Some boys are born with posterior urethral valves (PUV). These are extra flaps of tissue that are in the tube that carries urine out of the body. Babies with PUV may not be able to wee normally – both while growing in the womb and after they are born.

PUV may be suspected during pregnancy, on the antenatal ultrasound scan. PUV is diagnosed after birth with some tests. In some cases, they are diagnosed when a boy is older.

PUV need to be removed by surgery. Sometimes, other treatment is needed. Boys who have had PUV may have problems in their kidneys and/or bladder later in childhood, and will need follow-up throughout their lives.

About the urinary system and urine

The **urinary system** gets rid of things that the body no longer needs, so that we can grow and stay healthy.

The **kidneys** are bean-shaped organs. They filter blood to remove extra water and waste in urine (wee). Most of us have two kidneys. They are on either side of our spine (backbone), near the bottom edge of our ribs at the back.

The two **ureters** are long tubes that carry urine from the kidneys to the bladder.

The **bladder** is a bag that stores urine until we are ready to urinate (have a wee). It sits low down in the pelvis.

The urethra is a tube that carries urine from the bladder to the **outside** of the body.

» More about the urinary system and kidneys
Symptoms and complications
PUV can cause serious problems because they stop – or partially stop – urine flowing out of the bladder and through the urethra. After the PUV are removed by surgery, some boys have no symptoms or complications. In other cases, they can cause other problems shortly after birth or later in life, which need further treatment.

- Occasionally, newborn babies are unable to breathe properly because the PUV leads to problems with their lungs. They need special care after birth to help them breathe.
- PUV may cause bladder problems, and some boys have problems passing urine. A few will need to use a catheter (a long, thin tube that is placed through the urethra) to help empty their bladder.
- PUV may cause the kidneys to swell – this is called antenatal hydronephrosis, and is a common problem for babies in the womb. Occasionally, this causes more serious problems.
- Boys with PUV may be more likely to get urinary tract infections (UTIs) when germs get into the urine and cause infection and illness. These are treated with medicines. In some cases, boys with PUV get a serious infection in their kidneys or in their blood.
- PUV may cause vesicoureteral reflux (VUR) – when urine refluxes (goes back up) the wrong way towards or into the kidneys.
- Occasionally, PUV can cause, or happen with, more serious problems with the kidneys.

Will it affect other family members?
Doctors do not think that PUV runs in families. If one of your children has PUV, it is unlikely that another of your children or another family member will get them.

Test and diagnosis
PUV may be suspected or diagnosed (identified) at different times in a boy’s life.

Before birth
The 20 week antenatal ultrasound scan looks at your baby growing in the womb. It also measures the amount of amniotic fluid (or liquor), the liquid that surrounds your baby, which is partly made up of his urine. PUV cannot be seen in the scan. They may be suspected in a boy if there are signs of a bladder outlet obstruction – that urine is not flowing out of the bladder.

You may need to go back to the hospital for more ultrasound scans during pregnancy. You may be referred to specialist healthcare professionals, such as a paediatric urologist (a surgeon who treats children with problems of the urinary system) or a paediatric nephrologist (a doctor who treats children with kidney problems).

After birth
If the antenatal scan(s) showed signs of a blockage affecting the flow of urine, your child will need tests to find out whether he has PUV or another condition. Some babies show signs, such as dribbling urine or having a larger bladder that feels like a lump in the tummy.

Later in life
In some boys, PUV are not diagnosed until later in life. Signs of PUV include problems passing urine, wetting themselves or urinary tract infections. Your child will need some tests to find out whether he has PUV.

Causes
PUV are extra flaps of tissue that are in the back (posterior) part of the urethra. They develop while a baby boy is growing in the womb.

PUV are the most common type of bladder outlet obstruction in newborn babies. This means that they stop (obstruct) the flow of urine out of the bladder.

The urethra in boys and men is longer than in girls and women. The male urethra has three parts:

- the posterior urethra (or prostatic urethra) is the back part and starts at the bladder (“posterior” means at the back)
- the membranous urethra is the middle part
- the spongy urethra is the last part and ends at the tip of the penis.

Causes: Read more about the male urethra

Posterior urethral valves | Page 2
Before birth

In most cases, there is no treatment before birth. In a small number of cases, an operation may be recommended during pregnancy. This aims to drain urine out of the baby’s bladder and into the amniotic fluid (or liquor) that surrounds the baby.

