Hirschsprung's Disease

What is Hirschsprung's Disease?

Hirschsprungs's (pronounced HURCH-sprungz) disease is a condition that affects the bowel, usually the large bowel. Faeces (poo) moves through the bowel by a process called peristalsis. Special nerves cells control peristalsis and cause the bowel muscles to squeeze and relax regularly. In Hirschsprung's disease these nerve cells are missing from a section of bowel, and this means faeces (poo) cannot be pushed through the bowel in the usual way. The length of bowel affected by Hirschsprung's disease is different in every child.

What are the symptoms?

Symptoms of Hirschsprung's Disease usually appear in the first few weeks of life. Your baby may not be feeding well; they may vomit greenish fluid, and/or have a very swollen belly. Newborn babies may not pass meconium at all; (the dark poo passed in the first few days of life). In slightly older infants, toddlers and children the main symptom of Hirschsprung's Disease is constipation. Laxatives may not work as well as they should, as the affected bowel is unable to push the faeces (poo) through. It is recommended to seek a medical review if you child is having ongoing issues with constipation.

How is it diagnosed?

It is diagnosed by your child having a rectal biopsy, which means a small sample is taken from the bowel and examined under a microscope. If the sample doesn't have any nerve cells, your child will then be diagnosed with Hirschsprung's disease.

What causes Hirschsprung's Disease?

Hirschsprung's disease is what's known as a congenital condition, which means it was present at birth, although it wasn't caused by anything that occurred during pregnancy. While your baby was developing in the womb, the nerves failed to grow normally all the way to the anus. The cause is not known, but some families are more prone to having it, a genetics appointment may be organised for you if appropriate. Hirschsprung's disease affects around one in every 5000 babies, and is much more common in boys than in girls.

How is it treated?

Usually surgery is required to remove the abnormal bowel. This can be done in a one step or two steps depending on how severe the condition is, and how unwell your child is around the time of diagnosis. Initially your child may be managed with rectal washouts (enemas). You will be taught how to do the washouts. If washouts are unsuccessful then surgery may be done sooner. This process will be closely monitored by the medical staff.

Surgery involves the surgeon removing the section of bowel affected by Hirschsprung's disease, and then attaching the two healthy ends of bowel together. This should result in a healthy bowel which faeces (poo) can pass through without any problems.

Sometimes however a stoma is required, which involves the surgeon bringing out healthy ends of the bowel and stitching them onto your child's tummy. Depending on which part of your child's bowel has been affected, it will be called either an ileostomy or a colostomy. Faeces will pass through this stoma and into a bag placed over it, which can then be emptied. You will be taught to care for the stoma. After a period of time, your child will return to surgery and have the non-functioning bowel removed, and the working bowel brought down close to the anus. There are several types of operations to achieve this, depending on your baby/child and your surgeon. The operation will be discussed with you by the surgeon.

What happens after the operation?

- Your child will return to the ward to recover. Regardless as to which procedure they have had, they will have a drip (IV) in their arm or foot which allows us to give fluids until they are drinking well.
- Your child will also have medicine into the drip until they are able to take it by mouth. This allows the bowel to rest and help healing.
- If you have had a one-step procedure it is recommended you apply barrier cream to your child's bottom with every nappy change, as it can become quite sore afterwards.
- If your child has had a stoma formed it may or may not have a stoma bag over it when your child returns from their operation. The bowel should begin functioning within 1-3days.

If my child has a stoma, can I still take them home?

Yes. You will meet the Stoma/Urology Nurse Consultant (NC) who will initially provide the care and supplies for your child's stoma. They will then begin to teach you stoma care when you feel ready to do so. When you feel comfortable you will begin to provide stoma care for your child, initially with the help of your Stomal Therapy Nurse and ward nurses, and eventually independently. You will be provided with supplies to help care for your child's stoma at home on discharge from hospital. Your Stomal Therapy Nurse will continue to be a resource and support person once you are home.

What supports do I have when I take my child home?

There will be several supports for you on discharge. Your Stomal Therapy Nurse will organise the supplies you need and will join you to the appropriate association for ongoing supplies. Community nursing support is also available please discuss this with your Stomal Therapy Nurse or ward nurse. You will also be able to contact your Stoma/Urology NC Monday-Thursday 0830 - 1600 Pager 4099 to discuss any issues, or a member of the surgical team will be available 24 hours a day if you have any urgent concerns.

In Case of an Emergency

Hirschsprungs can cause some children to get severe gastroenteritis type symptoms and in this instance it is very important to present early to the Emergency Department for treatment as your child can become extremely unwell.

Potential long term Issues

While many children often go on to have normal bowel function, some still require help with bowel habit. If you have any ongoing concerns then contact the Surgical Team or the Stoma/Urology Nurse Consultant (NC) Pager 4099.

For more information

Women's and Children's Hospital
72 King William Road
North Adelaide 5006
Telephone: (08) 8161 7000
www.wch.sa.gov.au

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