

Hirschsprungs

What is it?

Hirschsprung's is a congenital (present at birth) disorder of the bowel. There is an absence of nerve cells (ganglia) in the wall of the bowel. The portion of bowel that is aganglionic (without nerve cells) does not work and normal peristalsis (movement along the bowel) does not occur. As a result, poo cannot pass and your baby may have any of the signs and symptoms listed below soon after birth. Older children may show a slower onset with a history of chronic constipation, poor growth and development, with a swollen tummy. The portion of bowel which is affected can vary.

The condition can be hereditary and occurs in the first few weeks of pregnancy. The cause is unknown, but it is not due to anything which has been done, or not done, by either parent. Approximately 1 in 6,450 births is affected¹, generally more boys than girls, and those children with Down's Syndrome.

Signs and symptoms

The following may be present at birth:

- ▶ a long delay or failure to pass meconium (the first bowel motion after birth)
- ▶ vomiting
- ▶ reluctance to feed
- ▶ distended (swollen) abdomen

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Reference:

1. EUROCAT (the European Surveillance System for Congenital Anomalies) <http://www.eurocat-network.eu/> @ 31.08.2010. Period covered 2000 - 2008 inclusive.

Hirschsprungs

Investigations

Abdominal x-ray

This is an x-ray of the abdomen, sometimes abbreviated to AXR.

Rectal biopsy

A small instrument is passed into your child's rectum and a biopsy is taken from the lining of the rectal wall which is then checked for the presence of ganglion cells. If none are found then Hirschsprung's disease is confirmed.

Contrast enemas

A special dye that shows up on x-ray that is put into your baby's bottom.



Treatment

Enemas / washouts

Treatment consists of regular washouts to enable the lower bowel to completely empty. This method is suitable for children who do not require immediate surgery.

Surgical

For most children born with Hirschsprung's disease, surgery is carried out soon after birth. Surgical procedures are often carried out at different stages according to the child's condition. The surgery involves removing the affected portion of bowel and 'pulling-through' the unaffected portion. This may result in a temporary stoma.

It must be remembered that the individual techniques will differ, depending on the hospital where the surgery is being carried out and the surgeon who is performing the operation.

Once all the surgery has been completed, many families assume that their child will have no further problems. However we know from experience that some children may have problems in establishing good bowel control.

Potential problems following 'pull-through'

A sore bottom can almost be predicted to occur in the immediate post-operative period, as a result of frequent loose poo. Consequently, the skin around your baby's bottom should be carefully protected using barrier products and frequent nappy changes. A positive approach to preventing sore skin should always be taken instead of waiting until the child's bottom becomes sore. There is a range of products that can help to protect the skin, which your nurse will advise you.

Loose poo

Loose poo is a common problem after surgery until the remaining bowel recovers. The number and consistency of poos your child may have will depend on their condition and type of operation performed.

Constipation

Constipation may occur as a result of a small amount of bowel remaining that is affected by the Hirschsprung's or rarely, stricture formation (a narrowing of the bowel occurring after the operation). Milder cases can be managed with oral laxatives.

Soiling

Soiling may occur due to delayed bowel control. With increasing maturity and the aid of a structured toileting programme, acceptable control can normally be achieved.

Enterocolitis

Enterocolitis is a complication that can occur at any time. The reason is not fully understood but it is thought that there may be overgrowth of bacteria in part of the bowel. It is important to be aware of the signs and symptoms as they can sometimes be confused with gastroenteritis.

- ▶ swollen Tummy (tight and shiny)
- ▶ vomiting (more than is usual after a feed)
- ▶ watery diarrhoea (may have blood visible)
- ▶ discomfort (drawing knees up)
- ▶ foul-smelling wind
- ▶ high temperature

If you are concerned about any of these symptoms, or your baby seems generally unwell, it is important to seek medical advice immediately and to explain that your child has Hirschsprung's disease. Your child should be taken to hospital as there is a risk of severe dehydration.

Bowel management programme

There is always the risk that your child may become constipated. You should seek advice if they have not pooped after a specified time, or if the poo is becoming difficult and hard to pass. Your Stoma Care Nurse or doctor will provide specific advice (encourage them to ask if they are unsure).

The majority of children experience some degree of constipation following reconstructive surgery. A structured programme should begin early, rather than waiting for problems to develop. By keeping your child under regular review until bowel control has been fully established, any major problems may be prevented.

For further advice on:

- ▶ diet
- ▶ fluid intake
- ▶ monitoring of stool habit
- ▶ toilet training

Please contact your specialist nurse.

