Emergency complications of Hirschsprung disease

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INTRODUCTION

Hirschsprung disease (HD) is a motor disorder of the colon caused by the failure of neural crest cells (precursors of enteric ganglion cells) to migrate completely during intestinal development. The resulting aganglionic segment of the colon fails to relax, causing a functional obstruction. In the majority of patients, the disorder affects a short segment of the distal colon and rectum, with a transition zone in the rectosigmoid. In other patients, the aganglionosis involves longer segments of the colon; in rare cases, the entire colon and parts of the small bowel may also be involved.

HD occurs in approximately 1 in 5000 live births, with an overall male:female ratio of 3:1 to 4:1. When the entire colon is involved, the sex ratio more nearly approaches 1:1 [1-3].

This topic review focuses on emergency complications of HD, the most important of which are mechanical intestinal obstruction and Hirschsprung-associated enterocolitis (HAEC). In most cases, mechanical obstruction and HAEC can be treated nonoperatively while the diagnostic work-up is completed. However, it is important to remember that patients with these presenting problems who do not improve with nonoperative treatment will likely need a laparotomy and diverting colostomy or ileostomy, depending on the level of the transition zone. Colonic volvulus, a rare complication, also is discussed. The pathogenesis, diagnosis, and clinical management of HD are presented separately. (See "Congenital aganglionic megacolon (Hirschsprung disease)".)

ACUTE OBSTRUCTION IN THE NEONATE

HD accounts for a substantial proportion of cases of neonatal bowel obstruction [4]. Neonates who become symptomatic during the first few days of life typically present with failure to pass meconium (present in approximately 95 percent of neonates with HD), abdominal distension and tenderness, poor feeding, vomiting (which may be bilious or feculent), and other signs of intestinal obstruction [5,6]. Their clinical condition may deteriorate rapidly unless they are treated promptly. Most newborns with HD can be resuscitated and stabilized by nonoperative means, allowing for a full diagnostic work-up and definitive elective surgery. Emergency surgery is seldom needed, and exploratory laparotomy in undiagnosed cases of HD is to be avoided. Those who present later in infancy have similar symptoms but are less likely to develop acute obstruction. (See "Congenital aganglionic megacolon (Hirschsprung disease)", section on 'Clinical features'.)

Occasionally, neonates and infants with undiagnosed HD may present with Hirschsprung-associated enterocolitis (HAEC), which is discussed below. The clinical presentation includes fever, lethargy, anorexia, vomiting, abdominal distension, and diarrhea; an explosion of gas and liquid stool may occur following digital rectal examination. Because HD is increasingly diagnosed in the first few months of life, fewer children present with HAEC than in the past. Most cases of HAEC now occur after a pull-through procedure, as discussed below. (See 'Enterocolitis' below.)

Diagnosis — The diagnosis of HD is suggested by a plain film of the abdomen, showing an airless rectum, a cone-shaped transition zone (the presence of which is diagnostic of HD), and dilation of the segment of colon immediately proximal to the aganglionic segment, best seen on lateral view. These findings are more clearly demonstrated by a barium or water-soluble contrast enema (image 1) (performed "unprepped," ie, without prior bowel evacuation), which should be done in any patient in whom the diagnosis of HD is suspected. The diagnosis should be confirmed by suction rectal biopsy. (See "Congenital aganglionic megacolon (Hirschsprung disease)", section on 'Evaluation'.)

In addition to these findings, infants with mechanical obstruction have marked bowel dilation, which may extend to the small bowel. Rarely, there may be evidence of perforation, which tends to occur in the cecum. Perforation is most likely to occur in patients with long-segment disease who do not improve with gastric drainage, rectal

irrigations, etc. Neonates and infants who present with intestinal perforation should be evaluated for HD by suction rectal biopsy or biopsy of the bowel during laparotomy. (See 'Treatment' below.)

The differential diagnosis of infants presenting with acute distal intestinal obstruction includes meconium ileus, intestinal atresia and stenosis, anorectal malformation, duplication cyst, meconium plug syndrome, and neonatal small left colon syndrome. Calcifications usually suggest meconium ileus or atresia but have also been reported with long-segment HD [7].

