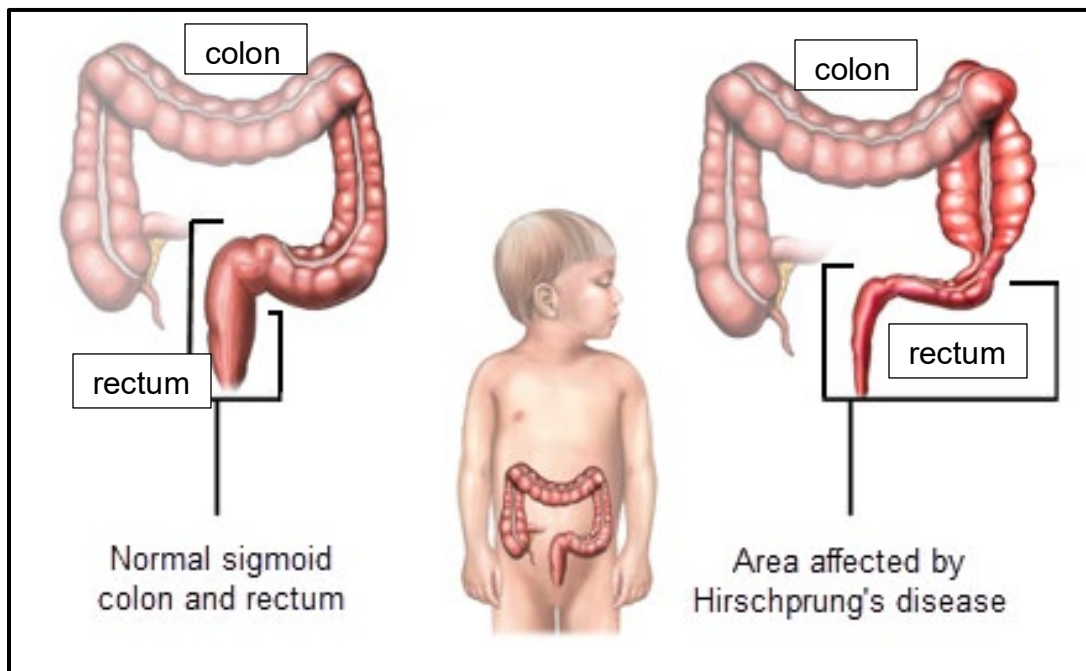


When Your Child Has Hirschsprung Disease.

The colon (large intestine) holds poop (stool) left in the body after digestion. The colon is lined with nerves. These nerves tell the colon when to contract (squeeze) and relax (open). The colon contracts and relaxes to push poop into the rectum. Poop leaves the rectum through the anus.

In a child with Hirschsprung disease, nerves are missing in the anus and rectum and in part or all of the colon. The part of the colon without nerves can't relax. So, poop can't leave the body.



What are the types of Hirschsprung Disease?

Hirschsprung disease is grouped based on how much the colon is affected. Your child's health care provider can tell you which type of Hirschsprung disease your child has. Types of Hirschsprung disease include:

- Short segment - where nerves are missing from the rectum and a small portion of the colon.
- Long segment - where nerves are missing from the rectum and a larger portion of the colon.
- Total colonic - where nerves are missing from the entire colon, rectum and sometimes part of the small intestine.

What are the signs of Hirschsprung Disease?

Hirschsprung disease is most often found in newborn babies. The most common sign is not pooping within the first 1 to 2 days (24 to 48 hours) of life. Other signs in newborns can include swelling of the belly, vomiting, and fever.

In some cases, the problem doesn't cause signs or symptoms until the child is a little older. When this happens, signs of a problem include constipation, poor appetite, and not growing as much as normal.

How is Hirschsprung Disease diagnosed?

First, the doctor takes a health history and does a physical exam. These help find the cause of the problem. The following tests may be done:

- **Contrast enema.** A liquid, called contrast, is inserted through the anus into the rectum using a soft tube. The liquid coats the inside of the colon so it shows up on an X-ray. The X-ray shows if the colon is backed up with poop or if it has an abnormal shape or size.
- **Anorectal manometry.** A tube with a small balloon on its tip is inserted through the anus into the rectum. The balloon is then inflated. Normally, the balloon would stimulate nerves in the rectum, causing the rectum to relax and open. With Hirschsprung disease, the rectum does not relax.
- **Rectal biopsy.** A small tube is inserted through the anus into the rectum. Through this tube, a tiny amount of tissue is removed. The tissue is studied under a microscope in the lab by a pathologist. The biopsy can be done in the clinic or in the hospital under anesthesia. In some cases, a larger amount of tissue is taken with a surgical biopsy done in the OR with anesthesia. If no nerves are found in the tissue, the child has Hirschsprung disease.

How is Hirschsprung Disease treated?

Hirschsprung disease is treated with surgery. The section of the colon without nerves is removed. The healthy colon is then reconnected to the anus. This surgery is called a pull-through.

Your child may need an ostomy. The ostomy provides a new way for poop to leave the body. A small opening is made in the belly and the colon is brought up to the skin. Waste leaves the body through this opening and empties into a bag. The ostomy may be temporary or permanent, depending on how much of the colon is affected by Hirschsprung disease. If your child needs an ostomy, the doctor will tell you more about it.

What are the long-term concerns?

If a large portion of the colon must be removed, the child may have long-term digestive problems. But in general, most children with Hirschsprung disease do very well after surgery.

Problems such as constipation and fecal incontinence may happen. These problems are often treated with medicines and a high-fiber diet. Also, children with Hirschsprung disease are more likely to develop an infection of the colon called enterocolitis. Your child's doctor can tell you more about your child's situation.

When to call your child's doctor

Call the doctor if your child has any of the following signs. These may be from an infection in the colon called enterocolitis:

- Temperature of 100.4°F (38°C) or higher
- Diarrhea that is very smelly, explosive, or has blood in it
- Throwing up (vomiting)
- Belly pain
- Swelling of the belly
- Has low energy, or not interested in playing or doing their usual activities