



NATIONWIDE CHILDREN'S
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Helping Hand™

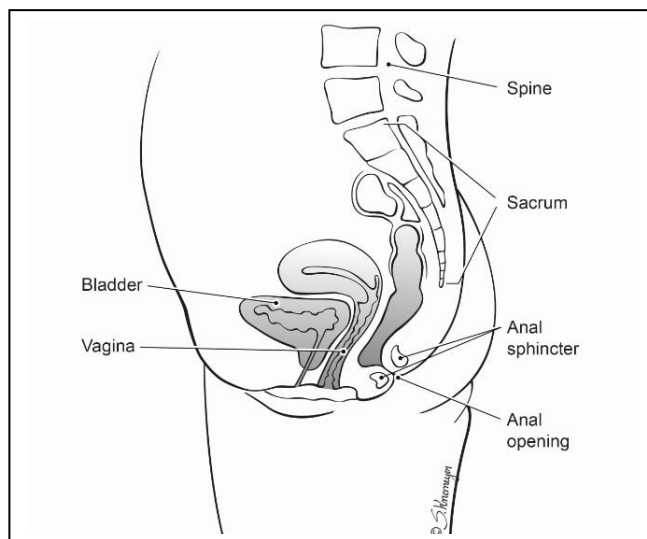
Health Education for Patients and Families

Anorectal Malformation (ARM) or Imperforate Anus: Female

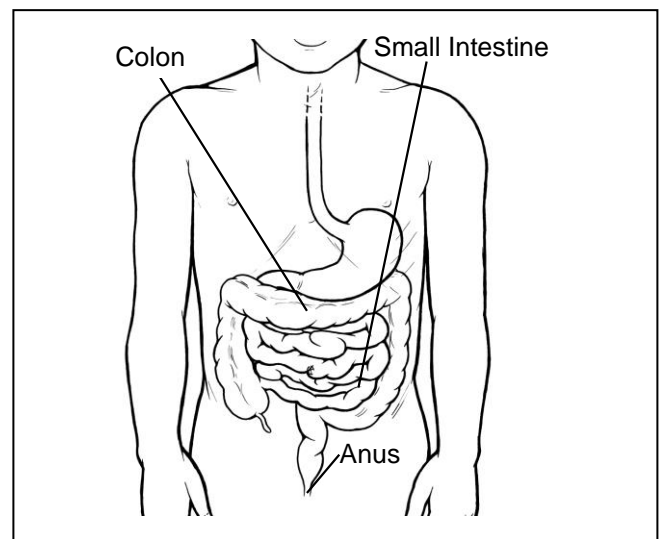
Anorectal malformation (ARM), also called imperforate anus (im PUR for ut AY nus), is a condition where a baby is born with an abnormality of the anal opening. This defect happens while the baby is growing during pregnancy. The cause is unknown. These abnormalities can keep a baby from having normal bowel movements. It happens in both males and females.

In a baby with anorectal malformation, any of the following can be seen:

- No anal opening
- The anal opening can be too small
- The anal opening can be in the wrong place
- The anal opening can open into another organ inside the body – urethra, vagina, or perineum



Picture 1 Normal organs and structures from the side.



Picture 2 Normal organs and structures from the front.

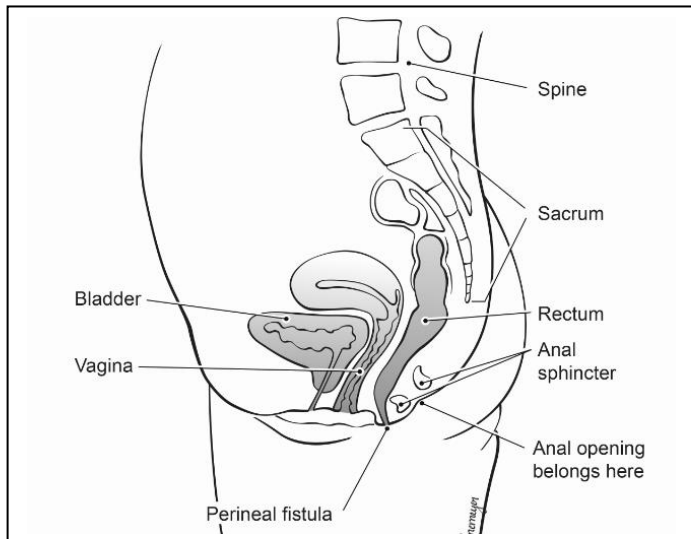
Signs and symptoms

At birth, your child will have an exam to check the position and presence of her anal opening. If your child has an ARM, an anal opening may not be easily seen. Newborn babies pass their first stool within 48 hours of birth, so certain defects can be found quickly.