Treatment — Children with HD seldom need to be brought to surgery urgently, even when they present with acute obstruction or HAEC. Neonates and infants with acute obstructive symptoms, and any child presenting with HAEC, should be resuscitated and stabilized with intravenous fluids, nasogastric or orogastric suction, broad-spectrum antibiotics, twice-daily rectal irrigations, and daily digital rectal examinations. Once stabilized, the patient should have a full diagnostic work-up, including a contrast enema and rectal biopsy to establish the diagnosis of HD. It should be noted, however, that the rectal manipulation from digital examinations or irrigation may occasionally decompress the bowel so effectively that it can cause the contrast enema to appear normal in patients with HD. (See "Congenital aganglionic megacolon (Hirschsprung disease)", section on 'Evaluation'.)

By stabilizing the patient and fully establishing the diagnosis prior to surgery, newly diagnosed cases can be treated with a one-stage, definitive pull-through rather than a diverting colostomy. (See "Congenital aganglionic megacolon (Hirschsprung disease)", section on 'Management'.)

In a minority of neonates with HD, the clinical manifestations do not resolve with the nonoperative measures noted above. When abdominal distention and tenderness persist and plain films show persistent dilation of the bowel, especially in the cecum and ascending colon, surgical intervention is indicated to relieve the obstruction and prevent perforation. This will almost always entail a diverting colostomy or ileostomy, ideally guided by use of frozen section pathology to localize the transition zone by confirming the presence of ganglion cells. Failure to relieve the obstruction is also the main cause of HAEC. (See 'Enterocolitis' below.)

ENTEROCOLITIS

Hirschsprung-associated enterocolitis (HAEC) is the most severe and potentially lethal complication of HD [8]. With advances in technique and medical care during the past 50 years, mortality rates for HAEC have decreased from as high as 33 percent [9,10] to less than 1 percent [1,11]. The incidence of HAEC ranges from 24 to 34 percent in different series of patients with HD [12-14]. Failure to recognize HD in the early perinatal period increases the risk for HAEC [15].

HAEC can occur prior to surgical intervention (eg, in a neonate), in the immediate postoperative period, or even years after definitive repair [9,16]. Postoperative HAEC usually occurs soon after definitive pull-through repair, ranging from three weeks to 20 months postoperatively [9]. Case series report that postoperative HAEC develops in up to 30 percent of patients and is more common in those with Down syndrome [10,14]. In a cohort study using the Pediatric Health Information System (PHIS) database of infants with HD in the United States, 13 percent had at least one episode of HAEC after the initial surgical repair and 7 percent had recurrent episodes [17]. However, due to the limits of the PHIS, HAEC patients managed in outpatient settings or in facilities other than freestanding children's hospitals may not be captured in these estimates.

Clinical presentation — Patients usually present with explosive, foul-smelling diarrhea; fever; vomiting; and abdominal pain and distension. Mild cases may be misdiagnosed as gastroenteritis. Rectal bleeding and shock may be present in severe cases [9]. The American Pediatric Surgical Association (APSA) has developed a decision support tool for diagnosis and severity grading, based on history, physical examination, and imaging studies [18]. Patients are categorized by the presence of one or more of the following findings (table 1):

- Possible HAEC (also termed grade I) – Suggested by a history of anorexia or diarrhea, mild abdominal distension on examination, and abdominal images with a normal gas pattern or mild ileus.
- Definite HAEC (grade II) – Suggested by a history of explosive diarrhea, fevers, or lethargy; signs of tachycardia, abdominal distension and tenderness, and/or explosive release of gas and stool on rectal examination; and/or imaging showing ileus, air-fluid levels, and dilated bowel on imaging.

- Severe HAEC (grade III) – Suggested by a history of obstipation or obtundation; signs of hypovolemia, altered mental status, or peritonitis; and/or radiographs demonstrating pneumatosis intestinalis and/or pneumoperitoneum.

On plain abdominal films, nonspecific signs of ileus or obstruction are most common. Findings specifically associated with HAEC include distension of the proximal colon and the "cutoff sign," or absence of air in the distal rectosigmoid colon, with an abrupt cutoff at the level of the pelvic brim [9,19]. A combination of the cutoff sign and at least two air-fluid levels has been strongly associated with a diagnosis of HAEC [13]. Diarrhea, explosive stools, abdominal distension, and radiologic evidence of bowel obstruction or mucosal edema are the most important clinical predictors of HAEC [20].

