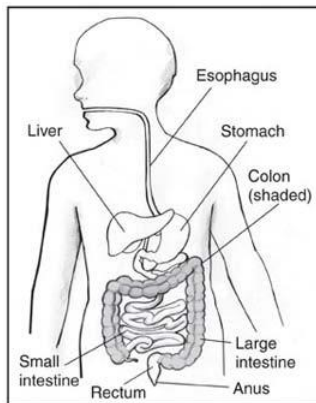


Your baby has recently been diagnosed as having Hirschsprung's Disease (HD). This information leaflet will help you understand more about the disease and how it is treated.

What is Hirschsprung's Disease?

Hirschsprung's disease occurs when some of the nerve cells (ganglion cells) that are normally present in the wall of the bowel (intestine) do not form properly while baby is developing during pregnancy.



The gastrointestinal tract

These nerve cells normally produce a wave-like squeezing motion of the intestine which pushes poo (stool) towards the anus and out of the body. This movement is called peristalsis.

In HD, these nerve cells are not present in the rectum. The cause is unknown but faulty genes cause some cases.

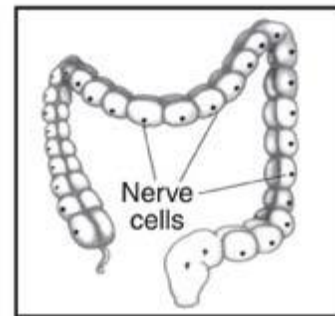
In HD, the healthy part of the intestine pushes the poo until it reaches the region without nerve cells. At this point the bowel stops moving the poo, which then begins to back-up and the intestine becomes partly or completely blocked (obstructed).

The length of the bowel affected by missing nerve cells varies between patients. In most cases, HD only affects the rectum and the very end of the colon (so called 'standard length' HD).

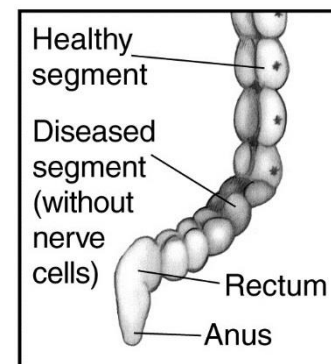
Sometimes the nerve cells are missing from the whole large intestine and even parts of the small intestine. This is called 'long segment HD' or 'total colonic HD'.

How common is HD?

About 1 in every 5000 babies are born with HD. It is more common in boys than girls.



Healthy large intestine. Nerve cells are found throughout the large intestine.



HD large intestine. Nerve cells are missing from the last part of the large intestine

Symptoms and Diagnosis

HD is often considered when a baby born at term has not passed meconium (the dark sticky poo of the newborn) within 24-48 hours after birth. Other symptoms may include poor feeding, vomiting, an enlarged abdomen and constipation.

Less commonly, the baby at first seems to be normal and may even breast feed normally but the baby may go several days between doing poos and their tummy (puku) may become a little

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swollen (distended). This can be difficult to tell apart from the normal bowel pattern in breast fed babies who often will not pass a stool for several days and then pass one or two large loose stools.

If HD is suspected the following investigations may be performed:

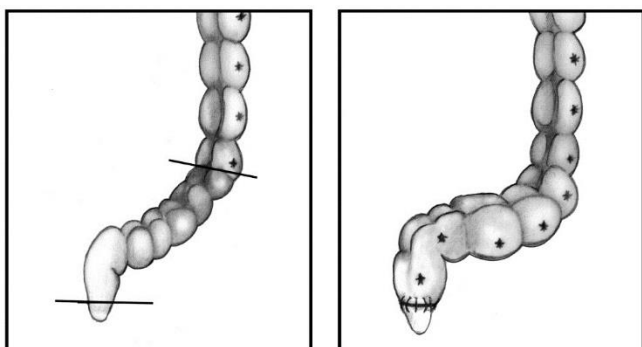
Contrast enema – a liquid dye is gently pushed into the anus through a tube and x-rays are taken. In HD the part of the bowel without nerve cells is usually narrower than the normal bowel.

