Let's Talk About...

Anorectal malformation

What is an anorectal malformation?

Anorectal malformation (aye-no-RECK-tull mal-for-MAY-shun) is a birth defect. It is also called **imperforate anus** (im-PER-for-at AYE-nuss). Imperforate anus is when there is no opening for the stool (poop) to exit the body. The rectum (the last part of the intestines) does not connect to the outside of the body. There may be other problems with the urinary tract, genital structures, heart, and spine.

- There can be a problem with muscles that control bowel movements. This can cause constipation or fecal incontinence (cannot control bowel movements).
- Sometimes the rectum connects to the urethra (where the urine or pee exits the body), the bladder, or in girls, the vagina. This is called a **fistula** (FISS-teu-lah). This means that stool may exit the body from a different place.
- Sometimes the anus is small or in a different place on the bottom. This can prevent stool from exiting the body normally.
- In girls, the rectum, vagina, and urinary tract can meet in a common space inside the body. This is called a persistent **cloaca** (cloh-ACHE-ah). When the doctor examines the child, there is only one opening, instead of three.

What causes anorectal malformation?

Anorectal malformations happen when the baby develops in the womb. Doctors do not know why this happens. It is not something that you can control. It is not caused by something you ate, drank, or did when you were pregnant. In rare cases, it is inherited.

How does the doctor know my child has anorectal malformation and does my child have other problems?

Your child's doctor probably found the imperforate anus during your baby's first physical exam. Other body structures may have developed differently, as well. These may also require surgery. Your child's doctor will look carefully for these problems:

- The spine, especially the lower spine, may not develop properly. The spinal cord (the nerves from the spine) may be attached wrong (tethered cord), or the backbone and spinal cord do not close before birth (spina bifida). This can cause problems with bowel (poop) and bladder (pee) control.
- Heart does not develop properly.
- Trachea (windpipe) or esophagus (tube that leads from mouth to stomach) may not develop normally.
- The kidneys (structure that makes urine or pee) and urinary tract (tubes from the kidney to the outside) may not develop normally.
- The female reproductive parts may not develop normally.
- Arms and legs may not develop normally.

When a child has three or more of these problems, doctors say they have VACTERL syndrome (V-vertebral, A- imperforate anus, C-cardiac, T- trachea, E- esophagus, R- renal, L- limbs). Your doctor will explain if this is the problem.

What tests does my child need?

Your child's doctor may order tests to understand your child's problems. These are some tests your child's doctor may order:

- **Ultrasound:** a machine with sound waves that can make a picture of the inside of the body.
  - **Abdominal (belly):** looks at kidneys and the bladder. In girls, it also looks at the vagina and uterus. It can also see if there are any other masses.
  - **Spinal:** looks for tethered cord.

- **Echocardiogram:** a machine with sound waves that looks for problems in the heart.
- **X-ray:**
  - **Abdominal:** looks for masses in the belly.
  - **Spinal:** looks at size and shape of the sacrum (tailbone). This helps predict if the child will have bowel control.
- **VCUG:** A test using dye placed into the bladder. It looks for urine backing up into the kidneys. It can see connections from the urinary tract to the bowel.
- **Cystoscopy:** A small camera looks at the urinary system (urethra, bladder, and ureters).
- **Vaginoscopy:** A small camera looks at the reproductive system (vagina and cervix).

How will my child be treated?

Most babies with anorectal malformation need one or more surgeries. Surgery will make a way for stool to come out of the body. Usually, the surgeon waits 24 to 48 hours after birth before doing surgery. This is to see if there is a fistula, where stool comes out a different way. If your child's belly is very full or has another urgent problem, she may have surgery right away. Your child's surgeon and her other doctors will decide the best time for your child to have surgery. Your child may need surgery for other parts that didn't develop properly, as well.

For anorectal malformation, there are two ways your child can be treated: primary reconstruction and staged reconstruction.

