 9,66±4,92 d. Two children (1,94%) died at the acute phase of disease. Using RIFLE criteria for ARF allow us decreased the time from developing oligoanuria to admission to dialysis center from 25,12±5,34h to 14,28±4,64h (p<0,05). The status of 66 children was analyzed at the end of treatment and outpatient's observation. The mean time of ambulatory observation was 3,2±2,3 y. End-stage renal failure was established in 1 child (1,54%), chronic renal disease at grades 2-4 was revealed in 2 (3,08%) patients, 77,5% of children had the arterial hypertension by discharge from the hospital, in 3 cases (4,6%) severe proteinuria was present.

**Conclusions:** The main method of RRT in children with HUS in Belarus is peritoneal dialysis. Using acute peritoneal dialysis and RIFLE criteria for ARF let us decrease considerably mortality in children with HUS for the last 5 years (from 22,1% in 2004 to 1,94 % in 2005-2010, p<0,001).

## Abstract# 724

**Interactive Music Therapy with Children during the Hemodialysis Sessions** L. Barcellos, M. Barcellos, F. Bandeira. *Fundação do Rim, RJ, Brazil; CDR Botafogo, RJ, Brazil.*

**Objectives:** Interactive music therapy (IMT) with children on HD sessions is a pioneering intervention with such patients (P) interacting with music therapists (MT) by means of voice, music instruments and their own bodies**.** Assessing the effects of IMT upon P on HD to favor the expression of inner contents; enhancing non traumatic, pleasant treatment aspects, provoking and activating the unhazardous creative skill by means of the music experiment unpredictability of improvisation and composition as opposed to the predictability of a chronic disease while favoring empowerment toward stress confrontation.

**Methods:** 7 P (3-20 y) undergo IMT 1 h/week in which MT and P interact on music experiences with employed music instruments built by the P according to their abilities with the MT collaboration. The work was designed at first as a group activity, but is also individually performed as well as in pairs depending on the willingness and conditions presented by P. MT creativity and flexibility are fundamental to understand P as the therapy core and to accept different age brackets lead into diverse musical preferences and choices. During music improvisations P, parents, nurses and MT interact.

**Results:** 4 P have individually composed lyrics and tunes and 2 more did the same collaboratively. The 3y P continuously improvises on the contents of a video referring to the existence of miracles. Through tune and lyrics content analysis the importance of expressing feelings turned toward future, strength, adversity facing, and transplant is observed. Music has favored interaction among staff, parents and P.

**Conclusions:** Duly-adapted, flexible music therapy can be employed during HD and can be another supporting aid to dialysis children.

## Abstract# 725

**Register of Children and Adolescents in Dialysis Treatment in Rio de Janeiro** E. Klen, N. Santos, F. Bandeira. *Fundação do Rim, Rio de Janeiro, RJ, Brazil.*

**Objectives:** There are few data on the incidence and prevalence of Chronic Renal Disease (CRD) under dialysis treatment (DT) in children and adolescents in Brazil. One non-profit organization which renders assistance to chronic renal children started a Register to know the epidemiology of CRD in DT, to analyse the profile of this population and the reasons for drop-out.

**Methods:** Since 2005, all the Dialysis Units (DU) of the state, with patients (P) of up to 20 yo receive a Registration Form containing personal identification and DT data. The DU take the responsibility to inform new P in DT as well as the reasons of drop-out. The data are analyzed each year.

**Results:** 42 DU participate in the Register. The number of P under DT in the end of each year was 105 in 2005, 145 in 2007, and 170 in 2009. The entry of new cases was 64 P in 2009. The incidence was of 11 new cases/million pop/age and the prevalence is of 24 cases/million pop/age. Distribution per gender: 61% for the males. In the distribution by DT, HD is more utilized 72% and PD - 28%. Distribution by age: 0-5yo - 6,5%, 5-10yo - 9,4%, 10-15yo - 26% and 58% above 15 yo. The survival of all those under DT, in the end of 36 m is of 80,8% (> 15yo - 83,1%, 10-15 yo - 81,8 % and 67,4% for those under 10 yo). The survival by dialytic modality was 77,6% for HD and 91% for DP in 36 m. Kidney transplantantion constitutes the most frequent reason for drop-out (36%) and death 24%.

**Conclusions:** Epidemiologic information of this register demonstrates the reality in our state and assists all those involved in the treatment of these P. The knowledge of these informations is important to guarantee the efficiency of assistance, to apply the best resources and to provide data for further research.

## Abstract# 726

**Indications, Risks, and Outcome of CRRT in Pediatric Intensive Care Unit over 5 Years** N. Stajic, R. Bogdanovic, J. Putnik, A. Paripovic. *Institute of Mother and Child Healthcare of Serbia, Belgrade, Serbia.*

**Objectives:** During the recent years CRRT became the therapy of choice in ARF and various metabolic disturbances in the ICU. We present our 5-years experience in this field.

**Methods:** 52 children who received CRRT over the last 5 yrs were retrospectively analyzed using: sex, age, body weight, primary diagnosis, indication for CRRT, number of failed organs.Severity scores (PRISM III and PELOD) were calculated at admission, on the day CRRT was started and 24h later. Outcome of CRRT and significance of severity scores were analyzed.

**Results:** 52 pts (29 m, 23 f) were treated (17 neonates,10 infants). Mean age was 48 mos (0-216) and weight 17.9 kg (1.7- 85).Primary diagnoses were: sepsis-16 children (31%), renal disease-11 (21%), and metabolic disturbances or intoxications-11 (21%), whilst 14 (27%) had other diseases. The most common indicationwas sepsis with MOD, with or without fluid overload in 13 (25%) and 10 (19%) pts, respectively. Mean number of failed organs was 3.2 (1-7). Mean PRISM III scores at the beginning of CRRT and after 24h of treatment was 8.8 and 7.7, respectively (p<0,01). Mean PELOD score at the beginning of the CRRT was 15.8, with consecutive mean mortality risk of 33.1%. The overall survival was 35/52 (67%). 21/22 pts with < 3 organs failed survived (95%), contrary to 14/30 (46%) pts with 3 and more organs affected. There is significant difference between PRISM III score at the beginning of the CRRT among survivors and non survivors (p<0,01).

**Conclusions:** Our experience has confirmed the important role of CRRT in treatment of the severe ill patients in PICU. Indications, risks and survival are comparable with the results published so far.

## Abstract# 727

**Dialysate CA-125 Levels after Five Years on Continuous Peritoneal Dialysis: Influence of Treatment Time and Glucose Exposure** C. Candan, P. Turhan, L. Sever, S. Caliskan, M. Civilibal, N. Canpolat, O. Kasapcopur, N. Arisoy. *Pediatric Nephrology, Cerrahpasa Medical Faculty, Istanbul, Turkey.*

**Objectives:** Our study was conducted to evaluate longitudinal changes in dialysate cancer antigen 125 (dCA125) levels over time and to analyze relationship between dCA125 and peritoneal glucose exposure (PGE) in children treated with long-term peritoneal dialysis (PD).

**Methods:** Peritoneal equilibration test (PET) was performed in 11 patients (7 boys) with a mean age of 13±5.1 years and a mean PD duration of 84.0±1.1 months. Peritoneal transport parameters were calculated, and dCA125 levels were measured after 4-hour dwells of PETs. Peritoneal appearance rates (AR) of dCA125, the velocity of the decrease in ARCA125 values and annual PGE levels were calculated.

**Results:** Final investigations were performed in 11 children at a mean of 63.3±3.5 (range 59.0-66.8) months after the initial ones. Both dCA125 and ARCA125 levels had shown statistically significant decreaments during the follow up period (p=0.003) and the velocity of decrease in ARCA125 was found to be 52.6±19.4%. No significant difference was found regarding peritoneal transport parameters. PGE values were significantly higher in the last year as compared to the first year (p=0,014). The velocity of the decrease in ARCA125 levels were not related to total PGE values.

**Conclusions:** We can speculate that after 5 years of PD treatment, mesothelial cell mass reduction is likely to occur. However, mesothelial cells are not likely to play a direct role in peritoneal transport during PD treatment. Furthermore, PGE may not have any influence on peritoneal transport parameters of patients with longer PD duration.

## Abstract# 728

**Low Infection Rates and Prolonged Survival Times of Hemodialysis Catheters in Infants and Children** I. Eisensetin,[1] D. Magen,[1] M. Tarabeih,[1] S. Pollack,[1] I. Kasis,[2] A. Ofer,[3] A. Engel,[3] I. Zelikovic.[1] *[1]Pediat Nephrol Div, Rambam Med Ctr (RMC),Fac Med-Technion, Haifa, Israel; [2]Pediat Infect Dis, RMC, Haifa, Israel; [3]Invas Radiol, RMC, Haifa, Israel.*

**Objectives:** Hemodialysis (HD) in children and infants with ESRD is performed mainly via central venous catheters (CVC). CVC- related infections are the leading cause of morbidity and mortality in this group of patients. We aimed to determine HD- CVC infection rates and survival times in a our Pediatric Dialysis Unit.

**Methods:** We analyzed the data of all children with ESRD who received HD therapy in our Unit between 2001 and 2009. Our strict care of HD-CVC makes no use of prophylactic (topical or systemic) antibiotic therapy.

**Results:** Twenty nine children with ESRD (mean age 8.3 yrs) received HD via a CVC for a total of 22892 days during the study period. Eleven (38%) were infants (<1yr of age) who received HD for cumulative 3779 days (16% of total). Fifty nine CVCs were inserted, of which 13 (22%) were in infants. There were 12 episodes of CVC infections- a ratio of 0.52/1000 CVC days. Four (33%) episodes occurred in infants- a ratio of 1.06/1000 CVC days. Only 3 (5%) CVCs were removed due to infection and additional 28 (47.5%) were removed due to obstruction/clot/tear. The remaining 28 (47.5%) CVCs were either removed while properly functioning (17) or still in place (11). Mean CVC survival time for all children was 388 days and for infants- 291 days.

**Conclusions:** Very low CVC infection rates- one infection per 5 catheter years - and prolonged CVC survival times- more than one year -, are achievable in infants and children with ESRD receiving HD therapy by strictly adhering to practices designed to prevent infections.

## Abstract# 729

**Home Visits in Children on Continuous Cycling Peritoneal Dialysis (CCPD)** E.N. Ellis,[1] C.K. Blaszak,[2] S.R. Wright,[2] A.E. Van Lierop.[2] *[1]Pediatrics, University of Arkansas for Medical Sciences, Little Rock, AR, United States; [2]Nursing, Arkansas Children's Hospital, Little Rock, AR, United States.*

**Objectives:** Home visits by trained personnel in patients undergoing home dialysis are required but no reports evaluating home visits in pediatric CCPD patients have been reported.

**Methods:** We retrospectively reviewed the initial home visit by a dialysis nurse in 22 children on CCPD. The type of community (urban vs. rural, city vs. small town) and type of home was recorded for each visit. Each home was evaluated for clean and working dialysis machine, correct dialysis prescription and supplies, thermometer, smoke detector, fire extinguisher, hand washing area, and prescribed medications.

**Results:** The children were aged from 0.6 to 19 (14±5) years and had been on CCPD for 2 to 60 (21±17) months. The nurse traveled an average of 111±78 miles one way and spent an average of 2 hours at each visit. 4 lived in rural and 18 lived in urban areas (8 in cities, 10 in small towns). 13 lived in single family homes, 3 in trailers, 5 in apartments, and 1 in a college dorm. Equipment was lacking: 27% smoke detectors, 73% fire extinghishers, 41% thermometers, 5% weight scale, 5% broken dialysis machine, and 9%dialysis record sheets. Problems in CCPD room were noted: 14% poor room configuration, 18% pets in the room during CCPD, 9% room dirty, 14% distant CCPD supplies, 5% incorrect CCPD prescription, 14% CCPD machine dirty. Some medications were not present in 36%, were out of date in 41%, and improperly stored in 18%.

**Conclusions:** Home visits are valuable in determining problems in the home care of children on CCPD but further evaluation of the follow-up benefits and costs are required.

## Abstract# 730

**Duodenal Microbiasis in Children with ESRD on Regular HD** A.M. El-Refaey,[1] A. Bakr,[1] G. Attia,[1] R. Yahya,[1] M. Matter,[1] S. Awad.[2] *[1]Pediatric Nephrology, Mansoura University Children's Hospital, Mnaoura, Egypt; [2]Parasitology, Faculty of Medicine, Mansoura, Egypt.*

**Objectives:** Screening uremic patients on HD by single diagnostic test Enterotest® for Helicobacter pylori and duodenal parasites and to elucidate their correlation to nutritional status and CD4 counts.

**Methods:** This study was performed on 23 children with ESRD on HD attending the nephrology unit of Children Hospital, Mansoura University, Egypt. All patients suffered from gastrointestinal symptoms as dyspepsia and diarrhea. 20 patients with ESRD on chronic hemodialysis and without any gastrointestinal symptoms were taken as control group. Duodenal fluid was studied by Enterotest® capsules for Helicobacter pylori and duodenal parasites.

**Results:** Our results show that 14% of patients were infected by H.pylory, 16% by C.parvum, 9% by I.Belli and 23% by G.lamblia. H.pylori has a positive correlation to CD4 (p=0.028) and a negative correlation to BMI, albumin, Hb and the duration of hemodialysis (p=0.025, 0.033, 0.013 and 0.042 respectively). Also, there is a negative correlation of each C.parvum, G.lamblia and I.belli to CD4 (p=0.029, 0.032 and 0,020 respectively), s. albumin (p=0.045, 0.038 and 0.034 respectively), hemoglobin (p=0.035, 0.042 and 0.024 respectively) and BMI (p=0.015, 0.025 and 0.038 respectively); and a positive correlation to the duration on hemodialysis (p=0.017, 0.022 and 0.041 respectively).

**Conclusions:** patients with ESRD submitted to renal transplant should be considered as a group at risk for opportunistic parasitic infection mostly Cryptosporidium infection and that it would be worthwhile to periodically submit these patients to tests for those parasites in order to avoid intrahospital transmission.

## Abstract# 731

**Significance of Acute Kidney Injury on Critically Ill Children Who Receiving Continuous Veno-Venous Hemofiltration** Y. Gao, H.Y. Deng, F. Zhong, Y.J. LI. *Guangzhou Children's Hospital, Guangzhou, China.*

**Objectives:** The study was to study significance of urea and creatinine levels on critically ill children who receving continuous veno-venous hemofiltration (CVVH).

**Methods:** A retrospective review of 33 cases with AKI treated by CVVH was done.
**Results:** 12 cases recovered, 10 cases improved, 11 cases died. The children including 2 cases with poisoning, 2 cases with grave pneumonia, 1 case with diarrhea, 3 cases with septic shock, 1 case with acute myocarditis, 1 case with empyrosis, 1case with extruding syndndrome and 1 case with grave influenza recovered. The children including 2 cases with nephrotic syndrome and adenovirus enteritis, 3 cases with actue poststreptococcal glomerulonephritis, 2 cases with rapidly progressive glomerulonephritis, 1 case with bronchial pneumonia and pneumorrhagia, 1 case with lymphoma and 1 case with poisoning improved. While other children including 2 infants after congenital heart surgery, 2 cases with pyemia,1 case with acute respiratory distress syndrome, 1 nephrotic syndrome with pyocyanic septimia,1 case with hemolytic uremic syndrome, 1 case with grave pneumonia, 1 lupus nephritis, 1 phago-hematocyte syndrome and 1 case with xanthogranuloma died. The PCIS scores were significantly higher in the patients who died than in the survivors. However, urea and creatinine levels did not effect the prognosis.
**Conclusions:** Correctly evaluate the severity of the clinical state of the children at the time of starting therapy, and grasp the indications of CVVH, early intervention with CVVH is beneficial to critically ill patients with AKI.

## Abstract# 732

**Alteplase (tPA) in Pediatric Hemodialysis (HD) Patients – Does It Reduce Catheter Dysfunction, Infections and the Need for Revisions? A Prospective Study** F. Hussain, R. Connell, A. Lunn. *Children's Renal Unit, NUH, Nottingham, United Kingdom.*
**Objectives:** Maintaining patent central venous access is essential for children requiring HD. Catheter dysfunction leads to morbidity from under-dialysis; long term sequelae include compromised future vascular access and inability to transplant. Following an untoward incident our practice of Heparin 1000u/ml line locks was changed to 100u/ml with Taurolock for infected lines with resulting increase in poor blood flow, line changes and use of Urokinase. tPA was introduced following reports from other units. Prospective audits were undertaken comparing all changes in unit practice and outcomes.
**Methods:** Eligible patients receiving HD between 1/12/09-28/2/10 had their central venous lines locked with tPA (1mg/ml). Target & achieved pump speeds and any side-effects or interventions noted.Comparisons were made to the previous audit.
**Results:** 14 patients underwent 454 HD treatments. Median age was 9.5 yrs median weight 30kg. 7 used pediatric lines and 7 adult lines. 99% achieved 80% of their target pump speed for all treatments (previously 82%), 53% achieved 100% (previously 52%). Only 2/14 patients developed line sepsis during the 3 months in comparison to 3/10. 2 patients required line changes in comparison to 3. 5/454 of sessions required urokinase lock/infusion in addition compared to 60/242 previously. There were no side-effects or complications related to the use of tPA.
**Conclusions:** tPA line locks can be used safely in pediatric HD patients. During the study period a reduction in catheter dysfunction was seen. The frequency of line infections and line revisions has declined. We have changed our practice to the routine use of tPA in our HD patients.

## Abstract# 733

**Is There Any Place for a Nephrologist in Treatment of Children's Poisonings?** B. Jachimiak, T. Jarmolinski, D. Runowski. *Department of Pediatrics, Nephrology and Toxicology, District Children's Hospital, Szczecin, Poland.*
**Objectives:** The aim of the study was to establish importance of extracorporeal blood purification (EBP) techniques in treatment and rate of renal complications after common intoxications.
**Methods:** The study group consisted of 555 children (259 male, 296 female; aged 12.4±5.2yr) who were admitted to our department between June 2007 and March 2010 were evaluated.
**Results:** Two main causes of poisoning were: medications (216 cases, 104 suicide attempts among them) and alcohol (206 cases). Less often were: chemical substances (44), carbon oxide (33), narcotics (20) and toxic plants (10). In 26 cases no toxic factor was found. Etiology of intoxication depended on sex and age. Only 7 patients required EBP: 3 girls aged 15-16yr and 3 boys aged 12-17yr. All of them were intoxicated with medicines (6 suicide attempts) and had clinical symptoms of poisoning and very high drug level in blood. Four of them used carbamazepine (blood concentration 33-50µg/ml), 2 – acetylsalicylic acid (ASA; 72-149mg/dl) and 1 – acetaminophen (346 µg/ml). Children poisoned with ASA were treated with hemodialysis and the other with hemoperfusion through a cartridge packed with activated charcoal (Adsorba, Gambro). Improvement was achieved after one session and all patients were cured after short term sequels. Only one patient, a 15-yr-old girl poisoned with ASA, had renal complication. She presented acute interstitial nephritis 3 days after drug ingestion. Symptoms disappeared without treatment in 5 days.
**Conclusions:** The indications to EBP in pediatric poisoning are rare. Renal complications of common intoxications appear incidentally (in less than 1% of cases).
**DISCLOSURE:** *Jachimiak, B.*: Other, Subinvestigator in Clinical Trial - Salary. *Runowski, D.*: Other, Subinvestigator in Clinical Trial - Salary.

## Abstract# 734

**Enema-Induced Hyperphosphatemia in a Newborn – Clinical Experience with Flow-Through Peritoneal Dialysis** V. Koch, D. Kostic, C. Metran. *Pediatric Nephrology, Children's Institute of School of Medicine of University of Sao Paulo, Sao Paulo, Sao Paulo, Brazil.*
**Objectives:** We report the case of a previously healthy 8-day-old newborn, who needed neonatal intensive care treatment after the inadvertent administration of an osmotically active hypertonic phosphate enema that was successfully managed with a modification of the continuous peritoneal dialysis technique. Considering that phosphate removal by peritoneal dialysis strongly depends on total dialysate turnover, continuous flow peritoneal dialysis (CFPD) was indicated.
**Methods:** A closed peritoneal dialysis system was created using two urethral catheters. Initially the peritoneal cavity was filled with 10ml/kg of peritoneal dialysis fluide (PDF) via the inflow catheter while the outflow catheter was kept closed. After completion of the infusion, the outflow catheter was opened and continuous flow of PDF was maintained at a rate of 5ml/min. The dialysate flow rate was adjusted for maximum efficiency by an infusion pump set to maintain flow at 300ml/hour. Serum electrolytes were collected every 3 hours.
**Results:** The dialysis could be discontinued after 6 hours, with a total ultrafiltration of 253 ml, with significant laboratory and clinical improvement and restoration of spontaneous diuresis.
This clinical experience is limited to a few case reports in adult patients in which the procedure was used for 4-8 hours. To our knowledge this is the first description of CFPD in a pediatric patient.
**Conclusions:** The modality of PD herewith described could be an alternative to current management options, as it is highly efficient, methodologically simple and low cost method without need for any sophisticated equipment.

## Abstract# 735

**The Prevalence and Predictors of Anaemia in European Children with Established Renal Failure** L.A. Krischock, K. van Stralen, K. Jager, E. Verrina, J.W. Groothoff, F. Schaefer, J. Tizard. *On behalf of the ESPN/ERA-EDTA Registry Study Group, Academic Medical Center, Amsterdam, Netherlands.*
**Objectives:** Determine the prevalence of anaemia in European children with Established Renal Failure (ERF), identify risk factors for anaemia in this population, and determine the adequacy of treatment of anaemia.
**Methods:** Data were collected from 3580 patients with ERF aged 17 years or under from 2000 to 2009, from 16 European countries. Percentages were weighted according to the number of haemoglobin (Hb) measurements.
**Results:** Comparing the results with European adult and UK NICE guidelines 65% of dialysis and 54% of transplant patients were anaemic. In dialysis patients predictors of anaemia were younger age, female gender, haemodialysis (HD), and a primary diagnosis (PRD) of pyelonephritis or Haemolytic Uraemic Syndrome. Factors significantly associated with being over the target range were older age and peritoneal dialysis (PD). In transplant patients predictors of anaemia were younger age, female gender and a PRD of glomerulonephritis, while being older, male and having a PRD of pyelonephritis were associated with a Hb level over the target. In both dialysis and transplantation groups, patients from Eastern Europe had the highest, and Western and Southern Europe had the lowest Hb levels, with a difference between these regions after adjustment of 1.5 mg/dl.
**Conclusions:** Anaemia is a common and significant problem in European children with ERF. Children on HD are more prone to anaemia than those on PD and those who have been transplanted. Younger children are most at risk. European guidelines are needed to help optimise the management of anaemia in children with ERF.

## Abstract# 736

**Safety, Efficacy and Convenience of Tinzaparin Anticoagulation in Pediatric Hemodialysis Patients** A.L. Lapeyraque, M.J. Clermont, A. Mérouani, V. Phan, J. Gagnon, J. Paquet, L Pelletier. *Service de Néphrologie, Département de Pédiatrie, CHU Sainte Justine, Montréal, QC, Canada.*
**Objectives:** Use of low molecular weight heparin for prevention of extracorporeal circuit (ECC) clotting during hemodialysis (HD) is recommended in adults. Our goal was to explore the safety, efficacy and convenience of a single bolus dose of tinzaparin in our pediatric HD unit.
**Methods:** We performed a prospective study where all stable HD patients were converted from unfractionated heparin (UFH) to tinzaparin. Initial dose of tinzaparin was 40% of the patient's total UFH dose. Patients were monitored for a minimum of 10 weeks after conversion. Hemorrhagic event, visual aspect of the

ECC, nurses satisfaction and anti-Xa activity (U/mL) were recorded.
**Results:** 172 HD sessions were evaluated in 7 patients aged from 2,3 to 17,4 (median: 13,1). 2 patients did not require any adjustment in tinzaparin dose while 5 needed increments of 20 to 62 % from the initial dose. Effective median tinzaparin dose was 73 UI/kg (range: 57-110). Clotting was observed in the air chamber traps in 46/172 sessions but only one was interrupted for massive clotting of ECC. Visual aspect scale of ECC showed significant clotting and coagulated filter in 16 and 2 /172 sessions. No hemorrhagic event was reported. Nurses preferred tinzaparin over UFH for ease of administration and lack of monitoring. AntiXa activity with effective dose of tinzaparin ranged between 0.26 to 0.68 at mid-time, <0.1 to 0.24 at the end of the session. No accumulation of tinzaparin activity was noted between HD sessions.
**Conclusions:** Our experience with tinzaparin was positive: it represents a safe, effective and convenient method of anticoagulation for ECC in a pediatric HD unit.

## Abstract# 737

**A Multicentre Prospective Cohort Study of Paediatric End Stage Renal Disease (ESRD) in Malaysia** M.-L. Lee, Y.-C. Yap, Y.N. Lim. *Hospital Tuanku Jaafar, Seremban, Malaysia; Paediatric Department, HKL, Kuala Lumpur, Malaysia.*
**Objectives:** The primary objectives of this study were to define the incidence and demographic features of paediatric ESRD in Malaysia and to determine the morbidity and mortality of these patients one year after diagnosis.
**Methods:** A prospective cohort study of children < 15 years old presenting with ESRD (eGFR <10ml/min/1.73m2) between January 2007 till December 2008 in Malaysia. One-year outcome in terms of death or renal replacement therapy (RRT) were collected for all patients. Clinical encounters and hospitalization data were collected for those who were started on RRT.
**Results:** 91 children were diagnosed with ESRD giving an incidence rate of 6.6 per million age related population. There were more boys (54%) and higher incidence among the older age groups. The commonest cause of ESRD was glomerulonephritis (29%), renal dysplasia (15%) and neurogenic bladder (11%). 78% of patients were started on renal replacement therapy; 22% of patients was treated conservatively. The commonest cause for conservative treatment (18 children) was because of young age < 5 years old (10 children) and severe co-morbid conditions (5 children). PD was the first treatment modality 86% compared to HD 14%. The commonest reasons for clinical encounters were fluid overload (19% )and hypertension (19% ). At 1 year follow-up; 12% (8 children) on dialysis had died. The main causes of death were fluid overload (3) and lack of dialysis access (2).
**Conclusions:** Paediatric renal replacement therapy in Malaysia is quite established by now but more need to be done for children < 5 years old. Fluid overload was the main cause of morbitiy and mortality among the cohort on renal replacement therapy.

## Abstract# 738

**Encapsulating Peritoneal Sclerosis (EPS) in Patients Treated with Peritoneal Dialysis (PD) in Childhood** M.A. Lewis, T. Smith, R. Lennon. *Paediatric Nephrology, Royal Manchester Children's Hospital, Manchester, United Kingdom.*
**Objectives:** To establish the incidence of EPS in patients treated with PD in childhood and examine the factors associated with EPS development.
**Methods:** Analysis of a cohort of children with data in the Manchester PD registry treated in the 20 years up to 1st January 2010.
**Results:** Over a 20 year period 177 patients <19 years received PD in our unit. 23 are still on PD therapy. The median age of commencement was 9.25 years (interquartile range of 2.3 to 13.2 years). 23% were below the age of 2 years at PD start.
The median duration of PD was 12.6 months in those who had ceased therapy (range 0.07 to 92.7 months) compared to a median of 11.6 months in those still on PD (range 2.8 to 35.5 months). 4 patients developed EPS. 1 was diagnosed at transplantation but was asymptomatic. She had been on PD for 19.9 months. The other 3 were male and were all symptomatic. Their durations of PD were 57.5, 70.5 and 92.7 months. 2 have died from the EPS. 1 has required peritonectomy. All 4 had lost peritoneal membrane function. 2 only recieved non biocompatible fluids, 2 were treated with both. Whilst 3 had a number of episodes of peritonitis the child who required peritonectomy had none.
**Conclusions:** In this cohort 2% of patients were known to have developed EPS but 27% of those treated >48 months developed it. All showed signs of membrane failure, 3 had recurrent peritonitis. PD is the mainstay of paediatric dialysis therapy. Patients who lose allografts may well return to PD. Carers need to know the risk of EPS increases with duration of PD. The length of PD in childhood needs to be recorded and passed onto adult clinics at transfer.

## Abstract# 739

**Complications of Vascular Access in Children and Adolescents in Hemodialysis: A Retrospective Cohort Study** R.A. Souza, E.O. Araujo, J.M.P. Silva, E.M. Lima. *Pediatrics, Federal University of Minas Gerais, Belo Horizonte, Minas Gerais, Brazil.*
**Objectives:** To study the type and incidence of complications of vascular access in children and adolescent who started hemodialysis (HD) treatment over 11 years period.
**Methods:** A retrospective cohort study tha evaluate the type of initial vascular access and the incidence of complications of vascular access in children and adolescents aged 0 to younger than 18 years who started HD in the city of Belo Horizonte, Brazil from 1997 to 2007.
**Results:** A total of 249 vascular accesses in 61 pts (33M, 28 F, median age 13.5 range 2.9-17.8 years), 97 permanent (94 arteriovenous fistula - AVF and 3 grafts) and 152 temporary accesses (uncuffed CVC) were studied. CVC was the first access in 51% of patients in the study. Patient weight did not influence the choice of CVC over AVF: 7 pts under 20 kg started HD using CVC and 7 AVF. However, for pts under 15 kg we detected a ratio of 3 CVC for 1 AVF. The main cause of CVC removal was infection in 35%, followed by the use of AVF in 37.8%. The median survival of the uncuffed CVC was 40 days. Primary failure of AVF was detected in 37.8% fistulas. The median survival time of AVF was 54 months; main cause of AVF failure was thrombosis (84%). No AVF was lost due to infection.
**Conclusions:** Infection was the major cause of uncuffed CVC removal and the risk of infection was 34 times higher in patients using uncuffed CVC vs AVF. Our results suggest that uncuffed CVC must be avoided for ESRD children on HD and replaced by AVF or cuffed CVC.

## Abstract# 740

**Predictive Factors for Survival of Arteriovenous Fistula in Children and Adolescents** R.A. Souza, E.A. Oliveira, J.M.P. Silva, E.M. Lima. *Pediatrics, Federal University of Minas Gerais, BeloHorizonte, Minas Gerais, Brazil.*
**Objectives:** To evaluate the survival of AVF and to correlate with demographic data, primary disease and time elapsed between formation and cannulation.
**Methods:** A retrospective cohort study that evaluated the survival of first AVF in 61 patients (pts) who started HD under 18 years old in the city of Belo Horizonte from 1997 to 2007.
**Results:** At initiation of HD 30 out of 61 (49%) pts studied were using a patent AVF and 31 (51%) were using an uncuffed central venous catheter (CVC).
The failure rate of the first AVF among patients who started HD with AVF or CVC was similar: 37% and 32%, respectively. The risk of AVF loss was 3.8 times higher in pts who started HD using a CVC compared to pts who have a patent AVF since the beginning of HD ($p = 0.025$), whose AVF survival rate at 1, 3 and 5 years was 73%, 46% and 27% respectively; pts who started HD using a CVC had a AVF survival rate at 1, 3 and 5 years of 40%, 20% and 10 % respectively. Pts with uropathy had 3.2 greater risk of AVF loss compared with other pathologies ($p = 0.012$). The maturation time less than 90 days was also predictive of loss of AVF: the risk was 4.4 ($p = 0.011$).
**Conclusions:** Our study identified three independent predictive factors for AVF loss. Pts with uropathy have a higher risk of AVF loss and therefore, placement and use of vascular access in this group of patients should receive special care. Our results also suggest avoid AVF cannulation before 90 days after placement and avoiding the use of CVC as a vascular access for chronic pts.

## Abstract# 741

**Successful Use of Continuous Venovenous Hemodialysis (CVVHD) in Children with Severe Acute Kidney Injury (AKI)** L.S. Milner,[1] A. Kausz,[2] L. Porter,[1] J. Nicoletta,[1] B. Files,[1] L. Courtemarche.[2] *[1]Pediatric Nephrology, Floating Hospital for Children,Tufts University School of Medicine, Boston, MA, United States; [2]Adult Nephrology, Tufts Medical Center,Tufts University School of Medicine, Boston, MA, United States.*
**Objectives:** The outcome of CVVHD in children with severe AKI and multiorgan dysfunction (MOD), receiving continuous renal replacement therapy (CRRT) was evaluated.
Normalization of biochemical abnormalities and fluid balance using convective or diffusive techniques is often required in severe AKI. CVVHD combines both techniques, precluding frequent large fluid volume replacement, and optimizes nursing care.
**Methods:** 17 children ( 18 treatments) were evaluated.The primary cause of AKI was Non Hematologic (NH), (2 atypical hemolytic uremic syndrome,2 hepatorenal, 1 post surgery,6 sepsis,1 drug overdose) or Hematologic (H), (5 bone marrow transplant and 1 hepatoblastoma).
A Baxter 25 or Next Stage dialyzer was used with a double lumen femoral or central line catheter with heparin/protamine or citrate anticoagulation.
**Results:** The mean age was 10.2 yrs and M: F ratio was 1.8:1.Survival for the whole group was 78%, being significantly higher in NH compared to H (82% vs

16%,p<0.01). MOD occurred in 77% of the whole group which was similar in both NH and H (75% and 83% respectively).Survival was significantly improved in children with NH compared to H with and without MOD.

Survival with and without MOD

| | NH (%) | H(%) | p* |
|---|---|---|---|
| Without MOD | 82 | 16 | <0.01 |
| With MOD | 82 | 0 | <0.01 |

* Fisher Exact

**Conclusions:** Combined diffusive and convective CRRT using CVVHD is effective therapy in children with severe AKI and MOD,except in those with a primary hematologic cause.

## Abstract# 742

**Low Density Lipoprotein Apheresis in Lebanese Pediatric Patients with Homozygous Familial Hypercholesterolemia** C.C. Mourani,[1] M. Hneidi.[2] *[1]Department of Pediatrics, Hotel Dieu de France, Beirut, Lebanon; [2]Department of Apheresis, Dahr El Bachek Governmental Hospital, Beirut, Lebanon.*

**Objectives:** The aim our study is to clarify the low density lipoprotein apheresis procedure for Lebanese pediatric patients with homozygous familial hypercholesterolemia (HFH) in terms of efficacy, adverse effects and difficulties.

**Methods:** This study has been realized in a governmental centre. LDL-cholesterol apheresis was performed by direct adsorption of LDL by Dali filtration from whole blood and performed every other week. The follow-up was carried out using an open, prospective uncontrolled clinical design. Data were collected from 18 patients (age 12 +/-6 years). Venous puncture was used for vascular access without any need of AV-fistula operation. Priming the extracorporeal volume was done by normal saline at the beginning of the the first 5 sessions in all patients.

**Results:** The follow-up covered five years, and more than 971 sessions were evaluated. The mean low density lipoprotein cholesterol (LDL-C) pre-treatment value was 680 mg/dL, and the post-treatment value was 270 mg/dL. The most frequently occurring technical problems were related to blood lines: puncture difficulties, insufficient blood flow and coagulation. The main clinical adverse effects were hypotension, chills, nausea and vomiting.

**Conclusions:** LDL apheresis, is a well tolerated, safe, effective and a simple way of treating Homozygous FH pediatric patients. Combined with lipid-lowering drugs, LDL apheresis provides a safe and effective lowering of the mean LDL-C levels in pediatric homozygous FH. This technique is more simple than hemodialysis and pediatric nephrologists should be more involved in realizing this procedure in the future.

## Abstract# 743

**Central Venous Catheters Complications in Pediatric Patients on Acute Hemodialysis** M. Mraz,[1] S. Gasparikova,[2] H. Kurcinova,[1] L. Podracka.[1] *[1]1-st Department of Pediatrics, Safarik University, Kosice, Slovakia (Slovak Republic); [2]Safarik University, Kosice, Slovakia (Slovak Republic).*

**Objectives:** To evaluate the complications associated with central venous catheters (CVCs) in pediatric patients on acute hemodialysis.

**Methods:** 82 CVCs in 49 children (24 boys, 25 girls; aged 11.4 ± 3.5 years) were analyzed.

**Results:** 4 different diameters of catheters were used: 11F (20 CVCs), 8F (53 CVCs), 7 F (5 CVCs) and 6 F (4 CVCs). The most common site of cannulation was right or left subclavian vein (66 CVCs, 80.5%). There were no complications seen in 57 CVCs (69.5%). At least one CVC-related complication was noted in 25 CVCs (30.5%). The lower was the diameter of CVC the higher was the risk of complications (r= -0,50, p<0.01) with 75.0% of all 6 F but only 20.0% of all 11 F CVCs experiencing at least one complication during the treatment. The most commonly seen (60.0%) complications were mechanical. 15 CVCs (18.3%) required recannulation due to kinking (6 CVCs), clotting (4 CVCs), malfunction (4 CVCs) or rupture (1 CVC). 2 CVCs (2.4%) were removed due to MRSE blood stream infection. 8 CVCs (9.8%) insertions were complicated with local bleeding (6 CVCs) or haematoma (2 CVCs). Patients with CVC-related complications were cannulated for a longer time comparing to those experiencing no catheter difficulties (11.5 and 6.8 days, respectively, p<0.01) and required more CVCs per treatment (2.15 and 1.24 CVCs, respectively, p<0.05).

**Conclusions:** The most commonly seen CVC-related complications in children on acute hemodialysis were mechanical with catheter kinking leading the way. From those complications not requiring CVC replacement local bleeding was the most frequently observed.

## Abstract# 744

**EBV and BK Virus PCR Monitoring in Kidney Transplantation as a Guide Line of Immunosuppressive Therapy** Q.H.T. Nguyen,[1] C. Azéma,[2] S. Decramer,[2] F. Bouissou.[2] *[1]Nephrology Service, National Hospital of Pediatrics, Hanoi Medical University, Hanoi, Viet Nam; [2]Nephrology Department, Hopital Des Enfants, Toulouse, France.*

**Objectives:** EBV and BKV PCR monitoring experience in kidney transplantation in Pediatrics Hospital in Toulouse France.

**Methods:** We report 12 children, out of 70 successive kidney transplantation, who experienced prolonged positive blood PCR test for 7 EBV (4 reactivation, 3 primary infection) and 5 BK virus (3 proven BKV nephropathy) undergoing classical triple IS therapy (steroid/AZA or MMF/ CsA or FK). In these patients we prospectively decided to decrease the drug dosage, first anti metabolite and second anticalcineurin, until a negative PCR test was obtained.

**Results:** The lowering of the IS drugs allowed to negative clinical virus symptoms and PCR test in all of the patients without any rejection (follow up from 1 to 5 years) and lymphoma. The drug dosage was continuously adapted to the level of PCR test and today 4 patients remain free of anticalcineurin, and 1 of them had only low dose of steroid, 8 continue triple therapy with half dosage of AZA or MMF and low level of anticalcineurin trough level in sera ( CsA 60 to 70 ng/ml, FK 3 to 5 mg/l).

**Conclusion:** Routinely virus detection is mandatory in tranplantaion. An EBV or BK virus blood PCR positive test is a sign of over IS and allows to decrease the IS regimen with no risk of rejection and to cure the viral disease and avoid EBV B lymphoma that we had previously experienced.

## Abstract# 745

**Peritonitis in Children on Peritoneal Dialysis (PD) in South-Africa: Epidemiology and Risk Factors** R. Raaimakers,[1] P. Gajjar,[2] C. Schroder,[3] P. Nourse.[2] *[1]Pediatrics, Radboud University Nijmegen Medical Centre, Nijmegen, Netherlands; [2]Pediatrics, School of Child and Adolescent Health, University of Cape Town, Cape Town, South Africa; [3]Pediatrics, Gelre Hospital, Apeldoorn, Netherlands.*

**Objectives:** To evaluate the peritonitis epidemiology in children in South Africa and to identify risk factors for peritonitis.

**Methods:** A retrospective cohort study was performed. All children enrolled in the peritoneal dialysis program between 2000 and 2008 were analyzed. Baseline characteristics and potential risk factors were recorded, including: housing, socio-economic circumstances, distance to PD centre, type of PD, mode of catheter placement, weight and height. Outcome indices for peritonitis were peritonitis rate, time to first peritonitis and peritonitis free patients. All peritonitis episodes were analyzed concerning causative organism, severity of peritonitis (fungal or gram negative), catheter policy, occurrence of technique failure and survival.

**Results:** 67 patients on PD for a total of 544 months were included. Total peritonitis episodes was 129. Median peritonitis rate was 0.23/patient month (range 0-1.76). Median time to first infection was 2.03 months (range 0-21.5 months). 28.4% of patients remained peritonitis free. Patients with good housing and good socio-economic circumstances had a significantly lower peritonitis rate and a longer time to first peritonitis episode. No other risk factors were associated with peritonitis outcome.

**Conclusions:** Peritonitis rate is high in this cohort compared tothe developed world. The characteristics of causative organisms are comparable. Most important risk factors for development of peritonitis are poor housing and poor socio-economic circumstances.

## Abstract# 746

**Continuous Partially Ambulatory Peritoneal Dialysis (CPAPD) in Underprivileged Indian Children** A. Ohri, R. Redkar, G. Kulkarni, U. Ali. *Nephrology Division, B J WADIA Hospital for Children, Mumbai, India.*

**Objectives:** To assess the feasibility and efficacy of CAPD in children from families living under socioeconomic constraints.

**Methods:** Retrospective study of children started on CAPD in the last 5 years at our hospital. Data was collected for the clinical profile, details of CAPD, financial status and living conditions of the families, complications and outcome.

**Results:** 13 children mean age 6.9 years (range 2-11) were on CAPD. Straight 2 cuffed Tenckhoff catheter was surgically placed in all. Mean break in period was 8.7 days. Manual CPD with 4 cycles/day and fill volume of 1200ml/m2 was given. Due to financial constraints and non availability of 1 litre bags all used 2 litre bags with 2 cycles from each bag making the CPD ambulatory for 2 cycles only.

Mean duration of CPD was 18.6months.All patients showed improvement in height and biochemistry, reduction in need for transfusions and antihypertensives on CAPD.7 were regularly going to school.

Mother was the caregiver in 12, father in 1.6 patients had an isolated room for dialysis, the rest used a common living room. Mean monthly family income was

Rs 14960 (325$) and expenditure on CAPD was Rs 14769(321$) which was reimbursed by employers in 4. In the rest, cost was borne by help from extended family members.
Peritonitis rate was 1 in 18.6 patient months.
Other complications were catheter block 2,haemorhagic outflow 2,catheter leak in 1,hernia 1.2 patient were transplanted, 2 partially recovered renal function and stopped CAPD, 2 died.7 are on CAPD.
**Conclusions:** CPAPD is a viable option of RRT in underprivileged children with family support and close medical supervision. Complication rates were comparable to those reported from developed nations.

## Abstract# 747

**Use of Tesio Catheters (TC) in Children on Chronic Hemodialysis (HD)** F. Nuzzi,[1] A. Musumeci,[2] G. Malgieri,[1] L. Marzano,[1] B. Minale,[1] A. Ferretti,[1] D. Molino,[1] C. Pecoraro.[1] *[1]Nephrology, Santobono Hospital, Naples, Italy; [2]University Federico II, Naples, Italy.*
**Objectives:** To report the experience with the use of TC in children on chronic HD.
**Methods:** 17 children (11 boys) received TC as vascular access; mean age 3.7 years (0.5-11.9). Primary renal diseases: FSGS (4), Renal Dysplasia (4), Polymarfomative Syndromes (3), Atypical HUS (2), Oxalosis (1), DMS (3). Indications for TC included: low body size (weight: 5 to 19 Kg) (n=10); main neurological complications (n=4); daily hemodialysis (n=1); peripheral venous vessel exhaustion (n=2). TC were inserted into the right internal jugular vein: a unique 10,5 F TC in 4 patients, twin single lumen 6.5 F TC in the remaining 13.
**Results:** 22 interventions in 17 children have been performed: 5 patients needed of removal with replacement of TC because of increase of body height with relative shortness of TC (n=2); thrombosis (n=2); dialysis inadequacy (n=1). Only in 2 cases a mild right pleural effusion developed as complication. The management of TC includes disinfection of the exit site weekly and "in situ" urokinase and heparine and oral warfarin without side effects. No episode of TC and/or tunnel and/or exit site sepsis occurred over 22 months mean follow up (3-79 months). Monthly single pool Kt/V was > 1.5 in all patients. Due to the lack of pain related to venipuncture and the confidence in one's movements, patient and parents compliance was very good.
**Conclusions:** Use of TC in children on chronic HD is very limited. Our experience in infants weighing less than 10 Kg is unique.TC are a reliable, effective in terms of dialysis adequacy and low infection long term rate of vascular access in infants too.

## Abstract# 748

**High Urinary and Dialysate Losses of Vitamin D Binding Protein May Contribute to Vitamin D Deficiency in CKD** A. Prytula,[1] D. Wells,[2] F. Balona,[1] A. Gullet,[1] L. Rees,[1] R. Shroff.[1] *[1]Renal Unit, Great Ormond Street Hospital for Children NHS Trust, London, United Kingdom; [2]Chemical Pathology, Ormond Street Hospital for Children NHS Trust, London, United Kingdom.*
**Objectives:** We hypothesise that low circulating levels of vitamin D metabolites (25-hydroxyvitamin D [25OHD] and 1,25-dihydroxyvitamin D [$1,25(OH)_2D$] are due to a vitamin D binding protein [VDBP] loss in urine and dialysate.
**Methods:** We studied serum, urine and dialysate VDBP levels and compared these with 25OHD and $1,25(OH)_2D$ levels. 16 children were on automated PD, 14 on HD and 10 in pre-dialysis CKD stage 4-5.
**Results:** Serum VDBP and 25OHD levels were significantly lower in children on HD than on PD or pre-dialysis patients (p= 0.03 and P=0.023, respectively). There were no significant differences in $1,25(OH)_2D$ serum levels, but all except 3 children in the HD group received oral 1-alfahydroxycholecalciferol. There was a strong positive correlation between the serum VDBP levels and time on dialysis (p=0.0004) in PD children, but not in HD. In the pre-dialysis group urine VDBP losses strongly correlated with urinary albumin losses. In PD children there was a correlation between serum VDBP levels and total dialysate and urine losses (p=0.03). VDBP losses in the long daytime dwell were higher than in the overnight drain (P=0.039), but did not correlate with the type of dialysis fluid used.
**Conclusions:** 85% of children were 25OHD deficient and 48% $1,25(OH)_2D$ deficient despite alfacalcidol supplementation, possibly as a result of VDBP losses. VDBP levels decrease with time on PD. This suggests that CKD and dialysis patients need careful monitoring of 25OHD and $1,25(OH)_2D$ levels and supplementation as appropriate.

## Abstract# 749

**Effect of Timing of Initiation of CRRT on Short-Term Mortality of Children Admitted to ICU: A Retrospective Nested Case-Control Study** M. Vinai,[1] M. Thompson,[1] D. Gollhoffer,[2] R. Quigley.[1] *[1]Pediatrics, University of Texas Southwestern Medical Center, Dallas, TX, United States; [2]Critical Care, Children's Medical Center, Dallas, TX, United States.*
**Objectives:** Critically ill children who develop renal failure requiring CRRT have an increased risk of mortality. The objective of our study was to evaluate the effect of timing of initiation of CRRT on short-term mortality in critically ill children requiring CRRT.
**Methods:** We reviewed all non-ECMO patients that underwent CRRT in our ICU between Jan 1, 2000 and Dec 31, 2008. Non-survivors were designated as cases and survivors as controls. Demographic data, diagnoses and timing of initiation (Ti, in relation to date of admission to ICU) of CRRT were obtained.
**Results:** A total of 230 non-ECMO CRRT were identified with a median patient age of 114.5 months (Range 0 to 269 months). 214 of these (with Ti $\leq$ 14 days) were included in the analysis. 95 patients did not survive the ICU admission and were designated as cases and the survivors (N=119) as controls. A significant difference in the Ti (mean $\pm$ SD) was noted (non-survivors 3.82$\pm$3.58 days Vs. survivors 2.11$\pm$2.61 days). Difference remained significant in the sub-set with Ti $\leq$ 7 days. No differences were seen in terms of age, PIM II scores at admission and PELOD scores at CRRT initiation. Odds of survival were higher in patients with Ti < 3 days (OR 4.15, 95% CI 2.31, 7.46). A positive correlation was noted between Ti and mortality rate ($r^2$ 0.7, p-value 0.0065).
**Conclusions:** There was a significant difference in the timing of initiation of CRRT between survivors and non-survivors in our cohort. A strong association between initiating CRRT within 3 days of admission to ICU and survival was found.

## Abstract# 750

**Risk Factors for Peritonitis in Ambulatory Peritoneal Dialysis (CAPD/CCPD)** S.Z.P Rigatto, R.C. Gonçalves, A.C.G. Brito, V.M.S. Belangero. *Pediatria, Medical Sciences Faculty/University of Campinas, Campinas, São Paulo, Brazil.*
**Objectives:** To evaluate risk factors for peritonitis in CAPD/CCPD.
**Methods:** A cross-sectional and longitudinal study of 83 chidren in CAPD/CCPD for ten years. Data analyzed are in table below.
**Results:** 83 children, 39 had peritonitis (44,8%), peritonitis rate of 0.48 per patient-year. Mean time free of peritonitis was 18.19±11.07 months. The table below shows the results in groups with and without peritonitis.

| Group | Peritonitis | No Peritonitis | |
|---|---|---|---|
| | Mean | Mean | P value |
| Age (y) | 9.64 | 9.52 | 0.794 |
| Sex n (m/f) | 24/15 | 22/22 | 0.294 |
| Previous Hospital IPD time (m) | 4.09±5.72 (1.8) | 1.99±3.83 (0.98) | 0.022* |
| Previous Peritonitis | 0.31± 0.46 | 0.11 ± 0.32 | 0.025 |
| APD time (m) | 35.39±21.12 (32.06) | 19.69±16.77 (18.0) | 0.001* |
| Observation time (m) | 13.96 | 19.69 | 0.206 |
| Center distance (km) | 130.78 | 69.48 | 0.293 |
| Parental Education n (<8/≥8 anos) | 26/13 | 18/26 | 0.031* |
| C cr ml/min/1.73m2 | 1.38 ± 1.76 (1.01) | 3.44± 3.89 (2.20) | 0.045* |
| Hemoglobin (g/dl) | 9.74 | 10.03 | 0.706 |
| Albumin (g/l) | 3.34 ± 0.69 (3.5) | 3.87 ± 0.46 (3.87) | 0.001* |
| CRP (mg/dl) | 0,675 | 0.524 | 0.514 |
| Kt/V urea | 2.46 ± 0.51 (2.35) | 4.20 ± 2.55 (3.37) | 0.033* |
| BMI | 16.92 | 18.05 | 0.633 |
| Urine vol (ml/kg/d) | 25.66 | 44.82 | 0.150 |

**Conclusions:** Time of previous intermittent peritoneal dialysis, previous peritonitis and parental education were risk factors for peritonitis and can represent socioeconomic difficulties. Albumin, residual renal function and Kt/v also were risk factors for peritonitis and agree with the literature.

## Abstract# 751

**Hemodialysis in a 31 Days of Age Patient with Prune Belly Syndrome: Case Report** C.A. Rogow, M.J. Barcia, M.L.D.M. do Val, S.L. Bergamo, Z.M. Andrade, J.T.A. Carvalhaes. *Pediatric Nephrology, Federal University of Sao Paulo, Sao Paulo, Brazil.*
**Objectives:** Prune Belly (PB) is a rare syndrome with urinary tract abnormalities, abdominal muscle deficiency and cryptorchidism, with high mortality in neonates. Chronic renal disease, including end stage (ESRD) occurs in 20-30%. In these patients, peritoneal dialysis (PD) may not be appropriate, because of technical issues like poor catheter anchorage, deficient exit healing and leakage.
**Methods:** We describe an indication of hemodialysis (HD) for a patient of 31 days of age, weighing 3.3 kg, with PB syndrome and renal abnormalities, progressing to ESRD. At newborn period, a septic shock lead to reduction in urinary output and deterioration of his exams (biochemistry at 30 days of age: creatinine 5.1 mg/dL; urea 179 mg/dL; P 9.8 mg/dL).
**Results:** Hemodialysis in children still remains a challenge, with few data in the literature. In infants < 2 years old or less than 10 kg, we find difficulties related to

patient's size, vascular access and hemodynamic instability. Nevertheless, it is an effective renal replacement therapy in small children, as seen in our patient, who kept hemodynamic stability using vasopressor drugs, with great improvement in post HD exams (creatinine 3.41mg/dL; urea 124 mg/dL; P 5.6 mg/dL).
**Conclusions:** The survival of PB patients depends mainly on renal impairment: as seen in our patient, dialysis was needed very early in life. As reported by previously data, one-year survival in patients younger than 1 year at the start of treatment is 83-89% for both PD and HD; based on that, the modality choice of treatment was limited by anatomical reasons, considering its risks and benefits for the patient.

## Abstract# 752

**Fifteen Years Experience with Pediatric Peritoneal Dialysis in R. Macedonia** E. Sahpazova, D. Kuzmanovska. *Nephrology, University Childrens Hospital, Skopje, Macedonia, The Former Yugoslav Republic of.*
**Objectives:** To describe our 15 years experience with pediatric patients on PD.
**Methods:** Retrospective study of 25 children (10 girls and 15 boys, mean age 10.10±4.32 years) who underwent PD treatment from January 1996 to Mart 2010.
**Results:** The cause of ESRD was uropathy in 11 children (44%), chronic glomerular disease in 6 children (24%) and others in 8 children (32%). The mean duration of PD was 32.24±25.83 months. 16 children received CAPD and nine received automated PD. The most common complication was peritonitis (60%) in 18 patients with 67 episodes, followed by exit site infection (23.21%). The over all rate of peritonitis was one episode in 12 patients/months. Staphilococcus aureus was the most prevalent pathogens and accounted for 41% of the peritonitis, and 40% of the ESI. The other complications (16.96%) were hernias in 4 patients, cardiovascular problem in 5 patients, omental capture of the catheter in 5 patients, dialysate leak in 3 patients, ileus and intraperitoneal pseudocyst formation in each 1 patient.
Technique survival after 1st, 2nd and 5 year was 94%,88% and 55% respectively. The cumulative survival of all children after 1stand 5 year was 90% and 60% respectively.
During follow-up period 4 patients (16%) died because cardiovascular problems, 7 (28%) received renal transplant from living donor. Persisting peritonitis and loss of ultrafiltration in 6 patients (24%) was the main cause for transferred to HD and seven children (28%) are still on PD therapy.
**Conclusions:** Despite the occurrence of peritonitis and death, PD is a good option of renal replacement therapy. Prevention and appropriate therapy of infection complications could prolong patients and technique survival.

## Abstract# 753

**Infancy and Extrarenal Comorbidity as Risk Factors for Mortality on Pediatric Peritoneal Dialysis** T. Sakai, Y. Hamasaki, K. Ishikura, H. Hataya, M. Honda. *Pediatric Nephrology, Tokyo Metropolitan Children's Medical Center, Fuchu-shi, Tokyo, Japan.*
**Objectives:** The long-term survival rate and baseline clinical characteristics of patients with end-stage renal failure who started peritoneal dialysis (PD) were analyzed to determine potential risk factors associated with PD in childhood.
**Methods:** A total of 135 patients under 16 years of age who started PD as the first dialysis modality in our hospital from 1990 to 2008 were grouped by age at the start of PD, and the 5-year survival rate was compared among age groups (0–1 year [n = 41], 2–5 years [n = 31], 6–12 years [n =44], 13–16 years [n = 19]). In addition, the 5-year survival rate in patients with life-limiting extrarenal comorbidity(LEC) at baseline was compared with that of patients without LEC. Patients lost to follow-up were censored.
**Results:** The median age at the start of PD was 4.80 years (range, 3 days to 15.2 years). Eight patients on PD (infection [n=3], cardiovascular disease [n=1], others [n=2], unknown [n=2]), 1 on hemodialysis switched from PD (unknown), and 3 after transplantation (infection [n=2], unknown [n=1]) died. The overall 5-year survival rate was 91.8%. The 5-year survival rate was 76.9%, 100%, 95.1%, and 100% in the 0–1 year, 2–5 year, 6–12 year, and 13–16 year groups, respectively, and 62.5% and 95.6% in patients with and without LEC at the start of PD (n=19 and 116), respectively. The number of patients who started PD at the age of 0–1 year has been increasing since 2000.
**Conclusions:** Survival is adversely affected by younger age and LEC at the start of PD. Improvements are needed to meet the increasing demand for care by patients who start PD at a young age and/or have LEC.

## Abstract# 754

**Reduced Efficacy with Molecular Adsorbent Recirculating System (MARS) Dialysis as Compared with Combined Plasma Exchange and Hemodialysis (PP/HD) in Children with Acute Liver Failure (ALF)** B. Schaefer, F. Schaefer, K.H. Heckert, G. Engelmann, C.P. Schmitt. *Center for Pediatric and Adolescent Medicine, Heidelberg, Germany.*
**Objectives:** MARS is an extracorporeal liver support system eliminating albumin-bound and water-soluble substances. It is increasingly applied in children with ALF, although a RC comparison with PP/HD is missing, pediatric data are scant.
**Methods:** We performed a retrospective analysis of 19 MARS and 29 PP/HD treatments in 7 children with ALF (0.1-18yrs). Adult MARS was used in 4 (24-66kg), MARS Mini set in 3 children (3-13kg). Mean session duration was 6.7 (4.5-10.5) h. Blood was sampled within 3 h before and after treatment.
**Results:** Treatment with adult MARS tended to decrease total serum bilirubin (19.2±5.4 to 14.4±3.1mg/dl per session); ammonia (72±35 to 78±34µmol/l) and INR remained unchanged (1.5±0.4 and 2.3±1.5, all p=ns). Mini-MARS did not reduce bilirubin or INR (19.7±3 to 20.5±3.2mg/dl and 2.5±0.7 to 2.9±1.1) and slightly reduced ammonia (70±24 and 56±9µmol/l, all p=ns). In contrast, PP/HD reduced bilirubin (22.9±6 to 13.5±4mg/dl), ammonia (88±33 to 64±25µmol/l) and INR (2.8±0.6 to 1.4±0.1, all p<0.05). Intraindividual comparison in 5 children yielded a reduction in bilirubin of 3.0±14 and 41±10% with MARS and PP/HD (p<0.001), a decrease in ammonia of 10±20 and 28±17% (p=ns) and an increase and decrease in INR of 40.2±46 and 47±12%, respectively (p=0.03). The treatments were well tolerated. 2 patients recovered, 1 with Wilson disease was successfully transplanted. 4 children died, including 3 children treated with Mini-MARS.
**Conclusions:** Our preliminary experience suggests superior efficacy of PP/HD compared to intermittent MARS therapy for treating ALF.