In neonates, HD usually presents with signs and symptoms of mechanical obstruction. HAEC seldom occurs in neonates, except when the diagnosis of HD is missed or delayed. (See "Congenital aganglionic megacolon (Hirschsprung disease)", section on 'Suspected Hirschsprung disease in neonates'.)

A contrast enema should **not** be performed if HAEC is suspected, because of the risk of intestinal perforation [9]. Computed tomography (CT) has been described as showing thickening of the wall of the entire large bowel and terminal ileum, with enhancement of the mucosa [21].

Pathogenesis — The mechanisms underlying HAEC are not fully understood. Contributing factors include stasis and reduced production of protective mucin, which lead to bacterial overgrowth in the lumen of the bowel proximal to the involved segment, and possibly translocation of bacteria through the mucosa [22]. HAEC is associated with intestinal dysbiosis, with reduced commensal bacteria (eg, bifidobacteria and *Lactobacillus*); in some cases, pathogenic bacteria (eg, *Clostridioides* [formerly *Clostridium*] *difficile*) are isolated, and rotavirus and *Staphylococcus aureus* may also have a role [9,23-26]. The bowel wall in the affected area is subsequently invaded by colonic organisms, which can lead to pneumatosis intestinalis, intestinal perforation, peritonitis, systemic sepsis, shock, and death [23,27].

Pathologic examination of involved colonic tissue reveals neutrophils within the crypts of the mucosa, with mucin retention, abscess formation, and, in the most severe cases, transluminal necrosis and perforation [9]. Two forms of HAEC can be differentiated on pathologic examination of colonic tissue: inflammatory enterocolitis, involving the mucosa

only, and necrotizing or ischemic enterocolitis, involving the entire thickness of the bowel wall [19].

Risk factors — Factors associated with increased risk for **preoperative** HAEC include [9,12,14,19]:

- Delay in diagnosis of HD (>1 week of age)
- Increased length of the aganglionic segment
- Trisomy 21 (possibly caused by an underlying immune deficiency in these children)
- Presence of other associated anomalies

Factors associated with increased risk for **postoperative** HAEC include:

- Coexisting intestinal neuronal dysplasia (IND) [28] or hypoganglionosis [29] proximal to the resected aganglionic segment. The utility of resecting these proximal segments is not yet established. One study failed to show any advantage of resecting a longer margin of ganglionated bowel (>5 cm) [11], whereas another study described reduced rates of HAEC in patients in whom the IND-involved segment of the colon was resected in addition to the aganglionic segment as compared with historical controls [28].

- Younger age at resection (within the first six months of life) [11]. This finding contrasts with the increased risk for **preoperative** HAEC among neonates diagnosed after the first week of life.

- The presence of an anastomotic stricture or leak, or of any form of intestinal obstruction, has been associated with up to a 3.5-fold increase in the incidence of HAEC [10,23,30]. Intestinal obstruction also can be caused by an aganglionic segment if it is unrecognized and left in place.

- History of preoperative HAEC before their definitive surgery [14].

- Patients with two or more crypt abscesses per high-power field on histologic examination of colonic mucosa, in both ganglionic and aganglionic segments, also appear to be at increased risk [31]. An alteration in the intestinal mucus and the intestinal immunologic defense system of HD patients has been described and may contribute to susceptibility [9,27,32].

Management — Management of HAEC includes volume resuscitation and intravenous antibiotics, which should provide broad-spectrum coverage against aerobic and anaerobic organisms.

Preoperative Hirschsprung-associated enterocolitis — For patients with HAEC arising prior to the initial colonic resection for HD, the traditional treatment has been to create a diverting colostomy or ileostomy, usually without resection of the aganglionic bowel, as soon as the patient has been stabilized and the diagnosis has been established with a view to performing the definitive repair at a later date [33]. However, the trend is to resuscitate and stabilize the patient using the nonoperative measures noted above so that a definitive one-stage repair can be accomplished. Surgical resection and colostomy are indicated in the presence of ischemic bowel, frank perforation, or failure to improve with nonoperative measures, which include nasogastric or orogastric decompression, intravenous antibiotics, and rectal irrigations. (See 'Treatment' above.)

Postoperative Hirschsprung-associated enterocolitis — Most cases of postoperative HAEC can also be successfully managed nonoperatively.

A general guideline for management of HAEC has been proposed by the APSA (table 1) [18].