Primary Reconstruction

This is when the surgeon fixes the imperforate anus with one surgery. If a baby has a **perineal** (pair-ih-KNEE-ahl) fistula (the rectum opens on the bottom in a different place), the surgeon creates a new anus in the correct location.

Staged Reconstruction (this is the usual surgery)

- **Step 1: Create a colostomy**
  This is the first surgery. This surgery usually happens in the first day or two after birth. The **colostomy** (coe-LOSS-toe-me) is a different opening for stool to exit the body. The surgeon cuts the intestine and brings both ends to the belly skin. He makes an opening to the outside for these ends. Your child will have a bag on her skin to collect stool that comes out. This end is called a stoma. The other end allows mucus from the intestine to come out. This end is called a mucus fistula. With the colostomy, your child can eat and grow before the next surgery.
- **Step 2: Anorectoplasty (aye-no-RECK-toe-plass-tee) or reconstruction of rectum and anus:**
  This is the second surgery. This usually happens 3–6 months after birth. Exactly when your child will have this surgery depends on your child's other problems. The surgeon separates the rectum from the urinary tract, if needed, and brings the rectum to the skin on the bottom to make a new anus. Your child will continue to have a colostomy while the new anus heals. This surgery is called a Posterior Sagittal Anorectoplasty or PSARP.

  Your child's doctor will teach you to do anal dilations a few weeks after surgery. This is a way of stretching the anus so that it stays the right size. Anal dilations prevent the anus from becoming too small, and unable to pass stool. The surgeon will tell you what size dilators to use and decide when to close the colostomy.
- **Step 3: Close the Colostomy**
  This is the third surgery. This usually happens 2–3 months after the anorectoplasty. At this surgery, the colostomy is closed. The surgeon reconnects the two ends of the colon and stool can pass through to the new anus. Your child will start passing stool through the new anus in about 2–3 days.

  At first, your child's stools will be loose and frequent. Your child may have a severe diaper rash. Your doctors and nurses will teach you how to prevent and treat diaper rash. The stools usually become more normal and happen less often. This may take a few weeks, or sometimes months. Some children become constipated when the

stool is less frequent. Your medical team will work with you to prevent and treat this problem.

Will my child have problems later in life?

Your child should do well after the surgeries. Your child will continue to see the doctors about her imperforate anus as a child and teen. Some children are followed by their doctor even when they are an adult. The most common problems your child may have are constipation or fecal incontinence (problems holding stool). You child may have other problems with the urinary tract or kidneys. Depending on your child's problem, she may have bowel or urine control problems. The Colorectal Center at Primary Children's Medical Center addresses these problems and coordinates care between different doctors to manage your child's needs. The Colorectal Center provides a Bowel Management Program. This program will work closely with you and your child. They can provide a way for your child to stay clean of stool and prevent accidents.

Constipation:

Constipation is a problem for children born with anorectal malformations who have a good chance to have bowel control. Once constipation develops the colon can become dilated, which makes constipation worse. This can result in liquid accidents. This is called overflow incontinence or **encopresis** (en-cah-PREE-sis). The stool gets hard and stuck in the dilated colon. Liquid stool leaks around the stool. This is called pseudo-incontinence. The goal is to prevent constipation by making sure your child empties her colon every day.

Incontinence:

Stool incontinence is a problem for children born with an anorectal malformation who have a poor chance of normal bowel control. This is when the child cannot control bowel movements and may have accidents or soil their underwear.

Fecal (stool) continence is when a child does not have stool accidents. To have stool continence, a child needs to be able to:

- feel the stool in the rectum
- have good movement of the colon
- have good control of the anus muscles

The best way to tell if a child will be continent is the type of anorectal malformation they have and if they have problems with the spine.

The Colorectal Center will work closely with you to keep you child clean and in normal underwear.

What do I do if I have other questions?

You can call your doctor or surgeon.

Web page:

- www.primarychildrens.org/colorectalcenter

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