ongoing help, when you and your baby are discharged home.

Dilatations

Two weeks after the pull-through surgery, you may need to begin a program of rectal dilatations in order to stretch the area, where the newly joined intestine is, to a normal size. You begin with a dilator that fits snugly, and perform the dilatations twice a day. Every 1-2 weeks the size of the dilator will be increased until the correct size is reached.
At this point if your baby has a colostomy it can be closed. The dilatations continue, but the frequency is gradually decreased until they are no longer required.

Colostomy Closure and Nappy Rash

When the colostomy is closed baby will start passing stool through the anus again, it will be frequent and loose. During this time, they are susceptible to severe nappy rash. Good bottom care is required with the use of a skin barrier cream.
There are a variety of barrier creams available and it may take some trial and error to find one that works well for your baby. Please ask your nurse for some advice.
It is advisable to start preparing baby's "bottom" a week/ fortnight prior to this operation. This can be done by placing some stool from baby's colostomy onto the nappy and having baby wear the nappy for 5-10 mins, and/or applying 'skin prep' wipes to baby's bottom. This should be done several times a day.

Adjusting after pull-through

Some children may have diarrhoea for a while, and good bottom care is required.
Eventually the stool will become more solid and the child will need to go to the bathroom less often.
Toilet training may be delayed, as the child learns how to use the bottom muscles only after the pull-through surgery.
Older children might stain their underwear for a while after the surgery. It is not their fault. They can not control this problem, but it improves with time supported by diet and regular toileting.

Infection

Infections can be very dangerous for a child with Hirschsprung's disease. Infection of the large and small intestines is called enterocolitis. It can happen before or after surgery to treat HD. Here are some of the signs to look for:

- fever
- diarrhoea – loose watery motions
- vomiting (especially bile (green) stained vomit)
- Tummy distended (swollen)
- Crying for no reason. Drawing knees up to tummy.
- Pale and lethargic (no energy), not moving arms & legs vigorously.

If the problem is enterocolitis, your child should be admitted to hospital.
Enterocolitis can be life threatening, so watch for the signs and do not delay in seeking medical help.

Adapted from the National Institute of Diabetes and Digestive and Kidney Diseases (USA), and Pull-thru Network.
www.digestive.niddk.nih.gov
www.pullthrough.org

Disclaimer: *Due to the dynamic state of the internet we cannot recommend or endorse these website.*

Parents Support Group:

Bowel Group for Kids Inc. (Australasian)
www.bgk.org.au
P.O. Box 40
Oakdale NSW 2570
Ph: 02 4659 6186
Fax: 02 4659 6381

Note: This is general information only and the situation can vary from case to case

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SSH/WI-3033/008– updated 01/06

Hirschsprung's Disease

A Parent's Guide

Department of Paediatric Surgery
Starship Hospital
Auckland

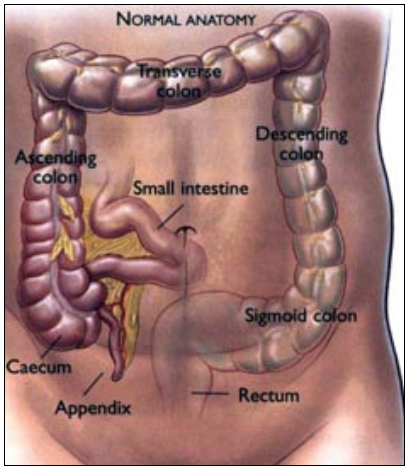
SSH/WI-3033/008– updated 01/06

Introduction

Your baby has been born with Hirschsprung's disease (HD). This brochure is intended to provide you with general information about HD and the operations that are done to deal with it.

What is Hirschsprung's Disease (HD)?

Hirschsprung's disease is a form of intestinal obstruction caused by the absence of *ganglion cells* (special nerve cells) in the wall of the intestine.



These ganglion cells produce the wave-like motion of the intestine, which pushes stool towards the anus where stool then leaves the body. This movement of the intestine is called *peristalsis*.

In HD normal peristaltic movement of the large intestine is not possible.