## Abstract# 755

**Peritoneal Dialysis-Associated Complications in Dutch and Belgian Children. Report from the RICH-Q Study** N.J. Schoenmaker,[1] J.H. van der Lee,[2] J.W. Groothoff,[1] on behalf of the RICH-Q working group. *[1]Paediatric Nephrology, EKZ/AMC, Amsterdam, Netherlands; [2]Clinical Epidemiology, AMC, Amsterdam, Netherlands.*
**Objectives:** The incidence rate of peritonitis in children treated with peritoneal dialysis (PD) has been reported as 1.04, 0.83 and 0.82 /patient- year at risk in studies from Germany (n=49), Finland (n=23) and USA (n=30), respectively. The incidence rate of exit site infection has been reported as 0.29 and 0.37 /patient- year at risk in studies from Japan (n=47) and Turkey (n=93), respectively. Here we describe the incidence of PD-associated complications in the Netherlands and Belgium.
**Methods:** The RICH-Q study is a collaborative project in which all Dutch and Belgian centres that provide paediatric chronic Renal Replacement Therapy (cRRT) collaborate to improve the quality of care. All Dutch and Belgian dialysis patients aged <19 years between October 2007 and 2010 were included (n= 159). Hospitalization and PD-associated complications were assessed. Data were assembled according to GCP guidelines including site monitoring.
**Results:** 40 of the 75 children (53%) who were treated with PD were admitted to the hospital at least once during the past year. The reasons were: PD catheter problems (24%), start of the therapy (21%) or peritonitis (17%), the other reasons were for other infection or investigation. The incidence rates were 0.62 and 0.70 / patient-year at risk for peritonitis and exit site infection, respectively.
**Conclusions:** Compared to other countries the incidence of peritonitis in the Netherlands and Belgium is low, of exit site infection high. This discrepancy might be due to different definitions of exit site infection.

## Abstract# 756

**Are We Late for the Diagnosis of Acute Kidney Injury in the Intensive Care Units?** O.D. Kara, N. Dincel, B. Sozeri, S. Mir. *Pediatric Nephology, Ege University Faculty of Medicine, Izmir, Turkey.*
**Objectives:** The p RIFLE criteria (acronym for risk, injury, failure, loss and end stage renal disease) has been used for the definition of AKI. The purposes of this study were, to emphasize the importance of p RIFLE in early diagnose and prognosis of AKI and to evaluate the practicability of the p RIFLE criteria in intensive care units (ICU) apart from pediatric clinics.
**Methods:** We evaluated 66 patients consulted from different from ICUs and applied acute peritoneal dialysis, between 1993 and 2008. Patients are classified at survivors and non survivors. Retrospectively, we evaluated all patients by p RIFLE criteria at consulted by pediatric nephrologists on intensive care units.
**Results:** Among these 66 (37 male, 29 female) patients, mean age was 16.4 ±31,6 months.There were 35 (53%) patients, suffered with cardiac causes while 31 (47%) patients with non cardiac causes.Mortality rate was found 63.6%. In survival group there were 10 patients in Risk, 7 in Injury and 7 in Failure while in non survival group,8in Risk, 6 reached Injury, and 28 reached Failure before consultation. The non- survivors had a longer interval between the onset of AKI and the day the PD was begun (4.3±-0.4 vs. 1.2±1.2 days, p=0.001), and longer duration of PD (13.0±3.5 vs. 6.6±2.7 days, p=0.036) than survivors.

**Conclusions:** When the patients were consulted with the pediatric nephrologists, most of them were in failure class. We suggest that if they were detected AKI earlier, the prognosis would be better. Therefore early diagnosis of AKI with p RIFLE criteria and early initiation of PD, will improve prognosis.

## Abstract# 757

**Continuous Renal Replacement Therapy (CRRT) in the Neonatal Intensive Care Unit (NICU): 5-Years Experience** N. Stajic, R. Bogdanovic, J. Putnik, A. Paripovic, B. Jankovic, M. Djordjevic, J. Nikitovic-Martic. *Institute of Mother and Child Healthcare of Serbia, Belgrade, Serbia.*

**Objectives:** Due to numerous risks, neonatal CRRT is the most challenging aspect of this procedure. We present our 5-years experience with CRRT in neonates.
**Methods:** A retrospective study was performed in 17 neonates treated with CRRT over the last 5 yaars, including their sex, age, body weight, primary diagnosis, indications for the CRRT, number of failed organs and severity scores.
**Results:** 11 male and 6 female neonates were treated, with mean age of 7.9 days (0-28) and weight 3,1kg (1.8-4.6). Mean PRISM III scores were 6.5, 7.2 and 5.5 on admission, at the beginning of CRRT, and after 24h of treatment, respectively. The difference of later two was significant ($p<0.05$). The mean number of failed organs was 2.5 (1-6). Primary diagnosis was congenital metabolic disturbances in 5 (hyperammoniemia 3, MSUD 2), isolated renal failure in 4, MOD with renal failure in 8.Ten pts received CVVHDF, 6 CVVHD, and 1 CVVH. The mean duration of CRRT was 5.7 days, mean circuit life span 2.6 days and ultrafiltrate rate 32.9 ml/kg/h. Overall survival rate was 70.5%; 100% in patient with 1-2 failed organs and 44.4% in patients with MOD (3 and more failed organs). Six patients had fluid overload (>10% BW), 4 of them having ARF without MOD survived, and both patients with fluid overload and MOD died.
**Conclusions:** CRRT is a safe and efficient procedure in the treatment of congenital metabolic disturbances and isolated renal failure in neonates. This procedure was less successful in the treatment of MOD with ARF. Our results are among the best of those published so far.

## Abstract# 758

**Ultrasound Evaluation of Peritoneal Thickness in Children and Young Patients on Peritoneal Dialysis (PD)** S. Testa, I. Borzani, F. Paglialonga, U. Matta, A. Castelli, M. Pavesi, G. Ardissino, A. Edefonti. *Pediatric Nephrology and Radiology, Fondazione IRCCS Osp. Maggiore Policlinico, Milano, Italy.*

**Objectives:** Sclerosing peritonitis is a rare but life-threatening complication of PD, characterized by peritoneal membrane (PM) proliferation, thickening and calcification. The role of ultrasound (US) evaluation in this condition is not well established: we propose US screening to assess PM thickening in pts on PD.
**Methods:** Between April and September 2009 we prospectively performed US peritoneal evaluation in 21 pts (16M) on PD, median age 6.4yrs (0.6-27); median PD duration was 27.6mo. (0.1-108). We studied the PM with high frequency probe at 3 different ventral windows, at a distance of at least 10cm from the peritoneal catheter insertion.
**Results:** 12 pts (57%) showed a PM thickness ≤1mm (reference normal upper limit), 5 pts (24%) between 1 and 1.5mm and 4 pts (19%) >1.5mm. 2 out of 4 pts in the last group also showed small bowel loops fixed and tethered posteriorly as well as symptoms of subocclusion. The regression lines between PM thickness and PD duration and the number of peritonitis, showed a statistically significant correlation (r:0,85, $p<0,0001$ and r:0,56, $p<0,001$, respectively).
**Conclusions:** CT is considered the gold standard in the diagnosis of sclerosing peritonitis, however it encompasses high radiation dose and is not suitable for repeated examinations. Our data suggest that US examination represents a safe tool to monitor PM changes during PD, allowing good peritoneal visualisation and dynamic evidence of small bowel distribution that can correlate with clinical symptoms even when they are still mild. We suggest to address all PD patients to US peritoneal screening once every 6 mo.

## Abstract# 759

**Tunneled CVC for Children on Chronic Hemodialysis** F. Paglialonga, G. Rossetti, A. Giannini, S. Testa, A. Edefonti. *Pediatric Nephrology and Dialysis Unit and Intensive Care Unit, Fondazione IRCCS Ca' Granda Osp Maggiore Policlinico, Milano, Italy.*

**Objectives:** Tunneled central venous catheters (CVC) are increasingly utilized as vascular access for chronic HD in children, but very limited data exist on the best CVC option to be chosen.
**Methods:** In 2006 and 2007, 11 Quinton Perm-cath CVC were surgically positioned in the internal jugular veins in 5 pts (age:2.7 [2.0-9.6] yrs). In 2008 and 2009, 10 Med-Comp Ash-split CVC were placed in the subclavian veins by percutaneous cannulation in 7 pts (age:11.7 [2.1-13.3] yrs). Both CVC types were tunnelled through subcutaneous tissue, the tip was positioned at the atrial-cava junction or just inside the right atrium. Indications for CVC placement were cardiac congenital malformations, low body weight (BW) or inadequacy of venous outflow, which contraindicated AV fistula creation.
**Results:** Survival of Perm-cath and Ash-split CVC at 100 days was 18% and 40% respectively. The access was lost because of bacteremia (1 case in each CVC group), malfunction (5 and 2) and dislodgement (4 and 2). Complications of CVC insertion were bleeding from the jugular vein which required surgical correction in 1 pts after Perm-cath placement and hemothorax due to subclavian artery puncture during Ash-split placement in 1 pts. Mean dialytic URR in pts with the Ash-split CVC and BW <20 kg was 77.1±8.5%, higher than that of pts with Ash-split CVC and BW >20 kg (53.1±7.0%, $p<0.005$) and not different from that achieved with Perm-cath (79.2±7.3%).
**Conclusions:** In conclusion, owing to the lack of venotomy (thus the possibility of preserving the access site), the better duration and the good dialytic performance, Ash-split CVC can be a good access option for children undergoing chronic HD.

## Abstract# 760

**The Effectiveness or the Effectiveless of CYP3A4 Polymorphism on Calsineurin Inhibitor Drugs Trough Levels in Pediatric Kidney Transplantations** E. Toroslu, B. Sozeri, S. Mir, A. Berdeli. *Ege Univercity Faculty of Medicine, Izmir, Turkey; Pediatric, Ege Univercity Faculty of Medicine, Izmir, Turkey; Pediatric Nephrolgy, Ege Univercity Faculty of Medicine, Izmir, Turkey.*

**Objectives:** Calsineurin inhibitor drugs (tacrolimus(tac) and CyclosporineA(CycA)) are the substrates of cytochromeP450 subgroup3A(CYP3A) metabolic enzymes. CYP3A4*1B 290A→G and CYP3A4 878T→C are the most common polymorphism at 10.exon of CYP3A4 genes.(7q21).We studied whether had an association with genotypes and clinic impacts.
**Methods:** 42 pediatric renal transplants were enrolled.We obtained 290AG genotype via PCR RLFP and 878TC genotype via direct DNA-sequencing by full 10.exon CYP3A4.Drug trough levels, serum urea and creatinin levels were collected at first 3,6,9,12 month(mo). Accepted trough levels were for CsA C0 250-300 ng/ml, C2 1100-1300 ng/m, Tac first 2 mo 10-15 ng/ml, further mo 8-10 ng/ml.
**Results:** The mean age 12±15years.(21=n CycA, 16=n Tac)We determinated 290AA,878TT genotype and also Ser201Ser, Val296Met, Met318Ile, Leu331Val, Glu333Lys, Ala337Thr genotypes in whole patients at exon 10 CYP3A4 gene. The mean trough level of CsA –C0 was lower than accepted levels all visits.(133± 67ng/dl,112±78.2,136±129 ng/dl,and129±65ng/dl,respectively,$p=0.00$).Also, the mean CsA-C2 was lower. The mean tac trough level was higher in all visits. (8.6±4.7ng/ml,19.9 ±39.4 ng/ml, 10.3±4.0 ng/ml,6.7±3,6 ng/ml,respectively). In tac group, the serum urea levels were higher at 3 and 6 mo than CycA group ($p=0.01$, $p=0.02$). no differences at creatinine($p>0.05$).
**Conclusions:** Wild-genotype caused lower CycA levels and higher tacrolimus levels than normal range. The drug regimens should be individualized by genetic evaluations.

## Abstract# 761

**Quality of Life in Dutch and Belgian Children with End Stage Renal Disease – Report from the RICH-Q Study** M. Tromp,[1] J.H. van der Lee,[2] J.W. Groothoff.[1] *[1]Pediatric Nephrology, Emma Children's Hospital, AMC, Amsterdam, Netherlands; [2]Pediatric Clinical Epidemiology, Emma Children's Hospital, AMC, Amsterdam, Netherlands.*

**Objectives:** The RICH-Q study (Renal Insufficiency therapy in CHildren – Quality assessment and improvement) is a collaborative project in which all centers in the Netherlands and Belgium that provide pediatric chronic Renal Replacement Therapy (cRRT) collaborate to improve the quality of care of pediatric cRRT. Here we report on Quality of Life.
**Methods:** All prevalent and incident Dutch and Belgian children on chronic dialysis and all incident transplanted patients aged <19 years between October 2007 and October 2011 were included. Quality of Life was investigated with the PedsQL.
**Results:** 159 patients were included; 75 PD, 57 HD and 27 pre-emptive transplantations, median age [range] 11 [0-18] years, median RRT duration (range) 10 [0-209] months. Quality of Life data were received from 91 children (response rate 57%). Mean [SD] total QoL score was 63 [16], which is low compared to pediatric ESRD patients (74 [15]; n=85) and healthy controls (87 [8]; n=131) in Texas.[1] Age, gender, country, treatment modality, duration of cRRT, general condition and parents' level of education, which might influence QoL, did not differ between responders and non-responders.
**Conclusions:** In children with ESRD in the Netherlands and Belgium Quality of Life is low compared to a similar pediatric ESRD population and compared to healthy controls in Texas. This may be due to cultural differences.

Reference
[1] Goldstein, SL et al. Health-related Quality of Life in pediatric patients with ESRD. Pediatric Nephrology 2006; 21: 846-850.

## Abstract# 762

**Current Management Policies for Children with End Stage Renal Disease in the Netherlands and Belgium – Report from the RICH-Q Study** M. Tromp, J.W. Groothoff. *Pediatric Nephrology, Emma Children's Hospital AMC, Amsterdam, Netherlands.*

**Objectives:** The RICH-Q study (Renal Insufficiency therapy in CHildren – Quality assessment and improvement) is a collaborative project in which all centers in the Netherlands and Belgium that provide pediatric chronic Renal Replacement Therapy (cRRT) collaborate to improve the quality of care of pediatric cRRT. At the start of the project a survey was performed to investigate the variation in management policies between the centers.

**Methods:** Twice a year representatives from all 9 centres convene to discuss patient data from all centres. Data on treatment and outcomes are registered centrally and monitored according to GCP standards. A questionnaire about ESRD patient management policies was sent to all centers at baseline. We compared reported policies with actual management in 2008.

**Results:** Questionnaires were filled in by 8 of the 9 centers. Indications to start RRT included clinical condition in all centers; additionally, some centers mentioned low GFR, uremia or hyperphosphatemia. The preferred treatment modality in all centers was renal transplantation, below the age of 3 years peritoneal dialysis. Contrary to the official local policy, in children older than 3 years with no residual diuresis hemodialysis was applied most frequently. Policies concerning PD care and transplantation policies regarding the offer of pre-emptive transplantation, the acceptance of non-HLA-identical donors, non-heart-beating donors and non-related living donors varied considerably, even within one country.

**Conclusions:** This multicenter study indicates that there is a large variation in treatment policies, showing the need for clear -preferably evidence based- guidelines.

## Abstract# 763

**Blood Purification for Pediatric Diseases: Usefulness of Partial Plasma Exchange and Continuous Hemodialysis** S. Tsuchida, H. Tamura, A. Noguchi, I. Takahashi, T. Takahashi. *Pediatrics, Akita University School of Medicine, Akita, Japan.*

**Objectives:** Due to recent advances, blood purification can now be performed safely in children including neonates. Depending on the target substances to be eliminated, Plasma-Dia-Filtration as partial plasma exchange and continuous hemodialysisbe performed for intractable pediatric disease and severe organ failure. We report cases and investigates desirable blood purification methods and equipment for children.

**Methods:** Four caces treated with Evacure EC-2A filter(Kuraray Medical Inc.) and Sublood-B(Fuso Pharmaceutical Co.)under CVVH. The patients were infused with albumin solution and fresh frozen plasma (FFP).

1. Influenza-related multiple organ failure; No improvement was seen with CHDF and Polymyxin B Column(PMX).
2. Fulminant hepatitis B; The recovery from hepatic failure was poor despite repeated plasma exchange (PE).
3. Macrophage activation syndrome secondary to systemic juvenile idiopathic arthritis; Because of poor response to steroid pulse therapy. PDF was performed as a PE procedure for excessive cytokine adjustment.
4. Severe pancreatitis accompanying intractable ulcerative colitis: CHDF using a PMMA membrane could not be continued because of DIC, and PDF was performed while continuing PE.

**Results:** All patients are alive, and their underlying conditions have improved. PDF was effective for removing substances with medium molecular weights, such as cytokines including IL-6 and TNF-a. The amount of FFP required for PE was one-fifth the normal.

**Conclusions:** In blood purification therapy in children, it is necessary to carefully select purification membranes and usage methods that match the treatment of the specific disease.

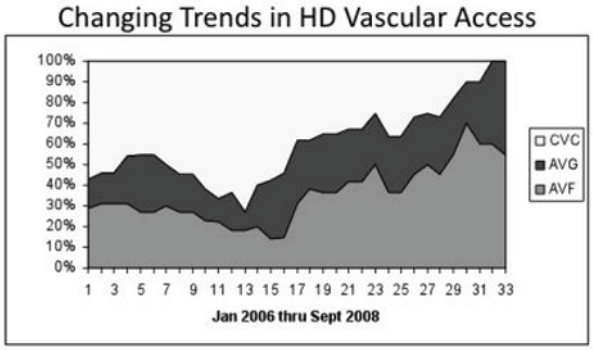
## Abstract# 764

**Preoperative and Perioperative Strategies To Increase Arteriovenous Fistula (AVF) Rates in Children** R.P. Valentini, M. West. *Children's Hospital of Michigan, Detroit, MI, United States.*

**Objectives:** Despite increased emphasis on AVF placement in adults on hemodialysis (HD), AVF rates in children remain low. The study purpose was to assess success of a comprehensive program (CP) on AVF creation in children.

**Methods:** Retrospective, single center review of access rates in chronic HD patients (pts) pre and post-intervention. Intervention began March 2006. AVF candidates underwent preoperative venography or ultrasound venous mapping, aspirin 81mg started, and vascular access surgeon referral. Surgery performed on non-HD day and BP medications reduced. Intraop BP monitoring by nephrologist; pts given crystalloid, 5% albumin and dopamine 3-5 mcg/kg/min to keep BP at 90th%tile for age. Postop: hospitalized overnight for BP monitoring. Dry weight and BP target increased. AVF maturation tracked as outpatient. Pts < 20 kg or AVF failures had arteriovenous graft (AVG) placement. Central venous catheters (CVC), if present, removed after 3 successful HD sessions with AVF/AVG.

**Results:** At outset, 14 pts on HD: 8 (57%) had CVC, 4 (29%) had AVF, and 2 (14%) had AVG. At 33 mo F/U, 12 pts on HD: 7 (58%) had AVF, 5 (42%) had AVG, and none had CVC.




Pts with AVF had mean age 14.5 yrs, weight 53.3 kg, and matured in 6.7 months. Pts with AVG were mean 7.3 yrs, 21.1 kg, and matured in 28.6 days.

**Conclusions:** CP led to increased AVF placement rates and decreased CVC prevalence in a pediatric HD program.

## Abstract# 765

**Determinants of eGFR at Start of Renal Replacement Therapy in Children** K.J. van Stralen,[1] J.E. Tizard,[2] K.J. Jager,[1] F. Schaefer,[3] E. Verrina.[4] *[1]Academic Medical Center, Amsterdam, Netherlands; [2]Royal Hospital for Children, Bristol, United Kingdom; [3]University Childrens Hospital, Heidelberg, Germany; [4]Gaslini Childrens Hospital, Genoa, Italy.*

**Objectives:** Few studies investigated determinants of the glomerular filtration rate (GFR) in paediatric patients starting on dialysis or with a transplant.

**Methods:** Data were collected as part of the ESPN/ERA-EDTA registry from fourteen European countries and referred to incident paediatric patients starting on RRT between 2002 and 2007 under the age of 18 year. eGFR was calculated using the Schwartz formula. Data were adjusted for age, gender, treatment modality at start, primary cause of renal failure (PRD) and regions in Europe ($eGFR_{adj}$).

**Results:** Median eGFR in the 938 patients starting renal replacement therapy (RRT) was 10.4 ml/min/1.73m$^2$ (5$^{th}$ & 95$^{th}$ percentile: 4.0-26.9). Twenty-six patients (2.8%), mainly infants with Finnish type nephropathy, started with eGFR levels over 50 ml/min/1.73m$^2$. Younger age, female gender, starting on dialysis and having a short time between the first visit to a paediatric nephrologist (PN) and start of RRT, were associated with lower eGFR at start of RRT. Gender differences were only present during adolescent age, and disappeared when using the same *K* value for both genders. The various PRDs showed large differences in the rate of decline in eGFR between the first visit to a PN and start of RRT, however this did not result in differences in $eGFR_{adj}$ at start of RRT.

**Conclusions:** The main determinants of eGFR at start of RRT were age, gender, treatment modality at start, and the time between the first visit to a PN and start of RRT. Research is needed to determine the consequences of these differences.

## Abstract# 766

**Proteome Profile of Peritoneal Effluents in Children on Chronic Peritoneal Dialysis** M. Bruschi,[1,2] G. Candiano,[2] L. Santucci,[1,2] K. Perri,[2] G.M. Ghiggeri,[2] E. Verrina.[2] *[1]Renal Child's Foundation, Genoa, Genoa, Italy; [2]Nephrology and Dialysis, IRCCS G.Gaslini, Genoa, Genoa, Italy.*

**Objectives:** We compared proteome profile of peritoneal effluent (PE) obtained with icodextrin (Ico) or glucose (Glu) peritoneal dialysis solution in paediatric patients and defined protein oxido-redox status (ORS).

**Methods:** Two 14-hour daytime dwells were performed on 2 subsequent days with 7.5% Ico and 3.86% Glu solutions in 16 patients. Protein composition was analysed by 2D-PAGE and mass spectrometry; oxidised products were evaluated from the amount of free SH labelled with new cyanines.

**Results:** Peritoneal transport kinetics of β2-microglobulin and cystatin C was linear for both solutions, but slightly higher with Ico than Glu suggesting a better efficiency for low-molecular weight molecules. There was a linear correlation between total protein removal during Ico and Glu dialysis in the same patient, suggesting that removal is a function of peritoneal membrane characteristics. The ratio between proteins removed by Ico and by Glu solutions was higher at low removal rate. Image gel analysis revealed a complex protein composition of PE (1064 and 774 spots respectively in Ico and Glu solutions, 524 of which were common to both conditions); 314 spots were higher in Ico compared to Glu PE. Analysis of protein ORS showed a greater amount of oxidised albumin in Ico PE that was correlated with lower serum levels.

**Conclusions:** Our results indicate better efficiency of Ico in removing small proteins. Removal of big proteins and of their oxidized isoforms reflects potentially opposite effects of the treatment that may lead to loss of substances with functional role and debris deriving from their conversion.

## Abstract# 767

**Morphological Study of Peritoneum in Children on Peritoneal Dialysis (PD)** E. Vidal, E. Benetti, M. Della Vella, L. Murer. *Pediatric Nephrology and Transplantation Unit, University of Padua, Padua, Italy.*

**Objectives:** The aim of this study is to examine the relationship between clinical, dialytic parameters and peritoneum histological characteristics in PD patients.

**Methods:** We retrospectively evaluated 17 children in CPD. Biopsies were performed after 6 mths from CPD initiation, during catheter removal due to malfunction or after treatment drop-out.

Parietal peritoneal biopsies were taken surgically. All samples were examined by light microscopy and sections stained by H&E and Masson's trichrome methods. Children were divided into 2 groups according to the peritoneal biopsy presence (Group P) or absence (Group A) of inflammatory infiltrate, vasculopathy and submesothelial sclerosis (>100 um).

**Results:** Median age at the start of CPD was 6.2 years (range 0-17) and median CPD duration was 18.7 mths (6-55). Peritonitis incidence was 1:17 episode:CPD-mths. Mean glucose concentration was 1.36% in 10 patients, while in the others mixed glucose concentrations were used. We found that Group P of children had significant greater:

- median duration of CPD: 22 (5-56) vs 5.5 mths (4-15); $p<0.05$;
- median number of peritonitis episodes: 2 (0-4) vs 0 episodes (0-1); $p<0.05$;
- peritonitis incidence: 1:13 vs 1:46 episode:CPD-mths; $p<0.05$.

Median age at the start of CPD was 12.9 years (0.8-17.8) in Group A children, significantly greater than that of Group P (4.2 years; 0-10.9). Moreover, the use of hyperosmolar glucose solutions was larger in Group P patients (45% vs 33%; n.s.).

**Conclusions:** Our study suggests that PD length and glucose load are mainly involved in mesothelial impairment. Peritoneal infectious episodes play a major role in determining chronic alterations.

## Abstract# 768

**Continuous Renal Replacement Therapy (CRRT) in Preterm Infants with Renal Failure** F. Yang. *Department of Pediatrics, The First Affiliated Hospital of Jinan University, Guangzhou, Guang Dong Province, China.*

**Objectives:** CRRT was as new treatment in our hospital for preterm infants admitted to the NICU in 2006. Whether CRRT in preterm infants effetive and safe?

**Methods:** CVVH were used in tow preterm infants with renal failure.5 F double lumaen venous catheter have been inserted into the umbilical cord vein.The machine (Prisma/JH2000) was placed in the CVVH mode.The circuit should be primed with blood.Heparin was used for anticoagulation ( loading dose of 25 units /kg, followed by a maintenance dose of 10 uints/kg). Blood flow rate were 5-10l/kg/min, replacement fluid rate was 30% of BFR.

**Results:** case 1, male 34 weeks gestation,body weight 1.6 kg. 3 days after birth because of shock, RDS and acute renal failure, artificial ventilation and life support management had been used, but the baby was still anuria.and creatinine increased to 426 umol/L. So began to CVVH. After 13.5 hours CVVH the patient's condition improves significantly.Oxygen was given only by a headbox, urine output increased to 160ml/24h., creatinine was reduced to152umol/L. During the time of CVVH, there was a little bleeding from the umbilical. at the end of CVVH the bleeding stopped. One month later the baby was dischaged from the hospital. Case 2.,male, 35 weeks gestation. body weight 2.5kg. Suffer from haemolytic uraemic syndrome, serum creatinine increased to 221umol/L. 3 days after birth began to CVVH. 10 days after CVVH, ( total 54 hours ). the patient 's urine output still less.After CVVH the serum creatinine of the patient can be reduced significantly,but creatinine increased again the next day. So the patient change to peritoneal dialysis.

**Conclusions:** Application of CRRT in critically ill preterm infants is effective and safe.

## Abstract# 769

**The Value of Ultrasonographic Measurement of Peritoneal Membrane Thickness in Children on Chronic Peritoneal Dialysis Treatment** O. Yavascan,[1] A. Bal,[1] M. Anil,[1] A.B. Anil,[1] F. Kamit,[1] N. Aksu,[1] S. Mir.[2] *[1]Pediatric Nephrolgy, Izmir Tepecik Teaching and Research Hospital, Izmir, Turkey; [2]Pediatric Nephrolgy, Ege University Faculty of Medicine, Izmir, Turkey.*

**Objectives:** Loss of peritoneal function due to peritoneal fibrosis is a major factor leading to treatment failure in chronic peritoneal dialysis(CPD) patients. The aim of the present study is to uncover the relationship between functional parameters of peritoneum and peritoneal thickness(PT) measured by US in children on CPD.

**Methods:** We recruited two groups of patients: 23(13 females,10 males) on CPD (patient group) and 26 (7 females, 19 males) on predialysis follow-up(CrCl: 20-60 ml/dak/1.73 $m^2$,control group). Age, sex, weight, height, body mass index(BMI), CPD duration, episodes of peritonitis and peritoneal equilibration test results were recorded. Hemoglobine(Hb), blood pressure(BP), left ventricular mass index(LVMI) and renal osteodystrophy(ROD) parameters were also obtained. Statistical analyses were performed by using student-t and Pearson correlation tests.

**Results:** Mean PT in CPD patients(1028.26±157.26µm) was significantly higher than control patients(786.52±132.33). Mean PT was significantly correlated with mean body height*($R^2=0.93,P<0.05$),* BMI *($R^2=0.25,P<0.05$),* CPD duration*($R^2=0.64,P<0.05$),* episodes of peritonitis*($R^2=0.93,P<0.05$),* $D/P_{creatinine}$*($R^2=0.76,P<0.05$),* $D4/D0_{glucose}$*($R^2=0.81,P<0.05$).* No correlation was found between peritoneal thickness and Hb, BP, LVMI and ROD parameters.

**Conclusions:** Ultrasonographic measurement of peritoneal membrane thickness is a simple and noninvasive method in CPD children. This diagnostic tool likely enables to assess peritoneal structure and function in these patients.

## Abstract# 770

**Encapsulating Peritoneal Sclerosis in Young Children Treated with Peritoneal Dialysis** N. Yildiz,[1] P. Turhan,[1] C. Candan,[1] B. Güclüer,[2] Mü. Erguven.[3] *[1]Pediatric Nephrology, Goztepe Research Hospital, Istanbul, Turkey; [2]Pathology, Goztepe Research Hospital, Istanbul, Turkey; [3]Pathology, Goztepe Research Hospital, Istanbul, Turkey.*

**Objectives:** Encapsulating peritoneal sclerosis (EPS) is the most serious and classical complication of peritoneal dialysis (PD). Prolonged duration of chronic PD is still the single most significant risk factor. However, many anecdotal cases exist of encapsulating peritoneal sclerosis developing within months of commencing dialysis. The clinical syndrome of EPS is characterized by intestinal adhesions that disturb mechanical intestinal motility.

**Results:** We present 2.25 years old boy with end-stage renal disease due to diffuse mezengial sclerosis, who was admitted to hospital with complaints of fever, nausea, vomiting, abdominal pain, cloudy dialysate, weight loss and nonfunctional dialysis catheter. He had been on automated peritoneal dialysis (APD) for 3 months. On the fifth day of the admission, he was operated because of the signs of intestinal obstruction. Peritoneal dialysis catheter was found adhered to peritoneal membrane and part of small intestines, and it was removed. Then, he was transferred to haemodialysis. After the surgery, the patient's clinical symptoms improved. A peritoneal biopsy revealed severe sclerosing peritonitis.

**Conclusions:** Because of its high mortality rate, patients on peritoneal dialysis (PD), it should be kept in mind that EPS may occur even in small aged children regardless of the duration of PD with impaired peritoneal ultrafiltration. These children should be followed closely for clinical symptoms of EPS.

## Abstract# 771

**RRT for Children from a East European Country 20 Years Following Political Transformation** I. Zagozdzon,[1] A. Zurowska,[1] C. Prokurat,[2] D. Drozd,[3] M. Szczepanska. *[1]Pediatric Nephrology & Hypertension, MUG, Gdansk, Poland; [2]Pediatric Nephrology & Transplantation, CZD, Warsaw, Poland; [3]Pediatric Nephrology, UJ, Cracow, Poland.*

**Objectives:** To assess the state of renal replacement therapy for children in Poland, where unrestricted treatment became available following political transformation in 1989.

**Methods:** A National RRT Registry set up in 2000 accumulated individual patient data of all children receiving dialysis or Tx at 13 pediatric dialysis centers. Data of 779 children aged 0-18 years was analyzed. Incidence rates, causes of CKD and choice of initial therapy were calculated for the different 5 year age groups.

**Results:** The standardized annual incidence of CKD5 was 6.4 per million age related population (pmarp) and was highest for the 0-4 age group (7.1 pmarp). CAKUT and genetic diseases accounted for 58% of all causes of CKD5. CAKUT was the predominant diagnostic category in all 5 year age groups (1.6-3.4 pmarp), followed by genetic diseases (2.0 and 1.2 pmarp) in 0-4 and 5-9 year olds and glomerular diseases (1.4 and 1.8 pmarp) in 10-14 and 15-18 year age groups. The preferred initial mode of therapy was PD (62%), followed by HD (32%) and PET (6%). HD was the most frequent choice (49%) only in the 14-18 age groups.

**Conclusions:** 1. There is unrestricted access to RRT for all pediatric age groups in Poland with a similar incidence rate to that of West European countries. 2. There is a free choice of both PD and HD modes of treatment with a disproportionately low choice of PET. 3. Slowing progression is the major procedure for decreasing the incidence of CKD5 as nearly 60% of its causes in east european children are congenital and/or genetic.

## Abstract# 772

**Serum Hepcidin Is Lower in Pediatric Hemodialysis *Versus* Peritoneal Dialysis Patients** J. Zaritsky, H.-j. Wang, T. Ganz, I. Salusky. *David Geffen School of Medicine at UCLA, Los Angeles, CA, United States.*

**Objectives:** Hepcidin, a key regulator of iron homeostasis, may be the molecular link between inflammation and anemia in chronic kidney disease. Given that hepcidin may be significantly cleared by hemodialysis (HD) we sought to compare serum hepcidin levels between pediatric peritoneal (PD) and HD patients (pts).

**Methods:** Serum hepcidin levels were measured in 26 PD and 30 HD pts by competitive ELISA and compared to indicators of anemia, inflammation and iron status. In addition, hepcidin clearance was determined by measuring serum hepcidin before, during and after HD in 8 pts.

**Results:** In HD pts, hepcidin levels were higher than in normals but were lower than in PD pts despite similar rhEPO dose and higher markers of inflammation, iron stores, and anemia.

| Biochemical Variables | | | |
|---|---|---|---|
| | PD (mean±SD) | HD (mean±SD) | p |
| Age (yrs) | 15.4±4.4 | 16±4.6 | NS |
| Hepcidin* (ng/ml) | 942±962 | 347±308 | <0.01 |
| Hgb (g/dL) | 13.3±1.7 | 12.1±1.7 | <0.01 |
| rhEPO (units/kg/week) | 283±235 | 302±233 | NS |
| hs-CRP (mg/L) | 2±3.8 | 10.2±21.4 | <0.002 |
| Ferritin (ng/mL) | 321±303 | 578±572 | <0.02 |
| % Iron Saturation | 37.7±23 | 32.2±12.1 | NS |
| Urea (mg/dL) | 46.7±11.4 | 53.3±21.7 | NS |
| Phosphorus (mg/dL) | 5.6±1.2 | 5.4±1.6 | NS |
| iPTH (pg/mL) | 824±721 | 529±678 | <0.02 |

*Normals=44±51 ng/ml

In HD pts, multivariate regression revealed that hepcidin was independently predicted by ferritin (β=5.48, p<0.01) and high sensitivity C-reactive protein (hs-CRP) (β=1.76, p<0.03). Similiarly in PD pts hepcidin was independently predicted by ferritin (β=0.82, p<0.01) and % iron saturation (β=0.68, p<0.01). Hepcidin levels decreased after HD from 532±297 to 292±171 ng/ml (p<0.01) with an average clearance of 141±40 ml/min.

**Conclusions:** The unexpected finding of lower serum hepcidin in HD *versus* PD pts despite similar associations on multivariate analysis suggests that hepcidin may be more efficiently removed via HD.

**DISCLOSURE:** *Ganz, T.*: Other, Co-founder and officer of Intrinsic Lifesciences, LLC. *Salusky, I.*: Consultant, Genzyme, Amgen and Cytochroma.

## Abstract# 773

**Nutrition Evaluation and Health Education for Children with Chronic Peritoneal Dialysis** Q. Zhou, J.-H. Tian, J. Ge, W. Feng. *Nursing Departemnt, Children's Hospital of Fudan University, Shanghai, China.*

**Objectives:** PD(peritoneal dialysis) is the most effective method for ESRD. The infection is dwindling because of the improvements of dialysis apparatus and operative technique. However, the nutrition problems of PD become more and more prominent. We conducted this study to evaluate children's nutrition status before and after PD with the aim to guide the treatment.

**Methods:** Included were 10 children receiving PD. For evaluating their nutrition status, the levels of blood BUN, Scr, Alb, prealbumin (PA), transferrin (TF), hemoglobin (Hb) and hematocrit (Hct) were all detected before and after PD. Also recorded were their heights and weights before and after PD. We elucidate the nutrition requirement of PD children to their parents, making them clear with the calculation of composition of food. Children's diets were formulated by discussing with their parents, and were urged to follow as often as possible.

**Results:** The average time receiving PD treatment was(12.5±9.28)months, ranging from 4 to 36months. Average level of BUN decreased significantly from (31.4±7.4)mmol/L before PD to (19.6±3.4)moml/L after PD(p<0.01). Hb and HCT levels increased respectively from (77.1±19.9)g/L, (22±6)% to (98.4±40.2) g/ L, (34±4)%. The average blood pressure was 14.6/9.6kPa after PD, comparing with 18.9/11.7kPa before PD (P<0.01). The average blood ALB after PD was (41.0±3.2)g/L. The average increase of heights was (0.67±0.59)cm/month.

**Conclusions:** Chronic PD can improve children's nurtrition status and growth. In the process of chronic PD, the nutrition guidance and health education play a key role.

Hypertension and Cardiovascular Disease

## Abstract# 774

**(O-97)**

**Hypertension in Pediatric Renal Replacement Therapy Patients in Europe** A.M. Kramer,[1] K. van Stralen,[1] K. Jager,[1] F. Schaefer,[2] E. Verrina,[3] J. Groothoff.[1] *[1]Academic Medical Centre, Amsterdam, Netherlands; [2]University Hospital, Heidelberg, Germany; [3]Gaslini Children's Hospital, Genoa, Italy.*

**Objectives:** Determine the prevalence of hypertension (HT) and the distribution of blood pressure (BP) in the European pediatric renal replacement therapy (RRT) population, for both renal allograft recipients and dialysis patients, and to identify potential determinants associated with HT.

**Methods:** Data on a cohort of 1987 children aged younger than 18 years and registered with RRT between 1999 and 2009 in 14 European countries were derived from the ESPN/ERA-EDTA registry. HT was defined as either systolic or diastolic BP ≥95$^{th}$ percentile for age, height and gender, according to the NHBPEP Fourth Report on the Diagnosis, Evaluation, and Treatment of High Blood Pressure in Children and Adolescents. Information on the use of antihypertensive medication was available for 54.2% of the patients.

**Results:** HT was present in 39.3% of hemodialysis, 28.1% of peritoneal dialysis and 21.1% of transplant patients. Of those on antihypertensive medication, 41.6% had uncontrolled HT (BP ≥90$^{th}$ percentile). Among those not receiving medication 17.9% had HT. In multivariate multilevel analysis, HT was associated with younger age, treatment with hemodialysis, and glomerulonephritis. No differences were found between genders. The prevalence of HT was 23.7% in Northern Europe, 28.9% in Eastern Europe and 34.5% in Southern Europe. Differences remained significant after adjustment.

**Conclusions:** In this large European cohort fewer patients suffered from HT compared to US studies. Despite the awareness of the possible consequences, HT is still inadequately treated in the pediatric European RRT population.

## Abstract# 775

**(O-98)**

**The CYP3A5 SNP and Risk of Hypertension after Kidney Transplantation in a Cohort of Young Adults** L. Ghio, M. Belingheri, M. Ferraresso, S. Turolo, S. Tirelli, E. Groppali, R. Villa, C. Civitillo, A. Edefonti. *Found. IRCCS Ca'Granda, Milano, Milano, Italy.*

**Objectives:** Calcineurin Inhibitors (CNIs) have a direct vasoconstrictive effect on afferent glomerular arterioles and may cause hypertension (Ht). CYP3A5 gene is involved in metabolism and transportation of CNIs. We analyzed the influence of CYP3A5 genetic polymorphisms on blood pressure (BP) values in the first 6 mos after kidney Tx.

**Methods:** 92 young kidney Tx recipients (median age of 15±7.3 yrs) with primary Tx, treated with CNIs were studied.

BP was assessed 3 times a day and the average of the measurements was recorded at 6th day and 6th mo post-Tx. HP was classified by WHO criteria for pts above 18 yrs, whereas for the others dedicated algorithms were used (Menghetti;J. Hypertens,1999). To account the effect of anti-hypertensive medications (AHM) BP values were normalized using Cui's method (Hypertens,2003): adjusted systolic and diastolic BP (AdjSBP; AdjDBP).

Patients were divided in groups according to their CYP3A5 SNP: CYP3A5 G/G (homozygous; non-expressing variant) present in 72 pts, CYP3A5 A/G and CYP3A5 A/A (heterozygous; expressing variant) in 20.

**Results:** AdjSBP and AdjDBP were significantly more elevated in pts carrying the A allele: at 6th day AdjSBP 161±23mmHg in heterozygous vs 140±23mmHg in homozygous (p<.001); AdjDBP 102±15mmHg vs 87±14mmHg (p<.001); at 6th mo AdjSBP 136±16mmHg vs 129±15mmHg; AdjDBP 89±15mmHg vs 80±12mmHg (p<.001). Besides heterozygous took more AHM than homozygous in both periods 2±1 vs 1.5±1.1 (p<.001).

**Conclusions:** Heterozygous for A allele is a risk factor for the development of HP in post-transplant recipients in CNIs therapy.

## Abstract# 776
**(O-99)**

**Correlates of Carotid Artery Intima Media Thickness (cIMT) in Children with Chronic Kidney Disease (CKD): A Report from the Chronic Kidney Disease in Children (CKiD) Cohort Study** T. Brady,[1] M. Schneider,[1] J. Flynn,[2] C. Cox,[1] T. Kimball,[3] S. Furth,[4] B. Warady,[5] M. Mitsnefes.[3] *[1]Johns Hopkins University, Baltimore, MD, United States; [2]Seattle Children's Hospital, Seattle, WA, United States; [3]Cincinnati Children's Hospital Medical Center, Cincinnati, OH, United States; [4]Children's Hospital of Philadelphia, Philadelphia, PA, United States; [5]Children's Mercy Hospital, Kansas City, MO, United States.*

**Objectives:** Determine factors associated with cIMT in the CKiD cohort study.
**Methods:** 92 children with carotid artery ultrasound (u/s) and complete data on exposures of interest 1 yr after enrollment in CKiD were studied. Standardized protocol imaging was done locally by B-mode u/s and read centrally. Multivariate linear regression analysis was conducted to determine associations with cIMT.
**Results:** The median age was 12.6 y; 71% male; 79% Caucasian; median GFR 43.4 ml/min/1.73m$^2$ and 17% had glomerular CKD. Median (IQR) cIMT was 0.44 (0.39, 0.49) mm; 43% had cIMT >95$^{th}$ %ile (0.453 mm). Multivariate analysis revealed that each 0.1 unit increase in systolic blood pressure index (SBPi = SBP/95$^{th}$ %ile BP) was associated with a 0.02 mm (95% CI: 0 to 0.04; p=0.09) increase in cIMT and the cIMT in children taking antihypertensive medications was 0.03 mm thicker (95% CI: -0.01 to 0.06; p=0.09) than those not receiving antihypertensive agents. CKD etiology, GFR, total cholesterol, Ca x P product, age, sex, and gender were not associated with cIMT.
**Conclusions:** Children with stage 2-4 CKD have impaired carotid artery structure. CIMT is associated with increasing SBPi and use of BP medications. Therefore, chronic exposure to elevated SBP may produce structural changes in the carotid artery among children with CKD.
**DISCLOSURE:** *Flynn, J.*: Consultant, Pfizer, Novartis, Gilead Sciences and the Lewin Group.

## Abstract# 777
**(O-100)**

**Skin Autofluorescence (AF) as a New Tool To Estimate a Potential Risk of Atherosclerosis Development in Children and Adolescents with Chronic Kidney Disease (CKD)** I. Makulska, D. Zwolinska, M. Szczepanska, D. Drozdz. *Department of Pediatric Nephrology, Wroclaw Medical University, Wroclaw, Poland; Department of Pediatric Nephrology, Wroclaw Medical University, Wroclaw, Poland; Dialysis Unit, Silesian Medical University, Zabrze, Poland; Dialysis Unit, University Children Hospital, Kraków, Poland.*

**Objectives:** Advanced glycation endproducts (AGEs) are thought to contribute to inflammatory processes and atherosclerosis in CKD population. We determine if skin AF, the measure of tissue AGEs, is increased and related to vascular changes.
**Methods:** 36 patients with CKD (stage 2-4) and 20 hemodialysed subjects were included in the study. We evaluated: AF using AGE-Reader, carotide intima-media thickness (IMT), aortic pulse wave velocity (PWV) using analyzer (SphygmoCor, Australia). Serum ADMA, MMP-9 and sE-selectin were measured by ELISA method. Study groups were compared to 26 healthy age-matched subjects.
**Results:** AF, IMT, PWV, serum ADMA, sE-selectin, MMP-9, SBP and DBP were significantly higher in all patients vs. controls. All parameters in HDs were significantly higher than in CKD group. Multiple regression analysis showed a significantly positive correlation between AF and IMT (p<0,004), PWV (p<0,03), ADMA (p<0,0001), sE-selectin (p<0,0001) and MMP-9 (p<0,008) in the group of all subjects.
**Conclusions:** Elevated AF in CKD indicate for the accumulation of tissue AGEs. A significant association between AF and investigated markers supports a role for AGEs as a contributor to vascular damage and shows that AF may be a good, easily applicable tool for assessing the risk of atherosclerosis development and cardiovascular events in CKD children.

## Abstract# 778
**(O-101)**

**Blood Pressure Control in Treated Hypertensive Children Using Ambulatory Blood Pressure Monitoring** T. Seeman, L. Dostalek, J. Dusek, J. Janda. *Dept. of Pediatrics, University Hospital Motol, Prague, Czech Republic.*

**Objectives:** Control of hypertension (HT) is important for prevention of cardiovascular diseases. The aim of our study was to investigate the control of HT in treated children using ambulatory blood pressure (BP) monitoring (ABPM).
**Methods:** We retrospectively reviewed all ABPM studies in our center. Controlled HT was defined as systolic and diastolic BP index at daytime and nighttime <1.0.
**Results:** A total of 195 ABPM studies fulfilled the inclusion criteria. The mean age was 13.6±4 years. 132 children had renoparenchymal HT (RPH), 10 children renovascular (RVH), 10 endocrine, 4 cardiovascular, 29 primary (PH) and 5 children other forms of HT. The mean number of drugs per patient was 1.6 ± 0.8. 109 children were on monotherapy. 150 children were on ACE-inhibitors. 53% of all children had controlled HT. There was no difference in the prevalence of controlled HT between primary and secondary HT (52% and 53%). Children with RPH had significantly better control of HT than children with RVH (58% vs. 20% p=0.02). In children with uncontrolled HT, 48% had isolated systolic HT and 43% isolated nighttime HT. The use of ACEI monotherapy was significantly more effective in controlling HT than the use of calcium channel blockers (CCB, p=0.018). In children with RPH, the use of ACEI monotherapy was associated with lower BP indexes than the use of monotherapy of CCB (p<0.0001). In children with PH there was no difference of maximal BP index between children receiving different drugs in monotherapy.
**Conclusions:** This is the first pediatric study on BP control in hypertensive children using ABPM. It indicates that the control of HT in treated children is inadequate in about 50% of them.

## Abstract# 779
**(O-102)**

**Role of Caveolin-1 and Heat Shock Protein 70 (Hsp70) Interaction in the Regulation of Nox4 Expression in Microdissected Proximal Tubules from Spontaneously Hypertensive Rats (SHR), as an Effect of Losartan** P. Vallés,[1,2] V. Bocanegra,[2] M. Lopez,[2] W. Manucha,[1,2] A. Gil Lorenzo.[2] *[1]Facultad de Medicina.Universidad Nacional de Cuyo, Mendoza, Argentina; [2]IMBECU, CONICET, Mendoza, Argentina.*

**Objectives:** We examined the AT1 receptor antagonist Losartan effect on Caveolin-1 and Hsp70 protein association in SHR proximal tubules (PT). Hsp70 involvement on Losartan oxidative stress regulation through the MAPK signaling pathway was also studied
**Methods:** Five-week-old SHR and controls Wistar-Kyoto rats(WKY)were randomized for receiving Losartan (40mg.kg.day) (SHRLos) or no treatment (SHRH2O) during 6 weeks. Interaction between proteins was determined by coinmunoprecipitation and by inmunocytochemical colocalization (confocal microscopy).
**Results:** The relative abundance of Cav-1 was 2-fold higher in microdissected PTmembrane fractions from treated SHR vs WKYH2O. Hsp70 membrane translocation was demonstrated in SHRLos through out the upregulation of Hsp70 expression in PT membrane fractions when compared with WKYH2O. Interaction between Cav-1 and Hsp70 was determined in SHRLos PT membranes. After membrane translocation of Hsp70, the decreased NADPH oxidase activity near controls demonstrated on SHRLos PT membranes, was reversed by the pre-incubation with anti-Hsp70 antibody. Interaction between Hsp70 and NADPHoxidase subunit Nox4 showed that PT membrane overexpression of Hsp70 was associated with decreased Nox4, involving ERK 1/2 activation in SHRLos.
**Conclusions:** Interaction of Cav-1 and Hsp70 was demonstrated in SHRLos proximal tubules.Translocation of Hsp70 to PT membranes in SHRLos might exert a cytoprotective effect by downregulation of Nox4 through the pERK1/2 pathway.

## Abstract# 780
**(O-103)**

**Arterial Stiffness in Healthy Children and Young Adults – Normal Values in a Large Cohort** D. Kracht,[1] A. Doyon,[3] C. Jacobi,[1] F. Schaefer,[3] B. Schmidt,[2] S. Sorrentino,[2] E. Wühl,[3] A. Melk.[1] *[1]Pediatric Nephrology, Medical School, Hannover, Germany; [2]Nephrology, Medical School, Hannover, Germany; [3]Pediatric Nephrology, University Hospital, Heidelberg, Germany.*

**Objectives:** Aortic pulse wave velocity (aPWV), an indicator of arterial stiffness predicts cardiovascular mortality risk in adults. Arterial stiffening advances with age and is accelerated in specific disease conditions. In childhood aPWV has not been investigated in larger cohorts. The aim of this study is to provide normal values and to prove the suspected increase with age.
**Methods:** Pulse waves were captured simultaneously by oscillometry on the right carotid and femoral artery (Vicorder) in 405 healthy school children aged 6 to 18 years. We also measured intima-media thickness (IMT) and elasticity on both carotid arteries by B- and M-Mode ultrasound.
**Results:** aPWV increased with age. 6-8 year olds (n=97): 4.2±0.4 m/s; 9-11 year olds (n=135): 4.5±0.4 m/s; 12-14 year olds (n=97): 4.9±0.5 m/s; 15-18 year olds (n=76): 5.2±0.5 m/s (p<.0001). We found no gender-related differences. aPWV significantly correlated with age (r=.63, p<.0001). Further significant correlations (normalized to age and height): mean systolic and diastolic blood pressure. We found no correlation of aPWV with IMT, but significant correlations with elasticity markers: incremental elastic modulus (r=.43, p<.0001), distensibility coefficient (r=-.42, p<.0001).

**Conclusions:** This study defines aPWV normal values in children and young adults using a new non-invasive oscillometric method. Even in healthy young individuals correlations to cardiovascular risk factors can be seen. Interestingly, a connection of aPWV to functional parameters of arterial elasticity was observed.

## Abstract# 781
**(O-104)**

**Cerebral Blood Flow Reactivity to Hypercapnia in Children with Hypertension** J.C. Kupferman,[1] M. Sharma,[1] Y. Brosgol,[1] K. Paterno,[1] S. Goodman,[1] M. Pagala,[1] I. Prohovnik,[2] S. Pavlakis.[1] *[1]Maimonides Infants and Children's Hospital, Brooklyn, NY, United States; [2]Mount Sinai School of Medicine, New York, NY, United States.*

**Objectives:** To determine the response of cerebral blood flow (CBF) velocity to hypercapnia in children with hypertension (HT).

**Methods:** We evaluated middle cerebral artery blood flow velocity in 5 groups of patients using transcranial doppler (TCD): 1: controls; 2: white-coat (Wc) HT, 3: preHT, 4: untreated HT, 5: treated HT. TCD was performed before and after re-breathing from a bag to increase carbon dioxide ($CO_2$) levels. Outcome variables were mean and peak blood velocity, end tidal $CO_2$ and mean reactivity. Reactivity slopes were generated using linear regression. ANOVA and unpaired t tests were used to compare among groups. We considered a $p<0.05$ as significant.

**Results:** There were no differences in baseline TCD velocities, baseline end tidal $CO_2$ and $CO_2$ change among groups. In regard to reactivity slopes, there were no significant differences between genders or age. All HT patients were treated with enalapril.

Table 1. Cerebral Blood Flow Reactivity Slopes

| | n | Age (mean±SD) | Slope (mean±SD) |
|---|---|---|---|
| Controls | 11 | 16.273±1.954 | 3.747±1.479 |
| WcHT | 16 | 13.438±3.265 | 3.511±1.775 |
| PreHT | 10 | 16.30±3.592 | 3.277±1.375 |
| HT untreated | 11 | 15.545±2.911 | 2.035±1.44 * |
| HT treated | 6 | 17.833±0.983 | 3.705±2.044 |

*p<0.05

Children with untreated HT had significantly lower CBF reactivity slopes ($p<0.05$), and reached control values with antihypertensive treatment.

**Conclusions:** This abnormal CBF reactivity to hypercapnia in children with HT may be a marker for potential cerebrovascular disease, which may be treated (or prevented) with antihypertensive medication.

## Abstract# 782
**Ambulatory Arterial Stiffness Index (AASI) and Pulse Pressure in Children with Diabetes Mellitus Type 1** T. Šuláková,[1] J. Janda,[2] J. Cerná,[1] V. Janštová,[1] J. Feber.[3] *[1]Pediatrics, University Hospital Ostrava and Faculty of Health Studies, Ostrava, Czech Republic; [2]Pediatrics, University Hospital Motol, Prague, Czech Republic; [3]Pediatrics, Children's Hospital of Eastern Ontario, Ottawa, ON, Canada.*

**Objectives:** The aim of the study was to assess pulse pressure (PP) and ambulatory arterial stiffness index (AASI) in children with diabetes mellitus type 1 (DMT1) compared to non-diabetic normotensive children.

**Methods:** We retrospectively evaluated ambulatory blood pressure monitoring (ABPM) records in 84 children (43 boys) with DMT1. Based on office BP and ABPM, patients with DMT1 were divided into 3 groups [true hypertension (DM HTN) 24/84, normotension (DM NT) 33/84, white coat hypertension (DM WCH) 27/84] and compared with 27 normotensive controls (NT). PP and AASI (regression analysis of diastolic on systolic BP values corrected by the regression coefficient) were obtained from ABPM.

**Results:** DM NT, DM WCH and DM HTN did not differ in metabolic control, but DM HTN had significantly longer duration of DMT1 and significantly higher 24h, day and night BP on ABPM than DM NT, DM WCH and NT controls ($p<0.0001$). DM WCH and DM HTN had significantly higher PP compared to DM NT and NT (47.6±27.3 and 47.4±38.7 vs. 41.4±54.4 and 41.9±25.9, respectively; $p=0.0045$). AASI was significantly elevated in both DM WCH (0.35±0.14) and DM HTN (0.36±0.15) compared to NT (0.22±0.16); $p=0.0021$.

**Conclusions:** Children with DMT1 and white coat or true hypertension had significantly higher PP and AASI compared to normotensive controls and normotensive diabetics. This suggests that these children may be at increased risk for developing cardiovascular complications later on their life.

## Abstract# 783
**Ambulatory Blood Pressure Monitoring after Renal Pediatric Transplantation** M. Aglony, M. Peredo, L. Bolte, A. Vogel. *Pontificia Universidad Catolica de Chile, Santiago, Chile.*

**Objectives:** Arterial hypertension (AH) is frequent after renal transplantation and risk factor for end-organ damage. We looked for blood pressure (BP) in renal transplant recipients with casual BP (CBP) measurement and ambulatory BP monitoring(ABPM).

**Methods:** Cross-sectional study of 10 children of our renal clinic who had ABPM (Spacelabs 90217).

**Results:** All had stable renal function and 4 mild proteinuria. Eutrophic 9, overweight 1.

Patient characteristics

| Age(yr) | Primary renal disease | Donor source/ Time post transplant(mo) | Immuno-suppressor | Estimated clear (ml/min/1.73m) | Syst/Diast CBP | ABPM (BP/ DIP) |
|---|---|---|---|---|---|---|
| 9 | FSGS | L/24 | R-M-P | 89 | H2/H2 | AH/P |
| 12 | Unknown | L/22 | R-P | 112 | H1/H1 | AH/P |
| 18 | RPGN | L/32 | Cy-P | 92 | PH/H1 | AH/A |
| 14 | Alport | L/20 | T-M-P | 108 | N/N | N/P |
| 14 | Obstruct nephropathy | C/24 | R-M-P | 140 | PH/N | AH/P |
| 19 | ARPKD | L/26 | R-M-P | 94 | N/N | AH/P |
| 13 | HUS | C/45 | R-M-P | 65 | H1/H2 | AH/P |
| 17 | HUS | L/20 | T-M-P | 94 | N/H1 | AH/A |
| 12 | Glomerular dis | L/38 | Cy-M-P | 90 | H1/PH | AH/P |
| 18 | FSGS | L/16 | T-R-P | 99 | H2/H2 | AH/P |

L:living;C:cadaveric;M:Mycophenolic acid/Mofetil Mycophenolate (Myfortic®/Cellcept®), R:Sirolimus(Rapamune®);Cy:Cyclosporine(Neoral®);T:Tacrolimus(T-Inmun®);P:Prednisone;N:normal;PH:pre-hypertensive;H1:hypertensive stage1;H2:hypertensive stage2;BP:blood pressure;P:present;A:absent

CBP was altered in 70%, abnormal ABPM 90% (all with AH and 2 non-dippers also). Previous AH with treatment (1-3 drugs):5 with regular control(2 normotensive). All were persistent AH by ABPM. 3/5 patients without treatment had altered CBP at their last evaluation. The ABPM was abnormal in 4.

**Conclusions:** The prevalence of AH in our renal transplant recipients was high, being ABPM more sensitive than CBP. These data represent a pioneer experience in our country.

