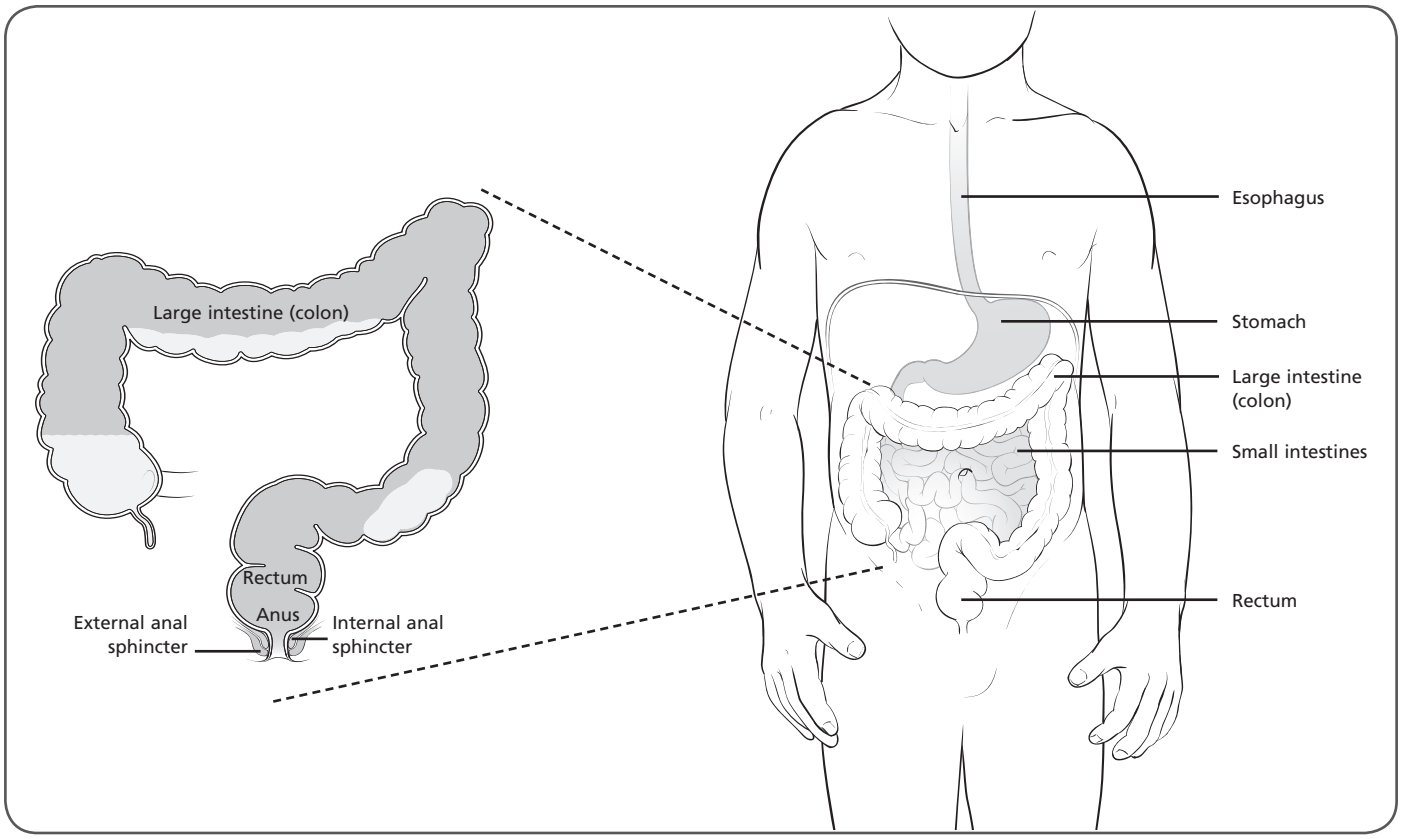
Let's Talk About...

Hirschsprung's Disease



What is Hirschsprung's Disease (HD)?

Hirschsprung's (HERSH-prungs) Disease or HD is a problem with the intestines (bowels) that children are born with. The intestines usually develop with muscles and nerves. In children with HD, the nerve cells which are usually in the last parts of the intestines (rectum) do not form the right way. These nerve cells are called ganglion (GANG-lee-on) cells, they help to move stool forward and relax the rectum so stool can pass from the body. If the child does not have ganglion cells in part of the intestine, stool backs up in the normal part of the colon. These children are constipated so badly that this could cause a severe infection or even create a hole in the intestines (a perforation).

The part of the intestines that does not have nerves could be short (just a part of the colon) or could be the whole colon and part of the small intestines. All children with Hirschsprung's disease need surgery to take out the part of the intestines that do not work.

How often does HD occur?

HD occurs in 1 out of every 5,000 live births. It occurs more frequently in boys than in girls. It is usually the only birth defect. It may occur in children with others syndromes, like Down syndrome.

Is HD inherited?

Sometimes HD is inherited and sometimes it is not.

How does the doctor know my child has HD?

All children with HD have constipation. Other signs may be present along with constipation.

Newborns can have the following signs of HD:

- Does not pass a stool (meconium) in the first two days of life.
- Belly gets bigger and fuller (abdominal distention).
- Refuses to eat.
- Vomits, has a fever with abdominal distension and/or failure to pass meconium.

