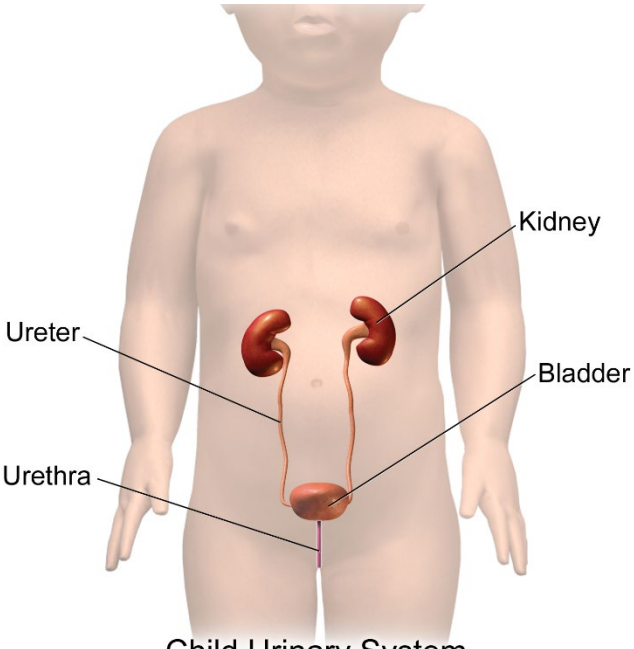
# Posterior Urethral Valves

## What are Posterior Urethral Valves?

The urethra is a tube that carries urine from the bladder to outside the body. Posterior urethral valves (PUV) is a condition where extra flaps of tissue grow inside the urethra and partially block the flow of urine, much like spinnakers on a sailboat block the flow of the wind. PUV is **a congenital anomaly** meaning babies are born with it. It may affect the baby even before it is born, while it is in the womb. (fetus).

BruceBlaus [CC BY-SA 4.0]. Via Wikipedia Commons
https://commons.wikimedia.org



Child Urinary System

## What causes PUV?

Posterior urethral valves are found almost exclusively in boys. The incidence of PUV is somewhere between 1 in 5,000 to 1 in 8,000 males babies. PUV is not a genetic condition. In other words, cases of PUV occur by chance (sporadic) and do not pass from one generation to the next. Cases where PUV affects several members of the same family are very rare.

## How do we diagnosis PUV?

The doctor will take a detailed medical history and a physical exam. They will also order a few medical tests such as:

- **A voiding cystourethrogram** – a special x-ray of the urethra and bladder

that can show the obstruction of the urethra. This test also shows if the patient has vesicoureteral reflux which occurs in about 30 to 50 out of 100 patients with PUV.

- **Kidney ultrasound** shows the dilation of the ureter and kidney and gives us an idea of the amount of kidney tissue.
- **Special blood tests** assess the function of the kidney.

### Antenatal (pre-natal) diagnosis

With the more common use of ultrasound during pregnancy more PUV patients are diagnosed in the uterus prior to birth. An antenatal ultrasound shows images of the baby and amniotic sac. It may also show problems that may be caused by PUV such as dilated kidneys and ureters and/or oligohydramnios which is the compression of the fetus in the uterus.

## What problems does PUV cause?

PUV is the most common cause of severe types of urinary tract obstruction (blockages) in children. It leads to several problems

- **Bladder hypertrophy** -the bladder wall muscle can thicken and enlarge from the extra work involved in voiding, like the muscles of a body-builder with exercise. Unlike the bodybuilder, the bladder may enlarge and stretch to the point the muscles become over stretched and too weak to work. This condition may require intermittent catheterization to empty the bladder.
- **Vesicoureteral reflux** - the valves may cause urine to move backwards from the bladder to the kidney. This leads to widening (dilation) of the ureters (the tube that connects the bladder to the kidney) and kidney. This greatly increases the risk of kidney infections and damage.
- **Renal dysplasia** - abnormal development of the kidney occurs in association with posterior urethral valves. Dysplastic kidneys function poorly or not at all.

For the fetus a severe case of posterior urethral valves may lead to life-threatening problems:

- **Body salt (sodium) imbalances and kidney failure** may occur if urine flow is obstructed or blocked.
- **Oligohydramnios** – compression of the fetus in the uterus may occur if the fetus's urine output is too small. This results in less fluid in the amniotic sac (fluid in the sac is fetal urine) which may lead to fetal compression. Oligohydramnios may cause bony abnormalities of the limbs and face, growth restriction, and underdeveloped lungs.

Patients may experience continuing difficulties even after the valves have been surgically opened (see later section). These persistent problems include partial renal failure, vesicoureteral reflux, and the inability of the bladder to void properly.

## How are patients with PUV treated?

To relieve a severe obstruction in an infant we place a small catheter into the bladder to take the urine out of the body. Infants may also receive extra fluid and sodium directly into the bloodstream with an IV. If lung function is poor we use mechanical ventilation until the baby is able to breathe on his own. If there is an infection, the baby receives antibiotics.

The surgery to remove posterior urethral valves is called **valve ablation**. This is an endoscopic surgery, meaning it does not require a surgical incision. The doctor inserts a cystoscope - a small tube with a light and a camera lens at the end - directly into the urethra. The doctor then inserts small knives or special hooks with exposed electrodes through the cystoscope and uses them to make incisions in the valves so they collapse and no longer obstruct the urethra.

If valve ablation does not improve kidney function, as sometimes occurs in severe cases, other surgical options are necessary in order to divert the urine

flow above the level of the urethra. There are several options for this:

- **Vesicostomy** is opening a small portion of the bladder directly to the skin
- **Ureterostomy** is the opening of a portion of the ureter directly to the skin.
- **Pyelostomy** is where the urine from the kidney is brought directly out of the skin overlying the kidney.

These procedures optimize the flow of urine from the kidneys and thus improve kidney function and minimize infections. The drawback is the need for further surgery in the future to bring the urine flow back down to the bladder.

Complications from PUV may lead to more surgeries.

1. **Bladder augmentation** - surgery to increase the bladder's capacity to hold urine safely. Some bladders lose flexibility and as more urine enters, it causes an increase in pressure which may cause urinary incontinence, infections, and continued damage to the kidneys. Bladder augmentation helps to prevent the development of these problems. It can be done in 2 ways:
    a) By using a piece of the bowel as a patch on top of the bladder.
    b) By cutting the wall of the bladder to allow its own inner lining to pouch out.
2. **Kidney transplantation** - may be needed if PUV leads to kidney failure Bladder augmentation may provide a safe urinary reservoir for a future transplanted kidney.

Author: Julian Wan, MD