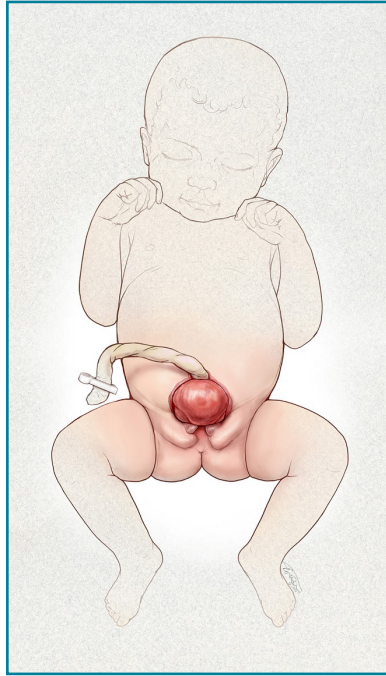


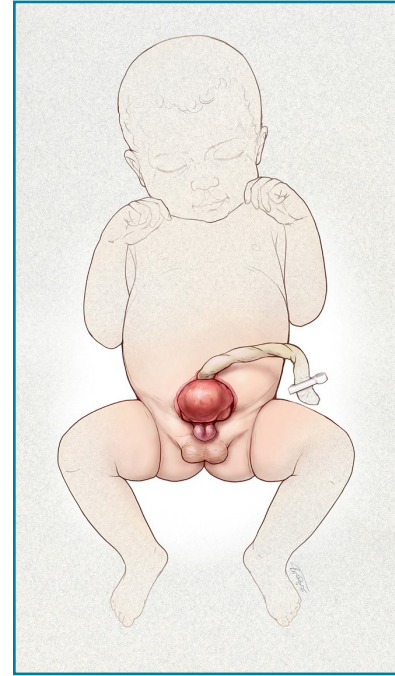
Bladder Exstrophy

What is bladder exstrophy?

Bladder exstrophy (X-stro-fee) is a bladder that forms outside the body. The bladder and genitals are split in half and turned inside out. There are many kinds of exstrophy.



Baby born with vagina



Baby born with penis

How common is bladder exstrophy?

Bladder exstrophy is rare. It happens in about 1 in 10,000 to 1 in 50,000 babies. It is more likely in babies born with penises. 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 pregnancy begins. This is when organs, muscles and tissues begin to form layers that separate, divide and fold. Exstrophy is not caused by something the pregnant person did or did not do while they were pregnant. It does not run in families (it is not hereditary).

To Learn More

- Reconstructive Pelvic Medicine
206-987-1240
- Ask your child's healthcare provider
- seattlechildrens.org/patient-education

Free Interpreter Services

- In the hospital, ask your nurse.
- From outside the hospital, call the toll-free Family Interpreting Line, 1-866-583-1527. Tell the interpreter the name or extension you need.



What other defects can babies with exstrophy have?

They may have some or all of these defects:

Genital

- **Epispadias** - In babies with penises, 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 sac of skin that holds the testicles (the scrotum).
- **Epispadias** - In babies with vaginas, 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 babies with penises, the penis may curve up.

Bladder

- Your baby may have an incomplete 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 pee (urine) than normal.

Vesicoureteral reflux (VUR)

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

Kidneys

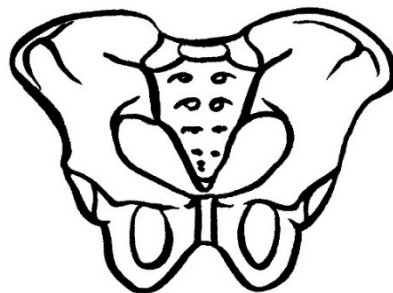
- Some children may have double kidneys, meaning they drain pee (urine) in 2 places or kidneys in a different location.

Bowel

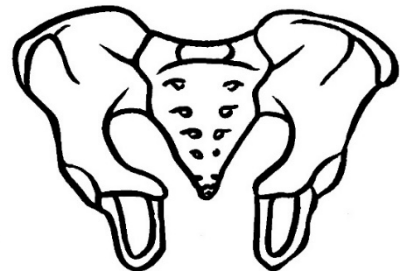
- Children with cloacal exstrophy may have poorly developed large bowels and no rectum (the hole where poop comes out). They often need surgery to make an opening from the intestines to outside the stomach so poop (stool) can come out (colostomy). Regular exstrophy does not usually involve the bowels.

Pelvis

- The front parts of the pubic bone are wide apart. This is called diastasis.



Normal pelvis



Pelvis with diastasis

Spine

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

How will we take care of our baby until they have surgery?

Your baby's open bladder does not cause them any pain. But it is important to protect it. Until your baby has their surgery to close the bladder, you will put a dressing over their bladder to protect it. You will wash and care for your baby as normal and avoid rubbing their bladder.

Babies with exstrophy are at risk for developing an allergy to latex later in life. Because of this, you should avoid using latex at home when you can, especially when it touches your baby's mouth, nose, urethra, etc. (mucous membranes). We will also avoid using latex products on your baby when they are in the hospital.

What surgeries will my baby need?

