Hirschsprung's Disease

Children's HOSPITAL • ST. LOUIS BJC HealthCare

Washington University in St. Louis Physicians

Call 911 or an ambulance if your child:

- Struggles to take each breath
- Is unable to speak or cry
- Grunts with each breath
- Has blue or white lips or nails

Call the Surgery Team night or day if your child has any of the following symptoms:

- Is less than 3 months old and has a temperature over 100.3˚F (38˚C).
- Temperature greater than 101˚F (38.4˚C)
- Is vomiting.
- Has diarrhea.
- Has bleeding from the anal opening.
- Has a swollen stomach.
- Looks or acts very sick.
- Is limp or very weak.

Note: Call the General Surgery Office 314.454.6022 during office hours. If after hours (nights, weekends, and holidays) call 314.454.6000 and ask for the Surgery Resident on-call to be paged.

Hirschsprung (HURSH-spruhng) is a disease of the large intestine. A portion of the large intestines is missing important nerve cells.

What is the Cause?

- It happens before the baby is born, while he or she is still growing.
- It is more common in boys than girls, children with Down's syndrome, and many other syndromes.
- It can be genetic.
- Some of the nerve cells or ganglion cells that are normally in the intestine do not form the way they should. The job of the nerve cells is to move stool forward during digestion, a movement called peristalsis.
- Stool will not move through the intestines like it should without these nerve cells. The healthy muscles in the intestine push the stool until it reaches the part without the nerve cells. The stool stops moving and new stool begins to build up behind it. This causes constipation or in severe cases an obstruction (a complete blockage of stool).

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What are the Signs and Symptoms?

80% of children have symptoms in the first 6 weeks of life. However, children with only a small part of intestine affected may not show symptoms for several months or years.

- No meconium stool (first stool all babies pass) within 24-48 hours after birth.
- Swelling/distention of the abdomen (stomach), that slowly increases.
- Gagging and onset of vomiting (often of green fluid or bile).
- Fever
- Constipation
- Fussy/irritable
- Delayed growth
- Loss of appetite
- Diarrhea
- Enterocolitis

How is it Diagnosed?

Abdominal x-ray: This may show a bowel blockage

Contrast Enema: This is an x-ray with contrast used to look at the large intestine (colon) for abnormalities.

Rectal biopsy: This is essential to diagnose. There are two types; a suction rectal biopsy and a full-thickness rectal biopsy. With both we take a small sample of cells in the rectum and look at them under a microscope for nerve (ganglion) cells.

What is the Treatment?

All children with Hirschsprung's disease need surgery. This surgery is referred to as the "pull-through" operation.

- This surgery removes the part of the intestine without the nerve cells and then joins the healthy bowel and colon together. If the child is sick from an infection, obstruction, and enlarged bowel, an ostomy is needed.
- An **ostomy** is a small hole made in the abdomen. The top part of the intestine comes through the small hole. Stool leaves the body through this opening, called a stoma. The stool goes into a bag that is attached to the skin by a wafer around the stoma. When your child is stable and the intestines have had a chance to get better, we can then perform the pull-through surgery. The opening in your child's abdomen should be closed at that time.

St. Louis Children's Hospital
One Children's Place
St. Louis MO 63110
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How do I Care for my Child after Surgery?

- After surgery most children are able to have a bowel movement normally.
- Some children may have diarrhea for a while, and infants may develop diaper rash. We recommend you use diaper cream, or prescription cream to help with the diaper rash.
- After a while the stool will become more solid and your child will need to use the bathroom less often. Toilet training may need to be pushed back as the child learns how to use their bottom muscles only after the surgery. This is because they need to get back control of their muscles again and will get better with time.
- The doctor/nurse will talk to you about wound care, bathing, and activity restrictions before you go home.
- Your child will need to drink more to make sure he/she is getting enough fluids.

  One job of the large intestine is to collect the water and salts the body needs. Since your child's intestine is shorter, it will absorb less. Tube feedings may be needed to make sure your child is getting enough food.
- Eating high fiber foods like cereal, bran muffins, and fresh fruit can help decrease constipation and diarrhea.

Frequently Asked Questions:

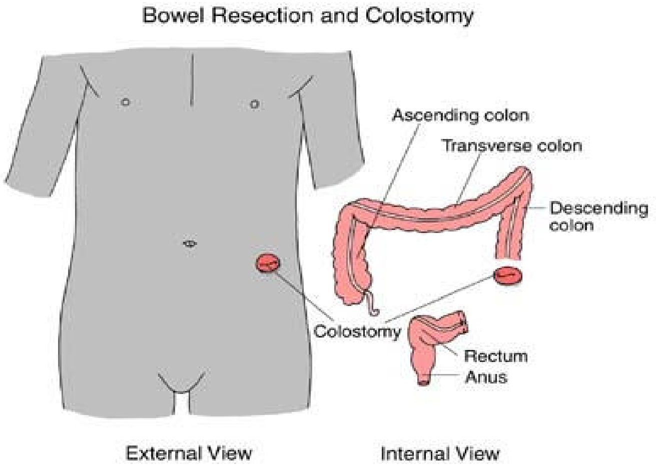
1. What is Enterocolitis?

- It is an infection of the large/small intestine. Infections can be very dangerous for a child with Hirschsprung's disease. This can happen before or after surgery.
- **Enterocolitis can be life threatening, so watch for the signs and symptoms of the following:**
  - Temperature over 101˚F (38.4˚C)
  - Swollen abdomen
  - Vomiting
  - Diarrhea
  - Bleeding from the anal opening
  - Sluggish behavior
  - Foul smelling stool
  - Explosive stools
  - Abdominal cramping
- Your child may have to stay in the hospital if they become sick, but only in severe cases.

- The treatment includes IV fluids to keep your child hydrated and antibiotics to help with infection. The intestine will be rinsed regularly with a salt-water solution (normal saline) until all remaining stool has been removed. This is called a rectal irrigation.
- Sometimes enterocolitis can be treated at home with antibiotics and rectal irrigations, but this will be decided by your medical provider.

2. What if my Child has an Ostomy? What will his or her life be like?

- Most children are more comfortable after they get an ostomy because they can pass gas more easily and are not constipated.
- Older children will be more comfortable too, but they may have some trouble getting used to an ostomy. They may be worried about being different from their friends.
- Constipation will not be a problem anymore.
- There are no restrictions in your child's activity.
- The nurses will teach you and your child how to care for a stoma and change the ostomy bag. Please talk with your nurse about any concerns or questions you may have.



For more information about Hirschsprung's Disease or Ostomies, contact:

The Family Resource Center on the 3rd floor at St. Louis Children's Hospital. Call at (314) 454 - 2350 or email at frc@bjc.org.

This handout is for your general information only. The lists above are guidelines and do not include all symptoms. This document is not a substitute for your child being seen by a doctor. Always call your child's doctor if you have any questions or problems. If your child's condition gets worse, call your child's doctor or go to the emergency department.

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