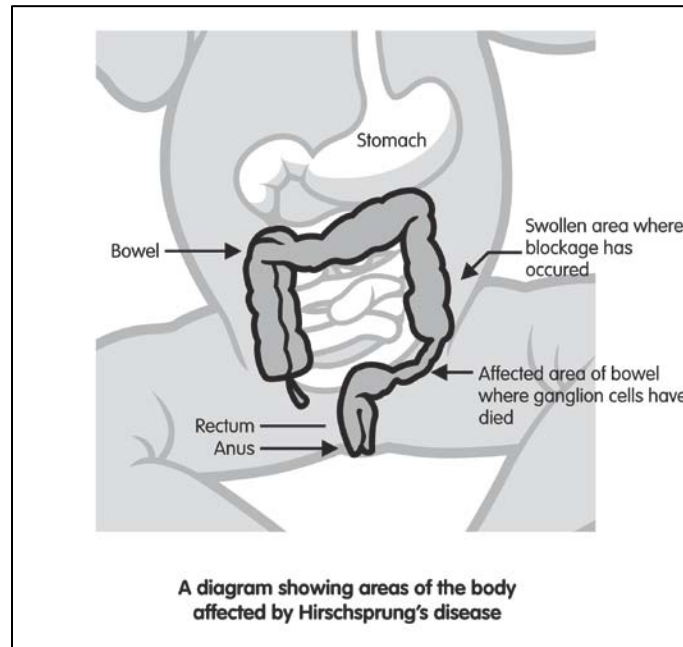


Hirschsprung's disease



Hirschsprung's disease (HD) is a rare condition which affects the nerve cells of the bowel causing severe constipation or blockage of the bowel.

In HD, nerve cells (called ganglion cells) are missing from the bowel. These cells control the muscles of the bowel, and these muscles push along the contents of the bowel. Contents are pushed along until they reach the affected part of bowel where the cells are missing, the passage of the contents slows and a blockage occurs.

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

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.

What are the symptoms of Hirschsprung's disease?

There is normally a delay in your baby passing meconium (black sticky stool normally passed within 24-hours of delivery).

Many babies with HD display symptoms at around three days of age, having never had their bowels open. Your baby may be reluctant to feed and may vomit green fluid, called bile, which would normally pass through the bowel.

Your baby may be uncomfortable because of constipation and trapped air in the bowel. The stomach may become swollen. Sometimes the bowels become inflamed and a very serious infection called enterocolitis can develop.

What is the initial treatment?

- Milk feeds will be stopped.
- A tube will be passed through the nose into the stomach to drain away any fluid and air collecting in the bowel.
- Fluids will be given through a drip inserted into a vein.
- Your baby will need to have regular bowel washouts to decompress the bowel and reduce discomfort. Usually the washouts will need to be done once or twice every day and will take 20-30 minutes.

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

How is Hirschsprung's disease diagnosed?

An x-ray of your baby's stomach will show swollen or enlarged bowels. The only way to test your baby for HD is by taking a sample of tissue from your baby's anus by inserting a lubricated instrument. This is called a 'rectal suction biopsy'. The sample is sent to the laboratory for examination.

Sometimes, following a rectal suction biopsy, bleeding occurs from the biopsy site. If this happens the blood will be passed through the anus into the nappy. Any bleeding is usually minimal and stops within 24-hours of the biopsy. Bowel washouts are continued while waiting for the results of the rectal suction biopsy. They will be continued if HD is confirmed until the major operation for HD is performed.

Is there any treatment while awaiting corrective surgery?

Your baby may need to remain in hospital while having bowel washouts and waiting for surgery. Sometimes your baby may go home and return to hospital daily for the washouts, or community nurses may do the washouts at your home. During this time your baby should be able to feed normally.

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 and regular bowel washouts.

What are the symptoms of enterocolitis?

- Swollen stomach
- Reluctance to feed
- Vomiting - this may be green in colour
- Explosive, watery stools
- A high temperature
- Dehydration (baby is pale and floppy)

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.

For more information, see our *Necrotising enterocolitis (NEC)* factsheet.

Corrective surgery

An operation called a 'pull through' is usually performed when your baby is a few weeks or months old. It involves taking out the part of the bowel affected by HD and connecting the healthy bowel to the anus. Sometimes this can be done using keyhole surgery.

What are the long term effects of Hirschsprung's disease?

Many babies with HD suffer with long term problems such as diarrhoea, constipation, soiling and abdominal pain and you may find that your child takes longer to toilet train. Follow up appointments will usually occur on a regular basis for some years.

More information on Bliss and its services

Further advice, support and information can be requested by phoning our Family Support Helpline on freephone **0500 618140**. The line is open from 9am to 9pm, Monday to Friday. You can also email us at enquiries@bliss.org.uk