Anorectal malformation (a-nor-rek-tul mal-for-may-shun) is a birth defect that occurs when there is no opening for the stool (poop) to leave the body. It is also called imperforate anus (im-per-for-ayt AYE-nuss). When your child has anorectal malformation, the rectum (end of the intestines) does not connect to the outside of the body.

What happens if my child has anorectal malformation?

Anorectal malformation can cause:

• Constipation or fecal incontinence (no control over bowel movements) caused by bowel muscle problems

• A fistula (FISS-teu-lah), which occurs when the rectum connects to the urethra (where the urine or pee exits the body), the bladder, or vagina. When this happens, stool tries to leave the body by passing through these organs.

• A small anus or anus formed in a different place

• A persistent cloaca (cloh-ACHE-ah) in girls, which occurs when the rectum, vagina, and urinary tract meet in one space inside the body

What causes anorectal malformation?

Anorectal malformations happen when the baby develops in the womb. Doctors do not know why this happens. In rare cases, it is inherited.

What are the signs of anorectal malformation?

A doctor will usually find an imperforate anus during your baby’s first physical exam. Other signs of anorectal malformation may include:

• Tethered cord (a neurological disorder where the spinal cord attaches to the wrong place)

• Spina bifida (backbone and spinal cord do not close before birth)

• Underdeveloped heart

• Trachea (windpipe) or esophagus (tube from the mouth to stomach) problems

• Kidney and urinary tract (tubes from the kidney to the outside) problems

• Female reproductive organ problems

• Arm and leg problems
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 child’s doctor will explain if this is the problem.

**How is anorectal malformation diagnosed?**

To diagnose anorectal malformation and understand your child’s health problems, the doctor may order these tests:

- Ultrasounds (picture using sound waves) to look at the spine, kidneys and bladder (and 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 tailbone’s size 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 look for urine backing up into the kidneys
- Cystoscopy (small camera that looks at the urethra, bladder, and ureters)
- Vaginoscopy for girls (small camera looks at the vagina and cervix)

**How is anorectal malformation treated?**

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

If your child has a perineal (pair-ih-KNEE-ahl) fistula, where the rectum opens on the bottom in a different place, the surgeon will create a new anus in the correct location in one surgery, called primary reconstruction. Otherwise, the surgeon usually repairs anorectal malformation with a three-stage surgery called staged reconstruction.

### What happens during staged reconstruction surgery?

#### 1 Colostomy: Usually 1–2 days after birth, the surgeon cuts the intestine, brings both ends to the belly skin, and makes an opening outside the body. This stool opening is called a colostomy (coe-LOSS-toe-me). A stoma (bag on the skin) collects stool that comes out, and the mucus fistula (other end of the opening) allows mucus from the intestine to come out. With the colostomy, your child can eat and grow before the next surgery.

#### 2 Posterior sagittal anorectoplasty:

Usually 3–6 months after birth, the surgeon separates the rectum from the urinary tract, if needed, and brings the rectum to the skin on your child’s bottom to make a new anus. This is called posterior sagittal anorectoplasty (ay-no-RECK-toe-plas-tee), or PSARP. Your child may have a laparoscopic surgery (done with a small camera) and will still have a colostomy 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 and prevent it from being too small so stool can’t pass through.
3 **Closing the colostomy:** Usually 2–3 months after the anorectoplasty, the surgeon closes the colostomy and reconnects the two ends of the colon. Stool can pass through the new anus in about 2–3 days. Your child may have a severe diaper rash, which the healthcare provider can help you treat and prevent. Stools will be loose and frequent at first, and it can take a few weeks or months to become more normal. Some children become constipated when the stool is less frequent, but providers can help you treat and prevent this problem.

**Will my child have problems after reconstruction surgery?**

Your child may have problems with:

- **Constipation:** When stool gets hard and stuck in the colon, the colon can become dilated and cause liquid stool to leak around the stool. This is called encopresis (en-cah-PREE-sis) or overflow incontinence. To prevent constipation and other problems, make sure your child poops and empties the 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 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.