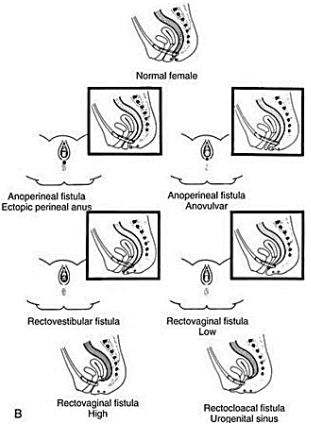
Indian Association of Pediatric Surgeons

Patient Information Sheet

FEMALE ANORECTAL MALFORMATIONS



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for & on behalf of the Indian Association of Pediatric Surgeons

What is female anorectal malformation ?

In girls anorectal malformation (ARM) is a congenital defect in which the anus and rectum (the passage from where stools are passed) is not properly formed in the baby. Either the passage is not formed at all or the stools pass through an abnormal passage into the urinary system, vagina or through an abnormal opening in the skin surrounding the genitalia.

What causes this problem and how common is it ?

During fetal development in the mother's womb or uterus, different organ systems are developing and maturing. The lower end of the intestinal and genitourinary tracts are joined together and separate in early pregnancy. When they do not separate then ARM results. The exact cause is not known. Some girls have multiple associated anomalies of other organ systems. They occur in ~ 1 in 5000 live births and are slightly less common than in boys (1:1.2).

What are the symptoms ?

There are several ways in which ARM can present in girls. The main ones are :

1. Normally there are separate openings for the urethra, vagina and anus. In ARM some of these openings may either be absent or may communicate with each other.
2. The stool and urine may come out from the same opening.
3. The baby may develop abdominal distension from non-passage of stool.
4. Some symptoms may become manifest when the girl grows up, particularly regarding discharge of menstrual fluid.

When to see your doctor ?

Most cases can be diagnosed at birth when the baby is examined by the doctors immediately after delivery. If not detected at birth then consult a pediatric surgeon if any abnormality is seen with the external appearance or for non-passage of stool or for passage of stool from any other orifice.

How is it diagnosed ?

- If the anal orifice is absent then x-rays in the new born period are done.

- If there are less than 3 orifices then x-rays with contrast material are done to establish which orifice is not developed or if there is a communication between any of the 3 tracts – urethral, genital and rectal.
- X-rays are also taken to determine any bony anomalies especially in the spine and sacrum. Ultrasound scan to look for any kidney malformation.
- ECHO cardiography to evaluate the heart.
- Any other associated anomaly may require specific investigations.

What are the treatments available ?

- A tight anal orifice can be treated with just dilatation.
- Absent or abnormally placed anal orifice require surgical treatment.
- Communicating tracts often require staged procedure or multiple surgeries.
- Some associated anomalies require specific surgical procedures.

Are there any alternatives to surgery ?

Almost all ARM in girls require surgical treatment. Even though stools maybe passed normally through an anal orifice in close proximity to the vagina, it may need to be shifted to its normal position to avoid infections of the vagina or serious injury during child birth.

What does the operation involve ?

The primary objective is to rectify the basic defect: -

- Absent orifices need to be created
- Abnormally placed orifices need to be relocated
- Communicating tracts have to be separated
- Absent vagina needs to be created

All the above objectives require complex operative procedures either through a cut between the buttocks (posterior sagittal ano-rectoplasty – PSARP or posterior sagittal ano-recto-urethro-vaginoplasty-PSARVUP) or this approach combined with part of the operation done through the abdomen (abdomino-perineal or AP-PSARP or AP-PSARVUP). The aim is to bring the rectum down to its

normal position through the muscles which give control to the passage of stool. Some surgeons would like to do the operation in one stage either by the open or the laparoscopic methods. However, for very complicated reconstructions most surgeons would prefer to do a colostomy as a preliminary procedure. A colostomy is a part of the large intestine which is brought out on to the abdomen so that stools can passed through it. This allows the distal part of the intestine to be cleaned thoroughly so as to minimise the incidence of infection in the major procedure. The appropriate reconstruction is then done. The colostomy is then closed by an operation so that the child can then pass stools from the newly created anus.

In girls in whom the initial assessment reveals that they are unlikely to develop acceptable control on their defecation a decision can be taken to provide a permanent colostomy and allow the stools to be collected in a changeable bag which is worn over the colostomy.

Some procedures eg reconstruction of the vagina may need a part of the intestines.

What are the possible complications / what happens after the operation ?

Postoperatively, the girl will have a few tubes placed in different parts for specific reasons and will receive antibiotics and pain relief medicines. If a colostomy is present then oral feeds can be started in a day or two. Feeding maybe delayed if there is no colostomy.

Approximately 3 weeks after surgery, dilatation of the newly created anus is started in a gradually increasing manner so that it does not shrink.

If a colostomy has been done before then it needs to be closed by another operation after 3-6 months.

What is the outlook or future of these children ?

These babies may develop constipation or fecal incontinence. These problems can be controlled if treatment is started early. Girls with extreme ARM eg cloaca often have difficulty in passing urine also. If the vagina is malformed then girls may have difficulty in coitus and in vaginal delivery.