



Endorectal Pull-Through for Hirschsprung's Disease

What is Hirschsprung's disease?

Hirschsprung's Disease is a condition where children are missing the nerve cells ("ganglion cells") within the wall of their colon or rectum. These cells are responsible for the normal wave-like motion of the bowel (peristalsis), and at the point where these cells are missing - the stool stops and a blockage (obstruction) occurs. This disease is also called "megacolon".

Who gets Hirschsprung's disease?

Hirschsprung's disease is a congenital disease which means that a person is born with it. It can be hereditary which means a parent can pass it on to a child. It happens to 1 out of every 5,000 babies born and is more common in boys than in girls.

How is Hirschsprung's disease diagnosed?

The diagnosis is made by barium x-rays and a rectal biopsy. Rectal biopsy involves removing a small piece of rectal tissue to learn whether the nerve cells that control the bowel muscles are present. Sometimes another test called rectal manometry is performed. This is where a small catheter with a balloon is placed in the rectum and records pressure changes and muscle movements.

How is the decision made that surgery is needed?

If the tests show that the nerve cells are missing in part of the bowel, surgery is recommended. The goal of surgery is to remove the abnormal bowel and attach the normal bowel to the anus just above the sphincter (the muscle that holds in stool). Three commonly performed operations known as pull-through procedures are called the Swenson, Duhamel, and Soave operations. The long term results seem to be the same whichever operation is used. Often these procedures can be done without a big incision in the belly by using a laparoscope and sometimes they can be done through the anus (transanal pull-through).

Depending on the place where the nerve cells stop in the bowel and the overall condition of your child, the surgeon may decide that your child needs to have a stoma created. This may be the only thing that is done at first or it may be done with the pull-through procedure. A stoma (ostomy) is where a small section of bowel is brought up to an opening in the abdomen so that stool can come out easily into a small bag. Your nurse will teach you how to care for the stoma and make arrangements for the supplies needed before you go home..

What can I expect from surgery?

Your child should wake up soon after surgery and will be given pain medicines for the next 2 – 3 days. They will have an I.V. (a tube that delivers fluids into the vein). S/he will probably have a small tube in her/his nose for a short time after surgery until s/he can start passing gas or have a stool. Once this happens the tube can be removed and the child can start to drink some water. If s/he does not start vomiting, s/he can move onto formula or regular foods. When s/he is eating well, passing stool, and does not have any fever s/he can go home. The nurses will begin putting special ointments or creams on your child's bottom right after surgery to protect the skin from a bad "diaper" rash that is common after pull-through procedures. You will continue doing this at home. After your first visit with the surgeon, you may need to begin rectal dilatations or stretching of the rectum where the normal bowel was sewn to the anus. Your surgeon will instruct you on how to do this and will provide you with the instruments to do this procedure at home.

Tips for the day of surgery.

Since you will be staying in the hospital for a few days – bring a change of clothes for yourself and whatever comfort toy that your child likes to have with her/him.

How much time should I plan off work?

Someone will need to be home with the child for several weeks or at least until the first visit with the surgeon.

What care is needed at home after surgery?

Care of the incision: Your child's surgeon will give you instructions

Activity: (dependent on age – no heavy lifting or school for older child)

Diet: Your child's surgeon will give you instructions

Bathing: Your child's surgeon will give you instructions

When can my child return to school or daycare?

Your child may return to school or daycare 4-6 weeks after surgery. Be sure to ask your child's surgeon when s/he may return to PE

What should I call my pediatrician for?

Call your pediatrician for regular checkups and other illnesses. If you see the pediatrician for an illness that has vomiting and diarrhea as a symptom – be sure to remind them that your child has Hirschsprung's disease. (Just because they had the corrective surgery – does not mean the disease goes away.)

Will this affect growth and development?

After the surgery is done your child should expect normal growth and development.

Is there anything else I need to know to care for my child?

IT IS VERY IMPORTANT TO CALL THE SURGEON FOR ANY EPISODE OF WATERY, EXPLOSIVE DIARRHEA WITH OR WITHOUT FEVER. Other symptoms to watch for will be a full, tight (drum-like) belly and constipation (no stools). Any of these symptoms could mean a serious condition called "Hirschsprung's enterocolitis". This is a dangerous condition that affects children with Hirschsprung's disease even after surgery and should always be watched for.

Although most children have excellent results following surgery for Hirschsprung's disease, almost 10 – 20% will have continued problems with bowel movements. Some may have constipation, while others may not be able to hold their stool and have "accidents" in their underpants (incontinence). There are several ways to treat these problems so it is VERY important that you talk to your child's surgeon or nurse about these problems if they are happening.

Please reproduce and distribute this sheet to your surgery families. This teaching sheet can also be downloaded at www.APSNA.org.

Copyright 2006, Melinda Klar. Copied with permission by Jones and Bartlett Publishers, Sudbury, MA."