

## Hirschsprung's Disease

### What is Hirschsprung's disease?

Hirschsprung's disease occurs when some or all of the large intestine's (colon) nerve cells called **ganglion cells** do not form properly during fetal development. During digestion, intestinal muscles move food forward in a movement called peristalsis. In order for this movement to occur ganglion cells are required. Without the ganglion cells the muscles in the colon cannot push stool out, which then builds up causing either partial or complete bowel obstruction.



Dr Monja Proctor



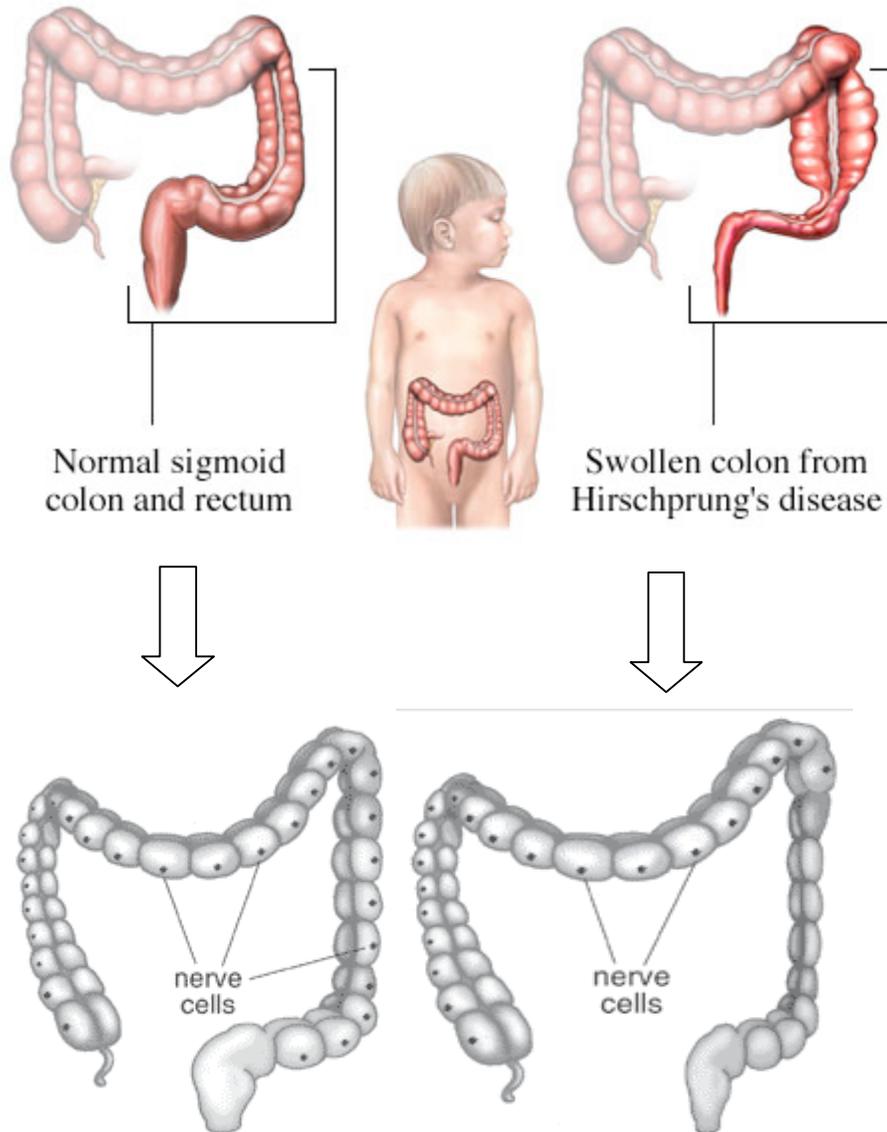
Dr Edwin Hatch



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### How common is Hirschsprung's disease?

This condition occurs in 1 out of every 5000 live births. It is more common in boys than girls. Although the exact cause is not known, it can be inherited.

### What are the symptoms?

Most babies with Hirschsprung's disease are symptomatic during the first 24-48hr of life. However there are some children that do not show symptoms for several months or even years. The most common symptoms are:

No bowel movement in the first 48 hours of life

Gradual swelling of the abdomen

Gradual onset of vomiting

Fever

Children who do not have early symptoms may have the following symptoms:

Fever

Constipation that worsens over time

Small watery stool

Foul smelling stool

Explosive stool

Loss of appetite

Delayed growth

### How is Hirschsprung's disease diagnosed?

There are three tests that may be done to find out if your child has Hirschsprung's disease:

#### Contrast enema

This is a procedure performed to examine the large intestine (colon) for abnormalities. A liquid is gently administered through a soft rubber tube into the rectum in order to coat the inside of the organs so they will show up on x-ray.

