

Camptodactyly, Arthropathy, Coxa Vara, and Pericarditis Syndrome Among Egyptians

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ABSTRACT. Objective. To highlight the clinical, radiological, and pathological presentations of 10 Egyptian patients with camptodactyly, arthropathy, coxa vara, and pericarditis (CACP) syndrome.

Methods. Ten cases underwent a full history, complete clinical examination, laboratory and radiological investigations (and magnetic resonance imaging, MRI, of knee joints); arthroscopic histopathological synovial studies were performed in 6 cases.

Results. Camptodactyly and large joint arthropathies were present in all cases. The onset was at birth in 5 cases, and consanguinity was present in all cases. Laboratory investigations were normal in all cases (normal complete blood cell count, erythrocyte sedimentation rate, absent rheumatoid factor, and antinuclear antibody). Synovial fluid analyses were acellular in 3, hypocellular in 4, and moderately cellular in 2 cases. Histopathology revealed noninflammatory synovial hyperplasia in the 6 synovial biopsies obtained, with multinucleated giant cells in 4 of them. Plain hip radiology revealed short broad femoral neck and widening of joint space in all cases, with coxa vara in 9 cases. MRI of the knees showed rim-like enhancement of the lining of the fluid filled bursae in 7 cases, and homogenous enhancement pattern in 2 cases. No evident cartilage destruction existed in any case.

Conclusion. Our cases represent a familial syndrome of noninflammatory arthropathies associated with camptodactyly and coxa vara. The complete picture of the syndrome may be related to disease duration, and MRI is a useful tool in diagnosis. Physicians should be aware of the syndrome, to avoid misdiagnosis with other pediatric connective tissue diseases. (J Rheumatol 2003;30:1081-6)

Key Indexing Terms:

CACP

CAMPTODACTYLY

FAMILIAL ARTHROPATHY

COXA VARA

Jacobs¹ was probably the first to recognize the familial association of congenital camptodactyly with arthropathy as a distinct clinical entity. Didier² was the first to describe a case of camptodactyly, arthropathy, coxa vara, and pericarditis syndrome (CACP). Several authors have reported a description of additional cases³⁻¹². Outstanding features of this syndrome include camptodactyly that is either congenital or early onset, together with noninflammatory arthropathy associated with synovial hyperplasia. Progressive coxa vara deformity^{6-9,11} and/or noninflammatory pericardial effusion^{4,5,7,8,10} has been observed in some patients.

Based on genetic analyses CACP is considered a genetically homogenous condition, despite clinical variability and differences in ethnic and geographic origins¹². An autosomal recessive genetic basis has been shown. Recently, etiology of the syndrome was attributed to a gene that was mapped to chromosome 1q¹¹. Marcelino, *et al*¹³ identified mutations in a

gene (CACP) encoding a secreted proteoglycan as the cause of CACP. The CACP protein has been also known as megakaryocyte stimulating factor precursor, superficial zone protein, and lubricin^{14,15}. This protein appeared to be a major joint lubricant and an intimal cell growth regulator. Clinically, when CACP protein is absent, it results in scarring of tendons to tendon sheaths, and hyperplasia of other intimal cells such as synovial and pericardial cells^{13,16}. The tissue distribution of this protein was recently described utilizing monoclonal antibodies, which may help define its roles in normal articular joints and other pathological conditions such as osteoarthritis, rheumatoid arthritis, and CACP¹⁷.

Verma, *et al*¹⁰ proposed a classification of familial arthropathies associated with camptodactyly according to presence of coxa vara (CAC) or pericarditis (CAP). Nevertheless, Bahabri, *et al*¹¹ presumed that congenital camptodactyly and noninflammatory arthropathy when associated with coxa vara deformity or pericarditis constitute a single syndrome with combined features of CACP syndrome instead of CAP or CAC syndromes.

We highlight the clinical and radiological features among Egyptian cases with familial arthropathy associated with camptodactyly, coxa vara, and pericarditis.

MATERIALS AND METHODS

This prospective study was conducted on 10 patients from 6 consanguineous kindred. They included 9-year-old monozygotic twin brothers and dizygotic 6-year-old twin brothers. In addition, 2 siblings, a 17-year-old

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brother and his 8-year-old sister, and another 2 siblings, a 16-year-old brother and his 3-year-old sister are described. A second-degree cousin of the last 2 siblings (7 years old) was also studied. Finally, a 7-year-old girl is described.

All were referred to the rheumatology and rehabilitation outpatient clinic of Cairo University Hospitals. They all had bilateral and symmetrical swelling of joints of upper and lower limbs and flexion contractures of the fingers. They were referred with the previous assumption that they had juvenile rheumatoid arthritis.

