balloonnotes

children'shealth

When your child has an Anorectal Malformation (ARM).

What is an ARM?

An anorectal [ā-nō-rek-tel] malformation (ARM) is sometimes also called imperforate [im·per·fo·rate] anus.

- It is a congenital defect (present at birth)
- It happens when the baby is developing early in pregnancy. This is when a baby's **rectum** (where poop is stored) and **anus** (where poop leaves the body) are forming.
- Also some of the muscles and nerves which help the baby poop may not work as well.

The cause is unknown. This defect is found in most babies shortly after birth when the baby does not poop, has a swollen belly, or when the doctor checks the baby and the anus is missing or in the wrong place.

What are the types of ARMs?

Malformations found in **both girls and boys** are:

- Recto-perineal fistula – the rectum opens in a hole on the skin between the anus and genitals (perineum). It may be mistaken for the anus, but it is not in the normal place for the anus.
- Anal or rectal atresia and/or stenosis – the anus or rectum is too small for poop to be passed normally.
    - **Atresia** means the structures are missing or not formed correctly.
    - **Stenosis** means the structures are too thin or only partly opened.
- Cloacal [clo·a·cal], exstrophy [ek´stro-fe] – the intestines are on the outside of the body. The lower parts of the reproductive system, urinary system and intestinal tracts may not be fully formed.

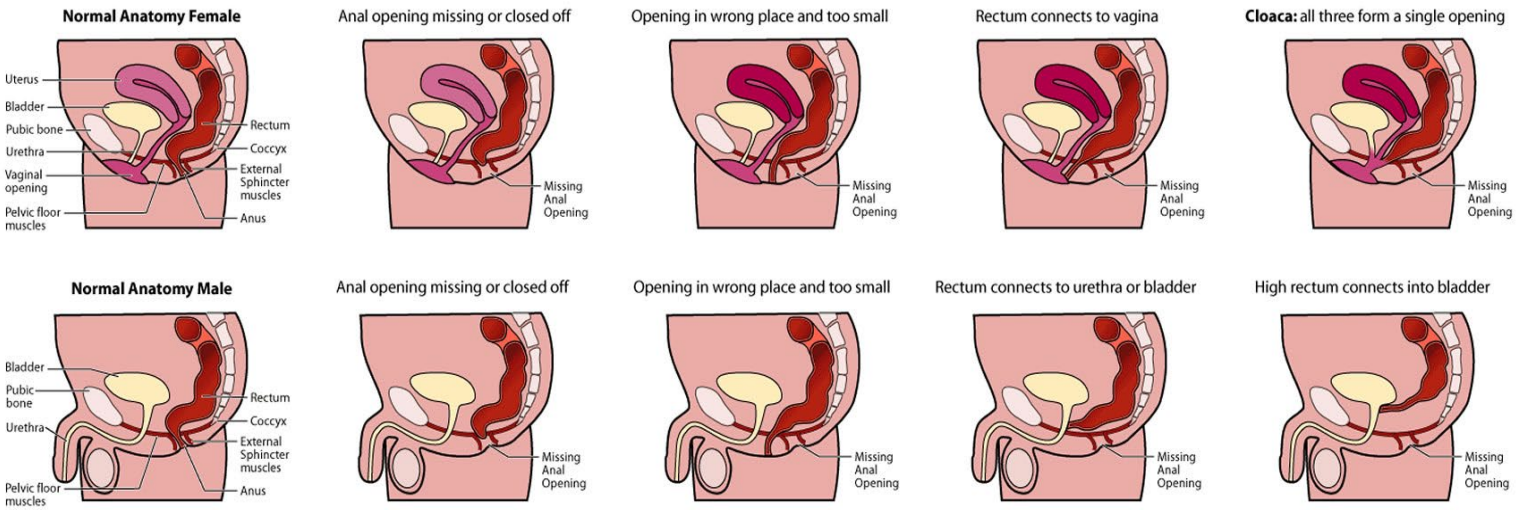
Malformations found **only in boys** are:

- Recto-bulbar urethral fistula – the anus is missing, and the rectum joins into the lower (bulbar) part of the urethra (the tube through which pee leaves the body).
- Recto-prostatic urethral fistula – the anus is missing, and the rectum joins into the higher part of the urethra, closer to the bladder (where pee is stored).
- Recto-bladder neck fistula – the anus is missing, and the rectum joins to the bottom of the bladder, where the urethra starts.

