What is an anorectal anomaly?

This is a disorder affecting the anus and the rectum, the last part of the digestive system. After food has been digested it passes through the small bowel into the large bowel. The faeces (poo) 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 two types of anorectal anomaly – low anorectal anomaly and high anorectal anomaly.

- A **low anorectal anomaly** is where the anus is closed over, in a slightly different position or narrower than usual. There may also be a connecting passage to the skin called a fistula.

- A **high anorectal anomaly** 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 bladder, urethra or vagina, through a fistula.

Girls can be affected by a very complex malformation where there is only one opening for the bladder, vagina and bowel. This is called ‘cloacal malformation’ and requires more specialist treatment – see our information sheet available online at www.gosh.nhs.uk/medical-information-0/search-medical-conditions/cloacal-malformation for details.

What are the symptoms of an anorectal anomaly?

Symptoms may vary according to the type of anorectal anomaly. Your child will not be able to pass meconium – the dark faeces passed in the first few days of life – in the usual way. This can cause a swollen abdomen and vomiting. If your child has an
anorectal anomaly with a fistula, the faeces will be able to partially pass out of the body, near the vagina in girls or through the urethra in boys. As the faeces can only be partially removed from the body, this will not relieve the pressure in the abdomen, causing swelling and vomiting. Anorectal anomalies need to be diagnosed quickly as your baby can become very unwell in a short period of time.

How is an anorectal anomaly diagnosed?
An anorectal anomaly 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 use the passing of meconium as part of the standard newborn checking procedure. The doctor may suggest scans to give a clearer picture of the type of anorectal anomaly and whether there is a fistula. These could include x-rays, ultrasound scans or rarely, MRI scans.

What causes an anorectal anomaly and how common is it?
An anorectal anomaly happens when the bowel does not form properly while the baby is developing in the womb. We do not know exactly what caused this, but it was not due to anything that happened during pregnancy. Research has indicated some possible causes of anorectal anomaly but these need further investigation. An anorectal anomaly can be associated with other problems, but the doctor will examine your child closely to check if this is the case. About one in 3,000 babies are born with an anorectal anomaly.

What treatments are available and are there any alternatives?
The treatment depends on the type of anorectal anomaly. All types of anorectal anomaly will need an operation under general anaesthetic – in some cases, only one operation (anorectoplasty) will be needed but most children may need a series of operations.

The first operation is to create a loop stoma (an artificial way of disposing of waste matter) usually in the days after birth. The second is a posterior sagittal anorectoplasty (PSARP) operation to join the bowel to a newly created anus. This usually happens when your child is a few months old and has gained weight. The final stage is to close up the stoma. This happens when your child's bowel and anus is working well a few months after the second operation. The three operations are usually completed by the time your child is six to nine months old. In most cases, the surgeon operates on the child's bottom, but occasionally an incision in the abdomen might be needed.

There are no alternatives to these operations, as your child needs to be able to pass faeces to prevent it building up in the bowel.

What happens before the operation?
Whichever operation is planned, you will receive information about how to prepare your child for the operation in your admission letter and your welcome booklet. Your child's 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 your child's anaesthetic in more detail. If your child has any medical problems, like allergies, please tell the doctors. As your child cannot take oral fluids normally due to the blocked bowel, he or she will need a ‘drip’ of fluids for a while before the operation. Your child will also need a nasogastric tube, which is passed up the nose,
down the foodpipe and into the stomach. This will drain off the stomach and bowel contents and ‘vent’ any air, which has built up, which will make your child more comfortable.

**What does the operation involve?**

**Anoplasty** – the surgeon will open up the anus (if it was closed over), move the anus (if it was in a slightly different place to usual) or widen the anus (if it was narrower than usual).

**Creation of loop stoma** – the surgeon will bring the bowel to an artificial opening in your child’s abdomen (stoma).

**Posterior sagittal anorectoplasty operation (also called PSARP)** – the surgeon will create a new anus and then separate the bowel from the fistula (if there is one) and bring it down to join the newly created anus. He or she will then close up the fistula. If the fistula is very high, the surgeon may need to make an incision in your child’s abdomen to correct it. Your child will continue to use the stoma so that the bowel and anus can rest and heal.

**Closure of loop stoma** – the surgeon will disconnect the bowel from the stoma and close it to form a fully working bowel.

**Are there any risks?**

All surgery carries a small risk of bleeding during or after the operation. There is a very small chance that the bowel could leak into the abdominal cavity. This could cause an infection called peritonitis, but we will give your child antibiotics to treat this, should it happen. If your child has a fistula, the urethra or vagina could be damaged when the bowel is disconnected but this is very rare. There is also a chance the muscles used for bladder and bowel control could be damaged, but this is also very rare. In many cases, the muscles are already affected to some degree before the anomaly is repaired. Every anaesthetic carries a risk of complications, but this is very small. Your child’s anaesthetist is an experienced doctor who is trained to deal with any complications.

**What happens afterwards?**

Your child will 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. When the surgeon is happy that the area is healing, you can start to feed your child again, starting with small amounts, and increasing the amount as he or she tolerates it.

If your child has had a new anus created, in some cases, you may be asked to stretch...
(dilate) it using a probe called a dilator. In many cases, the surgeon will check the area and carry out the first dilatation while your child is under a short general anaesthetic. If further dilatation is needed, your surgeon or stoma nurse specialist will show you how to do this before you go home. You will need to start with a small size dilator and gradually increase the size until the anus is the right size. Most children do not require dilatation.

If your child has had the stoma created, the stoma nurse specialist will teach you how to look after it. We will make sure you feel confident before you go home.

You will be able to go home once your 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

Your child’s abdomen may feel sore for a while after the operation, but wearing loose clothes can help. 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 your child can have a bath, but do not soak the area until the operation site has settled down.

After all the operations, your 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. After the operation, your child may have a very sore bottom. It is not a good idea to sit your baby upright or do anything that puts pressure on the healing area for several weeks after the operation. Whenever possible, leave your child’s bottom exposed to the air as this will help it heal too.

When the stoma has been closed and your child is in nappies, use nappy cream liberally 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 about six weeks after each operation. We will write to you with the appointment date.

What is the outlook for children with anorectal anomaly?

The outlook for children with anorectal anomaly depends on the type of abnormality. Many children go on to lead normal lives, working and raising a family. However, there are a couple of side effects which you should know about, should they arise in the future.

Your child may need extra bowel training at a later stage to learn when and how to empty his or her bowels. He or she may also need some extra help in the form of enemas, washouts or medicines, but these will be explained to you if they are needed. However, some children continue to have bowel problems for many years after the operations, but this depends on the original severity of the anorectal anomaly. If your child does not have full bowel control by the age of three years, please discuss this with the team at your next clinic appointment.

Your child will need regular check ups with your doctor at GOSH until he or she is well. 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.
You should call your family doctor (GP) or the ward if your child:
- has serious abdominal pain and/or diarrhoea
- has serious constipation

Seek urgent medical help if your child:
- vomits when feeding
- has a swollen stomach
- vomits green liquid
- is not gaining weight

Is there a support organisation?
There is no support group specifically for those with anorectal anomalies, but the following organisation may be able to help:

Contact a Family
Tel: 0808 808 3555
Email: info@cafamily.org.uk
Website: www.cafamily.org.uk

If you have any questions, please call Squirrel Ward on 020 7829 8818.