All patients were clinically evaluated. The full clinical assessment specifically included ophthalmic and cardiac consultations. Laboratory tests, including erythrocyte sedimentation rate, complete blood cell count, immunoglobulins, rheumatoid factor, and antinuclear antibodies, were done to exclude other connective tissue diseases. Skeletal survey was carried out for all candidates. Echocardiography to detect pericardial disease was performed.

Magnetic resonance imaging (MRI) studies were performed in 9 patients. Synovial hypertrophy, effusion, and the state of articular cartilage were evaluated with gadolinium diethelene triamine pentaacetic acid (Gd-DTPA) enhanced MRI with a GE SIGNA horizon 1.0 T instrument using a knee extremity coil. T1 weighted spin-echo sequence was performed with echo time (TE) 14 ms, repetition time (TR) 440 ms before and immediately after injection of Gd-DTPA 0.1 mMol/kg. T2 weighted spin-echo sequence with TE 90 ms, TR 2000 ms was performed.

Further, synovial fluid analysis and arthroscopically obtained synovial biopsies from the knee joints were performed in 6 patients.

RESULTS

Camptodactyly refers to congenital or acquired nontraumatic flexion contracture of the proximal interphalangeal joint of one or several fingers¹⁸. All patients had camptodactyly and arthropathy. Coxa vara were evident in 9 patients. No patient had pericarditis. Camptodactyly involved several digits in both hands (Table 1). Camptodactyly developed within the first decade of life in 5 cases, while it was present since birth in the other 5 cases. Onset of camptodactyly was at birth in cases 1 and 2 (monozygotic twins), and at 3 years in cases 3 and 4 (dizygotic twins). In cases 5 and 6 (siblings), onset was at birth; while onset was at 6 years and 1.5 years in the other 2 siblings (cases 7 and 8, respectively). Onset was at 3 years in case 9 and at birth in case 10. As regards joint involvement, painless and nontender symmetrical joint swellings were evident in all cases. On palpation, joint swellings were soft; they were made up of boggy synovial thickenings with or without effusion, yet without overt signs or symptoms of inflammation. The involved joints showed limited range of motion. The wrist and hip joints were invariably involved in all cases, associated occasionally with knees, elbows, and/or ankles (Table 1). Patients showed no associated or systemic symptoms and laboratory investigations were normal in all cases.

Samples of synovial fluid from knees of 9 cases, obtained by needle aspiration, revealed a clear, light yellow-colored viscous fluid. The fluid was acellular in 3 cases, hypocellular in 4 cases, and moderately cellular in 2 cases. Lymphocytes were the predominant cells. Culture of aspirated fluid revealed no growth on aerobic and anaerobic media. No crystals were detected by polarized light microscopy.

Radiographic skeletal survey showed camptodactyly in all patients (Figure 1), coxa vara and short broad femoral neck in 9 cases (90%) (Figure 2). Flattening of the femoral head was only evident in 3 cases, while widening of hip joint space was apparent in all cases. In contrast, widening of knee joint space (Figure 3) was evident in 8 cases (80%), and flattening of talar head (Figure 4) was observed in only 4 cases (40%).

MRI demonstrated various grades of joint effusion in all cases (Figure 5). Hyperplastic synovium was seen in all cases in images taken immediately after intravenous injection of Gd-DTPA. The enhancement appeared thin, uniform, and rim-like (Figure 6) in 7 cases and homogenous in only 2 cases. Popliteal lymph nodes were seen in 2 cases. A subarticular cyst was confirmed in only one case. MRI exhibited no abnormalities affecting menisci, cruciate ligaments, epiphyses, or infrapatellar fat pad.

Arthroscopically obtained synovial biopsies revealed profuse villous processes, synovial hyperplasia, and fibrin on surfaces of villi in all specimens (6 cases). No evidence of necrotic villi or inflammatory cells was detected. Hemosiderin deposits were evident in one case. Multinucleated giant cells with homogeneously eosinophilic cytoplasm were present in 4 cases. Their sizes varied from 12 to 20 μm in diameter, with 3–12 centrally placed nuclei.

DISCUSSION

We describe 10 patients with CACP, a relatively large series. CACP is characterized by wide clinical variability; while camptodactyly and noninflammatory arthropathy are consistent findings, the occurrence of pericarditis and coxa vara differ in various reports (Table 1). The clinical variability has raised the issue of possible genetic heterogeneity¹⁰; however, genetic analysis¹² suggests that CACP is a genetically homogenous condition despite clinical variability and differences in ethnic and geographic origins.