- Possible or mild HAEC (APSA grade I) – Patients should be treated with oral hydration and oral metronidazole, with or without rectal irrigations.
- Definite or moderate HAEC (APSA grade II) – These patients should generally be managed in an inpatient setting, with oral or intravenous hydration, metronidazole, broad-spectrum intravenous antibiotics, and repeated rectal irrigation through a rectal tube with saline; this decompresses the colon and often decreases the severity of the disease [5,9,23]. Occasionally, some patients with a relatively mild or indolent course may be managed as outpatients with metronidazole but with close follow-up.
- Severe HAEC (APSA grade III) – These patients should be managed with bowel rest, intravenous hydration, and broad-spectrum intravenous antibiotics, as well as rectal irrigations.

The rare patient that does not respond to this management may require urgent surgery to divert the fecal stream through a colostomy or ileostomy. Recurrent enterocolitis should

raise the suspicion of mechanical obstruction or a residual aganglionic segment [9].

Prevention — Some experts have advocated for routine use of rectal irrigations for the first six months after surgery in infants with HD who are at increased risk for developing HAEC, such as those with Down syndrome, long-segment HD, or prior episodes of HAEC [15]. Although this approach is not widely used, it is supported by some studies showing that rectal irrigations help to protect against the development of HAEC. In one study, rectal irrigations for six months after surgical repair significantly decreased the incidence and severity of HAEC compared with historical controls [34]. Similar results were found in a separate study that combined rectal irrigations with bowel decontamination with antimicrobial agents [35]. These studies were included in a meta-analysis that reviewed the effectiveness of strategies for management of obstructive symptoms and HAEC after surgical intervention. It concluded that routine postoperative irrigations significantly reduced the incidence of HAEC (relative risk [RR] 0.2, 95% CI 0.1-0.5) [36]. However, these findings must be interpreted with caution, given the paucity and low quality of the evidence, the temporal bias related to these reports encompassing several decades, and the heterogeneity of interventions. Moreover, irrigations represent a significant treatment burden for the patients and their families.

Anal dilatation regimens have also been proposed, based on the hypothesis that prevention of postoperative anastomotic strictures may decrease rates of HAEC and chronic constipation [37]. However, a meta-analysis concluded that routine anal dilatations did not reduce the incidence of postoperative strictures or HAEC [36,38]. Therefore, the value of routine anal dilatations as a preventive measure after surgery remains unclear.

Another approach that may reduce the short-term risk for HAEC is placement of a rectal tube for a few days after the endorectal pull-through operation. This was evaluated in a randomized multicenter study from China, which found that patients managed with a rectal tube for five days postoperatively were significantly less likely to develop HAEC during the first 30 postoperative days compared with those without a rectal tube (1.2 versus 6.8 percent, $p<0.05$) and also experienced less abdominal distention [39]. Similarly, some surgeons (including the senior author of this topic review) routinely leave a rectal tube in place postoperatively until the child passes stool, with favorable results.

Botulinum toxin injections have been used in the management of persistent obstructive symptoms after corrective surgery for HD. Relaxation of the internal anal sphincter may

improve obstructive symptoms and prevent fecal stasis, which may trigger the development of HAEC. In a systematic review and meta-analysis, botulinum toxin injections were found to be effective in treating obstructive symptoms but not helpful in reducing the occurrence of postoperative HAEC [40].

The administration of probiotics has also been proposed as a potential preventative strategy to reduce the incidence of postoperative HAEC. However, a meta-analysis of five studies, including two prospective multicenter randomized trials using four different strains, concluded that probiotics were not associated with a significant reduction in the risk of HAEC [41].

VOLVULUS

Volvulus is a rare complication of HD. It has been described in children and adults with previously undiagnosed short-segment HD [42-44] and occasionally in infants [45]. The volvulus usually involves the sigmoid colon and, less commonly, the transverse or cecal segments. The cause is thought to be related to torsion of an enlarged colonic segment filled with meconium or feces. Excessive mobility of the sigmoid mesentery may also have a role [46]. In one series, 2 of 11 consecutive children presenting with volvulus had underlying HD [47]. HD should be suspected in children presenting with otherwise unexplained colonic volvulus [43,46,47].