## Abstract# 784
**Aldosterone, Plasma Renin Activity and Aldosterone-Renin Ratio in Normotensive Pediatric Population as Screening for Primary Aldosteronism** M. Aglony, A. Martinez, C. Campino, H. Garcia, R. Bancalari, L. Bolte, C. Avalos, C. Loureiro, C. Carvajal, C. Fardella. *Pontificia Universidad Catolica de Chile, Santiago, Chile.*

**Objectives:** Primary aldosteronism(PA) is a frequent cause of secondary hypertension in adults and is suspected with an aldosterone-renin ratio(ARR)>25. In children its prevalence and normal ARR are unknown.We characterised serum aldosterone(SA), Plasma Renin Activity(PRA) and ARR in pediatric normotensive healthy population.

**Methods:** Prospective study, 211 healthy normotensive subjects(4-16 years). SA, PRA, ARR and DNA were obtained. They were divided in 2 groups: normotensive children with hipertensive parents(NH) and normotensive children with normotensive parents(NN). ARR cut off was established using the 97$^{th}$ percentile($P_{97}$) and mean plus two standard deviation(X+2SD).

**Results:** In total group, SA was 7.5±5.9 ng/dL, PRA 2.7±1.8 ng/mL/h and ARR 3.8± 6.2, subgroups values were similar. Systolic blood pressure was higher in NH than NN group.1 subject in the NH group had a chimeric CYP11B1/CYP11B2 gene. The $P_{97}$ and X+2SD values for ARR in NH group was 15 and 20.1 and in the NN group was 13.5 and 9.8 respectively. SA and PRA were negatively correlated with age and body fat percentage. ARR was not correlated with age or body fat in the 3 groups.

**Conclusions:** We demonstrated that the normal ARR in pediatric healthy normotensive population without hypertensive parents is surprisingly lower than those communicated for adult population. We propose a new cut off point for ARR that should be >10 to be used as screening in the diagnosis of PA in pediatric population

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## Abstract# 785
**Vascular Structure, Inflammatory Mediators and Oxidative Stress in Childhood Hypertension** M. Ahmed,[1,2] R. Touyz,[1] J. Feber,[2] K. Burns.[1] *[1]KRC,Ottawa Hospital Research Institute, Ottawa, ON, Canada; [2]Pediatric Nephrology, Children's Hospital of Eastern Ontario, Ottawa, ON, Canada.*

**Objectives:** The aim of the study was to evaluate biomarkers of inflammation and oxidative stress, and carotid intima-media thickness (CIMT) in childhood hypertension (HTN) and to assess the impact of obesity on these parameters.

**Methods:** Plasma levels of uric acid, IL- 18, IL-1β, ICAM-1, MCP-1, adiponectin, TBARS and 8-Isoprostanes and CIMT were measured in newly diagnosed, untreated obese (BMI>95th perc.) and nonobese children with primary HTN and compared with normotensive obese and nonobese children.

**Results:** 29 children (age14±2 yrs) were enrolled: 10 hypertensive (7 obese; 3 nonobese), 19 controls (9 obese, 10 nonobese). Plasma inflammatory and oxidative stress markers did not differ significantly between children with or without HTN; CIMT tended to be increased in hypertensive children compared to controls ($p=0.26$). Obese children with or without HTN had higher uric acid and increased CIMT compared to nonobese children.

Select biomarkers and CIMT in obese vs. nonobese children

| | Obese ± HTN (n=16) | Non obese (n=13) | P value |
|---|---|---|---|
| Uric acid (µmol/L) | 365.2±23.43 | 281.4±13.06 | 0.017* |
| TBARS (nmol/ml) | 2.83±0.15 | 2.47±0.17 | 0.14 |
| Adiponectin (mcg/ml) | 7.32±0.73 | 8.86±0.96 | 0.20 |
| CIMT (mm) | 0.57±0.02 | 0.44±0.03 | 0.005* |

**Conclusions:** Markers of inflammation, oxidative stress and vascular structure did not differ significantly between hypertensive and normotensive children. However, obese children with or without HTN had increased uric acid and CIMT. Obesity may confound the impact of HTN on markers of oxidative stress and vascular structure.

## Abstract# 786

**Ambulatory Blood Pressure Monitoring in Children with VUR: Do Microalbuminuria Levels Correlate with Changes in Blood Pressure?** A. Kilinç, H. Alpay, I. Gökçe, N. Biyikli, M. Benzer, A. Özen. *Pediatric Nephrology, Marmara Univercity, Faculty of Medicine, Istanbul, Turkey.*

**Objectives:** In this study we aimed to evaluate the variations in ambulatory blood pressure monitoring (ABPM) parameters and microalbuminuria (MA) levels in patients with vesicoureteral reflux (VUR) with respect to the extent of renal parenchymal scarring (RPS).

**Methods:** Eighty-eight patients who had been followed-up with VUR and or RPS were enrolled in the study. The grade of VUR and RPS were cumulated if bilateral. For each patient systolic, diastolic and mean blood pressures (SBP DBP, and MAP) for the three time periods -day, night and 24 hour time intervals- were transformed to standard deviation scores (SDS).

**Results:** Fifty-six patients had Grade I-IX VUR whereas 32 patients were without VUR. In 50 of the patients Grade I-III scar formation was noted while 38 patients had no RPS. When the association between ABPM parameters and degree of renal scarring was analysed, there was a significant difference in 24 hour systolic BP SDS values (p=0.042) and night systolic BP SDS values (p=0.015) among four groups (Grade 0 to III). When we analyzed the correlations between MA and ABPM parameters, 24 hour systolic BP SDS (p=0.021), day systolic BP SDS (p=0.042), systolic night BP SDS (p=0.014), 24 hour MAP SDS (p=0.009), day MAP SDS (p=0.043), night MAP SDS (p=0.01) values were significantly correlated with MA levels. In the logistic regression model, the elevation in MA was found to be independently associated with hypertension (p=0.013, O.R=25).

**Conclusions:** We conclude that MA might be a promising non-invasive diagnostic test to predict the risk of development of hypertension in patients with RPS.

## Abstract# 787

**Arterial Blood Pressure in Children Born Very Low Birth Weight** A. Mastrangelo,[1] S. Gangi,[2] S. Testa,[1] S. Ghirardello,[2] A. Edefonti,[1] F. Mosca,[2] G. Ardissino.[1] *[1]Pediatric Nephrology and Dialysis Unit, Clinica Pediatrica G. e D. De Marchi, Fondazione IRCCS Ca' Granda Ospedale Maggiore Policlinico, Milan, Italy; [2]Institute of Pediatrics and Neonatology, Fondazione IRCCS Ca' Granda Ospedale Maggiore Policlinico, Milan, Italy.*

**Objectives:** The aim of this study was to evaluate the prevalence of hypertension (HPT),early in life,in a cohort of children born VLBW.

**Methods:** Blood Pressure (BP),recorded during the follow up visit at age 5 yrs of 125 children (58 M) born VLBW (< 1.5 kg) between 1999 and 2003,was retrospectively analyzed.Mean±SD gestational age at birth was 29.6±2.6 wks and mean body weight (BW) was 1071±287 gr.BP was measured twice and the 2nd measurement was used for the present analysis.HPT was defined as a systolic (SBP) and/or diastolic (DBP) BP>95th centile (+1.65 SDS) for age, sex and height according to the 2004 Task Force on Hypertension in Children reference values.

**Results:** 14 children (11.2%) had SBP-DBP above the 95th centile,while 15 (12%) had borderline values (90-95th centile).In the group (n = 49) of children born with BW<1 kg,the prevalence of HPT was as high as 19.5%.The correlation between neonatal BW and z score of systolic and diastolic BP was r= 0.45 and 0.37,respectively.

Table 1. Distribution of abnormal blood pressure measurements by centiles in the study population

| | 90 -95 pc | 95 -97 pc | 97 -99 pc | >99 pc |
|---|---|---|---|---|
| SBP | 6 | 4 | 0 | 3 |
| DBP | 9 | 5 | 2 | 2 |

**Conclusions:** This cross-sectional study confirms the high prevalence of HPT in children born VLBW and the need for regularly monitoring BP in these patients. In our Center,all children born VLBW undergo BP measurement and urinary microlabumine and protein determination at 1,2,3,5 and 10 years of age.

## Abstract# 788

**Cardiomyopathy Due to Renovascular Hypertension in a Boy with Moyamoya Disease** A. Babaoglu,[1] K. Bek,[2] E. Ciftci,[3] B. Kara,[5] Z. Bircan.[2] *[1]Pediatric Cardiology, Kocaeli University Hospital, Kocaeli, Turkey; [2]Pediatric Nephrology, Kocaeli University Hospital, Kocaeli, Turkey; [3]Radiology, Kocaeli University Hospital, Kocaeli, Turkey; [4]Pediatric Neurology, Kocaeli University Hospital, Kocaeli, Turkey.*

**Objectives:** Moyamoya disease is a progressive carotid artery stenosis of unknown etiology often presenting in children as transient ischemic attack with focal neurological deficit.

**Methods:** Moyamoya disease was described in a boy who presented with dilated cardiomyopathy secondary to hypertension due to bilateral renal artery stenosis.

**Results:** An 8-year-old boy was admitted for the evaluation of abdominal murmur and hypertension. His weight and height were at 3rd percentile, blood pressure was 160/110 mmHg, abdominal murmur, walk disturbance, exaggerated deep tendon reflexes and clonus of the left lower limb were present. Echocardiography revealed hypertrophic and dilated left ventricle, moderate mitral regurgitation, decreased ejection fraction (50%). Abdominal aortoangiography detected stenotic segments on celiac trunk and renal arteries. Brain angio-MR findings were consistent with moyamoya disease with stenosis of cerebral arteries and collateral vessel development. Hypertension could be controlled hardly with atenolol, amlodipine and alpha methyl dopa. Aspirin was started for thrombosis prophylaxis. Renal arterial lesions were not suitable for balloon angioplasty and thus future surgery was planned.

**Conclusions:** Renal artery involvement in moyamoaya disease is an important clinical manifestation that may result cardiomyopathy secondary to severe renovascular hypertension.

## Abstract# 789

**Assessment of Left Ventricular Systolic and Diastolic Functions by Doppler Tissue Imaging in Children with End-Stage Renal Disease on Hemodialysis** H. Al-Marsafawy,[1] Z. El-Moursy,[1] A. Sarhan,[1] A. Bakr,[1] T. Mattoo,[2] A. El-Mougy.[1] *[1]Pediatrics, Mansoura University Children's Hospital, Mansoura, Egypt; [2]Pediatrics, Children's Hospital of Michigan, Detroit, MI, United States.*

**Objectives:** Cardiovascular disease (CVD) is a frequent cause of morbidity and mortality in children with end-stage renal disease (ERSD). Abnormalities of diastolic function precede those of systolic one. Few studies have assessed left ventricular (LV) diastolic function. The aim of this study is to evaluate LV systolic and diastolic functions in children with ESRD on hemodialysis and to identify the cardiac risk factors in these patients.

**Methods:** Twenty children with ESRD participated in this study, they were maintained on regular haemodialysis in Pediatric Nephrology Unit, Mansoura University Children's Hospital. A group of 20 age and sex matched children was served as a control group. Conventional echocardiography and Doppler tissue imaging studies (DTI) were performed for all patients and control.

**Results:** Left ventricular hypertrophy was found in 16 (80%) of the dialysis patients. Left ventricular mass index (LVMI) was greater in dialysis group than in control. We reported normal systolic functions in the studied patients, but, diastolic functions were affected. The LV Myocardial Performance Index is significantly higher in dialysis patients compared to controls ($p<0.001$).

**Conclusions:** Left ventricular global performance is impaired in pediatric patients with ESRD. LV systolic function is preserved, while diastolic function is impaired in these children. Hypertension, anemia, renal oesteodystrophy, acidosis, and hyperlipidemia are risk factors for development of CVD.

## Abstract# 790

**Prevalence of Obesity and Hypertension in South West Iranian Children** M. Basiratnia, D. Derakhshan, S. Ajdari. *Shiraz Nephrology Urology Research Center, Shiraz, Islamic Republic of Iran; Shiraz Nephrology Urology Research Center, Shiraz, Islamic Republic of Iran; Shiraz Nephrology Urology Research Center, Shiraz, Islamic Republic of Iran.*

**Objectives:** Obesity is likely one of the major causes of the increased prevalence of hypertension in children.The aim of this study was to investigate the prevalence of obesity and its association with hypertension in Iranian children.

**Methods:** A total of 2000 healthy students aged 11-17 years were screened. Data on weight, height, systolic and diastolic BP, parental history of hypertension and maternal educational level were obtained. Hypertension was defined as the average of three systolic and diastolic BP recordings ≥ 95th age-, sex- and height-matched percentile of the reference standard.

BMI ≥ 95% percentile was defined as obesity and between 85% and 95% percentile for age and sex considered overweight. Statistical analysis was done with Chi-square and multivariate multiple regression tests.

**Results:** Overall 7% and 11.8% of students were obese and hypertensive, respectively. 30.7% of obese compared with 8.4% of normal weight children were hypertensive. There was a strong association between obesity and hypertension ( p= 0.0001).
In multivariate analysis both BMI Z-score and age were associated with systolic and diastolic blood pressure, respectively ( p= 0.016, p< 0.001 for systolic BP, p=0.01, p< 0.001 for diastolic BP).
Parental history of hypertension and maternal educational level were not associated with hypertension.
**Conclusions:** These results confirm that obesity is an important risk factor for hypertension in children. The high prevalence of hypertension in obese children emphasizes the need for prevention of childhood obesity.

## Abstract# 791

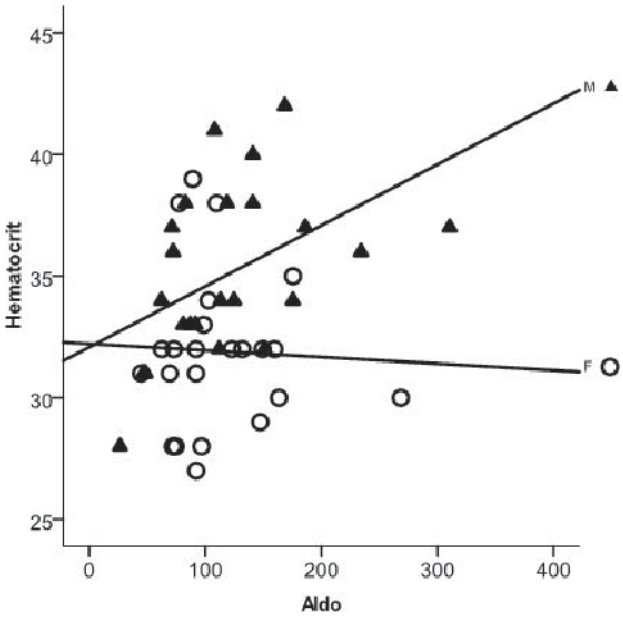
**Atherosclerosis and Arteriosclerosis Indicators in Childhood Dialysis Patients** Y. Uslu,[1] S. Yildirim,[2] S. Demir,[3] N. Cengiz,[1] K. Gulleroglu,[1] U. Bayrakci,[1] B. Varan,[2] N. Uslu,[3] E. Baskin.[1] *[1]Pediatric Nephrology, Baskent University, Ankara, Turkey; [2]Pediatric Cardiology, Baskent University, Ankara, Turkey; [3]Radiology, Baskent University, Ankara, Turkey.*
**Objectives:** In patients with chronic renal failure, vascular changes start to develop in the early phase of the renal insufficiency. Both the atherosclerosis and arteriosclerosis contribute to arterial disease in this group of patients.
**Methods:** 32 peritoneal dialysis(PD) and 26 hemodialysis(HD) patients (mean age 13.37 ± 4.14 years and 16.19 ± 2.91 years) and 20 healthy children (mean age 13.70 ± 2.49years) were included to the study. Ejection fraction(EF), left ventricular mass index(LVMI), left ventricular geometric construction, blood pressure elastic modulus and stiffness index β were measured with echocardiography. Two main carotid arteries, internal carotid artery and the intima media thickness(CAIMT) were screened with B mode doppler ultrasonography.
**Results:** The CAIMT values were increased in the patients group comparing to the control group (p<0.05). There was a positive correlation between LVMI and CAIMT (r = 0,25, p =0.039). In the patients group, EF values were lower, LVMI was higher than control. 93.6% of the patients group(89,3% concentric hypertrophy) had left ventricular hypertrophy. Measurements of stiffness index β and elastic modules were similar for two groups. There was no difference in atherosclerosis and arteriosclerosis indicators in PD and HD patients.
**Conclusions:** Although no arteriosclerosis indicators were determined in our study; the presence of atherosclerosis was shown in dialysis patients which is a risk factor for early cardiovascular disease.

## Abstract# 792

**The Relationship between Adipokines and Cardiovascular Disease in Childhood Dialysis Patients** E. Baskin,[1] Y. Uslu,[1] B. Unal,[2] S. Yildirim,[3] N. Cengiz,[1] K. Gulleroglu,[1] U. Bayrakci,[1] B. Varan.[3] *[1]Pediatric Nephrology, Baskent University, Ankara, Turkey; [2]Biochemistry, Baskent University, Ankara, Turkey; [3]Pediatric Cardiology, Baskent University, Ankara, Turkey.*
**Objectives:** Adipokines have many physiological and pathological effects on immunity and inflammation. In patients with renal failure, it is assumed that there is a low level of chronic inflammation.
**Methods:** The relationship between some of the adipokines (visfatin, adiponectin, TNFα) and cardiovascular diseases risk in dialysis patients was investigated. 58 dialysis patients (32 PD, 26 HD; mean age 14.63±3.88 years) and 20 healthy children (mean age 13.70 ± 2.49 years) were included in this study.
**Results:** In dialysis patients, serum adiponectin levels(25.85±5.17µg/ml) and serum TNF-α values(33.28±11.19 pg/ml) were higher than the control group(10.44±3.47µg/ml, 7.92±2.19 pg/ml, respectively) (p<0.05). Serum visfatin levels in dialysis patients were significantly lower than the controls(2.41±1.38ng/ml, 4,46±1.15ng/ml, p<0.05). In echocardiography, ejection fraction(EF) levels were lower but the left ventricular mass index(LVMI) was higher than the controls(p<0.05). Serum adiponectin and TNF-α levels of PD and HD patients were similar but visfatin values in PD patients were higher than HD patients(p<0.05). Visfatin had significantly negative correlation with adiponectin, TNF-α and LVMI.
**Conclusions:** This is the first study which expose the visfatin levels of pediatric patients with renal failure. Visfatin can be used as a marker of cardiovascular diseases in dialysis patients.

## Abstract# 793

**Hematocrit in Aldosterone-Mediated Cardiovascular Disease in Males** M. Beavers,[3] L. Ortiz,[1] J. White,[1] C. Hanevold,[2] G. Harshfield.[3] *[1]Pediatrics and Georgia Prevention Institute, Childrens Medical Center, Augusta, GA, United States; [2]Pediatrics, University of Washington, Seattle, WA, United States.*
**Objectives:** Previous studies have shown the role of aldosterone in adiposity-related cardiovascular (CV) risk. We studied whether hematocrit acts in aldosterone-mediated disease.
**Methods:** Subjects: 55 healthy, normotensive youths aged 15-19yrs (50% African-American, 54% male). Measures of adiposity: body mass index (BMI), subcutaneous (SAT) and visceral adipose tissue (VAT), and percent body fat (%BF). Measures of CV risk: casual systolic blood pressure (SBP), left ventricular internal dimension during diastole ($LVID_D$), left ventricular mass ($LVM/ht^{2.7}$), and hematocrit (HCT).
**Results:** Overall associations between adiposity, aldosterone, and CV risk showed significance, driven by males: VAT (r=.614) and BMI (.530)(p=.001), %BF (.495) and SAT (.442)(p=.05). Higher aldosterone was associated with higher casual SBP (r=.486), greater $LVID_D$ (r=.428), greater $LVM/ht^{2.7}$ (r=.397), and greater HCT (r=.468)(p<.05 for each) in males. No associations were noted in females.
**Conclusions:** Females have greater adiposity yet males suffer from greater adiposity-related disease. The aldosterone-hematocrit relationship (Fig 1) may imply a role for erythropoietin in the mechanism that defines these differences. Whether protective in females or pathologic in males, the mechanism remains unclear.




## Abstract# 794

**Association of eNOS T786C Promoter Polymorphism and Blood Pressure Response to Ramipril in Obese Hypertensive Children** C. Bereczki, P. Pasztor, B. Szucs, E. Endreffy, A. Barath, Z. Maroti, S. Turi. *Pediatrics, University of Szeged, Szeged, Hungary.*
**Objectives:** Several pharmacogenomic studies have showed the blood pressure (BP) response to given class of antihypertensive medications varies by genotype for different polymorphisms, but data on children are very limited. Levels of circulating nitrite/nitrate, the end-metabolites of nitric oxide, are significantly affected by ACE inhibition. The aim of this study was to determine, whether the eNOS T786C promoter polymorphism has any influence on blood pressure response to ramipril therapy in obese hypertensive children.
**Methods:** 68 obese (BMI: Z score >2.5 SD) (girl/boy:32/36) primary hypertensive (systolic or diastolic blood pressure >95th) children (aged 6-17 years) were enrolled. Patients received ramipril (0,1-0,15 mg/bwkg/day) once daily. Ambulatory blood pressure measurements (ABPM) were performed at start and after 1 month therapy.
**Results:** The reduction 24-h systolic BP depended on T786C promoter polymorphism in girls, but did not in boys (Fisher's exact test, p=0.033). The CC genotype was significantly more often observed in the girls with the treatment caused a decrease in blood pressure at least 5 mmHg.
The trend was similar in the night-time BP reduction, but it has not reached the

significant level. There was no any other significant difference in daytime and night-time diastolic blood pressure reduction in either boys or girls according to genotype.
**Conclusions:** The T786C polymorphism of the eNOS gene appears to be gender-related factor in the effect of ACE inhibitors. Further studies are necessary to examine whether other genes or hormones responsible for this gender-related effect.

## Abstract# 795

**Sleep Disordered Breathing in Children with Ambulatory Blood Pressure Monitoring and Renin Angiotensin System Evaluation** L. Bolte, M. Aglony, A. Martinez-Aguayo, N.L. Holmgren, C. Campino, C. Avalos, R. Bancalari, C. Loureiro, C. Carvajal, H. Garcia, C. Fardella. *Pontificia Universidad Catolica de Chile, Santiago, Chile.*
**Objectives:** Children with sleep disordered breathing (SDB) have higher cardiovascular risk. Our aim is to evaluate the risk of SDB in children with suspected arterial hypertension.
**Methods:** Cross sectional study in 49 children (44.9% male), median age 11.1 years. We measured arterial blood pressure (BP), 24 hour Ambulatory BP Monitoring (ABPM, Spacelabs 90217), Serum Aldosterone (SA), Plasma Renin Activity (PRA) and SA /PRA ratio (ARR). A validated SDB Risk Questionnaire (SDBRQ)was applied (score >13 was positive for SDB).
**Results:** Score >13 was present in 13/49 (26.5%) subjects, 3/13 were nondippers. We found no correlation between SDB and: nondipping pattern (p=0.5), average daytime systolic BP (p=0.4) or diastolic BP (p=0.7), nighttime systolic BP (p=0.4) or diastolic BP (p=0.95). No statistically differences were observed between subjects with or without SDB in median age (11.9 vs 10.3 years;p=0.05), BMI (94.4 vs 91.7 percentile; p=0.2), SA (5.4[4.7-10.5] vs 7.3[6.9-10.5] ng/dL; p=0.6), PRA (2.2[1.6-4.1] vs 2.2[2.1-3.1] ng/mL/h; p=0.9); and ARR (2.8[0.77-7.14] vs 3.4[3.1-4.8]; p=0.4).

SDBRQ AND ABPM

| ABPM | Normotension | Sustained hypertension | Masked hypertension | White coat hypertension | Total |
|---|---|---|---|---|---|
| N | 14 | 19 | 13 | 3 | 49 |
| SDBRQ >13(%) | 28.6% | 10.5% | 38.5% | 66.7% | 26.5% |

**Conclusions:** We could not demonstrate more SDB risk in hypertensive than normotensive children. SA, PRA and ARR between children with or without SDB were not statistically different.
Supported by Chilean Grant Fondecyt 1100356 and 1070876

## Abstract# 796

**Primary Familial Hyperaldosteronism Type 1 in Hypertensive Children: Prevalence, Clinical and Biochemical Characteristics** M. Aglony, L. Bolte, A. Martinez, C. Campino, C. Carvajal, C. Avalos, R. Bancalari, C. Loureiro, H. Garcia, C. Fardella. *Pontificia Universidad Catolica de Chile, Santiago, Chile.*
**Objectives:** Familial hyperaldosteronism type I(FH-1) or glucocorticoid-remediable aldosteronism is an autosomal dominant disorder caused by a chimeric gene(CG) CYP11B1/CYP11B2. Its prevalence and clinical presentation in pediatric population is not well defined. Our aim is to report the prevalence of FH-1 in hypertensive children and to describe their clinical and biochemical characteristics.
**Methods:** We studied 120 untreated hypertensive children aged 4 to 15 years. Plasma potassium(K), Plasma Renin activity(PRA) and serum aldosterone(SA) were measured. If aldosterone-renin ratio(ARR)>25,detection of CG was performed using long-extension PCR.
**Results:** In 4/120(3.3%) children ARR was>25. Genetic study confirmed a CG in all of them. They belonged to unrelated families, 21 first relatives were studied(9 affected), making a total of 13 patients. Severe hypertension 9/13, pre-hypertension 3/13 and normotension 1/13. PRA was suppressed 6/13, hypokalemia in 3/13, high SA level 10/13 (range 8.1-69.4 ng/dL) and ARR>25 in 11/13 (range 16.2 to 161.3).
**Conclusions:** The prevalence of FH-1 in pediatric hypertensive population was surprisingly high when ARR>25 was used as screening. Because all hypertensive children with ARR>25 were positive for a CG, we suggest that a lower ARR cut off needs to be used and shoud be determined in this population. The high variability in the clinical and biochemical characterics suggests that FH-1 is an heterogenous disease with a wide spectrum of presentation
Supported by Chilean Grant Fondecyt 1100356 and 1070876

## Abstract# 797

**Different Methods of Blood Pressure Measurements in Children** C.C. Corrado,[1] P.C. Carmina,[1] A.A. Anzelmo,[2] F.F. Lo Cascio,[2] M.M. D'Alessandro,[1] S.S. Maringhini.[1] [1]*Pediatric Nephrology, ARNAS Civico, Palermo, Italy;* [2]*School of Pediatrics, Palermo University, Palermo, Italy.*
**Objectives:** In the last years the detection of pediatric hypertension (HYT) has increased considerably. Blood pressure (BP) is not easy to measure in children. Auscultatory method of measurement of BP is considered the "gold standard". New methods need to be validated and their reliability in diagnosing hypertension is unknown. We investigated the variability of BP measurements in a school population with three different devices.
**Methods:** In 225 pupils (106 M/119 F), mean age 12.4 + -0.9 years, we recorded BP values using a sphygmomanometer device (A) and two oscillometric devices: OMRON M6 comfort, arm (B) and OMRON RX3, wrist (C). We performed a total 6 measurements of BP, two for each method, at a distance of 2 minutes. In all children we recorded: weight, height, abdominal circumference, triceps and subscapular folds.
**Results:** The average BP values (PAS/PAD) were 113.6±11.7/72.3±8.7 mmHg with A, 118.4±12.3/68.7±9.4 mmHg with B and 119.6±14.8/72±9.9 mmHg with C. Intraindividual variation with a single method were 0±7.4 (A), -2±11.9 (B), -3±14.8 (C). Intraindividual variations comparing the three methods were: 4.8±12.0 (A-B) 5.9 ± 14.1 (A-C). Both (A-B) and (A-C) were correlated ($p< 0.01$) with A values. The BP values registered with A,B,C were significantly correlated with age, BMI, waist circumference, triceps and subscapular folds ($p <0.001$).
**Conclusions:** Oscillometric devices overestimate BP and have a higher intraindividual variations particularly in children with high values on sphygmomanometer measurements.

## Abstract# 798

**Reference Values of Pulse Wave Velocity in Healthy Children** O. Cseprekál, É. Kis, A. Kerti, K. Kelen, O. Horváth, B. Göblyös, A.J. Szabó, G.S. Reusz. *1st Department of Pediatrics, Semmelweis University, Budapest, Hungary.*
**Objectives:** Pulse wave velocity (PWV) is an individual predictor of cardiovascular mortality. Normal values of PWV for pediatric patients have not been available so far. Our aim was to create a large database and to characterize determining factors of PWV in children.
**Methods:** 461 healthy children were investigated. Reference tables of PWV were created. Age- and height-specific estimates of the median (M), coefficient of variation (S) and degree of skewness (L) were obtained by maximum-likelihood curve-fitting technique. Effects of gender, age, height, weight, blood pressure and heart rate on PWV were assessed. The relation of the height-corrected PWV (PWV/h) to the normalized PWV data was assessed.
**Results:** Age and height specific reference tables and percentile curves were presented. Age, height, weight, blood pressure showed significant positive, heart rate a negative correlation with PWV. By multiple regression analysis, age and MAP remained the major predictors. A close correlation between PWV-Z values and PWV/h ($p<0.00001$) was found.
**Conclusions:** LMS reference tables permits calculation of appropriate percentiles for PWV in children. Age and MAP are the major determinants of PWV in children; however, height has to be considered in patients with growth deficit. PWV/h is offered as a simple, alternative measure of PWV. This study provides reference values for PWV in children, thereby constituting a suitable tool for longitudinal clinical studies assessing subgroups of children who are at long-term risk of cardiovascular disease.
Supported by OTKA-071730, TÁMOP-4.2.2-08/1/KMR-2008-0004, ETT 06-123/2009.

## Abstract# 799

**Cardiovascular Risk Factors in Children after Kidney Transplantation – The Impact of Steroid Dosage** M. Kaidar, M. Davidovits. *Institue of Nephrology, Schneider Children Medical Center, Petach-Tikva, Israel.*
**Objectives:** Risk factors for cardiovascular (CVR) disease present at higher rates in children after kidney transplantion (AKT) with mortality rates 100 higher than equivalent healthy children. Immunosuppressive protocol may play a role in CVR morbidity.K
**Methods:** Charts of 77 children AKT were reviewed. Group 1 (n=39) received high dose steroid protocol and Group 2 (n=38) received low dose steroid protocol. CVR parameters were evaluated at short-term (2,6m AKT) and long-term [2y and last follow-up (LFU 7.14±3.5y AKT)] follow-up. The CVR risk factors prevalence was compared overall and between groups.
**Results:** Significant reduction in CVR risk factors from 2m AKT to LFU was found: hyperglycemia 11.7% to 10%; hypercholesterolemia 48.7% to 33%; hypertriglyceridemia 50% to 12.5%; Hypertension 52.1% and 14%; need for antihypertensive treatment 54.8% to 42%, anemia 29.6% to 18.3%; hyperparathyroidism 32% to 18.3% and LVH 25.8% to 15%. Obesity rose from 12.12% to 24.56%; prevalence of CKD 3-4, rose from 4.75% to 24%.
Steroid dosage in group 1 was significantly higher than in group 2 ($p<0.001$) with higher prevalence of dyslipidemia ($p<0.001$), hypertension (52.8% to 45.5%) and need for antihypertensive treatment (65.7% to 44.7% p=0.01). No difference between groups GFR (81.03±16.7 and 78.14±16.4) was noted.
**Conclusions:** High prevalence of CVR risk factors was found in children after renal transplantation, mainly at short term follow-up. CVR risk factors are probably associated with pre transplant renal failure and intensive

immunosuppressive treatment (particularly steroids). Current treatment with rapid steroid dosage reduction can lessen the CVR morbidity without impairing long term renal function.

## Abstract# 800

**Targeted Control of Blood Pressure in Paediatric Renal Transplant Recipients** N.M. Dolan, K. Schumacher, J. Brierley, S.D. Marks. *Great Ormond Street Hospital for Children, London, United Kingdom.*

**Objectives:** Determine non-invasive measurement of CO and CI in renal transplant recipients(RTR) giving information for future studies regarding the choice of antihypertensive treatment to improve BP control.

**Methods:** Children attending renal transplant clinic were screened from July to October 2009.Cardiac output was measured transcutaneously using a continuous wave Doppler ultrasound probe at the suprasternal notch.A pilot study was initially undertaken to ensure minimal intra-observer bias.

**Results:** Thirty RTR were investigated.Two-thirds(20) received renal replacement therapy pre transplant.Follow-up since renal transplantation was 1-148(median 39.5)months.Estimated GFR was 22-74(median 46)mls/min/1.73m2.Sixteen(53%) patients were on 1-4(median1) antihypertensives during this study period with seven(23%) on at least two. Systolic BP centiles were 25th->95th(median 75th)centile.Of three patients with systolic BP above 95th centile, two were already receiving antihypertensive therapy;one(relapsed FSGS post transplant and clinically nephrotic) was receiving an ACEi, a diuretic and a calcium channel blocker, the other a calcium channel blocker.The third patient in this group had normal BP readings on subsequent visits and did not require antihypertensives.Cardiac output and cardiac index measured for these RTR were 2.5-6.8(median 4.5;normal 4-6) l/min and 2.0-4.5(median 3.3;normal 2.5-4.2)l/min/m2 respectively.

**Conclusions:** CO and CI were within low-normal limits with low-normal CO suggesting elevated SVR. Antihypertensives should therefore be targeted to reduce SVR in these patients.Non-invasive measurement of CO is simple to perform and may be a useful tool for more targeted BP management.

## Abstract# 801

**Oscillometric Measurement of Aortal Pulse Wave Velocity in Children** A. Doyon,[1] A.K. Bayazit,[2] A. Duzova,[3] D. Kracht,[4] B. Schaefer,[1] R. Zeller,[5] A. Melk,[4] U. Querfeld,[5] E. Wuehl,[1] F. Schaefer.[1] *[1]Pediatric Nephrology, University Children's Hospital, Heidelberg, Germany; [2]Cukurova University, Adana, Turkey; [3]Hacettepe Medical Faculty, Ankara, Turkey; [4]Hannover Medical School, Hannover, Germany; [5]Charite Children's Hospital, Berlin, Germany.*

**Objectives:** Aortal pulse wave velocity (aPWV) is a marker for arterial stiffness with prognostic value regarding cardiovascular morbidity and mortality. In the Cardiovascular Comorbidity in Children with Chronic Kidney Disease Study aPWV is determined yearly to evaluate progression of arterial stiffness in children with chronic kidney disease.

In this study, inter- and intraobserver reproducibility of an oscillometric device for aPWV measurements (Vicorder, SMT Medical) was evaluated in children.

**Methods:** Intraobserver reproducibility was determined by double measurements of aPWV in 81 children (age 6 - 18 y) in 5 German and Turkish centers by 6 examiners. In 16 children aPWV measurement was repeated by a different observer to determine interobserver variability.

**Results:** Mean standard deviation between 2 exams by the same observer was 0.32 ± 0.45 m/s, mean coefficient of variation 4.27%. Intraobserver variability was not significantly different between examiners (Kruskal-Wallis Test p = 0.86). Mean standard deviation for interobserver variability was 0.36 ± 0.45 m/s, mean coefficient of variation 5.2%, there was no significant interobserver variation (paired t-test p = 0.56).

**Conclusions:** Vicorder measurements are an easy non-invasive method to determine aPWV in children. High inter- and intraobserver reproducibility permit the detection of even small changes of aortal pulse wave velocity in longitudinal observations.

## Abstract# 802

**Blood Pressure Control in Casual and 24 Hour Measurements (ABPM) in Children with Chronic Kidney Disease (CKD)** D. Drozdz,[1] J.A. Pietrzyk,[1] K. Zachwieja,[1] M. Miklaszewska,[1] P. Kwinta.[2] *[1]Dialysis Unit, JUMC, Krakow, Poland; [2]Dpt. of Pediatrics, JUMC, Krakow, Poland.*

**Objectives:** Hypertension (HP) is an important cardiovascular risk factor in general population and in patients with CKD. The blood pressure (BP) control considerably influence the prognosis.

The aim of the study was an assessment of the BP control in casual and 24-hour measurements.

**Methods:** The examination covered 44 children (29 m, 15 f) with CKD at stages 1-5, mean age 12,3 years (SD=4,6), mean height 142,7 cm (SD=24), mean body mass – 39,9 kg (SD= 23) and eGFR – 32,7 ml/min/1,73m$^2$ (SD=24,8). The serum level of urea, creatinine and cystatin C was evaluated. A triple BP measurement was performed in the ward, followed by ABPM at home. Mean arterial pressure (MAP) and the percentage of SBP and DBP> 95 pc were analyzed.

**Results:** Based on casual measurements hypertension was diagnosed in 20 children. In ABPM the recommended values of MAP<75 pc were found in 19 children and hypertension in 10 children. The number of measurements of SBP >95 pc exceeded 25% of all measurements in 21 children, and for DBP in 18. As many as 33 children were diagnosed as *non-dipper*. Analyzing 3 methods of HP diagnosis, significant differences between the group of children with HP and without HP were observed only for MAP: creatinine level: 496 vs. 254 µmol/l (p=0,01), cystatin C: 5,1 vs. 2,5 mg/l (p=0,004), eGFR: 17,7 vs. 37,1 ml/min/1,73m$^2$ (p=0,03)and number of antyhipertensive drugs : 2 vs. 1 (p=0,02).

**Conclusions:** In advanced stages of CKD it is difficult to gain optimum BP control. Casual measurements do not ensure an adequate assessment of BP. Regularly performed ABPM should become an element of treatment control in children with CKD.

## Abstract# 803

**New Cardiovascular Risk Markers in Children with Chronic Kidney Disease (CKD)** D. Drozdz,[1] Z. Kordon,[2] P. Kwinta,[3] K. Sztefko,[4] J. Berska,[4] J.A. Pietrzyk,[1] A. Rudzinski.[2] *[1]Dialysis Unit, JUMC, Krakow, Poland; [2]Dpt. of Pediatric Cardiology, JUMC, Krakow, Poland; [3]Dpt. of Pediatrics, JUMC, Krakow, Poland; [4]Dpt. of Clin.Biochemistry, JUMC, Krakow, Poland.*

**Objectives:** Oxidative stress, chronic inflammation and endothelial dysfunction are causative agents of cardiovascular complications in patients with CKD. Prospective studies in adults showed that elevated levels of CRP, BNP and ADMA are all-cause and cardiovascular death predictors.

The aim of study was to find, are the above the risk factors of cardiovascular damage in children with CKD?

**Methods:** The study was conducted in 50 children (31 M, 19 F) with CKD stage 2-5. The basic patients' mean parameters were: age 11.6 yrs, height 136.7 cm and sCr 353 µmol/l, respectively. Serum cystatin C, thrombomodulin, hsCRP, oxyLDL, ADMA and BNP levels were additionaly measured. Left ventricular mass index (LVMI) and intima media thickness (IMT) were evaluated.

**Results:** Cystatin C levels correlated significantly with thrombomodulin (r=0.75, p<0.001), ADMA (r=0.33, p=0.02) and oxyLDL (r=0.35, p=0.014) levels. LVH (if LVMI >38 g/m$^{2.7}$) was diagnosed in 30 pts. Significant correlations between: 1. LVMI and BNP (r=0.63, p<0.001), hsCRP (r=0.31, p=0.032), thrombomodulin (r=0.44, p=0.02) and oxyLDL (r=0.36, p=0.01), 2. IMT and thrombomodulin (r=0.48, p=0.017), ADMA (r=0.51, p=0.011) and oxyLDL levels (r=0.44, p=0.03) were found.

**Conclusions:** Oxidative stress and endothelial dysfunction may cause cardiovascular damage in children with CKD. Thrombomodulin and ADMA seem to be reliable biomarkers of endothelial dysfunction and BNP of cardiac damage. All the above should be measured in those with LVH and increased IMT.

## Abstract# 804

**Which Are the Best Markers for Cardiovascular Risk Assesment in Children with CKD?** D. Drozdz,[1] J.A. Pietrzyk,[1] P. Kwinta,[2] Z. Kordon,[3] J. Bugajska,[4] M. Miklaszewska,[1] K. Zachwieja,[1] M. Drozdz.[5] *[1]Dialysis Unit, JUMC, Krakow, Poland; [2]Dpt. of Pediatrics, JUMC, Krakow, Poland; [3]Dpt. of Ped. Cardiology, JUMC, Krakow, Poland; [4]Dpt. of Clin. Biochemistry, JUMC, Krakow, Poland; [5]JUMC, Krakow, Poland.*

**Objectives:** Cardiovascular morbidity and mortaliy are most serious sequelae either in adults or in children with CKD but in the later group the risk factors may vary. The aim of the study was to show which of commonly assessed parameters correlate with LVH in children with CKD stage 2-5?

**Methods:** The study included 53 children (34 M, 19 F) with mean eGFR 46.2 ml/min/1,73m$^2$. The patients' mean age was 11.4 yrs, mean height 137 cm, mean sCr 349.7 µmol/l. Serum cystatin C, blood ions, albumin, PTH, Hb levels and lipid profile were measured. The patients were divided into 4 groups according to CKD staging: a) 2; b) combined 3+4; c) 5-dialysed and d) transplanted.

**Results:** Significant differences between 4 groups for systolic BP (0.01 vs. 0.2 vs 1.9 vs. 0.5 SDS; p<0.02), diastolic BP (0.9 vs. 0.8 vs. 2.8 vs. 1.1 SDS; p=0.01), Hb level (13.4 vs. 12.1 vs. 10.9 vs. 12.1; p=0.07), serum phosphorus (1.46 vs. 1.52 vs. 1.84 vs. 1.43 mmol/l; p=0.04) and PTH (28 vs. 100 vs. 245 vs. 88 pg/ml; p<0.001) were found. LVH (if left ventricular mass index >38 g/m$^{2.7}$) was diagnosed in 31 children. Significant correlations between LVMI and SBP SDS (r=0.51, p=0.001), DBP SDS (r=0.5, p=0.001), Hb (r=-0.31, p=0.02) and albumin level (r=-0.64, p<0.001) were found, respectively.

**Conclusions:** LVH is frequently prevalent in children with CKD. Hypertension, anemia and decreased albumin levels are associated with LVH, however all may be corrected during follow-up to prevent unfavorable outcome.

## Abstract# 805

**Causes, Presenting Symptoms and Pattern of Hypertension in Children; a Hospital Based Study** A.Y. Elmaghrabi,[1] S.A. Medani,[2] I. A/Azim.[1] *[1]Pediatrics, University of Medical Sciences and Technology, Khartoum, Sudan; [2]Renal Unit, Gaafar Ibn Auf Children's Hospiatal/ Alneelain University, Khartoum, Sudan.*

**Objectives:** Our objectives were to determine the causes, evaluate the presenting symptoms and identify the pattern of hypertension (HT) in children admitted to the hospital.

**Methods:** Our study is a descriptive case finding hospital based study. Children (<18 years) admitted with HT to the hospital during the study period (Dec.2008-Dec2009) were identified. Age, Gender, Presenting Symptoms and the stage of HT were assessed. Investigations obtained to identify causes and complications of HT in those children were also evaluated. Data was analyzed using SPSS version 15; Chi-square test was used to test significant relationship between variables with a p-value set at ≤0.05.

**Results:** 100 patients diagnosed with HT were identified of which 64% were males. 42% of the patients were 11-17 years with cause being significantly related to age (p=0.00). 54% of the patients were symptomatic. Vomiting and headache were reported in 26 and 23 of the symptomatic patients respectively. Vomiting was significantly related to high urea and creatinine (p=0.00). All patients were diagnosed as secondary HT with 80% being due to renal etiology, of those Glomerulopathy/Glomerulonephritis and Chronic kidney disease (CKD) were 60% and 22.5% respectively. 75% of the patients were stage 2 HT and 18% of the patients presented with Seizures. Left ventricular hypertrophy (LVH) was found in 50% of the 32 patients that had an echocardiography done.

**Conclusions:** HT in our study population was predominantly secondary to underlying renal disease and was associated with high rate of complications.

## Abstract# 806

**Delayed Diagnosis of Severe Hypertension in Children – Description of 3 Children with Fibromuscular Dysplasia and Renal Artery Stenosis** M. Faerch,[1] L. Winding,[1] N. Thrane,[2] S. Rittig.[1] *[1]Department of Pediatrics, Aarhus University Hospital, Skejby, Aarhus, Denmark; [2]Department of Pediatrics, Regional Hospital Herning, Herning, Denmark.*

**Objectives:** Renovascular hypertension is a rare, but none the less, important cause of hypertension in children. The diagnosis is often delayed because blood pressure is infrequently measured in children and high values are often dismissed as inaccurate.

**Methods:** We report the clinical course of three children with delayed diagnosis of renovascular hypertension. Two children with idiopathic renovascular hypertension and one was already diagnosed with neurofibromatosis type 1. Diagnosis was particularly delayed in the children with idiopathic disease. A 3,4 year old boy with symptoms of ADHD, admitted several times over a 4 month period with facial palsy, abdominal pain and vomiting. A 7,3 year old girl, admitted at the age of 11 months, observed for meningitis. High blood pressure was measured but disregarded. After this her left extremities developed hypoplasia. At the age of 7,2 when admitted for corrective surgery on her left leg she was found severely hypertensive. She had abdominal pain and vomiting as well as fatigue. A 4,5 Year old boy with NF1 had hypertension and symptoms of ADHD.

**Results:** In all three patients hypertension was uncontrollable by antihypertensive medication and was managed by percutaneous transluminal renal angioplasty.

**Conclusions:** Although symptoms of hypertension in children are generally thought to be few if any, these reported cases clearly demonstrate that unspecific symptoms such as abdominal pain, continuous vomiting, facial palsy and a behavioral pattern resembling that of ADHD are associated with severe hypertension.

## Abstract# 807

**Decreased Blood Pressure Rhythmicity Correlates with Increased Arterial Stiffness in Children with Primary and White Coat Hypertension** J. Feber,[1] A. Niemirska,[2] M. Litwin.[2] *[1]Dept. of Pediatrics, Children's Hospital of Eastern Ontario, Ottawa, ON, Canada; [2]Dept. of Nephrology & Arterial Hypertension, The Children's Memorial Health Institute, Warsaw, Poland.*

**Objectives:** To analyze blood pressure rhythmicity (BPR) in relation to target organ damage (TOD) and arterial stiffness in children with arterial hypertension (HTN).

**Methods:** We performed a retrospective analysis of ambulatory blood pressure monitoring (ABPM) and TOD (left ventricular hypertrophy, albuminuria) in children with primary HTN (secondary HTN was excluded). Left ventricular mass (LVM) was measured by echocardiography, ambulatory arterial stiffness index (AASI) was obtained from ABPM. BPR (24h, 12h, 8h amplitudes and acrophases) was analyzed with Fourier analysis (Chronos-Fit®) of ABPM.

**Results:** A total of 106 children aged 14.8±2.7 years were enrolled; 70 had true HTN (THTN) and 36 had white coat HTN (WCH). Both groups had similar prevalence of BPR except for a lower prevalence of 12h BPR in WCH (69%) compared to THTN (81%). There were no differences between patients with THTN and WCH in amplitudes/acrophases, LVM SDS and AASI. No significant correlation was found between BPR and TOD (LVM SDS, albuminuria); however, 24h and 12h amplitudes inversely correlated with the AASI ($r^2$=-0.39, $p<0.01$ and $r^2$=-0.29, p=0.01, respectively).

**Conclusions:** Children with primary and white coat hypertension had similar blood pressure rhythmicity, which was not correlated to target organ damage. However, decreased blood pressure amplitude was associated with increased arterial stiffness. This finding suggests that abnormal blood pressure rhythmicity may lead to early vascular dysfunction in children with hypertension.

## Abstract# 808

**Not Ready for Prime Time: Aliskiren for Treatment of Hypertension or Proteinuria in Children** J.T. Flynn. *Nephrology, Seattle Children's Hospital, Seattle, WA, United States.*

**Objectives:** Aliskiren, the first orally active direct renin inhibitor, has been shown to be effective in treatment of hypertension (HTN) in adults and may also reduce proteinuria in patients with chronic kidney disease (CKD). To date, no controlled studies of aliskiren have been conducted in children, but off-label use may be considered by some practitioners.

**Methods:** An e-mail survey of the PEDHTN and PEDNEPH listservs was conducted to solicit information on off-label use of aliskiren in children.

**Results:** Information on 9 patients was supplied from 5 centers. Mean age was 11.6y (range, 4-17y). Underlying diagnoses included renal disease (7), primary HTN (1)and unspecified (1). Indications for use of aliskiren included HTN (5) and proteinuria (6) (2 patients were treated for both HTN and proteinuria). Doses used ranged from 37.5-300 mg/day; no information was provided on dosing by body weight or surface area. All children were receiving at least 1 other antihypertensive medication at the time aliskiren was started; most were receiving multiple medications (range, 2-5). Aliskiren treatment was considered successful in 3 patients, partially successful in 2 patients, unsuccessful in one patient and not specified in 3 patients. Adverse effects reported included hypotension, angioedema and hyperkalemia (1 patient with each). Aliskiren was either withdrawn or the dose was reduced in 4 patients because of adverse effects or lack of clinical benefit.

**Conclusions:** Although conducted informally, the uneven success of aliskiren treatment and adverse effects of aliskiren reported in this small survey suggest that widespread pediatric use of aliskiren should be delayed until the results of properly conducted clinical trials become available.

**DISCLOSURE:** *Flynn, J.T.*: Consultant, Pfizer, Novartis.

## Abstract# 809

**Carotid Intima-Media Thickness in Children with End-Stage Renal Disease** A. Gheissari,[1] M. Sirous,[2] T. Hajzargarbashi,[3] R. Kelishadi.[4] *[1]Pediatric Nephrology Department, Isfahan University of Medical Sciences, Isfahan, Isfahan, Islamic Republic of Iran; [2]Radilogy Department, Isfahan University of Medical Sciences, Isfahan, Isfahan, Islamic Republic of Iran; [3]Pediatric Department, Isfahan University of Medical Sciences, Isfahan, Isfahan, Islamic Republic of Iran; [4]Pediatrics Department, Isfahan University of Medical Sciences, Isfahan, Isfahan, Islamic Republic of Iran.*

**Objectives:** There is scarce data on carotid and bulb intima-media thickness(IMT-C and IMT-B) as an early marker of atherosclerosis and related factors in children on dialysis. Since we had not enough information about our patients, this study was carried, on all ESRD children in a referral center.

**Methods:** Data was collected based on sixteen ESRD children under 18 years old containing 7 patients on PD and 9 patients on HD. Serum von Willebrand factor (vWF), homocystein, apo lipoprotein A, apo lipoprotein B and quantitative CRP were measured in fasting patients just before initiating dialysis. IMT-C and IMT-B were measured by gray scale ultrasound using 7.5 MHZ probe.

**Results:** Mean of systolic blood pressure in HD group was significantly higher than PD group, 135.55±25.54 mmHg versus 121.42±12.14 mmHg, $p<0.05$. Significant differences among all following parameters in ESRD patients, with normal laboratory values, were clarified : cholesterol, triglycerides, Apo A, Apo B, quantitative CRP, VWF,homocystein and IMT-C. After adjusting for age, partial correlation showed significant correlation between IMT-C and following factors: n-PTH and serum alkaline phosphatase.

**Conclusions:** Longitudinal studies with large size samples are needed to clarify the contributing factors with intima –media thickness in ESRD children.

## Abstract# 810

**A Four Year Old Girl with Both HIV Infection and Takayasu Arteritis**

T.M. Hanni, D. Hahn, W. Henson. *Pediatric Nephrology, University of the Witwatersrand, Johannesburg, South Africa; Pediatric Nephrology, University of the Witwatersrand, Johannesburg, South Africa; Pediatric Cardiology, University of the Witwatersrand, Johannesburg, South Africa.*

**Objectives:** Takayasu arteritis is the leading cause of renovascular hypertension in South Africa.This little girl presented with hypertensive encephalopathy and seizures. CT angiogram showed right renal artery stenosis and a hypoplastic right kidney. The kidney was nonfunctional. Histology of the resected kidney proved ischaemic glomerulopathy.The TB Mantoux test was strongly positive and the ESR was raised. The HIV elisa test was positive, with a CD4% 18.

**Conclusions:** This case demonstrates the coexistence of these two diseases in a child.

## Abstract# 811

**Hypertension in Williams Syndrome Due to Undescribed Vascular Lesions**

C. Herrero, E. Lara, M. Del Campo, Á. Madrid, R. Vilalta, G. Enríquez, P. Betrián, I. Barber, C. Aso, J. Nieto. *Hospital Vall d'Hebron, Barcelona, Spain.*

**Objectives:** Williams syndrome (WS) is a complex developmental disorder characterized by congenital cardiovascular disease, dysmorphic facies, mental retardation and infantile hypercalcemia. WS is due to microdeletion of chromosome 7q11.23 involving elastin gene and 25 others. Cardiac involvement is well known and often requires intervention in childhood. High blood pressure (HBP) is seen in 50% of patients, even in the absence of cardiac malformations, and goes often underdiagnosed.

**Methods:** We have evaluated a total cohort of 47 WS patients. In order to study their cardiovascular system, we have performed 41 ambulatory blood presssure monitoring (ABPM), 44 echocardiogram, 27 systemic vascular doppler ultrasound and 15 angioTC of the abnormal vascular lesions detected by ultrasound.

**Results:** We have found 29/41 abnormal ABPM. It has been diagnostic of HBP in 18 patients and has detected a loss of the nocturnal dip in 25 patients.

The echocardiogram study has detected a total of 31/44 abnormalities, being the most frequent finding the supravalvar aortic stenosis (19) and defining surgery in 4 patients.

Doppler ultrasound detected abnormalities (14/27) including infradiaphragmatic aortic narrowing and accelerated arterial flows in carotid, renal and abdominal branches. The angioTC performed confirmed the lesions showing midaortic diffuse narrowing and stenosis of the celiac and superior mesenteric arteria.

**Conclusions:** We described new vascular lessions in WS as cause of HBP that consist of the narrowing of the infradiaphragmatic aorta and some visceral branches.

## Abstract# 812

**The Correlation between Atherosclerosis Risk Factors and Carotid Intima Media Thickness in Children with Nephrotic Syndrome**

N. Hooman,[1] S.-H. Mostafavi,[2] F. Hallaji,[2] R. Isa-Tafreshi,[3] H. Otukesh.[1] *[1]Pediatric Nephrology, Iran University of Medical Sciences, Tehran, Islamic Republic of Iran; [2]Radiology, Iran University of Medical Sciences, Teharan, Islamic Republic of Iran; [3]Pediatric Cardiology, Iran University of Medical Sciences, Tehran, Islamic Republic of Iran.*

**Objectives:** To investigated the atherosclersosis parameters in children with nephrotic syndrome.

**Methods:** Thirty one children(22M,mean age 9yr) with history of nephrotic syndrome were enrolled into the study between 2008 and 2009. The inclusion criteria were: Idiopathic NS with normal serum complement, being at least one year on therapy,GFR>90ml/min/1.73m$^2$,Age>2 year at the time of study. Consent was taken from parents or patient. Fasting blood sample was drawn and 24 hour urine was collected. Carotid intima –media thickness (cIMT), left ventricular mass index (LVMI) and left ventricular hypertrophy (LVH) were studied in all children.P<0.05 was considered significant.

**Results:** Nine out of 31 patients was steroid responsive. The mean cIMT (mm) was 0.45(± 0.16) compared to 0.43(±0.1) in control group (P >0.0.05). There was no correlation between cIMT and serum homocystein, vitamin B12, and folic acid level neither in frequent nor infrequent relapses(P>0.05). Patients who received cyclosporine had significantly higher cIMT (0.5 vs 0.42 mm) (P=0.042), LVMI(42 vs 47 gr/m$^2$/ht$^{2.7}$), and lower vitamin B12 level (341vs 535 pg/ml) (P=0.02). LVH was detected in27.3% of patients on cyclosporine compared to 6.1% who had not received it (RR=4.5).

**Conclusions:** children with nephrotic syndrome who were under therapy with cyclosporine had higher rsik of athersclosrosis.

## Abstract# 813

**Left Ventricular Hypertrophy in Children with Chronic Kidney Disease**

A. Jankauskiene,[1,2] R. Cerkauskiene,[1,2] A. Malikenas,[1] V. Cerniauskiene,[1] V. Lukosiene.[1,2] *[1]Pediatric Department, Vilnius University Children Hospital, Vilnius, Lithuania; [2]Vilnius University, Vilnius, Lithuania.*

**Objectives:** The aim of the study was to evaluate influence of glomerular filtration rate (GFR) on the left ventricular hypertrophy (LVH) in children with chronic kidney disease (CKD).

**Methods:** The study population consisted of 56 patients with CKD and control group with no evident signs of renal or cardiovascular disease. Patients were devided into 4 groups: I group -GFR 60 - 30 ml/min/1.73 m$^2$; II group - GFR 29.9 - 15 ml/min/1.73 m$^2$, III group - patients on dialysis, IV group - patients after kidney transplantation. The left ventricular mass (LVM) was calculated from echocardiographic measurements and indexed to height$^{2.7}$.

**Results:** Regression analysis showed direct correlation between the GFR and LVMI (r = 0.45, p< 0.004).

Table 1.Demographic and echocardiographic data

| Variable | I group (n-16) | II group (n-10) | III group (n-13) | IV group (n -17) | Control group (n 15) |
|---|---|---|---|---|---|
| Age (y), mean st deviation | 9.2 ± 5 | 11.4± 5 | 11.2±6 | 13.6 ±4 | 9.7± 5.1 |
| Gender | 5/11 | 3/7 | 6 /7 | 10/ 7 | 7/8 |
| GFR (ml/ min/ 1.73m$^2$) | 47.5 ± 9 | 23.4± 5 | 12.8 ±6.2 | 62.5± 34.4 | 119± 20.1 |
| RWT | 0.36 ± 0.05 | 0.35± 0.09 | 0.41 ± 0.09* | 0.38 ±0.05* | 0.34 ± 0.04 |
| LVM (g) | 83.28 ± 43.33 | 140.09 ±155.32 | 118.7 ± 55.49* | 129.95 ± 49.3* | 80.71 ± 36.91 |
| LVMI (g/m2.7) | 43.77 ± 25.81 | 49.76 ±30.21* | 59.69 ± 45.3** | 41.91 ± 8.17** | 32.05 ± 5.43 |

* p < 0.05 ; ** p< 0.001 vs control group

**Conclusions:** Study showed negative influence of GFR on LVH with slight improvement after kidney transplantation. Further studies are needed for evaluating other risk factors and treatment effects.

