



## **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 tiny 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 balloon which records pressure changes and muscle movements is placed in the rectum.

### **How is the decision made that surgery is needed?**

If the tests show that the nerve cells are missing at a certain point of 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 (muscle that holds in stool). Often the surgery can be done without a big incision in the belly by using a laparoscope and sometimes they can be done just through the anus (transanal pull-through).

### **What can I expect from surgery?**

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 (belly) so that stool can come out easily into a small bag. (Ask for the teaching sheet on ostomies).

Your child should wake up soon after surgery and will be given pain medicines for the next 2 – 3 days. He/she will have an I.V. (a tube that delivers fluids into the vein) and a small tube in his/her nose until they start passing gas. Once this happens the tube can be removed and the child can start to drink some water. If they do not start vomiting, they can move onto formula or regular foods.

**When will my child be able to go home?**

When he/she is eating well, passing stool, and does not have any fever he/she 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.

**What should I bring to the hospital?**

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 him/her.

**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 kind of care is needed at home after surgery?**

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.

Care of the incision – your health provider will tell you how to care for the incision. It should remain clean and dry.

Activity limitations – Quiet, supervised activity for one to two weeks after surgery.

Frequent soaks and prescribed ointments for your child's bottom to treat diaper rash

Bathing – your health provider will tell you how to bathe your child.

Feeding: this will be discussed with you before you leave the hospital

Pain management: Your child will go home on oral pain medication

**When can my child return to school or day care?**

Your child can return to school/daycare 4-6 weeks after surgery, or as your surgeon tells you.

Your child should not have PE or play contact sports for at least six weeks. Your surgeon will tell you when he/she may resume PE. Ask us for a note to excuse your child from PE.

**What else do 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.

You can still call your pediatrician for regular checkups and other illnesses. After the surgery is done your child should expect normal growth and development. 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.)

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](http://www.APSNA.org).

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