Anorectal Malformations (ARM) in Girls
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 and genital tracts (wee tube and vagina) can also be affected.

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. If the anus is not in the normal position it is called a “fistula”.

Normal development

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

- rectum (lower bowel) and anus, which allow the passage of stool
- urethra, which allows the passage of urine, and
- vagina, which allows sexual intercourse and childbirth in later life.

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 three tubes in the middle of the baby’s perineum, from front to back. The first tube is the urethra, the second the vagina, and the anus is at the 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. Some ARMs are obvious: when you look at the baby’s bottom there is no bottom hole. However, if the bottom hole is there but slightly further forward than usual this may be missed for several months or until a child has problems with constipation.
Different types of ARM

1. **Rectoperineal fistula**
   In this type of ARM the anus is slightly closer to the vagina (further forwards) than it should be – this is 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.

   
   ![Diagram showing a normal urethra and vagina. The anus is closer to the vagina (further forward) than it should be. The opening is in front of the muscles that control continence.](image)

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

2. **Rectovestibular fistula**
   This is the most common ARM in girls. A rectovestibular fistula is usually diagnosed when the anus is not in the normal position or when stool is seen coming out of an area next to the vagina. Usually a rectovestibular fistula is diagnosed when the “baby check” is done, but occasionally it is not diagnosed until the baby is several months old. This is a moderately severe type of ARM.
This diagram shows a normal bladder and urethra (wee tube) at the front. The bottom hole is not present. Instead a fistula (narrow track) passes from the rectum to the skin next to the vaginal opening.

3. **Rectovaginal fistula**
   In this type of ARM the bowel opens directly into the vagina. This type of ARM is very rare.
4. Cloaca

The most severe ARM occurs when all three tubes are joined together. This disorder is called a “cloaca” and is rare. The length of the joined channel varies and the treatment is different according to the anatomy of your baby’s malformation.

This diagram shows urethra, vagina and anus all joined in one common channel. There is only one opening on the perineum.

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 4000 - 5000 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”.

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\begin{align*}
V & \quad \text{vertebra (spine)} \\
A & \quad \text{anus} \\
C & \quad \text{cardiac (heart)} \\
T & \quad \text{trachea (windpipe)} \\
E & \quad \text{esophagus (foodpipe)} \\
R & \quad \text{renal (kidneys)} \\
L & \quad \text{limb (arm or leg)}
\end{align*}
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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 ECHO = echocardiogram). Later, a contrast study of the urinary tract (dye is pushed into the bladder through a catheter while x-rays are taken) may be performed. Other investigations may be required and your doctor will explain these to you.

How are ARMs treated?

Most babies with an ARM need three operations. The first operation usually happens in the first days of life and allows the baby to pass stool. The ARM causes a partial or complete blockage to the passage of stool. The first operation creates an artificial opening for stool to pass to the outside world. This 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 3 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 7 - 10 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 3 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 her 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.

In babies with a cloaca different operations are usually necessary. The surgeon treating your baby will explain these in detail to you.

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 faeces 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. They 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 she 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 or she 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 to improve constipation and/or incontinence. Some children require additional surgery to help with bowel management. Very rarely patients are managed with a long term stoma.

Useful information can be found:
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 don’t 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

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@orh.nhs.uk

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