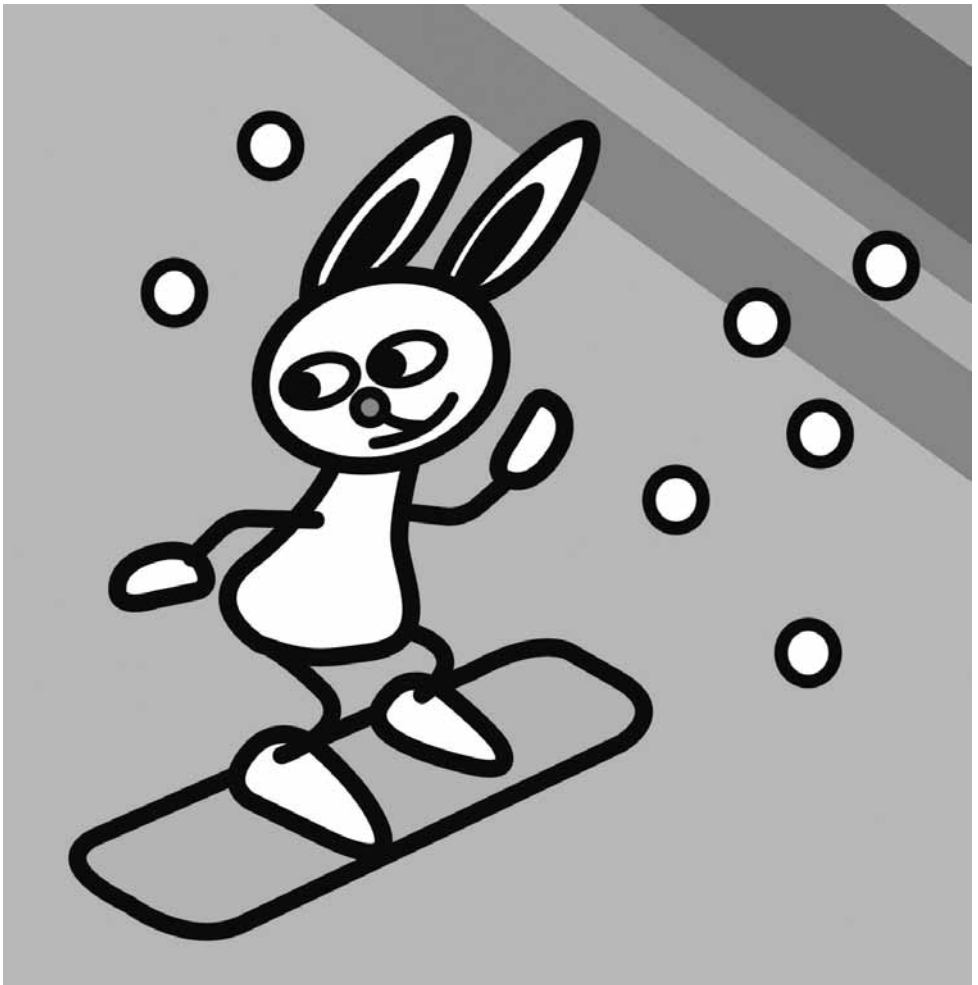


The Children's Hospital, Tom's Ward

Hirschsprung Disease

Information for parents

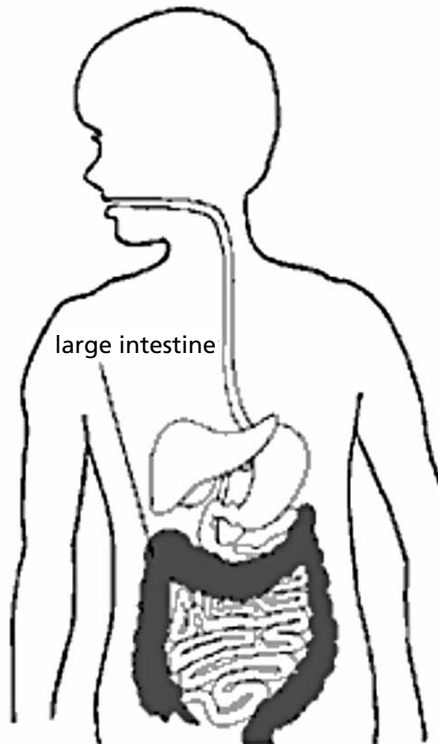


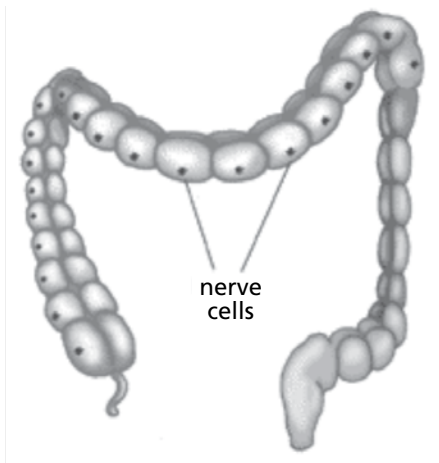
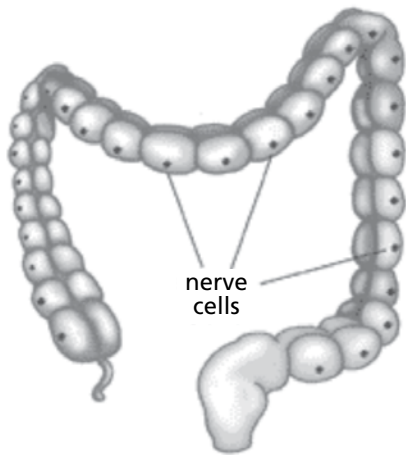
What is Hirschsprung disease?

Hirschsprung disease is a rare disorder of the bowels, most commonly the large bowel (colon).

Normally, the muscles in the bowel squeeze rhythmically to push faeces (poo) through to the rectum. In Hirschsprung disease, the nerves (ganglion cells) that control these muscles are missing from part of the bowel.

This means that faeces cannot be pushed through the bowel in the usual way. The length of the affected part of bowel varies from child to child.





What are the symptoms of Hirschsprung disease?

Often newborn babies with Hirschsprung disease do not pass meconium – the dark faeces passed in the first two days of life. Other symptoms include a swollen abdomen and vomiting green bile. Later on the main symptom of Hirschsprung disease is constipation, which cannot be treated using laxatives or softeners. This occurs because faeces are pushed through the bowel until they reach the affected part. As this part of the bowel cannot squeeze rhythmically to push the faeces through the bowel, the faeces cannot move any further. As more food is digested and turned into faeces, the bowel becomes blocked causing discomfort and a swollen abdomen.

How is Hirschsprung disease diagnosed?

It is diagnosed by taking a small piece of tissue from the bowel to examine under a microscope. This is called a rectal biopsy. If the piece of tissue does not have any ganglion cells, this means that Hirschsprung disease is confirmed.

What causes Hirschsprung disease and how common is it?

Hirschsprung disease is a congenital (present at birth) disorder. While your baby was developing in the womb, the nerve cells did not develop through the full length of the bowel. We do not know what caused this to happen, but as far as we know, it was not due to anything that happened during pregnancy.

Hirschsprung disease affects one in every 5,000 babies.

How is Hirschsprung disease treated and are there any alternatives?

How Hirschsprung disease is treated depends on the age at which your child is diagnosed and how well your child is generally.

Some children's constipation can be helped using bowel washouts, where a thin tube is inserted into your child's bottom and filled with a salt-water solution. This softens the faeces and flushes it from your child's bowel. If this is an option for your child we will teach you how to do bowel washouts before you go home.

All children will need an operation to treat Hirschsprung disease. If a child is newborn, the doctor may suggest the 'pull-through' operation. When a child is older, or for other reasons, the doctor may suggest creating an artificial opening (stoma) to remove faeces. This will allow your child's health to improve before the pull-through operation. The stoma is usually a temporary measure, which will be closed once your child has had the pull-through operation.

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.

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.

He or she may have laxatives or a bowel washout, and will be allowed to drink only clear fluids for 24 hours before the operation.

What does the operation involve?

Details of the operation will be provided by your surgeon, the following is a short summary.

