Gastrointestinal Motility Disorders in Children

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Abstract: The most common and challenging gastrointestinal motility disorders in children include gastroesophageal reflux disease (GERD), esophageal achalasia, gastroparesis, chronic intestinal pseudo-obstruction, and constipation. GERD is the most common gastrointestinal motility disorder affecting children and is diagnosed clinically and treated primarily with acid secretion blockade. Esophageal achalasia, a less common disorder in the pediatric patient population, is characterized by dysphagia and treated with pneumatic balloon dilation and/or esophagomyotomy. Gastroparesis and chronic intestinal pseudo-obstruction are poorly characterized in children and are associated with significant morbidity. Constipation is among the most common complaints in children and is associated with significant morbidity as well as poor quality of life. Data on epidemiology and outcomes, clinical trials, and evaluation of new diagnostic techniques are needed to better diagnose and treat gastrointestinal motility disorders in children. We present a review of the conditions and challenges related to these common gastrointestinal motility disorders in children.

Keywords
Gastrointestinal motility, children, achalasia, gastroparesis, gastroesophageal reflux disease, constipation, intestinal pseudo-obstruction

Gastrointestinal symptoms, many resulting from gastrointestinal dysmotility, are among the most common complaints in children. Gastroesophageal reflux disease (GERD), the most common motility disorder affecting children, is diagnosed clinically and treated primarily with acid secretion blockade. Surgery for GERD in children should be reserved for those with life-threatening complications. Esophageal achalasia, an uncommon disorder in children, is characterized by dysphagia, diagnosed by absent peristalsis, and treated with pneumatic balloon dilation and/or esophagomyotomy in accordance with physician experience and patient preference. Gastroparesis is poorly defined in children and associated with limited therapeutic options due to the lack of effective therapies and the potential adverse effects of most prokinetic agents. Chronic intestinal pseudo-obstruction (CIPO) is at the end of the spectrum of intestinal dysmotility and is also poorly characterized in children. Effective

treatments for pediatric CIPO are very limited, resulting in significant morbidity and mortality. Constipation is among the most common complaints in children and is associated with significant morbidity and poor quality of life, at times requiring intensive behavioral and/or medical therapy.

Gastroesophageal Reflux Disease

Gastroesophageal reflux, the passage of gastric contents into the esophagus, is a normal physiologic process; pathologic gastroesophageal reflux, or GERD, is a condition in which gastroesophageal reflux causes symptoms (frequent heartburn, regurgitation, and/or vomiting) and complications (esophagitis, strictures, and/or extraintestinal manifestations). GERD may be caused by mechanical factors, such as the increased frequency of transient lower esophageal sphincter (LES) relaxations or the presence of hiatal hernia or delayed gastric emptying, or by other factors, such as increased gastric acid secretion or overeating.

Evaluation

The diagnosis of GERD is clinical in the majority of patients and noted by the presence of classic symptoms to an extent that justifies the initiation of medical therapy. Diagnostic tests are typically reserved for patients whose symptoms do not respond to medical therapy and are used to evaluate patients for complicating factors and to rule out other diagnoses, such as eosinophilic esophagitis and *Helicobacter pylori* gastritis. Contrast imaging is useful in the evaluation of persistent vomiting in infants and children to assess for anatomic causes of symptoms, such as intestinal malrotation, achalasia, or hiatal hernia, and to evaluate for complications of GERD, such as peptic strictures.

Upper gastrointestinal endoscopy is recommended to evaluate for mucosal disease that may explain symptoms such as eosinophilic esophagitis and to evaluate for potential complications associated with GERD. Esophageal manometry (EM) is mainly used to rule out esophageal motor disorders and is discussed further in the section on esophageal achalasia. Esophageal pH monitoring and multiple intraluminal impedance combined with pH monitoring are useful in the evaluation of effective acid suppression and symptom correlation. Gastric emptying scintigraphy is reserved for patients with symptoms refractory to conventional therapy who may benefit from the use of prokinetic agents to accelerate the emptying of stomach contents into the small bowel.