Where is my baby or child treated?

Some babies need to go into a neonatal unit, an area of the hospital for newborn babies who need support, for tests and treatment. Older children are treated in a paediatric unit, a special part of your hospital, for children. Some children need to be transferred to another hospital where specialist services are available.

After birth

The first treatment is to drain urine from the bladder. This helps reduce the risk of long-term problems with the bladder and kidneys. A catheter (a long, thin tube) is placed through the urethra (from the tip of your child’s penis), or sometimes through the wall of the tummy, into the bladder, and urine flows out.

All boys need an operation to remove the PUV – this is called a PUV resection. This will happen shortly after diagnosis.

» More about treatment

Symptoms and complications

PUV is one type of bladder outlet obstruction – these extra flaps of tissue block (obstruct) urine flowing out of the bladder and through the urethra. This can cause complications in a newborn baby.

Once the PUV are confirmed, they need to be removed by surgery. Some boys have no symptoms or complications after this. A few boys have some problems in their bladder and/or kidneys – though these may not happen until later in life.

Newborn babies

The antenatal ultrasound scan cannot diagnose PUV, but can show signs of any problems that may be caused by an abnormality such as PUV. Your doctor will let you know as much as possible about what to expect when your baby is born.

PUV rarely causes any problems during pregnancy or in childbirth.

Antenatal hydronephrosis

PUV may be a cause of antenatal hydronephrosis, a common problem in pregnancy. One or both kidneys hold on to urine, and become stretched and swollen. Antenatal hydronephrosis may get better at a later stage in the pregnancy, but your doctor will check how your baby is affected.

Breathing difficulties

Occasionally, PUV can stop the lungs from fully developing. These babies need extra support to help them breathe while their lungs get better.

This happens when PUV cause there to be less amniotic fluid (or liquor), the fluid that babies float in when growing in the womb, than usual.

In extreme and rare cases, the lungs do not develop enough for the support to help, and the baby dies before, or shortly after birth.

» Read more about how PUV can affect the lungs on the next page

Follow-up

All boys who have been treated for PUV will need follow-up throughout their lives, to check for any long-term problems with their kidneys or bladder.

Long-term effects

Some boys will have no long-term problems. Some will need further care, such as using special equipment to pass urine and make sure their bladder is emptied.

Some boys with PUV may progress to later stages of chronic kidney disease (CKD) – their kidneys gradually stop working as well as they should; this usually happens slowly over many years. Some children with CKD may progress to final stages of CKD (kidney failure), and need specialist treatment.

If your child has CKD, you and your child will learn more over time about how to help manage the condition and what to expect.

» Read more about the stages of CKD in CKD - an introduction

» More information about the future
Other problems

PUV may happen with other anomalies or abnormalities in the urinary system – one or both kidneys or ureters may not develop properly. This varies from mild to very severe.

In some cases, the kidneys do not work as well as they should – there is reduced kidney function. The first treatment is to drain urine from the bladder. In many babies, the kidney function will get better after this.

Sometimes, the kidney function does not get better. In others, the kidney function slowly gets worse as the child gets older. Your child will need follow-up appointments to check his kidney function.

Kidney failure

Occasionally, babies or children have kidney failure, when their kidneys can no longer support their body. It is not usually possible to know whether and when kidney failure will happen. It may be:

- at birth – this is very rare
- during infancy or early childhood
- during puberty or the start of the teenage years, when children go through a growth spurt and the smaller kidneys are no longer able to work for the bigger body.


Children

Problems passing urine

Boys with PUV often have problems when passing urine, such as wetting the bed more than usual, feeling pain when weeing, weeing less often, or having a poor stream of urine.

However, these symptoms are common, and most young boys with these issues do not have PUV.

Urinary tract infections

Boys with PUV may be more likely to get urinary tract infections (UTIs) – when bacteria (germs) get into the urine and cause an infection, usually in the bladder. Babies and children with UTIs may become irritable, have a fever, have pain on weeing, feel sick or be sick.

Occasionally the bacteria that cause UTI travel all the way up to the kidneys, where they cause a more serious infection and illness – this is called pyelonephritis. If not treated, this can cause damage to the kidneys.

