



Bladder exstrophy and epispadias

This leaflet explains about bladder exstrophy and epispadias and what to expect when your child comes to Great Ormond Street Hospital (GOSH) for treatment. GOSH is one of only two hospitals in the UK carrying out bladder exstrophy repair operations, as research has shown that the outlook is better when a child is cared for in a specialist treatment centre.

How does the urinary system work?

The urinary system consists of the kidneys, the bladder and ureters. The kidneys filter the blood to remove waste products and form urine. The urine flows from the kidneys down through the ureters to the bladder. From here it passes through another tube called the urethra to the outside when urinating (peeing).

What is bladder exstrophy?

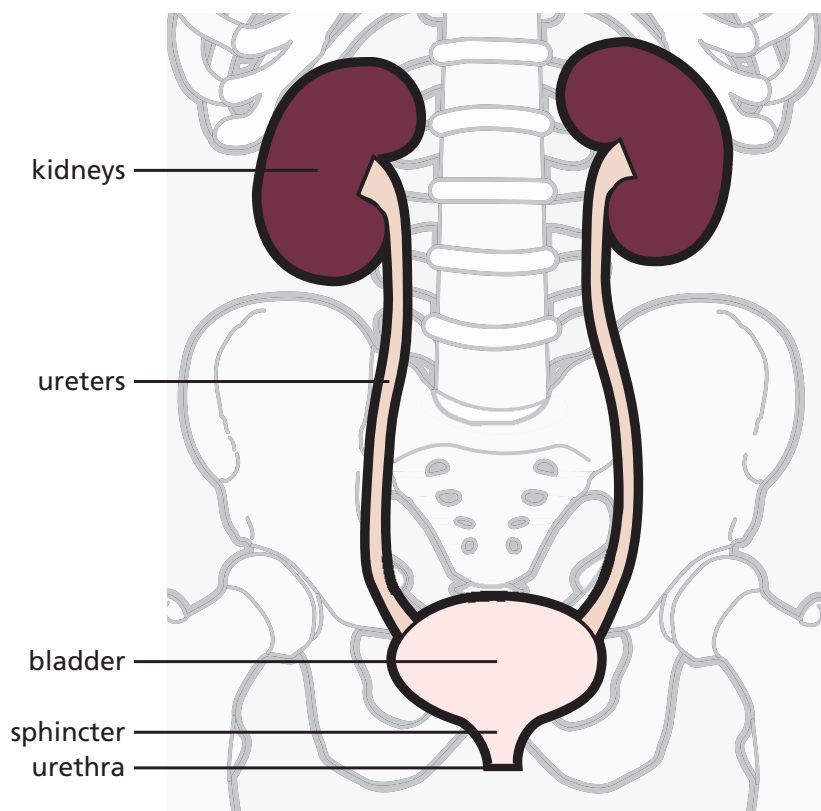
'Exstrophy' means 'turned inside out'. Bladder exstrophy is a congenital (present at birth) abnormality of the bladder. It happens when the skin over the lower abdominal wall (bottom part of the tummy) does not form properly, so the bladder is open and exposed on the outside of the abdomen.

What is epispadias?

In epispadias, the urethra does not form properly. All boys with bladder exstrophy also have epispadias, but it can also occur on its own.

In boys, the urethra may be very short and split, and as a result it emerges on the top surface of the penis rather than in its usual position at the end of the penis. The split may be small, or, when it occurs in boys born with bladder exstrophy, it may involve the full length of the penis, making the penis short and broad.

In girls, the opening of the urethra is higher and wider than usual, the labia (the lip-like folds encircling the vaginal opening) are further apart than normal and the clitoris (a small, very sensitive part of the female genitalia) is split in two.



What features are associated with bladder exstrophy?

