

Hirschsprung's Disease

What is Hirschsprung's disease?

Hirschsprung's disease is a disease of the large intestine (colon). Stool is normally pushed through the colon by muscles. These muscles are controlled by special nerve cells called ganglion cells. Children with Hirschsprung's disease are born without ganglion cells in the colon. In most cases, only the rectum is affected, but in some cases more of the colon, and even the entire colon, may be affected. Without these ganglion cells, the muscles in that part of the colon cannot push the stool out, which then builds up.

How common is Hirschsprung's disease?

Hirschsprung's disease occurs about once in every 5,000 babies born. It is slightly more common in boys than in girls.

What are the symptoms of Hirschsprung's disease?

Symptoms of Hirschsprung's disease usually show up in very young children but can sometimes show up in older children or teenagers.

Most babies with Hirschsprung's disease do not pass stool on the first or second day of life. After that, almost all babies with Hirschsprung's disease are constipated and have difficulty passing stool. These infants may vomit and their tummy enlarge because they cannot pass stool easily. Some babies have diarrhea instead of constipation. Children and teenagers with Hirschsprung's disease usually experience constipation their entire life. Normal growth and development may be delayed.

SPECIFIC INSTRUCTIONS:



How is Hirschsprung's disease diagnosed?

There are three tests that may be done to find out if a child has Hirschsprung's disease:

- ◆ contrast enema
- ◆ manometry
- ◆ biopsy

Contrast enema X-ray

A contrast enema is an X-ray of the abdomen taken after the doctor puts a liquid such as barium through the anus into the colon. The contrast makes the colon show up better on X-ray. The test is most useful in newborns when the baby cannot push the stool out and an obstruction to the lower end of the colon is suspected. Children with Hirschsprung's disease will have a narrow colon where the ganglion cells are missing. The diagnosis of Hirschsprung's disease is made by rectal biopsy, but the enema X-ray may have a role in determining examining the length of the affected colon.

Manometry

In ano-rectal manometry, the doctor places a small balloon into the rectum and measures how well the muscles around the anus respond to inflations of the balloon. Normally, when the balloon is inflated there will be a well-defined relaxation of the internal anal sphincter muscle (one of the anal muscles). Children with Hirschsprung's disease will not relax the muscles in the anus. If there is no relaxation, then this suggests that there are other parts of the large intestine that may not be able to push stool through. This results in stool building up and causing a blockage.

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Performing ano-rectal manometry requires special equipment and people skilled in its operation. Also, children having manometry need to be very still during the test. Manometry may not be very reliable in infants less than 12 days of age.

Biopsy

This is the most accurate test for Hirschsprung's disease. The doctor takes a very small piece of the rectum to look at under the microscope. Children with Hirschsprung's disease will not have any ganglion cells in the sample taken.

How is Hirschsprung's disease treated?

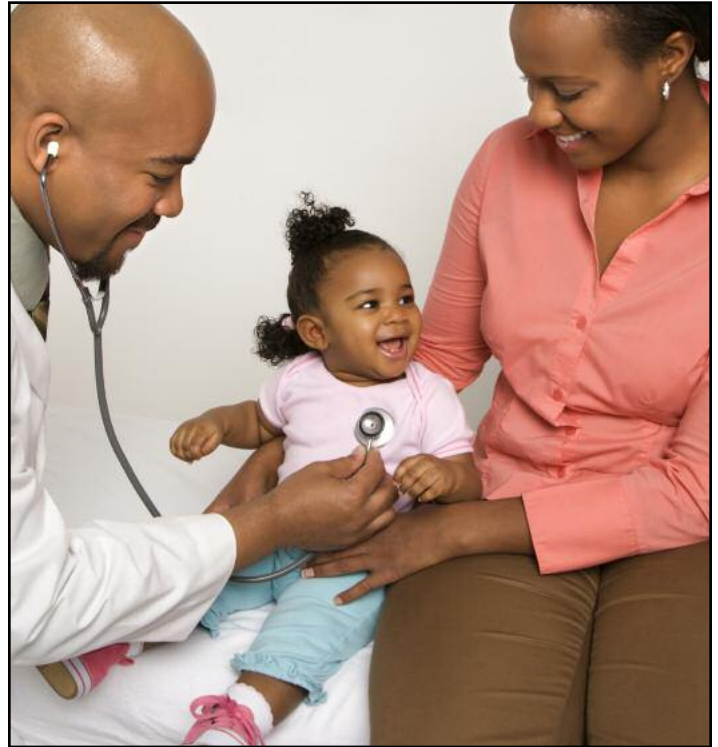
Hirschsprung's disease is treated with surgery. Surgery is done to remove that part of the colon that lacks the ganglion cells and then to connect the healthy colon above this to the anus. The operation can be done in two stages. In the first stage the surgeon separates the healthy colon from the affected colon. Then the healthy colon is brought out to the skin as a colostomy (opening of the colon to the abdomen) which then empties into a special bag that the parents can manage. Several months later the surgeon removes the affected colon and takes the healthy colon from the colostomy and connects this to the rectum just above the anus. In some babies the surgery can be done as a single operation without a colostomy. This can only be done safely if the baby is healthy and the colon is not full of stool. This operation can be done through the anus so that the baby does not require any incision on the front of the abdomen.

What can you expect after surgery?

Initially after surgery the stools may be loose, but over the longer term constipation is more likely to be a problem. With constipation, it is recommended that your child have a diet high in fiber and fluid. You may wish to speak with a dietician to help you provide high fiber food choices for your child. Some children require medications (such as laxatives) to manage the constipation. Most children will toilet train normally, although some may take longer than others. Over the long term children do well. Even after surgery for Hirschsprung's disease there is risk of developing a condition called enterocolitis. This is a serious infection of the colon where the children can be very sick with severe diarrhea. They often require intravenous fluids and antibiotics. This condition seems to be most common in the first few years of life and rarely occurs beyond the age of six.

For more information or to locate a pediatric gastroenterologist in your area please visit our website at: www.naspgghan.org

IMPORTANT REMINDER: This information from the North American Society for Pediatric Gastroenterology, Hepatology and Nutrition (NASPGHAN) is intended only to provide general information and not as a definitive basis for diagnosis or treatment in any particular case. It is very important that you consult your doctor about your specific condition.



LINKS :

<http://digestive.niddk.nih.gov>

www.iffgd.org

<http://www.mayoclinic.com>

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