UTIs are treated with medicines called antibiotics, which kill the germs. Some boys with PUV get UTIs that keep coming back, even after treatment – these are called recurrent UTIs.

If you think your child has a UTI, seek medical advice.

Reflux

PUV may cause vesicoureteric reflux (VUR). In VUR, some urine reflexes (goes back up) towards, and sometimes into, the kidneys. These children usually only need treatment if they get UTIs.

Infection in the blood

In some boys with PUV, a UTI can lead to a more serious infection in their blood – this is called septicaemia. Children with septicaemia are admitted to hospital for a few days, and treated with intravenous (IV) antibiotic medicines, which are given with a needle.

Other long-term complications are described in About the future.
Before birth

The 20 week antenatal ultrasound scan looks at your baby growing in the womb. It also measures the amount of amniotic fluid (or liquor), the liquid that surrounds your baby, which is partly made up of his urine.

PUV cannot be seen in the scan. They may be suspected in a boy if there is:

- a swollen bladder, ureters or kidneys – this may be a sign that urine is blocked from leaving the bladder
- a low amount of amniotic fluid (the liquid around the baby) – this may be another sign of blockage, or that the kidneys are not making as much urine as they should.

You may need to go back to the hospital for more ultrasound scans during pregnancy. You may be referred to specialist healthcare professionals, such as a paediatric urologist (a surgeon who treats children with problems of the urinary system) or a paediatric nephrologist (a doctor who treats children with kidney problems).

Antenatal hydronephrosis

Antenatal hydronephrosis is a problem found in antenatal scans. One or both kidneys hold on to urine. The affected kidney becomes stretched and swollen.

Occasionally, antenatal hydronephrosis is caused by a problem such as PUV.

After birth

In some boys, PUV are not diagnosed until later in life. Signs of PUV include problems passing urine, wetting themselves or urinary tract infections. Your child will need some tests to find out whether he has PUV.

Imaging tests

Imaging tests are used to help find out what is causing the problem. These use special scanners that take pictures of the inside of the body.

- Ultrasound scan – this is usually the first test, which looks at look at the shape and size of your child’s kidneys and other parts of his urinary system. A small handheld device is moved around your child’s skin and uses sound waves to create an image on a screen.

- MCUG (sometimes called a VCUG) – if your doctor finds a problem on the ultrasound, this test can check how your child is passing urine. A special X-ray machine takes a series of images of the bladder while your child passes urine.

- A DMSA scan or a MAG3 scan may also be arranged to find out well the kidneys are working. These are normally done after a baby is 3 or more months old. In each test, a chemical that gives out a small amount of radiation (energy) is injected into one of your child’s blood vessels – a special gel or cream can be used to stop your child feeling any pain. A special camera takes images of your child’s urinary system as the chemical passes through it.

Your doctor will talk to you about the tests, any risks and how to help your child prepare for them.

Blood tests

Blood tests can find out how well his kidneys are working (kidney function). A small amount of blood will be taken from a vein, with a needle and syringe. A special gel or cream can be used to help your child stop feeling any pain.

Urine tests

You, or a nurse, may need to collect some of your child’s urine in a small, clean container for a urine test. A dipstick will be dipped into the urine – this is a strip with chemical pads that change colour depending on what substances are in the urine. The sample may also be sent to a laboratory for more accurate tests.
Where will my child be treated?

Babies and children with PUV need to stay in hospital for monitoring and treatment.

- Newborn babies are admitted to a **specialist neonatal unit**, an area of the hospital for newborn babies.
- Older children are admitted to a **paediatric unit**, a special part of your hospital, for children.

They are discharged home when they are feeding or eating properly and are passing urine normally.

Hospital stay

A team of healthcare professionals will support your child and your family during his treatment. They include a **paediatric urologist**, a surgeon who treats babies, children and young people with problems of the urinary system.

» Read more on the next page

Treating before birth

In most cases, there is no treatment before birth. In a small number of cases, and only when the level of **amniotic fluid** (or **liquor**) around the baby has dropped, a surgical procedure called a **vesico-amniotic shunt** is recommended during pregnancy.

