

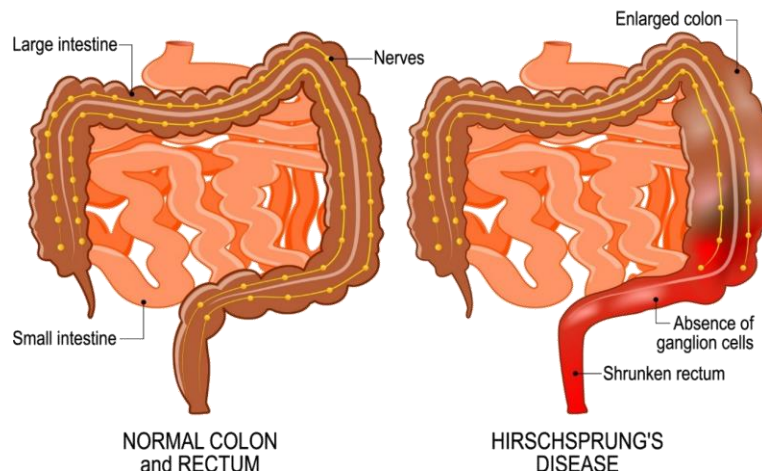
PATIENT AND FAMILY INFORMATION SHEET

Hirschsprung Disease

What is Hirschsprung Disease?

Hirschsprung Disease (HD) is a disorder where children are missing the nerve cells (“ganglion cells”) within the wall of the intestine (bowel). These cells cause normal wave-like motion of the bowel. Stool stops and does not move well where these cells are missing, causing distention (big belly) and sometimes vomiting.

HD is present at birth. It can be hereditary meaning a parent can pass it on to a child. It happens in 1 out of every 5,000 babies born and is more common in boys than in girls.



How do I know if my child has Hirschsprung Disease?

Here are some of the symptoms your child may have:

Constipation or intestinal obstruction (abdominal swelling and not stooling) are the most common signs that happen after birth. Constipation medications usually do not work on someone with HD.

If your child has HD, they can get an infection in the large intestine called Hirschsprung enterocolitis. Hirschsprung enterocolitis makes the child very sick.

Here are some of the signs a baby may have:

- Green or brown vomit
- Explosive stools after inserting a finger into the baby's bottom
- Swelling of the abdomen (belly)
- Diarrhea, often with blood

Here are some of the signs a child may have:

- Failure to grow
- Swelling of abdomen (belly)
- Unexplained fever
- Vomiting
- Not able to pass stools without a suppository (is a pill placed into the child's rectum) or enema (an enema involves flushing liquid into the child's anus using a special wash bottle).

Signs may vary on how much intestine (bowel) is affected.

How is Hirschsprung Disease treated?

The treatment of HD is surgery.

The surgeon will do tests before planning treatment options:

- **Contrast enema:** This test is done by placing a tube in the bottom and allowing contrast to fill the intestine.
- **Rectal biopsy:** This is when you remove a tiny piece of rectal tissue to learn whether the nerve cells within the intestines (bowel) are present.

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How is Hirschsprung Disease treated?

- If your child has Hirschsprung enterocolitis, he/she will need colonic irrigations.
 - A colonic irrigation is used to clear the bowels when there is a fear of enterocolitis.
 - This is done by using small amounts of salt water (saline) injected into the rectum through a large tube (catheter) to irrigate the colon.
 - It is different from an enema, which uses a large amount of saline with a stimulant, such as soap, to cause your child to have a large bowel movement.

Contrast enema showing the transition zone of a patient with Hirschsprung Disease



What happens before surgery?

- If the tests show that the nerve cells are missing, surgery is recommended.
- The goal of surgery is to remove the abnormal bowel that does not have the nerve cells and attach the normal bowel to the anus, just above the sphincter (muscle that holds in stool).

What happens during surgery?

- Often the surgery can be done without a big incision (cut) in the belly by using a laparoscope (special camera). Sometimes the normal bowel can be pulled through the anus (transanal pull-through) without a belly cut.
- Depending on the place where the nerve cells stop in the bowel and how your child is doing, the surgeon may decide that your child needs to have a stoma done.
- Sometimes, the pull-through procedure is done at the same time as the stoma. A stoma (ostomy) is a piece of bowel is brought up to an opening in the skin of the abdomen (belly). The stoma drains stool into a small bag rather than the child passing stool through the anus.

What happens after surgery?

- Your child should wake up soon after surgery and will be given pain medicines for the next 2 – 3 days.
- He/she will have a tube that delivers fluids into the vein (IV).
- A small tube may be put through the nose, into the stomach, to help gas and fluid to get out (“nasogastric/NG tube”).
- The NG tube will come out when it is no longer draining.
- Your child will be allowed small sips of clear liquids. He/She will slowly be given more if everything is going well.
- If they do not start vomiting, they can move onto formula or regular foods as directed by the surgical providers.

When can I be with my child again?

As soon as your child wakes up after surgery, someone from the recovery room will call you so you can be with your child again.



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Will my child have any pain?