Treatment

Lifestyle changes, such as avoidance of spicy and acidic foods, bed elevation, and weight loss, are the first line of therapy recommended for GERD despite the lack of significant evidence of benefit. Ample data support the use of acid secretion blockade, with most clinical trials demonstrating the superiority of proton pump inhibitors (PPIs) over H2 blockers and placebo. Uncontrolled studies have shown the efficacy of prolonged use of PPIs in healing esophagitis; however, prolonged use has recently been associated with increased respiratory and gastrointestinal infections in children[1] and an increased risk of fractures in adults.[2,3] Surgical procedures, such as fundoplication, are associated with higher symptom resolution compared with PPIs but are no different from PPI therapy in controlling esophagitis[4] and preventing adenocarcinoma.[5] Given this lack of clear superior benefit of surgery over medical therapy and the potential complications associated with fundoplication (particularly a higher incidence of complications and a lower survival rate in children with cerebral palsy[6-8]), we believe that surgery should be reserved for those with life-threatening complications of GERD.

Esophageal Achalasia

Esophageal achalasia is an uncommon disorder with an incidence of 0.18/100,000 pediatric cases per year,[9] a rate that has been increasing over the past 2 decades.[9,10] Esophageal achalasia has equal gender predilection and a mean patient age at diagnosis of 10.9 years,[9] with a higher incidence in adolescents and few reported cases in infants.[9,11,12]

Epidemiology

The clinical presentation of this condition varies with age.[13,14] Younger children frequently present with vomiting and respiratory symptoms,[13,15,16] whereas older children present with dysphagia, vomiting, and regurgitation.[13] Dysphagia progresses from solids to liquids in 70% of patients[13] and results in significant weight loss. Weight loss due to dysphagia may be confused with weight loss due to eating disorders and may lead to a delay in diagnosis and treatment.[17,18] Genetic disorders associated with achalasia include Allgrove or Triple A syndrome (achalasia, adrenal insufficiency, and alacrima), Alport syndrome, and Down syndrome.[16,19,20] The pathogenesis of esophageal achalasia is unknown, and proposed causes include decreased nitric-oxide synthase–containing nerve fibers and interstitial cells of Cajal in the distal esophagus.[21-23]

Evaluation

EM is the gold standard for the diagnosis of esophageal achalasia. Hallmark findings are absent esophageal peristalsis with abnormal LES resting pressure and relaxation. High-resolution manometry allows for easier study in children and therapy stratification in adults.[24,25] (Type 2, or panesophageal pressurization, demonstrates better response to therapy than type 1, or none/minimal esophageal pressurization, and type 3, achalasia with

distal esophageal spasm with or without pressurization.[24,25]) Such information is not available for children. Morera and Nurko reported difficulty in interpreting EM findings in 27.6% to 34.5% of children demonstrating heterogeneous LES parameters (resting pressure, residual pressure, relaxation, and duration of relaxation).[26] Barium esophagram may show the classic "bird beak" appearance of the distal esophagus, with proximal dilation and air-fluid levels.[27] Upper endoscopy may reveal a dilated esophagus and retained food products, but findings are often normal and of limited diagnostic utility.

Treatment

The goal of treatment is to facilitate bolus transfer by decreasing LES pressure.[14,19] Treatment options include pharmacotherapy, endoscopic LES botulinum toxin (BT) injection, pneumatic balloon dilation (PD), and surgical myotomy (with or without fundoplication). Randomized prospective studies evaluating the long-term efficacy of these treatments in children with esophageal achalasia are needed. Pharmacologic therapies (nitrates, calcium channel blockers, and sildenafil) are used in adults and rarely in children.[28] Hurwitz and colleagues reported an 83% response rate among children receiving BT.[29] The duration of the effect was 4.2 months, with more than 50% of those responders requiring additional procedures 7 months after receiving BT.[29] Besides its use as a diagnostic aid, BT is only recommended for those who are considered high-risk patients for anesthesia and surgery. The overall success rate for PD ranges from 70% to 90%.[30,31] Although significant short-term efficacy of PD has been reported, long-term efficacy data in children are lacking. A Cochrane review of adults demonstrated that PD is superior to BT in symptom remission at 6 and 12 months.[32] A recent meta-analysis demonstrated remission and relapse rates of 77.8% and 35.7%, respectively, for PD compared with 95% and 5.1%, respectively, for laparoscopic myotomy.[33] The surgical technique used in children with esophageal achalasia is largely center-dependent, with most studies reporting significant improvement[34-36] and low complication and recurrence rates[15,37]; however, long-term data are not available.

Gastroparesis

Gastroparesis is scintigraphically characterized by delay in gastric emptying associated with upper gastrointestinal symptoms in the absence of mechanical obstruction.

