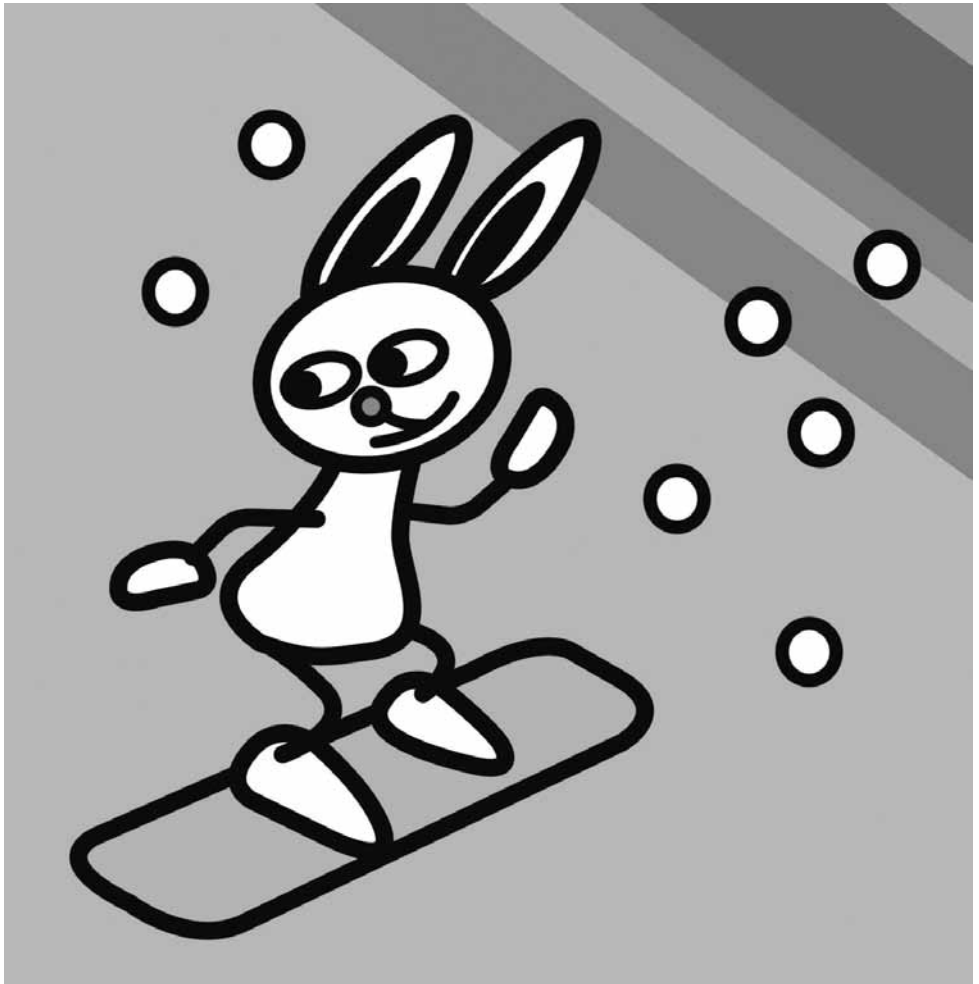


The Children's Hospital

Anorectal malformation (ARM) in boys

Information for parents



What is an Anorectal Malformation?

An anorectal malformation (ARM) is a congenital (it is present at birth) disorder in which the bottom does not develop normally. In ARMs the anus (bottom hole) does not open in the normal position. In addition the urinary tract (urethra (wee tube), bladder and kidneys) is affected most of the time.