## Abstract# 814

**Pulse Wave Velocity and Intima-Media Thickness in Renal Transplant Children**

E. Kis,[1] O. Cseprekal,[1] A. Kerti,[1] T. Horvath,[2] M. Kollai,[2] A. Szabo,[1] G.S. Reusz.[1] *[1]1st Dept. of Pediatrics, Semmelweis University, Budapest, Hungary; [2]Institute of Human Physiology and Clinical Experimental Research, Semmelweis Univerity, Budapest, Hungary.*

**Objectives:** Vascular calcification, accelerated by uraemia, could be characterized by pulse wave velocity (PWV), arterial distensibility (D) and intima-media thickness (IMT). These parameters can individually predict cardiovascular mortality in adults, but data is sparse in renal transplant (tx) children.

**Aims:** To describe functional (PWV, D) and structural (IMT) vascular disorders and to analyse vascular risk factors in children after tx.

**Methods:** In this cross-sectional study vascular parameters (PWV, IMT, D) of 24 tx patients aged (mean (SD)) 16,6(4,9) years, were measured by applanation tonometry and sonographic evaluation of carotid artery 4,5(3,1) years after tx. Results were expressed as standard deviation scores (SDS). Routine laboratory parameters were analysed.

**Results:** Carotid artery distensibility of tx children was in the normal range (D SDS:-0,01(0,98)). PWV SDS was increased (0,97(0,71)) and mean IMT SDS was around the 95th percentile (1,64(1,36)). There was a positive correlation between PWV SDS and creatinine, P and CaxP (r=0,51;r=0,40;r=0,46). In the subgroup of tx children with significantly increased IMT we found elevated P level and CaxP product at first year control following tx (IMT SDS:0,13 vs 2,61; P:1,24 vs 1,63mmol/l; CaxP:3,19 vs 4,18mmol$^2$/l$^2$).

**Conclusions:** There are structural (IMT) and functional (PWV) vascular alterations in tx children 4,5 years after tx. Our results suggest that decreased graft function and disturbed Ca-P metabolism might have a role in posttx vascular disorders.

OTKA 71730, TÁMOP-4.2.2-08/1/KMR-2008-0004, OTKA 49690

## Abstract# 815

**Hyponatremic Hypertensive Syndrome in Pediatric Patients: Is It Really so Rare?**

Y. Kovalski,[1] R. Cleper,[1] I. Krause,[1] A. Belinky,[2] M. Davidovits.[1] *[1]Institute of Pediatric Nephrology, Schneider Children's Medical Center of Israel, Petah Tiqwa, Israel; [2]Interventional and Vascular Radiology Unit, Rabin Medical Center, Petach tiqwa, Israel.*

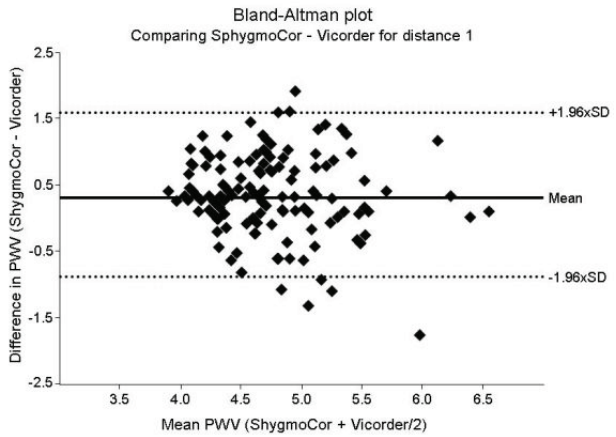
**Objectives:** To evaluate the prevalence of hyponatremic hypertensive syndrome (HHS) among children with renovascular hypertension and describe its clinical characteristics.

**Methods:** All children referred to the Institute of Nephrology, Schneider Children's Medical Center of Israel who were diagnosed with renovascular hypertension over a 5-year period (2003-2007) were included in the study. Patient medical charts were retrospectively reviewed and the relevant data analyzed. Children with accompanying hyponatremia (blood sodium <130 mEq/l) were diagnosed with HHS.

**Results:** Fourteen children were diagnosed with renovascular hypertension. Four (28%) were found to have HHS, all presented with polyuria and polydipsia, electrolyte disturbances, metabolic alkalosis, variable tubular dysfunction and nephrotic range proteinuria along with hypertension. In one patient, glomerular and tubular abnormalities preceded the development of hypertension. Symptoms resolved after resolution of ischemia following percutaneous angioplasty.
**Conclusions:** HHS is probably more common than previously thought and is reversible after correction of underlying renal artery stenosis.

## Abstract# 816

**Aortic Pulse Wave Velocity in Children and Young Adults – Validation of the Vicorder Device in Healthy Individuals** D. Kracht,[1] S. Baig,[2] A. Doyon,[3] C. Jacobi,[1] F. Schaefer,[3] E. Wühl,[3] B. Schmidt,[2] A. Melk.[1] *[1]Pediatric Nephrology, Medical School, Hannover, Germany; [2]Nephrology, Medical School, Hannover, Germany; [3]Pediatric Nephrology, University Hospital, Heidelberg, Germany.*
**Objectives:** Aortic pulse wave velocity (aPWV) is an estimate of aortic stiffnes. This study compares aPWV values of young individuals measured by applanation tonometry (SphygmoCor) and with a new oscillometric device (Vicorder).
**Methods:** aPWV in 131 healthy children (age 6-18 years) was measured 1. by sequential tonometry and 2. by oscillometry in a synchronic manner. SphygmoCor aPWV was calculated with distance 1 (dist.1) [=(Suprasternal notch(SSN) to Umbilicus(Umb))+(Umb to A.fem.)-(SSN to A.car.)]. For Vicorder 3 additional distances were used: dist.2 [=(SSN to Umb)+(Umb to A.fem.)]; dist.3 [=SSN to A.fem.]; dist.4 [=(SSN to A.fem.)-(SSN to A.car.)].
**Results:** Mean SphygmoCor aPWV was 4.9±0.6 m/s. Vicorder aPWV were 4.6±0.6 m/s (dist.1; r=.46 for correlation with Sphygmocor; p<.0001); 5.3±0.7 m/s (dist.2; r=.44; p<.0001); 5.2±0.7 m/s (dist.3; r=.45; p<.0001); 4.6±0.6 m/s (dist.4; r=.46; p<.0001). Using dist.1, the lowest deviation was seen (11.3±8.1%).




**Conclusions:** The variability between both methods is well acceptable using dist.1. Since the Vicorder is easier to handle, needs less operator training and measurements are done more quickly, it seems the appropriate device for larger cohort studies.

## Abstract# 817

**Low-Molecular Weight Adiponectin Is Associated with Left Ventricular Mass Index** M. Lo,[1] S. Furth,[2] B. Warady,[3] M. Mitsnefes.[1] *[1]Cincinnati Children's Hospital, Cincinnati, OH, United States; [2]CKiD Investigator, Philadelphia, PA, United States; [3]CKiD Investigator, Kansas City, MS, United States.*
**Objectives:** The mechanisms of left ventricular hypertrophy (LVH) in children with CKD are not fully understood. The known risk factors account for only 15-44% of variability in left ventricular mass index (LVMI) in pediatric studies. We investigated the associations of LVMI with adiponectin, an adipokine known to be cardioprotective.
**Methods:** We determined total adiponectin and its sub-fractions (high-molecular weight (HMW), low-molecular weight (LMW), and trimer) cross-sectionally in a sub-cohort of patients from the CKiD study (n=82). Echocardiograms were obtained using a standardized protocol at local sites and read centrally. Uni- and multi-variate linear regression analysis was used to quantify the relationships between LVMI and adiponectin and other risk factors of interest including BP.
**Results:** Mean age 12.3±3.5 years; 56% male; mean iGFR (iohexol) 45.3±17.4 mL/min/1.73m$^2$. Mean total adiponectin level was 17.2±6.4mcg/mL. Mean adiponectin sub-fractions were HMW 43.2±13.1%, LMW 34.9±7.9%, trimer 21.8±10.3%.

Multivariate analysis of LVMI predictors in the CKiD children

| Variable | Coefficient | 95%CI | p-value |
|---|---|---|---|
| % LMW adiponectin | -0.26 | -0.50, -0.03 | 0.03 |
| BMI Z-score | 2.25 | 0.49, 4.01 | 0.01 |
| Age, years | -0.68 | -1.23, -0.13 | 0.02 |

**Conclusions:** This study is the first characterization of adiponectin sub-fractions on intermediate cardiovascular outcome (LVMI) in CKD. Despite a higher total adiponectin level, decreased LMW fraction predicts a higher LVMI in the CKiD population. This effect was independent after adjustment for BMI, age, and casual BP.

## Abstract# 818

**Predictors of Adiponectin in CKiD** M. Lo,[1] S. Salisbury,[1] S. Furth,[2] B. Warady,[3] M. Mitsnefes.[1] *[1]Cincinnati Children's Hospital, Cincinnati, OH, United States; [2]CKiD Investigator, Philadelphia, PA, United States; [3]CKiD Investigator, Kansas City, MS, United States.*
**Objectives:** In contrast to the general population, total adiponectin is increased in patients with chronic kidney disease (CKD) despite increased cardiovascular disease (CVD). Adiponectin circulates as trimer, low-molecular-weight (LMW) and high-molecular-weight (HMW) complexes. The distribution and role of each subfraction in CKD is unknown. We hypothesized that imbalances in the subfractions rather then total adiponectin is associated with increased CV risk in children with CKD.
**Methods:** Total adiponectin and subfractions were measured cross-sectionally in a sub-cohort (n=105) of patients from the CKiD study. Uni- and multi-variate log-linear regression was used to quantify the relationships between adiponectin fractions and CV risk factors.
**Results:** Mean age 12.2±3.8 years; 56% male; mean iGFR (iohexol) 46.3±17.8 mL/min/1.73m$^2$. Total adiponectin 17.3±6.3mcg/mL. Mean HMW 43.2±13.1% and LMW 34.9±7.9%. In multivariate analysis, HMW was associated with age (-1.30, -1.90 to -0.70), BMI Z-score (-5.17, -7.28 to -3.06), and log urine protein to creatinine ratio (4.30, 2.28 to 6.32), all p<0.001. iGFR was not a significant predictor for HMW after adjusting for these factors. In contrast, LMW was associated with iGFR (0.12, 0.03 to 0.20, p=0.004) and was higher for pubertal vs pre-pubertal children (4.24, 1.34 to 7.14, p=0.005).
**Conclusions:** The increase in total adiponectin levels with progressive CKD is associated with decreasing percentages of the LMW fraction after adjusting for all other significant predictors. This imbalance in subfractions may be important for increased CV risk despite higher levels of total adiponectin in children with CKD.

## Abstract# 819

**Blood Pressure Association with Insulin Resistance and Adipokines in Obese Children and Adolescents** S. Málaga, J.J. Díaz, L. Somalo, I. Riaño, F. Alvarez. *Pediatric Nephrology, Hospital Universitario Central de Asturias, Oviedo, Asturias, Spain.*
**Objectives:** Pediatric obesity is reaching epidemic figures. It has been argued that insulin resistance (IR) is the fisiopathological link between obesity and hypertension. Several molecules produced by the adipose tissue (adipokines) have been implicated in the pathology of both processes, indicating the existence of a low grade inflammatory state in the base of atherosclerosis.We aimed to analyse the association between blood pressure (BP), insulin resistance and adipokines in obese children and adolescents.
**Methods:** 55 (40 boys) obese patients aged 6 to 16 years (mean 10,17 years). All subjects underwent physical examination, including automatic BP measurement. A 12 hour fasting blood sample including lipid profile, glucose, insulin, high sensitivity CRP, leptin and adiponectin was performed. HOMA index was calculated. IR was defined as having a HOMA value greater than 3.8. Obesity was defined according to body mass index (BMI) IOTF cut-off values for age and sex. Unpaired t-test, Pearson and Spearman correlation analysis were performed.
**Results:** Nineteen children showed IR (34,5%). Systolic BP was significantly higher in obese children with IR compared to those without (123,6 vs 115,5 mmHg; p= 0.049). No differences were observed for diastolic BP. Systolic BP showed a positive significant correlation with abdominal circumference, age, BMI, HOMA, insulin, CRP, and leptin. Diastolic BP only showed significant positive correlations with abdominal circumference and age.
**Conclusions:** Obese patients show high IR prevalence. There is a significant association between BP and adipokines, indicating a possible fisiopathological link.

## Abstract# 820

**Effects of Blood Pressure Variability on Left Ventricular Mass in Pediatric Hypertension** J. Mahgerefteh, K. Paterno, M. Pagala, P. Ramaswamy, J.C. Kupferman. *Pediatrics, Maimonides Infants and Children's Hospital, Brooklyn, NY, United States.*
**Objectives:** To determine whether systolic blood pressure (SBP) variability affects left ventricular mass in children with hypertension (HT), prehypertension (preHT) and white coat hypertension (wcHT).

**Methods:** BP variability was assessed by mean 24 hour SBP standard deviation (SD), daytime SBP SD and night time SBP SD. Left ventricular mass assessed by echocardiogram was used to calculate left ventricular mass index (LVMI). Differences in age and BMI z-scores were assessed by one way ANOVA. Correlation between LVMI and each SBP SD was assessed by linear regression analysis.
**Results:** Eighty-seven untreated children (6-19 years) underwent ambulatory BP monitoring. There were no differences in mean age (14.6±3, 14.2±3.8 and 13.5±3.3, p=0.41) or BMI z-scores (1.55±0.91, 1.77±0.76 and 1.74±0.76, p=0.62) among HT, preHT and wcHT groups.

Table 1-BP variability in HT, preHT and wcHT

| | 24h SBP SD | r value (p value) | Day SBP SD | r value (p value) | Night SBP SD | r value (p value) |
|---|---|---|---|---|---|---|
| HT (n=20) | 14.5±2.7 | 0.23 (0.33) | 13.3±2.8 | 0.32 (0.17) | 10.4±3.6 | 0.58 (0.007) |
| preHT (n=16) | 15.1±3.6 | 0.06 (0.79) | 13.8±3.3 | 0.09 (0.73) | 10.9±3.4 | 0.06 (0.82) |
| wcHT (n=51) | 12.8±2.3 | 0.17 (0.23) | 12.1±2.5 | 0.24 (0.08) | 9.8±3.6 | 0.10 (0.48) |

In HT group, LVMI had a significant (p=0.007) positive correlation (r=0.58) with night SBP SD, but no significant correlation with day or 24 hr SBP SD. In preHT and wcHT groups, there were no significant correlations.
**Conclusions:** In HT children, higher night SBP variability is associated with increased left ventricular mass.

## Abstract# 821

**Renal Fibromuscular Dysplasia (FMD) Is Unlikely To Be Familial and Is Not Caused by Smooth Muscle Alpha Actin (ACTA2) Mutations** S.D. Marks, A.M. Gullett, K. Tullus, R. Kleta, A.S. Woolf. *Department of Paediatric Nephrology, UCL Institute of Child Health and Great Ormond Street Hospital for Children NHS Trust, London, England, United Kingdom.*
**Objectives:** ACTA2 mutations have recently been reported in muscular malformations of extra-renal arteries. We investigated blood pressures (BP) of first-degree relatives of a cohort of non-syndromic FMD index cases and sought ACTA2 mutations by sequencing all exons.
**Methods:** Leucocyte DNA was obtained from index cases and ACTA2 sequenced. BP of first degree relatives were measured using standard devices and, when indicated, 24-hour ambulatory BP monitoring (ABPM) was performed.
**Results:** Thirteen unrelated index cases (62% female, 85% Caucasian) aged 2 - 32 (median 15) years were recruited. There were 40 first-degree relatives comprising 22 (55% mothers) parents aged 28 - 58 (median 44) years and 18 (55% male) siblings aged 3 - 30 (median 13) years, including two consanguineous families. Hypertension was evident in 27% of parents from different families (five [80% male] were on anti-hypertensive medications, with one mother discovered to be hypertensive by BP measurement and subsequent ABPM). All eight adult siblings were normotensive. Of the ten screened siblings aged <18 years, one female teenager was pre-hypertensive (90-95$^{th}$ centile), and the remainder were normotensive. No ACTA2 mutations were found in index cases.
**Conclusions:** Hypertension occurs in 25% of the general adult population, so the incidence of hypertension in parents of index cases was unremarkable. Familial cases of FMD are rare although none of the hypertensive parents have had renal angiography. ACTA2 mutations were not responsible for FMD in our cohort.

## Abstract# 822

**Decrease of Visceral Fat Determines Regression of Target Organ Damage in Boys with Primary Hypertension** A. Niemirska,[1] R. Janas,[1] E. Jurkiewicz,[1] J. Feber,[2] M. Litwin.[1] *[1]The Children's Memorial Health Institute, Warsaw, Poland; [2]University of Ottawa, Ottawa, ON, Canada.*
**Objectives:** The main intermediate phenotype of primary hypertension (PH) in adolescents is obesity and metabolic abnormalities of the metabolic syndrome. The aim of the study was to test the hypothesis that lowering of blood pressure (BP) and target organ damage (TOD) regression correlates with the decrease of visceral fat (VAT) in hypertensive boys.
**Methods:** 24h ambulatory BP and TOD were evaluated in 50 boys (13.6 ±2.5 yrs) with PH before and after 12 months of therapy with angiotensin converting enzyme inhibitors or angiotensin receptor blockers. TOD was assessed by left ventricular mass index (LVMi) and carotid intima media thickness (cIMT). Fat distribution was determined as waist circumference (WC), waist-to-hip ratio (WHR) and by magnetic resonance as subcutaneous fat, VAT, intraperitoneal VAT, and deep subcutaneous fat.
**Results:** Pts in whom BP lowered (n = 28) and pts with no change of BP differed only in intraperitoneal VAT decrease (-480 ±766 vs -93 ±478 mm$^2$; p = 0.04). In pts in whom cIMT decreased (n=32) serum leptin also decreased (-16.2 ±21.6 vs -3.0 ±1.6 ng/ml; p = 0.01) and groups did not differ regarding changes in BP, anthropometrical and biochemical variables. In 30 pts in whom LVMi decreased WC and WHR also decreased. 24h mean arterial BP decrease was predicted by the decrease of BMI (b = 0.458), WC (b = 0.653) and increase in adiponectin (b = -0.330; $R^2$ = 0.9; p = 0.0001). LVMi regression was predicted by the decrease of WHR (b = 0.709; p = 0.04; $R^2$ = 0.42).
**Conclusions:** In hypertensive boys the decrease of BP and regression of TOD are predicted by the decrease of visceral obesity.
**DISCLOSURE:** *Niemirska, A.*: Grant/Research Support, Clinical Study. *Litwin, M.*: Grant/Research Support, Clinical Study.

## Abstract# 823

**Low Efficacy of Office Blood Pressure in Detecting Hypertension in Transplant Recipients Comparing to Ambulatory Blood Pressure Monitoring** D. Paripovic, M. Kostic, B. Spasojevic, D. Kruscic, A. Peco-Antic. *Nephrology Department, University Children's Hospital, Belgrade, Serbia.*
**Objectives:** Our aim was to access the ability of office blood pressure (OBP) to predict hypertension in pediatric kidney transplants.
**Methods:** Study population consisted of 39 kidney transplant recipients (24 males) with stable functioning renal graft aged 6-20 years. Median age after transplantation was 14.5 years. OBP was performed at three different occasions and average value was used for further analysis. Office hypertension was defined when indexed office systolic and/or diastolic BP was ≥ 1. Ambulatory hypertension was defined as mean systolic or diastolic BP index at day-time ≥ 1 or BP load ≥ 25% and with appropriate values (mean 24h systolic BP ≥ 135 mmHg and mean 24h diastolic BP ≥ 85 mmHg) for those over 18 years. Kappa coefficient and receiver operator curve (ROC) were used to compare diagnostic efficacy of OBP and ABPM.
**Results:** Hypertension was detected in 44% of patients with CBP and in 62% with ABPM. Kappa coefficient (0.352) revealed a mild agreement between OBP and ABPM. The sensitivity and specificity of the OBP to predict hypertension were 58% and 80%. Diastolic OBP was a better predictor of ambulatory hypertension (ROC area under the curve (AUC) = 0.76 ± 0.08) than systolic OBP (AUC = 0.61 ± 0.09).
**Conclusions:** OBP often fails to detect hypertension in patients with kidney transplants. It is very important to perform regular ABPM in order to detect hypertension.

## Abstract# 824

**Risk Factors for Chronic Renal Disease in a Cohort of Pediatric Hypertensive Patients with Nephropathies** R.M. Pereira,[1] R.A.S. Gomes,[2] B.M. Vitor,[2] A.F. Leite,[2] E.A. Oliveira,[2] A.C. Simoes e Silva.[2] *[1]Hospital Foundation of Minas Gerais - FHEMIG, Belo Horizonte, Minas Geria, Brazil; [2]Pediatrics, Federal University of Minas Gerais, Belo Horizonte, Minas Gerais, Brazil.*
**Objectives:** The aim of this study was to identify risk factors for chronic renal disease (CRD) in a cohort of pediatric hypertensive patients with nephropathies.
**Methods:** In this retrospective cohort study, the records of 134 patients diagnosed with arterial hypertension due to renal diseases were included. These patients were followed at our Unit between 1994 and 2009. A data base was used for statistical analysis and patients were divided according to the development or not of CRD.
**Results:** A total of 21 patients (15.6%) among 134 developed CRD during a follow-up of 8.4±2.5 years. No differences were obtained in sex, race, age, etiology of renal disease, body weight, stature and mass index in the comparison between patients with versus without CRD. Blood pressure levels at admission was also similar in both groups, however the mean systolic and diastolic blood pressure remained significantly higher in patients that developed CRD. Despite similar at admission, serum creatinine levels progressively elevated in CRD group (3.8±2.8 vs. 0.6±0.2mg/dL). The frequency of target-organ injury was significantly higher in CRD group (43% vs. 9%). The logistic regression model showed that the elevation of serum creatinine, the persistence of hypertension and the presence of target-organ injury significantly and independently contributed to the development of CRD.
**Conclusions:** Our results were in accordance with the literature and showed the importance to evaluate pediatric hypertensive series.

## Abstract# 825

**Vitamin-D-Induced Calcification in Uremic Rats: Effect of Cinacalcet (CINA) and Parathyroidectomy (PTX)** S. Jung,[1] D. Müller,[1] S. Krämer,[2] H. Peters,[2] U. Querfeld.[1] *[1]Pediatric Nephrology, Charité, Berlin, Germany; [2]Nephrology, Charité, Berlin, Germany.*
**Objectives:** Both calcitriol and PTH are involved in the pathogenesis of vascular calcifications. We have studied the question to what extent vascular calcifications induced by high-dose calcitriol treatment can be prevented by parathyroidectomy (PTX) and cinacalcet (CINA) in a rat model of uremia (5/6 nephrectomy).
**Methods:** 5 groups of animals were studied: sham-operated (CON), uremic (U), uremic + calcitriol (U+vitD), uremic+calcitriol+PTX (U+vitD+PTX), uremic+calcitriol+CINA (U+vitD+CINA).
**Results:** PTH was significantly lower in U+D rats compared to U rats, and further suppressed to a similar degree in animals treated with PTX or CINA. The serum calcium-phosphate product was elevated in all groups receiving calcitriol,

without a significant difference between groups. Growth and weight gain was not suppressed in the U+vitD+CINA group.Vascular calcifications (%area of aortic wall) were not seen in CON and U animals, present (>20%) in U+vitD animals, and reduced to a similar degree (<10%) in both the U+vitD+PTX and the U+vitD+CINA group. In calcified aortas, the presence of chondrocytic cells, positive staining for chondrocyte matrix, collagen X and the transcription factor sox9 indicated endochondral bone formation in calcitriol-treated rats.
**Conclusions:** In uremic rats, CINA attenuates vitamin D- induced calcification; it has no suppressive effect on growth after 3 months of treatment. Neither CINA nor PTX can completely prevent vitamin-D induced calcification. Calcification resembling endochondral bone formation occurs by direct vascular effects of calcitriol, independent of serum calcium and serum phosphorus concentrations.

## Abstract# 826

**Dyslipidemia in Chronic Kidney Disease** J. Saland,[1] L. Satlin,[1] H. Ginsberg.[2] *[1]Pediatrics, Mount Sinai School of Medicine, New York, NY, United States; [2]Medicine, Columbia U College of Physicians & Surgeons, New York, NY, United States.*
**Objectives:** We hypothesized increasing degree of CKD is associated with impaired post-prandial triglyceride (TG) clearance.
**Methods:** A high-fat meal was provided to non-nephrotic subjects with CKD. TG area under the curve (AUC) over the next 8 hours was the measure of TG catabolic capacity. The hypothesis was tested by correlation to estimated GFR (eGFR) and urine protein:creatinine ratio (uPCR).
**Results:** 17 subjects with eGFR 12 to 173 ml/min/1.73m$^2$ participated. AUC was inversely correlated to eGFR ($R^2$=0.39) to about the same extent as fasting TG ($R^2$=0.43), both p<0.01. Univariate predictors of AUC were younger age ($R^2$=0.43), greater proteinuria ($R^2$=0.47) and lower serum albumin (Alb) ($R^2$=0.47), all p<0.01. The best 2-variable models predicting AUC were a) eGFR and Alb ($R^2$=0.66) b) uPCR and Alb ($R^2$=0.64), and c) age and Alb ($R^2$=0.79), all p<0.01. In each model, both variables were significant after adjustment for the other. Limited multivariable analysis suggested no improvement with >2 variables. While fasting TG was a dominant predictor ($R^2$=0.80, p<0.01) of AUC, AUC of TG excursion above the fasting level was also associated with the same variables and models (all p≤0.01). TG measured early (≤4 hours), but not later (≥6 hours) was associated with CKD variables to about the same extent as fasting TG.
**Conclusions:** Increasing degree of CKD and albumin variation within the normal range are independently associated with impaired post-prandial TG clearance. Overlapping or linked mechanisms contribute to fasting and post-prandial hypertriglyceridemia. Pending analysis of stored samples may discern effects of potential modulators such as insulin and ApoC-III.
**DISCLOSURE:** *Ginsberg, H.*: Grant/Research Support, Merck, Roche, AstraZeneca, Isis/Genzyme; Consultant, Merck, Aboot, Roche, AstraZeneca, BMS, Isis/Genzyme, Glaxosmithkline, Pfizer, Regeneron/Sanofi-Aventis; Speaker's Bureau, Pfizer; Other, Advisory Board: Merck/Schering Plugh, BMS/AstraZeneca.

## Abstract# 827

**Influence of Waist Circumference in Ambulatory Blood Pressure Monitoring of Overweight Children and Adolescents** J.C. Viana, C.M. Salgado, P.C.B.V. Jardim, T.S.B.V. Jardim. *School of Medicine, Federal University of Goiás, Goiânia, Goiás, Brazil; Department of Pediatrics and Hypertension League, Federal University of Goiás, Goiânia, Goiás, Brazil; Hypertension League, Federal University of Goiás, Goiânia, Goiás, Brazil; Hypertension League, Federal University of Goiás, Goiânia, Goiás, Brazil.*
**Objectives:** Comparing BP measurements using Ambulatory Blood Pressure Monitoring (ABPM) in overweight children and adolescents with and without increased waist circumference (WC).
**Methods:** Individuoals between 5 and 15 years of age were evaluated. Exclusion criteria: arm circumference >30 cm, diagnosis of secondary hypertension. A validated oscillometric method was used for the ambulatory BP measurements (SPACELABS 90207). Individuals with a BMI percentile ≥85 were divided into groups, according to cut-off points for WC. One group presented with a normal WC and the other with an increased WC.
**Results:** The study evaluated 46 children and adolescents with excess weight (9.09±2.89 years, 58.7% males and 58.6% non white). When comparing individuals from the normal WC and increased WC groups, no differences were found regarding weight, height, BMI, awake BP (p>0.05). Individuals with increased WC had higher night systolic BP (p=0.130) and diastolic BP (p=0.039) and smaller dipping (%) in systolic (p=0.003) and diastolic (p=0.0026) BP during sleep period.
**Conclusions:** In overweight children and adolescents, increased waist circumference is associated to a higher diastolic BP during sleep and lower night dipping of the systolic BP and diastolic BP. These changes in BP could be another marker for cardiovascular risk, in addition to the excess weight associated to an increased waist circumference.

## Abstract# 828

**Cardiovascular Morbidity in Dutch Children with ESRD** N.J. Schoenmaker,[1] I.M. Kuipers,[2] J.H. van der Lee,[3] J.W. Groothoff,[1] on behalf of the RICH-Q working group. *[1]Paediatric Nephrology, AMC/EKZ, Amsterdam, Netherlands; [2]Paediatric Cardiology, AMC/EKZ, Amsterdam, Netherlands; [3]Paediatric Clinical Epidemiology, AMC/EKZ, Amsterdam, Netherlands.*
**Objectives:** Cardiovascular disease is the main cause of death in patients with end stage renal disease (ESRD) since childhood. The RICH-Q study is a collaborative project in which all Dutch centres that provide paediatric chronic Renal Replacement Therapy (cRRT) collaborate to improve the quality of care. Here we report on factors associated with left ventricular hypertrophy (LVH).
**Methods:** All dialysis and transplanted Dutch patients aged <19 years between August 2007 and 2010 were included. Cardiovascular indicators like blood pressure, serum hemoglobin, phosphate and PTH were assessed every 3 months. Echocardiograms were performed once and were assessed by 2 observers centrally.
**Results:** 92 children were included. LVH was diagnosed in 20 children (22%). In 75%, LVH was asymmetrical septal. No significant differences were found in age, (median [range] age 15 [1-19] vs. 12 [0-18] years), duration of cRRT (median [range] duration of 2 [0-15] vs. 1 [0-15] years), gender, BMI, blood pressure, use of antihypertensive medication, serum hemoglobin, phosphate, PTH or treatment modality.
The first cRRT of the children with LVH vs. non LVH had been hemodialysis (HD) in 35% vs. 42%, peritoneal dialysis (PD) in 55% vs. 47% and transplantation (Tx) in 10% vs. 11%. The cRRT at time of the echocardiogram for LVH vs. non LVH was HD in 25% vs. 21%, PD in 45% vs.28% and Tx in 30% vs. 51%.
**Conclusions:** LVH, mostly asymmetrical septal LVH, is seen in 22% of the children with cRRT. This is considered to be a result of chronic volume overload.

## Abstract# 829

**Agreement between Cardiologists Diagnosing LVH in Children with ESRD Is Low** N.J. Schoenmaker,[1] I.M. Kuipers,[2] J.H. van der Lee,[3] J.W. Groothoff.[1] *[1]Paediatric Nephrology, EKZ/AMC, Amsterdam, Netherlands; [2]Paediatric Cardiology, EKZ/AMC, Amsterdam, Netherlands; [3]Paediatric Clinical Epidemiology, EKZ/AMC, Amsterdam, Netherlands.*
**Objectives:** Early detection and therapeutic intervention of cardiovascular disease is considered to be essential in children with end stage renal disease (ESRD) in order to prevent mortality in early adulthood.
Early signs of left ventricular hypertrophy (LVH) include interventricular septal (IVS) and posterior wall (PW) hypertrophy. The incidence of LVH appears to differ between different centres. The aim of this study was to assess the agreement between different observers of echocardiography with respect to the diagnosis of LVH.
**Methods:** We reviewed the echocardiograms, made by cardiologists from 4 different centres between August 2007 and 2010, of children aged 0-18 years with ESRD. Digital images that had already been analyzed at each centre were stored and analyzed offline by two independent observers. Kappa was calculated for 3 pairs of observers.
**Results:** In total 92 echocardiograms were reviewed. The cardiologists from the different centres diagnosed LVH in 28 children (30%), observer 1 in 20 children (22%), and observer 2 in 27 children (29%). All observers agreed on 8 children (9%) diagnosed with LVH, and on 46 children (50%) without LVH. Kappa ranged from 0.2-0.4, which is considered low.
**Conclusions:** There is limited agreement between cardiologists about the diagnosis of LVH in children with ESRD based on a single echocardiography. Research is needed to evaluate the use of repeated measurements over time.

## Abstract# 830

**The Early Cardiovascular Risk Markers in Primary Hypertensive Children** B. Sozeri,[1] O.D. Kara,[1] N. Dincel,[1] M. Deveci,[2] S. Mir.[1] *[1]Pediatric Nephology, Ege University Faculty of Medicine, Izmir, Turkey; [2]Pediatic Cardiology, Ege University Faculty of Medicine, Izmir, Turkey.*
**Objectives:** Hypertension impacts the vascular wall. Vascular morphology and function such as carotid artery intima-media thickness (cIMT), pulse wave velocity (PWV) and the pulse wave augmentation index (AIx) have been established as valid surrogate markers of arteriopathy in adults. We aimed to investigate these markers in vascular system showing cardiovascular damage before symptomatic period in hypertensive children.
**Methods:** Thirty-eight patients(28 male, 10 female) with primary hypertension and 15 healthy children were studied. cIMT(morphologic) was measured by 2-D ultrasonography. Radial and femoral PWV and AIx(functional ) were also investigated by vicorder system.
**Results:** The mean age was 12.7± 3.46 years. 20 of 38 patients were obese(BMI> 26.8). cIMTwas higher in patients(0.46 ±0.06 mm vs 0.35 ± 0.12 mm, respectively, P =0,01). Femoral PWV was higher in patients (5.7 ± 0.87 m/s, 4.1 ± 0.72 m/s, P =0.01). AIx was higher in patients(83.3 ± 6.6%,72.8 ± 7.4%,P =0.04).

There was a significant correlation between the higher PVW and AIx values and obesity(OR 2.2, p =0.01andOR 3.1, p =0.00, respectively). Also cIMT was higher in obese patients than others(p< 0.05).
**Conclusions:** We found that; in primary hypertensive child, the cardiovasculary risk markers are increased. Also additional obesity may aggregate this damage. In the light of these data, we suggest that simultaneously being of hypertension and obesity; increases vascular risk factors in children. Early determination of these parameters will lead us to know this risk group earlier and to prevent cardiovascular damage in adulthood.

## Abstract# 831

**Arterial Stiffness in Normotensive and Hypertensive Children** A. Stelcar, N. Marcun Varda, M. Miksic, A. Gregoric. *Pediatrics, University Medical Centre, Maribor, Slovenia.*
**Objectives:** The important part of investigations in hypertensive children is to establish the effects of elevated blood pressure on the hypertensive target organs. One potential early marker of this damage is the increase of arterial stiffness now recognized as a major driver of cardiovascular disease.
The aim of our study was to investigate if there are any differences between normotensive and hypertensive children according to the measures of arterial stiffness.
**Methods:** Twenty normotensive and 20 hypertensive children of both sexes, matched by age and sex, were included in the study. Carotid-radial pulse wave velocity (PWV) was measured in hypertensive children as an index of arterial stiffness using the applanation tonometry-SphygmoCor and compared to the values in normotensive children. Central aortic systolic pressure (AoSP), aortic pulse pressure (AoPP), augmentation pressure (AP) and the augmentation index (AIx@HR75) were also compared between the two groups.
**Results:** The hypertensives presented higher mean values of PWV (6.30 m/s), AoSP (131.9 mmHg), AP (0.35 mmHg) and the AIx@HR75 (2.80) than the controls (PWV (6.09 m/s), AoSP (125.5 mmHg), AP (-2.30 mmHg) and the AIx@ HR75 (-0.75)). The AoPP was higher in the normotensive group (55.9 vs. 51.1 mmHg). No significant differences in PWV (p= 0.57), AoSP (p=0.24), AP (p= 0.37), the AIx@HR75 (p=0.34) and AoPP (p=0.26) were found.
**Conclusions:** We found no significant differences of arterial stiffness between normotensive and hypertensive children. Nevertheless, we believe that with increasing size of studied population the difference would become more significant.

## Abstract# 832

**Melatonin Protects Hypertension and ADMA Accumulation in Young Spontaneous Hypertensive Rats** Y.-L. Tain,[1,2] L.-T. Huang,[1,2] J.-F. Hong,[1,2] I.-C. Lin.[1,2] *[1]Pediatrics, Chang Gung Memorial Hospital-Kaohsiung Medical Center, Kaohsiung, Taiwan; [2]Chang Gung University, College of Medicine, Kaohsiung, Taiwan.*
**Objectives:** Hypertension (HTN) is the major risk factor for the development of kidney disease. We investigated whether increased asymmetric dimethylarginine (ADMA, a NOS inhibitor) is involved in the development of HTN and melatonin can prevent it.
**Methods:** Using spontaneous hypertensive rat (SHR), a genetically hypertensive model, four groups (N=6/group) of 4-week-old SHR and their control WKY rats were sacrificed after 8-week treatment: Group 1, SHR without treatment; Group 2, SHR received 0.01% melatonin in drinking water (SHR+M); Group 3, WKY; and Group 4, WKY+M. ADMA levels and ADMA-related enzymes were determined.
**Results:** At 12 weeks of age, SBP of SHR were significantly higher than those in age-matched WKY rats (197±4 vs 157±3 mmHg), which were prevented by melatonin therapy (165±5 mmHg). We found plasma ADMA levels were higher in SHR than those in WKY from 4 wk (0.88±0.06 vs 0.66±0.01μM) to 12 wk of age (1.00±0.04 vs 0.63±0.03μM, P<0.05). In contrast, plasma L-arginine to ADMA ratio (AAR) was significantly decreased in SHR vs WKY at 4wk (153±17 vs 201±13μM/μM) and 12 wk of age (105±3 vs 199±8μM/μM). Increased ADMA concentrations in 12 wk-old SHR were prevented by melatonin (0.74±0.01μM), while melatonin significantly increased AAR (168±6μM/μM). In addition, data regarding ADMA-related enzymes in the kidney will be analyzed and presented.
**Conclusions:** High levels of ADMA precede hypertension. At 12 wk of age, SHR rats developed hypertension, and this was associated with increased ADMA and decreased AAR. Melatonin might decrease ADMA and restore AAR to prevent hypertension in young SHR rats.

## Abstract# 833

**Arterial Structure and Function in Pediatric Dialysis and Transplant Patients** H.K. Tawadrous,[1] K. Haroon,[2] A. Mongia,[1] L. Salciccioli,[2] M.J. Schoeneman,[1] J. Lazar.[2] *[1]Pediatric Nephrology, SUNY Downstate Medical Center, Brooklyn, NY, United States; [2]Cardiology, SUNY Downstate Medical Center, Brookly, NY, United States.*
**Objectives:** Higher augmentation index (AI), greater carotid intima-media thickness (CIMT) and endothelial dysfunction are present in patients (pts) with end stage renal disease and are predictive of cardiovascular morbidity and mortality. This is the first study whose objective was to compare these three indices of arterial integrity in children on dialysis(D) and after renal transplant ( Tx).
**Methods:** We studied 29 pts: 15 on D, 14 transplanted: 6 living related (LR) and 8 deceased donor ( DD), 15 healthy age, gender and ethnicity matched controls (C). CIMT and artery flow-mediated increase in arterial diameter (FMD) were measured by high-resolution ultrasound, augmentation index (AI) by applanation tonometry. Mean values between groups were compared using pairwise comparisons.
**Results:** AI (16±12, 6±11, 5±10) and CIMT (0.5±0.05, 0.49±0.05, 0.46±0.02) were significantly higher in the D group than in the Tx and C group (both p< 0.05). FMD was lower in D pts than tx and C (9 ±4, 11±4, 17 ± 5, p<0.002). CIMT and AI were similar between Tx and C. However, FMD remained lower in Tx vs C, p<0.001. Among the Tx group, AI was significantly lower in LRD than DDRT p=0.03and was significantly correlated with Hgb level (r= -0.35, p=0.05). There were no significant correlations between CIMT, AI, and FMD and the duration of D or time after Tx.
**Conclusions:** Pediatric Tx pts have fewer arterial abnormalities than D pts. While CIMT and AI were similar to C, FMD remained significantly impaired in Tx pts. LR may confer a more favorable arterial profile than DD.

## Abstract# 834

**Life Style Modifications (LSM) Have Limited Efficacy on Improvement of Blood Pressure and Cardiovascular (CVD) Risk Factors in Children with Hypertension** H.K. Tawadrous, S. Mongia, R. Gorgy, M. Schoeneman, A.K. Mongia. *Pediatric Nephrology, SUNY DOwnstate Medical Center, Broklyn, NY, United States.*
**Objectives:** Evidence of LSM for blood pressure (BP) reduction in children is limited.We determined effect on BP and CVD risk factors in children with HTN who received LSM alone or with medication.
**Methods:** Retrospective review, 87pts with new-onset HTN from July 02toOct09. Pts recieved LSM and medications were added in Stg1and 2HTN as needed. We used paired t-test to evaluate changes in each group in respect to BP, BMI, Chol, LDL, TG, HDL and microalbumin MA.
**Results:** Mean follow up was 2.1 ys.66% boys,83% AA, 51% were overweight.18% had pre hypertension (PH),38% stage 1 and 44% stg 2 HTN. At baseline, PH, stg 1and stg 2 HTN did not differ significantly as per age, sex, race, BMI, LDL,Chol, TG, HDL and MA. There was a significant improvement of systolic BP(SBP) in stg1and SBP and diastolic BP(DBP) in stg 2.Both SBP and DBP increased in pts with PH(Table). There was no significant improvement in, BMI, MA, Chol, LDL, TG, and HDL in any of the groups.

BP initial and F/U

| | SBPinitial (Mean ± SD) | SBP F/u (Mean ± SD) | p value | DBP initial (Mean ± SD) | DBP F/U (Mean ± SD) | p value |
|---|---|---|---|---|---|---|
| PH | 121.81 ±12.93 | 123.43 ±11.95 | 0.5549 | 68.5 ±10.66 | 70.43 ±8.91 | 0.5187 |
| Stage1 | 130.62 ±9.46 | 124.93 ±10.70 | 0.0042 | 74.06 ±8.63 | 75.86 ±9.45 | 0.48 |
| Stage2 | 144.61 ±14.17 | 127.63 ±13.79 | <.0001 | 81.13 ±12.98 | 74.94 ±9.26 | 0.0293 |

**Conclusions:** LSM may not be adhered to intensely, may not be practical and may be inffective in children. Newer innovative behaviour modification techniques or early institution of drug therapy may provide better outcome.

## Abstract# 835

**Results of Surgical Treatment for Renovascular Hypertension in Children: 30 Year Single Centre Experience** M.B. Stadermann,[1] G. Montini,[1] G. Hamilton,[3] D.J. Roebuck,[2] C.A. Mclaren,[2] M.J. Dillon,[1] S.D. Marks,[1] K. Tullus.[1] *[1]Paediatric Nephrology, Great Ormond Street for Children NHS trust, London, United Kingdom; [2]Radiology, Great Ormond Street Hospital for Children NHS trust, London, United Kingdom; [3]Vascular Surgery, Royal Free Hospital, London, United Kingdom.*
**Objectives:** We retrospectively reviewed the medical records of all patients who underwent surgery as part of the treatment of renovascular hypertension (RVH) at our centre between 1979 and 2008.
**Methods:** Thirty-seven children (65% male) with a median age of 7.6 (0.4–17.9) years were identified with a median systolic blood pressure (SBP) of 140 (105–300) mm Hg prior to surgery. Bilateral renal artery stenosis and intra-renal disease were present in 19 (51%) patients, mid-aortic syndrome in 15 (40%) and coexisting cerebral disease in eight out of 30 (26%) investigated patients.
**Results:** Surgical procedures ($n$ = 53) included (i) nephrectomy (18), (ii) renovascular surgery on the renal arteries (28) (iii) aortic reconstruction (7). Post-operative complications were haemorrhage (5), septicaemia (5) and chylous

ascites (1). There were no perioperative deaths; two children died during follow-up. The SBP post-surgery improved to a median value of 116 (range 90–160) mm Hg. Twelve months after surgery, 16 (43%) children had normal blood pressure without treatment, 15 (41%) normal or improved on one to four antihypertensive drugs and four (11%) unchanged.
**Conclusions:** Surgery effectively treated the hypertension of 90% of our children, when performed in conjunction with medical therapy and interventional radiology. In spite of aggressive surgical treatment, RVH is sometimes a progressive disease.

## Abstract# 836

**Is Solitary Kidney a Condition at Risk for Hypertension? a Follow up Study in a Pediatric Population** P.G. Vercelloni, E. Meregalli, A. Edefonti, M. Castellon, G. Marra. *Pediatric Nephrology Unit, Fondazione Ca'Granda, Milano, Italy.*
**Objectives:** Assessment of hypertensive risk in a population of children with congenital solitary kidney (SK) followed perspectively for 20 years.
**Methods:** We compared a population of 42 pts with SK with a control group (CK) of 46 pts with congenital uropathy (mild vescico-ureteral reflux, hydronephrosis) all with normal renal size and function. The groups were selected from a wide population of pts with congenital uropathy according to the following criteria: normal renal parenchyma as stated by US and renal scan, creatinine cl > 90 ml/min/1,73 mq, follow up longer than ten years. Age, BMI,proteinuria,sex,creatinine were comparable in the two groups. Blood pressure was measured at every visit and ABPM every 4 years or more often in case of hypertension.
**Results:** The percentage of hypertensive children was significantly higher in the SK than CK( 16.7 vs 6.5%; p-value 0,043 with $\chi$ sq test). All ABPM indices expressed as SD, except one,were significantly higher in the SK pts than in CK was significantly higher in the SK group.

| | SK mean±SD | NKmean±SD | p-value |
|---|---|---|---|
| 24 h DBP | 0.16±0.87 | -024±0.70 | 0.0178 |
| 24 h SBP | 0.56±1.19 | -058±0.76 | 0.0040 |
| day DBP | -0.27±0.79 | -057±0.72 | 0.0687 (NS) |
| day SDP | 0.17±1.03 | -0.26±0.86 | 0.0355 |
| night DBP | 0.58±0.82 | 0.13±0.77 | 0.0096 |
| night SBP | 1.03±1.24 | 0.44±0.84 | 0.0105 |

**Conclusions:** Solitary kidney represents a condition at risk for developing hypertension since childhood. Therefore blood pressure monitoring is mandatory in this population.

## Abstract# 837

**Blood Pressure in Juvenile Systemic Lupus Erythematosus Changes with Overall Disease Activity** L Watson,[1] C. Jones,[2] R.C.L. Holt,[2] M.W. Beresford.[1] *[1]Institute of Child Health, University of Liverpool, Liverpool, United Kingdom; [2]Paediatric Nephrology, AlderHey Children's NHS Foundation Trust, Liverpool, United Kingdom.*
**Objectives:** Juvenile-onset Systemic Lupus Erythematosus (JSLE) is a chronic autoimmune condition affecting children less than 17 years. A high proportion of children develop renal involvement [1]. Hypertension is linked to a worse renal prognosis [2]. We aimed to determine how systemic blood pressure changes with disease activity in JSLE.
**Methods:** Patients with JSLE, treated at Alder Hey Children's NHS Foundation Trust Hospital (2000-2009) were included. Informed consent and ethical approval was obtained. Standardised data forms were used to collect demographic and laboratory parameters. Blood pressure (BP) was converted to a standard deviation score (SDS) [3]. Renal parameters included GFR, urine albumin:creatinine ratio (UACR), renal biopsy findings and medication. An overall BILAG score and a renal BILAG category for each clinical episode was produced [4].
**Results:** Thirty children were recruited; 281 clinical episodes were captured. 10 were male. Median age 13.3 years (range 3.7-17.9). 73% were Caucasian. 21 episodes were hypertensive. BP SD scores correlated with overall disease activity (BILAG score), p=0.042; renal BILAG, p<0.01; corticosteroid dose (mg/kg), p=0.048 and if a renal biopsy had been performed, p=0.008. BP SDS didn't correlate with UACR.
**Conclusions:** Changes in blood pressure in JSLE are related to overall disease activity together with corticosteroid dose.The effect of systemic vascular inflammation, renal involvement and corticosteroid use may all be contributing to changes in blood pressure. Separating these variables may prove complex.

## Abstract# 838

**Prevalence of Prehypertension and Hypertension in an Asian Pediatric Population** C.-M. Quek,[1] D.T. Bautista,[2] E.S. Chan,[2] I.D. Liu,[1] H.K. Yap.[1] *[1]Pediatrics, Yong Loo Lin School of Medicine, National University Health System, Singapore, Singapore; [2]Singapore Clinical Research Institute, Singapore, Singapore.*
**Objectives:** Pediatric blood pressure (BP) norms vary in different populations and are yet to be established in multi-ethnic Singapore, comprising 74.7% Chinese, 13.6% Malay and 8.9% Indian.
**Methods:** A cross-sectional study was conducted to establish office BP norms in healthy Singapore school children. BP readings were measured following a standard protocol in 9936 children aged 6-19 years using the mercury sphygmomanometer, and the second and third readings for each child were analyzed. Blood pressure norms were constructed from a subset of children (n=7824, 3725 boys and 4099 girls) with normal weight, using restricted cubic spline quantile regression. Children with medical conditions or on drugs that could affect BP were excluded (n=84). BP normograms were established according to sex, age and height percentiles, based on international definitions for weight and height categories.
**Results:** The percentage of prehypertensive and hypertensive children by weight category is shown in the table below.

| | Normal Weight (n=7824) | Overweight/Obese (n=2028) |
|---|---|---|
| | No. (%) | No. (%) |
| Prehypertensive | 752 (10%) | 283 (14%) |
| Hypertensive | 699 (8.9%) | 316 (15.6%) |

Based on our normograms, overweight or obese children were 1.5 times more likely to be pre-hypertensive (relative risk 95% CI: 1.28, 1.65) and 1.7 times more likely to be hypertensive (relative risk 95% CI: 1.54, 1.97) than children with normal weight.
**Conclusions:** There is a fairly high proportion of prehypertensive and hypertensive children in our multi-ethnic population, and this risk increases significantly if the child is overweight.

## Abstract# 839

**Microalbuminuria and Elevated Blood Pressure in Obese Children** M. Zaniew,[1,3] B. Skowronska,[2] W. Stankiewicz,[2] A. Blumczynski,[3] P. Fichna,[2] J. Zachwieja.[3] *[1]Department of Pediatric Nephrology, District Children's Hospital, Szczecin, Poland; [2]Department of Pediatric Endocrinology and Diabetes, University of Medical Sciences, Poznan, Poland; [3]Department of Pediatric Nephrology, University of Medical Sciences, Poznan, Poland.*
**Objectives:** The objective of this study was to investigate albuminuria in relation to blood pressure (BP) level and other elements of metabolic syndrome (MS) in obese children.
**Methods:** In 50 children (mean age:13.5±2.8 yrs) with obesity, antropometrical data, lipid profile, fasting glucose, indices of insulin resistance [HOMA-IR, FIGR (fasting insulin to glucose ratio)], 24-h urinary albumin excretion (UAE), casual and ambulatory systolic BP (SBP) and diastolic BP (DBP) were analyzed.
**Results:** The prevalance of hypertension and MS was found in 32% and 40%, respectively. Twenty subjects (40%) fulfilled the criteria of MS. Mean UAE was 29.99±62.19 mg/24 h and mikroalbuminuria (MA) was present in 8 children (16%), of whom 6 had hypertension. In children with elevated BP, UAE was significantly higher in comparison to subjects with normal BP (p<0.01). Patients with MS and/or insulin resistance had similar UAE compared to those without these abnormalities. Children with UAE greater than median level had higher TG, HDL/TG ratio, uric acid, FIGR and a tendency for higher 24-h SBP (p=0.08) and SBP load (p=0.06). Univariate analysis showed correlations between UAE and 24-h SBP (r=0.27), SBP load (r=0.29), and SBP index (r=0.34). In a step-wise regression analysis, predictors for UAE were 24-h SBP and FIGR.
**Conclusions:** SBP is the strongest factor that influences UAE in obese children. Microalbuminuria could serve as an indicator of elevated BP in obese children.
**DISCLOSURE:** *Zaniew, M.*: Other, Subinvestigator in Clinical Trial - Salary. *Blumczynski, A.*: Other, Subinvestigator in Clinical Trail - Salary. *Zachwieja, J.*: Other, Subinvestigator in Clinical Trail - Salary.

## Abstract# 840

**Aortic Stiffness and Left Ventricular Hyperthrophy in Children with Chronic Kidney Disease** M. Zaniew,[1] D. Drozdz,[2] D. Runowski,[1] A. Blumczynski,[3] B. Jachimiak.[1] *[1]Department of Pediatric Nephrology, District Children's Hospital, Szczecin, Poland; [2]Department of Pediatric Nephrology, Jagiellonian University Medical College, Krakow, Poland; [3]Department of Pediatric Nephrology, University of Medical Sciences, Poznan, Poland.*
**Objectives:** To determine the association between aortic pulse wave velocity (PWV) and left ventricular mass index (LVMI) in children with chronic kidney disease (CKD).
**Methods:** Forty-six children (mean age: 13.5±3.1 yrs) with stages 2-5 CKD (GFR: 32±23 ml/min per 1.73m$^2$) were studied. LVMI was estimated by M-mode

echocardiography and left ventricular hyperthrophy (LVH) was defined as LVMI >95th percentile. Aortic PWV was determined by applanation tonometry. We used recently produced data of PWV by age from normal population to define abnormal PWV (ie >95% confidence interval).
**Results:** The prevalence of LVH and increased PWV was 43% and 48%, respectively. Subjects with increased PWV had higher LVMI (42.09±12.02 vs 33.79±7.86, p<0.01) and blood pressure (BP) than those with normal PWV. Similarly, children with LVH had greater PWV (6.0±0.99 vs 4.99±1.48, p<0.05) and BP than those without LVH. Higher PWV (p<0.001) and LVMI (p<0.01) were found in hypertensive children as compared to subjects with normal BP. PWV was positively correlated with BP and LVMI, while LVMI was associated with: height-SDS, BP, calcium, phosphorus, TG, HDL and PWV. After adjustment for all the above variables, diastolic BP (β=0.76) remained the only significant predictor for PWV and calcium (β=-0.42) and PWV (β=0.36) for LVMI.
**Conclusions:** These findings indicate that aortic stiffness measured by PWV could serve as an additional marker of increased cardiovascular risk in children with CKD.
**DISCLOSURE:** *Zaniew, M.*: Other, Subinvestigator in Clinical Trial - Salary. *Drozdz, D.*: Other, Subinvestigator in Clinical Trial - Salary. *Runowski, D.*: Other, Subinvestigator in Clinical Trial - Salary. *Blumczynski, A.*: Other, Subinvestigator in Clinical Trial - Salary. *Jachimiak, B.*: Other, Subinvestigator in Clinical Trial - Salary.

## Congenital Abnormalities

### Abstract# 841
**(O-105)**
**Matrix Metalloproteinase 9 and Tissue Inhibitor of Metalloproteinase 1 in Vesicoureteral Reflux** A. Yilmaz, I. Bilge, A. Kiyak, A. Gedikbasi, A. Sucu, B. Aksu, S. Emre, A. Sirin. *Pediatric Nephrology, Bakirkoy Maternity and Children Hospital, Istanbul, Turkey; Pediatric Nephrology, Istanbul University Istanbul Medical Faculty, Istanbul, Turkey; Biochemistry, Bakirkoy Dr. Sadi Konuk Training and Research Hospital, Istanbul, Turkey.*
**Objectives:** The aim of the study was to assess whether urine level of Matrix metalloproteinase 9 (uMMP9) and Tissue inhibitor of metalloproteinase 1 (uTIMP1) could represent novel markers of vesicoureteral reflux (VUR) and to determine the optimal cut-off level for these biomarkers to predict VUR in children.
**Methods:** Sixty seven patients with VUR and 20 healthy controls were enrolled the study. Urine MMP9 and TIMP1 were measured by enzyme-linked immunosorbent assay.
**Results:** Mean uMMP9 level was significantly higher in the VUR group than in the controls (1539,8pg/ml vs 256,4pg/ml, p= 0,0001) and using a cut-off 1054pg/ml for uMMP9 for diagnosis of VUR, sensitivity and specificity were 67.2% and 85%, respectively (AUC: 0.77). Mean uTIMP1 level was significantly higher in the VUR group than in the controls (182pg/ml vs 32.6pg/ml, p= 0,0001) and using a cut-off 18.7pg/ml for uTIMP1 for diagnosis of VUR, sensitivity and specificity were 74.6% and 65%, respectively (AUC: 0.73). Mean uTIMP1 level was also significantly higher in patients with high grade VUR than in those low grade VUR (927.4pg/ml vs 132.2pg/ml, p=0.012).
Mean uMMP9/creatinine and mean uTIMP1/creatinine ratios were also significantly higher in the VUR group.
**Conclusions:** Both uMMP9 and uTIMP1can be useful markers for identifing patients who have high risk for VUR to avoid unnecessary cystourethrografies.

### Abstract# 842
**(O-106)**
**Inhibition of mTor with Sirolimus Is Not Protective in PCK Rats** D.C. Fischer,[1] C. Renken,[1] G. Kundt,[2] N. Gretz,[3] D. Haffner.[1] *[1]Department of Pediatrics, University Children's Hospital Rostock, Rostock, Germany; [2]Department of Medical Informatics and Biometry, University of Rostock, Rostock, Germany; [3]Medical Research Center Medical Faculty Mannheim, University of Heidelberg, Mannheim, Germany.*
**Objectives:** To elucidate the therapeutic potential of sirolimus in PCK rats, an orthologous model of autosomal recessive polycystic kidney disease (ARPKD).
**Methods:** Weaned PCK- and SD rats (n=85 and 72) received drinking water without and with sirolimus (corresponding to a daily intake of 2 mg/kg body weight) for 4, 8, and 12 weeks, respectively. Renal and hepatic functions were monitored and both organs were investigated morphologically (fibrosis, number & size of cysts) and immunohistochemically (expression of Akt, mTOR, and S6K).
**Results:** Five out of 43 sirolimus treated PCK rats but none of the controls died during the study. Weight gain was slightly reduced in sirolimus treated rats. In PCK rats, grossly enlarged kidney and livers, hepatic fibrosis and enlarged bile ducts were noted. Whereas activation of Akt/mTOR signaling was hardly detectable in the kidneys of SD rats, strong signals were seen in the kidneys of PCK rats. Despite a significantly reduced relative kidney weight after 12 weeks of treatment (p<0.05), neither fibrosis and cyst area nor renal function improved during treatment. Sirolimus resulted in a minor inhibition of renal mTOR-specific phosphorylation of S6K only. Male PCK rats on sirolimus showed increased concentrations of bile acids and bilirubin compared to controls (each p<0.05 at 12 weeks). Similar, albeit none significant effects were noted in female PCK rats.
**Conclusions:** Sirolimus failed to attenuate progression of kidney and liver disease in PCK rats.

