

Introduction

Hirschsprung's disease (congenital megacolon) is caused by the failed migration of colonic ganglion cells during gestation. Varying lengths of the distal colon are unable to relax, causing functional colonic obstruction. Hirschsprung's disease most commonly involves the rectosigmoid region of the colon but can affect the entire colon and, rarely, the small intestine. The disease usually presents in infancy, although some patients present with persistent, severe constipation later in life. Symptoms in infants include difficult bowel movements, poor feeding, poor weight gain, and progressive abdominal distention. Early diagnosis is important to prevent complications (e.g., enterocolitis). A rectal suction biopsy can detect hypertrophic nerve trunks and the absence of ganglion cells in the colonic submucosa, confirming the diagnosis (*Parisi & Kapur, 2000*).

Hirschsprung's disease occurs in one out of 5,000 births. (*Amiel & Lyonnet, 2001*) The disease is caused by the failure of ganglion cells to migrate cephalocaudally through the neural crest during weeks four to 12 of gestation, causing an absence of ganglion cells in all or part of the colon Varying lengths of the distal colon are unable to relax, causing colonic obstruction over time. The aganglionic segment usually begins at the rectum and extends proximally. (*Feldman .,etal ,2002*)

Symptoms range from neonatal intestinal obstruction to chronic progressive constipation in older children. . Approximately 80 percent of patients present in the first few months of life with difficult bowel movements, poor feeding, and progressive abdominal distention. Typically 95% of normal term infants pass

meconium in the first 24 hours of life; less than 10% of children with Hirschsprung disease have passed meconium during that time (*Holschneider & Puri, 2000*) ; however, other causes of this delay also should be considered. (*Amiel & Lyonnet, 2001*).

After Hirschsprung's disease is diagnosed, surgery usually is needed. (*Coran & Teitelbaum, 2000*) Physicians should have a general knowledge of common procedures to help facilitate communication between the surgeon and the patient's family. In otherwise newborns with undistended colons and short-segment Hirschsprung's disease, the definitive transanal pull-through anastomosis can be performed (*Weidner & Waldhausen, 2003*). If the child has associated enterocolitis or a significantly dilated colon, a leveling colostomy can be placed for several months while the child recovers then pull-through procedure can be performed (*Langer., et al, 2003*).

There are several pull-through techniques, with complication rates ranging from 4 to 16 percent. Swenson's operation involves removing the rectum, pulling the healthy ganglionated colon through, and connecting it to the anus. (*Holschneider & Puri, 2000*) Other techniques (e.g., Duhamel operation, Soave operation) help preserve the nerve supply to the rectum and bladder. Rectal Dilatations are necessary for several months after the Soave operation; the patient's parents can do this at home. (*Hadidi , 2003*). The advent of laparoscopic primary one-stage pullthrough without the need for a covering enterostomy has completely revolutionised the management strategy of Hirschsprung's disease. All of these procedures have high success rates, and morbidity is minimal (*Saleh ., et al, 2004*).