Background/Incidence

HD was first described in 1886. HD develops before a baby is born. Normally the ganglion cells grow from the top of the gastrointestinal tract all the way to the anus. With HD, the ganglion cells stop growing before they reach the anus. No one knows why the ganglion cells stop growing.

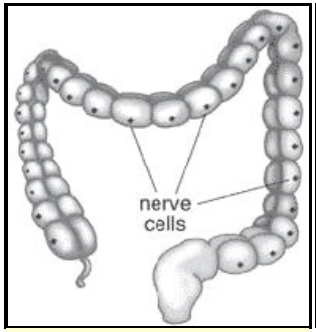
The incidence of HD is 1 in 5000 newborns and it occurs more frequently in boys than girls. The ratio being 4 males to 1 girl.

What happens in HD?

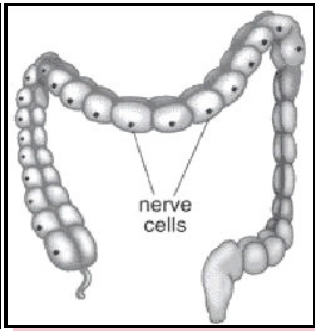
In a person with HD, the healthy muscles of the intestine push the stool until it reaches the part without nerve cells. At this point, the stool stops moving. New stool then begins to stack up behind it causing a blockage of the intestine.

The length of affected intestine varies. If the diseased section includes only part of the large intestine, it may be standard length or short segment HD.

Sometimes the ganglion 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.



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



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

Symptoms & Diagnosis

HD may be considered in a term baby who has not passed their first stool within 24 – 48 hours following birth. Other symptoms include poor feeding, vomiting, abdominal distension and constipation.

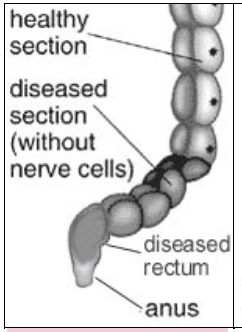
When HD is suspected the following investigations may be done:

- ***Contrast enema*:** Contrast solution is injected into the rectum via a catheter. This enables the anatomy & capacity of the large colon to be visualised. In HD the part of the bowel without ganglion cells is usually narrow compared to the normal bowel.
- ***Rectal Biopsy*:** This is the most accurate test for HD. The doctor removes a tiny piece of the intestine and a pathologist looks at it under a microscope. If the ganglion cells are missing, HD is the problem.

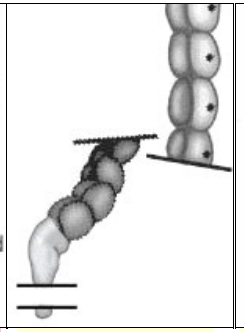
Operations

HD is treated with surgery called a pull-through operation. There are three common ways to do a pull-through operation, and they are called the Swenson, the Soave, and the Duhamel procedures. Each is done a little differently, but all involve taking out the part of the intestine that doesn't work and connecting the healthy part to the anus.

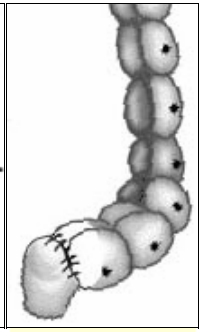
After pull-through surgery, the child has a working intestine.



Before surgery: The diseased section is the part of the intestine that



Step 1: The diseased section is removed



Step 2: The healthy section is attached

Management/Treatment

When the pull-through operation is done will depend on how baby is doing.

Babies who have been very sick may first need surgery to form a colostomy. This helps the child become healthy before having the pull-through operation.

Some doctors may form a colostomy in every child before doing the pull-through operation.

A colostomy is created by dividing the large intestine at the diseased section, and then bringing the two ends through the abdominal wall. This provides the child with a temporary way to pass stool.

The pull-through operation is usually done between 3 – 12 months later, but may be longer.

If baby is feeding and growing well, they may be managed with rectal washouts and rectal dilatations to help them empty their bowel, until they are older and bigger before having the pull-through operation.

Colostomy Care

Since you are dealing with a small baby, there may be some trial and error in handling the ostomy products. You will be provided with help from a stoma therapist and the nurses on the ward.

A referral will be made to a community stoma therapist for

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