This operation is always done by trained healthcare professionals in a specialist centre. A tube (the shunt) is inserted through the mother’s abdomen (tummy area) and into the baby’s bladder. This aims to drain urine out of the baby’s bladder and into the amniotic fluid. This reduces the effects of pressure and helps the baby’s lungs to develop normally.

Your healthcare team will talk to you about the risks of the procedure and the risks of not doing the procedure, so you can make an informed decision.

» Read more on the next page

Treating after birth

**Supporting breathing in newborn babies**

If there are concerns about your baby’s lung development, your healthcare team may give oxygen or use **ventilatory** (breathing) support equipment to help him breathe.

**Draining urine from the bladder (before surgery)**

The first treatment for babies and children is to drain urine from the bladder. This helps reduce the risk of long-term problems with the bladder and kidneys.

**Urinary catheterisation** drains urine from the bladder. This is done using a catheter, a thin, flexible tube. There are two types of urinary catheters:

- **urethral catheter** – placed through the urethra
- **suprapubic catheter** – inserted through the skin, using a needle.

» Read more on page 8

**PUV resection**

All boys need surgery to remove the PUV – this is called a **PUV resection** ("resection" means to cut away). It is done by a **paediatric urologist** using **cystoscopy**.

A cystoscope, a tube with a camera, is placed through the **urethra**. This allows the urologist to look inside it and remove the valves.

Babies usually need to weigh more than 2.5 kg before they can have this operation – depending on the size of their urethra. If your baby is not big enough, your urologist may recommend a temporary procedure – see below.

**Risks and complications**

Serious complications are very rare. However, no procedure is completely safe, and it is important you understand the risks. Your doctor will speak with you about the possible problems before you consent (agree) to the procedure. The risks include the following:

- Some boys have problems passing urine – this will be checked after the cystoscopy.
- Occasionally, boys get a **urinary tract infection** after the procedure, and will need to take antibiotic medicine to treat the infection.
- Some boys have visible blood in their urine (**haematuria**) for a short time after the operation.

**What happens**

- A **general anaesthetic** is used to make your child go to sleep so he does not feel pain anywhere in his body. General anaesthetic may be given as a gas that your child will breathe in or it may be injected into a blood vessel. You will see an **anaesthetist** (a specialist doctor who is trained in giving these medicines) before the procedure, who will explain more.
- The cystoscope is covered with a special gel, and gently passed through your child’s urethra and into his bladder.
- Sterile (completely clean) water is pumped through the cystoscope, so that the urologist can see inside the bladder using the camera and then remove the valves.

**After treatment**

- Your baby or child will stay in hospital until he is feeding or eating well, passing urine normally and tests suggest that his kidneys are working well.
- A urinary catheter will be left in place for 24–48 hours.
- Your baby or child may need to take antibiotic medicines to prevent or treat any infection caused by the cystoscopy.
Aim of the operation

Some babies with PUV and other types of blockages cannot pass urine into the amniotic fluid (the liquid around the baby). There is not enough amniotic fluid, which is needed to help the lungs and other parts of the body develop.

The aim of this operation is to drain fluid from the bladder into the amniotic fluid.

What happens

- After giving consent (agreement) for the operation, the mother is admitted to a hospital, usually for less than a day. She is given antibiotics, medicines that help prevent infection, as well as an anaesthetic, a medicine that makes sure she and the baby do not feel pain.
- A hollow needle is inserted through her tummy and uterus, and is guided into the baby’s bladder. An ultrasound scan helps to find the right place.
- A shunt, which is a flexible, shaped tube, is moved down the needle. The shunt is placed between the baby’s bladder and the amniotic fluid. The needle is removed, leaving the shunt inside, to allow any fluid in the bladder to drain away.
- When the baby is born, the shunt is removed.

Risks and complications

- The shunt may move or become dislodged – the mother may need another operation to put in a new shunt.
- The operation may cause problems for babies after birth – such as bladder problems or other developmental problems.
- There may be an increased risk of miscarriage (losing the baby before birth) or preterm birth.
- There may be risks to the mother, such as damage to her organs or infection.

» For more information, see the NICE Treatment of lower urinary tract blockage in an unborn baby using a vesico-amniotic shunt: understanding NICE guidance
About the future

Your healthcare team will speak with you and your child about any long-term effects your child may have from PUV.

Some boys have normal bladder and kidney function after the PUV are removed. Others will have long-term problems and may need specialist care.

