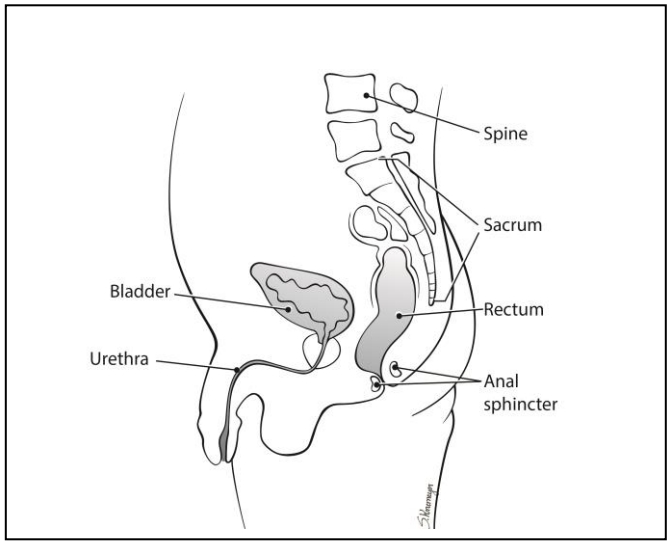
Anorectal Malformation (ARM) or Imperforate Anus: Male

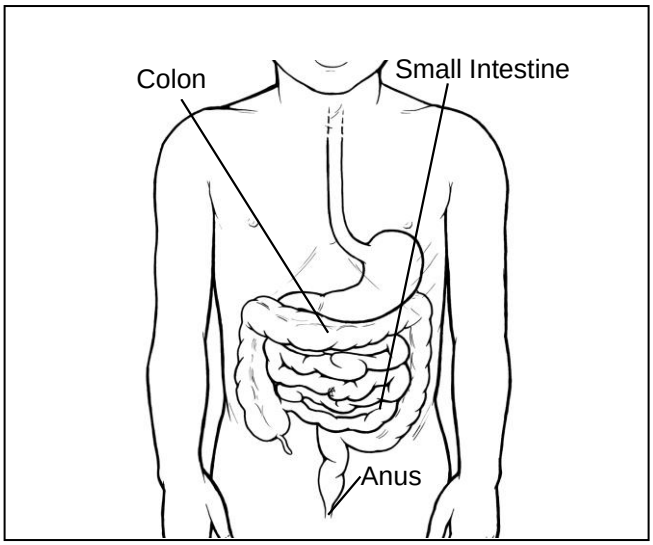
Anorectal malformation (ARM), also called imperforate anus, is a condition where a baby is born with an abnormality of the anal opening. This happens while the baby is growing during pregnancy. The cause is unknown. These abnormalities can keep a baby from having normal bowel movements. It happens in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

In a baby with an anorectal malformation, any of the following can happen:

- 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 the urethra (urinary system)



Picture 1 Normal organs and structures from the side.



Picture 2 Normal organs and structures from the front.

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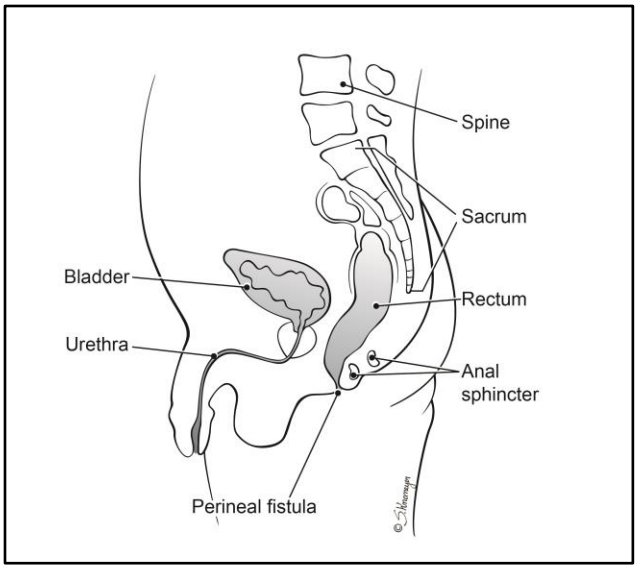
Signs and symptoms

At birth, your child will have an exam to check the position and presence of his anal opening. Newborn babies pass their first stool within 48 hours of birth, so certain defects can be found quickly.

