Pediatric Surgical Associates, Ltd.
Patient Education

Cloacal Malformations/Urogenital Sinus

What is a cloacal malformation?

A cloacal malformation is an extremely rare defect of development in which a female baby girl is born with one combined opening in their perinium area instead of three separate openings. The perinium is the area of skin between the female genitals and the anus. This malformation occurs in about 1 in 25,000 to 50,000 newborns. In urogenital sinus, only the vagina and urethra do not separate.

Development of the baby's intestinal, genital and urinary tract tissues during pregnancy come together in one unit known as cloaca. During normal development the one-unit breaks or splits into three single tracts as the baby continues to grow. The three tracts create the anus, vagina and urethra. If this process does not split correctly, the tracts do not separate from one another. These three tracts come together into what is called a common channel, with one opening-a cloaca.

Surgery is needed to correct the condition, creating three separate openings for the anus, vagina, and urethra. This surgery is usually performed during your baby's first year of life.

Different types of cloacal malformations

All the different types of cloacal malformations consist of one opening, but they can have different appearances. This one common opening will lead to three separate tracts. Severity and type of treatment depend on how long the one common channel is. The range can be from 1 centimeter to 10 cm in length.

- A short common channel happens when the tracts come together at a low point. The three tracts join near the opening. The baby can have their body's waste products leave easily out the common channel.
- When the three tracts come together at a higher point from the perineum/opening, this is considered more complicated. If the common channel is longer, it can make things more difficult for waste to be eliminated.

Symptoms and Causes

Children with cloacal malformation may have:

- Appearance of enlarged clitoris
- An abnormal appearing anus or no anus
- More than one cervix, uterus or vagina
- Urological problems with the kidney or ureter
- Heart or spinal cord issues
- Problems with the gastrointestinal tract

Are there complications related to being born with a cloacal malformation?

This malformation requires treatment shortly after birth so a newborn can get the waste products out of their body. Typically, children born with a cloacal malformation will grow and have normal urinary and bowel function. However, due to associated spinal or nerve issues, It is possible that some children may not have full control over their urination and stool passing. Some children will have a hard time with potty training and may have some incontinence issues with one or both systems as they grow.

How is a cloacal malformation diagnosed?

It is sometimes possible during pregnancy to see a cloacal malformation on a prenatal ultrasound. A MRI during pregnancy can reveal a cloacal malformation. After birth, a physical exam and further radiology imaging will help determine if it is a short or long common channel.

If the cloacal malformation is noted after birth, the baby may have abdominal swelling. Testing to further evaluate this condition includes:

- Ultrasound: This will look at the kidneys, bladder, female pelvic organs and spine
- X-rays: Fluoroscopy (dye instilled) will assess how the urinary tract is working.
- Echocardiogram: Evaluate for any heart problems

Treating cloacal malformation

Your child will see a pediatric urologist and a pediatric surgeon. If there are heart concerns, a pediatric cardiologist will also be a part of their care. The malformation requires surgery to separate and make three openings in your child's perineum. One for the urethra to eliminate urine, the vaginal opening and the anus. The goal is the ability for your child to be able to urinate and pass bowel movements normally.

Your baby may have surgery shortly after birth to create a colostomy or a loop colostomy to allow stool to pass. The babies usually need a catheter to help the bladder drain the urine, as the path may be obstructed. If the vagina has fluid accumulation, a pediatric surgeon may also drain your child's vagina.

Reconstruction is usually performed between the ages of 6 months to 1 year of age. This surgery separates the common area and creates a rectum. Tissue from the area is also used to create a vagina and opening along with a urethra. If the common channel is longer, these procedures can be more complicated.

The surgery to separate the common channel into three, they will eventually have surgery to close the colostomy or loop colostomy, and bring rectum thru perineum.

What are the expectations of having a cloacal malformation?

- Short common channels babies will have about a 90% chance of having complete normal urinary and rectal function with good control.
- More complicated malformations where the channel is longer will have about a 70% chance of having that control of their bladder and bowel. Urinary incontinence is a more frequent problem. Some children will need to catheterize their bladders to help empty the urine several times a day.

Future living when cloacal malformation has been diagnosed

- Be an advocate for your child's health and let your child's pediatrician and urologist know if you notice symptoms like urinary or bowel incontinence as they grow.
- Most children with this condition can have an enjoyable sex life in adulthood.
- It may be slightly difficult to become pregnant with this diagnosis compared to others, but this does not mean your child cannot have children, nor should they have fertility issues.

11/25 PSA RN