Malformations found **only in girls** are:

- Recto-vestibular fistula - the anus is missing, and the rectum opens just behind the vagina.
- Cloaca [clo·a·ca], Persistent Cloaca – the anus is missing, and the urethra, vagina, and rectum join into a single channel (tunnel) instead of 3 separate channels.

Below are examples of the different types of ARM:



Fahrion, C. (n.d.) Anorectal Malformations – all (digital image). UCSF Pediatric Surgery.

What are other related complications?

About half of children born with an ARM also have other problems. This is because these structures are developing all around the same time during the pregnancy.

- **VACTERL -** Children diagnosed with VACTERL often have 3 or more other problems of the structures listed below:
  - **V**ertebral – bones that make up the spine and sacrum (tailbone)
  - **A**nus – imperforate anus
  - **C**ardiac – heart
  - **T**rachea – windpipe
  - **E**sophagus – tube that joins the mouth to the stomach
  - **R**enal – kidneys or other parts of the urinary tract
  - **L**imbs – arms and legs

- **Tethered cord** – This occurs when the spinal cord is attached to tissue around the spine, most often at the lowest part of the spinal cord. It keeps a child from moving freely when they bend and move.

- **Sacrum and Spine anomalies** – Some children are missing part or all of the sacrum (the large group of bones at the lower spine).

- **Cardiac defect** – A congenital heart defect is a problem with the structure of the heart. It is the most common type of major defect found with imperforate anus. The type and how bad the defect is, will be different for each child.

- **Esophageal Atresia / Tracheoesophageal Fistula - Esophageal atresia** is a defect that occurs when the upper esophagus does not join with the lower esophagus and stomach. A **tracheoesophageal fistula (TEF)** is present when the esophagus joins with the trachea (windpipe).

- **Kidneys and Urinary tract** – The kidneys may not be formed or work normally. Pee (urine) may back up from the bladder towards the kidneys, or the bladder may be small and not drain normally.

- **Gynecologic anomalies** - Girls born with ARM may not have normal development of the inside female structures, also called gynecologic or reproductive structures (uterus, fallopian tubes, and vagina).

What tests will be done?

If an ARM is diagnosed, the doctor will order other tests to better understand the anatomy and check the baby for other known malformations. These tests are:

- X-ray of the belly to show where the rectum ends.
- Renal ultrasound to check for kidney or urinary problems.
- X-ray of the spine to check for spine problems.
- Spinal ultrasound to check for a tethered cord or other problems with the spinal cord.
- Spinal MRI to check for other spine problems or to learn more about those found on ultrasound.
- Echocardiogram to check the heart's structure.

What are the treatments/procedures needed?

A baby with an ARM will always need a surgery to fix the malformation. Which surgery is done will depend on the type of defect. For some cases, a primary repair (single surgery) will be done, and no ostomy is needed. However, if the malformation is complex, surgery will be done in stages, which will include an ostomy.

- **Ostomy** – An ostomy is a small opening made in the belly. This opening is connected to the intestine. Poop leaves the body through this opening and empties into a bag. A second opening is called the mucous fistula and lets the closed end of the rectum drain fluid or mucous. An ostomy may be done first to allow your child a couple of months to grow and get stronger.

- **Distal Colostogram** - Before the 2nd surgery your child may need to have this study to help find out if there is a fistula, or connection to the urinary or reproductive system, and to measure the distance from the end of the rectum to the skin.

- **Posterior Sagittal Anorectoplasty (PSARP)** – This is a type of surgery that creates a new anus for poop to come out. If your child has an ostomy, it will stay in place after the PSARP while this new anus heals. This will keep poop from contaminating the surgical site.

- **Anal/Rectal Dilation** - After surgery, it is likely that scar tissue will form in the new anal opening. This could cause the anus to close. You will need to learn how to do anal/rectal dilations to slowly stretch the anus until it reaches the size that is normal for your child's age. In some cases, if there is no ostomy, you will need to do anal dilations before the PSARP to keep your child pooping well before surgery. Please see our hand out "Parent's guide to rectal dilation" for instructions on how to do the dilations.

What are the long-term concerns?

Patients with ARM will be followed closely throughout their childhood and adolescence. The most common problem for these children is constipation (can't poop) or incontinence (not able to hold poop). Other problems may be with the urinary tract or function of the kidneys. The type and extent of the malformation can help predict if your child will have problems with their bowel or bladder function.

Ask your clinic staff specific information about your child. We are here to help you understand the information so you can be a partner in your child's care.