### Abstract# 843
**(O-107)**
**Long-Term Outcome of Bilateral Mild Isolated Antenatal Hydronephrosis** L.F. Alconcher, M.M. Tombesi, M.B. Meneguzzi. *Pediatric Nephrology Unit, Hospital Interzonal General de Agudos Dr Jose Penna, Bahia Blanca, Buenos Aires, Argentina.*
**Objectives:** To assess prevalence, outcome and incidence of urinary infection (UI) in newborns (NB) with bilateral mild isolated antenatal hydronephrosis (MIAH).
**Methods:** 491 NB with antenatal renal abnormalities were prospectively studied. Inclusion criteria**:** MIAH defined as antero-posterior pelvic diameter (APD) ≤ 15 mm confirmed by first postnatal ultrasound (US). Exclusion criteria: APD > 15mm, calyectasis, hydroureteronephrosis, renal or bladder abnormalities. US follow-up was performed. Neither voiding cystourethrography (VCUG) nor antibiotic prophylaxis (AP) were indicated. Parents were familiarized with the signs of UI. If UI was confirmed, VCUG was performed. The outcome was assessed as intrauterine involution (IUI), total involution (TI), partial involution (PI), stability (S) and progression (P). Outcome and incidence of UI was compared with NB with unilateral MIAH. Chi square test was applied.
**Results:** Of the 491 NB, 236 had MIAH (138 unilateral and 98 bilateral). Of the 138 NB with unilateral 38 (27%) showed IUI, 58 (42%) TI, 8 PI, 33 S and 1 P. Of the 98 NB with bilateral (196 renal units (RU)), 72 RU (37%) showed IUI, 82 (42%) TI, 12 PI, 26 S and 3 P. Mean follow-up was 17 and 14 months respectively. Fourteen patients with unilateral and 9 with bilateral MIAH had UI. Reflux was found in 2 patients with unilateral MIAH.
**Conclusions:** Bilateral MIAH represented 20% of the antenatal renal abnormalities. Outcome and incidence of UI were not statistically different between the two groups. NB with bilateral MIAH would have similar considerations as those with unilateral. Hence, we consider that neither AP nor VCUG are mandatory but follow-up is important.

### Abstract# 844
**(O-108)**
**Prevalence and Spectrum of Urinary Tract Abnormalities in Fetuses, Children and Adults with 22q11 Microdeletion** J. Harambat,[1] M.A. Delrue,[2] G. André,[3] B. Llanas,[1] D. Lacombe.[2] *[1]Pediatric Nephrology, University Hospital, Bordeaux, France; [2]Genetics, University Hospital, Bordeaux, France; [3]Pathology, University Hospital, Bordeaux, France.*
**Objectives:** The 22q11 deletion is the most frequent interstitial chromosomal aberration. This deletion is detected in various phenotypes including the majority of cases of DiGeorge syndrome, velocardiofacial syndrome and conotruncal anomaly face syndrome. We aim to describe the prevalence and clinical features of renal disease in 22q11 deletion.
**Methods:** All cases of 22q11 deletion, confirmed by fluorescent in situ hybridization at Bordeaux University Hospital, France, between 1995 and 2009 were reviewed.
**Results:** We identified 68 patients (43 males) from 58 families, and 15 fetuses. Five fetuses (33%) had renal malformations: 3 had multicystic kidney disease, 1 had renal agenesis, and 1 had bilateral pelvic dilatation. In patients with postnatal diagnosis, median age at diagnosis was 8.7 years (range 1 months-42 years). Main clinical features were congenital heart disease (61%), facial dysmorphism (92%), mental retardation (89%), velopharyngeal insufficiency (58%), growth retardation (50%), and hypocalcemia (18%). Five patients (7%) died. Among the 51 patients with documented renal exploration, 9 (18%) had urinary tract malformations including unilateral renal agenesis (2), obstructive megaureter (1), pelviureteral junction (2), and vesicoureteral reflux (4). Six patients had episodes of pyelonephritis, and 3 developed chronic kidney disease.
**Conclusions:** 22q11 deletion syndrome should be considered in patients with urinary tract abnormalities and additional signs such as dysmorphia, abnormal voice, or cardiac malformations.

## Abstract# 845

**(O-109)**

**Proteinuria as a Marker of Severe Unilateral Ureteral Obstruction in Children** O. Mansoor,[1] M. Sasso,[1] C.L. Abitbol,[1] G. Sfakianakis,[2] R. Gosalbez,[3] W. Seeherunvong,[1] J. Chandar,[1] G. Zilleruelo.[1] *[1]Pediatric Nephrology, University of Miami/Holtz Children's Hospital, Miami, FL, United States; [2]Nuclear Medicine, University of Miami, Miami, FL, United States; [3]Pediatric Urology, University of Miami/Miami Children's Hospital, Miami, FL, United States.*

**Objectives:** To examine radiologic and laboratory parameters of unilateral ureteral obstruction (UUO) and compare surgical intervention versus conservative management.

**Methods:** A retrospective analysis was performed on 106 children with high grade UUO. Proteinuria was assessed by random urine total protein to creatinine ratio (Upr/cr) (Normal <0.2 mg/mg). Albuminuria was assessed by random urine albumin to creatinine ratio (Ualb/cr) (Normal <30 mcg/mg). Renal pelvic diameter (RPD) was measured by digital real time ultrasonography in centimeters (cm). Renal scintigraphy measured differential renal function in percent.

**Results:** In the cohort 43/106 (41%) required surgical intervention. Current age is 5.5±3.1 years and there is a male predominance (73%). There was also a predominance of left kidney obstruction (67%). The table compares the RPD, Upr/cr and Ualb/cr for surgical (Sgy) versus the non-surgical (No-Sgy) patients:

| Group | N | RPD | Upr/cr | Ualb/cr | Split RF |
|---|---|---|---|---|---|
| Sgy | 17 | 2.9±1.2 | 0.7±0.4 | 191±172 | 44.7±14.4 |
| No-Sgy | 31 | 1.0±0.8 | 0.4±0.3 | 80±72 | 50.3±4.6 |
| p-Value | | <0.001 | 0.01 | <0.01 | 0.04 |

**Conclusions:** Both proteinuria and albuminuria distinquished those patients with UUO who required surgical intervention. This suggests that proteinuria is a sensitive marker of renal injury in patients with UUO.

## Abstract# 846

**Nocturnal Bladder Distention and Treatment in Myelodisplastic Children Who Performs Daytime Clean Intermittant Catheterisation** I. Akil, C. Taneli, O. Yilmaz, K. Genc. *Pediatric Nephrology, Celal Bayar University, Manisa, Turkey; Pediatric Surgery, Celal Bayar University, ManisA, Turkey; Pediatric Surgery, Celal Bayar University, Manisa, Turkey; Pediatric Surgery, Celal Bayar University, Manisa, Turkey.*

**Objectives:** Clean intermittent catheterization (CIC) treatment is a classical and successful method in resolving urinary retention and intravesical high pressure. In some cases, on daytime CIC protocol alone, recurring urinary infections, advancing incontinence, renal scarring and advancing hydronephrosis may be seen due to over distended bladder during night sleep.

**Methods:** In the present study, in 12 cases on CIC, with myelodisplasia and neurogenic bladder dysfunction, urinary osmolarity and nocturnal bladder capacity were measured. The urine volume acquired by CIC was measured in the evaluation of the bladder capacity.

**Results:** In 5 cases, nocturnal accidental voiding was seen between the last and the first ICI. These patients present an interval of 10-12 hours between the last CIC performed before bedtime and the first CIC in the morning. After adjusting the CIC and after an 8-hour sleep if the case awakes dry and the urine volume at first ICI is considered the nocturnal bladder capacity. After such an evaluation in 6 cases, a midnight CIC is suggested. Generally, CIC only during daytime is not found adequate.

**Conclusions:** In children with neurogenic bladder dysfunction, although under anticholinergic therapy and on CIC if urinary infections and bladder dysfunction ensue, nocturnal measurements of bladder capacity and if needed nocturnal bladder drainage should be instituted.

## Abstract# 847

**Height Growth in Children with Vesicoureteral Reflux and Renal Scarring** H. Alpay,[1] B. Ören,[1] M. Benzer,[1] Z. Atay,[2] T. Tarcan,[3] I. Gökçe.[1] *[1]Ped Nephrol Dept, MU Med Faculty, Ist, Turkey; [2]Ped Endocr Dept, MU, Ist, Turkey; [3]Urology, MU, Ist, Turkey.*

**Objectives:** The aim of this study was to evaluate the height growth in children with VUR and renal parenchymal scarring (RPS).

**Methods:** Eighty-five children with VUR were evaluated with respect to the clinical and radiological findings. Seventy-three age and sex matched children from pediatric outpatient clinics for mild upper respiratory problems served as control group. Height Z scores (HZ) and mean parental height values (MPH HZ) were calculated for patients with VUR and/or RPS and HZscores of the control group was calculated.

**Results:** DMSA scanning revealed RPS in 46 of 85 children with VUR.In 57 children, VUR was diagnosed before 1 y of age. HZ score means and standart deviations (SD) were 0.375±1.23 for VUR+ patients, 0.388±1.23 for control group and MPH HZ of VUR+ patients was - 0,269±0.95. When HZ values of VUR+ and RPS + group was compared with control there was no significant difference.

Table 1. HZ and MPH HZ of VUR+ and RPS+ patients

| Patients | HZ | MPH HZ | P |
|---|---|---|---|
| VUR+ patients (n: 85) | 0.375±1.23 | | |
| Control group (n:73) | 0.388±1.23 | - 0,269±0.95 | 0.008 |
| P | NS | | |
| RPS+ patients (n: 46) | 0.302±1.38 | | |
| Control group (n:73) | 0.388±1.23 | - 0,234±0.95 | 0.03 |
| P | NS | | |

MPH: Mean parental height, VUR: Vesicoureteral reflux, RPS: Renal parenchymal scarring, NS: Not significant

Surprisingly when HZ values of VUR+ and RPS+ groups was compared with their MPH HZ, there was statisticaly significant difference in favor of VUR+ group. There was no correlation between VUR degrees, duration of VUR and HZ values of patients.

**Conclusions:** Our study shows that VUR does not affect height growth in children.

## Abstract# 848

**The Effect of Ambulatory Status of Children with Meningomyelocele on Clinical Findings and Renal Outcome** M. Benzer,[1] H. Alpay,[1] I. Gökçe,[1] T. Tarcan,[2] N. Biyikli,[1] A. Özsen.[1] *[1]Ped Nephrol, MU Med Faculty, Ist, Turkey; [2]Urology, MU, Ist, Turkey.*

**Objectives:** Meningomyelocele (MMC) is a common neural development disorder. The objective of this study was to evaluate the influence of ambulatory status of children with MMC on clinical findings and renal outcome.

**Methods:** The records of 95 children with MMC followed up at least a year from 2005 to 2010 were reviewed retrospectively. Ambulatory status was classified as independent walkers (walks without assistive appliances), assisted walkers (requires walking aid), and non-ambulatory (wheelchair bound). Demographic, clinical and radiological characteristics of the patients were evaluated according to the ambulatory status.

**Results:** Ninety-five patients (53 boys and 42 girls) were enrolled to the study. The median follow-up of was 55 months ( 12 to199 months). Forty-six patients (49.5 %) were operated during the first three days of life. Ninety-two (97.9%) had lomber, 3 had (3.3%) thoracal defects. The technetium dimercaptosuccinic acid (DMSA) scan showed unilateral renal parenchymal scarring (RPS) in fourteen (21.5%) patients and bilateral RPS in 6 (9.2%) patients. Eighteen patients (26.9%) had vesicoureteral reflux (VUR)**.** Seventy-three children (76.8%) were non-ambulatory, 14 of the children (14.7%) were assisted walkers and 8 of them (8.4%) were independent walkers. Patients whose follow-ups started at an early age showed less hydronephrosis at USG and deformed bladder at VCUG. Ambulatory status was not found to have influence on operation time, frequent UTI, hydronephrosis, VUR, renal parenchimal scarring of the patients.

**Conclusions:** Our data shows that ambulatory status of children with MMC does not affect the clinical findings and renal outcome.

## Abstract# 849

**Audit of Posterior Urethral Valves at Red Cross Children's Hospital, South Africa** S. Antwi,[1] M. McCulloch,[2] P. Gajjar,[2] P. Nourse.[2] *[1]Dept of Child Health, School of Medical Sciences, Kwame Nkrumah University of Science and Technology/Komfo Anokye Teaching Hospital, Kumasi, Ghana; [2]Paediatric Nephrology Unit, Red Cross Children's Hospital, Cape Town, South Africa.*

**Objectives:** To determine the clinical presentation, timing of diagnosis, timing/type of surgical intervention, and disease progression among children with PUV at RXCH from January 2002 to January 2009.

**Methods:** A retrospective, folder review.

**Results:** 48 patients were reviewed. Median duration of follow-up: 52 months (IQR 24-107 months). 19 (39.6%) subjects diagnosed prenatally. Age at postnatal diagnosis: 1day to 2,950 days (median 17, IQR 1.0-90.5 days). 17 (58.6%) subjects diagnosed within first 3 months of age.

Commonest clinical presentations: UTI (10), palpable abdominal mass (4), voiding dysfunction (3), enuresis (2).

Surgical procedures employed: valve ablation alone 29 (60.4%), vesicostomy alone 12 (25.0%), cutaneous ureterostomies in combination with other surgeries 7 (14.5%) patients.

Median time from confirmation to primary surgery was 8.5 days (IQR 6-15days). At end of follow-up, GFR could be estimated for 37 patients, 20 (54.1%) of which were in stage 1 CKD. Only 1 (2.7%) had reached ESRF over 5.25 years; he has undergone successful renal transplant with functioning graft after 4 years 2 months (serum creatinine 48 µmol/l). No death recorded but significant numbers (29%) lost to follow up. Prognostic factors identified were serum creatinine at presentation and at 1 year of age, nadir serum creatinine 1-year post surgery.

**Conclusions:** PUV could be successfully managed in Africa to decrease disease progression to ESRF.

## Abstract# 850

**Congenital Malformations of the Urinary System in Fetuses and Deceased Newborns** M.I. Appasova. *Department of Pediatrie, Kazakh National Medical University, Tole Bi, Almaty, Kazakhstan.*

**Objectives:** The objective of the study was to perform a comparative analysis of the incidence and structure of congenital malformations of the urinary system in fetuses and deceased newborns in Almaty.

**Methods:** It was analysed 1014 protocols of autopsies on aborted fetuses and 1868 deceased newborns that died for three years in Almaty.

**Results:** The quantity in fetuses with congenital malformations of the urinary system from 1014 has made systems 17 (1,67%). In all 17 cases the basis for pregnancy interruption were prenatal the revealed anomalies of development. From 1868 died of children the quantity of newborns with developmental anomalies has made 46 (2,46 %). It was determined that the number of the newborns with congenital malformations of the urinary system significantly exceeds the number of the induced aborted fetuses. Among the most frequently diagnosed intrauterine malformations are hydronephrosis(50 %), aplasia and renal agenesis (16 %). At the died newborns numerous congenital malformations (40 %) and the combined congenital developmental anomalies urinary systems (27 %) more often were registered. Numerous congenital malformations represented a combination of congenital defects urinal system and congenital defects of gastroenteric system, cardiovascular system. In 2 cases congenital defects urinary systems have been diagnosed for children with defects of nervous system.

**Conclusions:** Among the deceased newborns it has been revealed frequently registered numerous congenital malformations and combined congenital breaches of the development of the urinary system that suggests an insufficient prenatal diagnostics of congenital malformations of the urinary system.

## Abstract# 851

**Pregnancy and Neonatal-Postneonatal Outcome in Maternal Hypokalemic Disoders: A Systematic Review of the Literature and a Case Series** L. Mascetti,[1] A. Bettinelli,[1] G.D. Simonetti,[2] F. Tammaro,[1] A. Tagliabue,[1] M.G. Bianchetti.[3] *[1]Pediatrics, Mandic Hospital, Merate, Italy; [2]Pediatrics, University, Berne, Switzerland; [3]Pediatrics, Hospital, Mendrisio-Bellinzona, Switzerland.*

**Objectives:** The term inherited hypokalemic disorder involves a set of renal-tubular diseases that present with normal blood pressure, hyperreninemia and hypokalemia. The management has improved over the last years and the issue of pregnancy has become important for the patients. Therefore, we extensively review reported information on pregnancies and neonatal-postneonatal outcome in children born to affected females. Furthermore we report our experience.

**Methods:** There were 93 pregnancies (literature, N=88; our experience, N=5) in 45 women with Gitelman syndrome (N=25), antenatal Bartter syndrome (N=1) or an unclassified hypokalemic disorder (N=19).

**Results:** There were 12 spontaneous abortions and 6 terminations. Pregnancy was associated with a decline in potassium level. Drug management included supplementation with potassium in all, with magnesium in at least 19, and with potassium-sparing diuretics in 11 pregnancies. A term or near term birth was noted in the 75 pregnancies. Babies exposed in utero to potassium sparing diuretics were also found to be normal. Somatic growth and neuropsychological development are currently normal in 5 subjects aged between 1 and 18, median 10 years born to 4 patients with the biochemical and molecular diagnosis of Gitelman syndrome, who are on follow up at our institutions.

**Conclusions:** Females affected by inherited hypokalemic disorders can become pregnant and the disorder may be managed without outward effect on the fetus and with an excellent long-term outcome.

## Abstract# 852

**Wilms Tumor Gene 1 (WT1) Mutation in Monozygous Twins with Congenital Nephrotic Syndrome (CNS)** K. Blahova,[1] F. Fencl,[1] D. Mixova,[2] K. Vondrak,[1] J. Zieg,[1] V. Stara,[1] J. Stejskal,[3] M. Malina.[1] *[1]Dpt. of Paediatrics, 2nd Faculty of Medicine Charles University and Motol Univ.Hospital, Prague, Czech Republic; [2]Dpt. of Anestesiology and Resuscitation, Dtto, Dtto, Czech Republic; [3]Dpt. of Pathology and Molecular Medicine, Dtto, Dtto, Czech Republic.*

**Objectives:** CNS is classified in two types: Finnish type and other,including diffuse mesangial sclerosis (DMS).WT1 gene mutations have been demonstrating in some pts with DMS.Monozygous twins with CNS and early onset of ESRD and their genetic analysis are presented.

**Methods:** Female twins from first pregnancy without polyhydramnion and normal placenta were born at 32 g.w. with 46 XX karyotype, normal female phenotype. On admission at the age of 2 months edema, ascites, oliguria/anuria, hypoproteinaemia, hypoalbuminaemia, low IgG, low ATIII, proteinuria (12g/L in a girl with oliguria) were presented. USG showed a small kidney enlargement,parenchymal hyperechogenicity and poor corticomedullary differentiation. Girls reached ESRD within a few weeks. Peritoneal dialysis (PD) was started with a shift to CVVH/CVVHD in one of them. This one died at the age of 23 weeks.Preliminary light microscopy showed DMS.

**Results:** After excluding the most common CNS mutations (NPHS1,2), direct sequencing of a WT1 gene (exon 8 and 9) revealed the presence of an identical heterozygous mutation R417R in boths girls.

**Conclusions:** NPHS1, 2 gene mutations are not the only cause of CNS. The genetic analysis of WT1 gene should be applied in all CNS pts, expecially with presence of DMS. Kidney Tx in a twin who is still on PD is the only therapeutic alternative. Because of the risk of Wilms tumor, prophylactic nephrectomy at the time of ESRD/Tx should be performed.

## Abstract# 853

**Set Up *In Vitro* Polycystin 1 Knock-Down Cell Line by miRNA Method** Y.-Y. Chiou,[1] S.T. Jiang.[2] *[1]Pediatric Nephrology, National Cheng-Kung University Medical Colledge and Hospital, Tainan, Taiwan; [2]Institute of Molecular Biology, Academia Sinica, Taipei, Taiwan.*

**Objectives:** Autosomal dominant polycystic kidney disease (ADPKD) is one of the most -common inherited cystic kidney diseases and can lead to chronic renal failure. In this study, we utilized siRNA technology to knock down the polycystin 1 protein expression in immortalized renal tubular cell line to investigate the future study.

**Methods:** Stable knockdown of PC1 in M-1 mouse CCD cells were achieved by lipo-polymeric formulation with a specific small interfering RNA for Pkd1. After transfection, stable clone of miRNA PKD1 cell lines were sorted by FACS and selected by blasticidin. For immunodetection, cells were lysed and detected by rabbit anti-PC1 polyclonal antibody. Proliferation assay was performed and tubulogenesis and cysts formation were investigated in an ice-cold matrigel solution.

**Results:** Polycystin 1 knock-down cell miRNA 6820 exhibited significant effects on cell proliferation at 24h later to 120h cell cultures. According to the GFP fluorescence intensity, miRNA 6820 cells could be sorted by high and low fluorescence. Down regulation of polycystin 1 protein in transcriptional level were confirmed by RT-PCR and real time PCR and these were parallel to the intensity of GFP. Cysts formation presented in 3-D matrigel system and low expression of polycystin 1 cell line would produce larger cysts.

**Conclusions:** Polycystin 1 knock-down cell lines were set up successful and their cellular biologic behaviors would present as human ADPKD. These cell lines will provide as a good *in vitro* cellular model to investigate further cellular mechanical signal pathway of cyst formation in future.

## Abstract# 854

**The Importance of Duplex Collecting System Diagnosed by Ultrasonographic Screening in Infants** K. Drnasin,[1] M. Saraga,[2] M. Saraga-Babic.[3] *[1]Pediatric Outpatient Clinic, Solin, Croatia; [2]Pediatric, University Hospital, Split, Croatia; [3]School of Medicine, University of Split, Split, Croatia.*

**Objectives:** To perform ultrasonographic screening (USc) of clinically asymptomatic infants in order to determine the frequency of duplex collecting system (DCS) and follow up the urinary tract infections (UTI) and/or other coexisting urinary tract anomalies (CUTA).

**Methods:** USc of urinary tract (UT) was performed on an unselected population of 2471 healthy infants (1276 boys and 1195 girls). All of them had normal result of prenatal USc of UT. Out of a total number of postnataly screened infants, 1749 of them were followed up for 5-60 months regarding the presence of UTI or urinary tract anomalies (UTA).

**Results:** In the examined group, 58 infants (2.3%) had UTA, while 33 (1.3%) infants with UTA had DCS. UTI was found in 16 (48.5%) infants with DCS, and in 146 (8.5%) out of 1716 followed infants without DCS. Other CUTA were found in 6 (18.1%) infants with DCS. Pyelocaliceal dilation (PCD) was found in 13 infants with DCS. In that group UTI was found in 10 (77%) infants, while in the rest of 20 infants with DCS, UTI was found in 5 (25%) of them.

**Conclusions:** Postnatal USc of UT in children is an useful and valuable method for detecting DCS. Our results indicate that DCS in infants increases the risk for UTI approximately 5.7 times and for other CUTA 7.9 times, compared to an unselected population. Moreover, children with DCS associated with PCD have a triple greater risk for UTI than those with DCS only. Early diagnosis of DCS warns about the increase of UTI and CUTA possibility, so we recommend the postnatal USc of UT in all children during the first months of life in order to prevent kidney damage.

## Abstract# 855

**Outcome of Prenatal Hydronephrosis in Infants Born at Sultan Qaboos University Hospital, Oman** I.B. Elnour,[1] Z.S. Reyes,[2] M.M. Florideza,[3] M.A. Sadoon,[4] S.S. Hussein,[5] D.K. Sankhla.[6] *[1]Child Health, Sultan Qaboos University, Muscat, Muscat, Oman; [2]Child Health, Sultan Qaboos University, Muscat, Muscat, Oman; [3]Child Health, Sultan Qaboos University, Muscat, Muscat, Oman; [4]Child Health, College of Medicine, Sultan Qaboos University, Muscat, Muscat, Oman.*

**Objectives:** We carried a prospective hospital based study to assess the out come of mild to sever antenatal hydronephrosis in the first 8 weeks of life in our population, to highlight the diagnostic and therapeutic dilemma encountered and determine the future mode and frequency of necessary radiological investigations.

**Methods:** Babies born at SQUH with the diagnosis of prenatal hydronephrosis between 1998- 2007 were included in a prospective study. Infants were accessed with combination of renal ultrasonography, micturating voiding cystogram (MCUG) and a well tempered diuretic renogram. Hydronephrosis was classifies following Gringnon et al classification.

**Results:** 17 babies had vesicoureteric reflex with total of 22 refluxing unit. 4 babies had sever pelviureteric junction obstruction (PUJ). 7 babies had unequivocal findings of mild PUJ on ultrasonography and renogram. 16 patients (10.4%) had IVU for persistent hydronephrosis and confirmed 11 units of PUJ. 20 babies had equivocal finding of mild PUJ/idiopathic HN. 20 babies had unequivocal finding suggesting idiopathic hydronephrosis.
Other pathological ginding will be discussed.

**Conclusions:** With wide use of antenatal ultrasonography, more case of congenital renal formation are detected. Babies with prenatal hydronephrosis should be promptly investigated to determine the aetiology, counsel parents and prevent future reflex nephropathy.

## Abstract# 856

**Liver Involvement in Autosomal Rezessive Polycystic Kidney Disease (ARPKD)** M. Feldkötter,[1] T. Ronda,[1] S. Habbig,[1] B. Beck,[1] F. Körber,[2] B. Hoppe.[1] *[1]Pediatric Nephrology, University Hospital, Cologne, Germany; [2]Pediatric Radiology, University Hospital, Cologne, Germany.*

**Objectives:** Autosomal recessive polycystic kidney disease is always accompanied by congenital hepatic fibrosis (CHF), characterized by hyperplastic bile ducts and portal fibrosis. We here describe diagnostic criteria and the clinical course of liver involvement in 18 patients now aged 0.3-21 years.

**Methods:** Duration of clinical observation varied between 0.3 to 17.9 years. Type and degree of liver involvement was determined by laboratory tests and (Duplex) sonography, in some by MR-cholangiography, oesophago-gastroscopy or liver biopsy.

**Results:** In all patients with liver biopsy (9/9), CHF was histologically confirmed, however, clinical follow up was heterogenous. Caroli syndrome was identified with ultrasound and confirmed by MR-cholangiography in 5 patients. A portal hypertensive form was found in 4 children. Patients with Caroli syndrome demonstrated at younger age a hepatic manifestation with portal hypertension in two patients expressed as gastro-esophageal varicosis. Also, prominent accompanying complications were found in the other patients with portal hypertension. Duplex sonography showed re-canalized umbilical as well as periportal veins in 2 patients. Endoscopically, esophageal varices were found in 6 of 11 patients. Reasons for liver or liver-kidney transplantation were either severe esophageal hemorrhage requiring sclerotherapy, or recurrent cholangitis and esophageal hemorrhage, or cholangitis and renal failure.

**Conclusions:** Hence, specific attention has to be paid to liver involvement in patients with ARPKD, as it adds significant long term problems and morbidity.

## Abstract# 857

**Parenchymal Thickness in Children with a Solitary Kidney** H. Flögelová,[1] K. Michálková,[2] J. Langer,[3] Š. Dolezalová.[3] *[1]Faculty of Medicine, Palacký University, Olomouc, Czech Republic; [2]Department of Radiology, University Hospital, Olomouc, Czech Republic; [3]Department of Pediatric, General University Hospital, Prague, Czech Republic.*

**Objectives:** Reduction of parenchymal thickness (PT) detected by ultrasound examination is one of the criteria for surgery in obstructive uropathy. In a solitary kidney (SK) with obstruction, determination of severe reduction of the parenchyma is difficult. This is due to the fact that as the SK develops, hypertrophy of the parenchyma occurs. As a result, the reduction may be misrepresented. Moreover, there is no comparison with a healthy kidney. Therefore, a study was performed to determine normal PT in children with a healthy functionally SK.

**Methods:** The prospective multicenter study comprised 163 children, aged 11 days to 18.96 years (mean age 6.46 years; 64% males). The function of the SK was confirmed by a DMSA scan. All the children had normal glomerular filtration rates. In addition to other parameters, the thickness of the renal parenchyma was measured during an ultrasound examination of the SK. The study assessed the dependence of PT on the children's age, height and weight (non-parametric Spearman's correlation analysis).

**Results:** The correlation analysis showed a positive correlation of PT with all the studied parameters. Linear dependence was only revealed between the thickness of the renal parenchyma and body height. The correlation coefficient for height was 0.837. The linear regression equation was y=0.087x+5.902. The coefficient of determination was $R^2$ =0.665.

**Conclusions:** Parenchymal thickness in a functionally SK is well correlated with the body height. The 5th and 95th percentiles were estimated for PT in a SK depending on the body height.

## Abstract# 858

**Mental Retardation, Agenesis of Vena Cava Inferior and Sacrum, Associated with VACTERL Anomalies: Unusual Features or a New Syndrome?** D.O. Hacihamdioglu,[1] D. Gul,[2] S. Vurucu,[3] C. Yildiz,[4] M. Kocaoglu,[5] Y. Kibar,[6] F. Gok.[7] *[1]Pediatric Nephrology, GATA Medical School, Ankara, Turkey; [2]Genetics, GATA Medical School, Ankara, Turkey; [3]Neurology, GATA Medical School, Ankara, Turkey; [4]Orthopedics, GATA Medical School, Ankara, Turkey; [5]Radiology, GATA Medical School, Ankara, Turkey; [6]Urology, GATA Medical School, Ankara, Turkey; [7]Pediatric Nephrology, GATA Medical School, Ankara, Turkey.*

**Objectives:** We report a 3 years old female who was born from unconsanguineous Turkish parents.

**Results:** On the physical examination; head circumference, weight and length were under the 3rd percentiles and high blood presure. Her psychomotor and mental development was severely delayed. She's malformations include long face, frontal bossing, bitemporal depression,asymmetric ears, radial deviation of both wrists, clinodactyly, rudimentation and ulnar deviation of first finger of left hand, aplasia of first finger of right hand, bilateral radial hypoplasia, sacral agenesis, developmental hip dsyplasia and bilateral equinovarus. On the laboratory findings echocardiography demonstrated a secundum ASD, abdominal US showed bilateral pelvicaliectasy, MCUG showed bilateral grade 5 VUR. Abdominal CT demonstrated the absence of inferior vena cava.

**Conclusions:** In our knowledge vena cava inferior agenesis association with skeletal anomalies and mental motor retardation has not been reported in lecture so far. We offer stillborn or infants with limb anomalies would better evaluated for especially urinary tract anomalies. Because early diagnosis is crucial in such cases as potential targeted therapy (either surgical or conservative) might prevent irreversible damage of renal parenchyma.

## Abstract# 859

**Longitudinal Study of Renal Function in Children Diagnosed with Simple Renal Pyelectasis** M.J. Hernández-González,[1] M.I. Luis-Yanes,[1] M. Monge,[1] S. González-Cerrato,[1] F.M. Claverie-Martín,[2] V.M. García-Nieto.[1] *[1]Pediatric Nephrology Uniy, Hospital Nuestra Señora de Candelaria, Santa Cruz de Tenerife, Canary Islands, Spain; [2]Unidad de Investigación, Hospital Nuestra Señora de Candelaria, Santa Cruz de Tenerife, Canary Islands, Spain.*

**Objectives:** The simple renal pyelectasis (SRP) is defined as a moderate dilatation of the renal pelvis in the absence of vesicoureteral reflux or urinary tract obstruction. Recently, we described that might indicate a genetic predisposition to suffer from urolithiasis in adulthood. We have studied the longitudinal renal function in a group of children with SRP.

**Methods:** We included 44 children (34M, 10F). The age at initial study (S1) was 104.8 ± 93.1 days (range: 4-330 days). The age at the final study (S2) was 3.6 ± 2.5 years (range: 1.02-10.5 years). We determined the maximum urine osmolality (Uosm) after stimulation with desmopressin and urinary albumin/creatinine (Alb/cr) and N-acetilglucosaminidasa/creatinine (NAG/cr).

**Results:** In S2, Uosm values were significantly higher than in S1 (938.3 ± 138.4 vs. 574.3 ± 135.3, p<0.001). Similarly, in S2 values of MAU/cr (1.4 ± 1.7 vs. 5.6 ± 4.5 µg/µmol, p<0.001) and NAG/cr (4.2 ± 3.6 vs. 18.9 ± 19.6 U/g, p = 0.001) were significantly lower than in S1. In S1, 18/44 infants (40.9%) showed renal concentrating defect and S2 only 8/44 children (18.2%). In S1, 11/35 (31.4%) had the ratio Alb/cr high and S2 only 4/36 (10%). In 26/44 (59.1%) of families, family history of urolithiasis were in first and/or second generations.

**Conclusions:** The SRP is a benign process. It is unknown why some patients have impared renal function initially although a progressive improvement of the same one exists.

## Abstract# 860

**An Exceptional Case Neonatal Acute Renal Failure, ACE Inhibitors Nephropathy** M. Giménez, A.D. Madrid, C. Herrero, C. Yolanda, L. Enrique, V. Ramon, N. Jose, M. Garrido. *Pediatric Nephrology Department, Hospital Vall d'Hebron, Barcelona, Barcelona, Spain; Pathological anatomy, Hospital Vall d'Hebron, Barcelona, Barcelona, Spain.*

**Objectives:** These drugs now have been shown to be fetotoxic causing profound fetal hypotension, renal tubular dysplasia, anuria-oligohydramnios, growth

restriction, hypocalvaria, and death when used in the second and third trimesters of pregnancy.
The target is to show a case born in our service with all his clinical characteristics and his renal biopsy.
**Methods:** we check his clinical chart and his renal biopsy.
**Results:** This was a 35-week, 1,995-g infant of a 35-year-old pregnant woman who had been treated with lisinopril throughout her pregnancy for hypertension. Spontaneous rupture of membranes and fetal distress led to the premature delivery of the infant who was noted to have severe oligohydramnios, intrauterine growth retardation, hypocalvaria, profound hypotension, and renal failure with a renal ultrasound that showed big-sized kidneys.[/bold]
Renal biopsy at 8 weeks, which showed tubular focal dilation and absent differentiation of proximal convoluted tubules. These finding are known like disgenesia tubular renal typical from the deficit of angiotensina during the fetal period.
**Conclusions:** We conclude that ACE inhibitor induced renal tubular dysgenesis is an unusual but important cause of acute renal failure neonatal.
ACE inhibitors have been shown to cause fetal toxicity, including profound fetal hypotension, anuria, oliguria, and growth restriction.
The best strategy to prevent ACE fetopathy is to aggressively educate physicians and the public that these drugs should not be used during the second and third trimester of pregnancy.

## Abstract# 861

**A Case of Congenital Nephritic Syndrome with Prenatal Brain Infarction** K. Hine,[1] H. Saito,[1] A. Yoshida,[1] J. Suzuki,[1] M. Ishige,[1] T. Urakami,[1] K. Nakanishi,[2] S. Takahashi.[1] *[1]Pediatrics, Nihon University School of Medicine., Tokyo, Japan; [2]Pediatrics, Wakayama Medical University, Wakayama, Japan.*
**Objectives:** Congenital nephritic syndrome (CNS) is often complicated with thrombosis. We had a patient with CNS complicated with brain infarction developed during fetal period. Since complication of congenital brain infarction with CNS has not been reported before, the possibility of prenatal onset and the time point of brain infarction is discussed.
**Methods:** A 10-month-old boy without family history was born at 38 weeks of gestation and weighed 2510g. Giant placenta was noted at birth. No abnormalities were noted in his growth and development. He was transferred at 2 months of age to our hospital suspected of nephritic syndrome. A high level of proteinuria was noted, and extensive brain infarction was identified by diagnostic imaging. Genetic analysis revealed no mutations. Renal biopsy established the diagnosis of CNS of the Finnish type.
**Results:** Although MRI/MRA identified old brain infarction in the left middle cerebral artery region, no neurological abnormalities were observed before hospitalization. Since formation of a secondary cerebral sulcus was observed at the infarction site, onset of infarction was estimated to be 32 weeks of gestation or later. The fractional excreation of total protein (FETP) was 0.5, and the prenatal FETP was estimated to be 0.05, assuming a prenatal GFR of 10 ml/min. This suggested a theoretical possibility that hyperproteinuria and hypoproteinemia might have developed during fetal life to trigger prenatal brain infarction.
**Conclusions:** We had a patient with CNS of the Finnish type complicated with brain infarction occurred during fetal period.

## Abstract# 862

**The Importance of Postnatal Ultrasonography in the Management of Antenatal Hydronephrosis** N. Karakurt,[1] N. Besbas,[2] S. Tekgul,[3] Y. Bilginer,[2] A. Bakkaloglu,[2] A. Bakkaloglu.[2] *[1]Pediatric Hematology, Ankara Pediatric Hematology and Oncology Training and Research Hospital, Ankara, Turkey; [2]Pediatric Nephrology, Hacettepe University Medical School, Ankara, Turkey; [3]Urology, Hacettepe University Medical School, Ankara, Turkey.*
**Objectives:** We aimed to examine the importance of postnatal renal USG in the follow-up of children with antenatal hydronephrosis.
**Methods:** The clinical and USG findings, diuretic renograms, DMSA sintigraphies and VCU of 76 patients with prenatal hydronephrosis are recorded. 126 kidney units are included in the study.
**Results:** Regarding to the renal pelvis anteroposterior diameter in the postnatal USG on the 7th day, 48 kidney units had mild, 31 had moderate, 24 had severe, and the rest had no hydronephrosis. 40% of kidney units with mild hydronephrosis and 32% of kidneys units with moderate hydronephrosis were found to be normal at the end of the follow-up. UPJO was dedected in 38% of kidney units with severe hydronephrosis.
Of the 23 kidney units which were found to be normal in the first postnatal USG, only 11 were found out to be normal in the second USG.
There was no significant relation in between the degree of the hydronephrosis of the kidneys with or without VUR.
**Conclusions:** Predicting postnatal genitourinary pathologies in patients with antenatal hydronephrosis by means of renal USG is very important for the clinician. Even if the first postnatal USG is normal, USG should be repeated in 4 weeks. Mild and modarate hydronephrosis should be followed with USG. For mild, moderate an even severe hydronephrosis, we recommend to delay performing a VCU as much as possible, considering the electromagnetic radiation exposure.

## Abstract# 863

**Does Anthropometric Development Affect the Spontaneous Closure of Vesicoureteral Reflux?** M. Bayram,[1] S. Kavukcu,[1] E. Serdaroglu,[2] M. Turkmen,[1] B. Kasap Demir,[1] A. Soylu.[1] *[1]Pediatrics, Dokuz Eylul University Medical Faculty, Izmir, Turkey; [2]Pediatrics, Behcet Uz Children's Hospital, Izmir, Turkey.*
**Objectives:** Primary vesicoureteral reflux (VUR), especially low grade, resolves in time spontaneously in most cases. We aimed to evaluate if the rate of anthropometric development of children affects the closure of VUR.
**Methods:** Children with primary low grade (I to III) VUR were grouped as those with spontaneous closure (Group 1) and those without spontaneous closure (Group 2). All children were evaluated retrospectively for height SDS, weight SDS and BMI at the time of diagnosis and yearly thereafter until the last voiding cystography. In addition, the rate of increase in height and weight for each year was evaluated for all children. Both groups were compared with respect to these parameters.
**Results:** Group 1 (n=32) and Group 2 (n=10) were similar for sex, age at diagnosis and follow up period. With regard to anthropometric development, only height SDS was different between the Group 1 and Group 2 at the time of diagnosis (-0.10±1.18 vs 0.93±1.20, p=0,022). At the end of follow up period, none of the parameters were different between these groups. In addition, no difference was determined for the evaluated parameters within each group at the time of diagnosis and at the last cystography.
**Conclusions:** Although VUR resolves in time, we could not demonstrate that the rate of anthropometric development affect the closure of low grade VUR.

## Abstract# 864

**Long-Term Renal Outcome in Patients with Lumbar Meningomyelocele** C.-Y. Lee, C.-Y. Lin. *Pediatric Nephrology, Children's Medical Center, China Medical University and Hospital, Taichung, Taiwan.*
**Objectives:** The most common complication of lumbar meningomyelocele (LM) is progressive loss of renal mass and subsequent renal failure (RF). The aim of this study focused on long term follow-up in children with LM and factors influence renal progression.
**Methods:** Forty-six LM patients were diagnosed at birth and received surgical repair of meningomyelocele for twenty or more years. Basic data were evaluated for associated abnormalities, urodynamic studies and laboratory results, then filed in the National Health Insurance database (one file per person), with data for 1997-2009 analyzed.
**Results:** There were 20 cases with spinal bifida (SB) and 23 cases with bladder dysfunction, 3 cases progressed to RF and on dialysis, showing that SB itself was not associated with long-term renal progression. Only bladder dysfunction and vesicoureteral reflux (VUR) strongly correlated with long-term renal progression. Frequent urinary tract infection exacerbated long-term outcome. In SB patients, as Hb decreased, systolic BP elevated and serum creatinine fell; as body weight decreased, diastolic BP rose.
**Conclusions:** Further renal progression of LM throughout follow-up correlated with bladder dysfunction, VUR, numbers of urinary tract infection and hypertension. Close cooperation among pediatric nephrologists, urologists and transplant surgeons is vital.

## Abstract# 865

**A Clinico-Genetic Study of Congenital Renal-Coloboma Syndrome** J. Leviashvili, N. Savenkova, L. Lysenko, I. Anihkova, E. Pankov, E. Fedotova. *Pediatric Nephrology, St-Petersburg State Pediatric Medical Academy, Saint-Petersburg, Russian Federation.*
**Objectives:** Report on a patient with congenital Renal-Coloboma syndrome.
**Methods:** Clinical, histological and genetic methods.
**Results:** Report on a patient, a 10-year-old girl, diagnosed with optic disk coloboma with chorioretinal degeneration at birth. On clinic examination she showed hematuria, increasing proteinuria and arterial hypertension. Doppler ultrasound and angiographic findings revealed stenosis of the renal arteria of right multicystic displastic kidney. At the age of 9 years it was performed right nephrectomy. The sizes of the removed right kidney - 5 sm x 2,5 sm x 1,5 sm. Histological examination of the renal tissue revealed multicystic dysplasia. Work-up revealed chronic renal failure at the age of 11: Glomerular Filtration Rate by creatinin clearance was decreased (35-47ml/min), creatinine was increased - 0,135 mmol\L, hypostenuria, renal tubular acidosis presented. Ophthalmological examination showed a coloboma of the optic nerve and nistagm. Genetic testing showed nucleotid change in intron 6 gene PAX2: IV 6-1 g->c in heterozygote state which is a mutation responsible for the development of the disease. Genetic

testing of her brother (6 years old) did not show a PAX 2 mutation. Treatment strategies: routine treatment of arterial hypertension and syndroms of chronic renal failure, follow-up by ophthalmologist to monitor vision.
**Conclusions:** The association of unilateral multicystic dysplastic kidney, optic disk coloboma and gene PAX2 mutation is consistent with the diagnosis of congenital Renal- Coloboma syndrome in girl.

## Abstract# 866

**Ultrasound of Urinary System and Urinary Screening in 14 256 Asymptomatic Children in China** Q. Li, H.P. Yang. *Children's Hospital of Chongqing Medical Universtiy, Chongqing, China; Centre for Lipid Research, Chongqing Medical University, Chongqing, China.*
**Objectives:** The aim of this study is to assess the characteristics of urinary system diseases and the role of the ultrasound screening and urinalysis screening for chronic kidney disease (CKD) in asymptomatic children in China.
**Methods:** Between September 2008 and November 2008, 14 256 children excluding those with obvious symptoms and signs were enrolled in our study. All the subjects accepted ultrasound and urinary screening. A case– control study was performed to evaluate the relative risk of having stones in those children exposed to melamine formula.
**Results:** Of the enrolled children, 6.10% (869 of 14 256) showed abnormalities, of which 409 (2.87%) were established by ultrasound, 572 (4.01%) by urinalysis and 112(0.79%) by both ultrasound screening and urinalysis. The abnormalities included congenital anomalies of kidney and urinary tract, urinary stones and/or hydronephrosis, leucocyturia and haematuria and/or proteinuria. Children exposed to melamine formula were 5.17 times as likely to have kidney stones as children exposed to no-melamine formula (95% confidence interval, 3.28–8.14; $P < 0.001$); the probability of kidney stones in melamine-fed infants were 6.28 times as likely as those no melamine-fed (95% confidence interval, 3.71–10.65; $P < 0.001$).
**Conclusions:** Ultrasonography and urinalysis could complement each other and play important roles in the early diagnosis of anomalies of the urinary system, but urinalysis is a more cost-effective screening tool for CKD in children in China. Exposure to melamine-contaminated formula associated with urinary stones, especially in infants, was significantly higher than the control group.

## Abstract# 867

**Nephrogenic Diabetes Insipidus (NDI) Associated with Fetal ARB Exposure** Y. Murano, A. Endo, N. Nishizaki, S. Hara, T. Someya, Y. Ohtomo, T. Shimizu. *Department of Pediatrics and Adolescence, JUNTENDO University Faculty of Medicine, Tokyo, Japan.*
**Objectives:** Angiotensin-receptor blockers (ARBs) and angiotensin converting enzyme inhibitors (ACEIs) are widely used drugs for hypertension and heart failure.
Recent reports show that the maternal use of them may induce ACEI/ARB fetopathy, characterized by the symptoms such as oligohydramios, neonatal pulmonary hypoplasia, renal insufficiency, limb contracture, fetal hypotension, and so on.
To date, there are few reports on ARB fetopathy. We report a case with NDI followed by neonatal renal failure due to fetal ARB exposure.
**Results:** Case: a 2-year-old boy
His 28-year-old mother had treated hypertension with valsartan throughout pregnancy. He was born at a gestational age of 34 weeks with 2308g body weight, 47cm length, APGAR score 7/8. He was transferred to our NICU, where transient tachypnea due to mild pulmonary hypoplasia, renal dysfunction and hypocalvaria was managed with oxygen supply and intravenous fluid support for several days. These outcomes were favorable and he was discharged on day 35.
At the age of 2 years, he developed polydipsia and polyuria and admitted to our hospital to evaluate his renal function. His plasma / urinary osmolality were 281 / 189 mOsm respectively, which suggested diabetes insipidus. Water deprivation test revealed maximum urine osmolality of 167mOsm/kg with no response to the injection of arginin vasopressin. As a result, we could diagnose him as nephrogenic diabetes insipidus.
**Conclusions:** Among a few reports of ARB fetopathy, cases with NDI are rare. More attentions should be paied for the occurrence of late-onset complications on the cases with ACEI/ARB fetopathy.

## Abstract# 868

**Audit of Pelviureteral Junction Obstruction at Red Cross Children's Hospital** I. Ocheke,[1] M. McCulloch,[2] P. Gajjar.[2] *[1]Paediatrics, JUTH, JOS, Plateau, Nigeria; [2]Renal, Red Cross Hospital, Cape Town, Western Cape, South Africa.*
**Objectives:** To describe the characteristics, mode of diagnosis, clinical course and outcome of management of children with pelvi-ureteric junction obstruction (PUJo) managed at the Red Cross Children's Hospital over a six year period (2002-2007).
**Methods:** A retrospective folder review.
**Results:** 100 children with PUJo were reviewed. This constituted 133 renal units with PUJo. The male: female ratio was 4:1; the left kidney was most commonly affected in 40%. 70% of those with hydronephrosis confirmed to be secondary to PUJo regressed spontaneously at 6 months of follow up while the rest progressed at variable interval over the 12 months of review of each case. Mostly all the children (99%) with the disease were antenatally diagnosed.
19 (14.3%) had surgery in the form of pyeloplasty, 31% of this number had surgery in the first 6 months of life.
11 children had confirmed UTI and the commonest organism isolated was *Klebsiella pneumoniae* followed by *E coli*.
Outcome of surgical management showed that there was overall improvement in function (MAG3) renogram and decrease of AP pelvis diameter at 12 months of follow up.
**Conclusions:** Pelviureteric junction obstruction is an important and common cause of prenatal hydronephrosis. Significant proportion of children had spontaneous regression of hydronephrosis, however a few had surgical intervention with appreciable improvement of renal function.

## Abstract# 869

**The Challenges of Management of Posterior Urethral Valve in a Resource Limited Setting** I.O. Odetunde,[1] H.U. Okafor,[3] O.A. Odetunde,[2] N.E. Obianyo,[2] J.C. Azubuike.[1] *[1]Paediatrics, Enugu State University Teaching Hospital, Enugu, Enugu, Nigeria; [2]Paediatric Surgery Unit, Surgery, Enugu State University Teaching Hospital, Enugu, Enugu, Nigeria; [3]Paediatrics, University Nigeria Teaching Hospital, Enugu, Enugu, Nigeria.*
**Objectives:** Objectives: This study was conducted to ascertain the pattern of presentation, frequency and complications in patients with posterior urethral valve at initial presentation, duration of symptoms before presentation and outcome of management in Enugu, South -East Nigeria.
**Methods:** MATERIALS AND METHODS: A retrospective study of children presenting at the University of Nigeria teaching hospital, Enugu from 1997-2004 (7years) and the Enugu State University Teaching Hospital 2005 -2009 (4years) with the diagnosis of posterior urethral valve.
**Results:** RESULTS: Twenty-one patients were seen during this period 1997-2009. The ages ranged between two (2) days and thirteen (13) years, with mean age of 3 years. The duration of symptoms before presentation is a mean of 2.6years. 2(9.52%) of cases presented at the neonatal age, 10(47.62%) were aged 1month to 1 year, 5(23.81%) were aged between 1 – 5 years and 4(19.05%) were aged 5-13 years. 19(91%) of cases presented with urosepsis while 8(38%) presented with significant renal insufficiency. Laboratory findings varied from slight raise to marked elevation in serum creatinine level.
**Conclusions:** Early presentation with prompt intervention will improve the renal function and prevent the renal complication especially in the neonatal period, in a setting where renal replacement therapy is not readily available.

## Abstract# 870

**Cystatin C at Birth in Neonates with Congenital Kidney Malformation Diagnosed on Prenatal Ultrasound Compared to a Normal Population** P. Parvex,[1] M.H. Billieux,[2] C. Combescure,[3] R. Robyr,[4] E. Girardin.[1] *[1]Pediatric Nephrology, HUG, Geneva, Switzerland; [2]Obstetrics, HUG, Geneva, Switzerland; [3]Clinical Research, HUG, Geneva, Switzerland; [4]Obtstetrics, La Tour Hospital, Geneva, Switzerland.*
**Objectives:** Congenital abnormality of kidney and urinary tract (CAKUT) accounts for 20% of all anomalies diagnosed on prenatal ultrasound(US). As Cystatin C (Cys C) is a sensible marker for renal function in neonates, we compared Cys C at birth in neonates with prenatal diagnosed of CAKUT with Cys C in normal neonates.
**Methods:** In the control group, 100 neonates with normal prenatal US were recruited and Cys C was measured at birth to obtain reference interval. In the same period, Cys C was measured in 19 neonates, with CAKUT, diagnosed on prenatal US (13 dilated kidneys(K) >10mm ±dilated ureters; 3 dysplastic K, 2 multicystic K disease, 1 megabladder). Among these, 11 had one kidney anomaly the 8 others have two kidneys involved.
**Results:** The 19 patients(pts) with CAKUT were comparable in gestational age, weight and size to the control group. In the control group, Cys C reference interval was [1.55-2.66 mg/l] with a mean (M) value of Cys C of 2.04 ± 0.27SD. In the 19 pts with CAKUT, M Cys C was 2.18 ± 0.44SD and the difference between the 2 groups was not significant (P=0.069). However the M Cys C (2.43 ± 0.38SD) was significantly higher in pts with bilateral kidney anomalies compared to pts with only one kidney involved (M Cys C of 1.9 ± 0.4SD) (p=0.03), and to the pts from the control group (P=0.00037).
**Conclusions:** Cys C is a sensible marker of renal function in neonates with CAKUT. Preliminary results showed significant increased of Cys C in neonates with both kidney involved compared to one or to the control group.

## Abstract# 871

**Correlation of Hypertension with Renal Function in Children with Obstructive Uropathy** H. Rahman, A. Begum, M. Hossain, G. Muinuddin, A. Rahman. *Pediatric Nephrology, Bangabandhu Sheikh Mujib Medical University, Dhaka, Bangladesh.*

**Objectives:** The aim and objective of the study was to see the correlation of hypertension with renal function and renal function with obstructive uropathy.

**Methods:** In this study 41 different types of obstructive uropathy patients with age range from 4 months to 14 years (Mean age: 5.7 ± 2.3 years) were enrolled. Patients were diagnosed by detailed history, physical examination and relevant imaging studies like ultrasound of the kidney, ureter, bladder region with post voidal residue, micturating cystourethrogram, DMSA and DTPA renogram with total and split renal function. Chronic kidney disease (CKD) Stagings were done by Schwarts formula.

**Results:** Important observation of this study were out of 41 patients 11 (26.88%) had hypertension 30(73.17%) were normotensive. Among the hypertensive patients 5 (45.45%) were in stage V (mean Ccr 10.7 ± 2.57 ml/1.73 $m^2$/min) and 2 (18.18%) were in stage IV (mean Ccr 22.4 ± 7.58 ml/1.73 $m^2$/min) CKD, which is statistically significant (P<0.001). When normotensive patients were considered 13(40.62%), 5(15.62%) and 4(13.33%) were in stage III, IV and V CKD respectively. On the other hand, when CKD status were correlated with types of obstructive uropathy posterior urethral valve 25 (61%) were present in the majority cases and all (100%) of the patients had different grades of CKD.

**Conclusions:** It was observed from this study, patients with CKD due to obstructive uropathy may be normotensive ever in stage V CKD, and posterior urethral valve was the commonest cases of obstructive uropathy in children.

## Abstract# 872

**Postnatal Outcome of Antenatal Echogenic Kidneys – A Retrospective Study** R. Lea,[1] S. Russell,[2] M. Lewis,[3] S. Sukthankar.[1] *[1]Paediatric Medicine, Royal Manchester Children's Hospital, Manchester, United Kingdom; [2]Foetal Medicine Unit, Saint Mary's Hospital, Manchester, United Kingdom; [3]Paediatric Nephrology, Royal Manchester Children's Hospital, Manchester, United Kingdom.*

**Objectives:** Antenatal echogenic kidneys suggest differential diagnoses of polycystic (PKD), cystic dysplastic (CDK) or multicystic dysplastic (MCDK) kidneys. Postnatal prognosis is guided by the liquor volume, any other system anomalies and presence of parental renal cysts. We aim to study the postnatal outcome in children with antenatally diagnosed echogenic kidneys.

**Methods:** A retrospective case note review was undertaken. The spectrum of postnatal investigations (renal function, ultrasonography of kidneys) and clinical course (blood pressure, need for medication, or renal replacement) over a five year follow up period is presented here.

**Results:** In the 4 year recruitment phase, 47 foetuses with echogenic kidneys were seen in the Foetal Medicine Unit (19 with macrocysts). Twenty (10F, current age 5-9 years) of the 29 live births (7 with macrocysts) were included in this study (data unavailable in 9). Six died in infancy (3 PKD, 2 bilateral MCDK, 1 congenital nephrotic syndrome). Survivors were followed up for at least 5 years. Eight remained normotensive with normal renal function, 5 had renal impairment (3 ARPKD, 2 CDK) - two were also hypertensive. Three of the 4 with ADPKD had family history of PKD.

**Conclusions:** The postnatal outcome of antenatally diagnosed echogenic kidneys is variable. The morbidity in survivors beyond the neonatal period is significant, with high incidence of hypertension and/ or severe renal impairment on 5 years' follow up. Long term studies with larger numbers are required to further delineate the prognosis.

## Abstract# 873

**Prune Belly Syndrome: Burden and Outcome** H.H. Twahir. *Department of Paediatrics, Coast Provincial General Hospital, Mombasa, Coast Province, Kenya.*

**Objectives:** In the last 48 months, the Coast Provincial General Hospital (CPGH) in Mombasa admitted about 18,000 patients in the department of paediatrics. Some of these admissions were congenital disorders.

Prune Belly syndrome is a congenital disorder defined by a characteristic clinical triad; urinary tract abnormalities, abdominal muscle deficiency and bilateral cryptorchidism.

Prognosis is quite favourable for the less severe types, best management is to do nothing surgically. Prophylactic antibiotics may be indicated for the urinary stasis.

**Methods:** Retrospective audit of the last two years (48 months) looking for patients that fit the clinical criteria of prune belly syndrome and describe their clinical characteristics.

**Results:** Five patients fitting the criteria of prune belly syndrome were identified. Age at presentation ranged from one day to 60 months. Three of the patients were identified at the neonatal unit having been admitted after birth for abdominal wall defects. The remaining two were admitted to the general Paediatric ward, one with sepsis and the other severe malnutrition.

One of the patients had other anomalies; a patent urachus and skeletal anomalies. Echocardiography was not done in any of the patients, as all patients were haemodynamically stable and also because of financial constraints

Average creatinine at presentation was 139ummol/l (range 40µmol/l - 420µmol/l)

Two of the patients are lost to follow up. Two have been referred to Kenyatta National Hospital (KNH) for surgically intervention because of gradually deteriorating renal function and one to the surgical team locally.

**Conclusions:** Prune belly syndrome appears to be a fairy common abnormality locally compared to international statistics of 1 in 35,000 to 1 in 50,000.

## Abstract# 874

**Phenotypic and Genetic Characterization of 13 Colombian Families with Bartter and Gitelman's Syndrome** J.J. Vanegas,[1,2] L.M. Serna-Higuita,[1,2] L. Urbano,[2] L.M. Betancur,[1] C.M. Medina,[1] A.M. Garcia-Cepero,[3] N. Pineda-Trujillo.[2] *[1]Medicine Faculty, Pontificia Bolivariana University, Medellin, Colombia; [2]Medicine Faculty, University of Antioquia, Medellin, Colombia; [3]Biological Research Corporation, Medellin, Colombia.*

**Objectives:** To determine the phenotypic and genetic characteristics of 13 colombian families with Bartter and Gitelman's syndrome.

**Methods:** 13 colombian families were phenotypically characterized. 2 SNPs were typed in 6 genes (*KCNJ1*, *SLC12A1*, *SLC12A3*, *BSDN*, *CASR, CLCNKB*) previously associated with the syndrome. Association, linkage and haplotype analyses were applied.

**Results:** 17 patients were analyzed. 47% with Gitelman's, 47% with neonatal Bartter's and 6% with classical Bartter. 38% of the children were consanguineous. Average age at diagnosis was: Neonatal Bartter 18.4 mo, classical Bartter 9.6 y and Gitelman 15.5 y. Main clinical symptoms were: polyhydramnios, preterm delivery, polyurea and hydroelectrolytic disturbances. All of the patients presented with hypokalemia and required treatment. Nephrocalcinosis was found in 33% of the patients with classical Bartter, 66% with neonatal Bartter.