Follow-up

Boys who have been treated for PUV have follow-up appointments. It is important to go to these appointments even if your child seems well. You will also have the opportunity to ask any questions.

Checking bladder function

Bladder function tests check how well your child’s bladder is working. These are usually done by a paediatric urologist, a surgeon who treats babies, children and young people with problems of the urinary system, including the bladder and kidneys, and a specialist nurse.

- An MCUG (sometimes called a VCUG) – this helps confirm whether the PUV are completely removed, and also checks for vesicoureteral reflux (VUR), when some urine goes back up towards the kidney. A catheter is passed through your baby’s urethra and a dye is put through to reach the bladder – this does not hurt your baby. A special X-ray machine takes a series of images (pictures) of your child’s urinary system while he passes urine.
- Urinary flow rate – this measures how much urine your child is passing and the flow of urine. Your child needs to urinate (wee) into a special device fitted to a toilet.
- Ultrasound scans – these look at the bladder, often before and after your child passes urine.
- Urodynamics – this checks how well the bladder and urethra are working. One catheter (a long, thin tube) is passed through the urethra into the bladder, and fills the bladder with water. A second catheter is passed into the back passage (rectum). Both catheters measure pressure in the bladder as it fills and empties.

Checking kidney function

Kidney function tests check how well your child’s kidneys are working. They include:

- blood tests
- urine tests
- imaging tests, such as DMSA and MAG3 scans – to look at their kidneys.

Bladder problems

PUV can cause pressure to build up inside the bladder. If this is severe, the bladder swells, or gets larger. The wall of the bladder may get thicker.

Sometimes these problems continue even after the PUV resection. Some boys cannot empty their bladder – this can make toilet training difficult. Problems may include:

- going to the toilet more often than usual (frequency)
- wetting the bed more often than usual, or wetting themselves because they cannot get to the toilet in time
- feeling like they need to go to the toilet more than usual (urgency).
Medicines
Some boys may benefit from taking medicines. These include:

- oxybutynin, solifenacin and tolterodine, which relax the wall of the bladder. This means the bladder can hold more urine to help your child control it.
- desmopressin, which reduces the amount of urine your child’s kidneys make.

Using a catheter to empty the bladder
Some boys need to use special equipment to empty their bladder – this may be for a short or long time. If your child needs to use this treatment, you will be given training and support.

Clean intermittent catheterisation (CIC)
Some boys can use a catheter – a long, thin tube that is placed through the urethra and into the bladder.

- Several times a day, the catheter is placed through the opening of the urethra.
- Urine flows out of the bladder through the catheter and into a bag.
- When the urine stops flowing, the catheter is removed. The bag can be emptied into a toilet.
- Some children leave a catheter in at night while they sleep.

Mitrofanoff
Some boys need a surgical procedure that creates a channel from the bladder to the outside of their tummy. This is called a Mitrofanoff and is created out of another part of the body, such as the appendix and/or bowel.

The opening is usually in the tummy button so it is well hidden. A catheter is placed into the opening to drain urine out of the bladder.

Kidney problems
Some boys will have long-term problems with their kidneys. They may progress to later stages of chronic kidney disease (CKD) – their kidneys gradually stop working properly – this usually happens slowly over many years. These children will need specialist monitoring and treatment.

Some children with CKD progress to final stages of CKD (kidney failure), and need dialysis, which uses special equipment to clean the blood, and/or a kidney transplant, to receive a new kidney from a donor.

If your child has progressed to later stages of CKD, you and your child will learn more over time about how to help manage the condition, and what to expect.

Impact on your child and your family
Boys who have been successfully treated for PUV can usually do all of the things that other children their age do. They can go to, or return to, school or nursery. They can play with other children and stay active.

Most children will not need to change their diet, unless they have problems with their kidneys.

Some children need further support. Your doctor or nurse will give you more information, and let you know if you need to speak with someone at your child’s school or nursery.

Further support
This can be a difficult and stressful experience for your child and the whole family, including other children.

- If you have any concerns or need additional support, speak with your doctor or nurse.

Further information
This is the end of the information on PUV. If you would like to read more about other kidney conditions, tests and diagnosis, treatment or supporting information, you can find a list of topics covered on the infoKID website at www.infoKID.org.uk.