

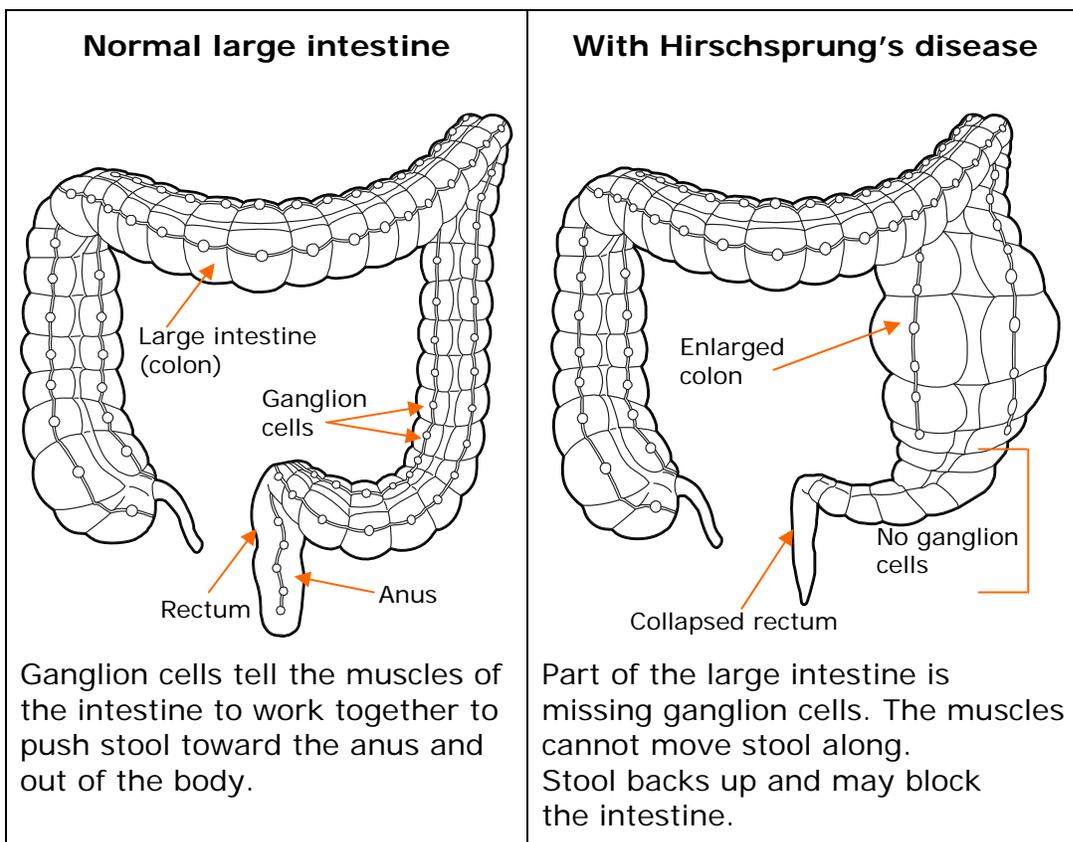
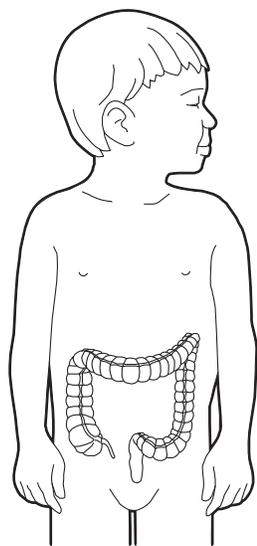
Hirschsprung's disease

A glossary of words is on page 4

What is Hirschsprung's disease

Hirschsprung's disease is a disease of the large intestine. It can sometimes affect the small intestines as well. It occurs when some of the ganglion cells in the large intestine are missing.

Without these cells, the muscles of the intestine cannot push stool towards the anus, where it should leave the body. As a result, the large intestine fills up with stool and becomes partially or completely blocked (obstructed). The intestine becomes enlarged and may get infected (enterocolitis).



What are the symptoms of Hirschsprung's disease?

Hirschsprung's disease may cause problems such as:

- no bowel movements at all (obstruction)
- difficulty having bowel movements (severe constipation)
- gas, bloating (distension)
- delayed growth

Some children may get a serious infection called enterocolitis.

The symptoms include:

- explosive, watery bowel movements (diarrhea)
- vomiting
- abdominal distension
- fever due to infection

Hirschsprung's disease usually just affects the rectum. In some cases, it can affect all of the colon or even the small intestine. The problems that your child has will depend on how much of the intestine is affected.

Most symptoms of Hirschsprung's disease are seen soon after birth. However, if only a short section of the intestine is affected, your child may not show symptoms for several months or years.

What causes Hirschsprung's disease?

Hirschsprung's disease develops before a baby is born. Nerve cells don't grow in all parts of the large intestine.

We don't know what causes the nerve cells to stop growing. Nothing has been shown to cause the problem, including medication or what the mother ate during pregnancy.

Hirschsprung's disease:

- 1 out of 5,000 children is born with this disease.
 - Occurs more often in boys than girls.
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How do you know my child has Hirschsprung's disease?

The doctor will examine your child and review the results of these tests:

- **Contrast enema and x-ray**

A soft tube is placed in your child's anus to put a liquid contrast material into the large intestine. The enema is followed by an x-ray. The contrast makes the large intestine show up better in the x-ray.

- **Biopsy of the rectum**

A tiny piece of tissue from the rectum is checked for ganglion cells. A biopsy is not painful and is usually done without an anesthetic.

How is Hirschsprung's disease treated?

Treatment begins with enemas (rectal irrigation) to clean out the colon. You may be asked to do this at home.

Surgery is needed for all children with Hirschsprung's disease. The surgeon will remove the parts of the intestine that are not working (lack ganglion cells). Some children need a temporary colostomy.

After surgery, you may be asked to pass a small, smooth, metal dilator into your child's anus daily, for several months, to prevent scar tissue from forming.

What will happen in the future?

Some problems are possible, depending on the amount of intestine that is affected and how much was removed.

Your child may have problems with:

- frequent bowel movements and diaper rash
- sensing when he or she needs to have a bowel movement
- toilet training
- constipation

The health care team at McMaster Children's Hospital will continue to help you and your child at regular follow-up visits.

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Glossary

Anus The opening of the rectum where stool leave the body ('bum').

Colostomy Surgery to create an opening from the healthy end of the intestine to the outside of the abdomen. Stool passes through the opening (called a stoma) into a pouch.

Large intestine A long tube that moves waste from digested food (stool) to the anus, where it leaves the body. The large intestine is also called the colon.

Ganglion cells Ganglion cells and nerves in the intestine carry messages to and from the brain. Nerves tell the muscles in the intestine when to contract. The muscles squeeze and move the stool through the intestine.

Rectum The last part of the large intestine near the anus.

Stool Waste from digested food that is passed as a bowel movement ('pooh').
