

Bladder Exstrophy

What is bladder exstrophy?

Bladder exstrophy (*x-tro-fee*) is a bladder that is not formed right. The bladder and genitals are split in half, turned inside out and sit outside the body. There are many kinds of exstrophy.

How common is bladder exstrophy?

Bladder exstrophy happens in about 1 in 30,000 babies. It is more likely in boys than girls. Cloacal exstrophy (a form of bladder exstrophy) happens in about 1 in 50,000 to 100,000 births.

What causes bladder exstrophy?

We don't know what causes exstrophy. The problem occurs 4 to 8 weeks after a woman gets pregnant. This is when organs, muscles and tissues begin to form layers that separate, divide and fold. Exstrophy is not caused by something the mother did or did not do while she was pregnant. It does not run in families (it is not hereditary).

How is bladder exstrophy diagnosed?

Sometimes, exstrophy is diagnosed when a pregnant woman has an ultrasound. The ultrasound doctor (radiologist) might notice the bladder is missing. But unborn babies pee often, making the bladder hard to see, and it is easy to miss seeing exstrophy. That's why many babies are diagnosed after they are born. The defect is seen right after birth. Sometimes, the diagnosis of exstrophy is not made right away, because it is a rare defect that most health-care providers have never seen before. Sometimes, it will take a

specialist to confirm the diagnosis and to tell if the baby is a boy or a girl.

What other defects can children with bladder exstrophy have?

Babies with bladder exstrophy may have some or all of the defects listed below.

Genital:

- Epispadias – In boys, the tube that carries urine from the bladder to the outside of the body (urethra) may be short and split. It opens on the upper surface of the penis. The split may also involve the two halves of the testicles (scrotum).
- Epispadias – In girls, the tube that carries urine from the bladder to the outside of the body (urethra) is located between a split clitoris and labia minora.
- Chordee – In boys, the penis may curve up.

Bladder:

- Missing bladder neck and sphincter. The bladder neck is the lower part, or door, of the bladder. The sphincter is a ring of muscles around the bladder neck. These parts control urine flow when they open and close.
- The bladder may hold less urine than normal.

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Vesicoureteral reflux (VUR):

- Urine travels from the kidneys down tubes called ureters into the bladder. Normally, this flow is one way. Reflux is a condition in which urine can flow back up from the bladder to the kidneys.

Pelvis:

- The front part of the pubic bone doesn't join. This is called diastasis.

Spine:

- Some children with cloacal exstrophy may have a fatty growth (lipoma) on their spinal cord. This may cause problems with their legs, bladder and rectum.

Kidneys:

- Some children may have double kidneys or kidneys in a different location.

Bowel:

- Children with cloacal exstrophy may have poorly developed, large bowels and no rectum. This will usually require a surgery to make an opening from the intestines to outside the stomach for passage of stool (colostomy). Regular exstrophy does not usually involve bowels.

What kind of treatment will my baby receive?

All babies with exstrophy are put on "latex precautions." This means we avoid using products made with latex when your baby is in the hospital. You should also avoid using latex at home when possible, especially when it touches your baby's mouth, nose, urethra, etc. (mucous membrane). Avoiding latex may prevent your child from developing a latex allergy later in life.

Within the first week of life, your baby may have one or more surgeries to:

- Close the separated pubic bones and place the bladder into the pelvic cavity
- Create a bladder neck
- Repair the epispadias
- Close the abdomen
- Create an ostomy (for children with cloacal exstrophy)

After surgery

Your baby will probably be in the hospital for 1 to 3 weeks and will have:

A spica cast:

- This special cast will be placed by the orthopedic surgeons. The cast extends from the ankles to the mid-chest, with a hole for the diaper area. This cast will stay on for 4 to 6 weeks or until your baby outgrows it. Your nurses will teach you how to take care of the cast and give you special tips for diapering. Occupational therapists will help you pick the right type of car seat to use while your baby is in the cast. Special car seats for spica casts are available on loan.

Catheters:

- Catheters are tubes to drain urine from your baby. Your baby will have a catheter called a supra pubic (S-P) tube in the stomach that will drain urine from the bladder into a bag. This keeps the bladder empty so that it can heal. Your baby will have the S-P tube in for 3 to 4 weeks at home. Your baby may also have 2 ureteral catheters in place that exit through the urethra. These will usually be taken out before your baby leaves the hospital.

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Intravenous line (IV):

- Your baby will have a tube inserted into a vein to give fluids and antibiotics for several days.

Cardiac respiratory monitor:

- Your baby's heart rate, breathing and oxygen levels may be measured for several days.

Antibiotics:

- Your child will be on a daily, low dose of antibiotics to help prevent urinary tract infections.

Going home

Your nurse will teach you all you need to know to care for your baby at home. Also, the Urology Clinic can answer your questions at any time. You will have weekly appointments in the Urology Clinic for the first month after discharge and then visits every 2 to 3 months. Your baby will have X-rays or ultrasounds during many of the visits. You will also need to see your child's regular doctor (pediatrician) on a regular basis for well-child checkups.

Later years

All children with reflux take antibiotics. Children with exstrophy who have reflux often need surgery. Some children will need surgery to tighten the bladder neck to prevent leaking of urine (incontinence). Some children will need their bladders enlarged with a surgery called an augmentation. Boys may need additional surgery on the penis. Surgeries can often be combined to reduce the number of times your child is operated on. Your child may get more urinary tract infections than other children. Your child's primary care doctor can help monitor and treat these. Your school-age child should see the urologist at least once a year to check kidney growth. If your child is having urinary problems, they will need to see the urologist more often.

Children and adults with exstrophy can lead normal, healthy, active lives. Our goal is to help your child have bladder control and good bladder health. As an adult, your child should have normal sexual sensation, function and potential for fertility.

Each person with exstrophy is different, and their care and treatment will vary based on their needs. You and your child's urologist will develop a plan of care just for your child.

Where can I get more information?

The Urology Clinic nurses can provide you with more information on:

- Bladder exstrophy resources
- Other families of children with exstrophy via the NW Exstrophy Support Group
- A national exstrophy organization called Association for the Bladder Exstrophy Community (ABC)
- Latex
- Vesicoureteral reflux
- Spica casts
- Urinary tract infections

Children's Hospital and Regional Medical Center is a Center of Excellence for Exstrophy. This means we are internationally known for our expertise, research and specialists in caring for children with exstrophy. Please contact us for more information.

FOR MORE INFORMATION

- Urology Clinic (206) 987-2509
- Exstrophy Nurse (206) 987-2509 ext. 4
- Your Child's Health Care Provider

Children's will make this information available in alternate formats upon request. Please call Marketing Communications at (206) 987-5205.

This handout has been reviewed by clinical staff at Children's Hospital. However, your child's needs are unique. Before you act or rely upon this information, please talk with your child's health-care provider.