

THE CHALLENGES OF HIRSCHSPRUNG DISEASE

Hirschsprung Disease Affects the Large Intestine of Newborns, Babies and Toddlers

Hirschsprung Disease is More Common in Boys Than Girls

Hirschsprung Disease (HD) is a congenital defect that occurs when nerve cells are absent from the lower part of the colon. The absence of these cells, called ganglion cells, leaves a portion of the colon essentially paralyzed and unable to function properly.

The following summary will provide information on the cause of HD, along with symptoms, diagnosis and treatment.

01 | How Hirschsprung Disease Develops

HD is caused by the failure of ganglion cells to reach the bottom end of the colon. Nerve cells develop early in the embryo. They begin in the esophagus and migrate along the intestinal tract to the stomach, small intestine and finally the colon. For unknown reasons, the genes will sometimes function abnormally and the nerve cells do not reach the lower part of the colon, leading to Hirschsprung Disease.

02 | Symptoms and Diagnosis

Newborns with HD are usually unable to have a bowel movement in the days after birth. Signs and symptoms vary with the severity of the condition and may include:

- Distended belly
- Vomiting
- Constipation or gas
- Diarrhea
- Fatigue
- Jaundice
- Failure to thrive

When symptoms are present, the child's doctor will perform an exam and recommend the following tests to make a conclusive diagnosis:

- Abdominal X-ray with contrast

- Manometry test to measure muscle control
- Colon or rectal biopsy

03 | Treatment

Treatment will vary based on many factors including the severity of the disease and the child's overall health. Surgery is the most effective treatment for HD. The most common surgery involves removing the defective section of the colon and can often be done laparoscopically. In more severe cases a colostomy or ileostomy may be required.

Symptoms improve or disappear in most children after surgery. A small number of patients may experience continued symptoms. Children who get treated earlier or who have a shorter segment of the intestine removed have a better outcome.

For more information and to find support on Hirschsprung Disease, please visit:
<https://www.reachhd.org/>

Did You Know?

1 in 5,000 newborns are affected by Hirschsprung Disease

References

<http://www.mayoclinic.org/diseases-conditions/hirschsprung's-disease/home/ovc-20214664>

<https://www.niddk.nih.gov/health-information/digestive-diseases/hirschsprung-disease>

<http://www.stanfordchildrens.org/en/topic/default?id=hirschsprungs-disease-90-P01999>

<https://my.clevelandclinic.org/health/articles/behcets-disease-what-you-need-to-know>