

Bladder Exstrophy

What Parents Should Know



What is Bladder Exstrophy?

Bladder exstrophy is a rare birth defect. On average, it occurs in about 1 out of every 50,000 live births. It is slightly more common in males than females. It is a condition where the bladder and parts around it form inside-out. The bladder muscles do not develop properly for children with bladder exstrophy. The skin, muscle and pelvic (hip) bones at the lower part of the belly or abdomen are not joined. As a result, the inside of the bladder pokes outside the belly. Instead of its normal round shape, the bladder is flat. There are also problems with the abdominal muscles and pelvic bones.

What Causes Bladder Exstrophy?

There is no clear cause for this condition. It is thought to happen during the 11th week of pregnancy, as the organs develop. Some experts believe that the bladder defect occurs when the tissues in the lower wall of the belly or abdomen develop. At the same time, the developing muscles and pelvic bones are affected too.

A temporary tissue called the cloacal membrane covers the lower belly wall and is replaced by maturing and developing abdominal muscles. If the cloacal membrane bursts before the abdominal muscles fully form, this may result in an “exstrophied” bladder. Another tissue called the urorectal septum helps to separate the developing bladder from the bowels or intestines.

Whether the child is born with epispadias, classic bladder exstrophy or cloacal exstrophy depends on when the cloacal membrane bursts and if the bladder and intestines are separated by the urorectal septum.

- An epispadias is a rare birth defect of the penis, where the urethra ends in an opening on the top side of the penis. For girls, the urethral opening may develop between the clitoris and labia or even in the belly area.
- Cloacal exstrophy (EC) is a severe birth defect where the abdominal organs (the bladder and intestines) are exposed. The penis or vagina is split, and the anus may be sealed.

How is Bladder Exstrophy Diagnosed?

Bladder exstrophy can often be found before birth during a routine sonogram. This condition will be clear at birth as the bladder will be seen outside of the baby's belly.

If this condition has been seen before birth, plans are often made for treatment right away after birth. If the condition is not found until the time of birth, the baby will be moved to the specialist unit. The doctor will note the bladder size and quality, the shape of the pelvis, and the state of the outer sex organs.

Treatment for Bladder Exstrophy

Bladder exstrophy is treated with surgery. The type of surgery used depends on how severe the defect is. It is of great value to work with a surgeon who has experience treating exstrophy.

The main goals of treatment are to:

- Close the bladder, the back of the urethra and the pelvis
- Rebuild a penis that looks normal and works in boys; Rebuild the outer sex organs in girls
- Fix the bladder so it can hold urine until it is time to urinate (“urinary continence”) without harming kidney function

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One form of treatment is “staged reconstruction.” This involves parts of the above surgeries done over the early childhood years:

- The first surgery is to close the bladder and pelvis. This is sometimes done just after the baby is born, but many times the surgery will be done after a few months to allow the baby and the bladder to grow.
- About 6 months after the bladder is closed, surgery is done to rebuild the epispadiac urethra and penis.
- When the bladder has grown large enough and the child is ready for potty-training, surgery is done on the bladder neck to achieve continence.

When the bladder’s quality and penis size (for boys) is good at birth, closing the bladder and penile reconstruction can be done in a “single operation” at an early age. Both early and staged reconstruction have good results.

If the bladder has grown enough and the surgeon is skilled, continence (control over urination) is possible. Often, further surgeries are needed over time to improve the child’s ability to urinate. More surgery may also be needed to rebuild and/or improve the outer sex organs.

In some harder cases, longer-term management is needed. Modern reconstructive surgery can still allow a baby to reach his/her late teens with successful results.

Frequently Asked Questions

What is the outlook for a baby born with this condition?

Children with exstrophy have a normal life expectancy. They can often take part in normal activities without restrictions.

Are there other health problems linked to bladder exstrophy?

Yes. These may include:

- Epispadias
- Vesicoureteral reflux
- Pubic bone separation or diastasis
- Small bladder capacity
- Missing bladder neck and sphincter
- Inguinal hernias

Patients often walk with a “waddle” gait, where the legs are pointed out. This gets better with age. Bowel function and control are normal.

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