Rectal Biopsy - the surgeon removes a tiny piece of the lining of the rectum by inserting a small tube into the baby's bottom. This is looked at under a microscope. If the nerve cells are missing then HD is diagnosed. This is the most accurate test for HD.

Surgery

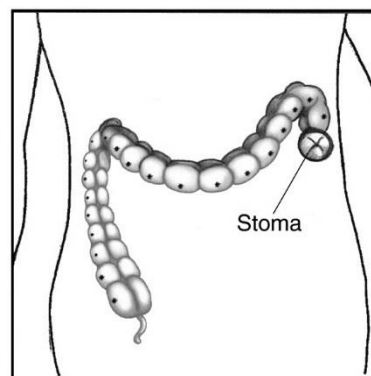
Once a diagnosis of HD is made surgery is the only treatment option. Depending on the circumstances this may involve one or two operations.

If the baby can be successfully treated by daily washouts of the large bowel with salty water (saline) through a tube inserted into the bottom, then a single operation is performed within the first few months of life. The baby usually goes home once the parents have been taught how to do the washouts and then baby returns for the surgery a few weeks later. The diseased bowel is removed and healthy bowel is brought down and joined to the anus.



Surgery for HD. The diseased bowel is removed and healthy bowel brought down to the anus (the pull-through operation)

If the washouts don't work well or if the baby is not suited to this approach the HD is first treated by an operation to make a temporary stoma (colostomy) through which the baby passes poos into a bag on the tummy.



A temporary stoma. The healthy intestine opens on to the surface of the tummy. (There may be a second smaller opening next to the main stoma which is the part of the bowel that goes to the rectum).

A few weeks or months later, the diseased bowel is removed in a second operation and the stoma is brought down and joined to the anus. The baby is then able to poo through his/her bottom and no longer has a stoma. At this time baby's skin is susceptible to severe nappy rash and use of a good skin barrier cream is essential.

The two common operations for removing the diseased bowel and connecting the healthy bowel to the anus (pull-through operations) are named after the surgeons who invented the operations. They are called the Duhamel procedure and the Soave procedure. In both operations, the diseased bowel is removed and replaced by healthy bowel. The outcomes of both operations are similar.

Management / Treatment

If a stoma is made then you will be given advice on how to manage and care for it. If your baby is treated with rectal washouts, you will be shown how to do this safely.

After surgery

Some infants have diarrhoea for a while after the pull-through surgery. Eventually the stool will become more solid and there will be fewer stools passed.

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Hirschsprung enterocolitis

Some children with HD can develop a bowel infection called enterocolitis. This can develop before or after surgery. If your baby/child shows **any** of these signs or symptoms of infection you need to **seek urgent medical attention** as enterocolitis can be very serious.

Signs and symptoms of infection:

- Severe diarrhoea (loose, watery motions)
- Fever
- Vomiting (especially bile green vomiting)
- Swollen tummy
- Crying persistently in pain
- Pale skin and lethargy (no energy)

Images adapted from the National Institute of Diabetes and Digestive and Kidney Diseases (USA).

Contact us

After hours medical centres

Kenepuru Hospital Accident & medical
Open 24hrs a day everyday (04) 918 2300

Paraparaumu -Team Medical Coastlands Shoppingtown
Open 8am -10pm everyday (04) 298 2228

Waikanae
Mon-Thurs 7pm-8am, Fri 5pm-8am, and at anytime during weekends or Public Holidays
(04) 293 6002

Accident & Urgent medical centre Wellington
Open 8am-11pm every day. No appointment necessary (04) 384 4944

Healthline
If you are unsure if your child needs to visit your GP or After hours medical centre you may wish to call Healthline for free advice. Call free anytime on 0800 611 116.

Remember In an emergency call 111