Clinical manifestations are similar to those seen in patients with volvulus caused by other conditions, including abdominal pain, distension, and vomiting (which may or may not be bilious). Typically, patients also have historical features, such as failure to pass meconium (in the neonate) or chronic constipation (in older children), suggesting the underlying disease.

The diagnosis is suggested on plain abdominal radiographs, which will reveal distended intestinal loops and air-fluid levels consistent with obstruction [46]. The diagnosis can be confirmed with a contrast enema, which may also detorse the volvulus. In the stable patient, the initial approach to detorsion may include contrast enema or colonoscopy with rectal tube placement. Intravenous fluids and antibiotics should be administered. Surgery is indicated if detorsion is unsuccessful, there are signs of peritonitis, or bowel necrosis or perforation is suspected.

INFORMATION FOR PATIENTS

UpToDate offers two types of patient education materials, "The Basics" and "Beyond the Basics." The Basics patient education pieces are written in plain language, at the 5th to 6th grade reading level, and they answer the four or five key questions a patient might have about a given condition. These articles are best for patients who want a general overview and who prefer short, easy-to-read materials. Beyond the Basics patient education pieces are longer, more sophisticated, and more detailed. These articles are written at the 10th to 12th grade reading level and are best for patients who want in-depth information and are comfortable with some medical jargon.

Here are the patient education articles that are relevant to this topic. We encourage you to print or e-mail these topics to your patients. (You can also locate patient education articles on a variety of subjects by searching on "patient info" and the keyword(s) of interest.)

- Basics topics (see "Patient education: Hirschsprung disease (The Basics)")

SUMMARY AND RECOMMENDATIONS

- Hirschsprung disease (HD) is a common cause of neonatal bowel obstruction. Neonates who become symptomatic during the first few days of life typically present with failure to pass meconium, abdominal distension, vomiting (which may be bilious or feculent), and other signs of intestinal obstruction. Most can be treated nonoperatively with gastric drainage and rectal irrigations while awaiting the results of the contrast enema and suction rectal biopsy. Rarely, surgery will be required when the obstruction does not improve. (See 'Acute obstruction in the neonate' above.)

- Hirschsprung-associated enterocolitis (HAEC) is the most severe and potentially lethal complication of HD. It can occur prior to surgical intervention (preoperative HAEC), in the immediate postoperative period, or long after definitive repair (postoperative HAEC). Young infants and those with Down syndrome are at increased risk for HAEC. (See 'Enterocolitis' above.)

- Patients with HAEC usually present with explosive, foul-smelling diarrhea; fever; vomiting; abdominal pain and distension; and sometimes rectal bleeding and shock.

Mild cases may be misdiagnosed as gastroenteritis. Therefore, HAEC should be considered in any child known to have HD presenting with symptoms suggestive of gastroenteritis. Radiographs show an obstructive picture, with air-fluid levels and dilated and edematous bowel loops. (See 'Clinical presentation' above.)

- Management of HAEC includes volume resuscitation and intravenous antibiotics. Repeated rectal irrigation with saline decompresses the colon and may decrease the severity of disease. (See 'Management' above.)
    - Infants presenting with HAEC prior to definitive surgery for HD should be stabilized and then treated with surgical resection of the aganglionic segment. Emergency surgery and a diverting ileostomy or colostomy are rarely necessary, except in the presence of ischemic bowel or a frank perforation, or failure to improve with nonoperative treatment.
    - Postoperative HAEC generally responds to antibiotics and irrigation and rarely requires surgical intervention. Patients with recurrent or refractory postoperative HAEC should be evaluated for mechanical obstruction or a residual aganglionic segment.
- Volvulus is a rare complication of previously undiagnosed short-segment HD. The cause is thought to be related to torsion of an enlarged colonic segment filled with meconium or feces. Clinical manifestations are similar to those seen in patients with volvulus caused by other conditions, including abdominal pain, distension, and vomiting. (See 'Volvulus' above.)

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REFERENCES

1. Suita S, Taguchi T, Ieiri S, Nakatsuji T. Hirschsprung's disease in Japan: analysis of 3852 patients based on a nationwide survey in 30 years. J Pediatr Surg 2005; 40:197.
2. Best KE, Addor MC, Arriola L, et al. Hirschsprung's disease prevalence in Europe: a

register based study. Birth Defects Res A Clin Mol Teratol 2014; 100:695.

