

Surgery for Children with Hirschsprung's Disease A Guide for Patients & Parents

What is Hirschsprung's Disease?

Hirschsprung's disease is a birth defect that affects about 1 out of 5,000 newborn babies. Babies with this birth defect are born without special intestinal nerve cells called ganglion cells. These nerve cells allow the intestine to relax so waste or stool (poop) can pass through the intestine and out of the body. Without these special nerve cells, the intestine cannot relax and it becomes very narrow. When the intestine is narrowed, stool can't easily pass. As a result, babies with Hirschsprung's disease cannot have bowel movements on their own and have severe constipation.

In most babies with Hirschsprung's disease, only a small part of the large intestine near the rectum is missing ganglion cells. In some babies, all of the large intestine is missing ganglion cells. Very rarely, babies can be born without ganglion cells in both the large and small intestine.

How will the diagnosis of Hirschsprung's disease be made?

Most babies have a bowel movement in the first one to two days after birth. Hirschsprung's disease is usually suspected when a baby does not have a bowel movement for several days following birth. A baby with Hirschsprung's disease often has a large, swollen appearing abdomen and may vomit green bile after feeding.

Two tests commonly are used to diagnose Hirschsprung's Disease. The first is a contrast enema in which a special liquid called contrast fills the large intestine through a small tube placed in the anus. X-ray pictures are taken as the liquid enters the intestine. If Hirschsprung's disease is present, the pictures of the intestine will show a wide or dilated area next to a narrow segment of intestine. The narrow area is the part of the bowel without ganglion cells. The wide area of the intestine is healthy bowel filled with stool that cannot be passed.

In the second study, a pediatric surgeon removes two or three tiny pieces of intestine using a narrow suction tube placed in the anus. This procedure is not painful. These samples of intestine are examined under a microscope for the ganglion cells. If ganglion cells are seen in the biopsy samples, the intestine is normal and there is no Hirschsprung's Disease. If no ganglion cells are seen on the biopsy samples, the diagnosis of Hirschsprung's disease is made.

While most babies are diagnosed very soon after birth, some children may not be found to have Hirschsprung's disease until much later in life. These children often have severe constipation, diarrhea, slow weight gain or a life-threatening infection of the bowel called "enterocolitis."

If my child has Hirschsprung's disease, how will this be treated?

The part of the intestine without ganglion cells must be removed with an operation. Usually this is done in one operation called a primary repair. In most babies, we operate without making an incision (surgical cut) on the abdomen. The operation is done with telescopic (laparoscopic) instruments through the anus. In

some babies, more than one operation may be required. This is called a staged repair. In the first operation -- or first stage -- the part of the intestine without ganglion cells is removed and a temporary stoma made. A stoma is an opening on the abdomen where the intestine is brought out to the skin. If your child has a stoma, stool will drain into a bag worn on the outside of the body. Several months later, the stoma is closed and the intestine is sewn together. Your child's pediatric surgeon will talk to you about the best operation for your child.

After the operation, if your child does not have a stoma, he or she will have a lot of diarrhea. It will be important to protect the skin around the anus at all times. Your child's nurse will teach you how to mix and apply a cream called "butt balm." Use this cream at all times to protect the skin from the irritating effects of diarrhea. The recipe for butt balm is: a four-ounce tube of Desitin® ointment, a one-ounce bottle of Stomahesive® powder and a 1/2-ounce bottle of Mycostatin® powder combined to create a cream the consistency of peanut butter. The surgical nurse will order the prescription powders needed to mix the balm. Please give us the phone number of your pharmacy once you go home.

How long will my child remain in the hospital after the operation?

If your baby has anal surgery without a stoma, he or she will go home in one to three days. If your baby has a stoma, he or she may stay in the hospital longer. Your baby will be ready to go home when he or she is having bowel movements, eating and drinking without vomiting, and is comfortable on pain medication taken by mouth.

How do I take care of my child at home following discharge from the hospital?

Pain: Prescription pain medication (narcotics) usually isn't needed once your child is home. Most children only need Acetaminophen (Tylenol®) or Ibuprofen (Motrin®). Give the medication according to the dosage directions on the label. If your child is still uncomfortable, call our office and we may prescribe something stronger.

Dressings: If your baby has an incision on the abdomen there will be a gauze pad and clear plastic dressing in place. This dressing is to be removed two days after the operation. There may be a small amount of blood on pieces of tape called Steri-strips® under the dressing. This is normal. Your child can bathe with the Steri-strips® in place. These can be pulled off one week after the operation. The skin around the incision may be red and bruised and slightly swollen. This can last several weeks. There will be no visible stitches to remove because they are under the skin. The stitches will dissolve after several weeks. Sometimes these stitches are irritating and will come out of the skin through the incision. If this happens, the incision will look red and may drain white, yellow or red fluid. This can be normal for some children and will get better with time and daily bathing.

Healing ridge: If your child has an abdominal incision, you will feel a firm ridge just under the incision once it is healed. This is called a healing ridge and it is normal. The healing ridge lasts for several months before it softens and disappears.

Bathing: Your child may bathe or shower as soon as two days after surgery. Once your child is feeling better, either in the hospital or at home, he or she may bathe or shower. Bathing is a good way to soothe the anal area and gently clean the skin without injury. Some parents prefer to use a hair dryer, on the low (cool) setting, to dry the anal area well before putting on more protective cream.

Skin Care: Continuously apply the recommended protective skin care products. Begin as early as the day of surgery. Do not stop using the skin protective products until there are fewer bowel movements, usually after many weeks. If your child develops a rash that does not get better, please call our office.

Activity: Your child can return to doing his or her usual activities without any special restrictions, unless the pediatric surgeon tells you otherwise. If your child is school age, school activities can begin again as soon as he or she feels well enough. If you need a letter sent to your child's school about the operation and recovery, please call our office. If your child has a stoma, supplies will be ordered for your baby. You will be taught how to take care of the stoma and use the supplies before going home.

Do I see the surgeon again after the operation?

If your child has a stoma, you will need to make an appointment to be seen one after discharge. If your child has a primary surgery or stoma closure, he or she will need to be seen in the Pediatric Surgery office two weeks after the operation. At this visit, his or her anus will be checked to make sure the opening is large enough for stool to pass easily. Sometimes a child's anus has to be dilated, or stretched, for several weeks or months following surgery. If this is necessary, your child's surgeon or surgical nurse will teach you how to do dilations at home.

When do I call your office?

Call our office at (415) 476-2538 if:

- You have any concerns you have about your child's recovery
- Your child has a temperature of 101 degrees or higher
- Your child's incision is red
- Your child has severe pain at the incision
- The incision that is painful to touch
- Fluid is coming out of the incision
- The number of bowel movements changes each day
- There is no bowel movement for one day
- Red rash around the anus is not getting better
- You have any questions about your child's stoma

This information is for educational purposes only and is not intended to replace the advice of your physician or health care provider. We encourage you to discuss with your physician any questions and concerns you may have.