

Hirschsprung's Disease

Hirschsprung's disease is a congenital disorder wherein there is limited or absent intestinal motility due to a lack of ganglion cells in the intestine. The absence of ganglion cells prevents the intestine from receiving appropriate signals from the nervous system, resulting in constant sympathetic stimulation; and, therefore, increased bowel tone. Because the intestine is in a contracted state, peristalsis does not occur. Newborns with Hirschsprung's disease do not pass meconium within the first 24 to 48 hours of birth and may also present with a distended abdomen and feeding intolerance, or refusal to feed, with bilious vomiting. A digital rectal exam may reveal no stool on the examiner's finger; however, the exam can stimulate the bowel and subsequently cause forceful expulsion of stool. Interventions to treat the disease include surgical resection of the bowel with the possibility of a temporary colostomy.



PLAY PICMONIC

Mechanism

Congenital Aganglionic Megacolon

[Present-from-birth A-gang-lions Mega-colon](#)

Hirschsprung's disease is a congenital disorder in which there is limited or absent intestinal motility due to a lack of ganglion cells in the intestine.

No Peristalsis

[No-sign Pear-on-stilts](#)

The deficiency of ganglion cells present in Hirschsprung's disease prevents the intestine from receiving appropriate signals from the nervous system, resulting in constant sympathetic stimulation; and, therefore, increased bowel tone. Because the intestine is in a contracted state, peristalsis does not occur.

Assessment

No Meconium Within 24 Hours

[No-sign Meconium-macaroni and 24hr-clock](#)

A newborn's first stool is called meconium. In a healthy newborn, meconium should be passed within the first 24 hours after birth. An infant that does not pass meconium within 24 to 48 hours after birth, or one that requires digital stimulation to pass meconium, should be evaluated for Hirschsprung's disease.

Refusal to Feed

[Refusing to Feed](#)

Neonates with Hirschsprung's disease may exhibit a feeding intolerance, or refusal to feed, with bilious vomiting.

Abdominal Distention

[Abdominal Distention](#)

Infants with this disease may have a distended abdomen due to the retention of stool.

No Stool on Rectal Exam

Clean Finger at Rectum-rectangle Exam

There may be no stool present on the examiner's finger after a digital rectal exam; however, the exam can stimulate the bowel and subsequently cause forceful expulsion of stool.

Forceful Expulsion of Fecal Material After Exam

Splattered Fecal Material

A digital rectal examination (DRE) may produce enough stimulation to cause forceful expulsion of fecal matter after the exam is complete.

Interventions

Surgical Resection

Surgeon

Surgical resection of the bowel is typically performed to remove the aganglionic section of the bowel, thus restoring motility and relieving obstruction. Anal stricture and incontinence may occur following a pull-through surgical procedure. Chronic problems of constipation and fecal incontinence may occur after surgery.

Colostomy

Colon-star-mouth

The patient may need a temporary colostomy after surgery is performed to resect the bowel. A colostomy will facilitate waste removal while the bowel heals. The earlier the diagnosis of the condition is made, the more likely the proximal bowel may not distend extremely. This allows for a primary pull-through (Soave) surgery and the elimination of needing to have a temporary colostomy.