3. Ieiri S, Suita S, Nakatsuji T, et al. Total colonic aganglionosis with or without small bowel involvement: a 30-year retrospective nationwide survey in Japan. J Pediatr Surg 2008; 43:2226.
4. Fitzgerald JF. Constipation in children. Pediatr Rev 1987; 8:299.
5. Pearl RH, Irish MS, Caty MG, Glick PL. The approach to common abdominal diagnoses in infants and children. Part II. Pediatr Clin North Am 1998; 45:1287.
6. Khan AR, Vujanic GM, Huddart S. The constipated child: how likely is Hirschsprung's disease? Pediatr Surg Int 2003; 19:439.
7. Cowles RA, Berdon WE, Holt PD, et al. Neonatal intestinal obstruction simulating meconium ileus in infants with long-segment intestinal aganglionosis: Radiographic findings that prompt the need for rectal biopsy. Pediatr Radiol 2006; 36:133.
8. Imamura A, Puri P, O'Briain DS, Reen DJ. Mucosal immune defence mechanisms in enterocolitis complicating Hirschsprung's disease. Gut 1992; 33:801.
9. Sherman JO, Snyder ME, Weitzman JJ, et al. A 40-year multinational retrospective study of 880 Swenson procedures. J Pediatr Surg 1989; 24:833.
10. Bill, AH, Chapman, ND. The enterocolitis of Hirschsprung's disease: Its natural history and treatment. Am J Surg 1962; 103:70.
11. Haricharan RN, Seo JM, Kelly DR, et al. Older age at diagnosis of Hirschsprung disease decreases risk of postoperative enterocolitis, but resection of additional ganglionated bowel does not. J Pediatr Surg 2008; 43:1115.
12. Teitelbaum DH, Qualman SJ, Caniano DA. Hirschsprung's disease. Identification of risk factors for enterocolitis. Ann Surg 1988; 207:240.
13. Elhalaby EA, Coran AG, Blane CE, et al. Enterocolitis associated with Hirschsprung's disease: a clinical-radiological characterization based on 168 patients. J Pediatr Surg 1995; 30:76.
14. Menezes M, Puri P. Long-term outcome of patients with enterocolitis complicating Hirschsprung's disease. Pediatr Surg Int 2006; 22:316.
15. Frykman PK, Short SS. Hirschsprung-associated enterocolitis: prevention and therapy. Semin Pediatr Surg 2012; 21:328.
16. Roorda D, Oosterlaan J, van Heurn E, Derikx JPM. Risk factors for enterocolitis in

patients with Hirschsprung disease: A retrospective observational study. J Pediatr Surg 2021; 56:1791.

17. Pruitt LCC, Skarda DE, Rollins MD, Bucher BT. Hirschsprung-associated enterocolitis in children treated at US children's hospitals. J Pediatr Surg 2020; 55:535.
18. Gosain A, Frykman PK, Cowles RA, et al. Guidelines for the diagnosis and management of Hirschsprung-associated enterocolitis. Pediatr Surg Int 2017; 33:517.
19. Blane CE, Elhalaby E, Coran AG. Enterocolitis following endorectal pull-through procedure in children with Hirschsprung's disease. Pediatr Radiol 1994; 24:164.
20. Pastor AC, Osman F, Teitelbaum DH, et al. Development of a standardized definition for Hirschsprung's-associated enterocolitis: a Delphi analysis. J Pediatr Surg 2009; 44:251.
21. Sheth J, Nour S, Dickinson F. CT images of Hirschsprung's associated enterocolitis: a rare finding. Arch Dis Child 2009; 94:816.
22. Mattar AF, Coran AG, Teitelbaum DH. MUC-2 mucin production in Hirschsprung's disease: possible association with enterocolitis development. J Pediatr Surg 2003; 38:417.
23. SWENSON O, NEUHAUSER EB, PICKETT LK. New concepts of the etiology, diagnosis and treatment of congenital megacolon (Hirschsprung's disease). Pediatrics 1949; 4:201.
24. Shen DH, Shi CR, Chen JJ, et al. Detection of intestinal bifidobacteria and lactobacilli in patients with Hirschsprung's disease associated enterocolitis. World J Pediatr 2009; 5:201.
25. Hardy SP, Bayston R, Spitz L. Prolonged carriage of Clostridium difficile in Hirschsprung's disease. Arch Dis Child 1993; 69:221.
26. Pierre JF, Barlow-Anacker AJ, Erickson CS, et al. Intestinal dysbiosis and bacterial enteroinvasion in a murine model of Hirschsprung's disease. J Pediatr Surg 2014; 49:1242.
27. Teich S, Schisgall RM, Anderson KD. Ischemic enterocolitis as a complication of Hirschsprung's disease. J Pediatr Surg 1986; 21:143.
28. Estevão-Costa J, Fragoso AC, Campos M, et al. An approach to minimize postoperative enterocolitis in Hirschsprung's disease. J Pediatr Surg 2006; 41:1704.
29. Wu X, Feng J, Wei M, et al. Patterns of postoperative enterocolitis in children with