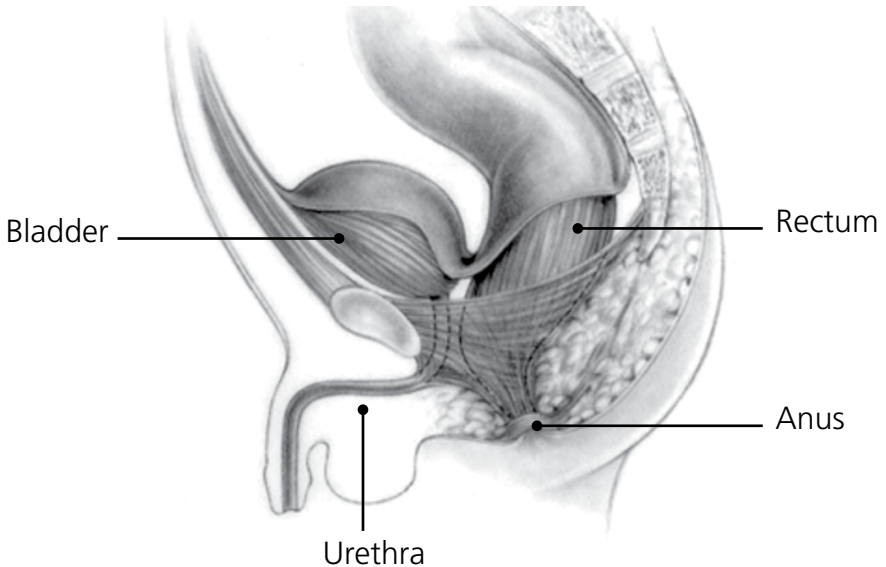
There are different types of ARMs, which are of different severity and might require different treatment. They are classified according to the place where the bowel ends and whether there is an abnormal connection between the bowel and the urinary tract. This is the case in most boys. The abnormal connection is called a **fistula**.

Normal development

Normally, two separate tubes develop when a baby boy is growing in the womb. Both tubes should have an opening to the outside world. The tubes, openings and functions are as follows:

- rectum (lower bowel) and anus, which allow the passage of stool (poo) out of the body
- urethra, which allows the passage of urine (wee) out of the body

The region where these openings are placed is called the perineum. It extends from bone at the front to bone at the back.



This diagram shows the baby's perineum, from front to back.

How is an Anorectal Malformation detected?

It is not usually possible to detect ARMs on antenatal scans so you may first be told that something is wrong at the postnatal check. Most ARMs in boys are obvious: when you look at the baby's bottom there is no anus or your baby passes meconium (black poo) from his urethra. However, if the anus is present, but slightly further forward than usual this subtle abnormality may be missed until a child has problems with constipation.

The point where the bowel joins the urinary tract (fistula) is usually not obvious by looking at your baby and can only be determined with a special X-ray. This X-ray (micturating cystourethrogram (MCUG) and loopogram) is usually done when your baby is six weeks to three months old. The X-ray involves passing X-ray dye (contrast) into the bowel and urinary tract through catheters and this allows us to work out where the two meet.

Different types of ARM

1. Rectoperineal fistula

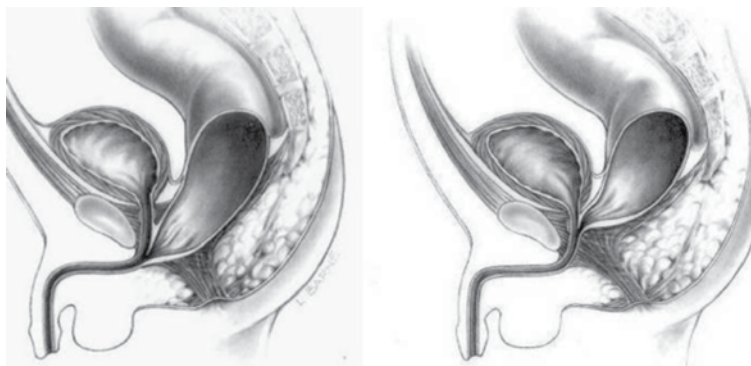
In this type of ARM the anus is slightly closer to the scrotum (further forwards) than it should be (sometimes called an **anterior anus** or **anterior ectopic anus**). This may be obvious at birth or it may only be diagnosed when the baby is older and develops constipation (difficulty passing stool). This is a mild type of ARM. Sometimes the anus is covered with a thin layer of skin.

This diagram shows a normal urethra. The anus is closer to the scrotum and urethra than it should be. The opening is in front of the muscles that control continence.