Symptoms of a child with anorectal malformation may include:

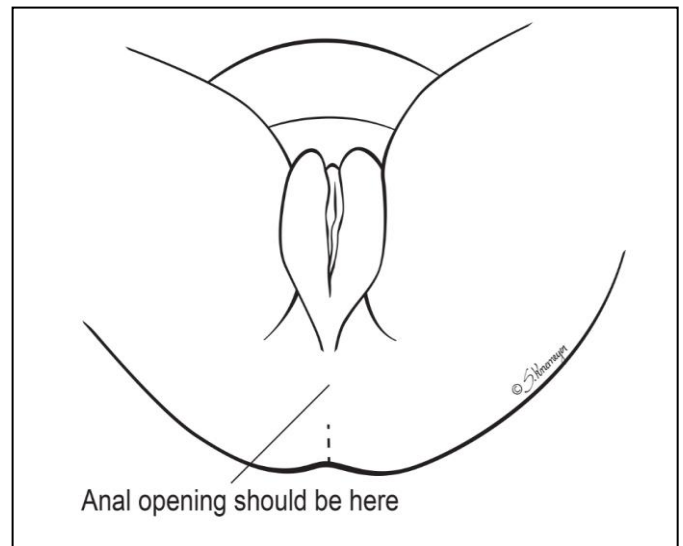
- Belly swelling
- Vomiting
- No stool within the first 48 hours
- Stool coming out of the vagina or urethra

Types of anorectal malformations

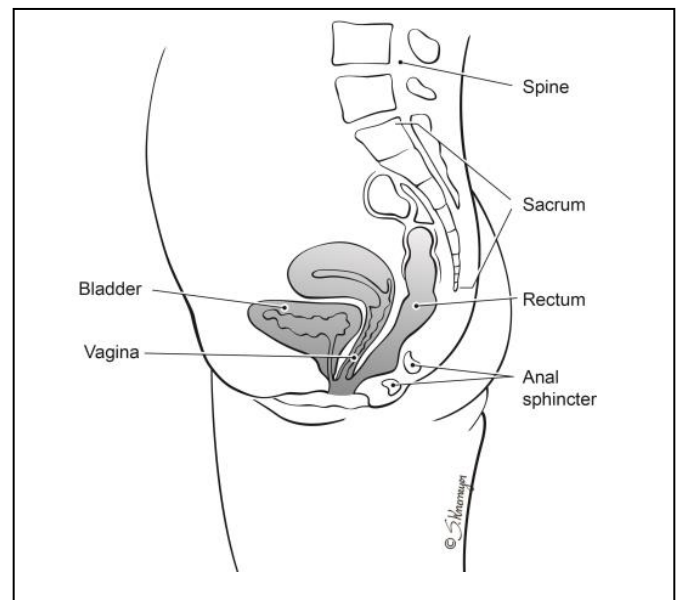


Picture 3 Perineal fistula at birth, view from side

- **Perineal fistula** – the anal opening is in the wrong place (Picture 3). The anal opening opens into the perineum. This is the place between the vagina and anal sphincter. It may also be too small.
- **Vestibular fistula** – (vess TIB u lar FIS too la) the anal opening is in the wrong place. The anal opening is behind the vagina opening into the vestibule. It may also be too small.
- **Cloaca** – (klo AY ka) the lower intestine (rectum), urethra, and vagina come together to make one opening (Pictures 4 and 5).
- **No fistula** – the anal opening is too narrow or ends before it reaches the skin. The rectum does not connect to the vagina or urethra.



Picture 4 Cloaca at birth, view from the bottom



Picture 5 Cloaca at birth, view from the side

Other possible problems

If your child has been diagnosed with ARM, she may be at risk for other possible problems. These problems include:

- **Urology problems** – problems with the kidneys and other organs that help with peeing (urination). These problems can cause infections and permanent kidney damage if left untreated. The doctor may order tests to check on these problems. Some of these problems include:
 - Only having one kidney
 - Hydronephrosis – swelling of the kidneys caused by a backup of urine
 - Vesicoureteral reflux – urine from the bladder goes back up into the kidneys
 - Neurogenic bladder – having trouble controlling the bladder
- **Gynecology problems** – Girls with anorectal malformations can have changes with their female anatomy. These changes can cause issues if left untreated. The doctor may order tests to check your child's anatomy. Some changes that may happen include:
 - Uterine didelphus – two uteruses with two separate cervixes
 - Bicornate uterus – the uterus is heart-shaped
 - Vaginal septum – vagina is divided into two sections
- **Spinal problems** – problems with the lumbar (lower) portion of the spine that assists with bowel and bladder function. Some of these problems include:
 - Tethered cord – tissue attachments of the spinal cord cause stretching of the spinal nerves. This stretching can cause loss of nerve function to the bowels and bladder.
 - Fatty filum – tissue that connects the spinal cord to the backbone is thickened.
 - Myelomeningocele – spinal cord does not form properly.
- **Sacral problems** – problems with the sacrum and coccyx (lowest) portion of the spine that assists with bowel and bladder function. Some of these problems include:
 - Caudal regression – end of the spinal cord does not fully develop
 - Hemisacrum – a portion of the sacrum does not fully develop
 - Sacral hemivertebrae – a part of the sacrum does not fully develop
 - Presacral mass – a mass between the rectum and the sacrum