It was found association to two SNPs in *KCNJ1 (p=0.0186)*. Haplotype analysis did increase the significance of association (p=0.007). This gene has been previously reported as associated with type II Bartter. In addition two sibs with Gitelman's presented clear inheritance implicating the SLC12A3.

**Conclusions:** This is the first study in Colombia looking for a genetic component of Bartter and Gitelman's syndrome. It is remarkable that only association to *KCNJ1* was found. It may correspond to phenotypic variability. Sequence analysis of both *KCNJ1* and *SLC12A3* genes is underway.

## Abstract# 875

**Urinary Levels of Transforming Growth Factor β1 and of Inflammatory Cytokines in Patients with Fetal Hydronephrosis** M.A. Vasconcelos,[1] M.C.F. Bouzada,[1] K.D. Silveira,[2] L.R. Moura,[1] F.F. Santos,[1] J.M. Oliveira,[1] F.F. Carvalho,[1] M.M. Teixeira,[2] A.C. Simoes e Silva,[1] E.A. Oliveira.[1] *[1]Pediatrics, Federal University of Minas Gerais, Belo Horizonte, Minas Gerais, Brazil; [2]Biochemistry and Immunology, Federal University of Minas, Belo Horizonte, Minas Gerais, Brazil.*

**Objectives:** The aim of this cross-sectional study was to identify noninvasive biomarkers of clinically significant uropathies in patients with antenatal hydronephrosis.

**Methods:** We have evaluated spot-urine levels of interleukin-6(IL-6), transforming growth factor-β1(TGF-β1) and tumoral necrosis factor-α(TNF-α) of 100 patients with intra-uterine hydronephrosis. Patients were divided into three groups: idiopathic renal pelvic dilatation (n=47), uropathies (n=35), and dysplastic kidneys (n=18).

**Results:** The most frequent uropathies were ureteropelvic junction obstruction (n=13) and vesicoureteral reflux (n=11). Dysplastic kidneys included multicystic dysplasia (n=14) and hypodysplasia (n=4). No significant differences of urinary TGF-β1, IL-6 and TNF-α levels were found in the comparison between the groups. Of 100 patients, 29 presented a reduction in DMSA uptake at the first renal scintigraphy. Both absolute urinary concentration of TGF-β1 and TGF-β1 levels standardized to urinary creatinine presented significant elevation in patients with reduced DMSA uptake in comparison to those with normal DMSA (p<0.05). Urinary concentrations of IL6 and TNF-α did not differ in the comparison between these groups.

**Conclusions:** Although urinary cytokine measurements seemed not to be useful as screening test for clinically significant uropathies, increased concentrations of TGF-β1 pointed out to renal damage.

## Abstract# 876

**How Long Should We Follow Children with Unilateral Multicystic Dysplastic Kidneys (MCDK)?** W. Hayes, A.R. Watson. *Children's Renal & Urology Unit, NUH NHS Trust, Nottingham, United Kingdom.*

**Objectives:** Current consensus is that unilateral MCDK is a benign condition with few patients suffering complications of urinary tract infection, proteinuria, hypertension or malignant transformation. Long-term follow-up has shown the persistence of the MCDK in some patients. We studied whether this was related to initial size of the MCDK. We also examined whether patients could be discharged if involution had occurred by 5 yrs.

**Methods:** Data were retrieved from a prospective regional registry of patients with MCDK between 1985 and 2009. Children were followed using a common protocol of investigation with follow up clinical assessments and ultrasound scans (USS) at 2, 5 and 10 years.

**Results:** Of 323 initial patients 249 had USS at 2 years, 180 at 5, 94 at 10 and 17 at 15 years follow up. Survival analysis of MCDK kidneys >5cm at birth showed persistence on USS at 10 years in 85% compared to 40% in those where the initial kidney size was <5cm ($p < 0.0001$).Of 180 patients followed to 5 yrs, 126 demonstrated compensatory hypertrophy of the contralateral kidney and 59 of the 126 were associated with complete involution of the MCDK. 20 of the 59 had been followed for 10 yrs with a mean eGFR of 88ml/min/1.73m$^2$ (range 46–108). None of the patients have had hypertension, significant proteinuria or developed malignancy.

**Conclusions:** MCDK kidneys are less likely to involute if they are large at birth. However the clinical progress appears to be benign and conservative management is justified. The data would suggest that up to 1/3 of patients could be discharged at 5 yrs with compensatory hypertrophy of the contralateral kidney in association with involution of the MCDK and benign clinical status.

## Abstract# 877

**Prenatal Diagnosis and Epidemiology of Multicystic Kidney Dysplasia in Europe** L. Winding,[1] E. Garne,[2] D. Wellesley.[3] *[1]Århus University Hospital, Skejby, Århus, Denmark; [2]Lillebaelt Hospital, Kolding, Denmark; [3]Princess Anne Hospital, Southampton, United Kingdom.*

**Objectives:** To describe prevalence, prenatal diagnosis and epidemiology of multicystic kidney dysplasia (MCKD) in Europe.

**Methods:** Data from a large European database for surveillance of congenital malformations (EUROCAT). The registries are based on multiple sources of information including live births, fetal deaths, terminations of pregnancy, prenatal diagnosis and gestational age at diagnosis. Included were all cases form 20 registries with MCKD born in 1997-2004.

**Results:** There were 1206 cases with MCKD, giving an overall prevalence of 3,78 per 10.000 births, with an increasing trend over the period.

Among all cases there were 698 male (58 %) and 471 female (39%). Similar sex distribution were seen when cases were divided into uni- and bilateral MCKD. The majority of cases were unilateral (76%) and of these most were live born (84 %) and had isolated renal malformation (79%). Of the bilateral MCKD the majority of the pregnancies were terminated (61%) and only half had isolated renal malformation (52%).

Time of diagnosis were prenatal for 1087 cases (87%) and the prenatal detection rate for all registries together were high (81-93%) with no trend seen over the years. There were a large regional difference in both prevalence (0,2 - 9,5 per 10.000) and in prenatal diagnosis.

**Conclusions:** Cases with unilateral MCKD were mainly live births, while pregnancies with bilateral cases often were terminated. There were large regional differences in prevalence in Europe. There seems to be an increasing prevalence of MCKD in Europe that cannot be explained by increasing prenatal detection rates.

## Abstract# 878

**Prospective Follow up of the Patients with Antenatal Hydronephrosis** F. Duzenli,[1] F. Yalcinkaya,[1] S. Fitoz,[2] Z.B. Ozcakar,[1] M. Ekim.[1] *[1]Pediatric Nephrology, Ankara University Medical School, Ankara, Turkey; [2]Radiodiagnostic, Ankara University Medical School, Ankara, Turkey.*

**Objectives:** The aim of this study was to define clinical features and follow up of the patients with antenatal hydronephrosis.

**Methods:** The study consisted of prospective data of 136 infants (27F, 109M) with antenatal hydronephrosis. Hydronephrosis was graded in accordance with the Society of Fetal Urology (SFU).

**Results:** The mean follow up period was 18 months (min 6 month, max 3 years). Antenatal hydronephrosis was detected with the ultrasonographies performed in the second and third trimester in 80% and 20% of the patients, respectively. Only 10% of the patients had a family history of urinary tract disease. Grade 2 and 3 hydronephrosis were seen in 56% and 43% of the patients, respectively. Half of the patients (50%) had unilateral hydronephrosis. Vesicoureteral reflux was detected in 14% of the patients. During the follow up antimicrobial prophylaxis was given to 36% of the study group. Urinary tract infection occured in 22% of the patients and renal scarring was observed in 8%. Surgical correction was performed in 10% of the patients.

**Conclusions:** Antenatal hydronephrosis, which was more frequently seen in boys, has a low incidence of urinary tract infection. However, surgical correction although rarely, might be indicated. Non- invasive longterm postnatal follow up should be justified in the majority of the patients.

## Abstract# 879

**Severe Form of Schimke Immuno-Osseous Dysplasia with Unilateral Kidney Agenesis** J. Zieg,[1] M. Balascakova,[2] J. Dusek,[1] N. Simankova,[1] E. Seemanová,[2] J. Lebl,[1] F. Fencl,[1] K. Blahova,[1] A. Krepelova.[2] *[1]Dpt. of Paediatrics, University Hospital Motol, Second Faculty of Medicine, Charles University, Prague, Czech Republic; [2]Dpt. of Biology and Medical Genetics, University Hospital Motol, Second Faculty of Medicine, Charles University, Prague, Czech Republic.*

**Objectives:** Schimke immuno-osseous dysplasia (SIOD) is a disease affecting many organ systems. Severe form of genetically confirmed SIOD with unique features is presented.

**Methods:** 3 year old girl with severe form of SIOD presented initially with growth failure, solitary kidney, nephrotic syndrome, T-cell immunodefficiency and characteristic physical features. Subsequently she developed hematologic abnormalities- rituximab resistant idiopatic trombocytopenic purpura (ITP) and central nervous system symptoms- reccurent transient ischemic attacks. To our current knowledge it is the first described association of SIOD, unilateral renal agensis and rituximab resistant ITP.

**Results:** Finally we analysed DNA and confirmed mutations in SMARCAL 1 gene. The patient is compound heterozygous for mutations c.2542G>T (p. Glu848X) in exon 17 and c,1439>T (p.Pro480Leu) in exon 8. According to mutation type- nonsense, missense, the patient was expected to have milder form of SIOD. Genotype-fonotype variability in SIOD is apparent.

**Conclusions:** SIOD affects many organ systems. Severe forms lead to death of patient in early childhood. Therapy of SIOD is only symptomatic. Our efforts to influence the course of SIOD associated ITP by rituximab and neurologic manifestations of this disease was unsuccessful. Molecular genetic testing can be done. Mutations our found in 50-60% of cases. Prenatal diagnosis is possible in these families.

## Perinatal Nephrology

## Abstract# 880

**(O-110)**

**Intrauterine Growth Restriction and Postnatal Overnutrition Affect the Proteomes of the Kidneys in Adult Rats** Q. Shen, H. Xu, J. Chen, H.-M. Liu, W. Guo. *Department of Nephrology and Rheumatology, Children's Hospital of Fudan University, Shanghai, China.*

**Objectives:** Intrauterine growth restriction (IUGR) is associated with hypertension, diabetes and chronic kidney disease in adulthood. Postnatal overnutrition following IUGR may be of pathogenic importance for the development of diabetes and cardiovascular disease. The aim of this study was to identify the possible pathogenesis of kidney disease in IUGR and the effect of postnatal overnutrition by comparative proteomic approach.

**Methods:** IUGR was induced in rats by isocaloric protein restriction in pregnant dams. IUGR pups were divided into two groups, fed either standard-protein diet (IUGR group) or high-protein diet (HP group). At the age of 12 weeks, kidney proteins were obtained from each group. 2-DE, staining, mass spectrometry and database searching were used.

**Results:** The differential proteomic expression analysis between IUGR and control group found 12 proteins had significantly differential expressions, which were transcription regulators including prohibitin and ribonuclease UK114, structural molecules including capping protein, enzymes and so on. Subsequently, the differential proteomic expression analysis between IUGR and HP group found 16 proteins had significantly differential expressions, which were transcription regulators including ribonuclease UK114 and NADH-ubiquinone oxidoreductase, structural molecules including ezrin and gamma-actin, enzymes and so on.

**Conclusions:** Data from this study may provide, at least partly, valuable experimental evidence of proteins involved in the pathogenesis of kidney disease in IUGR and the effect of postnatal overnutrition.

## Abstract# 881
**(O-111)**

**Urine Biomarkers of Acute Kidney Injury (AKI) Predict Mortality in Very Low Birthweight (VLBW) Infants** D.J. Askenazi,[1] R. Koralkar,[1] P. Devarajan,[2] C. Parikh,[3] S. Goldstein,[4] N. Ambalavanan.[1] [1]*Pediatrics, U of Alabama, Birmingham, AL, United States;* [2]*Pediatrics, U of Cincinatti, Cincinatti, OH, United States;* [3]*Medicine, Yale, New Haven, CT, United States;* [4]*Pediatrics, Baylor, Houston, TX, United States.*

**Objectives:** AKI is a strong predictor of mortality in VLBW infants. Novel urine biomarkers of AKI predict mortality in other populations. We evaluate the utility of 6 biomakers to predict mortality in VLBW infants.

**Methods:** Urine samples were collected on postnatal days 1 through 6 for 105 VLBW infants in whom 13 died at (age 26 +/- 6 days). The maximum concentration for each biomarker was compared between survivors and non-survivors.

**Results:** Neutrophil gelatinase-associated lipocalin (NGAL), kidney injury molecule 1 (KIM-1), interleukin 18 (IL-18) and osteopontin (OPN) levels were higher in non-survivors vs. survivors, while cystatin C and B2 Microglobulin (B2 Mgb) were not statistically different.

| | Survived ( n = 94 ) | Died (n =13 | p | ROC AUC |
|---|---|---|---|---|
| NGAL (ng/mL) | 376 ( 181,569) | 555 (300,1176) | <0.05 | .68 |
| KIM-1 (pg/mL) | 315 ( 146,611) | 384 ( 252,1215) | <0.01 | .66 |
| IL-18 (pg/mL) | 93 (54,353) | 114 (40,213) | <0.05 | .50 |
| OPN (ng/mL) | 229 (169,354) | 480 (280,631) | <0.05 | .78 |
| Cystatin C (ng/mL) | 1938 (570,4469) | 1884 ( 400,4588) | 0.39 | .47 |
| β2 Mgb(ug/mL) | 1.7 (1.1,2.5) | 1.7 ( 0.9 ,2.9 ) | 0.33 | .51 |
| BW (gm) | 1000 + 304 | 681 + 237 | < 0.01 | |

Median (25-75%) , AUC for Mortality

**Conclusions:** Urine AKI biomarkers predict survival in VLBW infants. Single or panel biomarkers may improve our ability to detect predict outcomes, and design intervention studies in this population.

**DISCLOSURE:** *Devarajan, P.*: Consultant, Abbott Diagnostics, Biosite Inc. *Parikh, C.*: Consultant, Abbott Diagnostics. *Goldstein, S.*: Grant/Research Support, Gambro Renal Products/Merck; Consultant, Gambro Renal Products/Baxter; Speaker's Bureau, Gambro Renal Products.

## Abstract# 882
**(O-112)**

**Urine Biomarkers Can Detect Acute Kidney Injury (AKI) in Very Low Birth Weight (VLBW) Infants** D.J. Askenazi,[1] R. Koralkar,[1] P. Devarajan,[2] C. Parikh,[3] S.L. Goldstein,[4] N. Ambalavanan.[1] [1]*Pediatrics, University of Alabama, Birmingham, AL, United States;* [2]*Depatrment of Pediatrics, University of Cincinatti, Cincinatti, OH, United States;* [3]*Department of Medicine, Yale School of Medicine, New Haven, CT, United States;* [4]*Department of Pediatrics, Baylor College of Medicine, Houston, TX, United States.*

**Objectives:** AKI is a strong predictor of mortality in VLBW infants (birthweight 500-1500g) Novel urine biomarkers predict AKI in others. We evaluate the utility of 6 urine biomarkers to predict AKI in VLBW infants.

**Methods:** Urine samples were collected on postnatal days 1 through 6 in 78 VLBW infants. Maximum concentration for each biomarker was determined. AKI is defined as a rise in serum creatinine (SCr) by 0.3 mg/dl within 48 hours or SCr rise by > 50% at any time.

**Results:** Urinary concentrations of neutrophil gelatinase-associated lipocalin (NGAL), kidney injury molecule-1 (KIM-1), Interleukin 18 (IL-18), and osteopontin (OPN) were increased in infants with AKI. No difference was seen in B2 Microglobulin or Cystatin C.

| | No AKI ( n = 60 ) | AKI (n =18 ) | p | ROC AUC |
|---|---|---|---|---|
| NGAL (ng/mL) | 349 ( 168,651) | 562 (358,847) | <0.05 | .65 |
| KIM-1 (pg/mL) | 313 ( 129,592) | 711 ( 307,1190) | <0.01 | .73 |
| IL-18 (pg/mL) | 75 (26,385) | 192 ( 87,810) | <0.05 | .67 |
| Osteopontin (ng/mL) | 214 (136,329) | 379 ( 231,536) | <0.05 | .72 |
| Cystatin C (ng/mL) | 1776 (335,4560) | 2884 ( 1373,4545) | 0.39 | .58 |
| B2 Microglobulin (ug/mL) | 1.6 (0.9,2.4) | 1.7 ( 1.1,3.3 ) | 0.33 | .59 |

**Conclusions:** Candidate biomarkers are elevated in VLBW infants with AKI and may improve our ability to detect AKI, predict outcomes, and design interventions in this population.

**DISCLOSURE:** *Devarajan, P.*: Consultant, Abbott Diagnostics/Biosite, Inc. *Parikh, C.*: Grant/Research Support, Abbott; Consultant, Abbott. *Goldstein, S.L.*: Grant/Research Support, Gambro Renal Products/Merck; Consultant, Baxter Healthcare/Gambro Renal Products.

## Abstract# 883
**(O-120)**

**Outcome and Prognosis for Prenatally Diagnosed, Isolated Echogenic Kidneys from a Single Tertiary Centre over a 10 Year Period** S. Edwards,[1] N. West,[2] J. Dudley,[2] T. Overton.[1] [1]*Women and Children's Health, St Michael's Hospital Fetal Medicine Unit, University of Bristol, United Kingdom;* [2]*Child Health, Bristol Royal Hospital for Children, Bristol, United Kingdom.*

**Objectives:** To establish clinical outcomes of antenatally detected isolated, fetal echogenic kidneys to aid prenatal counselling.

**Methods:** The database of a UK tertiary fetal medicine unit was used to identify all cases of fetal echogenic kidneys from 1998-2008. Cases with renal macrocysts, hydronephrosis and other fetal anomalies were excluded, and antenatal renal size was recorded. Paediatric notes were reviewed to identify clinical diagnosis and renal function.

**Results:** 17 cases of isolated renal echogenicity were identified. 2 were lost to follow up and 1 resulted in stillbirth. Postnatal renal investigations found 4 patients with no abnormality, 1 with autosomal dominant polycystic kidney disease (with a family history), 2 with autosomal recessive polycystic kidney disease (in the same family), 2 with HNF1Beta mutation, 1 with Bardet-Biedel syndrome (with polydactyly), 2 with unilateral multicystic dysplastic kidney, 1 with a dysplastic kidney which involuted in pregnancy and 1 with a unilateral dysplastic kidney. There was 1 neonatal death, 2 cases with chronic kidney disease and 2 patients with hypertension but normal renal function.
In the 10 cases with postnatally confirmed disease 8 had kidneys >97$^{th}$ centile prenatally. Only 1 of the 4 normal patients had kidneys >97$^{th}$ centile prenatally.

**Conclusions:** Isolated echogenic kidneys are an uncommon prenatal finding and overall prognosis appears to be good. Family history and detailed ultrasound for extra-renal anomalies are important and kidney size >97th centile appears to predict worse outcome.

## Abstract# 884

**The Significance of Fetal Renal Pelvic Dilatation as a Predictor of Postnatal Outcome** A.I.M. Al-Shibli, F. Chedid, H. Mirgani, W. Al Safi, M.K. Al Bassam. *Pediatrics, Tawam Hospital, Al Ain, United Arab Emirates.*

**Objectives:** To improve parents counseling and help in better understanding the clinical significance of FRPD.

**Methods:** We analyzed data from all infants who were diagnosed with FRPD during intrauterine life from January 1, 2005 to February 29, 2008 at Tawam Hospital, Al Ain, UAE.
FRPD was graded according to the sonographic anterior-posterior diameter (APD) measurement of fetal renal pelvis.

**Results:** Of 89 FPRD (64 patients), 36% had normal postnatal ultrasound, 22.5% significant uropathy and 41.5% had isolated hydronephrosis. Severe FRPD (≥15 mm) predicted significant postnatal uropathy with a sensitivity of 65% and a specificity of 98.6%. Moderate FRPD (≥10 mm) increased the sensitivity to 95% but decreased the specificity to 60.9%, mild FPRD (5-10 mm) was seldom (4%) associated with significant postnatal pathology.

Correlation between the severity of fetal renal pelvic dilatation and postnatal outcome.

| Postnatal diagnosis | Mild | Moderate | Severe | Total |
|---|---|---|---|---|
| Normal | 20 (46.5%) | 11 (34.4%) | 1 (7.2%) | 32 (35.9%) |
| Isolated hydronephrosis | 22 (51.2%) | 15 (46.8%) | 0 (0%) | 37 (41.6%) |
| Pelvi-uretric junction obstruction | 1 (2.3%) | 4 (12.5%) | 6 (42.8%) | 11 (12.5%) |
| Vesico uretric reflux | 0 (0%) | 1 (3.1%) | 4 (28.5%) | 5 (5.6%) |
| Posterior urethral valve | 0 (0%) | 1 (3.1%) | 3 (21.5%) | 4 (4.4%) |
| Total | 43 (100%) | 32 (100%) | 14 (100%) | 89 (100%) |

**Conclusions:** Third trimester fetal renal pelvis measurement is useful in predicting postnatal outcome. Patients with sever and moderate RPD had a high prevalence of uropathy, although they rarely needed surgical intervention and the renal pelvic dilatation tended to resolve spontaneously.

## Abstract# 885

**Perinatal Factors Associated with Acute Kidney Injury in Preterm Infants – A Matched Case Control Study** S. Bhojani, J. Banerjee, L Holmqvist, A. Shrivastava. *Paediatrics, Southend University Hospital, Southend on Sea, United Kingdom.*

**Objectives:** Acute kidney injury (AKI) is an established contributing factor of morbidity and mortality in preterm infants but the influence of perinatal factors on AKI is unclear. This study aimed to evaluate the perinatal risk factors in infants born before 32 weeks gestation with AKI.

**Methods:** Data was collected retrospectively from a level 2 neonatal unit at a District General Hospital in the UK identifying all preterm infants born between January 2007 and January 2010. Infants with AKI were identified using the Acute Kidney Injury Network criteria. Each case was matched by birth weight (BW) (±100gms) and gestational age (GA) (± 1week) with up to 2 controls. Data on perinatal factors and serum creatinine was collected from the case notes for the first week of life.

**Results:** 55 cases were identified and matched to 65 controls.

Perinatal Factors

| | Case | Control | P value |
|---|---|---|---|
| GA (weeks) | 29.2±1.7 | 29.5±1.7 | 0.44 |
| BW (grams) | 1293±312 | 1337±312 | 0.44 |
| Antenatal steroids | 43 (78.2%) | 61 (93.8%) | 0.01 |
| Cord pH | 7.3±0.01 | 7.3±0.02 | 0.46 |
| 5 min Apgar | 8.21±0.27 | 8.73±0.15 | 0.1 |
| Ventilation | 39 (73.6%) | 27 (41.5%) | 0.001 |
| Infection | 22 (40%) | 16 (24.6%) | 0.08 |
| Inotropes | 12 (25%) | 7 (11.9%) | 0.12 |

Cord pH, Apgar±S.E, others S.D, column%

Antenatal steroids were found to make significant difference with OR of 0.23 (95%0.07-0.78,p 0.01). Infants needing ventilatory support were more likely to have AKI (p 0.001).

**Conclusions:** Antenatal steroids are likely to have a protective effect against AKI in preterm infants. Large prospective randomised studies are needed to confirm the relation between antenatal steroids and AKI.

## Abstract# 886

**Influence of Indomethacin (IN), Ibuprofen (IB), and/or Furosemide (FU) on Aquaporin Gene Family in Kidneys of Neonatal Rats** N. Chorny,[1] J. Sharma,[1] H. Tawadrous,[1] M.J. Schoeneman,[1] A. Mongia,[1] A. Nada,[1,4] G. Valencia,[2] J.V. Aranda,[2] C. Cai,[2] D. Kumar,[2] K.D. Beharry.[3]
*[1]Pediatric Nephrology, SUNY Downstate Medical Center, Brooklyn, NY, United States; [2]Neonatology, SUNY,Downstate Medical Center, Brooklyn, NY, United States; [3]Neonatology, University of California, Irvine, CA, United States; [4]Pediatric Nephrology, University of Alexandria, Alexandria, Egypt.*

**Objectives:** We examined effect of IN,IB and/or FU on AQP gene family in neonatal rat kidney.

**Methods:** At birth P0, neonatal rats(12/group) were randomly assigned to receive IP injections:1) 0.2 mg/kg IN on P0 and 0.1 mg/kg on P1,2; 2) 10 mg/kg IB on P0 and 5 mg/kg on P1,2; 3) 5 mg/kg FU on P0,1,2; 4) IN+FU on P0,1,2; 5) IB+FU on P0,1,2; or 6) same volume saline (S) on P0,1,2. PCR examined expression of AQP gene family in kidneys at P3.

**Results:** IB upregulated all AQP genes. IN and FU upregulated all AQP genes except 2 and 12. FU+IB downregulated AQP2,8,12. FU+IN downregulated AQP1-3,8,9,12. AQP4 gene expression was increased 30fold by IN and 25fold by IB. This effect was blunted with FU, FU+IN and FU+IB,as with AQP5,9. IB upregulated all AQPs similarly in males and females. IN and FU were gender-specific. In males IN downregulated AQP2,12. FU downregulated AQP1,2 3. IN+FU downregulated AQP2-4,9. IB+FU downregulated AQP2,9. In females, IN downregulated AQP12; FU downregulated AQP2,3,9. IN+FU downregulated all except AQP11,7,8. IB+FU downregulated AQP1,2,8,12.

**Conclusions:** IN&IB consistently downregulated AQP2 suggesting that diuresis by FU involves regulation of AQP2 gene expression. Robust upregulation of AQPs with NSAIDs may be related to decreased urine output.

## Abstract# 887

**Ontogeny of Aquaporin Genes in Newborn, Suckling and Weanling Rat Kidneys** N. Chorny,[1] H. Tawadrous,[1] J. Sharma,[1] A. Nada,[1,4] M. Schoeneman,[1] C. Cai,[2] D. Kumar,[2] J. Aranda,[2] G. Valencia,[2] K. Baherry.[2,3]
*[1]Pediatric Nephrology, SUNY Downstate Medical Center, Brooklyn, NY, United States; [2]Neonatology, SUNY Downstate Medical Center, Brooklyn, NY, United States; [3]Neonatology, University of California, Irvine, NY, United States; [4]Pediatric Nephrology, University of Alexandria, Alexandria, Egypt.*

**Objectives:** Aquaporins (AQPs) belong to superfamily of membrane proteins that control transfer of water and small solutes across cellular membranes.We examined the ontogenic profiles of the AQP gene family in kidneys of newborn, suckling and weanling rats.

**Methods:** Rat kidneys were collected at birthP0,4,7,14, 21 for mRNA expression of AQP1-9,11,and12 using PCR. Gender differences were determined at P14,P21.

**Results:** Compared to P0, AQP1, 4, and 5 were robustly expressed at birth and increased with advancing age from 24fold, 52fold, and 19fold at P4, respectively, to 300fold at P21 for AQP1 and 5. AQP4 peaked at P14, declined at P21. At P14, AQP1 expression was 30% less in males, but by P21, no differences were seen. AQP2 was expressed at birth but progressively increased until P14, then declined at P21. AQP3 was consistently elevated from birth to P21, but highest expression was P7. AQP6 was expressed at birth and P4, but was not seen at P7 to 21. AQP7, 9, and 12 mRNA expression was present at birth, did not change with age. AQP8 was not present at birth and P4, mildly expressed at P7 to 21. AQP11 was not detected.

**Conclusions:** AQP1,2,4, and 5 mRNA are expressed at birth but gene expression dramatically increases during suckling in rats, crucial for water homeostasis during kidney development. Data emphasize importance of lactation for kidney development and function.

## Abstract# 888

**Risk Factors Effects Associated with Development of Acute Kidney Injury in Neonates According to the pRIFLE Criteria** d.P. Fonseca L,[1,2] A. Pereyra M,[1,2] P.B. Mandeville,[2] F. Escalante.[1,2]
*[1]Pediatric Nephrology, Hospital Central Ignacio Morones Prieto, San Luis Potosi, Mexico; [2]University of San Luis Potosi, San Luis Potosi, Mexico.*

**Objectives:** To analyze the frequency and association of AKI risk factors with pediatric RIFLE (pRIFLE) criteria in neonates.

**Methods:** Retrospective, observational study. From 340 admissions to NICU, 140 files were randomized, 100 fulfilled inclusion criteria. AKI cases were identified as having high serum creatinine and/or pRIFLE criteria. Multivariate logistic regression and polythetic analyses were performed with R 2.10.1 software.

**Results:** AKI was identified in 47% of the patients, 39% with pRIFLE criteria. In the polythetic analysis sepsis (p=0.005), gestational age (p=0.02) (figure1) asfixia (p=0.01), oliguria (p<0.001), Apgar at 1 (p=0.007) (figure 2) and 5 minute (p=0.007) were significant.

**Figure 1. Gestational age effect on pRIFLE**

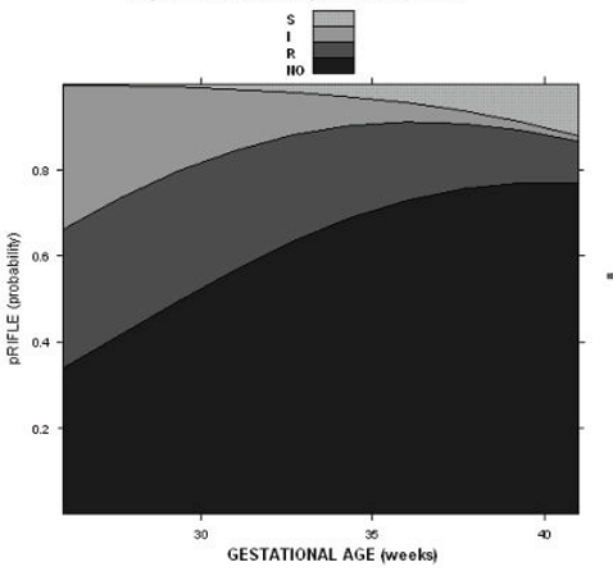
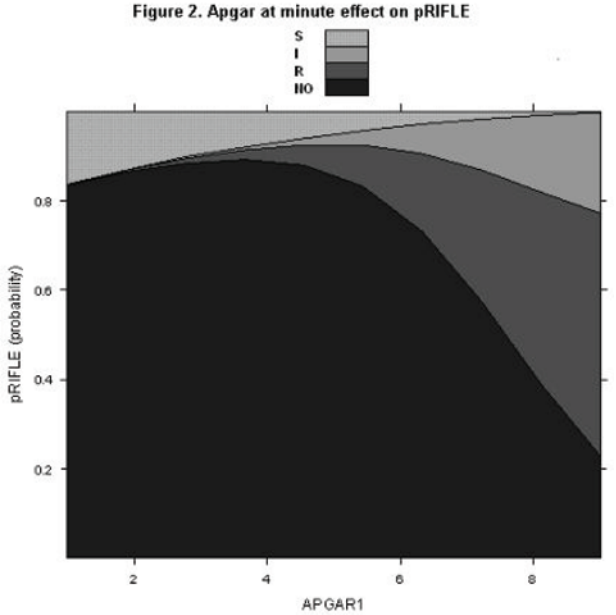



**Figure 2. Apgar at minute effect on pRIFLE**




**Conclusions:** pRIFLE criteria is useful in evaluating AKI in newborn. The effects of the risk factors are different for each criteria.

## Abstract# 889

**Risk Factors Associated with the Development of Acute Kidney Injury in Critically Ill Newborn Infants** d.P. Fonseca L,[1,2] A. Pereyra M,[1,2] P. Mandeville,[2] F.J. Escalante.[1,2] *[1]Pediatrics, Hospital Central Ignacio Morones Prieto, San Luis Potosi, Mexico; [2]School of Medicine, University of San Luis Potosi, San Luis, Mexico.*

**Objectives:** Determine the possible associations between the development of acute kidney injury (AKI) in critically ill newborns and perinatal factors.

**Methods:** The study was designed as a retrospective, observational study. From 340 admissions to NICU from January 1 to December 31, 2008, 140 files were randomized, and 100 fullfilled inclusion criteria. Cases of AKI were identified as having high serum creatinine (values of > 1.33 mg/dl and > 1.0 mg/dl in patients of < 33 weeks and ≥ 33 weeks respectively), and/or pRIFLE criteria. Univariate and multivariate logistic regression analyses were performed with R 2.10.1 software.

**Results:** AKI was identified in 47%, mean maternal age in AKI and non-AKI were 24.38 ± 6.79 and 25.56 ± 7.70 years (p<0.05), maternal urinary tract infection in 38.3% and 41.5% (p = 0.007), mean gestational age 32.59 ± 3.78 and 33.57 ± 2.95 weeks (p = 0.03), need of neonatal reanimation in 55.3% (p = 0.007), OR 3.28 (CI 95% 0.95 – 12.25). Mean Apgar score at 1 minute 6.42 ± 1.91 versus 7.18 ± 1.42 (p < 0.001), OR 1.81 (CI 95% 1 – 3.06) and 5 minute 7.57 ± 1.52 versus 8.47 ± 0.89 (p < 0.001). Sepsis was present in 76.6% (<0.001) and phototherapy in 59.5% (p = 0.008) of AKI patients. Length of stay was 29.9 ± 31.84 days in AKI (< 0.001). There were 8 deaths in AKI (p = 0.02).

**Conclusions:** There was a larger AKI frequency than expected and a lower Apgar score at 1 minute was a significant risk factor for AKI. It's very important to detect AKI neonatal given the high risk in this population of developing chronic kidney disease later.

## Abstract# 890

**Surviving after Multi-Organ Dysfunction (MOD) and Extreme Azotaemia – A Neonatal Case Report** H. Georgaki-Angelaki,[1] N. Lipsou, C. Petropoulou,[2] N. Stergiou, M. Anagnostakou.[2] *[1]Pediatric Nephrology, "Agia Sophia" Children's Hospital, Athens, Greece; [2]2nd Neonatal Intensive Care Unit2, "Agia Sophia" Children's Hospital, Athens, Greece.*

**Objectives:** In neonates<28 days old MOD is an important risk factor for acute renal injury and carries a poor prognosis.A severely ill neonate is presented who survived following PD despite MOD and extreme azotaemia.

**Methods:** A 12day old full-term neonate,BW 4.3Kgr,admitted to ICU with a 2day history of bloody stools and breast feeding refusal.Hyperbilirubinaemia,18.6mg/dl,was reported on day 4 of life treated by phototherapy.On admission the baby was in lethargy, dehydrated, and feverish.BP was 120/80mmHg.No urine production was reported for the last six hours.

**Results:** Laboratory test revealed leukocytosis(30.99x$10^3$/µl, netrophils 64.6%) procalcitonin109 ng/ml (normal<0.5)urea395mg/dl,creatinine8.33mg/dl,glucose12mg/dl,urates 48.5mg/dl,$Na^+$174mmol/L,$K^+$7.5mmol/L, $HCO_3^-$5mmol/L,pH7.29,Phosphate14.9mg/dl,SGOT 744U/L,SGPT 376U/L,γ-GT 133U/L.Ultrasonography shown two edematous,hyperechogenic kidneys. Attempts for renal perfusion restoration with appropriate fluids and inotropic agents plus conservative management of metabolic derangements failed and six hours post admission PD treatment was conducted.In two weeks urine production started,renal function improved remarkable,and creatinine decreased (2.8 mg/dl). Three weeks post admission PD discontinued.At the latest follow-up,9 months old, urea was 62 mg/dl and creatinine1.3mg/dl.

**Conclusions:** In a life threatening situation with MOD and extremely high levels of azotaemia early therapeutic intervention by PD changed the otherwise poor prognosis.To our knowledge not a similar case in a neonate is reported.

## Abstract# 891

**Acute Renal Failure in Neonates** H. Georgaki-Angelaki,[1] K. Naoum, F. Anatolitou, A. Kapoyiannis, M. Anagnostakou. *[1]Pediatric Nephrology, "Agia Sophia" Children's Hospital, Athens, Greece; [2]2nd Neonatal Intensive Care Unit, "Agia Sophia" Children's Hospital, Athens, Greece.*

**Objectives:** Major predisposing risk factors of acute renal failure (ARF) in neonates are congenital urinary tract (CUT) or cardiovascular system (CVS) abnormalities and acquired conditions such as perinatal asphyxia (PA), septicemia (S) and respiratory distress syndrome (RDS).Aim of the study: To retrospectively determine the incidence, major causes, treatment and outcome of ARF in one of our neonatal intensive care units (ICU) over a period of eight years.

**Methods:** Medical records of neonates in ARF were reviewed according to the following criteria:1) Increased for age blood urea nitrogen (BUN) and creatinine (>50% above baseline level), 2) Concomitant reduction in urine output,< 0.5-1 ml/Kgr/hour. Analyzed data included gestational age (GE),birth weight (BW),causes,treatment,and outcome.

**Results:** Of 3199 on ICU hospitalised neonates, 39 ARF cases were identified (1.2%). According to their GE, neonates were grouped into three categories: Group I, GE :< 32 w, n =15, Group II, GE: 32-35 w, n=7, GE III: >37 w, n=17. Major causes was RDS (100%) and S 73.3% in group I, CUT42.8%, CVS 28.5%, PA 42.8%, S 28.5% in group II, CUT 17.6%, CVS 35.2%, PA 35.2%, S 35.2% in group III. Treatment by Peritoneal Dialysis (PD) required 3/15 (20%) in group I and 5/17(29.4%) in group III. Outcome: 11/15 (73.3%) deaths in group I, 4/7 (57.14%) group II and 8/17(47%) group III.

**Conclusions:** Among a high risk group of neonates the incidence of ARF, by using certain criteria, was only 1.2%. Although no statistically significant it seems that prematurity remains of the most important predisposing factors affecting mortality.

## Abstract# 892

**Renal Impairment in ELBW Infants Can Be Defined on Day 3** I. George, E. Levtchenko, M. Rayyan, K. Allegaert, D. Mekahli. *Pediatrics, University Hospital Gasthuisberg, Leuven, Belgium.*

**Objectives:** Due to successful neonatal care, survival of extremely low birth weight (ELBW) neonates continues to increase. Recent data showed that very low birth weight (VLBW) neonates are at high risk of acute renal failure, however data on renal function of ELBWgroup is scare. Furthermore, serum creatinine (Scr) remains a widely used clinical tool to estimate GFR, however normal values in this population are not yet defined. We aim to assess Scr of this group and define predictors of renal impairment.

**Methods:** Retrospective chart review of ELBWneonates assessed in a single centre between 2000-2005. Neonates with congenital renal abnormalities and/or death ≤7 days were excluded. Neonatal Scr (mg/dl) was analysed according to prenatal, peripartal and postnatal variables. Results are described as median (range).

**Results:** In 151 ELBW (66 females), gestational age(GA) at birth was 27 (23-33)w, birthweight(BW) was 810 (330-1000)g with 34% SGA. Peak Scr level was reached on day 3:1.3 (0.4-2.5). We defined severe renal impairment as peak Scr on day 3>75$^{th}$ percentile. Thirty seven (24.5%) neonates had severe renal impairment. Their Scr remains higher on day 14 and 28 (p<0.001).They had lower BW, lower GA, more ibuprofen and steroid exposure and longer ventilation period. Multivariate analysis confirmed that GA and ventilation period (p<0.0001) were significant risks factors, while SGA was not (p=0.17).

**Conclusions:** ELBW neonates are at risk for severe renal impairment in the neonatal period with similar risks factors as the VLBW peers. Our data showed that peak Scr on day 3 was a good predictor for renal impairment in this weight category. Prospective studies for long term renal function of this group are needed.

## Abstract# 893

**Has Maternal Preeclampsia an Influence on Neonatal Blood Pressure Level?** M. Reveret,[1] B. Marin,[2] A. Bedu,[1] V. Guigonis.[1] *[1]Pediatrics, CHU, Limoges, France; [2]Statistics and Epidemiology, CHU, Limoges, France.*

**Objectives:** Recent data give evidence that preeclampsia could be an antibody mediated disease. Therefore, pathogenic antibodies (i.e. directed against angiotensine II receptor type 1) (AT1R-AA) could have an impact in neonates. It has been reported that blood in neonates less than 1350g of birth weight and 29 SG and born from preeclamptic mothers, blood pressure was significantly higher than in controls (neonates born from normotensive mothers). We conducted a retrospective study in premature newborn - older than 29SG and with a weight higher than 1350g - in order to compare blood pressure levels whether their mother presented with preeclampsia or not.

**Methods:** Blood pressure levels of 24 premature newborns from preeclamptic mothers and their matched controls were retrospectively studied and compared during their first three days of life.

**Results:** Mean gestational age (31.1 vs. 31.2 SG) and birth weight (1774 vs. 1865g) were not different between the two groups. The only difference was the delivery mode with more caesarian section in the preeclampsia group. Diastolic blood pressure was significantly higher in the preeclampsia group of newborn than in the control group (mean difference: + 3.13 mmHg, p<0.05). No difference was present between the two groups considering systolic or mean blood pressure.

**Conclusions:** Even if not clinically relevant, these data seem to confirm that newborns from preeclamptic mothers are susceptible to have higher blood pressures than newborns from normotensive mothers. This difference could be explained by the transplacental transfer of AT1R-AA to fetuses.

## Abstract# 894

**Maturation of Renal Functions & Renal Volume in Normal and Growth Restricted Neonates** A. Gulati, I. Ado, R. Agarwal, V. Paul, A. Gupta, R. Lakshmy, A. Bagga. *All India Institute of Medical Sciences, Delhi, India.*

**Objectives:** There is limited data on anatomic & functional renal development in growth restricted neonates.We compared glomerular function on day 3 in appropriate for date (AFD) & small for date (SFD) neonates.Glomerular,tubular functions & renal volumes were also assessed in first 6-wks.

**Methods:** After consents, 103 neonates (32-41 wk gestation) delivered at this center included. Serum creatinine, cystatin, Schwartz GFR, urine osmolality & fractional excretion of sodium (FENa), potassium (FEK) measured starting from 12-24 hr of life until 6 wk. Renal volume assessed by ultrasonography at 2- 4 wk after birth.

**Results:** No significant difference in serum creatinine (mg/dl) observed in preterm AFD, term SFD& term AFD at day 3 (0.85±0.3, 0.80± 0.3 & 0.82±0.2 respectively, P=0.9).

Renal function, volume at 6 wk

| | Preterm AFD (n=26) | Term SFD (n=37) | Term AFD (n=40) | P |
|---|---|---|---|---|
| GFR (ml/min/1.73m2) | 27±8 | 30±7 | 44±10 | <0.01 |
| Serum cystatin (ng/ml) | 1761±577 | 1716±345 | 1701±364 | 0.9 |
| Urine osmolality*(mOsm/kg) | 400 | 317 | 408 | 0.2 |
| FENa*(%) | 0.6 | 0.3 | 0.5 | 0.3 |
| FEK*(%) | 10.8 | 13.6 | 17.2 | 0.5 |
| Combined kidney volume (mm3) | 9±2 | 10±2 | 13±3 | <0.001 |

Mean ±SD;*Median values

In each group, GFR correlated positively to postnatal age (P<0.01).Levels of cystatin did not change (P=0.8) while urine osmolality rose with postnatal age (P<0.001).On multivariate analysis, smaller kidney volume correlated significantly with prematurity and low weight for gestational age.

**Conclusions:** Renal function and volume in preterm AFD and term SFD babies is compromised. Follow up studies are required to determine implications on long term renal outcome.

## Abstract# 895

**The Roles of Antenatal and Postnatal 2nd Week USG on Prediction of Severe Hydronephrosis** M. Bayram, A. Soylu, D. Alaygut, B. Kasap, M. Turkmen, S. Kavukcu. *Pediatrics, Dokuz Eylul University Medical Faculty, Izmir, Turkey.*

**Objectives:** To compare the predictive powers of antenatal USG and postnatal 2nd week USG for obstructive uropathy and/or VUR.

**Methods:** Patients with antenatal hydronephrosis were evaluated by serial postnatal USG (2nd week, 3-6-12-24 months) and, if needed, dynamic renal scintigraphy and/or voiding cystourethrography. Each renal unit (RU) was classified as normal, non-obstructive dilatation, obstructive uropathy or VUR. The predictive powers of antenatal and postnatal 2nd week USG for obstructive uropathy ± VUR were determined according to these data.

**Results:** Evaluations were performed on 92 patients (184 RU) having both antenatal USG (hydronephrosis in 120 RU) and postnatal USG (hydronephrosis in 113 RU). The diagnoses of RU with vs without hydronephrosis in antenatal USG were normal (1 vs 48), non-obstructive dilatation (70 vs 12), obstructive uropathy (33 vs 2) and VUR (16 vs 2), respectively (p<0.001) (OR 10, sensitivity 93%, specificity 46%, PPV 41%, NPV 94% for obstructive uropathy ± VUR). The diagnoses of RU with vs without hydronephrosis in postnatal USG were normal (8 vs 41), non-obstructive dilatation (59 vs 23), obstructive uropathy (33 vs 2) and VUR (13 vs 5), respectively (p<0.001) (OR 6, sensitivity 87%, specificity 49%, PPV 41%, NPV 90% for obstructive uropathy ± VUR).

**Conclusions:** Antenatal and postnatal 2nd week USG were sensitive but not specific for the prediction of significant hydronephrosis. Furthermore, presence of hydronephrosis in antenatal or postnatal USG did not differ with regard to the prediction of final diagnosis.

## Abstract# 896

**Renal Tubular Dysgenesis: Clinical Case** I. Kazyra, J. Grygorenko, A. Sukalo, T. Letkovskaja, S. Bayko, V. Savosh, N. Tur. *Pediatrics, Belarus State Meducal University, Minsk, Belarus; Chair of Pathology, Belarus State Medical University, Minsk, Belarus.*

**Objectives:** We report the first case of intravital diagnosis of renal tubular dysgenesis in neonate in our Republic of Belarus probably associated with Nimesulide using in the second and third trimester of pregnancy.

**Methods:** A 2880g male neonate of 36-37 weeks gestation was born to a 28 year-old prima gravida mother (non-consanguineous couple). The pregnancy was complicated by the intrauterine fetal hypoxia and oligohydramnios. There was a history of acute respiratory illness and urogenitaly tract infection in the first trimester of pregnancy. The baby was delivered by cesarean section because of mother's symphysitis. Symphysitis was diagnosed on 29 weeks gestation and she was treated by Nimesulide. Third trimester sonographic (US) demonstrated of oligohydramnios with structurally normal kidneys. At delivery Apgar scores were 6 and 7 at 1 and 5 min respectively. He was intubated after 3 hours after birth because of poor oxygenation and transferred to our hospital for evaluation of anuria at 3 days-old age.

**Results:** Our examination revealed nornal face, peripheral edema, high blood urea and creatinin, acidosis, anemia. His kidneys were normal on size and shape on US, there was no evidence of thrombosis and urological problems. The peritoneal dialysis was started. At the age of 11 days-old kidney's biopsy was undertaken. No markers of proximal tubular function were detected immunohistochemically (AB to EMA).

To date, our patient 1 year and 10 months, he is on peritoneal dialysis, and continues to be in anuria.

**Conclusions:** We should be extremely careful in the prescription of drugs during pregnancy.

## Abstract# 897

**Long-Term Follow-Up of Antenatal Oligohydramnios of Renal Origin (ROH)** I. Klaassen,[1] S. Schmidtke,[1] G.F. Laube,[2] D.E. Mueller-Wiefel,[1] M.J. Kemper.[1,2] *[1]Pediatric Nephrology, Universitiy Children's Hospital, Hamburg, Germany; [2]Pediatric Nephrology, University Children's Hospital, Zuerich, Switzerland.*

**Objectives:** Prognosis of ROH has been regarded as unfavorable, although clinical data are scarce. Due to progress in neonatal intensive care and the treatment of infants with chronic kidney disease, the overall prognosis of these children has improved considerably. The aim of this study was the evaluation of complications and long-term follow-up in patients with ROH.

**Methods:** Data on 39 fetuses (26 male, 13 female) with ROH from 1990 to 2010 were evaluated.

**Results:** Primary diseases included urinary tract malformations (n=26), ARPKD and ADPKD (n=9) and other (n=4). Of 9 non-survivors (23%), 6 died within the neonatal period. 27 patients required mechanical ventilation (16 with associated pneumothorax). The surviving 30 children have a current median age of 5.6 (0,1-16) years. All developed CKD, which could be managed conservatively in 14 patients (median GFR 50.1 (range 19.9–130) ml/min/1.73m²). 16 patients reached ESRD at a median age of 3 months (range 2 days to 8.2 years); two received a preemptive kidney transplantation (KT), and 14 started peritoneal dialysis. 11 of these patients underwent successful KT at a median age of 3.1 (range 1.1-12) years. Two of them had a combined hepatorenal transplantation due to ARPKD. Cognitive and motor development was normal in 25 of 30 patients (83%) and showed a delay in 5 children.

**Conclusions:** In ROH long-term prognosis is encouraging and range from CKD stage 1 to combined hepatorenal transplantation. The rate of perinatal problems and complications is high and may increase, since more severely affected fetuses are actively treated.

## Abstract# 898

**One Year Follow-Up Results of Patients Prenatally Diagnosed Hydronephrosis** S. Yel, H.M. Poyrazoglu, Z. Gündüz, S. Tülpar, R. Düsünsel. *Pediatric Nephrology, Erciyes University Medical Faculty, Kayseri, Turkey.*

**Objectives:** We aimed to determine the etiological reasons and frequency of prenatal determined hydronephrosis, and to evaluate the association between some parameters and renal functions during study.

**Methods:** Forty-eigth patients born between April 2006-October 2007 with prenatal hydronephrosis were followed prospectively. The urinary ultrasound scan and renal functioning tests were performed on day 3-7 of life and repeated on week 4-6, months 3, 6 and 12.

**Results:** In 29.3% of all 76 renal units with prenatal hydronephrosis, transient hydronephrosis was diagnosed. Ureteropelvic junction obstruction was the most common cause of prenatal hydronephrosis. Thirty-two patients (66.6%) had urinary tract infection during study. Eshericia coli was the most common cause of urinary tract infection. There were negative correlations between GFR of at the end of study and fractione $K^+$ excretion at the 3th month and 1st year. There were negative correlations between GFR of at the end of study and fractione $Mg^{++}$ excretion at the 3th and 6th months. There were positive correlation between GFR of at the end of study and tubular reabsorbtion of phosphate 1st year and negative correlation between GFR of at the end of study and serum BUN and creatinine levels 1st year. Only one patient had stage 2 chronic renal disease at the end of study.

**Conclusions:** Transient hydronephrosis is one of the most important reasons of prenatal hydronephrosis. Tubular functioning tests may be impaired early stages. The patients with prenatal hydronephrosis must be followed-up closely and the episodic evaluation of tubular functions may predict renal damage before renal failure development.

## Abstract# 899

**Omega 6 PREVENTS Hypertension and Renal Damage Induced by Intrauterine Exposure to Cadmium** J.L. Reyes, M.P. Avendano-Huerta, D. Martin, M.C. Namorado, O.C. Barbier. *Physiology, Toxicology and Pharmacology Depts., Center for Research and Advanced Studies. National Polytechnic Institute (Cinvestav-IPN), Mexico, Distrito Federal, Mexico.*

**Objectives:** Acute exposure to cadmium induces nephrotoxicity. Chronic Cd intoxication leads to renal damage and arterial hypertension (HT), in humans and animal models. Tabaquism is a chronic exposure source to Cd and long half-life (10 to 30 years in humans) accounts for in utero exposure to Cd in offsprings to smoking women. In this study we analyze the time course of renal damage and HT during postnatal life in offsprings from rats gestationally exposed to Cd and beneficial effect of omega 6.

**Methods:** Pregnant Wistar rats were allocated in 4 groups: a) control, b) borrage oil (source of omega 6, 10 g/kg po, daily), c) Cd (500 mg/kg bw, po, daily), and d) Cd plus borrage oil (same doses as groups b and c). Offsprings were studied at 45 and 60 PND. Mean, systolic and diastolic pressures were measured. Na (FeNa) and K (FeK) fractional renal excretions and creatinine clearances (GFR) were estimated.

**Results:** Mean, systolic and diastolic pressures augmented at 45 and 60 PND in Cd-treated rats. Changeswere prevented by borrage oil-treatment. There was no change in GFR, FeNa or FeK at 45 PN in Cd- or Cd+borrage oil-treated rats. At 60 PND GFR was reduced in Cd-treated rats, while FeNa and FeK were increased. These changes were absent in borrage oil+Cd-treated rats, showing protection against HT and renal damage.

**Conclusions:** Cd exposure to pregnant rats induced HT and renal damage in offsprings. HT developed earlier than renal damage. Borrage oil, as source of omega 6, prevented both, HT and renal damage. Supported by CONACYT Mexico grant 51755M

## Abstract# 900

**Evaluation of Glomerular and Tubular Functions in Neonates with Birth Asphyxia** A. Saha,[1] S. Kaur,[2] S. Jain,[2] D. Chawla,[2] V. Parmar.[2] *[1]Pediatrics, PGIMER & Associated Dr Ram Manohar Lohia Hospital, New Delhi, India; [2]Pediatrics, Government Medical College & Hospital, Chandigarh, India.*

**Objectives:** To evaluate glomerular and tubular dysfunction in neonates with moderate to severe birth asphyxia.

**Methods:** Subjects were inborn neonates born at 34 or more completed weeks of gestation and Apgar score <7 at 1 min after birth. Renal function test (RFT) with serum electrolytes were done daily till 96 hrs of life, FeNa (Fractional excretion of sodium), RFI (Renal failure index), urinary myoglobin and CrCl (creatinine clearance) were calculated, using timed urine collection. Renal failure was defined as serum creatinine > 1.5mg/dl.

**Results:** A total of 2196 neonates were born during the study period, of which 44 met the inclusion criteria. 36 babies were available for final analysis. Incidence of ARF was found to be 10/36 (27.8%). Acute renal failure (ARF) was present in 1/11(9%) of moderately asphyxiated (Apgar score 4-6 at 1 min) and 9/25(36%) of severely asphyxiated babies (Apgar score 0-3 at 1 min). Serum creatinine (1.23±0.64 mg%), which is an indicator of glomerular function was found to become normal by 72 h of life in all babies with ARF except 3 (1 had normal by 96 hrs of age, 1 died, 1 unknown). FeNa (6.5±12.2)%, RFI (9.9±15.6) and urinary myoglobin (954±1283) µg/L which are indicator of tubular function, were found to be significantly high at 72-96 h of life. Tubular function were found to be deranged till 72 hrs of life in 70% of ARF cases by which time glomerular function had normalized in these cases.

**Conclusions:** There is differential recovery of glomerulo-tubular dysfunction in neonates with birth asphyxia, with tubular dysfunction persisting beyond 72-96 h after birth.

## Abstract# 901

**Renal Eicosanoid Synthesis in Formula-Fed Rats Exposed to Hyperoxia with Brief Hypoxia** J. Sharma,[1] A. D'Souza,[1] L. Forjour,[1] D. Kumar,[1] G. Valencia,[1] A. Ahmad,[1] C. Cai,[1] M. Schoeneman,[1] J.V. Aranda,[1] K. Beharry.[2] *[1]Department of Pediatrics, Division of Nephrology and Neonatology, SUNY Downstate, Brooklyn, NY, United States; [2]Department of Pediatrics, Division of Neonatology, University of California, Irvine, CA, United States.*

**Objectives:** To exam if formula enhanced with prebiotics (PRE), probiotics (PRO), and /or synbiotics (Syn) preserves renal Prostaglandin (PG) synthesis in a NEC neonatal rat model and hyperoxia with brief hypoxia effects on PG levels.

**Methods:** At birth (P0), neonatal rats pups (n=18pups) were either exposed to normoxia or hyperoxia (50%O2) with brief hypoxia (10%O2) and either maternally-fed (MF); hand-gavaged with Similac, FF; or hand-gavage enhanced with PRO, PRE or Syn from P0 to P3. Kidneys were examined for PGE2, PGF 2α, 6-keto-PGF1α, and TromboxaneB2 (TXB2), 12(S)-HETE, and 15-HETE.

**Results:** On RA, renal PGE2 was elevated in FF (981.2_+81.9, p <0.001), Pro (940.3_+ 102.4, p <0.001), Pre (11009.0-+146.0, p<0.001), and Syn (942.8_+97.6, p<0.001) compared to MF (217.0_+ 25.9) groups. Also noted in PGF2α, 6-keto-PGF1α, 12(S)–HETE, and 15(S)-HETE. TXB2 levels had not change. Hyperoxia elevated PGE2 in the MF group (217.0_+25.9, p<0.05), but reduced it in the FF (179.2_+16.6, p<0.001), Pro (247.4_+72.3, p<0.01), Pre (604_+43.2, p<0.01), and Syn (342.2_+4638, p<0.01) groups. Similarly was for PGF2α, 6-keto-PGF1α, TXB2, and 12(S)–HETE. PGF2α was decreased in all hyperoxic FF groups. 6-keto-PGF1α and TXB2 responses were variable, and 15(S)-HETE was abolished.

**Conclusions:** Renal eicosanoids are altered in FF despite supplementation, thus demonstrating the importance of the maternal milk for normal renal development and function.

## Abstract# 902

**Single Versus Multiple Doses of Early Postnatal Indomethacin with Furosemide Treatment on Renal Eicosanoids and Drug Metabolizing Enzymes in Neonatal Rats** J. Sharma,[1] H.K. Tawadrous,[1] A. Nada,[1] N. Chorny,[1] A. Monia,[1] M. Schoeneman,[1] C. Cai,[2] D. Kumar,[2] J. Aranda,[2] G. Valencia,[2] K. Baherry.[3] *[1]Pediatric Nephrology, SUNY Downstate Medical Center, Brooklyn, NY, United States; [2]Neonatology, SUNY Downstate Medical Center, Brooklyn, NY, United States; [3]Neonatology, University of California, Irvine, CA, United States.*

**Objectives:** Indomethacin (IN), a cyclooxyginase inhibitor, closes patent ductus arteriosus (PDA). IN inhibits renal prostanoids, and decreases GFR Furosemide (FU) with IN prevents indomethacin nephrotoxicity by mutual antagonism. We examined single vs multiple IN dose with FU on renal eicosanoid synthesis in neonatal rat kidneys,determining gene expression of cytochrome P450 enzymes (Cyps).