2. Rectourethral fistula

This is the most common ARM in boys. The rectum ends in a narrow tube (fistula) that joins the urethra, rather than ending at his anus on his bottom. It is usually obvious that the anus is not present (often called an imperforate anus).



This diagram shows the rectum joining the urethra. This join can be at different levels. The anus is not present.

3. Rectovesical fistula

This is a more severe form of ARM. The rectum ends in a narrow tube (fistula) that joins the bladder (organ where urine is stored) rather than ending at his anus on his bottom. It is usually obvious that the anus is not present (often called an imperforate anus).

This diagram shows the rectum joining the bladder. The anus is not present.



4. Rare ARM in boys

If the anus or rectum does not form normally, there is usually an abnormal connection (fistula) between the bowel and urinary tract. Rarely there is no such fistula. This can only be determined by MCUG. This type of ARM is sometimes called anal agenesis or rectal atresia.

What causes Anorectal Malformations and how common are they?

The cause of ARMs is not known. As far as we know, there is nothing that you could have done to cause or prevent your baby from having an ARM. ARMs are more common in babies with chromosomal abnormalities (genetic / inherited conditions). About one in 4,000 - 5,000 babies has an anorectal malformation.

Will my baby have any other problems?

ARMs can be associated with other anomalies. The most common are anomalies of the urinary tract, genital tract and heart. A well-known combination of anomalies is called **VACTERL association**. Each letter stands for an anomaly, but not all of these have to be present to diagnose **VACTERL**.

V – vertebra (spine)

A – anus

C – cardiac (heart)

T – trachea (windpipe)

E – esophagus (food pipe)

R – renal (kidneys)

L – limb (arm or leg)

Will any investigations be necessary?

As ARMs can be associated with other anomalies your baby will need investigations to find out if this is the case. The most common investigations are an ultrasound scan of the kidneys, spine and heart called an ECHO (echocardiogram). Later, a contrast study of the urinary tract (MCUG) and lower bowel (loopogram) will be performed. Other investigations may be required and your doctor will explain these to you.

How are ARMs treated?

Because of the abnormal connection between the bowel and the urethra or bladder there is a risk of urinary tract infection (UTI). To minimise the risk of urinary tract infection your baby will be started on prophylactic antibiotics. Until the contrast study is done, it is not possible to tell the difference between a rectourethral and a rectovesical fistula type of ARM.

Most babies with an ARM need three operations. The first operation happens in the first days of life and allows the baby to

pass stool. This is a life saving operation. This operation brings the bowel to the surface of your baby's tummy. The artificial opening is called a stoma. Stool passes directly from the bowel onto the skin of the lower tummy. A bag is stuck to the skin to protect the skin and catch stool; this is called a stoma bag. Most parents quickly learn how to look after their baby's stoma.

The second operation reconstructs the tubes in the correct positions. This operation is called an anorectoplasty, and usually happens at about three months of age. The anorectoplasty can be a long delicate operation as it involves placing the rectum (lower bowel that stores stool) within the muscles (sphincters) that will help to keep your child continent and create an anus in the normal position.

After the second operation it is important to make sure that the new anus does not narrow while it is healing. It will therefore need dilatations (stretching). This starts seven - ten days after the operation and needs to be done every day. A metal rod is inserted into the anus for about 30 seconds. Your doctor will determine which size dilator is right for your baby and show you how to do the dilatations yourself at home. Increasing sizes of dilators are used until your baby's anus is a normal size.

The third operation happens when the reconstruction has healed and the anus is a normal size. This is usually about three months after the anorectoplasty. The operation involves assessing the reconstructed anus. If the new anus is big enough to allow stool to pass, the stoma will be closed so that the baby will be able to pass stool out of his bottom and not into a bag.

Babies with a rectoperineal fistula may only require one operation. The surgeon treating your baby will explain if this is the case.

What are the risks?

All operations have a small risk of bleeding and infection.

The operation to form a stoma carries the risk that the stoma may become too tight (stenotic) to allow stool to pass. The stoma may also prolapse (fall out of place) so that a length of bowel that should be on the inside slides outside. These problems are rarely severe enough to require another operation to treat them.

