

Failure to thrive

Constipation

Abdominal distention

Episodes of diarrhea and vomiting

Signs of enterocolitis

Explosive, watery diarrhea

Fever

Appears significantly ill

Childhood

Constipation

Ribbonlike, foul-smelling stools

Abdominal distention

Visible peristalsis

Easily palpable fecal mass

Undernourished, anemic appearance

In infants and children, the history is an important part of diagnosis and typically includes a chronic pattern of constipation. On examination, the rectum is empty of feces, the internal sphincter is tight, and leakage of stool and accumulated gas may occur if the aganglionic segment is short. To confirm the diagnosis, rectal biopsy is performed either surgically to obtain a full-thickness biopsy specimen or by suction biopsy for histologic evidence of the absence of ganglion cells.

Therapeutic Management