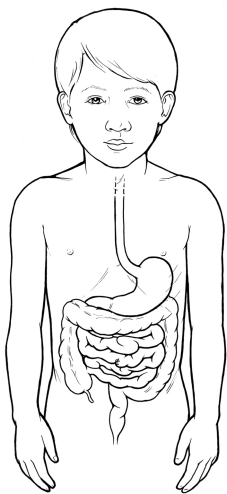


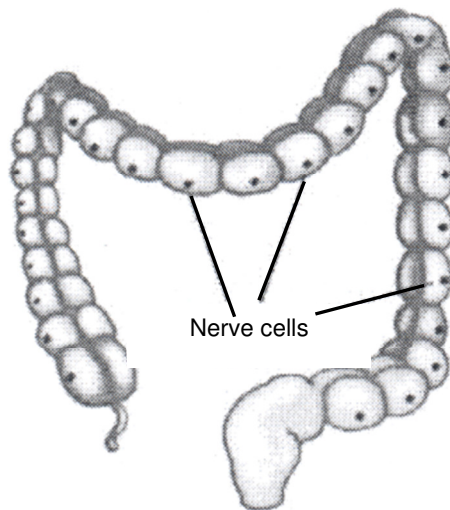
Hirschsprung Disease (HD)

Hirschsprung (HERSH prung) disease (HD), affects the part of the bowel also known as the large intestine, or the colon. It can affect the entire colon and part of the small intestine, but this is rare. With HD, the colon did not form the nerves needed for it to work as it should.

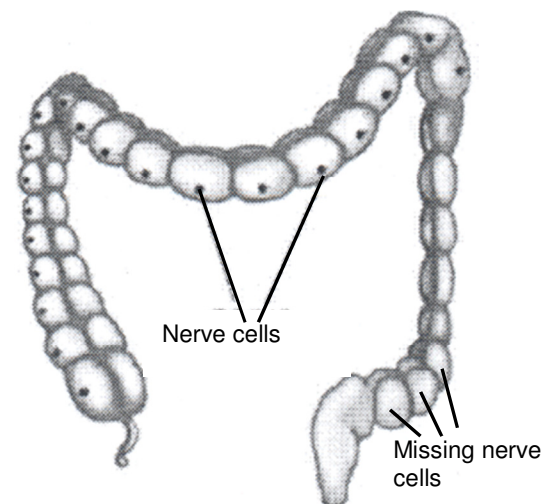
Usually, the part of the bowel that has no nerve cells is in the lower part of the colon (Pictures 1, 2 and 3). This section gets very narrow, and the bowel above it gets larger. This is because stool (bowel movements) and gas cannot pass through the narrow area. In a person with HD, the healthy muscles of the bowel push the stool and gas until it reaches the part with no nerve cells. At this point, the stool stops moving. New stool then begins to build up behind it. Stool can fill up the large intestine and cause serious problems like infection, a burst colon and even death.



Picture 1 The digestive system inside the body.



Picture 2 Healthy large intestine: Nerve cells are found throughout the intestines.



Picture 3 HD large intestine: Nerve cells are missing from the last part of the intestine.

Symptoms

The symptoms of HD depend on the person's age:

Newborns and Infants

- No bowel movement in the first 48 hours of life
- Swollen abdomen (belly)
- Vomiting (throwing up) a green liquid called bile
- Fever
- Abdominal pain
- Diarrhea

Young Children, Teens and Adults

- Severe infection
- Severe constipation
- No appetite
- Delayed growth
- Watery stools
- Abdominal pain

How HD Is Diagnosed

Several tests* may be done to see if a child has this disease.

(See page 3 for a list of Helping Hands.)

- **Rectal exam** - The doctor examines the rectum with a gloved finger.
- **Contrast enema*** - Contrast material is put into the rectum through a small tube. Then an X-ray is taken. The contrast shows up on X-rays. It allows the doctor to see the outline of the bowel.
- **Anal manometry*** - (Ay nul mah NAH meh tree) A small tube with a balloon on the end is inserted into the rectum. The balloon is inflated with air. This allows the doctor to see if the nerves and muscles of the colon are working right.
- **Rectal biopsy*** - A small tube is inserted into the rectum. A very small piece of the lining of the bowel is taken out through the tube. This small piece of bowel is studied under a microscope in the lab to see if there are nerve cells.

Treatment and Surgery

Hirschsprung disease is treated with surgery. Symptoms may first be treated with irrigations of the colon. This can flush out the stool blocked behind the part of the colon without nerves. It can also help relieve your child's pain and swollen belly. Your child will have the irrigations until he has surgery.

During surgery, part of the bowel without nerve cells is taken out. This surgery can be done entirely through the opening in the anus, with no incisions. Rarely, small incisions are made, and surgery is done with a scope. Most of the time, a colostomy (koe LOSS toe mee) is not necessary. A temporary colostomy is only needed if the child is very ill. This lets stool pass out of the colon through an opening in your child's belly, into a bag.

Infection

Infections can be very dangerous for a child with HD. Infection of the large and small intestines is called enterocolitis (EN ter oh koe LIE tis). It can happen before or after surgery.

Watch for these signs. Call your child's doctor immediately if you see:

- Underarm temperature over 101°F
- Vomiting or diarrhea
- Child acts "sluggish" – no energy
- Swollen belly
- Bleeding from the rectum

After the Surgery

After surgery, most children have normal bowel function. Constipation can occur, but it can be easily treated with changes in diet and laxatives. If your child does have accidents and soiling after potty training, the doctor will order a Bowel Management Program.

Helping Hands

For more information, please ask your nurse for these Helping Hands:

Bowel Management Program, HH-II-222.

Contrast Enema, HH-III-2.

Anal Manometry, HH-III-10.

Rectal Biopsy (Suction Procedure), HH-III-75.

Ostomy Care: Ordering Supplies, HH-II-99, and one of the following:

Ostomy Care: One-Piece Pouch with Attached Skin Barrier, HH-II-107;

Ostomy Care: One-Piece Pouch without Attached Skin Barrier, HH-II-4; or

Ostomy Care: Two-Piece Pouch, HH-II-33

If you have any questions, be sure to ask your doctor or nurse, or call _____.