#### Rectal Manometry

This test involves inflating a small balloon in the rectum. Ano-rectal Manometry measures pressures of the anal sphincter muscles.

### Rectal Biopsy

This involves removing a tiny piece of rectal tissue and looking at it under a microscope. The absence of ganglion cells confirms the diagnosis of Hirschsprung's disease. This can often be performed at the bedside or in the clinic with minimal discomfort.

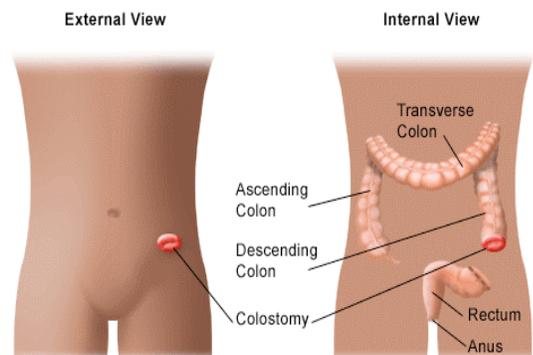
### How is Hirschsprung's disease treated?

Hirschsprung's Disease is treated with surgery. The portion of the intestine that does not have ganglion cells is removed and the areas that are healthy are connected. This is called a **primary repair** or a "pull through".

Some babies or children will require more than one operation called a **staged repair**. This involves the creation of a colostomy.

A colostomy involves connecting the colon to the surface of the abdomen. Stool will drain into a collection bag placed around the ostomy. Eventually a "pull through" is performed and the colostomy is closed.

### Bowel Resection and Colostomy



### **What to expect after surgery.**

We will review care of the incision etc. before your discharge home.

### **Diaper rash**

After the pull though surgery, the first bowel movements can cause the baby's diaper area to get very sore. This can be minimized with good peri-anal care. Stool must be gently removed while avoiding vigorous scrubbing as this can lead to more irritation. Skin is cared for by frequent warm soaks in a tub. Patting dry and applying a barrier to the skin is the best way to avoid breakdown of skin. We recommend using Ilex, Desitin or a thick diaper cream with diaper changes. We will work with you to find the best regimen for your child.

### **Pain**

Managing your child's pain is one of our priorities. By the time of discharge his/her pain will be controlled with oral medications.

### **Diet**

Your child will likely be on an age appropriate diet by the time of discharge home. Some children require additional nutrition guidance which will be arranged.

### **Anal Dilatation**

Surgical healing and scarring occur after any cutting of tissue. This scarring in intestinal surgery is particularly important because it can cause stricture or narrowing of the passage. As a precaution the surgeon may begin to dilate the rectum with a series of dilators. We will teach you how to perform the daily dilatations. This is usually initiated about 2 weeks after surgery and we will provide you with a schedule for dilations at home. The dilators are small tubular instruments which come in advancing sizes to gradually stretch the anal canal to a normal size.

### **When do I come back to the office?**

You will have frequent follow-up appointments with the surgeon. The first appointment is usually made two weeks after your child is discharged from the hospital.

### **When do I call the doctor?**

Fever (greater than 101 degrees Fahrenheit)  
Redness, swelling or persistent pain at the incision.

Vomiting

Full, tight (drum like), Distended abdomen

Constipation

Foul smelling liquid stool

Diarrhea

Explosive gas

### **Long Term Potential Issues:**

Most children have good quality of life after surgery.

We will work with you to optimize stool frequency and other issues that may arise.

### **Enterocolitis**

Enterocolitis is an infection within the intestine. It occurs occasionally in children with Hirschsprung's Disease and can happen after surgery. It can be very serious, sometimes requiring emergent hospitalization. However, if detected early Enterocolitis can be treated as an outpatient with a fast recovery.

### **Things to watch for:**

Fever

Irritability or Lethargy

Refusing to feed

Abdominal distension

Very foul smelling, loose diarrhea

Explosive gas with dilations

### **Constipation/Toilet Learning/Bowel Management**

It is important to remember that surgery for Hirschsprung's Disease needs to be complemented by close follow up for the best outcome. Children may continue to have difficulty with stool retention and constipation. This is especially true during toilet training when withholding stool is common. Toilet learning itself can be somewhat delayed.

***Please call the pediatric surgery office at 206-215-2700 if you have any questions about pain control, infection or other concerns.***

Notes

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