Epidemiology

Most of the mechanisms associated with gastric emptying gradually mature with gestational age, with the presence of the gastric emptying function emerging as early as age 24 weeks and a normal pattern of gastric emptying occurring at around age 32 weeks; hence, delayed gastric emptying is a common occurrence in premature infants. The most common symptoms of gastroparesis in children include vomiting (42%-68%), abdominal pain (35%-51%), and nausea (28%-29%).[38,39] Children commonly present with vomiting, whereas adolescents primarily report nausea and abdominal pain. These symptoms appear to have a male predominance in infancy and a female predominance in adolescence.[38] In 2 large pediatric series, no cause was found in up to 70% of cases; gastroparesis was associated with viral gastroenteritis (18%), medications (18%), surgical procedures (12.5%), mitochondrial disease (8%), and diabetes mellitus (2%-4%).[38,39]

Evaluation

Gastric emptying scintigraphy demonstrating a 10% or greater retention of solids at 4 hours is diagnostic for gastroparesis in adults.[40] Most pediatric institutions define delayed gastric emptying as 60% or greater retention at 1 hour or a gastric emptying half-time of greater than 90 to 100 minutes. These institutions use their own standards, given the lack of protocol standardization and pediatric normative data. However, recent evidence suggests that adult standards can be applied to the pediatric population.[41] Breath testing, in which 13C is used to label the meal substrate and the exhalation of 13C in breath over time reflects the emptying of the substrate from the stomach, has been used as a noninvasive and nonradioactive alternative to scintigraphy. The half-emptying of 13C-sodium acetate correlates with scintigraphy findings in children with gastroparesis symptoms[42,43] and discriminates between healthy volunteers and children with gastroparesis symptoms.[42] Antroduodenal manometry (ADM) can be used as an adjunct in the evaluation of gastroparesis and may demonstrate abnormal antral contractions during fasting and antral postprandial hypomotility in children with postviral[44] and diabetic gastroparesis.[45] Recently, a wireless motility capsule (SmartPill, Given Imaging), which simultaneously measures pressure and transit, has shown a good correlation with scintigraphy and can reliably identify gastroparesis in adults.[46] Validation studies in children are underway (Figure 1).

Treatment

Oral nutritional support is recommended in patients with gastroparesis. If the oral route is not tolerated, nutritional support should be delivered via an enteral tube. Despite a lack of association with symptom improvement, prokinetic agents are used to accelerate gastric emptying.[47] Response to prokinetic agents has been reported in up to 55% of children with gastroparesis.[38] Erythromycin, the most commonly used prokinetic agent, is a moti-