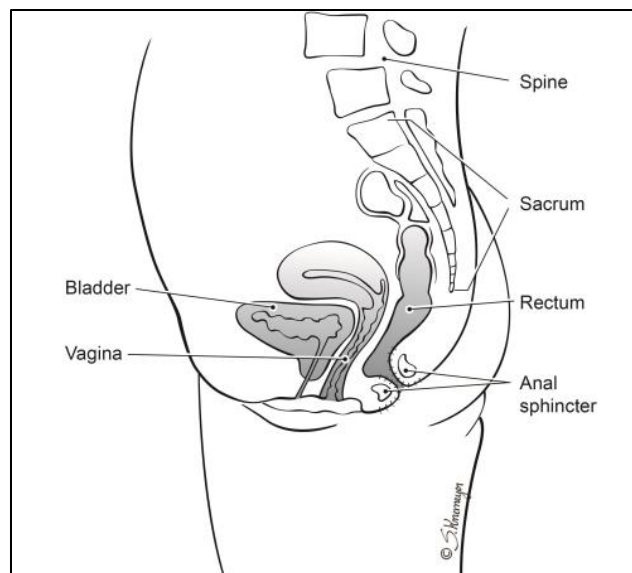
Other tests

The doctor may order tests to better understand your child's malformation. These tests will also help the doctor know if she has any other problems related to the diagnosis.

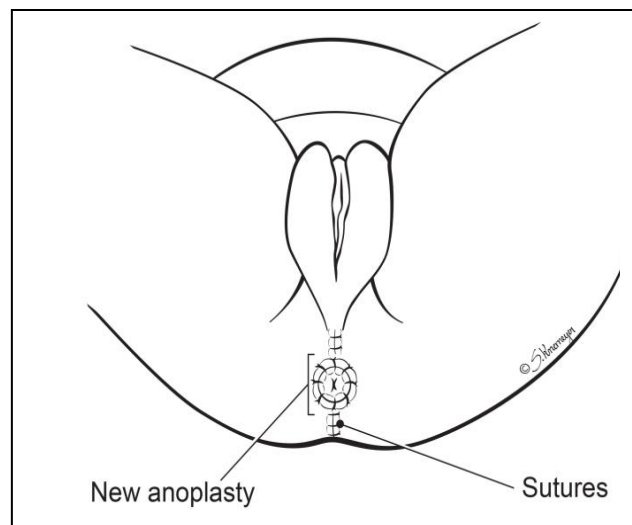
- **X-ray** – This test shows the bowel gas pattern, vertebrae of the spine, and the sacrum.
- **Ultrasound** – This test looks at your child's belly (abdomen) to find problems in the urinary tract system and vagina. It also looks to see if the spine formed in the right way.
- **MRI of the spine** – This may be done if a problem is found on the spinal ultrasound or if your child is 6 months of age or older when she needs to have her spine assessed.
- **Echocardiogram** – This test checks to see if there are any problems with your child's heart.
- **Labs (renal function panel, cystatin C)** – These labs tell the doctor more about your child's kidney function.
- **Video urodynamics** – This test shows how your child's urinary tract works, including the bladder and kidneys.

Treatment and surgery

The treatment for anorectal malformations is surgery. The repair for ARM is called a posterior sagittal anorectoplasty (PSARP). This surgery will either create your child's anal opening or put it in the right place so she can have bowel movements (Pictures 6 and 7). The type of surgery and how many surgeries your child has depends on the type of ARM and previous surgical history. If your child has a cloaca or an ARM with no fistula, she will first need a colostomy (Picture 8, next page), then a PSARP. If your child is born with a perineal or vestibular fistula, then a colostomy is not usually needed.



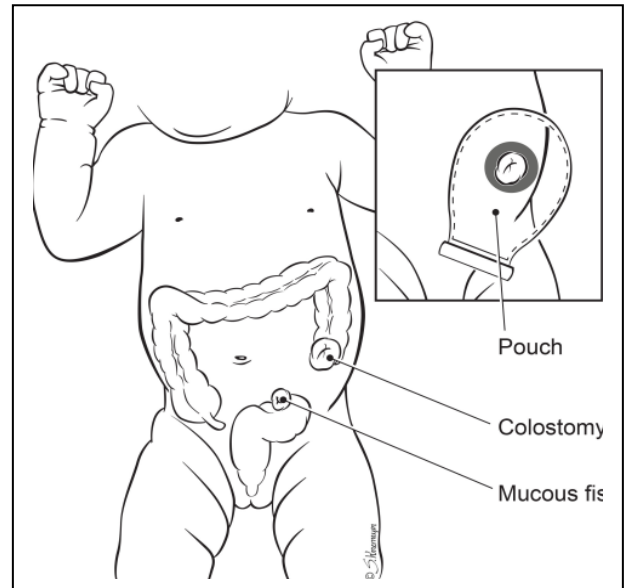
Picture 6 A side view of a PSARP repair with stitches (sutures).



Picture 7 A close-up view of the PSARP repair from below.

Long-term care

Children born with an anorectal malformation need long-term care. They typically need help to manage their bowel movements. This may mean taking part in a week-long bowel management program. It is also very important to watch for problems with the urological and gynecological system.



Picture 8 A close-up view of the PSARP repair from below.