The main risk of the anorectoplasty is narrowing of the new anus (this problem is called anal stenosis). Unusual, but significant risks of anorectoplasty are that the original opening re-opens (this is called re-fistulation), so that there are two openings. Rarely the reconstruction can break down (this is called a dehiscence). Re-fistulation and dehiscence are generally treated with a second anorectoplasty.

After the third operation, closing the stoma, there is a very small chance that the join may leak. Leaks happen in the first few days after the operation and will require another operation to treat them. The join can become too narrow (stenotic) and this may require another operation. Both these complications are rare.

What happens before the operation?

Whichever operation is planned, you will receive information on how to prepare your child for the operation in your admission letter and our welcome booklet.

Your child's surgeon will explain the operation in more detail and discuss with you any concerns you may have. The surgeon will ask you to sign the consent form giving your permission for the operation to go ahead.

An anaesthetist will also visit you to explain about your child's anaesthetic in more detail and discuss options for pain relief afterwards. If your child has any medical problems, like allergies,

please tell the doctors. Please also bring in any medicines your child is currently taking.

If you are at home before the operation you and your child might need to come into hospital one day before the operation. This is so your child's bowel can be empty of stool ready for the operation. Your child may have laxatives or a bowel washout.

In the anaesthetic room

A nurse and parent can accompany your child to the anaesthetic room. Your child may take a toy.

It may be possible to give the anaesthetic with your child sitting on your lap. Your child may either have anaesthetic gas to breathe or an injection through a cannula (a thin plastic tube that is placed under the skin, usually on the back of the hand).

If the anaesthetic is given by gas, it will take a little while for your child to be anaesthetised. He may become restless as the gases take effect.

If an injection is used, your child will normally become unconscious very quickly indeed. Some parents may find this frightening.

Your child will then be taken into the operating theatre to have the operation or investigation.

Anaesthetic risks

In modern anaesthesia, serious problems are uncommon. Risk cannot be removed completely, but modern equipment, training and drugs have made general anaesthesia a much safer procedure in recent years.

Most children recover quickly and are soon back to normal after their operation and anaesthetic. Some children may suffer side effects like sickness or a sore throat. These usually last only a short time and there are medicines available to treat them if necessary.

The exact likelihood of complications depends on your child's medical condition and on the nature of the surgery and anaesthesia your child needs. The anaesthetist can talk to you about this in detail before the operation.

What happens afterwards?

You will be able to rejoin your child in the recovery room as soon as he is awake; your child will then come back to the ward to recover.

We will keep your child as comfortable as possible by giving regular pain relieving medications. For the first few days the pain relief will usually be given through a 'drip' and then, when your child is more comfortable, in the form of medicines to be swallowed.

For the first few days, your child will need a drip of fluids until he feels like eating and drinking again. This will also allow the bowel to rest and start to heal. You will be able to go home once your child is comfortable and the bowel working.

What is the outlook for children with ARMs?

Babies and children with ARMs will need long term follow up in a surgical clinic. A few children with mild ARMs have an entirely normal bowel function and do not become constipated and are continent without medicines or surgery. Most children require medicines or washouts to improve constipation and/or incontinence. Some children require additional surgery to help with bowel management or for problems with their urinary tract. Very rarely patients are managed with a long term stoma.

Useful information can be found on the following websites:

www.vacterl-association.org.uk (a UK support group)

www.pullthrunetwork.org (an American based website)

Further information

If you have any questions, or there is anything you do not understand, please ask one of the doctors or nurses.

Your child will be followed up regularly in the outpatient clinic, but if you have any problems while at home you can either contact your GP or you can phone the ward for advice on:

Tom's Ward: 01865 234108 or 234109

Reference

The Royal College of Anaesthetists (RCoA) (2003) Your child's general anaesthetic: information for parents and guardians of children London: RCoA www.rcoa.ac.uk

If you need an interpreter or need a document in another language, large print, Braille or audio version, please call **01865 221473** or email **PALSJR@ouh.nhs.uk**

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