Other signs of an anorectal malformation may include:

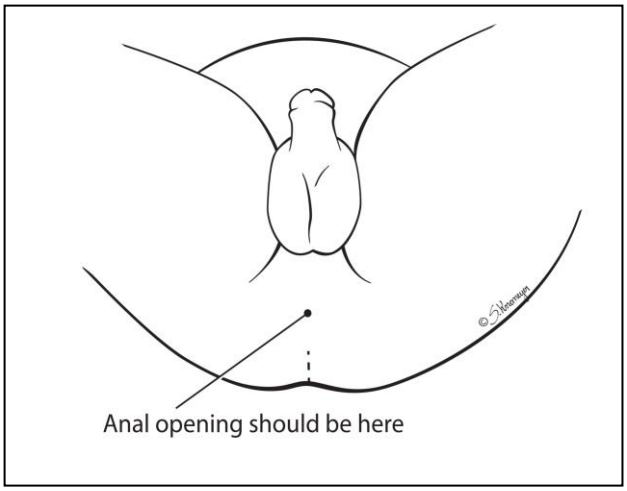
- Belly swelling
- Vomiting
- No stool within the first 48 hours
- Stool coming out of the urethra



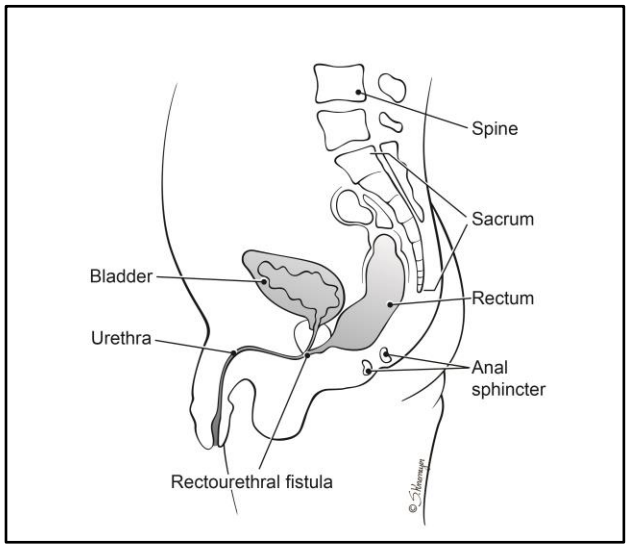
Picture 3 Perineal fistula, view from the side

Types of anorectal malformations

- **Perineal fistula** – the anal opening is in the wrong place (Picture 3). The anal opening opens into the perineum, which is the place between the scrotum and anal sphincter. It may also be too small.
- **Rectourethral fistula** – the anal opening is in the wrong place. The anal opening opens into the urinary tract (Pictures 4 & 5). There is no visible anal opening on the skin. The opening is classified by name based on where it opens into the urethra:
  - **Bulbar** – rectum opens into the lower portion of the urethra
  - **Prostatic** – rectum opens into the middle portion of the urethra
  - **Bladder neck** – rectum opens into the upper portion of the urethra near the bladder
- **No fistula** – the anal opening is too narrow or ends before it reaches the skin. The anal opening may look very small or you may not be able to see it at all.



Picture 4 Rectourethral fistula at birth, view from the bottom



Picture 5 Rectourethral fistula, view from the side

Other possible problems

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

- **Urology** – problems with the kidneys and other organs that help urination (peeing). These problems can cause infections and/or permanent kidney damage if left untreated. 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
- **Spine** – 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
- **Sacrum** – 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 portion of the sacrum does not fully develop
  - Presacral mass – a mass between the rectum and the sacrum