Infants can have the following signs of HD:

- Does not grow normally
- Does not have a good appetite
- Belly gets bigger and fuller
- Has diarrhea and vomiting
- Has an infection in the intestines (enterocolitis) and may have explosive watery diarrhea, fever, vomiting, and look very sick

Children can have the following signs of HD:

- Ribbon-like, stools that smell bad
- Belly gets bigger and fuller
- Does not eat enough, does not grow enough
- When the doctor examines the child, the doctor can feel a mass (which is stool)

How does the doctor diagnose HD?

The doctor will examine your child and order some other tests. These may include:

Abdominal x-ray: This is an x-ray of the belly and can show if the intestines are blocked. This may give some helpful information, but the doctor will order other tests, too.

Contrast enema: This is a special enema with contrast in it. This makes the intestines easier to see on x-ray. In HD, the colon may be narrower or larger than normal at certain points.

Rectal biopsy: This test must be performed to diagnose HD. The doctor will take a small bit of tissue from the rectum. It is examined with a microscope to see if it has nerve cells or not. In infants, "a suction rectal biopsy" may be done right in the child's hospital room or in the doctor's office. This does not hurt, because children cannot feel that part of the intestines.

It is possible the child may need another biopsy in the operating room. This procedure tells the doctor whether the child has HD or not.

Anorectal manometry: This test may be done in older children. It tells the doctor if there are normal reflexes in the rectum and anus (the last part of the intestines).

How is HD treated?

HD is treated with surgery. Your child may have surgery through a small cut with the use of a camera called "minimally invasive" or it may be a regular surgery with a larger opening. The surgeon removes the part of the intestine that does not have ganglion cells. Then the surgeon attaches the normal intestine to the anus (called a pull through procedure). The surgeon may do this with one or sometimes with two surgeries (called "one stage" or "two stage" surgery).

One stage surgery is commonly used for newborns. The intestine without ganglion cells is removed and the normal intestine attached to the anus.

In two stage surgery, the child receives a temporary colostomy and then the surgeon closes up the colostomy in the second surgery. In the first stage, the surgeon makes a small hole in the belly and attaches part of the colon to it. This lets stool leave the body through this opening and empty into a bag. The child can eat and grow while the bowel heals and returns to a normal size. The second surgery happens later. In the second stage surgery, the surgeon removes the part of the intestine without ganglion cells. Then he closes the colostomy with stitches and attaches the normal intestine to the anus.

The surgeon will carefully consider whether your child needs one stage or two stage surgery. The choice is complicated and is based on your child's individual needs. Your surgeon will discuss this with you.

If the surgeon chooses one stage surgery, you may have to give your child rectal irrigations for a while

before surgery. Your surgeon will let you know which surgery is best for your child.

What kind of care will I give my child after surgery?

If your child has the two stage procedure, the ostomy nurses at the hospital will teach you how to take care of the colostomy and change the bag that collects the stool.

You may need to protect your child's skin from loose stool with creams and powders.

You may need to change your child's diet or have her drink more water.

Your doctor and nurses will help you learn all the care your child will need.

What are some of the problems children with HD have?

Diarrhea: After the pull-through surgery, most children have diarrhea for a while. This may give a baby a diaper rash. You can help your child by using special creams and powders on her skin. After a while, your child's stools will get more solid and she will have bowel movements less often.

Anus is too tight: Some children may have trouble with bowel movements because the anus (the area where normal intestine was sewn to the anus) is tight. The surgeon may teach you how to do anal dilations to prevent scar tissue from forming and increase the anus size.

Toilet training issues: Some children toilet train later than usual. This is normal for these children.

Need more water: One of the things the colon does is absorb water and salts from the stool. The body needs the water and salts. Because the colon is shorter, HD children may need to drink more liquids.

Tube feedings: Some children have long-segment or total colon HD (more of the colon does not have nerve cells). These children may need special tube feedings.

Infection: HD children may develop enterocolitis (infection of the intestines). This is serious, and could be life threatening. This can happen if stool and bacteria back up in the colon. Children with enterocolitis may have:

- Fever
- Swollen Belly
- Vomiting
- Diarrhea
- Bleeding from the Rectum
- Sluggishness.

If your child has these signs, tell your child's doctor immediately. The doctor will want your child to receive rectal irrigations to rinse stool and bacteria out of the colon. The surgeon may admit your child to the hospital for IV fluids and antibiotics.

What happens to children with HD after surgery?

Most children with HD have a small amount of colon that does not have ganglion nerves. This is called short segment disease. These children usually have good bowel control over time. After surgery, these children have frequent stools (5–10 times a day). This usually changes so by one year after surgery, the child has one to four stools per day.

Enterocolitis can happen after surgery, but usually only for one to two years after surgery.

Most children (three of every four children) have some amount of stool incontinence (not being able to hold stool properly) or constipation. Common ways to treat these issues are a special diet, laxatives, or enemas. A small number of children (1 in every 10) have bad constipation or stool incontinence (called "fecal incontinence") for a long period of time. These children may need a bowel management program. Children who are neurologically impaired or have Down syndrome have more problems with fecal incontinence and severe constipation.

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