- **Stoma**

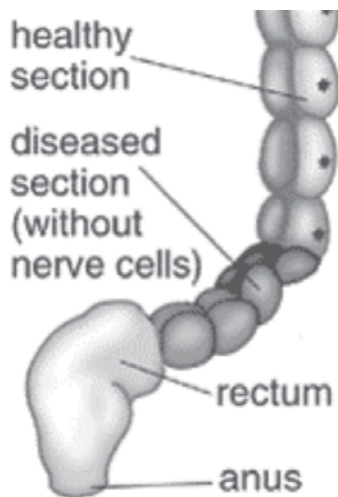
Biopsies are taken to see how much bowel is affected. During the operation the surgeon will bring the healthy end of the bowel to an artificial opening in your child's abdomen called a stoma. This means faeces can be pushed through the bowel to the stoma, where they are collected in a bag to be disposed of later.

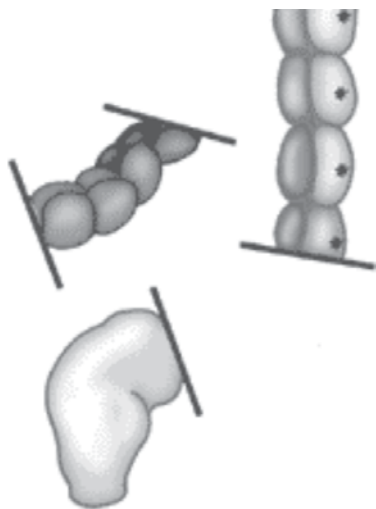
Some weeks later, in another operation, the surgeon will remove the part of the bowel missing the ganglion cells as in the 'pull-through' operation (see below). The stoma might be closed at the same time or in a third operation.

- **Pull-through operation**

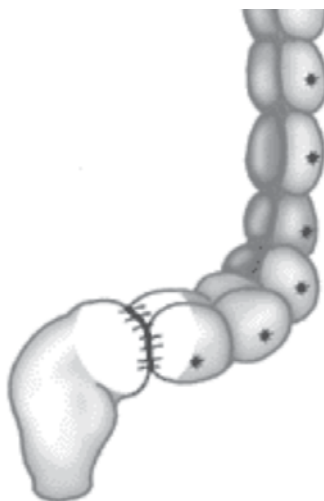
The surgeon will remove the part of the bowel missing the ganglion cells and attach the healthy end to the bottom. This creates a working bowel, with enough nerve cells to control the muscles so that your child can pass faeces as usual. If your child does not have a stoma, biopsies will be taken during this operation to see how much bowel is affected. The operation will partly be performed from the bottom and partly from the abdomen. This part will be performed by laparoscopy ("keyhole surgery") if possible.

*Before surgery:
The diseased section is
the part of the intestine
that doesn't work.*





Step 1:
The doctor removes the diseased section.



Step 2:
The healthy section is attached to the rectum or anus.

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. If this occurs, your child will need a stoma as this reduces the risk of developing a serious infection called peritonitis.

What happens afterwards?

You will be able to rejoin your child in the recovery room as soon as they are 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. One week from the operation a gentle anal stretch may be needed – we will advise you at the time of discharge.

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.

When you get home

After the pull-through operation, your child may have a very sore bottom when passing faeces normally. You can try using baby oil to clean your child's bottom, as this will also ease any discomfort. Whenever possible, leave your child's bottom open to the air as this will help it heal too.

We will write to your child's GP to tell them about your child's operation.

Are there any other complications?

Some children with Hirschsprung disease develop enterocolitis. The symptoms include fever, a swollen abdomen and diarrhoea, sometimes with very smelly stools. This is potentially a serious complication and needs to be treated in hospital with antibiotics, washouts of the bowel and fluids through a drip. Children who develop enterocolitis usually do so before the operation, but it can happen afterwards also.

What is the outlook for children with Hirschsprung disease?

The outlook for children with Hirschsprung disease is good, with the majority growing up to live normal lives, working and raising a family. They may however experience long-term problems with constipation or bowel control. Some children have ongoing problems and need more operations.

Is there a support group?

The Hirschsprung's & Motility Disorder Support Network

www.hirschsprungs.info

The Willows
Forest Road
Narborough
Leicestershire LE19 3LD

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**

We hope that this information is useful to you and would welcome any comments about the care or information you have received.

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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Fiona Mills, NP/SSN, Tom's ward
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The Hirschsprung's & Motility Disorders Support Network
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