**Methods:** At birth (P0), rat pups (n=18 ) were randomly assigned to receive IP injections of either IN, 0.2 mg/kg on P0 and FU, 5 mg/kg on P0, P1 and P2; IN and FU on P0, P1, and P2 or equivalent volume saline (Sal) on P0, P1 and P2. At P3, kidneys assessed for $PGE_2$, $PGF_2$, 6-keto-$PGF_1$ ( stable metabolite of prostacyclin), $TxB_2$ ( stable metabolite of $TxA_2$), 12(S)-HETE, regulator of renin release, and gene expression of Cyp enzymes.

**Results:** Multiple-dose IN with FU decreased $PGE_2$ (157.02, p<0.01), $PGF_2$ (137.91, p<0.01), and $TxB_2$ (823.31, p<0.05) compared to Sal (724.31, 1132.21, 1012.52 ) and single-dose IN (733.41, 1506.62, 1863.23). No changes in 12(S)-HETE.Cyps were downregulated with multiple-dose IN. Data expressed as pg/mg protein(meanSEM).

**Conclusions:** FU blunts effects of single dose IN on renal vasoactive eicosanoids, but not multiple doses IN. FU may inhibit Cyp-dependent metabolism and potentiate nephrotoxic effects of IN.

## Abstract# 903

**The Influence of Erythropoietin and Hypothermia on the Damaged Kidneys of Rats with Perinatal Asphyxia** V. Stojanovic,[1] S. Spasojevic,[1] N. Barisic,[1] A. Doronjski,[1] N. Vuckovic.[2] *[1]Institute for Child and Youth Healt Care of Vojvodina, Novi Sad, Serbia; [2]Clinical Centre of Vojvodina, Novi Sad, Serbia.*

**Objectives:** Objectives: The aim of this research was to determine the effects of erythropoietin and moderate hypothermia on the damaged kidneys of rats with perinatal asphyxia.

**Methods:** Methods: Experimental animal population encompassed Wistar rats. Perinatal asphyxia was induced by immersing the uterus containing foetuses into the water (38°C). The uterus was removed from the female rats by caesarean section, on the last day of gestation. The period of asphyxia until delivery of newborn rats has continued for 15 minutes. After birth, the newborn rats were distributed into four groups of 15 animals: G1 - asphyxia; G2 - asphyxia + erythropoietin (2,5µg) + hypothermia (32°C); G3 - asphyxia + hypothermia (32°C); G4 - asphyxia + erythropoietin (2,5µg). The rats were sacrificed on day 7 of life and histo-pathological evaluation of kidney preparations was performed.

**Results:** Results: The results revealed that damage of proximal tubules was significantly higher then that of the distal tubules (p<0.01). Statistically significant differences were established with respect to damage of proximal tubules in group G1, which were significantly higher as compared to the group G2, as well as to groups G3 and group G4 (p<0.01). The zone of immature glomeruli in group G4 was significantly wider then that of the groups G1, G2 and G3 (p<0.01).

**Conclusions:** Conclusions: Erythropoietin and hypothermia, as well as the combination thereof have a protective effect on rat kidney damaged during perinatal asphyxia. Moreover, erythropoietin also enhances maturation of immature kidney glomeruli.

## Abstract# 904

**Perinatal Exposure to Ultrafine Carbon Black Causes Renal Failure to Offspring** M. Sugamata,[1] T. Ihara,[1] M. Sugamata,[1] M. Umezawa,[1,2] K. Takeda.[2] *[1]Tochigi Institute of Clinical Pathology, Tochigi, Japan; [2]Tokyo University of Science, Chiba, Japan.*

**Objectives:** Various nanoparticles are produced with development of nanotechnology. The safety is not confirmed precisely at present. We have already reported that some nanoparticles exposure to pregnancy animal exert severe damages on nervous and respiratory systems of offspring. In this study, the influence of exposure to ultrafine carbon black (UfCB) in fetal period on renal system of offspring was examined morphologically.

**Methods:** Pregnant ICR mice were exposed to a total of 100 μg of UfCB (particle size; 14nm) by intranasal instillation on gestational days 5 and 9 for exposure group (CB group); comparative control animals were treated with vehicle. After delivered their pups, kidney tissues were collected from 12-week-old male pups, and examined with light and electron microscope. All experimental animals were handled in accordance with institutional and national guidelines for the care and use of laboratory animals.

**Results:** In renal cortex of CB group, collagen fibers were diffusely observed surroundings of renal corpuscle and tubule, and significantly larger than in control group. In addition, specific and interesting renal failure, that is similar to characteristic findings in a part of genetics diseases, were found in tubular epithelial cell of renal cortex of CB group.

**Conclusions:** These observations indicate that perinatally-exposed ultrafine carbon black caused renal failure in offspring. Our findings indicate existence of a severe health hazard to infants borne from mothers exposed nanomaterials during pregnancy. It is possible that these inborn damages carry a severe disorder, which is recognized as a genetic disorder.

## Abstract# 905

**Is There Any Relationship between Maternal BMI and Neonatal Kidney Size?** S. Sultana,[1] S. Rahman,[1] B.K. Basak,[1] S. Ferdaus,[1] M. Hanif,[2] N. Hossain.[1] *[1]Paediatrics, Uttara Adhunik Medical College Hospital, Dhaka, Bangladesh; [2]Paediatric Nephrology, Dhaka Shishu Hospital, Dhaka, Bangladesh.*

**Objectives:** The study was done to find out any relationship between maternal BMI with kidney length and volume of term neonate.

**Methods:** Fifty eight (58) healthy, term, appropriate for gestational age (37 to 41 wks) newborn were examined prospectively by sonography within 72 hours of birth by a single sonologist. In 30 boys and 28 girls, body weight(BW), supine length(SL) and occipitofrontal circumference(OFC) were collected from delivery room records. Maternal BMI was calculated by the formula BMI=wt in kg.ht$^2$ in meter.Scanning was performed with 6.5 MHZ transducer. Maximum length of each kidney was determined. Volume of the kidneys were determined by in built formula of software. Kidney length and kidney volume were then correlated with maternal BMI. This two were also correlated with different parameters of newborn.

**Results:** There were no significant differences in mean kidney length and volume between right (38.41mm, 9.67cc) and left (38.43mm, 9.79cc) kidney. The length of kidneys between boys and girls showed no difference, but volume of right kidney between boys and girls showed significant difference (<0.01). Kidney length (KL) was correlated better with infant BMI (<0.01), infant body weight (<0.01) than infant body surface area (BSA) (<0.05).Kidney volume was also correlated with BMI (<0.05).body weight (<0.05) and BSA (<0.05).
But kidney length and volume showed no significant correlation with maternal BMI.

**Conclusions:** Despite expectation that maternal BMI might have some correlation with neonatal kidney size but our study showed no significant correlation with maternal BMI.

## Abstract# 906

**Ultrasound Determination of Kidney Length and Volume in Premature Newborn** S. Sultana, S. Rahman, B.K. Basak, S. Ferdaus, N. Hossain. *Paediatrics, Uttara Adhunik Medical College Hospital, Dhaka, Bangladesh.*

**Objectives:** To determine the normal range of kidney length and volume in premature Bangladeshi newborn.

**Methods:** Forty one inborn, appropriate for gestational ages, preterm infant (31 to 36 weeks) were prospectively examined by sonography within 72 hours of birth by a single sonologist, who were unaware about the gestational age of the baby. In 23 boys and 18 girls, gestational age was estimated by Ballard score. Body weight (BW), supine length (SL),occipito frontal circumference (OFC) were collected from delivery room records. Body surface area (BSA) and BMI were calculated using the formula. Scanning was performed with 6.5 MHZ transducer. Maximum length of each kidney was determined. Kidney length and volume were then correlated with different parameter of infant.

**Results:** There were no significant differences in mean kidney length and volume between right (35.45 mm, 7.55cc) and left (34.30mm, 7.23cc) and kidneys in boys and girls. Kidney length correlated more with body wt (<0.001), BMI (<0.001) and BSA (<0.01) than gestational age (<0.05) and OFC (<0.05). Here no correlation found with length.
Kidney volume also correlated better with body wt (<0.001), BSA (<0.001) and BMI (<0.01) than height (<0.05) and OFC (<0.05). Here no correlation found with gestational age.

**Conclusions:** The present ongoing study provides an important baseline data in preterm babies for kidney dimension in Bangladeshi neonate as well as neonate of Indian subcontinent. BW, BMI, BSA all are correlated with kidney length and volume. So other two along with BW can also be selected as the independent variable for preparation of normogram for kidney length and Volume in premature infant.

## Abstract# 907

**Effects of Total Parenteral Nutrition on Renal Function in Preterm Neonate** Y. Tabel,[1] M. Oncul,[2] I.M. Akin,[2] A.B. Karabulut,[3] S. Gungor.[2] *[1]Pediatric Nephrology, Inonu University, Malatya, Turkey; [2]Pediatrics, Inonu University, Malatya, Turkey; [3]Biochemistry, Inonu University, Malatya, Turkey.*

**Objectives:** The aim of the study was to evaluate the effect of total parenteral nutrition on renal functions of prematrue infants by comparing the serum cystatin C (cysC) levels and urinary N-acetyl- β-D glucosaminidase (NAG), β 2 microglobulin (β2M), gluthatione S transferase (GST) π levels in premature infants receiving total parenteral nutrition or enteral feding.

**Methods:** Premature infants hospitalized in neonatal intensive care unit of Inonu University between January 2007 and July 2008, with a gestational age of 28 and 34 weeks were included in the study. Exclusion criteria consisted of presence of congenital malformations, parental refusal, onset of TPN or death prior to 72 hours. Prenatal, natal and postnatal characteristics of infants including the presence of respiratory distress syndrome, early neonatal sepsis, late-onset neonatal sepsis and the use of aminoglycosides were recorded. on the 3$^{rd}$ and 30$^{th}$ day of life, blood samples of all patients were obtained for evaluating biochemical parameters and cysC, and urine samples for the evaluation of NAG, GST π, β2M, sodium, creatinin levels, density and pH of the urine.

**Results:** Serum cysS, urinary β2M, NAG and GST π excretion were significantly higher in samples of patients receiving TPN both on 3$^{rd}$ and 30$^{th}$ days (p<0.05 for each parameter on each day).

**Conclusions:** This study have shown for the 1$^{st}$ time in premature infants that, TPN together with the presence of sepsis and use of aminoglycosides can have adverse effects on glomerular and tubular functions of the kidney which can be manifested with cysC, β2M, NAG and GST π.

## Abstract# 908

**Early Appearance of Hypokalemia in Gitelman Syndrome** F. Tammaro,[1] A. Bettinelli,[1] D. Cattarelli,[2] C. Colombo,[3] S. Tedeschi,[4] M.L. Syrén,[5] M.G. Bianchetti.[6] *[1]Pediatrics, Mandic Hospital, Merate, Italy; [2]Pediatrics, Gavardo Hospital, Brescia, Italy; [3]Intensive Care, S.Gerardo Hospital, Monza, Italy; [4]Laboratory of Medical Genetics, Fondazione IRCCS Ca'Granda Policlinico, Milano, Italy; [5]Scienze Materno-Infantili, Università degli Studi, Milano, Italy; [6]Pediatrics, S.Giovanni Hospital, Bellinzona, Switzerland.*

**Objectives:** Inactivating mutations in the *SLC12A3* gene that encodes the thiazide-sensitive co-transporter cause Gitelman syndrome whose features include normal-low blood pressure, hypokalemia, metabolic alkalosis, hypomagnesemia, hypocalciuria, and hyperreninemia. These patients are at low risk for preterm birth and the condition is usually diagnosed in childhood or in adult life. We report on 4 patients, 2 pairs of prematurely born twins, in whom hypokalemia was demonstrated early in life.

**Methods:** We evaluated 2 pairs of twins born after 28 and 34 weeks of gestation. In the 4 children, hypokalemia was noted during the third week of life, associated with normal blood pressure, normal total plasma magnesium level, hypochloremia and hyperreninemia. A marginal tendency towards hypomagnesemia (0.70 mmol/L) was observed after 2 years of life.

**Results:** In both pairs of twins we demonstrated compound heterozygous mutations in the *SLC12A3* gene: a frame shift mutation in exon 10 (c.1196_1202dup7bp), leading to the truncated protein p.Ser402X, and a missense mutation in exon 11, p.Ser475Cys (c.1424C>G) were disclosed in the first pair and two missense mutations, p.Thr392Ile (c.1175C>T) in exon 9 and p.Ser615Leu in exon 15 (c.1844C>T), in the second pair.

**Conclusions:** Gitelman syndrome may be considered as an additional cause of hypokalemia in the neonatal period.

## Abstract# 909

**Effects of Co-Administraton of Indomethacin, Ibuprofen and Furosemide on Renal Prostanoid Synthesis in Neonatal Rats** H.K. Tawadrous,[1] J. Sharma,[1] N. Chorny,[1] A. Nada,[3] M. Schoeneman,[1] C. Cai,[2] D. Kumar,[2] J. Aranda,[2] G. Valencia,[2] K. Baherry.[4] *[1]Pediatric Nephrology, SUNY Downstate Medical Center, Brooklyn, NY, United States; [2]Neonatology, SUNY Downstate Medical Center, Brooklyn, NY, United States; [3]Pediatric Nephrology, University of Alexandria, Alexandria, Alexandria, Egypt; [4]Neonatology, University of California, Irvine, CA, United States.*

**Objectives:** Indomethacin(IN),for closure of PDA,has renal side effects. Furosemide(FU) is used with IN to lessen them. Ibuprofen(IB)is as effective as IN for closure of PDA with fewer side effects.We examined IN and IB administed with FUon prostanoid synthesis in neonatal rat kidneys.

**Methods:** Rat pups P0(n=12)randomly injected IP1)0.2mg/kg INonP0and0.1 mg/kg on P1&P2;2)10mg/kg IB P0 and 5mg/kg on P1&P2;3) 5mg/kg FU on P0, P1&P2;4) IN+FUonP0,P1,P2;5)IB+FUonP0,P1,P2;or6)equal volume saline(S)on P0,P1,P2. P3, urine and kidneys assessed for $PGE_2$,$PGF_2$,6-keto-$PGF_1$,$TxB_2$.

**Results:** IN+FU decreased $PGE_2$(157,p<0.01) vs.S(862). FU increased urinary $PGE_2$ (2478,p<0.001) vs.IN(1603), IB(1781), IN+FU(2002) and IB+FU(1781). IB+FU increased $PGF_2$(3443p<0.01) vs.IN(308), IB(947), and IN+FU(39). IB and IB+FU increased renal 6-ketoPGF$_1$(10577and10313,p<0.01) vs IN(1141), IN+FU(2344),and FU(687). Urinary 6-ketoPGF$_1$ increased with IB(7411, p<0.01)vs.all groups. FU increased $TxB_2$(2195,p<0.05)and IB increased urinary $TxB_2$(8934,p<0.01)vs.S(3353),IN(2502),FU(3018),IN+FU(1897) and IB+FU(2959).

**Conclusions:** FU with IB increase prostanoid excretion suggesting predominance of FU over IB on renal prostanoids. IN and FU decreased $PGE_2$ most important renal prostanoid. FU does not reverse the renal effects of multiple doses of IN.

## Abstract# 910

**Ontogeny of Arachidonic Acid Metabolites and Gene Expression of Cytochrome P450 Enzymes in Kidneys of Suckling and Weanling Neonatal Rats** H. Tawadrous,[1] N. Chorny,[1] J. Sharma,[1] M. Schoeneman,[1] A. Mongia,[1] G. Valencia,[2] J.V. Aranda,[2] C. Cai,[2] D. Kumar,[2] K.D. Beharry.[2,3] *[1]Pediatric Nephrology, SUNY Downstate Medical Center, Brooklyn, NY, United States; [2]Neonatology, SUNY Downstate Medical Center, Brooklyn, NY, United States; [3]Neonatology, University of California, Irvine, CA, United States.*

**Objectives:** We examined ontogeny of renal arachidonic acid(AA) metabolites and gene expression of CYPs during renal maturation in newborn,suckling and weanling periods in neonatal rats. We determined gender differences in CYP enzymes.

**Methods:** Kidneys from rat pups were examined at birth P0,P4,P7,P14,P21 for levels of PGE2,PGF2,6-ketoPGF1,thromboxane(Tx) B2,8-isoPGF2,leukotriene (LKT) B4, and (15-HETE) using ELISA, and the gene expression of CYPs using PCR.

**Results:** All AA metabolites were high at birth(PGE2:412611;PGF2:44298;6-ketoPGF1: 47196;TxB2:350548;8-isoPGF2:1181725;LKTB4:59983114;and 15-HETE: 41026109. During low Na suckling, metabolites exhibited different maturation. All decreased by(%)90,40,95,100,98,100; p<0.01,respectively,except TxB2 3390609, which declined at P7(46145) and P14(12248) compared to P0. At high Na weaning, metabolites increased with vasoconstrictors,TxB2(1364.09),8-isoPGF2(126545859), and 15-HETE(3063165) being highest. Ontogenic patterns of CYP enzymes paralleled metabolites. The main renal isoform of CYP3A regulates BP and R-A system, downregulated during suckling, and upregulated during weaning, 5-fold higher in females than males.

**Conclusions:** Renal AA metabolites favor vasoconstrictors at weaning and may be important for drug disposition/Na excretion. Higher expression of CYP3A5 in females has early gender-specific difference.

## Abstract# 911

**Angiotensin AT1 Receptor Inhibition-Induced Apoptosis by Rho A Activation Associated with Na+/H+ Exchanger 1 NHE1 Downregulation in Neonatal Unilateral Ureteral Obstruction (UUO)** P.G. Vallés, V. Bocanegra, W. Manucha, A. Gil Lorenzo, M. Rinaldi. *Facultad de Medicina.Universidad Nacional de Cuyo, Mendoza, Argentina; CONICET, Mendoza, Argentina.*

**Objectives:** We examined the involvement of NHE1 associated with RhoA in the regulation of epithelial cell apoptotic response after AT1 receptor inhibition in obstruction. NHE1 modulation of the ERK1/2 kinase signal pathway was also evaluated.

**Methods:** Neonatal rats subjected to complete UUO within the first 48 hours of life and sham received saline, Losartan, or PD-123319 AT2 inhibitor for 14 days.For apoptosis study TUNEL assay confirmed by electron microscopy, Bax/BcL2 expression and caspase 3 expression and activity, were performed. Western blotting and immunofluorecence were conducted for proteins expression.

**Results:** Tubular cell apoptotic response associated with mitochondrial signaling pathway through the increased proapoptotic ratio Bax/BcL-2 and consequently increased caspase3 expression and activity was demonstrated in Losartan obstructed kidney.

Stimulation of RhoA activity that in turn reduced NHE1 expression was demonstrated in 14 day Losartan UUO kidney.Interaction between NHE1 and pERK was determined by coimmunoprecipitation showing that membrane downregulation of NHE1 was associated with pERK overexpression. Absence of increased AT2 protein expression after Losartan on day 14 of obstruction was shown. PD-123319 had no protective effect on the renal response to complete 14 day UUO.

**Conclusions:** Our findings suggest a role of RhoA on the negative regulation of NHE1 inducing phosphorylation of ERK 1/2 as events involved in tubular cell apoptosis regulation after AT1 receptor inhibition in neonatal UUO.

## Developmental Nephrology

## Abstract# 912

**(O-113)**

**Inhibition of Branching Morphogenesis and Developmentally Regulated Signaling Pathways by Maternal Nutrient Restriction** M. Awazu, M. Hida. *Department of Pediatrics, Keio University School of Medicine, Tokyo, Japan.*

**Objectives:** Hypertension and chronic kidney disease in the adulthood are related to prenatal insults that usually result in low birth weight and reduced nephron number. In animals, maternal nutrient restriction produces offspring with fewer nephrons. We studied whether the reduced nephron number is due to the inhibition of ureteric branching and developmentally regulated signaling pathways.

**Methods:** The offspring of dams given food ad libitum (CON) and those subjected to 50% food restriction throughout pregnancy (NR) were examined. Embryonic day 13 (E13) to day 15 (E15) metanephroi were stained with Dolichos biflous lectin. Lysates of E15 metanephroi were analyzed by immunoblot.

**Results:** At E13, there was no difference between NR and CON in either body weight or kidney surface area. Ureteric buds branched once in both NR and CON. At E14 and E15, body weight and kidney surface area were significantly reduced in NR compared with CON. Ureteric bud tip numbers per kidney were significantly reduced in NR (11.2±1.3 vs 21.3±1.0 at E14 and 31.0±1.7 vs 59.6±3.5 at E15). Activated forms of ERK, p38, PI3-kinase, and Akt and protein expression of β catenin were decreased 0.6-, 0.8-, 0.7-, 0.5-, and 0.7-fold, respectively, in NR. The expression of PCNA, a marker of proliferation, was not different between NR and CON, whereas that of cleaved caspase 3, a marker of apoptosis, was markedly decreased (0.4-fold of CON).

**Conclusions:** Ureteric branching and metanephros growth were inhibited by maternal nutrient restriction after E13. Downregulation of developmentally regulated signaling pathways may play a role in abnormal kidney development in this pathophysiological setting.

## Abstract# 913

**HNF1B and FXYD2 Co-Expression Helps Explain Renal Magnesium Wasting in the Renal Cysts and Diabetes Syndrome** S. Adalat,[1] J. Papakrivopoulou,[1] A.S. Woolf,[2] D. Bockenhauer.[1] *[1]Institute of Child Health, London, United Kingdom; [2]University of Manchester, Manchester, United Kingdom.*

**Objectives:** *Hepatocyte nuclear factor 1B* (*HNF1B*) mutations cause kidney malformations in the renal cysts and diabetes (RCAD) syndrome. We reported hypomagnesaemia in children with *HNF1B* mutations and found that, *in vitro*, HNF1B transactivated *FXYD2* encoding a distal convoluted tubule (DCT) $Na^+K^+$ATPase subunit itself mutated in another inherited hypomagnesaemia. However, whether HNF1B protein exists in DCT is unknown and we accordingly investigated expression of this and fxyd2 in mouse kidneys.

**Methods:** Immunohistochemistry and western blotting in adult mouse kidneys and an established DCT cell line.

**Results:** In tissue sections, hnf1b transcription factor was immunodetected in nuclei of kidney epithelia, being most prominent in collecting ducts and distal tubules, the latter characterised by expression of the transporter slc12a3. Hnf1b was weakly expressed in proximal tubules but glomerular epithelia, interstitial and vascular cells were negative. Fxyd2 itself had an overlapping distribution with hnf1b, being expressed in the distal tubule and collecting ducts. The DCT cell line expressed hnf1b as assessed by western blotting.

**Conclusions:** Hnf1b and fxyd2 co-expression in distal tubules support the hypothesis that HNF1B mutations cause magnesium wasting by altering DCT physiology. Interestingly, expression of hnf1b in proximal tubules is consistent with a role in uric acid handling, as hyperuricaemia can be part of the syndrome.

## Abstract# 914

**Renal Prognosis of Children Born from Hemodialysis Mothers** P. Abou-Jaoude, L. Bessenay, L. Dubourg, A. Jolivot, P. Cochat, J. Bacchetta. *Hospices Civils de Lyon et Universite de Lyon, Lyon, France.*

**Objectives:** Pregnancy during dialysis is a high risk condition that is more and more frequent. The renal outcome of children issued from such pregnancies needs to be investigated since renal development may be influenced (i.e., exposition to uremic toxins and therapies, hemodynamic changes during sessions, prematurity, growth retardation).

**Methods:** We performed prospective global and renal evaluation (inulin clearance or locally-adapted Schwartz formula in younger children) in a single-centre cohort of 10 children from 7 hemodialysis mothers.

**Results:** The mean age of mothers at the beginning of pregnancy was 28 years, with average maximal weekly hemodialysis duration of 18.9 hours (12-30). Four mothers had hypertension; two polyhydramnios and one oligohydramnios were observed. Seven children were born before 36 (26-39) weeks of gestation. The mean birth weight was 1820 g (930-3430), and eight children had a birth weight below 2500 g.

One child had a PAX2 mutation requiring early renal transplantation during childhood, and was thus excluded for further analysis.

Even if GFR and blood pressure were normal in all other children, an increased urine albumine/creatinine ratio was found in five children, questioning the presence of an underlying silent reduction in nephron number. Moreover, one premature baby with severe necrotizing enterocolitis showed proximal tubular dysfunction and nephrocalcinosis.

**Conclusions:** Despite a small number of children, these preliminary results underline the interest of performing a regular follow-up of children born from mothers undergoing hemodialysis during pregnancy, so as to early refer children when minor abnormalities are highlighted.

## Abstract# 915

**Prevalence of Renal Disease in HIV Infected Children at Kenyatta National Hospital** D.D. Galgallo, R. Nduati, E. Obimbo, G. Irimu, D. Wamalwa. *Paediatrics, University of Nairobi, Nairobi, Nairobi, Kenya; Paediatrics, Universtiy of Nairobi, Nairobi, Nairobi, Kenya; Paediatrics, University of Nairobi, Nairobi, Nairobi, Kenya.*

**Objectives:** 1. To determine prevalence of Renal disease in Hiv infected children age between 18 months and 12 years as detemined by persistent proteinuria and or decreased glomerular filtration rate.

**Methods:** Children between ages 18 months to 12 years were enrolled.December 2005 and April 2006. At baseline urine protein, serum creatinine, bicarbonate, albumin and CD4% were determined. Among children with proteinuria,a repeat urine assay for protein was performed 2 weeks later.Renal disease was defined as persistent proteinuria and/or decreased GFR.

**Results:** A total of 87 subjects were recruited. Forty six (52.8%) were females and 41 (47.1%) males.Overall, 35.6% of the patients had evidence of nephropathy based on presence of persistent proteinuria and/or abnormal glomerular filtration rate.Proteinuria was found in 28(32.2%) of the subjects, persistent proteinuria in 14 (16.1% )subjects,deranged glomerular filtration rate in 23 (26.4%).Forty four (50.6%) of the subjects were in WHO clinical stage 3 of the disease,17(19.5%) in stage 2,14 (16.1%) in stage 4 and 12 (13.8%) were in stage 1.

**Conclusions:** Renal disease is common among Kenyan HIV infected children and may present as persistent proteinuria and/or abnormal GFR. A third of HIV infected children were found to have renal disease. Persistent proteinuria was more frequent among children with early clinical stage of HIV/AIDS disease than those with later stages in this study population.

## Abstract# 916

**Nephrocystin 1 Interacts with the Centrosomal Protein CEP170** S. Habbig,[1,2] M.C. Liebau,[2] F. Fabretti,[2] S. Zank,[2] H. Zentgraf,[4] I. Schmedding,[2] T. Benzing,[2] B. Schermer.[2] *[1]Pediatric Nephrology, University Childrens Hospital, Cologne, Germany; [2]Renal Division, University Hospital, Cologne, Germany; [3]Institute of Developmental Biology, Cologne, Germany; [4]DKFZ, Heidelberg, Germany.*

**Objectives:** After molecular genetics identified many crucial genes in nephronophthisis, the characterization of the underlying proteins, called nephrocystins, and especially their localisation at the primary cilium were important steps in understanding this most common genetic dialysis-requiring renal disease in childhood. However, the physiological cellular role of the nephrocystins is poorly understood. In a proteomics screening approach, we identified CEP170 as an interacting protein of nephrocystin-1 (NPHP1). Interestingly, another protein of the same famlily, CEP290, has already been identified as nephrocystin-6.

**Methods:** Interaction was confirmed by independent immunoprecipitation experiments and mapped using several truncations and mutants of NPHP1 and CEP170. Novel monoclonal antibodies against CEP170 were generated and optimized for immunoblot, immunofluorescence and immunoprecipitation.

**Results:** The interaction was confirmed and mapped to the SH3-domain of NPHP1 and to the N-terminus of CEP170. Immunofluoresence studies with the generated antibodies in ciliated cells revealed colocalisation of endogenous CEP170 with NPHP1 at the base of primary cilia.

**Conclusions:** We identified and characterised the novel interaction between CEP170 and NPHP1. Interaction as well as colocalisation of CEP170 and NPHP1 suggest a common functional role of both proteins at the ciliary base and at the centrosome.

## Abstract# 917

**Toilet Training in Iranian Children – Multicenter Study** N. Hooman,[1] A. Safaii,[2] E. Valavi, Z. Amini,[1] E. Gerami,[1] M. Shams.[1] *[1]Pediatric Nephrology, Iran University of Medical Sciences, Tehran, Islamic Republic of Iran; [2]Pediatric Nephrology, Gylan University of Medical Sciences, Tehran, Islamic Republic of Iran; [3]Pediatric Nephrology, Ahwaz University of Medical Sciences, Tehran, Islamic Republic of Iran.*

**Objectives:** To determine the beliefs of parents about the appropriate age, the true age, and the methods used of toilet training.

**Methods:** 209 questioners contained fourthly five items were distributed among parents of healthy children aged 1-5 years who brought to the clinic for routine checkup between August 2008 and December 2009 in Tehran(capital), Ahwaz (south), and Rasht(north). The data are presented as mean, frequency and correlation. $P < 0.05$ was considered significant.

**Results:** 113 girls and 96 boys with mean age of 4.06 years were entered to study. The parents believed that the appropriate age of staring toilet training is less than 12 month in 18.2%, 1-2 years in 64.1%, age > 2 years in 9%, and 8.6% had no idea. 19% of parents punished and 6.7% teased their children if they could not keep dry. There was strong reverse correlation between the level of education of father with applying punishment for training (P=0.005). The reactions of the children during training were fear of washing room 9.6%, hate going to toilet 12.4%, prefer to continue the game 55%, constipation 12.4%, and temper tantrum 20.8%. There was reverse significant correlation between temper tantrum and the age of starting training. There was correlation between punishment with showing temper tantrum and hate of going to toilet (P=0.0001).

**Conclusions:** The age and the method of toilet training have strong influence on the success of this process.

## Abstract# 918

**Role of Focal Adhesions (FAs) in Rat Developing Kidneys** S. Matsuura, S. Kondo, K. Suga, Y. Kinoshita, M. Urushihara, S. Kagami. *Dept. of Pediatrics, Tokushima Univ., Tokushima, Japan.*

**Objectives:** FAs play a critical role as the central to transduce signals by cell-matrix interactions and regulate cell bahaviors. FAK, ILK, paxillin and Hic-5 are major proteins to contribute these signaling events. We investigated the expression of these proteins in developing kidneys.

**Methods:** We performed western blotting and immunohistochemistry in rat developing kidneys which removed from embryos on day 14 to 18 of gestation, and on 1 to 42 days after birth.

**Results:** Western blotting revealed that the expression of FAK (p-FAK$^{397}$), paxillin (p-paxillin$^{118}$ ) and Hic-5 were high in embryonic kidneys. Meanwhile, ILK expression retain from embryonic to matured kidney. Immunohistochemistry revealed that FAK and paxillin were strongly expressed in mesenchymal cells and ureteric bud, following detected in elongating tubular epithelial cells, collecting duct, immature glomerular endothelial and mesangial cells. Compared with distribution of PCNA positive cells, FAK and paxillin might be involved in renal growth and morphogenesis. Hic-5 was dominantly expressed in mesenchymal and immature glomerular endothelial and mesangial cells, similar to α-SMA positive cells, suggesting that Hic-5 might be related to mesenchymal cell behavior. In mature kidneys, FAK and paxillin were limited in tubules and Hic-5 was limited in vascular smooth muscle cells. Although ILK expression similar to FAK in developing stages, it strongly expressed at matured podocytes and tubules. It suggests that ILK plays a role in epithelial cell differentiation as well as kidney growth and morphogenesis.

**Conclusions:** Temporo-spatially regulated expression of FAs might play a role in kidney morphogenesis and differentiation.

## Abstract# 919

**Morphologic and Functional Analyses of Two Infants with Obstructive Renal Dysplasia** K. Miura,[1] T. Sekine,[1,2] R. Nishimura,[1] Y. Kanamori,[3] A. Yanagisawa,[4] K. Sakai,[5] M. Nagata,[6] T. Igarashi.[1] *[1]Department of Pediatrics, Tokyo University Graduate School of Medicine, Tokyo, Japan; [2]Department of Pediartics, Toho University School of Medicine, Ohashi, Tokyo, Japan; [3]Department of Pediatric Surgery, Tokyo University Graduate School of Medicine, Tokyo, Japan; [4]Department of Pediatrics, Ohta Nishinouchi General Hospital, Kohriyama, Fukushima, Japan; [5]Department of Urology, Miyagi Children's Hospital, Sendai, Miyagi, Japan; [6]Department of Pathology, Institute of Basic Sciences, University of Tsukuba, Tsukuba, Ibaraki, Japan.*

**Objectives:** To discuss the pathogenesis of obstructive renal dysplasia (ORD), a rare condition associated with urinary tract obstruction.

**Methods:** Serial morphologic and functional analyses were performed in the two patients with ORD from early gestational weeks to infantile periods.

**Results:** Fetal ultrasonography revealed marked dilatation of unilateral ureter at early gestational weeks in both patients. They manifested unilateral kidney enlargement with multiple cortical cysts, mild hydronephrosis, and marked dilatation of the ipsilateral ureter at birth. These manifestations were consistent with a diagnosis of ORD. Renal scintigraphy revealed that the functions of the affected kidneys were severely impaired but still remained detectable. Most of the cortical cysts disappeared within a few months, while the functions of the affected kidneys were further deteriorated.

**Conclusions:** The present study illustrates the pathophysiology underlying in the development of ORD. It also indicates that this phenotype is distinct from multicystic dysplastic kidney, another form of renal dysplasia associated with urinary tract obstruction.

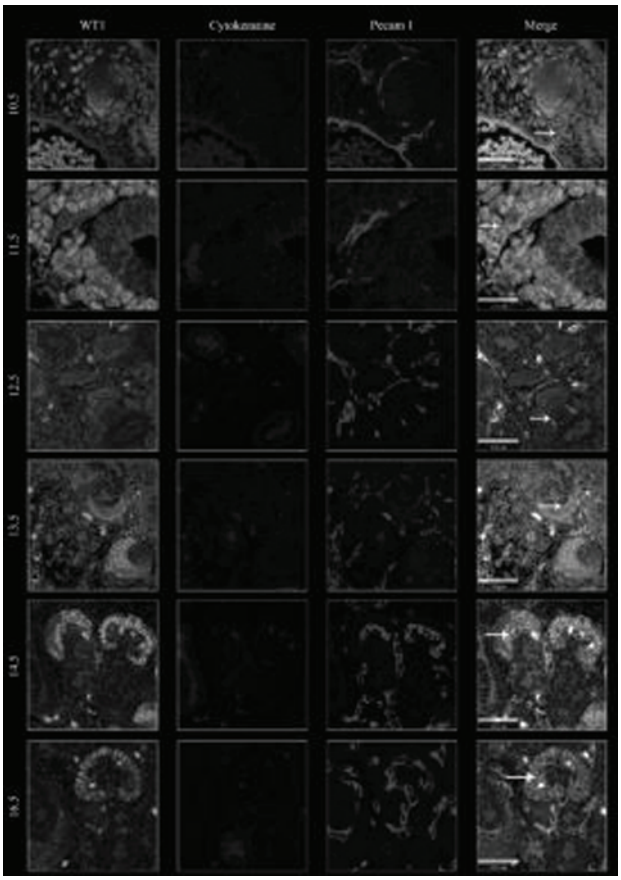
## Abstract# 920

***In Vivo* and *In Vitro* Analysis of Vascular Development in Kidneys: Towards Renal Engineering** O.R.P. NIEL,[1,2] S. Bradford,[2] E. Cassuto,[1] E. Berard,[1,2] A. Schedl.[2] *[1]Néphro-Pédiatrie, Centre Hospitalier Universitaire Archet II, Nice, France; [2]Génétique et développement Rénal, Inserm U636, Nice, France.*

**Objectives:** We'll characterize vasculogenesis and angiogenesis *in vivo* in kidney development. We'll analyse *in vitro* vascular development limitations. We'll propose a novel kidney culture method, to improve vascular development.

**Methods:** We used B6 mice. IF was done on Zeiss LSM 510 Meta. qRT-PCR conducted on LightCycler 1.5 (Roche). P values of less than 0,05 considered significant.

**Results:** *In vivo*, endothelial cells (PECAM1+) bud from the aorta at 10.5 dpc and migrate towards the metanephric mesenchyme (WT1+) to form the primitive renal artery at 11.5 dpc. Angioblasts (FLT1+ and KDR+) gather among the metanephric mesenchyme at 10.5 dpc and participate in renal vascular network development. Vascular tuft formation occurs between 12.5 and 14.5 dpc.




*In vitro* vascular development is impaired. It requires the PGC-1α pathway, HIF-independent, activated by hypoxia. *In vitro* kidney cultures with vascular microperfusion in 18.5 dpc embryos show significant renal vasculature, and fluid excretion.

**Conclusions:** We showed that angiogenesis and vasculogenesis occur early *in vivo* in kidney development. *In vitro* vascular development requires PGC-1α pathway in addition to HIF. We proposed a novel kidney culture method, to optimize vasculature, with encouraging preliminary results.

## Abstract# 921

**Early vs Late Antenatal Betamethasone Effects on Renal Eicosanoids in the Newborn and Suckling Neonatal Rats** J. Sharma,[1] A. Ahmad,[1] A. D'Souza,[1] C. Charles,[1] D. Kumar,[1] L. Forjour,[1] G. Valencia,[1] N. Anwar,[1] J.V. Aranda,[1] M. Schoeneman,[1] K. Beharry.[2] *[1]Departement of Pediatrics, Division of Pediatric Nephrology and Neonatology, SUNY Downstate Medical Center, Brooklyn, NY, United States; [2]Departement of Pediatrics, Division of Neonatology, University of California, Irvine Medical Center, Irvine, CA, United States.*

**Objectives:** Exposure to AB (170µg/kg IM) influences renal eicosanoids in the newborn and suckling neonatal rat and determine the effects of hyperoxia with brief hypoxia on renal prostanoids.

**Methods:** Pregnant rats received a single 2-dose course of: 1) AB on E17 and E18 (early); 2) AB on E19 and E20 (late); 3) equivalent volumes of early and late saline (Sal). Pups were randomly kept on room air or placed in hyperoxia (50% O2) with random, brief hypoxia (O210%) until P4. Kidneys were analyzed for PGE2, PGF2α, 6-keto-PGF1α, TXB2, and 12(S)-HETE.

**Results:** Early AB increased PGF2α (327.7±48.7 v. 96.6±25.2, p<0.01). At P4, it increased PGE2 (1039.3±66.1 v. 719.8±74, p<0.05) and 12 (S)-HETE (1426.6±92.2 v. 508.5±94.8, p<0.001) and decreased TxB2 (585.5±70.9 v. 1247±126.2, p<0.01), and suppressed PGE2, PGF2α, 6-keto PGF1α, and TxB2 in hyperoxia. Late AB increased PGE2 (412.9_+93.0 v. 176.3_+14.0, p<0.05), and PGF2α (157.7±21.2 v. 96.1±16.0, p<0.05). But suppressed 6-ketoPGF1α (831.0±128.7 v. 1972.2±213.6, p<0.01), TxB2 (248.3±57.7 v. 785.2±117, p<0.01), and elevated 12(S)-HETE (1108.5±88.6 v. 602.4±30.8, p<0.001).

**Conclusions:** Antenatal steroid tx results in alterations in renal vasoactive eicosanoids which may potentially contribute to high BP in theoffspring. Hypoxia-hyperoxia appears to have a more deleterious effect on renal eicosanoids.

Renal Physiology

## Abstract# 922
**(O-114)**

**Functional Characterization of Six Alternatively Spliced Claudin-10 Isoforms Found in Renal Epithelia** L. Haisch,[1] M. Stuiver,[1] P.J. Kausalya,[3] S.M. Krug,[2] R. Rosenthal,[2] I. Meij,[4] W. Hunziker,[3] M. Fromm,[2] D. Günzel,[2] D. Müller.[1] *[1]Pediatric Nephrology, Charité, Berlin, Germany; [2]Institut of Clinical Physiology, Charité, Berlin, Germany; [3]Institut of Molecular Biology, Singapore, Singapore; [4]Max-Delbrück-Center, Berlin, Germany.*

**Objectives:** Tight Junctions regulate paracellular permeability in epithelia. Claudins, a family of TJ proteins, provide an intercellular seal but may also convey specific ion permeability. Claudin-10 has been described as existing in 2 isoforms. In the present study, we have identified another four splice variants: one lacking 57 nucleotides in exon1 and for each of these three splice variants a further version without exon4.

**Methods:** Mouse and human cldn-10 isoforms were transfected into high-resistance MDCK-C7 cells. Immunostainings were carried out to investigate cldn-10 localization. Ion selectivity was determined by measuring dilution and biionic potentials in Ussing-type chambers.

**Results:** Confocal laser scanning microscopy revealed a junctional distribution of three isoforms, while the isoforms lacking exon4 were retained in the endoplasmatic reticulum. Electrophysiological investigations showed that Cldn10a changed paracellular anion selectivity. In contrast, Cldn10b greatly increased cation selectivity changing Eisenman sequence from IV to X. This indicates the presence of a high field-strength binding site within the paracellular pathway.

**Conclusions:** According to the immunostainings exon4 determines subcellular claudin localization. Overexpression of both Cldn10a and b changed the ion selectivity of the paracellular pathway by increasing the interactions between the permeating ions and the paracellular pore.

## Abstract# 923
**(O-115)**

**Role of a Novel Slit Diaphragm Component, SIRPα, as a Nephrin-Interacting Protein** Y. Harita,[1,2,5] A. Matsunaga,[3,5] H. Tsurumi,[2,5] S. Kanda,[2,5] T. Sekine,[2] T. Igarashi,[2] S. Hattori,[3,5] H. Kurihara.[4] *[1]Molecular Biology, Yokohama City Univ. Grad. Sch. of Med., Yokohama, Kanagawa, Japan; [2]Pediatrics, Univ. of Tokyo, Bunkyo-ku, Tokyo, Japan; [3]Biochemistry, Kitasato Univ., Minato-ku, Tokyo, Japan; [4]Anatomy, Juntendo School of Medicine, Bunkyo-ku, Tokyo, Japan; [5]Cellular Proteomics (BML), IMSUT, Minato-ku, Tokyo, Japan.*

**Objectives:** Podocyte Slit diaphragm (SD) serves as a structural framework for glomerular filtration. SD components, such as Nephrin and Neph1, also function as a signaling platform through their dynamic tyrosine phosphorylation. Recently, we found signal regulatory protein (SIRP) α, a transmembrane glycoprotein that recruits phosphatases and adaptor proteins to plasma membranes, as a novel SD component.

**Methods:** The present study investigates the role of SIRPα in podocytes by immunohistochemical and biochemical analysis.

**Results:** SIRPα and Nephrin are colocalized at SD in developing and adult kidneys. Tyrosine phosphorylation of SIRPα was found to be dramatically decreased in podocyte injury models in vivo, which was inversely correlated with Nephrin phosphorylation. Using cultured cells, we demonstrated that through its extracellular domain SIRPα forms hetero-oligomer with Nephrin. Upon tyrosine phosphorylation of cytoplasmic domain of SIRPα, which is known to recruits phosphatase SHP-1 and SHP-2, SIRPα negatively regulates Nephrin phosphorylation. A specific SHP-1/2 inhibitor, NSC-87877, increased Nephrin phosphorylation, further supporting the involvement of SIRPα-SHP-1/2 in the regulation of Nephrin phosphorylation.

**Conclusions:** The expression and phosphorylation of SIRPα at the SD may play a critical role in regulating SD signaling and in glomerular disease.

## Abstract# 924
**(O-116)**

**Genetic Deletion of Angiotensin-(1-7) Receptor Mas Produces Renal Fibrosis and Dysfunction in FVB/N Mice** J.A. Magalhaes, A.C. Simoes e Silva, A.J. Ferreira, G.T. Kitten, K.D. da Silveira, N.S. de Assis, D.A. Rodrigues, R.A.S. Santos, S.V.B. Pinheiro. *Federal University of Minas Gerais, Belo Horizonte, Brazil.*

**Objectives:** It was shown that genetic deletion of Mas receptor produces glomerular hyperfiltration and albuminuria in C57BL/6 mice. Other authors suggests that Mas deletion might be nephroprotective in models of renal disease. To address those issues, we evaluated renal physiology and morphology of FVB/N *Mas*-knockout mice (MasKO), that present hypertension and metabolic syndrome.

**Methods:** Urine and plasma samples were collected for measuring diuresis, free-water, osmolal and creatinine clearances, sodium and albumin excretion. Kidneys were harvested for histology, confocal immunofluorescence of matrix proteins, RT-PCR for ACE, ACE2, $AT_1$ receptor and TGF-β.

**Results:** Compared to wild-type controls, FVB/N MasKO mice have reduced daily urine volume (38.8±3.4 *vs* 50.1±3.9 ml/g BW; $p<0.05$) and sodium fractional excretion (1.2±0.2 *vs* 2.2±0.5%; $p<0.05$), without changes in osmolal and free-water clearances. The 1.8-fold higher creatinine clearance and the 2.9-fold increase in albuminuria detected in FVB/N MasKO mice suggest glomerular hyperfiltration, as previously observed in C57BL/6 mice. Kidney histology showed reduction of glomerular tuff, glomerulosclerosis with perivascular matrix deposition, proliferation of mesangial and Bowman's epithelial cells and increased collagen IV and fibronectin.Those changes were associated with upregulation of mRNA for $AT_1$ receptor, TGF-β and ACE2.

**Conclusions:** Our results suggest that receptor Mas is critical for inhibiting renal fibrosis and dysfunction, possibly counterbalancing Ang II effects in the kidney.

## Abstract# 925
**(O-117)**

**Mechanism of TRPC6 Activation by FSGS-Causing Mutations: Antagonistic Role of Nephrin in Its Surface Expression** S. Kanda,[1,2] Y. Harita,[1,2,5] T. Sekine,[2] T. Igarashi,[2] T. Inoue,[4] S. Hattori.[1,3] *[1]Div. of Cell. Proteom., Inst. of Med. Sci., Tokyo, Japan; [2]Dep. of Pediatr., Univ. of Tokyo, Tokyo, Japan; [3]Dep. of Biochem., Sch. of Pharm. Sci., Kitasato Univ., Tokyo, Japan; [4]Dep. of Life Sci. and Med. Biosci., Waseda Univ., Tokyo, Japan; [5]Dep. of Mol. Biol., Yokohama City Univ., Yokohama, Japan.*

**Objectives:** While mutations of a podocyte slit diaphragm component, TRPC6, cause FSGS, how its Ca2+ channel activity is involved in the pathogenesis remains unclear. Meanwhile, TRPC6 activity is upregulated by tyrosine phosphorylation. Here we investigated the mechanism by which phosphorylation regulates its activity, and examined its involvement in the pathogenesis.

**Methods:** Surface expression was evaluated by biotinylation assay. Intracellular calcium concentration was measured by the fluorescence of Fura-2/AM loaded cells.

**Results:** Tyrosine phosphorylation of TRPC6 promoted its surface expression. Nephrin and PLC-γ1 competitively bound to a phosphorylated tyrosine residue of TRPC6, which was essential for its membrane insertion. While PLC-γ1 was necessary for TRPC6 trafficking, Nephrin disturbed TRPC6-PLC-γ1 interaction and interfered surface expression and activation of TRPC6. FSGS-causing mutations weakened Nephrin-TRPC6 interaction, whereby increasing its membrane expression and channel activity.

**Conclusions:** TRPC6 activity is regulated by a balance between Nephrin and PLC-γ1, and the disease-causing mutations shift the balance toward its excess activity. Regulation of its channel activity by Nephrin may underlie the pathogenesis of proteinuria.

## Abstract# 926
**(O-118)**

**Altered Renal Tubular Ultrastructure and Electrophysiology Caused by KCNJ10 Mutations in EAST Syndrome** D. Bockenhauer,[1] M. Reichold,[2] A. Zdebik,[1] E. Lieberer,[2] K. Schmidt,[1] M. Rapedius,[3] S. Bandulik,[2] C. Sterner,[2] I. Tegtmeier,[2] T. Baukrowitz,[3] S.A. Hulton,[4] B. Ben-Zeev,[5] A.J. Howie,[1] R. Warth,[2] R. Kleta.[1] *[1]UCL, London, United Kingdom; [2]Universitaet, Regensburg, Germany; [3]Universitaet, Jena, Germany; [4]Children's Hospital, Birmingham, United Kingdom; [5]CSMC, Tel-Hashomer, Israel.*

**Objectives:** Recently, we described a new multi-organ disorder caused by mutations in the potassium channel gene KCNJ10 which we named EAST syndrome (Epilepsy, Ataxia, Sensorineural deafness, Tubulopathy). We aimed to investigate the expression of KCNJ10 in mouse and human kidney, as well as electrophysiological and anatomical consequences of the mutations identified in our patients.

**Methods:** Immunohistochemical analysis of KCNJ10 expression in sections of adult mouse and human kidney. Expression of wild type and mutant KCNJ10 in experimental cells for whole cell and single channel currents; electronmicroscopy (EM) of a kidney biopsy of an EAST patient.

**Results:** KCNJ10 is expressed in cortical tubules post macula densa in mouse kidney. In human kidney, however, staining was also observed in thick ascending limb. EM showed reduced basolateral infoldings in distal cortical tubules in an EAST patient. The mutations R65P, G77R and R175Q strongly reduced KCNJ10 currents with decreased opening time, altered pH dependency and complete loss-of-function with R199X.

**Conclusions:** Expression of KCNJ10 in human kidney includes cortical thick ascending limb. Mutations reduce currents mainly by altered channel gating. The reduced reabsorption in distal convoluted tubules is reflected in altered cellular ultrastructure.

## Abstract# 927

**NHE3, the Epithelial Sodium-Proton Exchanger Is Required for Renal and Intestinal Calcium Absorption** R.T. Alexander, J. Borovac, W. Pan. *Pediatrics, University of Alberta, Edmonton, AB, Canada.*

**Objectives:** Passive paracellular proximal tubular and intestinal calcium ($Ca^{2+}$) flux is driven by active sodium ($Na^+$) absorption. NHE3 is necessary for $Na^+$ flux at these sites. However, its role in $Ca^{2+}$ homeostasis has yet to be evaluated. We thus setout to determine whether NHE3 is necessary for $Ca^{2+}$ absorption.

**Methods:** Wild-type ($NHE3^{+/+}$) and NHE3 knockout ($NHE3^{-/-}$) mice were housed in metabolic cages for 24 hours. Serum and urine was collected. mRNA expression was quantified by real-time PCR and protein expression by western blot. Intestinal $Ca^{2+}$ absorption was assessed by $^{45}Ca^{2+}$ flux across duodenum under conditions of voltage clamp in Ussing chambers. µCT analysis was used to quantify bone mineral density.

**Results:** Serum $Ca^{2+}$ was unaltered however 1,25-dihydroxyvitamin D was elevated in $NHE3^{-/-}$ mice. Urinary $Ca^{2+}$:Cr ratio was significantly increased in $NHE3^{-/-}$ mice (0.5 ±0.1 vs 0.8 ±0.1). Renal TRPV5 mRNA expression was down regulated while NCX, PMCA1b and Calbindin-$D_{28K}$ were unaltered. Both TRPV5 and Calbindin-$D_{28K}$ protein expression were decreased. Claudin-16 mRNA expression was unchanged while claudin-19 expression was decreased. Duodenal TRPV6 and PMCA1b mRNA expression was unaltered, however calbindin-$D_{9K}$, claudin-12 and claudin-15 mRNA expression were significantly decreased. Calbindin-$D_{9K}$ protein expression was also decreased. $NHE3^{-/-}$ mice had decreased $Ca^{2+}$ flux across the duodenum (61 ±12 vs 13 ±2 nmol/hr/cm$^2$) and significantly decreased cortical bone mineral density (1.09 ±0.03 vs 0.86 ±0.04 g/cm$^3$).

**Conclusions:** Our results demonstrate decreased renal tubular and intestinal $Ca^{2+}$ absorption in $NHE3^{-/-}$ mice and provide the molecular identity of a gene linking $Na^+$ to $Ca^{2+}$ absorption.

## Abstract# 928

**Estimating GFR with Equations Derived from Creatinine and from Cystatin C: Reliability in Clinical Pediatric Practice?** O. Dolomanova,[1] J. Bacchetta,[1] B. Ranchin,[1] P. Cochat,[1,2] A. Hadj-Aissa,[1,2] L. Dubourg.[1,2] *[1]Hospices Civils de Lyon, Lyon, France; [2]INSERM820, Lyon, France.*

**Objectives:** Estimation of glomerular filtration rate (GFR) in children is challenging. Reference methods are cumbersome and formulas used to estimate GFR have limitations. The aims of this study were to evaluate 1/ the new creatinine-based formula recently proposed by Schwartz and 2/ the cystatin C-derived formula proposed by Le Bricon in a cross-sectional cohort of 646 French children (882 measurements) with moderate CKD or normal GFR.

**Methods:** Formulas estimating GFR (eGFR) with creatinine (2009 Schwartz bedside formula) and cystatine C (2000 Le Bricon formula) were compared to the reference standard (inulin clearance, iGFR). Plasma creatinine was measured by a Roche compensated Jaffe technique which closely agreed with an enzymatic method; cystatin C was measured by a nephelemetric technique.

**Results:** Mean body weight, height, creatinine and cystatin C were 39±16 kg, 143±20 cm, 62±32 µmol/L and 1.0±0.4 mg/L, respectively. Mean±SD iGFR was 94±33 mL/min/1.73m². Good agreements between eGFR with both Schwartz 2009 or Le Bricon formulas and iGFR were found with a mean eGFR - iGFR of +5±25 and – 5±19 ml/min/1.73m$^2$, respectively. Accuracies 10 and 30% of Schwartz 2009 and Le Bricon formulas were 39/85% and 41/87%, respectively. The thresholds of Cystatin C for predicting iGFRs below 90 and 60 mL/min/1.73 m² at 0.9 and 1.3 mg/L showed a sensitivity/specificity of 85/79 and 83/95 %, respectively.

**Conclusions:** In a general pediatric population, the 2009 Schwartz bedside formula (k=36.5) and the Le Bricon cystatin C-derived formula are accurate and simple to use for estimating GFR in clinical practice.

## Abstract# 929

**Glomerular Filtration Rate Determined by Simultaneous Investigation of Inulin, Iohexol, Creatinine, and Formula Clearance** U.B. Berg,[1] R. Bäck,[2] G. Celsi,[1] S. Edström Halling,[1] R.T. Krmar,[1] K. Åsling-Monemi,[1] H. Öborn,[1] M. Herthelius.[1] *[1]Dept of Clinical Science, Intervention and Technology, Karolinska Institutet, Division of Pediatrics, Stockholm, Sweden; [2]Dept. of Laboratory Medicine, Karolinska Institutet, Division of Clinical Chemistry, Stockholm, Sweden.*

**Objectives:** Estimated GFR (eGFR) is often validated against plasma clearance of iohexol (Ciohexol) but Ciohexol has seldom been compared with renal clearance of inulin (Cinulin), which is the aim of the study.

**Methods:** 61 children, mean 11.6 years old, with various renal disorders were investigated simultaneously for Cinulin, Ciohexol, clearance of creatinine (Ccreat), and eGFR based on S-creatinine and height. Cinulin was investigated with continuous infusion during water diuresis. Ciohexol was based on the slope and on single sample. Iohexol was analyzed by HPLC. The data were compared with the correlation coefficients, Bland-Altman plot, and the accuracy % of Cinulin.

**Results:** The correlation coefficient between C iohexol and Cinulin was 0.92 and the mean difference between the methods 2.6 ml/min per 1.73 m$^2$. 52% of patients belonging to CKD 1-3 (GFR >30 ml/min/1.73 m$^2$) showed Ciohexol within ± 10% and 88% within ± 30% of Cinulin, but in patients with a GFR below 30 ml/min/1.73 m$^2$ (CKD 4-5), the accuracy was worse with 13% within ± 10% and 60% within ± 30%. In patients with CKD 1-3, eGFR showed 26% within ± 10% and 76% within ± 30% and in CKD 4-5 13% and 73% respectively. Ccreat overestimated Cinulin with a mean difference of 18 ml/min per 1.73 m$^2$.

**Conclusions:** Ciohexol showed good agreement with Cinulin with some overestimation in CKD 4-5. eGFR could be used in clinical practice but Ccreat overestimates GFR.

## Abstract# 930

**Molecular Expression and Electrophysiological Effects of TRPC6 Channel Over-Expression in Podocytes** L. Jiang,[1] J. Ding,[1] H. Cai,[2] L. Li,[2] J. Miao,[1] Q. Feng,[1] Q. Fan.[1] *[1]Department of Pediatrics, Peking University First Hospital, Beijing, China; [2]The Central Laboratory, Peking University First Hospital, Beijing, China.*

**Objectives:** It is known that transient receptor potential cation channel 6 (TRPC6) is one of the key molecules for maintenance the structure and function of filtration barrier in podocytes. It is important for the study on pathogenesis of podocyte injury as well as mechanism of proteinuria. This study aimed to establish a stable technique to test TRPC6 not only the expression level but also the electrophysiology in podocytes.

**Methods:** The protein expression of over-expression TRPC6 was evaluated with western blot. The intracellular $Ca^{2+}$ in podocyte was measured by laser scanning confocal microscope. The current recording was utilized in the whole-cell mode to detect the channel function of over-expression TRPC6.

**Results:** Exposure of the podocyte to carbachol (CCh) and/or 1-oleoyl-acetyl-sn-glycerol (OAG) after initiating whole-cell dialysis via the patch pipette, caused a time-dependent activation of membrane current. The current density increased dramatically in over-expression group than in control by using CCh and OAG ($P$<0.05). After inhibiting the TRPC6 ion channel by treated with U73122, the current density was decreased in both over-expression TRPC6 group and control group ($P$>0.05).

**Conclusions:** Functional changes of TRPC6 were detected for the first time in podocytes of MPC5. It was found that TRPC6 ion channel could be activated excessively with increasing of $Ca^{2+}$ concentration via over-expression of TRPC6 in cultured podocytes, which might provide a cell model for further study on TRPC6 ion channel in podocytes.

## Abstract# 931

**Chronic Vasodilation Results in Plasma Volume Expansion (PVE) – Support for the Underfill Theory** A. Fekete,[1,2] J.M. Sasser,[2] C. Baylis.[2] *[1]1st Department of Pediatrics, Semmelweis University, Budapest, Hungary; [2]Department of Physiology and Functional Genomics, University of Florida, Gainesville, FL, United States.*

**Objectives:** In pregnancy, systemic vasodilation results in renal Na and water retention and consequent PVE. Previously we showed that increased renal phosphodiesterase 5 (PDE5) activity inhibits the natriuretic response, contributing to the PVE in pregnancy.