As well as the bladder being exposed, babies with bladder exstrophy may also have related problems affecting their urinary system and pelvic bones. These related problems vary in severity and do not affect every baby. These will be confirmed using ultrasound scans and x-rays, and may be corrected during a series of operations. They include:

- Problems with the neck of the bladder and sphincter (ring of muscle that squeezes and relaxes to let urine flow from the bladder)
- The bladder has a smaller capacity than usual, so cannot hold much urine
- The ureters join the bladder in a different place to normal
- The middle part of the pelvic bones are separated

Bladder exstrophy can be associated with other problems, but the doctor will examine your child closely to see if this is the case. Some may need to be corrected with an operation, but others do not. For more information about the operations, please see our leaflets. The more common problems include:

- The anus is further forward than usual
- The belly button is lower down than usual
- Umbilical and inguinal hernia, where part of the abdominal lining and sometimes a section of intestine bulges out through a weak area in the abdominal wall muscles
- Undescended testes, where the testicles are not in their usual place in the scrotum

How are bladder exstrophy and epispadias diagnosed?

Bladder exstrophy is sometimes diagnosed before birth using ultrasound scans. However, it is often not picked up before birth, but will be obvious once your baby is born. Epispadias in boys is usually identified at birth, but in girls the diagnosis is usually made later when they develop bladder control problems or infections.

What causes bladder exstrophy and epispadias?

We do not know why bladder exstrophy occurs. It affects the developing baby during very early stages of development, at about four to six weeks into the pregnancy. This is when organs, muscles and other tissues start to form. It is not the result of anything either parent did or did not do, and is not simply an inherited condition.

How common is bladder exstrophy?

Bladder exstrophy occurs in 1 in every 40,000 births, affecting two to three times more boys than girls. If you have a baby with bladder exstrophy the chance of having another baby with bladder exstrophy is increased to about 1 in 100. People born with bladder exstrophy who go on to have children have a 1 in 70 chance of giving birth to a baby with bladder exstrophy. If you have any concerns, we can arrange for you to see a genetic counsellor.

How are bladder exstrophy and associated problems treated?

Babies born with bladder exstrophy and epispadias are admitted to GOSH in the first few days after birth by ambulance from where he or she was born. His or her abdomen will be covered in 'cling film', which protects the bladder surface, and also allows staff to observe the area closely too.

Bladder exstrophy and epispadias are corrected in a series of operations over the first few years of life. The overall aim of treatment is to prevent any kidney damage and correct the abnormalities so that your child's urinary system and genitals work properly and look as normal as possible.

Your child's urology surgeon (specialist in problems affecting the urinary system) will explain the treatment plan for your child; this can vary from child to child. Before each operation, your child's surgeon will visit you to explain about the operation in more detail, discuss any worries you might have and ask you to give your permission for the operation, by signing a consent form. An anaesthetist will also visit you to explain about the anaesthetic and pain relief after the operation.

What do the operations involve?

Bladder and abdominal wall repair operation – first few days after birth

This operation closes the bladder and abdominal wall, so that the bladder is inside the body and in the correct position. After the operation, urine will drain from the bladder through a number of catheters (plastic tubes) placed in the bladder.

Your child will come back to the ward to recover. For the first day or so, they will have an intravenous infusion (drip) giving fluids and medications until the bladder starts to recover. Your child will need to have regular pain relief after the operation. Initially, pain relief will be given through an epidural.

They may also have ureteric stents in place, which are thin tubes inserted through the abdomen into your child's bladder and up each ureter. These drain away urine while the bladder recovers from surgery. A urethral stent will also have been inserted into the urethra to keep it open while the area heals.

After the first week, some of the tubes will be removed. The drip will be removed when your child starts feeding again. The epidural is usually removed three to five days after the operation. Seven days after the operation, the nurses will remove the ureteric stents if they were inserted. This will be done on the ward – we will give your child pain relief beforehand although it may still be uncomfortable.

You and your child will be able to go home once your child is recovering and has been reviewed by the doctors.

Around three months later, your child will have a cystoscopy to check how the bladder is healing. Please see our *Cystoscopy* information sheet for further details.

Kelly procedure – at one to two years old

After the initial closure of the bladder exstrophy, there is no sphincter at the junction of the bladder and urethra. The Kelly procedure (also called a soft tissue reconstruction of the bladder neck) uses existing muscle and soft tissue to create a ring of muscle that acts like a sphincter. This holds urine in the bladder allowing it to stretch and gain more capacity and also helps form a strong stream of urine when weeing.

