Hirschsprung’s disease

Hirschsprung’s disease

Genetic defects (e.g. RET , EDNRB, EDN3) can a ff ect the - migration of neural crest-derived intestinal neurones (neuro -), cristopathy), leading to aganglionosis and thickened nerve trunks in the distal bowel. There may be a family history . Agan - glionic bowel fails to relax, causing a functional obstruction. Aganglionosis extends from the an us to the sigmoid colon in 75%, the proximal colon in 15%, and the terminal ileum in 10% of cases. A transition zone lies between dilated, proximal, normal bo wel and narrow , distal aganglionic bowel. Neonatal Hirschsprung’s disease presents with delayed passage of meco - - nium, abdominal distension and bilious vomiting requiring resuscitation, gastric decompression, antibiotics and a bowel - washout. The diagnosis is made on a cot-side suction rectal - biopsy . A contrast enema may show the narrow aganglionic segment, a cone and dilated pr oximal bowel (Figure 18.15). Daily rectal washouts may allow a period of growth at home before surgery . If decompression fails, a stoma is fashioned using frozen section histopathology to identify ganglionic bowel. Definitive surgery removes the aganglionic segment

(b) (c)

and brings ganglionic bowel to the anus; Swenson, Duhamel, Yancey–Soave and transanal ‘pull-throughs’ are options. Most children achieve reasonable bowel control, but some have residual constipation, incontinence or episodes of enterocolitis.

Figure 18.15 Barium enema in an infant, showing a ‘transition zone’ in the proximal sigmoid colon between the dilated proximal normally innervated bowel and the contracted aganglionic rectum.

Hirschsprung’s disease

Genetic defects (e.g. RET , EDNRB, EDN3) can a ff ect the - migration of neural crest-derived intestinal neurones (neuro -), cristopathy), leading to aganglionosis and thickened nerve trunks in the distal bowel. There may be a family history . Agan - glionic bowel fails to relax, causing a functional obstruction. Aganglionosis extends from the an us to the sigmoid colon in 75%, the proximal colon in 15%, and the terminal ileum in 10% of cases. A transition zone lies between dilated, proximal, normal bo wel and narrow , distal aganglionic bowel. Neonatal Hirschsprung’s disease presents with delayed passage of meco - - nium, abdominal distension and bilious vomiting requiring resuscitation, gastric decompression, antibiotics and a bowel - washout. The diagnosis is made on a cot-side suction rectal - biopsy . A contrast enema may show the narrow aganglionic segment, a cone and dilated pr oximal bowel (Figure 18.15). Daily rectal washouts may allow a period of growth at home before surgery . If decompression fails, a stoma is fashioned using frozen section histopathology to identify ganglionic bowel. Definitive surgery removes the aganglionic segment

(b) (c)

and brings ganglionic bowel to the anus; Swenson, Duhamel, Yancey–Soave and transanal ‘pull-throughs’ are options. Most children achieve reasonable bowel control, but some have residual constipation, incontinence or episodes of enterocolitis.

Figure 18.15 Barium enema in an infant, showing a ‘transition zone’ in the proximal sigmoid colon between the dilated proximal normally innervated bowel and the contracted aganglionic rectum.

Hirschsprung’s disease

Genetic defects (e.g. RET , EDNRB, EDN3) can a ff ect the - migration of neural crest-derived intestinal neurones (neuro -), cristopathy), leading to aganglionosis and thickened nerve trunks in the distal bowel. There may be a family history . Agan - glionic bowel fails to relax, causing a functional obstruction. Aganglionosis extends from the an us to the sigmoid colon in 75%, the proximal colon in 15%, and the terminal ileum in 10% of cases. A transition zone lies between dilated, proximal, normal bo wel and narrow , distal aganglionic bowel. Neonatal Hirschsprung’s disease presents with delayed passage of meco - - nium, abdominal distension and bilious vomiting requiring resuscitation, gastric decompression, antibiotics and a bowel - washout. The diagnosis is made on a cot-side suction rectal - biopsy . A contrast enema may show the narrow aganglionic segment, a cone and dilated pr oximal bowel (Figure 18.15). Daily rectal washouts may allow a period of growth at home before surgery . If decompression fails, a stoma is fashioned using frozen section histopathology to identify ganglionic bowel. Definitive surgery removes the aganglionic segment

(b) (c)

and brings ganglionic bowel to the anus; Swenson, Duhamel, Yancey–Soave and transanal ‘pull-throughs’ are options. Most children achieve reasonable bowel control, but some have residual constipation, incontinence or episodes of enterocolitis.

Figure 18.15 Barium enema in an infant, showing a ‘transition zone’ in the proximal sigmoid colon between the dilated proximal normally innervated bowel and the contracted aganglionic rectum.

Revision #1
Created 2025-12-31 15:09:53 UTC by Omar Ayman
Updated 2025-12-31 15:09:53 UTC by Omar Ayman