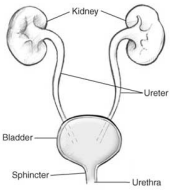


Posterior Urethral Valves

Monash Children's Hospital

What is it?

Posterior urethral valve (PUV) is a disorder in which an abnormal leaflet of tissue (valve) exists in the urethra, causing an obstruction (blockage) to urine flow. It is a developmental anomaly, present before birth and only affects boys.

PUV is rare, occurring in 1 in 4000-6000 boys.

Background

The urethra is the channel through which urine passes out from the bladder. In boys with PUV, urethral blockage near the bladder makes it hard for the bladder to expel urine, so the bladder has to push harder to try to empty. This increases pressure in the urinary tract. The pressure may push the urine back through the ureters to the kidney and cause the ureters, kidneys and bladder to dilate (expand).

The bladder wall may become thickened from trying to force the urine out against an obstruction. The ureters often do not work properly due to distension and back-pressure. PUV is also strongly associated with kidneys that have not developed normally (dysplastic) and do not work properly.

What causes it?

It is not known what causes the PUV.

There is as yet no identified genetic cause, although it is sometimes seen in families.

How is it diagnosed? How does it present?

The severity of obstruction will affect the severity of symptoms and the timing of presentation.

When there is a blockage or impairment of urine flow, the urine builds up behind the blockage. This can be identified as urinary tract dilatation on ultrasound scans performed during pregnancy.

If the blockage is not identified in pregnancy, it may present soon after birth with:

- no urine or poor urine stream
- poor feeding and failure to thrive
- urinary tract infections
- abdominal mass due to distended bladder or kidneys

In the older child, PUV may present with:

- recurrent urinary tract infections
- difficulty voiding
- poor urinary stream
- frequent voiding
- protracted bed wetting
- poor weight gain or growth

What tests are performed?

Ultrasound

This shows distension of bladder, ureters, kidneys, and upper part of urethra. The ultrasound can suggest the diagnosis. The ultrasound can also help detect the impact of obstruction on the kidneys.

Blood tests

These are performed to assess and monitor the kidney function.

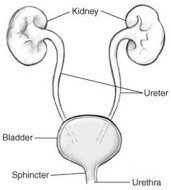
Micturating cystourethogram (MCU)

This is the definitive test for PUV. It is performed with a small tube ('catheter') placed through the urethra into the bladder. Contrast material is injected through the catheter to outline the bladder. An X-ray is taken which will show the shape of the bladder, whether there is any back flow up the ureters (reflux). Xrays are taken to identify the contrast outlining the urethra and any internal obstruction (valve).

Cystoscopy

This is both a confirmatory and therapeutic intervention. It is a surgical procedure, performed under anaesthesia. A small telescope is placed inside the urethra to identify the obstruction. Treatment of the valves can be performed at the same time.

Other tests may be needed to check the function of the kidneys, the drainage, and to monitor treatment.



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What are the treatment options?

Principles of treatment focus on relief of urinary tract obstruction, support of kidney function, prevention of infection and management of bladder dysfunction.

Urinary tract obstruction

A catheter is inserted initially, to drain the bladder and relieve obstruction, while stabilisation of kidney function is occurring. This is particularly critical in newborns with this condition.

Surgical relief of the obstruction is performed with valve ablation via cystoscopy. This should be delayed until the baby's medical condition has stabilised and surgery is as safe as possible. Catheter drainage may also be required in the immediate post-operative period.

Rarely, other surgical procedures are needed if the valve ablation and catheter do not achieve sufficient drainage. These include forms of urinary diversion; where part of the urinary tract is opened directly onto the skin surface ('stoma') to allow passive continuous drainage. These manoeuvres are usually short- to medium-term measures in critically ill babies or children. They include:

- 'vesicostomy': bladder opened to skin, usually low on the abdomen
- 'ureterostomy': ureter opened to skin, usually in the groin
- 'pyelostomy': renal pelvis (kidney-level) opened to skin, usually on the back

Kidney function

Boys born with PUV may have varying degrees of kidney damage. This needs to be assessed, managed and monitored. Specialist care under a paediatric nephrologist (kidney specialist) is critical to the ongoing care of these patients. Impaired kidney function can progress to end-stage renal failure with time and growth. About 1/3 of boys with PUV eventually require dialysis or renal transplantation at some stage in their life.