The surgeon can create a tummy button (omphalo-plasty) during the same operation if preferred.

In boys, the Kelly procedure may also involve a reconstruction of the urethra and penis or it may be done in a separate operation at a later stage.

During the same operation, the ureters may be re-positioned within the bladder if they are not joining the bladder in the correct place. This can cause a condition called vesico-ureteric reflux (VUR) where the valves can fail, allowing urine to flow backwards from the bladder to the kidney. Depending on the severity of the VUR, sometimes the urine can flow backwards as far as the kidneys. This can damage the kidney and eventually lead to kidney failure. The ureteric re-implantation operation involves disconnecting the ureters and re-attaching them in the correct place. For more information, please see our *Ureteric re-implantation* information sheet.

Are there any risks with these operations?

All surgery carries a small risk of bleeding during or after the operation. Every anaesthetic carries a risk of complications, but this is small. There is a small risk of infection, but your child may be given antibiotics as a precaution.

After the first operation to repair the bladder and abdominal wall, there is a risk that the wound will not heal properly and open up again. This can cause the bladder to move out of position. This happens more often if the area to be repaired is large, as the skin needs to stretch to cover it. If the wound opens up again, your child will need another operation to repair the bladder and abdominal wall. The surgeons may correct the pelvic bones during this operation as well.

There is also a risk of kidney damage in children with bladder exstrophy. The abnormal join between the ureters and bladder allows urine to flow backwards towards the kidneys. This is called vesico-ureteric reflux (VUR) it can sometimes lead to a condition called hydronephrosis, where the kidneys become swollen. Both these conditions will be monitored closely throughout your child's treatment. For more information, please see our *Vesico-ureteric reflux and Hydronephrosis* leaflets.

What is the outlook for babies born with bladder exstrophy?

The outlook for your baby is good, although around 20 per cent of all children born with bladder exstrophy need some further treatment later in childhood if they are having problems keeping dry. Following the Kelly procedure, if the ring of muscle around the base of the bladder is too weak, urine could dribble out all the time. This also happens if the bladder cannot hold enough urine.

This can be improved with an operation called bladder augmentation, which involves making the bladder larger, and therefore able to hold a larger volume of urine, using a section of intestine. These children then empty their bladder using a catheter to drain away the urine. The catheter can be inserted either into the urethra or a specially made channel called a Mitrofanoff.

For more information about this, please see our Bladder augmentation and Mitrofanoff leaflet.

The external genitalia of an individual born with bladder exstrophy or epispadias will always look different from others. In males the penis tends to be shorter and broader, but this does not usually cause any problems with their sex lives. Men born with bladder exstrophy have fathered children, although they may need fertility treatment. In many cases, sperm production is normal and sperm are healthy but ejaculating may be a problem. Women born with bladder exstrophy have also had children, although pregnancy should be supervised by a specialist obstetrician and their babies are usually delivered by caesarean section.

Long term follow up and transferring to adult services

Your child will need regular check ups at GOSH to make sure that everything is working properly. At various life stages, such as starting primary school, transferring to secondary school and finishing school, support from our psychologist will be offered. These will happen until the age of sixteen or so, when young people transfer to adult urology services. We generally transfer them to the Adolescent Unit at University College Hospital London (UCLH), where they will continue to be under the care of a urology surgeon. They will still need to have regular follow up appointments but often less frequently than before. The team at GOSH will work with you and your child to make the transfer from children's to adult services as smooth as possible.

Is there a support group?

Unfortunately, there is no support group for people affected by bladder exstrophy. The organisation Contact a Family may be able to put you in touch with another family. Call their helpline on 0808 808 3555 or visit their website at www.cafamily.org.uk

If you have any questions, please telephone the Urodynamics Unit on 020 7405 9200 Ext 5916 or 5917. Out of hours, please call Squirrel Ward on 020 7829 8814.