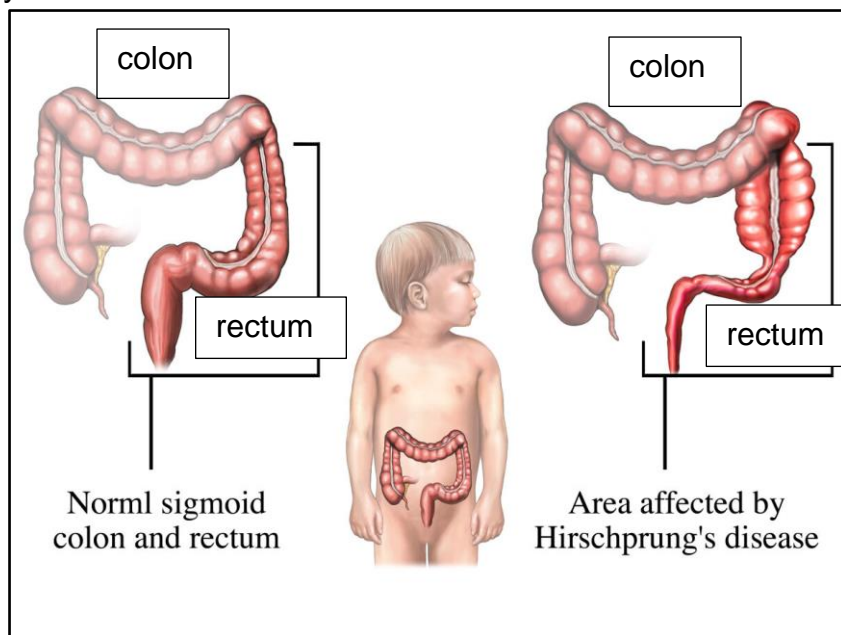


When Your Child has Hirschsprung Disease.

The colon (large intestine) holds poop (stool) left in the body after digestion. The colon is lined with nerves. These 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 [Hirsch-sprung] 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 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:

- Ultra-short segment where nerves are missing from a very small portion of the rectum.
- 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.
- Very long segment 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 (hard, dry poop that can be hard to pass), 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:

- **Barium [bar-i-um] or contrast enema.** A white liquid containing barium is inserted through the anus into the rectum. The liquid coats the inside of the colon and lower part of the small intestine, so they show up on an X-ray. The X-ray shows if the colon and lower part of the small intestine are blocked by poop. In some cases, a substance other than barium may be used, but the test is done in the same way.
- **Rectal biopsy [bi-op-sy].** 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 in the lab. In some cases, a larger amount of tissue is taken with a surgical biopsy. If no nerves are found in the tissue, it is a sign of Hirschsprung disease.
- **Anorectal [ano-rec-tal] manometry [ma-nom-e-try].** 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 colon, causing the colon to relax and open. With Hirschsprung disease, the colon contracts and closes instead.

How is Hirschsprung Disease treated?

Hirschsprung disease is treated with surgery. The section of colon without nerves is removed. The healthy ends of the colon are then reconnected. This surgery is called resection [re-sec-tion] and pull-through.

Your child may need a stoma [sto-ma]. The stoma provides a new way for poop to leave the body. A small opening is made in the belly. This opening is connected to the colon. Waste leaves the body through this opening and empties into a bag. The stoma may be temporary or permanent, depending on how much of the colon is affected by Hirschsprung disease. If your child needs a stoma, 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.

Your child's poop may not be normal. Due to Hirschsprung disease, the rectum and anus may be collapsed. If so, poop can't be pushed out normally. Minor problems such as constipation and leakage may happen. These problems are often treated with medicines and a high-fiber diet. Also, children with Hirschsprung are more likely to develop an infection of the colon (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 [en·tero·co·li·tis]:

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