Cloacal Anomalies
(Cloaca, high imperforate anus)

Patient and family information, brought to you by the Education Committee of APSA

Overview - "What is it?"

A cloacal anomaly is a result of abnormal development of the urinary, vagina and anal openings. Instead of three separate openings, there is only one opening where urine and stool comes out. (Figure 1). This opening connects to the urethra/bladder (urine), vagina/uterus and rectum (stool). This problem occurs only in girls. It is the most complex malformation of this area and is rare, occurring in 1 in 20,000 live births.

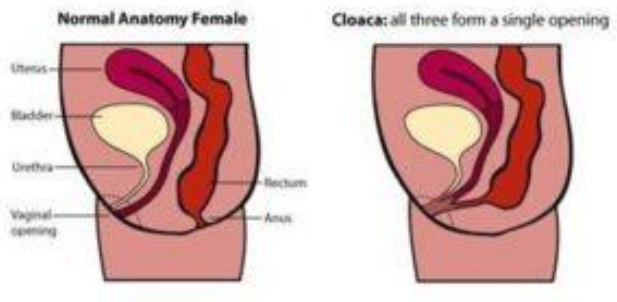

Figure 1: Comparing normal anatomy to cloaca

Signs and Symptoms - "What symptoms will my child have?"

There is an abnormal appearance of the area where the vagina, urinary opening (*urethra*) and opening for stool (*anus*) looks abnormal. There is only one hole instead of three separate holes for these body functions. Urine and stool will be noted coming from same one hole. The abdomen may be distended (look and feel "full").

Diagnosis - "What tests are done to find out what my child has?"

Physical exam by an experienced professional is critical in making the correct diagnosis.

Labs and tests: Babies with cloaca have higher risk of certain types of other abnormalities. It is important to find out if the baby has these abnormalities so nothing is missed that can impact the baby's health moving forward. The most common abnormalities are grouped into the "**VACTERL**" association (named after the first letter of the most common anomalies).

- **V**ertebral (spine): Spine X-ray to look for abnormalities of the bones and ribs. Spine ultrasound or MRI may be obtained.
- **A**nus: Baby has high imperforate anus/cloaca.
- **C**ardiac (heart): An ultrasound of the heart (echocardiogram) is needed to check for problems such as abnormal holes, problems with valves, etc.
- **T**racheo-**E**sophageal fistula (TEF): Abnormal connection between airway (trachea) and esophagus (tube that connects mouth to stomach) and a blind-ending esophagus in neck.
- **R**enal (kidney): Ultrasound of kidneys to look for abnormalities.
- **L**imbs (arms and legs): Examine arms and legs for deformity. Arms most common place for abnormal bones.

These babies usually need **ultrasound or MRI pelvis** to look at ovaries and uterus.

Treatment - "What will be done to make my child better?"

There is no medical treatment for cloaca, only surgery. The ultimate goal is to separate the urinary tract, stool opening (rectum) and the vagina. These surgeries are done in stages, as they are so complicated that the best results are achieved when the baby is bigger.

First Surgery

- The goal of the first operation is to define the anatomy, as cloacal anomalies are not all the same. This is done by placing a scope in the opening and following where the channels lead—the urinary tract (bladder), the vagina and uterus, and the intestinal tract (rectum)
- Next, it is important to separate urine flow from stool output, since mixing of stool and urine can lead to infections of the bladder and can damage the kidneys. If the drainage of the bladder is not adequate, a tube may be placed through the belly to drain the bladder (*vesicostomy*)
- Often, adequate passage of stool is accomplished by creating a colostomy. A colostomy is when the large intestine is brought out through the belly and stool comes out into a bag. A mucus fistula is another hole where the lower portion of the intestine can drain out.
- Sometimes, the vaginal opening is so narrow that the womb (uterus) is filled with fluid, making the belly big. If so, the womb also needs to be drained of fluid to allow it to

normalize in size. If the drainage of the uterus is not adequate, a tube may be placed through the belly to decompress the womb (*vaginostomy*).

Postoperative care for the first surgery includes starting feeds when there is poop coming out of the colostomy. Parents will be taught colostomy care.

Risks for the first operation include infection, bleeding, possible dislodgement of tube if they were placed, and anesthesia risks.

Benefits for the first operation include figuring out the specific anatomy of the cloaca. Another benefit is that the urine flow is separated from the stool, decreasing chances of urinary infection and kidney damage.