Between the first 4 to 6 months of life, your baby may have 1 or more surgeries to:

- Close their bladder and place it in the pelvis. This may include reconnecting the ureters.
- Bring the front parts of the pubic bones back together (called "reapproximate") to fix the diastasis.
- Create a tube that goes into the bladder (a bladder neck).
- Fix the epispadias.
- Close the abdomen.
- Create a surgical opening in the belly so poop can come out (ostomy) for children with cloacal exstrophy.

Your child's urologic surgeon and team will help you know what to expect before, during and after each surgery.

What can we expect before surgery?

Your baby will need regular visits at Seattle Children's to prepare for surgery.

2 months before surgery

- You will have a clinic visit with your child's urologic surgeon.
- Your baby will need a blood test for anemia by their primary care provider.

2 weeks before surgery

You will need to:

- Have a clinic visit with your child's urologic surgeon.
- Have a clinic visit with your child's orthopedic surgeon. They will help you order a special car seat for your baby to use after their surgery.
- Visit the Pre-Anesthesia and Surgical Service clinic (PASS Clinic), where the anesthesia team will check that your baby is ready for surgery.
- Your baby may need some lab tests after this clinic visit.

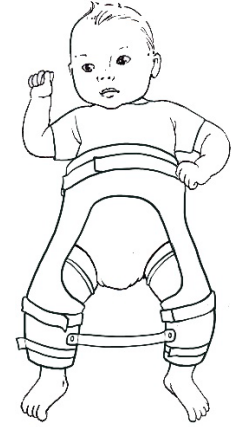
What can we expect after surgery?

Right after surgery, your child will have:

A spica brace

Orthotic clinicians will measure and fit your child for this custom spica brace (or just spica) before surgery and place the spica on your child immediately after the bladder surgery.

This spica will stay on for about 6 weeks. The spica will impact how your baby moves and how you hold and care for them. Your child's nurses will teach you how to take care of the spica and give you special tips for diapering. We will give you a special type of car seat (at no cost to you) to use while they have the spica brace.



Catheters

Catheters are tubes that drain pee (urine) from your baby. Your baby will have a tube called a suprapubic catheter in for about 6 weeks at home. We call this the "SP" tube. Your baby may also have 2 ureteral catheters or stents in place that come out through the urethra or belly. These will fall out on their own about 10-14 days after surgery.

Intravenous line (IV)

Your baby will have a tube inserted into their vein to give them fluids and antibiotics for several days after surgery.

Cardiac respiratory monitor

Your baby may have a cardiac respiratory monitor to check their heart rate, breathing and oxygen levels for several days.

When will we be able to go home from the hospital?

Your child will have weekly appointments in the Urology clinic for the first 6 weeks. While you are home, we can answer your questions at any time.

6 weeks after surgery:

Your child will return to the hospital for:

- Clinic visit with their orthopedic surgeon to remove their spica
- Radiology studies including:
 - X-ray to look at how well their bones are healing.
 - Cystogram or dye study to look at how well their bladder is healing and whether your child has vesicoureteral reflux.
 - Ultrasound to look at their kidneys.
- Clinic visit with their Urologic surgeon to remove the SP tube.

You will also need to see your child's primary care provider for their ongoing well-child checkups.

Will my child need future surgeries?

Some children who had surgery for exstrophy gain complete control over urine flow, and they have no other issues later that need treatment.

Some children will need other procedures. These may include:

- Children with urine that flows back up from the bladder to the kidneys (vesicoureteral reflux) sometimes need antibiotics. This helps prevent urinary tract infections (UTIs). As they get older, they may need a procedure to reduce reflux.
- If leaking of urine (incontinence) is a problem, your child may need surgery to help them stay dry.
- Some children need surgery to enlarge their bladder (bladder augmentation) or to place a tube (Mitrofanoff channel) so they can empty their bladder by putting a catheter through an opening in their belly.
- Babies with a penis may need surgery if the opening where pee comes out is on the underside, not at the tip (hypospadias).

We can often do these surgeries at the same time.

Will the exstrophy impact my child as they grow up?

- Your child may get more UTIs than other children. Your child's primary care provider can help monitor and treat these.
- When your child reaches school-age, they should see their pediatric urologist at least once a year. We will give your child an ultrasound to make sure their kidneys are growing well and to look for any signs of kidney problems.
- We partner with adult urologists at University of Washington and Harborview Medical Center to ensure your child gets proper care when they become an adult. Life-long follow-up care is important for good urinary health.
- Children and adults with exstrophy can lead normal, healthy, active lives. Our goal is to help your child have healthy kidneys and good bladder health. As an adult, your child should have normal sexual sensation, function, and ability to have babies.
- 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?

We can provide you with more information on:

- Bladder exstrophy resources
- A national exstrophy organization called Association for the Bladder Exstrophy Community (ABC)
facebook.com/AssociationfortheBladderExstrophyCommunity/

See our handouts:

- “Latex Allergy: Who is at risk and how to lower risk of infection”
seattlechildrens.org/pdf/PE001.pdf
 - “Urinary Reflux”
seattlechildrens.org/pdf/PE181.pdf
 - “Urinary Tract Infections (UTIs)”
seattlechildrens.org/pdf/PE179.pdf
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