To our knowledge, our study is the first to report the occurrence of disease in twins. Interestingly, the 2 pairs of twins (Figure 7) in the study are strikingly similar in the clinical, radiological, and pathological findings of each pair.

The onset of camptodactyly in our study was at birth in 5 cases, while the other 5 cases developed camptodactyly within the first decade. Camptodactyly was bilateral in all cases, yet with variable distribution (Figure 8). This finding agrees with Bahabri, *et al*¹¹, as they propose that camptodactyly in CACP syndrome is usually bilateral and congenital, although it can develop in early childhood. The degree of camptodactyly need not be equal in both hands. The interphalangeal joint of the thumb was affected in all our cases except one. The thumb was spared in some cases in various reports^{3,8,9,11,19}. The spared thumb in some cases contradicts the proposition of Hammoudeh and Siam²⁰ that thumb involvement is the first diagnostic sign of the disease. Arthropathy primarily affects large joints (e.g., wrists, elbows, hips, knees, and ankles).

Table 1. Clinical features of cases of present work and in previous reports.

Study	Pt	Age*/Sex	Camptodactyly	Arthropathy	CV	PC
Athreya ³	1	6 F	Both 3rd	Polyarticular	NR	-
	2	8 M	Both 3rd	Polyarticular	NR	-
	3	4 F	Right 3rd	W, E, H, K, A	NR	-
Malleison ¹⁹	1	35 F	Both 2nd, 3rd, 4th, 5th	W, E, S	NR	-
	2	13 F	All	W, E, H	NR	-
	3	12 M	Both 2nd, 3rd, 4th, 5th	K, W, A, E, S	NR	-
	4	8 M	Both 2nd, 3rd, 4th, 5th	K, W, A, E, S	NR	-
	5	5 M	—	W, A	NR	-
	6	4.5 M	All	W, A	NR	-
Martinez-Lavin ⁴	1	9 F	Both 5th	K, W, E	-	+
	2	12 M	Both 4th, 5th	K, W, E	-	-
	3	6 F	Both 1st	K, W, E	-	+
	4	5 M	Both 1st, 4th, 5th	K, W, E, A	-	+
	5	4 M	Both 1st	K, A	-	-
Ochi ⁵	1	9 F	Both 3rd	W, F, K	-	-
	2	7 F	Both 3rd	W, K	-	-
Martin ⁶	1	13 F	All	E, H, K, A	NR	-
	2	23 M	All	E, H, K	+	-
	3	13 F	All	W, E, H, K, A	+	-
	4	2 F	All	W, E, K, A	-	-
Bulutlar ⁷	1	12 F	Both 1st	W, E, H, K, A	+	+
	2	14 F	—	W, E, H, K, A	+	-
	3	13 F	Both 5th	W, E, H, K, A	+	+
	4	7 F	3rd, 4th	W, E, H, K, A	+	-
Hammoudeh ²⁰	1	14 M	Both 1st, 3rd, 4th, 5th	W, E, H, K	-	-
Verma ¹⁰	1	10 F	All	W, E, K	NR	+
	2	16 F	All	W, E, K, A	NR	+
Bahabri ¹¹	1 [§]	12 F	+ (Distribution NR)	W, E, H, K, A	+	-
	2 [§]	10 F	+ (Distribution NR)	W, E, H, K, A	+	-
	3 [§]	5 F	+ (Distribution NR)	W, E, H, K, A	+	+
	4	13 M	+ (Distribution -NR)	W, E, H, K, A	+	-
	5	3 F	2nd, 3rd, 4th, 5th	W, E, H, K, A	+	-
	6	2 M	+ (Distribution -NR)	K, H	+	-
	7	3 M	2nd, 3rd, 4th, 5th	W, E, H, K, A	+	-
Faivre ¹²	8 [†]	5 M	All	W, E, H, K, A	-	+
	1	6 F	+ (Distribution NR)	E, H, K, A	+	-
	2	3 M	+ (Distribution NR)	W, H, K, A	+	-
	3	4 M	+ (Distribution NR)	W, E, H, K, A	+	-
	4	2 M	+ (Distribution NR)	W, E, H, K, A	-	-
	5	13 F	+ (Distribution NR)	W, E, H, K	+	-
	6	9 M	+ (Distribution NR)	W, E, H, K	-	-
	7	15 M	Both 3rd, 5th	W, E, H, K	-	-
	8	9 M	+ (Distribution NR)	W, E, H, K, A	+	-
	9	6 M	+ (Distribution NR)	W, H, K, A	-	-
	10	3 M	+ (Distribution NR)	E, H, K, A	+	-
	11	6 M	+ (Distribution NR)	E, K	-	-
Current study	12	3 M	+ (Distribution NR)	W, E, K, A	-	-
	1	9 M	Both 1st, left 3rd, 4th	W, E, H, K, A	+	-
	2	9 M	Both 1st, right 3rd	W, E, H, K, A	+	-
	3	6 M	Both 1st, left 3rd, right 4th	W, E, H, K, A	+	-
	4	6 M	Both 1st, left 3rd, right 4th	W, E, H, K, A	+	-
	5	17 M	Left 1st, both 5th	W, E, H, K, A	+	-
	6	8 F	Both 2nd, left 3rd, 4th	W, H, K, A	+	-
	7	16 M	All fingers	W, E, H, K, A	+	-
	8	3 F	Both 1st, left 3rd	W, E, H	+	-
	9	7 F	Both 1st, right 5th	W, E, H, K, A	-	-
10	7 M	Left 1st, 3rd, 4th	W, H, K, A	+	-	