Other tests

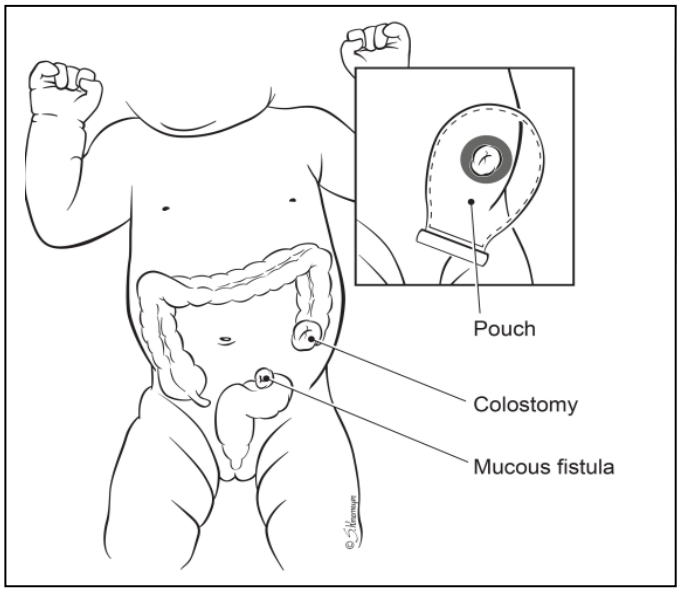
The doctor may order tests to better understand your child's malformation. These tests will also help the doctor know if there are any possible problems related to the diagnosis.

- **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. It also looks to see if the spine formed 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 his spine assessed.
- **Echocardiogram** – This test checks to see if there are any problems with your child's heart.

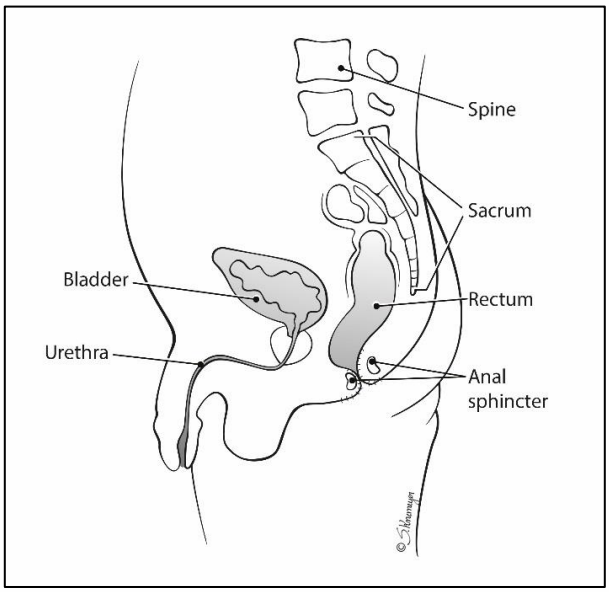
- **Labs (renal function panel, cystatin C)** – These labs tell the doctor more about your child's kidney function.
- **Video Urodynamics** – This test shows how your child's urinary tract works, including the bladder and kidneys.

Treatment and surgery

The treatment for anorectal malformations is surgery. The repair for ARM is called a posterior sagittal anorectoplasty (PSARP) (Pictures 7 & 8, next page). This surgery will either create your child's anal opening or put it in the right place so he can have bowel movements. The type of surgery and how many surgeries your child has depends on the type of ARM and previous surgical history. If your child does not have a fistula or has a rectourethral fistula, he will first need a colostomy (Picture 6), then a PSARP repair. If your child is born with a perineal malformation, then a colostomy is not commonly needed.



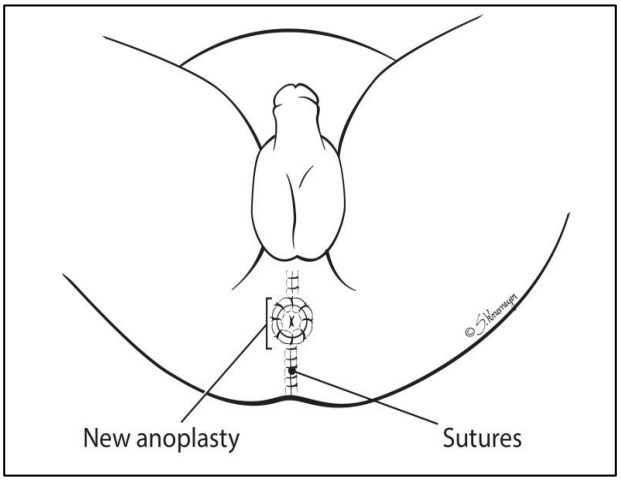
Picture 6 A child with a colostomy, colostomy pouch, and a mucous fistula.



Picture 7 A side view of a PSARP repair with stitches (sutures).

Long-term care

Children born with an ARM 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 system.



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