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Anorectal Malformation (ARM): Female

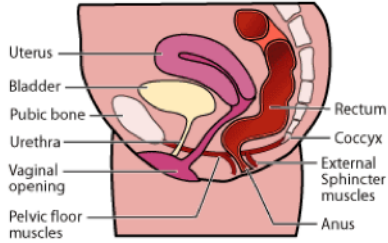
An anorectal malformation (ARM) is a disorder affecting the anus and the rectum, the last part of the digestive system. This page explains about ARMs and how they can be treated and what to expect when a child comes to Royal Belfast Hospital for Sick Children (RBHSC)

After food has been digested it passes through the small bowel into the large bowel. The poo (faeces) is stored in the rectum until the muscles receive a message from the brain to empty the bowel. The faeces then pass out through the anus.

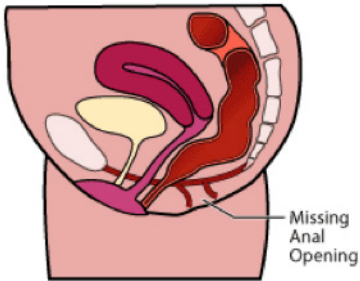
There are broadly types of ARM– low and high.

- A low ARM is where the anus is in a slightly different position or narrower than usual. There may also be a connecting passage to the skin called a fistula.
- A high ARM is where the bowel has a closed end at a high level and does not connect with the anus, or it may connect with another part of the body, usually the vagina, through a fistula.
- Girls can be affected by a complex and rare malformation where there is only one opening for the bladder, vagina and bowel. This is called 'cloaca' or 'cloacal malformation' and requires more specialist treatment than covered in this information.

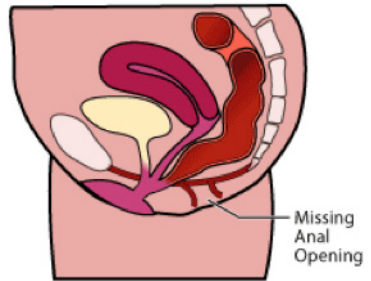
Normal Anatomy Female



Rectum connects to vagina



Cloaca: all three form a single opening



1.

Your consultant will discuss which of these problems your child has and how best to treat them.

What are the symptoms of an ARM?

ARMs in girls are rarely picked up on scanning before birth. Symptoms after birth may vary according to the type of ARM. The child will not be able to pass meconium – the black/ green faeces passed in the first few days of life – in the usual way. This can cause a swollen abdomen and vomiting. Sometime in girls these are picked up later. Usually ARMs need to be diagnosed quickly as the baby can become very unwell in a few days.

How is an ARM diagnosed?

An ARM is usually diagnosed soon after birth, on examination, when the baby fails to pass meconium at all, or the meconium comes from a different place. Health care professionals will examine your baby and note the absence of an anal opening. In your daughter this maybe the presence of one or two opening instead of the usual three. They may detect the presence of a fistula or passage of meconium in urine.

The doctor may suggest scans to give a clearer picture of the type of ARM and whether there is a fistula. These could include X-rays, ultrasound scans and rarely, MRI scans.

An ARM can be associated with other problems particularly the VACTERL association; a syndrome in which there can abnormalities of the spine (vertebrae), anus, heart (cardiac system), wind-pipe (trachea), food pipe (oesophagus), waterworks (renal system) and limbs. We will look for these during our investigations.

What causes an ARM and how common is it?

An ARM happens when the bowel does not form properly while the baby is developing in the womb. We do not know exactly what causes ARMs, but it is not due to anything a mother did or ate during pregnancy. Certain steps take place whilst your baby is in the womb for the rectum and anus to separate from the bladder and pee-pipe.

What treatments are available and are there any alternatives?

The treatment depends on the type of anorectal anomaly. Most types of ARM will need an operation under general anaesthetic. Your consultant will discuss the options in detail with you. In some cases only one operation (anoplasty) or an anal transposition will be needed, but most children may need a series of operations.