* Age at time of referral, years; CV: coxa vara; PC: pericarditis; NR: not reported; +: present; -: absent; W: wrist; E: elbow; S: shoulder; H: hip; K: knee, A: ankle. [§] Previously reported in Bahabri, *et al*, 1994⁹. [†] Previously reported in Laxer, *et al*, 1986⁸.



Figure 1. Hand radiograph showing camptodactyly.



Figure 4. Flattened head of talus.



Figure 2. Coxa vara and short broad femoral neck.



Figure 3. Wide knee joint space.



Figure 5. Sagittal T1 MRI of camptodactyly showing distension of the suprapatellar and popliteus bursae by homogenous low intensity joint fluid.

Involvement is bilateral and symmetrical. Clinically, the affected joint is swollen, with synovial thickening, but no overt signs or symptoms of inflammation. Synovial fluid was noninflammatory. However, it was moderately cellular in 2 out of the 9 synovial samples studied.

Histopathologically, profuse villous processes, synovial hyperplasia, and fibrin deposits on the surface were present in all 6 cases that underwent arthroscopic synovial biopsy of the knee. Multinucleated giant cells with centrally placed nuclei were present in 4 cases. Many authors^{3-6,8-10,20} have



Figure 6. Sagittal T1 MRI of camptodactyly with subsequent intravenous contrast injection resulting in a uniform, thin rim-like enhancement of the fluid filled bursae.

reported similar pathologic findings. Ochi, *et al*⁵ extended the synovial findings to include tendon abnormalities. The authors found tendon involvement that included the tenosynovium, and histopathologic abnormalities between chronic fibrosing tenosynovitis and the fibrosing camptodactyly. They suggested that camptodactyly represented the result of tenosynovitis that occurred *in utero*.

Coxa vara were evident radiologically in 50% of published cases¹¹. In our study, it was radiologically evident in 90% of cases, while clinical hip involvement was present in all cases. Bahabri, *et al*¹¹ suggested that coxa vara deformity becomes clinically apparent with increasing age and cited that the actual percentage of the previously described patients in which the hip ultimately becomes involved is uncertain.

Widening of joint space and short femoral neck reported in all our cases were previously described^{3,21}. Abnormal modeling of bones in the form of flattening of the head of femur and talus in the eldest 2 cases with the longest duration supports the observation that radiological changes appear fairly consistent but are age related²¹.

There is no comprehensive description of MR imaging in congenital camptodactyly and familial arthropathy affecting major joints. Hugosson, *et al*²¹ carried out MRI studies of the hips and knees in 4 patients with T2 weighted signal