Informed consent: A consent form is a legal document that states the tests, treatments or procedures that your child may need and the doctor or practitioner that will perform them. Before surgery, your doctor should tell you what the operation is, the goal of the surgery and other possible treatment options that are available. Your doctor should explain the risks and benefits of the surgery. You give your permission when you sign the consent form. You can have someone sign this form for you if you are not able to sign it. You have the right to understand your child's medical care in words you know. Before you sign the consent form, make sure all of your questions are answered. It is important to know that during surgery, there are things that can happen that your doctor may have not predicted before going in. He or she will explain these to you after the surgery.

Second Surgery

- The goal of the second surgery is to repair the combined urinary, vaginal and rectal anomaly and create a separate hole is for each system. Depending on how complicated the abnormalities are, the repair may be done from the bottom only, or a combination of bottom and belly approaches. The timing of the second surgery is dependent on many factors: baby must growing and gaining weight, other abnormalities may need to be addressed first (heart, spine, etc). Other studies may need to be done prior to the surgery. Your surgeon will decide on this.
- The second surgery starts with a scope inserted through the single opening to look at the channel to the bladder, vagina and anus. A decision can then be made as to whether the operation can take place from the bottom or whether there needs to be an incision on the bottom and another incision through the abdomen. The repair then consists of making an opening for the urethra (urine), one for the vagina, and one for the new anus (stool). The colostomy remains in its same location.

Postoperative care involves remaining in the hospital for pain control, antibiotics, and to monitor for proper wound healing. A bladder catheter will be kept in place. Stool comes out of the colostomy so that stool does not contaminate the new incisions. The baby will return to clinic in 2-3 weeks to start learning dilations of anal opening. Parents will then perform dilations at home to keep the hole from closing up.

Risks of the second operation include infection, bleeding, and risks of anesthesia, in addition to a risk of damaging the vagina or urethra when trying to separate and create the three holes.

Benefit of the second operation is that now there are three distinct holes (normal anatomy) for the child's urinary and stool evacuation, and for eventual sexual function.

Third surgery

- The goal of the third and final surgery is colostomy closure to reconnect the intestine so the baby can poop out of the new anal opening and not have to use a colostomy anymore.
- The timing for colostomy closure is dependent on whether the sites for the previous operation are completely healed. Before this surgery, the new anus needs to be completely healed. Your surgeon will order a bowel prep day before surgery—usually clear liquids and magnesium citrate or golytely (surgeon preference).
- The third surgery starts with an exam under anesthesia to look at anus and then the ostomy will be taken down to reconnect colostomy and mucus fistula so stool can now come out the new anus.

Postoperative care includes antibiotics after surgery and continuing anal dilations. Diaper rash cream must be applied to the buttocks since the baby's bottom has not yet seen stool and the skin is very prone to breakdown.

Risks of colostomy closure include a small risk of bleeding and risks from the anesthesia. Also, the previous colostomy wound will closely be watched for signs of infection. Dressing changes may be needed. There is also a risk of leak from anastomosis (where the colostomy was connected to the other part of the colon downstream).

Benefits: The colostomy is gone and the baby's intestine is now all connected.

Home Care - "What do I need to do once my child goes home?"

Diet: Your child may eat a normal diet after surgery.

Activity: Your child should avoid straddling after the first two operations.

Wound care: Surgical incisions should be kept clean and dry for a few days after surgery. The stitches used in children are absorbable and do not require removal. Your surgeon will give you specific guidance regarding wound care, including when your child can shower or bathe.

Medicines: Medicines for pain such as acetaminophen (Tylenol) or ibuprofen (Motrin or Advil) or something stronger like a narcotic may be needed to help with pain for a few days after surgery.

What to call the doctor for: Call your doctor for worsening belly pain, fever, vomiting, diarrhea, problems with urination, decreased stool output or if the wounds are red or draining fluid.

Follow-up care: Your child should follow up with his or her surgeon 2-3 weeks after surgery to ensure proper post-operative healing. IT IS IMPORTANT TO KEEP ALL APPOINTMENTS WITH THE SURGEON, AS HEALING IS CRITICAL IN THE FIRST FEW MONTHS AFTER SURGERY.

Long Term Outcomes - "Are there future conditions to worry about?"

Children after repair still may face long term problems with fecal and urinary incontinence (inability to control when they poop or pee). Sixty percent may have soiling (accidents), 35% may have fecal incontinence, 39% may have constipation, and 70% may have urinary incontinence. Patients with fecal incontinence who are involved in a bowel management program have success rates of 88-93%. They may have to see a specialist (urologist) for problems with urinary incontinence.

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