Let's Talk About...

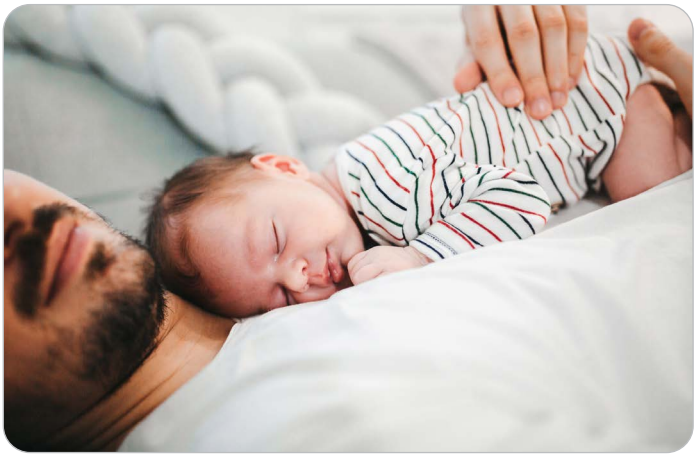
Anorectal Malformation

What is anorectal malformation?

Anorectal [aye-no-RECK-tuhl] **malformation** [mal-for-MAY-shun] is when the rectum (the end of the large intestine or colon) does not connect to the outside of the body, opens in the wrong place on the baby's bottom, or is too small.

What causes it?

Anorectal malformation happens as the baby develops in the womb. Doctors do not know why it happens. In rare cases, it is inherited.



What can happen if my child has an anorectal malformation?

Anorectal malformations can cause:

- **Constipation** (can't poop) or **fecal** [FEE-kuhl] **incontinence** (no control of bowel movements) caused by bowel muscle problems
- A **fistula** [FISS-teu-lah], which is when the rectum connects to another body part, such as the urethra (where the urine or pee exits the body), the bladder, or vagina.
- A **small anus** (opening to the rectum) or an anus formed in a different place
- A **persistent cloaca** [cloh-ACHE-ah] in girls. This is when the rectum, vagina, and urinary tract connect into a single channel inside the body

How is it diagnosed?

In most cases, imperforate anus can be diagnosed during your baby's first physical exam. In some cases, an exam needs to be done while the baby is under anesthesia to confirm the diagnosis.

What other problems are associated with anorectal malformation?

Anorectal malformation is associated with the following problems:

- Tethered cord: When the spinal cord attaches to the wrong place.
- Spina bifida: When the backbone and spinal cord do not close before birth.
- Underdeveloped heart.
- Problems with a baby's:
  - Trachea (windpipe) or esophagus (food tube)
  - Kidneys and urinary tract (tubes that carry pee from the kidney to the outside)
  - Female reproductive organs
  - Arms or legs

When a child has 3 or more of these problems, doctors say they have **VACTERL association**:

- V-vertebral
- A-imperforate anus
- C-cardiac
- T-trachea
- E-esophagus
- R-renal
- L-limbs

How is VACTERAL association diagnosed?

The doctor may order these tests to look for problems commonly found with VACTERL association:

- Ultrasounds (picture using sound waves) to look at the spine, kidneys, and bladder, as well as the vagina and uterus in girls
- Echocardiogram (heart ultrasound) to look for heart problems
- Abdominal x-rays to look for masses in the belly
- Spinal x-rays to look at the size of the tailbone and shape of the sacrum (which can help determine bowel control)
- Voiding cystourethrogram [SIS-tow-you-REE-thro-gram], or VCUG, which uses dye in the bladder to see if urine is backing up into the kidneys
- Cystoscopy [sis-TOSS-kuh-pee], which uses a small camera on a thin, flexible tube to look at the urethra, bladder, and ureters
- Vaginoscopy [vaj-in-OSS-kuh-pee] (for girls), which uses a small camera on a thin, flexible tube to looks at the vagina and cervix

How is anorectal malformation treated?

Most babies with anorectal malformation need one or more surgeries to make a pathway for poop to leave the body. Usually, the surgeon waits 24 to 48 hours after birth to see if a fistula is causing poop to come out a different way. Children who have full bellies or urgent problems, however, may need surgery right away.

Some babies may have a **perineal** [pear-eh-KNEE-ahl] **fistula**. This is when the rectum opens in a different place on their bottom. In this case, the surgeon will create a new anus in the correct location in a single surgery. This is called **primary reconstruction**. Otherwise, anorectal malformation is usually repaired with a 3-stage surgery called **staged reconstruction**.

What happens during staged reconstruction surgery?

1. **Colostomy** [koh-LOSS-tuh-mee]: Usually within 1 to 2 days after birth, the surgeon performs a colostomy. During this surgery, the colon (large intestine) is cut in 2 places. The cut ends are attached to the skin of the belly, creating an opening for poop to leave the body. The openings are called **stoma** [STOW-mah]. The stoma are covered with an ostomy bag that collects the poop. With a colostomy, your child can eat and grow before the next surgery.
2. **Posterior sagittal** [SAJ-jit-tuhl] **anorectoplasty** [ay-no-REK-toh-plass-tee] **(PSARP):** This is a surgery to separate the rectum from the urinary tract (if needed) and route the rectum to the skin on your child's bottom to make a new anus. It is usually done within 3 to 6 months after birth. In some cases, a **laparoscopic** [lap-ah-roh-SKOP-ik] **surgery** is also needed. This part of the surgery is done with small openings and small cameras in the belly. The colostomy remains in place while the new anus heals. A few weeks after surgery, your child's doctor will teach you to do **anal dilations**. These dilations stretch the anus so that it's large enough for poop to pass through.
3. **Closing the colostomy:** The colostomy is closed by the surgeon usually within 2 to 3 months after the anorectoplasty. During this surgery, the 2 ends of the colon are reconnected. Poop will begin passing through the new anus in about 2 to 3 days. Your child may have a severe diaper rash at first. Your child's healthcare provider will help you prevent and treat the rash. Expect their poop to be loose and frequent at first. It can take a few weeks or months to become more normal. Some children become constipated when the poop is less frequent, but providers can help you prevent and treat this problem.

Will my child have problems after reconstruction surgery?

Your child may have problems with:

- **Constipation:** When poop gets hard and stuck in the colon, the colon can become dilated and cause liquid poop to leak around the hard poop. This is called **encopresis** [en-cuh-PREE-sis] or **overflow incontinence.** To prevent constipation and other problems, make sure your child poops and empties their colon every day.
- **Bowel control:** Some anorectal malformations and spinal cord problems can make it hard to control bowel movements. Your child's surgeon will help you know whether your child will have bowel control problems after surgery.

If your child has bowel problems, work with their healthcare providers to create a bowel management program. You can learn ways to prevent accidents and help your child stay in regular underwear. Healthcare providers can also help if your child has kidney or urinary tract problems.

What if I have other questions about anorectal malformation?

If you have more questions, call your child's doctor or surgeon, or visit: **Primarychildrens.org/colorectalcenter**.

Notes