Urinary tract infection

As well as the kidney impairment present at birth, these boys are particularly prone to urinary tract infections (UTIs) due to poor drainage and emptying of their abnormal bladder and ureters. Any infection that involves the kidney can cause secondary damage, resulting in greater impact on already fragile renal function. Prevention, recognition and treatment of urinary tract infection are critically important to protect the existing kidney function.

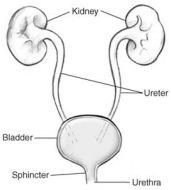
Prevention measures may involve use of low dose antibiotic prophylaxis and/or circumcision. In boys with abnormal urinary tracts who are prone to infection, circumcision has been shown to significantly reduce the incidence of UTIs.

Early recognition of signs of infection, and prompt treatment is also very important in reducing further kidney damage from infection.

Bladder dysfunction

Bladder function is also affected in patients with posterior urethral valves. In infancy the bladder tends to be small, and the pressure inside the bladder is often high. In older children the bladder tends to be overactive, trying to empty even when the volume in the bladder is very small, and in even older children the bladder tends to become distended, with minimal contractions. Bladder dysfunction causes incontinence, upper urinary tract dilatation and damages kidney function. The principles in treatment are to allow urine storage and emptying without elevation of pressure in the urinary tract. The techniques for management of the bladder include

- behaviour modification
- appropriate fluid intake
- regular emptying of the bladder (timed voiding)
- catheterization (see CIC information sheet)
- management of constipation
- nocturnal drainage of the bladder
- anticholinergic medication to reduce bladder overactivity
- bladder surgery, including bladder augmentation to increase the volume of the bladder



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What are the outcomes?

The outcome for children with PUV will depend on the degree of kidney and bladder abnormality. While removing the obstruction is an important part of the treatment, it does not reverse the damage that has occurred during development

About one third of patients with PUV will develop kidney failure at some stage in their life, requiring renal replacement therapy (dialysis or renal transplant).

About one third of patients will have a degree of renal impairment, managed with medication and diet.

About one third of patients will have normal kidney function, but may have ongoing issues with urinary tract drainage, infection and bladder function.

What is the follow-up?

A team of specialists, including a paediatric urologist and a paediatric nephrologist, looks after patients with posterior urethral valves. The paediatric urologist is a surgeon, responsible for relieving the urinary tract obstruction and managing bladder dysfunction. The paediatric nephrologist is a kidney specialist, responsible looking after the renal (kidney) function and the health consequences of renal impairment and failure.

The aim of follow-up is to identify any problems as early as possible, so that they can be investigated and/or treated as required.

What happens at follow-up?

At different times, you will need to see the urologist or the nephrologist more than the other. We will endeavour for you to have a joint appointment for those times you need to see both teams (nephron-urology clinic).

Regular blood and urine tests and ultrasounds will be performed prior to many of these appointments.

At follow-up appointments, the doctors will take a history and examine your child. They will ask about:

- urinary tract infections
- growth – height and weight
- continence (dryness) in older boys
- bed wetting
- urinary stream

The doctors will want to measure your child, check the blood pressure, and examine their abdomen.

Specialised tests may be performed depending on symptoms or concerns. These would include nuclear medicine studies, urine flow studies, and urodynamic studies.

How often?

The frequency of follow-up visits will depend on how old your child is, and the type of problems he is having.

After the initial diagnosis and treatment, your son will be followed closely to monitor his response to treatment.

Once the outcome of the initial treatment is apparent, he may be followed less frequently to monitor for the development of further problems.

If your son develops problems between clinic visits, call the hospital and ask to talk to someone from your specialist team. This is particularly advised for urinary tract infections, changes in continence, or urine flow. If you are worried about your son's health between appointments, it is better to seek help or advice than to wait.

Further information can be found at:
www.chop.edu/healthinfo/posterior-urethral-valves-puv.html

For parents' experiences, see: www.puvs.org