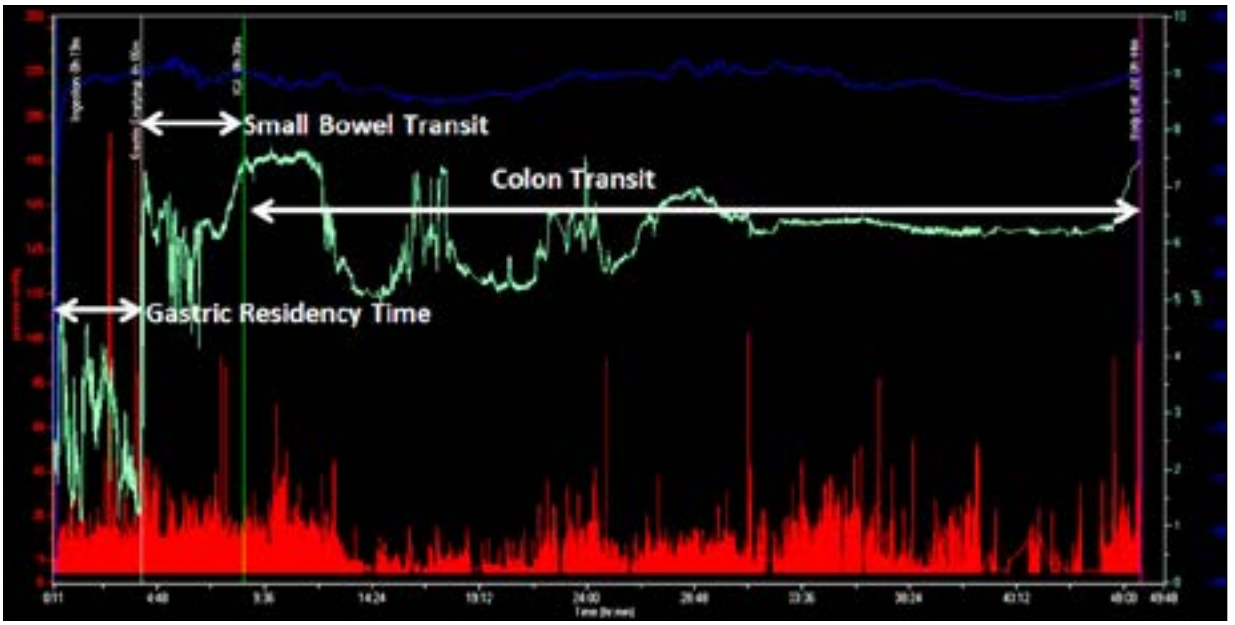

Figure 1. A trace of a normal wireless motility capsule study in a 10-year-old girl with nausea and vomiting. The green line represents pH, the blue line represents temperature, and the red line represents pressure. Note that gastric emptying occurs in less than 4 hours, small bowel transit occurs in less than 5 hours, and colon transit occurs in less than 40 hours. The capsule exits the body in 48 hours.

lin receptor agonist that stimulates gastric emptying, increases the amplitude of antral contractions, induces phase III of the migrating motor complex (MMC), and improves antroduodenal coordination.[47-50] A systematic review of adults with gastroparesis showed that, compared with other prokinetic agents, erythromycin significantly improved symptoms and gastric emptying[47]; however, in a recent report in children, erythromycin demonstrated low efficacy.[38] Given its good safety profile, erythromycin is recommended as a first-line prokinetic agent. Erythromycin, however, has been associated with QT interval prolongation and cardiac arrhythmias, especially when used in conjunction with CYP3A isozyme inhibitors.[51,52] Prolonged use of erythromycin may result in tachyphylaxis that can be overcome by cycling therapy.[53]

Other macrolides, such as azithromycin, have been shown to improve gastric emptying and antral motility patterns in adults,[54-56] but such data are not available in children. Cisapride and tegaserod are serotonin agonists that improve gastric emptying and antral/small intestinal motility and coordination.[53,57-60] Both, however, were withdrawn from the US market due to QT prolongation, cardiac arrhythmias, and sudden death. Cisapride is available in a limited access program. Metoclopramide and domperidone are dopamine receptor antagonists with antiemetic and prokinetic properties. Metoclopramide has not demonstrated significant symptom improvement[38] and is not recommended for long-term use due to an increased risk of central nervous system adverse effects (acute dystonic reactions and irreversible tardive dyskinesia).[53] Domperidone improves gastric emptying[61,62] and symptoms in children.[38] The agent was reported to be superior to cisapride in children with diabetic gastroparesis.[63] Domperidone has a better neurologic safety profile than metoclopramide,[64] but it also has been associated with prolonged QT, cardiac arrhythmias, and sudden death.[65,66] It is not approved for use in the United States.

Endoscopic pyloric BT injection has been reported to improve symptoms and gastric emptying in uncontrolled open-labeled adult studies,[67] but 2 placebo-controlled trials did not find sufficient evidence to support its use.[68,69] Rodriguez and colleagues reported an overall 67% response rate for BT in children, with a median duration of 3 months with no significant adverse effects.[70] Older age and vomiting were predictive of response to the initial injection, and male sex predicted response to repeated injections. The use of BT should be limited to those patients who fail medical therapy before invasive surgical interventions are considered.

Gastric electrical stimulation has emerged as an alternative therapy for medically refractory cases. Long-term follow-up studies report improvements in symptoms, quality of life, length of hospital stay, and medication use[71] with no significant change in gastric emptying. The use of gastric electrical stimulation in children is limited to small case series that report improvements in symptoms.[72,73] Short- and long-term outcomes and safety profiles in children remain to be elucidated.