If your child has a cloaca abnormality the surgical team will explain the treatment plan which is different to other types of ARMs. The first operation is to create a temporary stoma (colostomy) usually in the days after birth. Once recovered, the baby can then go home for a few months.

The second is a posterior sagittal anorectoplasty (PSARP) operation to join the bowel to a newly created anus. This describes the operation and the approach taken by the surgeons. This usually happens when the child is a few months old and has gained weight. Rarely, a PSARP can be done without a stoma. In most cases, the surgeon operates on the child's bottom, but occasionally a wound in the tummy might be needed.

The final stage is to close the stoma. This happens when the child's bowel and anus are working well a few months after the second operation. The three operations are usually completed by the time the child is six to nine months old.

There are no alternatives to these operations as the child needs to be able to pass faeces to prevent it building up in the bowel. The only exception to this would be a baby with lots of other health problems, where sometimes the colostomy is the only operation needed.

What happens before the operation?

Whichever operation is planned, you will receive information about how to prepare your child for the operation. The surgeon will explain the operation in detail, discuss any worries you may have and ask your permission for the operation by asking you to sign a consent form. An anaesthetist will also visit you to explain about anaesthetic in more detail. If your child has any medical problems, like allergies, please tell the doctors.

As your child cannot drink they will need a drip for intravenous fluids for a while before the operation. They will also need a nasogastric tube which will stop your child being sick.

Are there any risks?

Your surgeon will go through the risks of each type of operation with you in detail before the operation. Every anaesthetic carries a risk of complications but this is very small. The anaesthetist is an experienced doctor who is trained to deal with any complications.

What happens afterwards?

Stoma: You will be contacted by the stoma nurse specialist and trained to look after your child's stoma before you go home.

PSARP: If your child has had a new anus created you may be asked to gently stretch (dilate) it using an instrument called a dilator. All new anuses require dilatation. Your surgeon or stoma nurse specialist will show you how to do this 2 weeks after the PSARP is done. You will need to start with a small size dilator and gradually increase the size until the anus is the right size.

You will be able to go home once the child is feeding well and starting to gain weight. We will contact your health visitor and family doctor (GP) to tell them about the operation.

When you get home

The stitches used during the operation will dissolve on their own so there is no need to have them removed. If possible, keep the operation site clean and dry to let it heal properly. The surgeon will let you know when the child can have a bath, but do not soak the area until the operation site has settled down.

After all the operations, the child may have diarrhoea or constipation. This is an after-effect of the operation and we will discuss this with you at the follow up appointment. Occasionally some children need medicines to help with any bowel problems. Whenever possible, leave the child's bottom open to the air as this will help it heal too.

When the stoma has been closed and your child is in nappies, use lots of nappy cream to prevent nappy rash. Almost all babies develop nappy rash at some point after this operation, usually as a reaction to the bowel starting to work. You will need to come back to hospital for an outpatient appointment after the operation which will be arranged before you go home.

You should call your family doctor (GP) or the ward if the child:

- Has serious abdominal pain and/or diarrhoea
- Has serious constipation

Seek urgent medical help if the child:

- Vomits when feeding
- Has a swollen stomach
- Vomits green liquid
- Is not gaining weight

What is the outlook for children with anorectal anomaly?

The outlook for children with ARM depends on the type of abnormality. Almost all children need close follow up and bowel management once they are toilet trained. Many could need support in the form of laxatives, suppository, enema or washouts. However, they should be able to go to school and work, and have a social life, just like anyone else.

Girls who have had an anorectal anomaly repair may be better having a caesarean section rather than vaginal childbirth. This will put less strain on the operation site and is less likely to cause problems in the future. Your daughter should discuss her previous medical history with her doctor when planning a pregnancy.



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1. Anorectal Malformation

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<https://surgery.ucsf.edu/conditions--procedures/anorectal-malformation.aspx>

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