

ROCKY MOUNTAIN PEDIATRIC SURGERY

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Hirschsprung's Disease

What is Hirschsprung's Disease?

Hirschsprung's Disease (congenital aganglionosis) occurs once in approximately 5000 live births and more in boys than girls. There may be a hereditary influence with a risk of ~5% of a second child having the disorder.

During intrauterine development, the nerves to the intestine do not migrate to the rectum. Normal coordinated intestinal motion, or "peristalsis," depends on nerve cells embedded within the intestinal wall. Without these nerve cells, the bowel cannot propel the foodstuff or waste products. Instead of normal peristalsis, the segment of bowel without nerves often does not move correctly, resulting in a functional obstruction or blockage. In Hirschsprung Disease, nerve cells are usually absent just in the lower part of the large intestine (from the rectum upward). Occasionally however, nerves may be absent (or abnormal) in the entire large intestine, or rarely the small intestine as well.

What are the Symptoms and Signs of Hirschsprung's Disease?

- **The majority of patients with Hirschsprung's Disease are diagnosed in the neonatal period.**
 - Failure to pass a first stool without assistance within 2 days and followed by abdominal distension and vomiting.
- **Sometimes the disease is subtle, and is not suspected until later.**
 - In older children, there is almost always a history of never having had a normal bowel movement.
 - Most of these children have a markedly distended abdomen.
 - Watery diarrhea (as only liquid can get by the obstructed stool)
 - Enterocolitis: bacterial overgrowth and inflammation of the colon)
 - Life-threatening sepsis (blood borne infection).

How is Hirschsprung's Disease Diagnosed?

- **Prior coming to the Pediatric Surgery Office:**
 - Your baby's doctors may order an abdominal x-ray and a contrast enema (an x-ray study performed with contrast placed into the rectum).
 - Occasionally, anorectal manometry is performed which measures the pressures within the anorectal area.
- **The definitive diagnosis of Hirschsprung's Disease is made by taking a small piece of the lining of the bowel called a biopsy.**
 - This is usually performed in the newborn by a suction biopsy instrument. This does not require anesthesia and is usually done at the bedside in the nursery or office.

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- **In older patients**, a deeper biopsy is needed and is usually done in the operating room under general anesthesia.
 - The tissue is examined by the pathologist who determines whether nerve cells are present or absent.

What is the Treatment for Hirschsprung's Disease?

The basic principle in the treatment of Hirschsprung's Disease is to remove the segment of intestine without nerve cells and replace it with normal bowel

- **Stable child:** after confirmation of the diagnosis, a laparoscopic pull-through procedure using three to four small incisions may be performed.
 - In some cases, rectal irrigations (injecting saline through a small flexible catheter inserted up the bottom) may be performed first and then the pull-through operation performed when the child has grown.
- **Unstable child:** your child might have many health concerns; therefore a definitive surgery cannot be performed right away. In this case a colostomy, where a piece of the bowel is brought to the surface of the skin, is performed. This relieves the obstruction and allows the patient to recover after which the pull-through procedure may be done safely.
 - If the pull-through procedure is difficult to perform, your surgeon may elect to do a colostomy with the pull-through to protect the operation by diverting the stool. The colostomy is then closed in four to six weeks.
- After the procedure, you may be given instructions to order anal dilators. These need to be brought to your child's first post-operative appointment. Do not use the dilators prior to this appointment. You will be given instructions for dilations at your follow up visits.

What are the Post-operative and Long-term Complications?

All operations under general anesthesia incur the risks of the anesthesia, bleeding, infection, and post-operative bowel obstruction.

- As the new segment of intestine is sutured or sewn to the anal tissue, scarring and narrowing may occur which may require dilatations or operative revision of the connection.
 - The majority (75-80%) of the patients do quite well eventually with normal bowel movements and no soiling.
 - About 5% may continue to have severe constipation or incontinence which is usually treated by medical means of stool softeners, laxatives, enemas, and dilations.
- **It is important that all patients that have had a pull-through procedure avoid constipation.** Constipation may lead to enterocolitis or an infection of the intestine which is quite serious and may be life-threatening.
 - If an infant or child becomes constipated, has small, watery stools with abdominal distension, and appears lethargic or has a fever, it is imperative that you contact us or your pediatrician.
 - It is particularly important for children to avoid constipation when their diet changes (like on vacation, starting school, etc).
 - You should always have glycerine suppositories and Pediatric Fleet® enemas with you when traveling.
- **Patients with Down's syndrome and neurologic impairment will have a higher incidence of constipation and incontinence and can only be treated with medical management and a great deal of patience.**

***Disclaimer:** Your child's condition is unique. The information contained on this web site is not intended to substitute for advice from a doctor or nurse. If you are unsure about any aspect of your patient's care, please contact us at 303-839-6001, or talk to your pediatrician.*

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