Paediatric urology

Posterior urethral valves

Also known as congenital obstructing posterior urethral membranes

Aim

We hope that this leaflet helps to provide information to parents and families if your baby has been diagnosed with posterior urethral valves (PUV) and aims to support the discussion you have had with your baby’s consultant and team.

What are posterior urethral valves?

- Membranes in the posterior urethra cause a narrowing of the passage way between the base of the bladder and the tip of the penis.
- Frequency one in 4000/8000 male infants.
- May be diagnosed before birth may not become obvious until later in childhood.
- Can have an effect on the development and function of the urinary tract. As a result of poor bladder drainage before birth/during pregnancy, the kidney system develops under high pressure. This can result in dilatation (swelling) of the kidneys and the tubes that drain them. If this is severe then it usually shows up on the 20 week ultrasound scan.

This diagram shows the difference between the flow of urine in the normal system and the obstruction caused by the posterior urethral valves.
What causes PUV?

- The presence of membranes in the urethra is due to remnants of tissue remaining from an early stage of fetal development, which would usually become incorporated into the internal structure of the urinary tract.
- This takes place at an early stage in a baby’s development sometime between seven and 11 weeks of the pregnancy.
- There are no recognised risk factors, although the condition is seen occasionally in twins or brothers, which suggests possible familial influence.

Signs and symptoms

If the blockage is significant it will show up earlier. Below you will find a list with some examples of the signs and symptoms and the time they may be noticed.

<table>
<thead>
<tr>
<th>Before birth/during pregnancy</th>
<th>At birth</th>
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<tbody>
<tr>
<td>• thick walled bladder</td>
<td>• usually there are no external signs of a problem however there may be:</td>
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<tr>
<td>• dilatation of both kidneys and tubes leading to bladder</td>
<td>o breathing difficulties</td>
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<tr>
<td>• reduced fluid around your baby</td>
<td>o swelling of tummy due to very full bladder</td>
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<tr>
<td></td>
<td>o urine infection</td>
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<td></td>
<td>o weak stream of urine</td>
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<table>
<thead>
<tr>
<th>Early childhood</th>
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<tr>
<td>• poor weight gain</td>
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<tr>
<td>• urine infection</td>
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<tr>
<td>• weak urine stream</td>
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<tr>
<td>• pain and frequency passing urine</td>
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<td>• bedwetting</td>
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Investigations/tests

Your baby’s care will be managed by one of our two consultant urologists. They are experienced surgeons who will organise a number of tests to confirm the diagnosis and undertake any surgery required.

Ultrasound scan (USS) of your baby’s kidneys, bladder and ureters (tubes which run from kidney to bladder one from the left kidney and one from the right) is usually undertaken a few days after birth.

Micturating Cystourethrogram (MCUG) uses a contrast fluid which is put into the bladder with a thin tube or catheter to show whether there is reflux and an x-ray is taken. Reflux means the urine is pushed up the ureters towards the kidneys causing poor drainage. This x-ray will also show an enlarged posterior urethra where the urine is pooling above the blockage.

Cystoscopy is an investigation which is carried out under a general anaesthetic, allowing a tiny camera in a narrow tube to look inside the bladder and see the membranous valves in the urethra.

Blood tests will show how well the kidneys are working and whether there has been any damage to them. The kidneys act as a filter to maintain the fluid and electrolyte balance (salts and water ratios) of the body and ensure that your baby remains healthy.
Any ill effect caused by the blockage will show in these tests. The level of creatinine in the bloodstream is particularly important in helping to show how efficiently the kidneys are working. This test is only used 24 hours or more after birth.

**Treatment/management**

This depends on the symptoms and how ill your baby is when diagnosed. Emergency treatment will be carried out before there has been a diagnosis if necessary for those babies with breathing problems, infection or electrolyte abnormalities. A very fine tube or catheter will drain the distended bladder and aim to prevent further damage to the kidneys.

Urinary tract infection and electrolyte imbalance will be treated with intravenous fluids and antibiotics. Once your baby’s condition is stable, a surgical procedure will be carried out to remove the extra membranes which have been causing the blockage and backflow.

The procedure is known as ‘ablation of the posterior urethral membranes’ and is carried out in the operating theatre with your baby fully anaesthetised. There is usually no need to cut the skin, as the operation is done with a cystoscope which is a tiny camera in a tube with an integral instrument to remove or resect the extra membranes.

This is a very delicate procedure done with great care as there is a small risk in the order of less than 1% of causing incontinence if the external sphincter is damaged. The external sphincter is a muscle which controls the outflow of urine from the bladder and is situated just behind the area where the membranes are found, so it could be damaged in the resection process. It is therefore preferable to resect too little than too much, which means that occasionally a second resection may be required at a later date.

In severe cases where a baby has an extremely low birth weight, a temporary urinary diversion may need to be considered in order to drain urine from the bladder above the level of the valves, or from higher up the urinary tract, for example, the ureter or the kidney. This would take the form of a surgically placed short tube to allow the urine to drain from higher up the normal drainage pathway reducing the pressure within the system.

**Privacy and dignity**

Same sex bays and bathrooms are offered in all wards except critical care and theatre recovery areas where the use of high-tech equipment and/or specialist one to one care is required.

**What happens after the operation?**

Your baby will be closely monitored by the urology team to check for any long term problems the severity of which will depend on how serious the blockage was before treatment.

The potential problems include:

- Continued reflux of urine from the bladder into the upper urinary tract resulting in infection and increasing damage to the kidneys.
- Delay in development of toileting skills.
- Reduced kidney function.
Regular monitoring by the consultant and team at our outpatient clinic will ensure early recognition of problems, and involvement of a wider team of professionals can ensure the best outcome for your child.

We hope this explanation will inform you and encourage you to ask questions which we will be very happy to answer. The team looking after your baby / child includes:

**Contacts/further information**

<table>
<thead>
<tr>
<th>Professionals/Title</th>
<th>Name</th>
<th>Telephone number</th>
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<tbody>
<tr>
<td>Consultant</td>
<td></td>
<td>Secretary:</td>
</tr>
<tr>
<td>Nurse specialist</td>
<td></td>
<td>Monday to Friday 08:00 - 18:00 01223 586973</td>
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We are now a smoke-free site: smoking will not be allowed anywhere on the hospital site.

For advice and support in quitting, contact your GP or the free NHS stop smoking helpline on 0800 169 0 169.

**Other formats:**

If you would like this information in another language, large print or audio, please ask the department where you are being treated, to contact the patient information team: patient.information@addenbrookes.nhs.uk.

Please note: We do not currently hold many leaflets in other languages; written translation requests are funded and agreed by the department who has authored the leaflet.