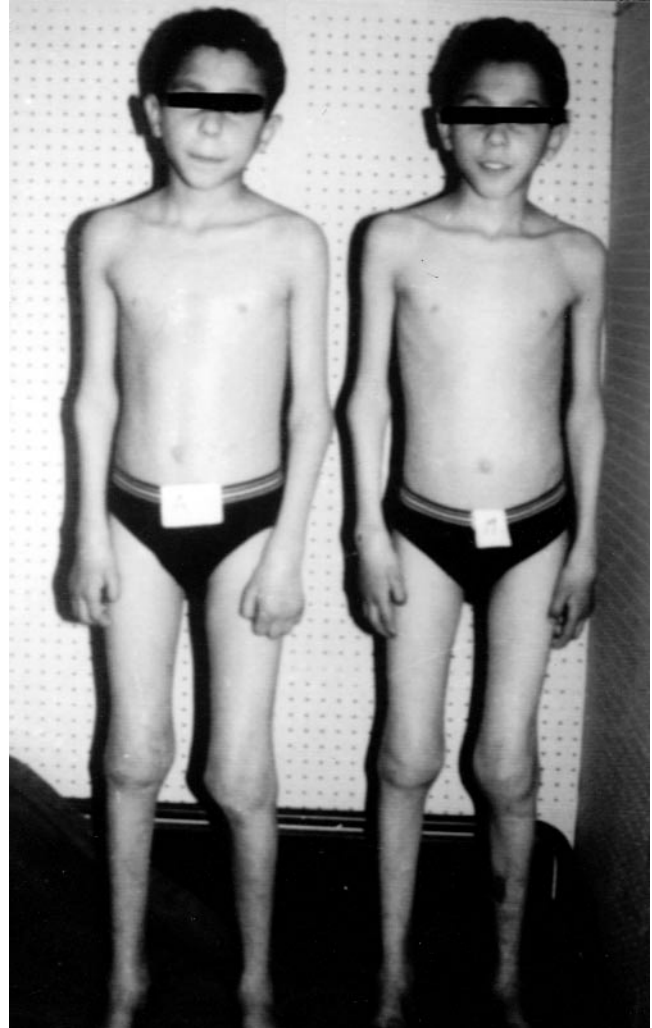


Figure 7. Monozygotic twin brothers with striking similarities in general appearance and affected joints.

intensity and observed only prominence of cartilage with normal menisci and cruciate ligaments. However, in our study, images were taken in T1 weighted signal intensity before and after injection of contrast material. Such a technique would allow precise distinction between effusion and synovial hypertrophy. The hyperplastic synovial enhancement appeared in 7 cases as a uniform rim enhancing the walls of fluid-filled bursae. Since enhancement is related to the presence of inflammatory tissue^{22,23}, the presence of rim-like enhancement supports the noninflammatory nature of the syndrome. Additionally, the rim-like enhancement could be easily distinguished from the homogenous or multinucleated enhancement seen in juvenile rheumatoid arthritis^{22,24}.

Two of the cases (dizygotic twins) had different pathologic features than other cases, despite the clinical and laboratory similarities. They had moderately cellular synovial fluid with less marked hyperplasia of the synovium and absent giant cells. Also, they had homogenous synovial enhancement on MRI, which is suggestive of inflammatory

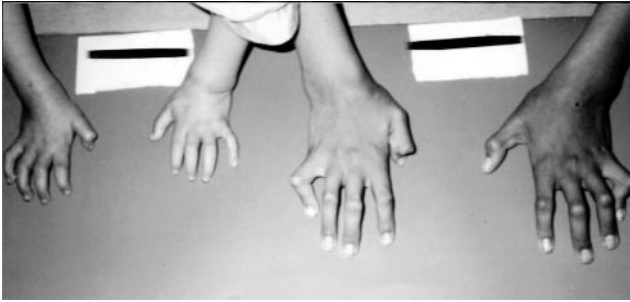


Figure 8. Distribution of camptodactyly in a boy (right) and in his younger sister.

changes²². These findings, together with the relatively short disease duration (3 years), would suggest a low grade inflammatory process. This picture is similar to the synovial inflammatory changes described by Malleon, *et al*¹⁹, termed familial arthritis and camptodactyly; however, their cases had autosomal dominant inheritance, and iridocyclitis was present in 2 of them. Ochi, *et al*⁵ cited that early in the disease, there is early and persistent low grade inflammation, followed by degeneration of tendons that are later replaced by fibrous tissue. Therefore, it might be assumed that these 2 cases would represent the early inflammatory stage of disease, while other cases (with disease duration of 7–17 yrs) represent the late postinflammatory stage.

No case with pericarditis was found in our study. The term CACP syndrome was used to verify that camptodactyly and noninflammatory arthropathy when associated with either coxa vara or pericarditis constitute a single syndrome. Noninflammatory pericarditis has been described in up to 30% of published cases^{4,7,8,10}. Indeed, pericarditis may be mild and self-limited¹¹, or life-threatening, necessitating pericardiocentesis or pericardiotomy^{4,7,8,10}. Therefore, all cases with this syndrome should be observed for emergence of other features of pericardial disease.

We describe 10 cases, which is a relatively large number with camptodactyly and noninflammatory arthropathy. Such a syndrome is clearly distinct from other childhood arthropathies. Cases referred as juvenile rheumatoid arthritis, in particular with abnormal presentation or with lack of overt clinical or laboratory evidence of inflammation, ought to be thoroughly investigated. MRI would be considered among the investigative tools that can differentiate the syndrome from other childhood connective tissue diseases.

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