- Pain varies depending on the type of surgery needed.
- With a pull-through, the incision does not cut through muscles and nerves which means there is little pain and usually only requires 1-2 days of pain treatment.
- Sometimes there is a need for an incision (cut) to be made on the belly. This cuts through muscle and nerves which will require more pain treatment.

When will my child be able to go home?

- When he/she is eating well, passing stool, pain is well controlled with medications taken by mouth, and does not have any fever.
- If your child does not have a stoma, 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.
- If your baby has a "stoma" you will be taught how to take care of it before you go home.

How do I care for my child at home?

- After your first visit with the surgeon, you may need to begin rectal dilatations or stretching where the normal bowel was sewn to the anus. Your surgeon will instruct you on how to do this. Your surgeon will provide you with the instruments to do this at home. Not every child needs dilations.

Care of the Incision:

- For pull-through (bottom) -- Clean with warm soapy water. It may be helpful to use a syringe to squirt soapy water over the child's bottom and then carefully pat the area dry with a soft cloth. Never rub or use baby wipes that have chemicals unless your surgeon gives you the okay.
- For belly incisions, follow your surgeon's instructions for care.
- No tub bathing for 1 to 2 weeks- or as directed by your surgeon.

Activity Restrictions:

- Nothing such as medicine or thermometers should be placed into the bottom until your surgeon allows.
- **Return to school or day care:**
 - Your child can return to school/daycare 4-6 weeks after surgery
 - Your child should not have physical education (PE) or play contact sports for at least six weeks.
 - Your surgeon will tell you when he/she may resume PE.

Diaper rash:

- Frequent soaks and prescribed ointments for your child's bottom to treat diaper rash.
- It is easier to prevent diaper rash than to treat it once the rash is developed. Barrier cream should be applied with each diaper change.

Diet:

It is important to learn which foods cause your child's stool to be either more formed or more loose. This will allow you to adjust the diet to help promote the right consistency of stool. Daily intake of water and foods such as fruits, vegetables and whole grains are often helpful to prevent constipation. Discuss the appropriate dietary needs with your surgeon or nutritionist.

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How do I care for my child at home?

Bowel Pattern:

- It is important to watch for signs of constipation (not stooling). Your child should have a soft stool every day.
- Laxatives may be needed to have a daily soft bowel movement. Laxative use should be discussed with your doctor before starting.
- If your child does not have the ability to potty train, discuss bowel management options with your care team.

Medications:

- Give pain medications as directed by your surgeon.
- Give laxatives as directed by your surgeon.

Anal Dilations

- Anal dilations are often started at 2 to 4 weeks after surgery. This is to prevent narrowing of the area repaired as it heals.
- Important things to know:
 - Dilators are smooth medical rods used to keep your child's new anus from closing up.
 - The surgeon will pass the first dilator through the repaired area 2-4 weeks after surgery.
 - The surgeon or nurse will teach you how to do the anal dilation for your child at home.
 - It is important to do these correctly for best healing and prevention of another surgery.
- To find the final size of the anal dilator your child will use, see chart below.



When Starting Dilatations, Your Child Is:	Final Desired Dilator Size
1-4 months	#12
4-8 months	#13
8-12 months	#14
1-3 years	#15
3-12 years	#16
13 years of age and older	#17

Anal Dilations

Once you find that the dilator goes in easy two times a day and does not cause discomfort, you may start to taper (reduce) how often you dilate your child's anus. While you taper, you will still use the goal size dilator.

- Month 1: One time a day for a month.
- Month 2: One time every other day for a month.
- Month 3: One time every third day for a month.
- Month 4: Two times a week for a month.
- Month 5: One time a week for a month.
- Month 6-8: One time a month for three months.



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Anal Dilations

If the dilatation gets harder, causes discomfort, or is bloody at any time during the above schedule, go back to dilating two times a day. When you can easily put in the dilator without discomfort, start the taper schedule again from the beginning (one time a day for a month).

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 OR VOMITING WITH OR WITHOUT FEVER. Other symptoms to watch for will be a full, tight (drum-like) belly, decreased eating/ drinking, decreased urine or constipation (no stools). Any of these symptoms could be a sign of a serious condition called “Hirschsprung enterocolitis.” This is a dangerous condition affecting children with HD even after surgery. Any diarrhea, vomiting, abdominal distention (big belly), decreased oral intake or fever should prompt phone call to your surgeon.

You can still call your pediatrician for regular checkups and other questions. Remind your pediatrician that your child has HD especially if he or she is having vomiting, diarrhea or fever. Even after surgery for HD, your child is still at risk for Hirschsprung enterocolitis.

Although most children have excellent results following surgery for HD, 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.

When should I call the office?

If your child experiences any of the following, please call our office:

- Fever over 100.5 F
- Full, tight (drum-like) belly
- Pain not relieved by medications as recommended
- Redness or swelling of the incisions
- Increase or decrease in bowel movements
- Serious diaper rash
- Vomiting or decreased oral intake of fluids/ foods
- Decreased number of wet diapers (urine)

Your child will need to follow up with the surgeon. You will receive specific instructions for follow up when your child is discharged.

Please don’t hesitate to call our office if you have any problems or concerns.

Surgical provider: _____

Office Number: _____

After hours number, if applicable: _____

Thank you for allowing us to care for your child.