

Hirschsprung's Disease

Neonatal surgery

Patient information

What is Hirschsprung's Disease?

Hirschsprung's Disease (HD) is a rare condition which affects the nerve cells of the bowel, causing a blockage of the bowel.

In HD, nerve cells (called ganglion cells) are missing from a section of the bowel. These cells normally control the muscles of the bowel, which push along the contents (poo). The contents move along the bowel until they reach the part where the cells are missing. This causes a blockage or the passage of poo to slow down.

HD affects the rectum and a variable length of the bowel above it. This length is usually a few centimetres but sometimes may affect the entire bowel.

There is no known cause for HD, although it can run in families.

What are the symptoms of Hirschsprung's Disease?

Some infants with Hirschsprung's Disease (HD) do not pass meconium-the dark poo normally passed in the first two days of life. Other symptoms include a distended (swollen) abdomen (tummy) and vomiting bile (green fluid).

The main symptom of Hirschsprung's 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 distended abdomen. Your baby may be uncomfortable because of constipation and trapped air in the bowel.

How is Hirschsprung's Disease diagnosed?

An x-ray of your baby's abdomen will show enlarged loops of bowel.

The only way to diagnose HD is by taking a small piece of tissue from the lining of the bowel. This is called a rectal biopsy. The sample is sent to the laboratory for examination under a microscope. The

laboratory will be looking for a type of cell called a ganglion cell. The results will be available in approximately five working days.

If the piece of tissue does not have any ganglion cells, this means Hirschsprung's disease has been diagnosed. Bowel washouts are continued while waiting for the results of the rectal biopsy. If HD is confirmed, they will be continued until your baby has an operation.

What is the initial treatment?

- Milk feeds will be stopped
- A tube will be passed through the nose into the stomach to remove any fluid and air collecting in it
- Fluids will be given through a vein (drip)

Your baby will need to have rectal washouts to decompress the bowel and reduce discomfort. Usually the washouts will need to be done once or twice every day and will take 20 to 30 minutes.

Bowel washouts empty the lower large bowel of air and stool and reduce the risk of enterocolitis developing. Occasionally the abdomen cannot be decompressed adequately by washouts. Your baby would then need an operation (colostomy formation) to ensure that the bowel remains decompressed.

How is Hirschsprung's Disease treated?

All babies will need an operation to treat Hirschsprung's Disease. This operation is called a 'pull through' and is usually performed at a few weeks/months of age. It involves taking out the part of the bowel affected by HD and connecting the healthy bowel to the anus. Your baby will be in hospital for five to seven days after the operation. Some surgeons may suggest creating an artificial opening on the abdominal wall (stoma) to remove faeces while waiting for the 'pull through' operation.

Your baby may need to remain in hospital while having bowel washouts and waiting for surgery. Sometimes your baby may go home and the nurses will teach you how to do the rectal washouts. During this time your baby should be able to feed normally.

If your baby is not able to feed normally for any reason, if you wish to breast feed, the nursing staff will teach you how to express and store your milk to feed your baby when they are ready.

While awaiting surgery your baby remains at risk of developing enterocolitis.

What is enterocolitis?

Enterocolitis is an infection of the large bowel and is a potentially very serious complication of HD. It is treated with antibiotics (given through a drip into a vein) and regular bowel washouts.

What are the symptoms of enterocolitis?

- Distended abdomen
- Reluctance to feed
- Vomiting - this may be green in colour

- Explosive / offensive watery stools
- A high temperature
- Tummy ache
- Dehydration (baby is pale and floppy)

We can provide you with a laminated medical alert card if you wish.

Enterocolitis can occur both before and after surgery but the risk after surgery is much less.

If you are at all concerned about your baby please seek medical advice urgently.

What does the operation involve?

Pull-through operation

The surgeon will remove the part of the bowel with the missing ganglion cells and bring the healthy bowel with ganglion cells down to the rectum. This creates a working bowel, with enough nerve cells to control the muscles so that your baby can pass faeces as usual.

What are the long-term effects of Hirschsprung's Disease?

Many babies with HD experience long-term problems such as diarrhoea, constipation, soiling and abdominal pain and you may find that your child takes longer to toilet train. Sometimes medicines are needed to help with these problems. You will have specialist advice and support to manage and improve any symptoms in the longer term.

Following discharge from hospital, your baby will be seen in the outpatient's department by members of the surgical team, where help and advice will be available to you. It may be possible for this follow up to take place in your local hospital. Follow up appointments will usually occur on a regular basis for some years.

If St George's Hospital is not your local hospital

Once your baby has had surgery and made a good recovery, i.e. when their specialist medical and nursing requirements are less, the baby will be transferred back to the care of your local hospital. This transfer is a sign of progress and will not occur until the baby is ready. It will allow you to be closer to home and become familiar with your local healthcare professionals.



May 2013

Review May 2016