**Methods:** To test the "underfill hypothesis" of pregnancy, we produced chronic vasodilation by 14 days po. nifedipine (NIF, 10 mg/kg/day) or sodium nitrite (NaNO2, 70 mg/kg/day) to female Sprague Dawley rats vs those on control diet (CON). Mean arterial blood pressure (MAP) was monitored by telemetry, PV was determined by the Evans-blue method, hematocrit (Hct), osmolarity (Posm), Na (PNa), and total protein (CA) concentrations were also measured. mRNA expression and protein abundance of PDE5A and its isoforms were determined in kidney and aorta.

**Results:** MAP was reduced in NIF by 6.7%, in NaNO2 rats by 7.6% vs. baseline and CON ($p$<0.05). NIF and NaNO2 lowered Hct and increased PV. Posm and PNa both fell, as did CA (*$p$<0.05 vs. CON).Renal medullary PDE5A and PDE5A1 mRNA expression increased both in NIF and NaNO2 group vs CON ($p$<0.05), while PDE5A2 isoform was the same in all groups. PDE5A protein level increased both in NIF and NaNO2 vs CON ($p$<0.05), however, neither renal cortical nor aortic PDE5 level was changed by vasodilator treatment.

**Conclusions:** A primary vasodilation produced by NIF or NaNO2 drives increased medullary PDE5 expression, renal Na retention causing volume expansion and partial "refilling" of the vasculature. These responses closely resemble the changes in normal pregnancy with hemodilution due to marked PVE.

**DISCLOSURE:** *Baylis, C.*: Other, Forrest Pharma.

## Abstract# 932

**ANGPTL3 Directly Binds and Activates Integrin αVβ3 in Podocytes**

X. Gao,[1] H. Xu,[1] C. Feng,[2] H.-M. Liu,[1] J. Rao.[1] *[1]Department of Nephrology and Rheumatology, Children's Hospital of Fudan University, Shanghai, China; [2]Institute of Nutritional Sciences, Shanghai Institutes for Biological Sciences, Chinese Academy of Sciences, Shanghai, China.*

**Objectives:** We investigated whether glomerular can express ANGPTL3 and whether ANGPTL3 can directly or indirectly bind to integrin αVβ3 in podocytes.
**Methods:** We studied normal and MCD kidneys from humans and rats to evaluate the location and sub cellular features of ANGPTL3 in the podocytes. Using ELISA and quantitative real-time PCR, we evaluated ANGPTL3 produced by cultured podocytes. And we evaluated the bind model of ANGPTL3 and integrin αVβ3 in podocytes by confocal microscopy and immuno-precipitation.
**Results:** Our results showed that ANGPTL3 were specially localized to the podocyte foot processes in glomerular. Quantitative analysis revealed a significantly higher concentration of colloid-gold particles in the nephrotic podocytes compared to normal podocytes ($P<0.05$). Results of ELISA and real-time PCR suggested that the ability of cultured podocytes to synthesize ANGPTL3, which was greatly enhanced after ADR treatment. ANGPTL3 could directly bind to integrin β3. The active integrin β3 expression was significantly up-regulated in podocytes over-expressing ANGPLT3 compared with control cells ($P<0.01$). Compared with an un-knockdown ANGPTL3 podocyte, ANGPTL3 knockdown cell did not show significant the active integrin β3 up-regulation after ADR-stimulation, suggesting that ANGPTL3 could regulate integrin β3 expression under certain pathological conditions.
**Conclusions:** It is first report to demonstrate that ANGPTL3 is found mainly within the foot processes of podocyte. And ANGPTL3 is a new key ligand that induces integrin β3 activation in podocytes.

## Abstract# 933

**Chinese Medicine Extractum Trametes Robiniophila Murr Improves Actin Rearrangement of Podocytes**

X. Gao, H. Xu. *Department of Nephrology and Rheumatology, Children's Hospital of Fudan University, Shanghai, China.*

**Objectives:** Chinese medicine Extractum trametes robiniophila murr is a kind of fungus. Recent study has suggested that it could be capable to efficiently improve the effacement of foot process in Adriamycin (ADR) induced rats. The aim of this study was to identify whether Extractum trametes robiniophila murr modulates actin rearrangement of podocytes and signaling molecules such as ACTN4, nephrin and podocin.
**Methods:** The podocytes were divided into four groups as follows: Extractum trametes robiniophila murr group (Extractum trametes robiniophila murr 10mg/ml + ADR 0.5umol/L); DXM group (DXM 0.1umol/L + ADR 0.5umol/L); ADR group (ADR 0.5umol/L) and control group. F-actin of podocytes in each group were stained by fluorescent phallotoxins of invitrogen and observed by confocal microscope. The expression levels of ACTN4, nephrin and podocin in each group were tested by real-time PCR and western blot.
**Results:** ADR group had significant actin rearrangement. However, in Extractum trametes robiniophila murr group, podocytes showed a more powerful resistance to actin rearrangement of ADR induction than DXM group. In ADR group, the expression levels of ACTN4, nephrin and podocin were significantly higher than those in control group. The expression level of ACTN4 in Extractum trametes robiniophila murr group was almost same as that in control group, while in DXM group, the expression level of ACTN4 was higher than that in control group. Extractum trametes robiniophila murr could not regulate the altering expression of nephrin and podocin after ADR treatment.
**Conclusions:** Extractum trametes robiniophila murr is a powerfully potential modulator to actin rearrangement of podocytes.

## Abstract# 934

**Over-Expression of Prohibitin Suppresses Renal Interstitial Fibroblasts Extra-Cellular Matrix Expression and Intercepts Smad3 Nuclear Translocation**

W. Guo, H. Xu, J. Chen, Q. Shen, H.-M. Liu, W.-Y. Huang. *Department of Nephrology and Rheumatology, Children's Hospital of Fudan University, Shanghai, China.*

**Objectives:** To investigate effects of PHB on cell growth and ECM expression induced by TGF-β1 in renal interstitial fibroblasts, and ascertain whether PHB can suppress TGF-β1-mediated Smad signaling passway.
**Methods:** Cell growth curves were detected. PHB, ECM proteins fibronectin (Fn) and collagen type III (Col III), and matrix metalloproteinase-1(MMP-1) protein and mRNA expression were detected. Phosphorylation and activation of Smad3 by TGF-β1 and Smad7 expression were detected, and phosphorylation of Smad3 and its association with Smad4 in cells after TGF-β1 stimulation were further investigated by co-immunoprecipitation.
**Results:** Stimulation of cells with 1ng/ml TGF-β1 induced a remarkable cells growth. Transfected with PHB plasmid suppressed cells growth, and the cell growth was 27%, 48% and 35% inhibited after transfection for 24, 48 and 72 h, respectively. TGF-β1 induced Col III and Fn proteins and mRNA expression and the effects were dose-dependent. Over-expression of PHB substantially attenuated the up-regulation of Col III and Fn, although it had almost no effect on the up-regulation of MMP1 induced by TGF-β1. Smad3 was phosphorylated after TGF-β1 treatment. Co-immunoprecipitation revealed that p-Smad3 was physically associated with Smad4. However, Over-expression of PHB did not significantly affect Smad3 phosphorylation and its association with Smad4.
**Conclusions:** Extraneous PHB suppresses renal interstitial fibroblasts proliferation and ECM expression induced by TGF-β1, and suppresses profibrotic cytokine TGF-β1 by intercepting Smad3 nuclear translocation.

## Abstract# 935

**PEA3 Synergistically Enhance Smad2 Regulate TGF-beta-Induced RGC-32 Gene Transcription in Epithelial-Mesenchymal Transition**

W.-Y. Huang,[1] H. Xu,[1] P.A. Jose,[2] S. Chen.[2] *[1]Department of Nephrology and Rheumatology, Children's Hospital of Fudan University, Shanghai, China; [2]Department of Pediatric Nephrology, Georgetown University Medical Center, Washington, DC, United States.*

**Objectives:** This study was to investigate molecular mechanism of RGC-32 gene regulation in TGF-beta-induced EMT.
**Methods:** (1) Plasmid Construction;(2) RNA interference;(3) Transfection and luciferase activity assay;(4) EMSAs.
**Results:** (1) TGF-beta1 can active RGC-32 gene expression driven by pLuc-1500, pLuc-2107, or pLuc+844, but not by pLuc-85, pLuc-228, pLuc-600, or pLuc-1021 ($P<0.001$) after treated by TGF-beta1. It is indicated binding site which involved TGF-beta1 has located at RGC-32 promoter region from -1021 to -1500. SBE (GTCTGGAC) is located at the site from -1344 to -1337 of RGC-32 promoter, as well as a PEA3 binding site (AGGAAG) align at 35bp upstream of SBE from -1379 to -1374. Mutation of Smad site significantly inhibited RGC-32 promoter activity ($P<0.01$), double mutations of Smad and PEA3 sites completely abolished promoter activity. (2) Smad2, Smad4 and PEA3 proteins are contribute to regulation of TGF-beta-induced RGC-32 expression: Smad2 and Smad4 antibody shown the same degree of supershift, Smad2, Smad4 and PEA3 proteins are contribute to regulation of TGF-beta-induced RGC-32 promoter activity. Smad2 was essential for TGF-beta induction of RGC-32 promoter, knockdown of PEA3 alone did not significantly inhibit RGC-32 promoter, but knockdown of both Smad2 and PEA3 completely blocked TGF-beta function.
**Conclusions:** SBE and PEA3 binding site were found at the upstream of RGC-32 promoter. Smad2 and PEA3 synergistically regulate TGF-beta induction of RGC-32 transcription during EMT.

## Abstract# 936

**Hyperfiltration Affects Accuracy of Creatinine but Not Cystatin C or Beta Trace Protein eGFR Measurement**

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**Objectives:** Surrogate markers, such as creatinine, cystatin C, and beta trace protein have been used to estimate Glomerular Filtration Rate (eGFR). The accuracy of eGFR for these markers may be altered with hyperfiltration and differences in filtration fraction. We hypothesize that the accuracy of creatinine for eGFR may be affected by hyperfiltration and estimated renal plasma flow (ERPF).
**Methods:** A total of 127 pediatric patients with various renal diseases underwent simultaneous measurements of both GFR using $^{51}$Cr-ethylenediamine tetra-acetic acid (EDTA) renal scan and ERPF ($^{131}$I Hippurate clearance) to calculate the filtration fraction (FF=GFR/ERPF). The eGFRs were calculated using the commonly used Schwartz's (creatinine), Filler's (cystatin C) and Benlamri's (beta trace protein) formulae. Agreement of the various eGFRs with the measured isotope GFRs was assessed by Bland&Altman plots. Correlation analysis was performed using non-parametric tests to compare FF with eGFR-GFR.
**Results:** The 127 children at a median age (with 25$^{th}$ percentile, 75$^{th}$ percentile) of 11.9 (8.5, 14.9) years had a mean $^{51}$Cr EDTA GFR of 100.6 ± 32.1 ml/min and a median $^{131}$I hippurate clearance (ERPF) of 588 (398, 739) mL/min/1.73 m$^2$. Mean filtration fraction was 17.7± 4.5% with no correlation between the FF and the error for Cystatin C eGFR and BTP eGFR, whereas there was a significant negative correlation between the error for the Schwartz eGFR and the FF.
**Conclusions:** There is a significant negative correlation between the error for the Schwartz eGFR and the FF. Cystatin C and BTP are not affected by differences in filtration fraction.

## Abstract# 937

**mTORC1 Activation Induces Unfolded Protein Response of Podocytes in Nephrotic Syndrome**

N. Ito, K. Yan. *Pediatrics, Kyorin University School of Medicine, Tokyo, Japan.*

**Objectives:** The pathomechanism of minimal change nephropathy (MCN) has remained unclear. We previously showed that unfolded protein response (UPR) caused by lack of ATP in the endoplasmic reticulum might underlie the pathomechanism of MCN. The purpose of this study was to further investigate

whether ATP consuming signaling cascade, mammalian target of rapamycin complex 1 (mTORC1), precedes UPR in MCN.
**Methods:** Rat MCN was induced by puromycin aminonucleoside (PAN). Isolated glomeruli and kidney tissue from rats treated with or without mTOR inhibitor everolimus were subjected to immunohistochemistry, dual-immunofluorescence and confocal microscopy, and Western blot analysis for mTORC1 and UPR phosphorylation. Immortalized mouse podocytes were treated with PAN in a time dependent manner, and subjected to Western blot analysis for mTORC1 and UPR phosphorylation.
**Results:** Rats developed significant proteinuria from day 2 after injection of PAN. Activation of both UPR and mTORC1 in glomerular podocytes was reveled at day 1 whereas expression of nephrin and podocin was reduced from day 2. Interestingly, mTORC1 activation was revealed to precede UPR when cultured podocytes were treated with PAN. Pre-treatment of everolimus at day 1 before PAN injection interfered with proteinuria compared with placebo-treated rats. The activation of both mTORC1 and UPR in the glomerular podocytes of rats treated with everolimus was decreased compared with placebo-rats.
**Conclusions:** mTORC1 may be a potential signaling cascade that initiates UPR through dysregulation of energy system. Investigation of precise regulatory mechanism of ATP in podocytes may contribute to develop new therapeutic reagents for MCN.

## Abstract# 938

**Characterization of the Constitutively Active V2 Receptor Mutants Conferring NSIAD** T. Julie,[1,2] A. Mohammed,[2] R. Bruno,[3] M. Christiane,[2] D. Thierry,[2] M. Denis.[1,2] *[1]Néphrologie Pédiatrique - CHU, Montpellier, France; [2]CNRS UMR 5203,IGF and INSERM 661, UM 1, Montpellier, France; [3]Néphrologie Pédiatrique - HFME, Lyon, France.*
**Objectives:** Patients having the nephrogenic syndrome of inappropriate antidiuresis (NSIAD) exhibit hyponatremia and inappropriate elevated urinary osmolality associated to low plasma vasopressin levels. All patients reported to date present either the R137C or R137L V2 mutated receptor. While the clinical features have been characterized, the molecular mechanisms of functioning of these two mutants remain elusive. In the present study, we compare the pharmacological properties of R137C and R137L mutants with the wild-type and the V2 D136A receptor, the latter being reported as a highly constitutively active receptor.
**Methods:** We have performed binding studies, second messenger measurements and BRET experiments in order to evaluate the affinities of V2 receptor ligands, their agonist and antagonist properties and the ability of the mutated receptors to recruit β-arrestins, respectively.
**Results:** The R137C and R137L V2 receptors exhibit small constitutive activities regarding the $G_s$ protein activation. In addition, these two mutants induce a constitutive β-arrestin recruitment. Of interest, they also exhibit weak sensitivities to V2 -agonist and to V2 - inverse agonist in term of $G_s$ protein coupling and β-arrestin recruitment.
**Conclusions:** The small constitutive activities of the mutants and the weak regulation of their functioning by agonist suggest a poor ability of the antidiuretic function to be adapted to the external stimuli, giving to the environmental factors an importance which can explain the phenotypic variability in patients having NSIAD.

## Abstract# 939

**Population Pharmacokinetics of Mizoribine in Pediatric Patients with Renal Disease** H. Kaneda, O. Uemura, K. Ushijima, K. Ohta, Y. Gotoh, K. Satomura, M. Shimizu, T. Nagai, M. Fujieda, M. Morooka, T. Yamada, M. Yamada, N. Wada, M. Takaai, Y. Hashimoto. *The Japanese Pediatric Pharmacokinetics Study Group for Kidney disease, Nagoya, Japan.*
**Objectives:** Mizoribine (MZR) is an inhibitor of purine synthesis for an immunosuppressive agent. We empirically noted that the clinical efficacy of MZR is relatively lower in pediatric patients compared with adults, but there have been few pediatric pharmacokinetic studies on MZR reported.
**Methods:** One hundred and five pediatric patients in our institutions were enrolled. The most frequent disease was idiopathic nephrotic syndrome in 74 patients, followed by secondary glomerulonephritis and IgA nephropathy. The single dose of MZR was 1-14 mg/kg. Population pharmacokinetic (PPK) analysis was performed based on the blood MZR levels in the patients.
**Results:** The mean absorption lag time and absorption rate constant estimated to be 0.501 h and 0.761 $h^{-1}$, respectively, showing that MZR slowly appeared in the circulation and blood level elevation was also slow compared to those in adults. Using the calculated PPK parameters, the $AUC_\infty$ and $C_{max}$ were compared based on Bayes' theorem among individual patients. A significant correlation was noted between the age and $AUC_\infty$ ($P$<0.0001), and the $C_{max}$ was also significantly different among the age groups ($P$<0.05). Based on these findings, the required MZR dose per body weight increased as the age decreased.
**Conclusions:** The regular dose of MZR (2-3 mg/kg/day) is insufficient for pediatric patients. To obtain the expected clinical effect, high-dose treatment should be applied using the MZR-PPK model established in this study.

## Abstract# 940

**Inappropriate Antidiuresis without Elevation of Anti-Diuretic Hormone** A. Kogon, R. McDonald. *Pediatrics, Seattle Childrens Hospital, Seattle, WA, United States.*
**Objectives:** To describe a child with hyponatremic seizures and apparent syndrome of inappropriate antidiuresis (SIADH), with a mutation in the arginine vasopressin receptor 2 (AVPR2) gene.
**Methods:** A 13 month old previously healthy boy presented with a generalized tonic clonic seizure. On exam, the patient was post-ictal, euvolemic and normotensive. His only laboratory abnormality was a serum sodium of 122 mEq/L. Initially, he was treated with a normal saline infusion for 24 hours and serum sodium improved to 128 mEq/L. His intravenous fluids were then held and he maintained hydration with oral fluids. Twelve hours later, his serum sodium was 118 mEq/L. Concurrently, all other serum electrolytes, blood urea nitrogen and creatinine were normal. His fractional excretion of sodium was 1.2%, plasma and urine osmolalities were 252 and 392 mOsm/L respectively, anti-diuretic hormone level was 1.8 pg/ml (range:1-13.3), serum aldosterone was 13 ng/dl (range:2-37) and serum renin was <0.1 ng/ml/hr. Subsequently, he was successfully treated with mild fluid restriction and table salt supplements approximating 10 meq/kg/day of sodium chloride, to maintain serum sodium >130 mEq/L.
**Results:** AVPR2 gene sequencing revealed a p.Arg137Cys mutation. This gene mutation, located on the long arm of the X chromosome, causes a gain of function of the arginine vasopressin receptor that induces constitutive receptor activation in the absence of arginine vasopressin. Over the subsequent 6 months, the patient's table salt supplements were decreased gradually, maintaining serum sodium in the low normal range.
**Conclusions:** AVPR2 mutations should be considered in infants and children presenting with symptomatic hyponatremia without laboratory evidence of SIADH.

## Abstract# 941

**Mutations and Expression of TRPC6 Gene in Chinese Children with Steroid-Resistant Nephritic Syndrome** X.-Y. Kuang, Y. Shi, X.-L. Zhang, W.-Y. Huang, H. Xu. *Department of Nephrology and Rheumatology, Children's Hospital of Fudan University, Shanghai, China.*
**Objectives:** Autosomal-recessive Steroid-resistant nephritic syndrome is a common cause of ESRD in children. Mutations in the TRPC6 gene are a major cause of focal segmental glomerulosclerosis. However, whether the mutations of TRPC6 are also responsible for Chinese children with SRNS? And what's the differentiation of the TRPC6 expression between SRNS children and SSNS children? It's still unknown.
**Methods:** Genomic DNA was isolated from blood samples of 37 SRNS patients, 22 steroid sensitive (SSNS) patients and 16 normal children, and we used PCR-amplified each of 13 exons of TRPC6 gene sequence analysis. Immunohistochemistry method was used to detect the variance of TRPC6 expression between SRNS and SSNS children who had renal biopsy.
**Results:** One novel synonymous mutation G42G (1151C>A) was identified in three SRNS patients, which had not been found in the other two groups. Three biallelic SNPs were also found, including P15S (40C>T, rs3802829), -254C>G (rs3824934), and 237G>A. P15S and -254C>G were identified in the promoter region, maybe they could take part in the regulation of this gene, and it's need further study. Immunohistochemistry showed that the expression of TRPC6 in nephric tubule were significantly higher in SRNS patients than in SSNS children ($P$<0.01).
**Conclusions:** As far, we had not found causative mutation in TRPC6 gene in Chinese children with SRNS. Maybe TRPC6 mutations were not a major cause of SRNS children in China, though the cohort was small. But, the expression of TRPC6 was higher in SRNS children than in SSNS children. And the two SNPs might act like a regulator of the promoter region of this gene.

## Abstract# 942

**Isolation of Human Renal Mesenchymal Stem Cells from the Urine of Pediatric Patients with Nephropathy** N. Kumagai,[1] M. Satou,[1] N. Sugawara,[1] F. Kamada,[1] T. Morimoto,[1] Y. Kondo,[2] S. Tsuchiya.[1] *[1]Department of Pediatrics, School of Medicine, Tohoku University, Sendai, Miyagi, Japan; [2]Department of Medical Informatics, School of Medicine, Tohoku University, Sendai, Miyagi, Japan.*
**Objectives:** CD133-CD146+ cells in renal the interstitium are mesenchymal stem cells that can undergo multipotent differentiation. We developed a highly effective method to culture renal cells from voided human urine. We aimed to culture CD133-CD146+ cells from the voided urine of pediatric patients with nephropathy to determine whether these cells could differentiate into osteogenic and adipogenic cells.
**Methods:** Cells were cultured from the voided urine of pediatric patients with Alport syndrome, Henoch-Schonlein nephritis, IgA nephropathy, post-streptococcal acute glomerulonephritis, and lupus nephritis. mRNA was

extracted from cultured cells, and RT-PCR detection of CD133 and CD146 mRNAs was performed. CD133-CD146+ cells were sorted by FACS and cultured to determine their ability to differentiate into osteogenic and adipogenic cells on media specifically suitable for differentiation into each cell type.
**Results:** Cells could be cultured very effectively from the urine of patients with hematuria and/or proteinuria. RT-PCR showed expression of CD133 and CD146 mRNAs irrespective of whether the patients had nephropathy. Histochemical staining showed that the cells could differentiate into adipocytes and osteoblasts on the appropriate culture media.
**Conclusions:** Renal mesenchymal stem cells can be cultured from the urine of pediatric patients with nephropathy. Urine may be a useful tool for regenerative medicine for kidney disease.

## Abstract# 943

**Mechanisms of Interleukin-1beta Increase Lipids Accumulation in Podocytes** Q. Li, G. Zhang. *Children's Hospital of Chongqing Medical Universtiy, Chongqing, China.*
**Objectives:** Low-density lipoprotein receptor (LDLr) and 3-hydroxy-3-methylglutaryl coenzyme A (HMG-CoA) reductase are normally regulated via a feedback system that is dependent on intracellular cholesterol levels. This study was designed to investigate the mechanism by which interleukin-1beta dysregulation of the LDLr and HMG-CoA reductase expression, causing lipids accumulation in podocytes.
**Methods:** We explored the effect of 5~20 ng/ml IL-1beta on the levels of intracellular cholesterol and the expression of LDLr, HMG-CoA reductase, sterol regulatory element binding protein (SREBP)-2, SREBP cleavage-activating protein (SCAP) and Insulin induced gene-1 (Insig-1) in podocytes in the presence of 200μg/ml native LDL.
**Results:** LDL loading increased intracellular cholesterol levels and lipid droplets in podocytes, thereby reduced LDLr and HMG-CoA reductase expression. However, IL-1beta increased the ratio of SCAP to Insig-1 and SREBP-2 expression, and overroded the suppression of LDLr and HMG-CoA reductase in the presence of 200μg/ml LDL, which resulted in further increased cholesterol levels in podocytes.
**Conclusions:** These observations demonstrated that IL-1beta could increase the rate of exogenous LDL-cholesterol uptake and the rate of endogenous cholesterol synthesis in podocytes by blocking the downregulation of LDLr and HMG-CoA reductase that normally occurs in response to the increased intracellular cholesterol levels.

## Abstract# 944

**Role of Altered Renal Lipid Metabolism and Mechanism for Renal Disease in Inflammatory Stress** Q. Li, Z. Xu. *Children's Hospital of Chongqing Medical Universtiy, Chongqing, China; Centre for Lipid Research, Chongqing Medical University, Chongqing, China.*
**Objectives:** To investigate the mechanism by which inflammatory mediators interfere with cholesterol sensor SREBP cleavage-activating protein (SCAP) and its biological consequences.
**Methods:** C57BL/6J mice that were fed a diet a western diet (WD) and were randomly assigned to receive either injections of 0.5 ml 10% casein or vehicle (control) for 14 weeks, terminal blood samples were taken for plasma cholesterol, triglycerides, LDL, HDL, amyloid A (SAA) and IL-6 assays. The lipid accumulation in kidney was evaluated by Oil Red O staining. Human mesangial cell line(HMCs) were transiently transfected with pCMVSport6-SCAP, and subjected to cholesterol loading or inflammatory stress by adding LDL or IL-1β. Confocal microscopy was used to investigate the translocation of SCAP in human mesangial line (HMCS).The expression of SCAP,SREBP2,LDL receptor(LDLr),and HMGCoA reductase (HMGCoA) were examined by real-time PCR and western blot.
**Results:** We demonstrated that cholesterol loading retained SCAP in endoplasmic reticulum (ER), thereby decreasing mRNA and protein expression of LDLr and HMGCoAR. However, exposure to imflammtion caused over-expression of SCAP, enhancing its translocation from the ER to the Golgi. This resulted in increased cholesterol accumulation with increased LDLr and HMGCoAR activities in HMCs and kindey. Over-expression of SCAP in HMCs overrode the ER retention induced by cholesterol and resulted in increased translocation of SCAP from ER to Golgi.
**Conclusions:** Inflammation promotes abnormal translocation of SCAP from ER to Golgi and plays an important role in lipid accumulation in HMCs and kidney.

## Abstract# 945

**Mechanisms of Deposition of ApolipoproteinB100-Containing Lipoprotein in Paediatric Primary Nephrotic Syndrome** Q. Li, G. Zhang. *Children's Hospital of Chongqing Medical Universtiy, Chongqing, China.*
**Objectives:** This study was designed to investigate the mechanisms abnormal deposition of apoB100-containing lipoprotein in paediatric primary nephrotic syndrome(PNS).
**Methods:** The kidney tissues specimens were obtained by percutaneous needle biopsy from 31 patients with PNS in children, which consisted of minimal change nephrotic syndrome (n=4) and non-minimal change nephrotic syndrome (n=27). Local expression of interleukin-1beta (IL-1beta), transforming growth factor- beta1(TGF- beta1), low-density lipoprotein receptor (LDLr), 3-hydroxy-3-methylglutaryl coenzyme A (HMG-CoA) reductas, sterol regulatory element binding protein (SREBP)-2, SREBP cleavage-activating protein (SCAP) and apoB100 in kidney were detected by immunohistochemistry, renal histopathology were evaluated by HE and PAS staining. Six normal control kidney tissues were obtained from compeletly normal regions of surgical biopsy form children with early renal tumor.
**Results:** The expression of IL-1 beta, TGF- beta1, LDLr, HMGCoA reductase, SREBP-2, SCAP and apoB100 in kidney of NMCNS group were higher than that of normal group and MCNS group respectively($p<0.05$). Deposition of apoB100 was significantly correlated with the expression of IL-1beta, TGF-beta1, LDLr, HMG-CoA reductase, SREBP-2, SCAP respectively($p<0.05$), but not with their plasma lipids levels. The expression of IL-1 beta, TGF-beta1 was significantly correlated with the expression of LDLr, HMGCoA reductase, SREBP-2 and SCAP respectively($p<0.05$).
**Conclusions:** Inflammation can upregulate expression of the pathway of SCAP-SREBP-2–LDLr/HMGCoA reductase, which may be the cause of the increasing apoB100-containing lipoprotein accumulating in kidney.

## Abstract# 946

**Various Glomerular Filtration Rate Measurements Using Cystatin C Level in Korean Children with Renal Disease** I.S. Lim. *Department of Pediatrics, College of Medicine, Chung-Ang University, Seoul, Korea.*
**Objectives:** Glomerular filtration rate (GFR) is a fundamental parameter in assessing renal function and predicting the progression of chronic renal disease. Because the use of serum creatinine has several disadvantages, many studies have investigated the use of cystatin C for estimating GFR. This study was performed to investigate the clinical usefulness of GFR estimation by various formulas with serum cystatin C level.
**Methods:** We retrospectively analyzed 416 patients with various renal diseases and classified them into two groups according to creatinine clearance (Group 1: CrCl >90 mL/min/1.73m$^2$, Group 2: CrCl <90 mL/min/1.73m$^2$). We compared GFRs calculated by the Schwartz and Counahan formula using serum creatinine and those by Filler et al, Bokencamp et al, and Grubb et al formula using serum cystatin C.
**Results:** GFR determined by the Schwartz formula had the highest correlation to creatinine clearance (r=0.627, *P*=0.00). GFR determined by various formulas using cystatin C had lower correlation to creatinine clearance (r=0.171, r=0.182 and r=0.228, *P*<0.01). The Schwartz and Counahan formulas showed greater diagnostic accuracy in detecting decreased GFR than formulas using cystatin C, especially in group 2 (areas under the curve: Schwartz, 0.625; Counahan, 0.594; Filler, 0.532; Bokencamp, 0.512; and Grubb, 0.524).
**Conclusions:** GFRs determined by serum creatinine showed higher correlation coefficient than those determined by cystatin C. The formulas using cystatin C were not superior to those using serum creatinine in detecting decreased GFR. So we concluded that cystatin C measurement was not satisfactory for assessing GFR in pediatric patients whose renal function was not severely decreased.

## Abstract# 947

**Effect of Angptl3 and Dexamethasone on Podocyte Cytoskeleton and Podocyte Associated Molecules** H.-M. Liu, X. Gao, H. Xu, W.-Y. Huang, J. Rao. *Department of Nephrology and Rheumatology, Children's Hospital of Fudan University, Shanghai, China.*
**Objectives:** To explore the effect of Angiopoietin-like 3 (ANGPTL3) and dexamethasone (Dex) on reorganizing the podocyte cytoskeleton and podocyte associated molecules changing in vitro.
**Methods:** Defined cultured murine podocytes to 7 different groups in vitro. PAN–induced podocyte injury, pretreatment with or without Dex (PD group, P group). Transfected podocyte by using pcDNA3.1 ANGPTL3 pretreatment with or without Dex (TD group, T group). Knock-down ANGPTL3 was S group. C group was control group and Tc group was empty-transfected group.
**Results:** The fluorescence intensity of F-actin was lower and disassembling of cytoskeleton architecture was observed in PAN induced podocyte injury–P group and paracelluar permeability to FITC- BSA was increased. The expression of ANGPTL3 was increased and changes of podocyte associated molecules were observed. The similar changes in F-actin (398.4±66.20), FITC-BSA (24.5%±5.3%) and podocyte associated molecules could be found in T group. Pretreatment with Dex can protect podocyte from PAN-induced injury. The fluorescence intensity of F-actin returned to stronger (1030.0±101.24) and less disassembling of cytoskeleton architecture was observed in PD group and paracelluar permeability to FITC- BSA was decreased (6.8%±2.6%). The expression of ANGPTL3 was also decreased. In TD group we also found the protection effects of Dex on podocyte injury.

**Conclusions:** The expression of podocyte associated molecules were changed in PAN–induced podocyte injury. High expression of ANGPTL3 may induce the similar podocyte injury as PAN. ANGPTL3 may take part in the process that Dex protects and repairs podocyte injury.

## Abstract# 948

**Role of NKCC in BK Channel-Mediated Net K Secretion (JK) in the Cortical Collecting Duct (CCD)** W. Liu, Y. Henandez, B. Zavilowitz, L.M. Satlin. *Pediatrics, Mount Sinai School of Medicine, NY, NY, United States.*

**Objectives:** Apical SK/ROMK and Ca-activated BK channels mediate baseline and flow-stimulated JK (pmol/min.mm), respectively, in the CCD. Whereas the SK/ROMK channel is restricted to principal cells (PCs), BK channels are detected in both PCs and intercalated cells (ICs). An unresolved question is whether ICs, with little functional basolateral Na-K-ATPase, can sustain K secretion. Immunodetectable 'secretory' Na-K-2Cl cotransporter (NKCC1) is present along the basolateral membrane of ICs in rat medullary CD (Ginns et al, JASN 7:2533, 1996). We sought to examine the contribution of NKCC1 to BK channel-mediated JK in the CCD of the NZW rabbit. Quantitative PCR was performed on pooled samples of microdissected CCDs ( 10 mm total length/sample) using NKCC1- and 18S-specific primers and probes and the TaqMan assay; gene-specific transcripts were identified in all 4 samples. Next, JK and net Na absorption (JNa) were measured in CCDs microperfused in vitro at fast (~5 nl/min.mm) flow rates in the absence and presence of bumetanide (10 μM) added to the bath. In 3 CCDs, this low concentration of bumetanide completely inhibited flow-stimulated JK (-15.8± 2.1 to -8.4± 3.8; $p<0.05$), and had no effect on JNa (p=NS). Bumetanide (100 μM) did not block the flow-induced [Ca2+]i transient necessary for BK channel activation (n=4). These results suggest that BK-mediated flow-stimulated JK is dependent on a basolateral bumetanide-sensitive transport pathway, proposed to be NKCC1, a cotransporter identified, at least at the level of mRNA, in the CCD. Studies are underway to more precisely define the cell type to which this cotransporter is localized.

## Abstract# 949

**The Matrix Metalloproteinases and Tissue Inhibitors Its in Urine as an Indicator of Ongoing Chronic Renal Process** V.N. Luchaninova, O.V. Semeshina, A.N. Nee, O.V. Bykova. *Children Diseases, Vladivostok State Medical University, Vladivostok, Russian Federation.*

**Objectives:** The matrix metalloproteinases (MMPs) are a large family of enzymes that break down different components of the extracellular matrix. The action of these enzymes regulates by tissue inhibitors of metalloproteinases (TIMPs). The purpose- to study a level excretion with urine MMPs -9- gelatinase and to define ratio MMPs / TIMPs at children with various renal diseases.

**Methods:** 38 children with diseases of kidneys range in age from 3 to 16 years old (middle age - 8, 4±0, 27) are surveyed. 11 persons (1 gr.) with a acute pyelonephritis, 19 (2 gr.) - with a chronic pyelonephritis, 8 (3 gr.) - with chronic glomerulonephritis. The control group is presented by 14 healthy children.

**Results:** Level of activity MMPs-9 at healthy children 0,909±0,17 ng/ml [0,244-2,408]. Ratio MMPs / TIMPs was 0,025±0,004. At children 1 gr. the significant increase in level MMPs-9 (1,550±0, 28) with simultaneous reduction of ratio MMPs / TIMPs (0,021±0,005) are determined. Uric excretion proteolytic peptides at children with chronic process in kidneys authentically decreased. Values were accordingly: 2 gr.- MMPs -9 - 0,938±0,2 ng/ml [0,042-2,743], p=0,053; 3 gr.- MMPs-9 - 0,076±0,03 ng/ml [0,010-0,251], p=0,0004 in comparison with parameters in control group. At the same time ratio MMPs / TIMPs increased (0,029±0,004; 0,031±0,005), that indicates on the decrease tissue inhibitors of TIMPs at the nephrosclerosis.

**Conclusions:** Disbalance in system MMPs-TIMPs depending on duration of inflammatory process in kidneys was revealed. Level urinary TIMPs and ratio MMPs/TIMPs are a sensitive markers of progressing CRD and nephrosclerosis.

## Abstract# 950

**Sleep Deprivation Induces Diuresis and Natriuresis in Healthy Children** B. Mahler,[1] M. Scroeder,[2] K. Kamperis,[1] S. Rittig,[1] J.C. Djurhuus.[2] *[1]Pediatrics, Aarhus University Hospital, Skejby, Aarhus, Denmark; [2]The Institute of Clinical Medicine, Aarhus University, Skejby, Aarhus, Denmark.*

**Objectives:** During sleep urine production is down regulated. This study was undertaken in order to show the effect of sleep deprivation on urine production in children at an age where enuresis is still prominent.

**Methods:** Twenty healthy children underwent two 24-hour in-patient studies, they were randomly assigned to either normal sleep or sleep deprivation in the two study periods. Diet and fluid intake were standardized. Blood pressure and heart rate were non-invasively monitored. Blood was analyzed for plasma antidiuretic hormone(AVP), atrial natriuretic peptide(ANP), angiotensin II, aldosterone, and renin.

**Results:** Successful sleep deprivation(SD) was achieved in all participants with a minimum of 4h50min postponement of time to sleep. During SD markedly larger amounts of urine was produced(477±145 vs 291±86 ml, $p<0.01$). SD dramatically increased urinary excretion of sodium(0.17±0.05 vs 0.10±0.03 mmol/kg/h) and induced a significant fall in night-time plasma AVP($p<0.01$), renin($p<0.05$), angiotensin II($p<0.001$) and aldosterone($p<0.05$) whereas plasma ANP levels were not influenced by SD(p=0.807). The night-time blood pressure and heart rate was significantly higher during SD(MAP:78.5±8.0 vs 74.7±8.7 mmHg, $p<0.001$).

**Conclusions:** Sleep deprivation leads to natriuresis, thus increasing the nocturnal diuresis. The underlying mechanism may be the reduced night-time dipping in blood pressure and a decrease in RAAS levels during sleep deprivation. Our findings elucidate the need for a reevaluation of sleep quality as part of the pathophysiology behind polyuria in enuresis.

## Abstract# 951

**Single versus Multiple Doses of Early Postnatal Indomethacin on Prostanoid Production and Cox Gene Expression in the Neonatal Rat Kidneys** J. Sharma,[1] H. Tawadrous,[1] N. Chorny,[1] D. Kumar,[1] C. Cai,[1] G. Valencia,[1] J.V. Aranda,[1] A. Mongia,[1] M. Schoeneman,[1] K. Beharry.[2] *[1]Department of Pediatrics, Division of Nephrology and Neonatology, SUNY Downstate, Brooklyn, NY, United States; [2]Department of Pediatrics, Division of Neonatology, University of California, Irvine, CA, United States.*

**Objectives:** To exam the effects of a single versus multiple IN dose co-administered with FU on renal eicosanoid synthesis in the neonatal rat kidneys and determine the gene expression profiles of various cytochrome P450 enzymes (Cyps).

**Methods:** At birth (P0), neonatal rat pups (n=18 pups) were randomly assigned to receive IP injections of either indomethacin (IN, 0.2 mg/kg) on P0 and furosemide (FU, 5 mg/kg) on P0, P1 and P2; IN and FU on P0, P1, and P2 or equivalent volume saline (Sal) on P0, P1 and P2. At P3, whole kidneys were assessed for PGE2, PGF2α, 6-keto-PGF1α, TxB2, 12(S)-HETE,, and gene expression of various Cyp enzymes.

**Results:** Multiple-dose IN in combination with FU substantially decreased PGE2 (157.0±29.8, $p<0.01$), PGF2α (137.9±103.8, $p<0.01$), and TxB2 (823.3±109.1, $p<0.05$) compared to Sal (724.3±140.7, 1132.2±154.2, 1012.5±245.7, respectively) and single-dose IN (733.4±128.1, 1506.6±269.3, 1863.2±351.6, respectively). There were no appreciative changes in 12(S)-HETE. All Cyps were robustly downregulated with multiple-dose IN. Data are expressed as pg/mg protein (mean±SEM).

**Conclusions:** Furosemide may be effective for blunting the effects of a single dose of IN on renal vasoactive eicosanoids, but not those of multiple doses of IN. The concomitant use of FU in infants may inhibit Cyp-dependent metabolism and potentiate the nephrotoxic effects of IN.

## Abstract# 952

**CDK2 Dependent G1/S Checkpoint Mechanism Initiating an Redifferentiation Proliferated Process** Q. Shen,[1] Q. Shen,[1] H. Xu,[1] X.-L. Zha.[2] *[1]Department of Nephrology and Rheumatology, Children's Hospital of Fudan University, Shanghai, China; [2]Department of Biochemistry and Molecular Biology, Shanghai Medical College, Fudan University, Shanghai, China.*

**Objectives:** During repair of injured renal epithelium, a critical process is thought to be redifferentiation processe that indicate the transition from a mesenchymal- like proliferation cell to an epithelial-like proliferation cell. However, what control factors implicated in this process are still not clear.

**Methods:** We used highly synchronized HK-2 cells with proteomics to address a relative mechanism of redifferentiation, because this cellular model resemble an adaptive regeneration of injured renal epithelium after early hypoxic damage.

**Results:** Although 50% intracellular ATP was reduced in G0-phase cells and initiation of DNA replication was delayed during G1/S trasition, synchronized HK-2 still could complete whole of cell cycle and remain normal morphology. Early S-phase cells restored expression of CK18 downregulated during G1 later stage. Furthermore, CK18 was detected from anti-Cyclin A immunoprecipitate compounds(IPs) in early S-phase cells, but not in G1 later stage. However, a Ubiquitin-Ribosomal Protein S27a (UBRPS27A) was abundance highly in anti-Cyclin A IPs during later G1 and not in early S-phase cells.

**Conclusions:** UBRPS27A fusion protein may be an injuried response protein. Downregulated CK18 expressing in later G1-phase is possible of resulting from that UBRPS27A correlated with Cyclin A and inhibited the function of Cyclin A-CDK2. When UBRPS27A "released" Cyclin A in early S-phase, Cyclin A correlated with CK18 may result in restored expression of CK18 due to disclosed function of Cyclin A-CDK2 compounds.

## Abstract# 953

**Fluid Flow Shear Stress (FFSS) in Podocytes Is Mediated by Akt/GSK-3β/β-Catenin Signaling Pathway** T. Srivastava,[1] R. Sharma,[2] P.A. Cudmore,[1] M. Sharma,[2] E.T. McCarthy.[3] *[1]Children's Mercy Hospital, Kansas City, MO, United States; [2]Kansas City VA Medical Center, Kansas City, MO, United States; [3]University of Kansas Medical Center, Kansas City, KS, United States.*

**Objectives:** Prostaglandin $E_2$ ($PGE_2$)-mediated Wnt/β-catenin signaling has been shown to be important in osteocytes exposed to FFSS. Podocytes are subjected to FFSS due to hyperfiltration associated with various chronic kidney diseases. We have recently shown that podocytes respond to FFSS by increased synthesis of $PGE_2$ [JCCS: In press]. The aim of the present study was to determine if $PGE_2$ activates Wnt/β-catenin signaling in podocytes exposed to FFSS.

**Methods:** We exposed immortalized mouse podocytes to FFSS (2 dynes/cm$^2$ for 2hr) with and without pre-treatment with 2.5 μM indomethacin, and evaluated the (a) changes in phospho-Akt, phospho-GSK3β and phospho-p38MAPK by Western blotting (WB) and, (b) nuclear translocation of β-catenin by immunoflorescence (IF) at 2hr and 24hr post-application of FFSS.

**Results:** FFSS caused increased phospho-Akt/total Akt and phospho-GSK3β/total GSK3β compared to controls at 2 and 24 hrs following cessation of FFSS, and this change was abrogated by indomethacin. There was no change in phospho-p38MAPK/total p38MAPK following FFSS. Nuclear staining for β-catenin was increased at 2 and 24 hrs following FFSS compared to controls. The nuclear translocation of β-catenin was abrogated by indomethacin.

**Conclusions:** $PGE_2$ activates Akt/GSK-3β/ -catenin signaling *in vitro* in podocytes exposed to FFSS. Manipulation of the $PGE_2$-$EP_2$-Akt/ GSK-3β/β-catenin signaling pathway is an exciting new avenue to explore in the treatment of chronic kidney diseases associated with hyperfiltration.

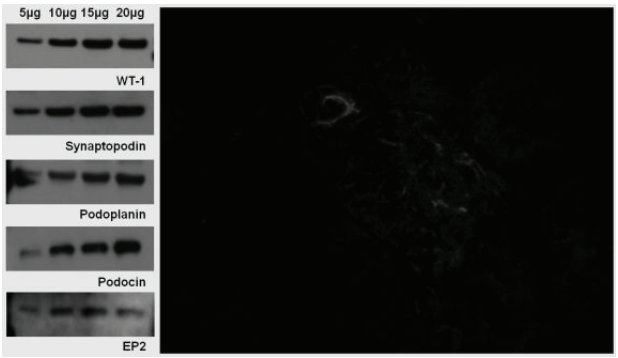
## Abstract# 954

**Prostanoid Receptor $EP_2$ Is Induced by Fluid Flow Shear Stress (FFSS) in Podocytes** T. Srivastava,[1] R. Sharma,[2] P.A. Cudmore,[1] A. Kats,[1] M. Sharma,[2] E.T. McCarthy.[3] *[1]Children's Mercy Hospital, Kansas City, MO, United States; [2]Kansas City VA Medical Center, Kansas City, MO, United States; [3]University of Kansas Medical Center, Kansas City, KS, United States.*

**Objectives:** Podocytes express prostanoid receptors $EP_1$ and $EP_4$. Hyperfiltration may cause podocyte injury through FFSS. We have shown that cultured podocytes secrete $PGE_2$ following exposure to FFSS [JCCS: In press]. The aim of the present study was to determine if $EP_2$ is present and induced by FFSS in mouse podocytes.

**Methods:** Using standard RT-PCR, qRT-PCR and Western blotting techniques we studied the expression of $EP_2$ in mouse podocytes under basal conditions and after exposure to FFSS (2 dynes/cm$^2$ for 2hr). $EP_2$ expression was analyzed in kidney by immunofluorescence (IF). Two separate primer sets were used for PCR studies. PCR products were sequenced to confirm $EP_2$. Specific $EP_2$ antibody (sc-20675) was used for WB and IF.

**Results:** Podocytes express podocyte specific proteins. $EP_2$ expression was confirmed by RT-PCR, qRT-PCR, WB and IF. IF showed presence of $EP_2$ in mouse kidney. FFSS caused an increase in $EP_2$ expression on both qRT-PCR and WB.




**Conclusions:** We show for the first time that $EP_2$ receptor is present in mouse podocytes and kidney, and is further induced by FFSS. We conclude that FFSS on podocyte secondary to glomerular hyperfiltration result in altered eicosanoid metabolism, and this pathway maybe amenable to drug intervention.

## Abstract# 955

**Modulation of Transient Receptor Potential Canoninal-6 (TRPC6) Ion Channel in Cultured Podocytes** T. Szabó,[1] Lí. Ambrus,[2] G. Czifra,[2] N. Kedei,[3] P.M. Blumberg,[3] T. Bíró.[2] *[1]Department of Pediatrics, University of Debrecen, MHSC, Debrecen, Hungary; [2]Department of Physiology, University of Debrecen, MHSC, Debrecen, Hungary; [3]MMTP, LCBC, NCI, National Institutes of Health, Bethesda, MD, United States.*

**Objectives:** The TRPC6 ion channel has recently been implicated in the pathogenesis of familiar proteinuric glomerular diseases. In the current study our aim was to study the function and modulation of TRPC6 in cultured podocytes.

**Methods:** TRPC6 activation on mouse immortalized podocytes was assessed by fluorimetric Ca-imaging whereas expression of TRPC6 was followed by immunolabeling and quantitative "real-time" QPCR techniques.

**Results:** We have shown that mouse podocytes express TRPC6 (both at protein and mRNA levels) and that the activation of TRPC6 by diacylglycerol derivatives results in an increase of intracellular Ca-concentration. We have also found that 30 min pre-treatment of podocytes with insulin markedly sensitized TRPC6. Interestingly, pre-application of bradykinin inhibited whereas inflammatory mediators (such as arachidonic acid, prostaglandin E2, and histamine) did not alter TRPC6 function. Of further importance, when applied for 24 hrs, insulin and arachidonic acid significantly up-regulated the expression of TRPC6 in podocytes whereas the other agents employed did not affect the level of the ion channel.

**Conclusions:** These data suggest that various signaling molecules may directly alter expression and function of TRPC6. Since these agents were implicated in the pathogenesis of acquired podocytopathy-associated proteinuric glomerular diseases, our findings argue for the putative central role of TRPC6 in podocytopathies of various origins.

## Abstract# 956

**Expression of Chemokine Receptors on Peripheral Blood T Cells in Children with Chronic Kidney Disease** M. Szczepanska,[1] L. Sedek,[2] I. Makulska,[3] K. Szprynger,[1] B. Mazur,[4] J. Bulsa,[2] D. Zwolinska,[3] J. Karpe,[5] T. Szczepanski.[2] *[1]Dialysis Division, Dept. of Pediatrics, Medical University of Silesia, Zabrze, Poland; [2]Dept. of Pediatric Hematology/Oncology, SUM, Zabrze, Poland; [3]Dept. of Pediatric Nephrology, Wroclaw, Poland; [4]Dept. of Microbiology/Immunology, SUM, Zabrze, Poland; [5]Dept. of Anestesiology/Intensive Care, SUM, Zabrze, Poland.*

**Objectives:** Children with chronic kidney disease (CKD) show features of altered cellular immunity. Only limited data are available on chemokine receptor expression on peripheral blood (PB) T-cell subsets in children with CKD on renal replacement therapy.

The study aimed at multiparameter analysis of the absolute numbers and percentages of T-cell subsets with CCR5, CCR7, CXCR3, and CXCR4 in CKD children on hemodialysis.

**Methods:** The expression of surface antigens was evaluated on PB mononuclear cells using multicolor flow cytometry. The study group consisted of 12 children and young adults with CKD on hemodialysis. Forty-one healthy individuals served as a control group.

Absolute and relative values of particular subsets were correlated with age, anthropometrical parameters, creatinine, BUN, hemoglobin, adequacy markers and mean arterial pressure (MAP).

**Results:** The CKD patients showed diminished absolute numbers of T cells with CXCR4 receptor as compared to the controls. The expresssion of CCR5, CCR7, CXCR3 was comparable to healthy children. During HD session the percentage of CXCR3+ lymphocytes was decreased with the lowest value at the end of session.

**Conclusions:** Diminished expression of some chemokine receptors on T-cells in patients with CKD in hemodialysis might results in impaired inflammatory response.

## Abstract# 957

**Intravenous Contrast Media and Fractional Excretion of Sodium in Children with Obstructive Uropathies** E.M. Tsygina, M.I. Bakanov, A. Tsygin. *Institute of Pediatrics NCZD RAMS, Moscow, Russian Federation.*

**Objectives:** Visualization of kidney and urinary tract with the use of contrast media (CM) is one of most useful tools in the workout of obstructive uropathies. Despite contrast induced nephropathy (CIN) is rare with non-ionic hypo- and iso-osmolar CM, it still may happen. So, studies of CIN pathogenesis are important in respect of changes in renal function, sodium and volume homeostasis.

**Methods:** We've studied sodium fractional excretion (FENa) before and 24 hours after intravenous pyelography (IVP) in 18 children aged 0,8-13 years with hydronephrosis and uretherohydronephrosis with normal glomerular filtration rate (GFR).

**Results:** A significant decrease of FENa was observed after IVP (0,66 ± 0,33% vs 0,88 ± 0,36%; $p<0,05$). No changes were seen in GFR, cystatin C, serum sodium and potassium. The blood pressure profile had a characteristic decrease

4 hours after IVP, slow increase thereafter, low physiologic level at night and achievement of an average level before 24 hours after IVP. Urine output was higher than previous days since patients were advised to keep sufficient hydration.
**Conclusions:** We suppose, that the decrease of FENa after IVP may result from renin-angiotensin-aldosterone system (RAAS) activation in response to hypotension and renal hypoperfusion following CM injection. That may also support a hypothesis of predominant role of prerenal mechanism possibly responsible for a precipitation of CIN.

## Abstract# 958

**Effect of Metabolic Acidosis on Neonatal Mouse Proximal Tubule Acidification** K. Twombley, J. Gattineni, M. Baum. *UT Southwestern, Dallas, TX, United States.*
**Objectives:** NHE8 is the predominate sodium hydrogen exchanger in neonates and there is an isoform change at weaning to NHE3. The serum bicarbonate in neonates is lower than adults largely due to a lower rate of proximal tubule acidification. It is unclear if the neonatal proximal tubule is functioning at maximal capacity or if it can respond to metabolic acidosis as in adults. The purpose of this study was to examine the effects of metabolic acidosis in neonatal proximal tubules.
**Methods:** Mice were gavaged orally with 1 mMol/100grams of body weight starting at 5 days of age with either $NH_4Cl$ or NaCl twice daily for 7 doses. The kidneys were harvested for brush border membrane (BBM) vesicle isolation, RNA for cDNA synthesis, or for proximal tubule isolation for *in vitro* microperfusion.
**Results:** Acid gavaged neonates decreased their serum bicarbonate from 19.5 $\pm$ 1.0 to 8.9 $\pm$ 0.6 mEq/l ($p<0.001$). Proximal convoluted tubule $Na^+/H^+$ exchanger activity (dpH/dt) was 1.68 $\pm$ 0.19 pH units/min in control tubules and 2.49 $\pm$ 0.60 pH units/min in acidemic neonatal mice ($p<0.05$) indicating an increase in $Na^+/H^+$ exchanger activity. There was an increase in both NHE3 (vehicle 0.35 ± 0.07 vs acid 0.73 ± 0.07; $p<0.01$) and NHE8 BBM protein abundance (vehicle 0.41 ± 0.05 vs acid 0.73 ± 0.06; $p<0.001$) in acidemic neonates compared to controls, but no significant difference in mRNA levels in either exchanger.
**Conclusions:** Neonatal mice can adapt to metabolic acidosis by increasing the BBM expression of both NHE3 and NHE8 as well as increasing proximal tubule acidification with an increase in proximal tubule $Na^+/H^+$ exchange activity. This study shows that in neonates, NHE8 can play an adaptive role in response to acidosis.

## Abstract# 959

**The Pendrin Gene, PDS, Is Transcriptionally Regulated by the Intestinal Natriuretic Peptide Uroguanylin** J. Rozenfeld,[1] O. Tal,[1] L. Adler,[1] E. Efrati,[1] A. Stewart,[2] S. Carrithers,[3] S. Alper,[2] I. Zelikovic.[1] *[1]Div Pediat Nephrol, Rambam Med Ctr, Technion, Haifa, Israel; [2]Ren Div and Mol Vasc Med Div, Beth Israel Deac Med Ctr, Boston, MA, United States; [3]Sequela, Pewee Valley, KY, United States.*
**Objectives:** Pendrin (SLC26A4), a Cl-/HCO3- exchanger, encoded by the gene PDS and expressed in the CCD, is involved in blood pressure regulation. Uroguanylin (UGN), a peptide produced in the intestine, functions in the kidney as "intestinal natriuretic hormone". We investigated whether UGN exerts its effect on electrolyte homeostasis partly by modulating pendrin activity.
**Methods:** qRT-PCR, immunofluorescence, transfection experiments.
**Results:** qRT-PCR analysis of HEK293 cells exposed to UGN showed a decrease in endogenous pendrin mRNA.
Mice received UGN intravenously. Kidneys were harvested for pendrin mRNA quantification and pendrin protein visualization. UGN- injected animals displayed a decrease in pendrin mRNA and protein expression.
Luciferase reporter plasmids containing different length fragments of human PDS (hPDS) promoter were transfected into HEK293 cells. Exposure of cells to UGN decreased hPDS promoter activity compared to activity in control cells. Furthermore, the findings provided evidence for the presence of a hypothetical UGN response element within the 96bp region between -1140bp and -1044bp on the hPDS promoter. This region overlaps with the previously demonstrated hypothetical pH response element.
**Conclusions:** UGN inhibits pendrin expression at the transcriptional level in vitro and in vivo. Characterization of the UGN response element on the PDS promoter and its possible role in extracellular fluid homeostasis and blood-pressure regulation awaits future studies.

## Abstract# 960

**IPNA Fellowship Program Feed-Back in an African Centre** M.I. McCulloch, P.J. Gajjar, P. Nourse, P.J. Sinclair. *Red Cross Children's Hospital, University of Cape Town, Cape Town, South Africa.*
**Objectives:** Introduction: IPNA assisted in establishing a Fellowship Program in 2003 to allow paediatricians from various parts of Africa to train within Africa in the field of paediatric nephrology.
**Methods:** Review feedback by questionnaire of IPNA Fellows who have trained at a single centre, Red Cross Children's Hospital (RXH), Cape Town, South Africa from September 2003 to March 2010.
**Results:** Ten fellows were accepted onto the program of which 2 are still completing their training. All 8 trainees who completed their fellowship have returned to work in their country of origin and 7/8 completed the questionnaire. Countries of origin of trainees included Nigeria (3 fellows), Kenya (3), Benin (1), Ghana (1) and Uganda (1).
Time spent in training ranged from 6 months to 2 years (mean 15months, median 18 months). On return to countries of origin; 4/7 are based in state practice only, 1/7 in private practice only and 2/7 both state and private. Their nephrology work-load is judged as 4/7(50-75% of total work), 2/7(25 – 50%) and 1/7(10-25%).

| | |
|---|---|
| Training useful | Yes 7/7(100%) |
| Specific areas useful | Clinical approach and hands-on skills 7/7 (100%)<br>Biopsies 5/7(71%)<br>Dialysis 7/7(100%) |
| Areas to improve in training | More Haemodialysis 2/7(29%)<br>More Histology 2/7(29%)<br>More on-calls 1/7(14%)<br>More structured program 1/7(14%)<br>No improvement needed 1/7(14%) |
| SA Paeds Nephrol Certificate(need 18mths training to qualify- not required by IPNA) | Successful 4/7(57%) |
| On-return to home Institution | Support by Institution 5/7(71%) |
| Challenges | Poor Staffing 7/7(100%)<br>Lack of facilities and equipment 6/7(86%)<br>Radiology – Ultrasound only 6/7(86%)<br>Histology support poor in 4/7(57%) |
| Fellows presenting papers at Congresses | 7/7 presented papers at RXH and Local African meeting (100%)<br>5/7 International Congress (71%) |

**Conclusions:** IPNA sponsored fellows feel that this is a successful program in an African setting, with appropriate clinical teaching and "hands-on" learning of biopsy and basic acute peritoneal dialysis skills. They also developed oral presentation skills at academic meetings.
All the fellows in this program returned to their countries of origin, often facing difficult circumstances requiring personal motivation and imagination to adapt skills to their environment. They felt that further support was needed on return, in terms of institutional support, staff and resources in particular.

# Author Index

**Numerals refer to abstract number.**

# Keyword Index

## Numerals refer to abstract number.