Hirschsprung's disease combined with hypoganglionosis. J Pediatr Surg 2009; 44:1401.

30. Hackam DJ, Filler RM, Pearl RH. Enterocolitis after the surgical treatment of Hirschsprung's disease: risk factors and financial impact. J Pediatr Surg 1998; 33:830.
31. Wilson-Storey, D. Microbial studies of enterocolitis in Hirschsprung's disease. J Pediatr Surg 1994; 9:248.
32. Elhalaby EA, Teitelbaum DH, Coran AG, Heidelberger KP. Enterocolitis associated with Hirschsprung's disease: a clinical histopathological correlative study. J Pediatr Surg 1995; 30:1023.
33. Ghosh A, Griffiths DM. Rectal biopsy in the investigation of constipation. Arch Dis Child 1998; 79:266.
34. Marty TL, Seo T, Sullivan JJ, et al. Rectal irrigations for the prevention of postoperative enterocolitis in Hirschsprung's disease. J Pediatr Surg 1995; 30:652.
35. Núñez R, Torres A, Agulla E, et al. [Rectal irrigation and bowel decontamination for the prevention of postoperative enterocolitis in Hirschsprung's disease]. Cir Pediatr 2007; 20:96.
36. Soh HJ, Nataraja RM, Pacilli M. Prevention and management of recurrent postoperative Hirschsprung's disease obstructive symptoms and enterocolitis: Systematic review and meta-analysis. J Pediatr Surg 2018; 53:2423.
37. Pontarelli EM, Ford HR, Gayer CP. Recent developments in Hirschsprung's-associated enterocolitis. Curr Gastroenterol Rep 2013; 15:340.
38. Aworanti O, Hung J, McDowell D, et al. Are routine dilatations necessary post pull-through surgery for Hirschsprung disease? Eur J Pediatr Surg 2013; 23:383.
39. Zhang X, Li L, Li SL, et al. Primary laparoscopic endorectal pull-through procedure with or without a postoperative rectal tube for hirschsprung disease: a multicenter perspective study. J Pediatr Surg 2020; 55:381.
40. Roorda D, Abeln ZA, Oosterlaan J, et al. Botulinum toxin injections after surgery for Hirschsprung disease: Systematic review and meta-analysis. World J Gastroenterol 2019; 25:3268.
41. Nakamura H, Lim T, Puri P. Probiotics for the prevention of Hirschsprung-associated enterocolitis: a systematic review and meta-analysis. Pediatr Surg Int 2018; 34:189.
42. DEAN GO, MURRY JW. Volvulus of the sigmoid colon. Ann Surg 1952; 135:830.

43. Neilson IR, Youssef S. Delayed presentation of Hirschsprung's disease: acute obstruction secondary to megacolon with transverse colonic volvulus. J Pediatr Surg 1990; 25:1177.

44. Tan FL, Tan YM, Heah SM, Seow-Choen F. Adult Hirschsprung's disease presenting as sigmoid volvulus: a case report and review of literature. Tech Coloproctol 2006; 10:245.

45. Khalayleh H, Koplewitz BZ, Kapuller V, et al. Neonatal sigmoid volvulus. J Pediatr Surg 2016; 51:1782.

46. Sarioğlu A, Tanyel FC, Büyükpamukçu N, Hiçsönmez A. Colonic volvulus: a rare presentation of Hirschsprung's disease. J Pediatr Surg 1997; 32:117.

47. Venugopal KS, Wilcox DT, Bruce J. Hirschsprung's disease presenting as sigmoid volvulus in a newborn. Eur J Pediatr Surg 1997; 7:172.

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