Outcome

Waseem and colleagues reported symptom improvement in 60% of pediatric patients at 2-year follow-up, with the greatest improvement seen in adolescents.[39] Rodriguez and colleagues reported resolution of symptoms in 52% of patients, with 22% reporting resolution at 6 months, 53% at 18 months, and 61% at 36 months.[38] Younger age and response to prokinetics were associated with eventual resolution of symptoms in contrast to longer duration of symptoms, presence of mitochondrial dysfunction, and older age.[38]

Chronic Intestinal Pseudo-Obstruction

CIPO is a rare disorder with significant morbidity and mortality. It is characterized by severe and disabling repetitive episodes or continuous symptoms and signs of bowel obstruction, including radiographic evidence of dilated bowel with air-fluid levels, in the absence of a fixed, lumen-occluding lesion.[74]

Epidemiology

CIPO is classified as a primary or secondary cause of gastrointestinal dysmotility. Primary CIPO is further subclassified into neuropathic, myopathic, or idiopathic causes. Secondary CIPO is associated with a myriad of systemic disorders, including metabolic disorders, mitochondrial myopathies, muscular dystrophies, diseases of the nervous system, endocrinopathies, and connective tissue disorders. The diagnosis of CIPO is made in utero in approximately 16% of patients,[75] in the neonatal period in 55% to 67% of patients,[75,76] and within the first year of life in 76% of patients.[76] The most common symptoms are abdominal distention (98%), vomiting (91%; bilious in 80%), abdominal pain (58%-70%), failure to thrive (62%), diarrhea (31%-42%), constipation (42%-77%), feeding intolerance (39%), and urinary symptoms (11%).[75-78]

Urologic abnormalities and malrotation are the most common associated conditions of CIPO. Urologic involvement is present in up to 44% of children with CIPO.[75-78] Megacystis and megaureter associated with recurrent urinary tract infections develop in 32% of patients.[75-77] Malrotation may be present in 28% to 36% of patients in whom symptoms persist despite surgical correction.[75,77]

Evaluation

The diagnosis of CIPO is clinical, and the initial work-up should aim at ruling out conditions that mimic CIPO, such as mechanical obstruction,[79] pain-associated disability syndromes, and Munchausen by proxy (medical child abuse). Transit studies may help establish the degree and extent of gastrointestinal dysfunction that can be confirmed with manometry studies, including EM, ADM, colonic manometry (CM), and anorectal manometry (ARM). EM is abnormal in most adults with CIPO but is not specific for CIPO. Common abnormalities seen in ADM include abnormal or absent MMC and fed response.[59,79-81] ADM findings in children with CIPO have been associated with prognostic outcomes. Low-amplitude phase III MMCs with a low motility index are associated with dependence on parenteral nutrition (PN) and higher mortality.[82] Normal intestinal phase III of the MMC is a positive predictor of tolerance of jejunal feeds,[83] whereas its absence has been associated with an increased need for PN support and decreased response to cisapride.[60] A normal ADM study in the presence of symptoms should raise concern for other conditions such as pain amplification disorders and Munchausen by proxy[80,84] (Figure 2).

Treatment

A multidisciplinary approach that includes primary care clinicians, gastroenterologists, surgeons, dieticians, social workers, and mental health providers is recommended. Appropriate nutritional support remains the cornerstone of therapy. When appropriate, oral, gastric, or jejunal feeds should be used. Trophic feeds are recommended despite PN dependence, and the inability to tolerate enteral feeds necessitates the initiation of PN. Approximately two-thirds of children with CIPO require PN, and a quarter of these become PN-dependent.[75,76] Factors associated with the need for PN include neonatal presentation, acute onset, association with megacystis, and a history of surgical interventions.[76] PN-associated complications (hepatic failure, central line infections, and thromboembolic events) are a significant determinant of morbidity and mortality.[75-77]

The use of prokinetic agents in the management of patients with CIPO is limited. Cisapride is the only prokinetic agent that has been shown to improve enteral tolerance.[85] Erythromycin induces intestinal phase III of the MMC and antroduodenal coordination,[48,49] but its efficacy in CIPO has not been evaluated. The use of metoclopramide and domperidone has been limited due to their neurologic and cardiac adverse effects. Octreotide, a somatostatin analogue that induces phase III MMCs in the small intestine,[86] has been shown to benefit adults with scleroderma-associated CIPO.[87,88] Its use in children is limited to small case reports.[89]

Up to 68% of patients with CIPO have been reported to undergo surgical procedures,[76] including gastrostomy (38%-73%), ileostomy (25%-50%), fundoplication (19%-25%), colostomy (6%-16%), and jejunostomy (3%).[76-78] Gastrostomies and jejunostomies are used to provide continuous feeds distally and to vent dilated stomach and bowel. Ileostomies and colostomies are primarily used to decompress the bowel by decreasing distal

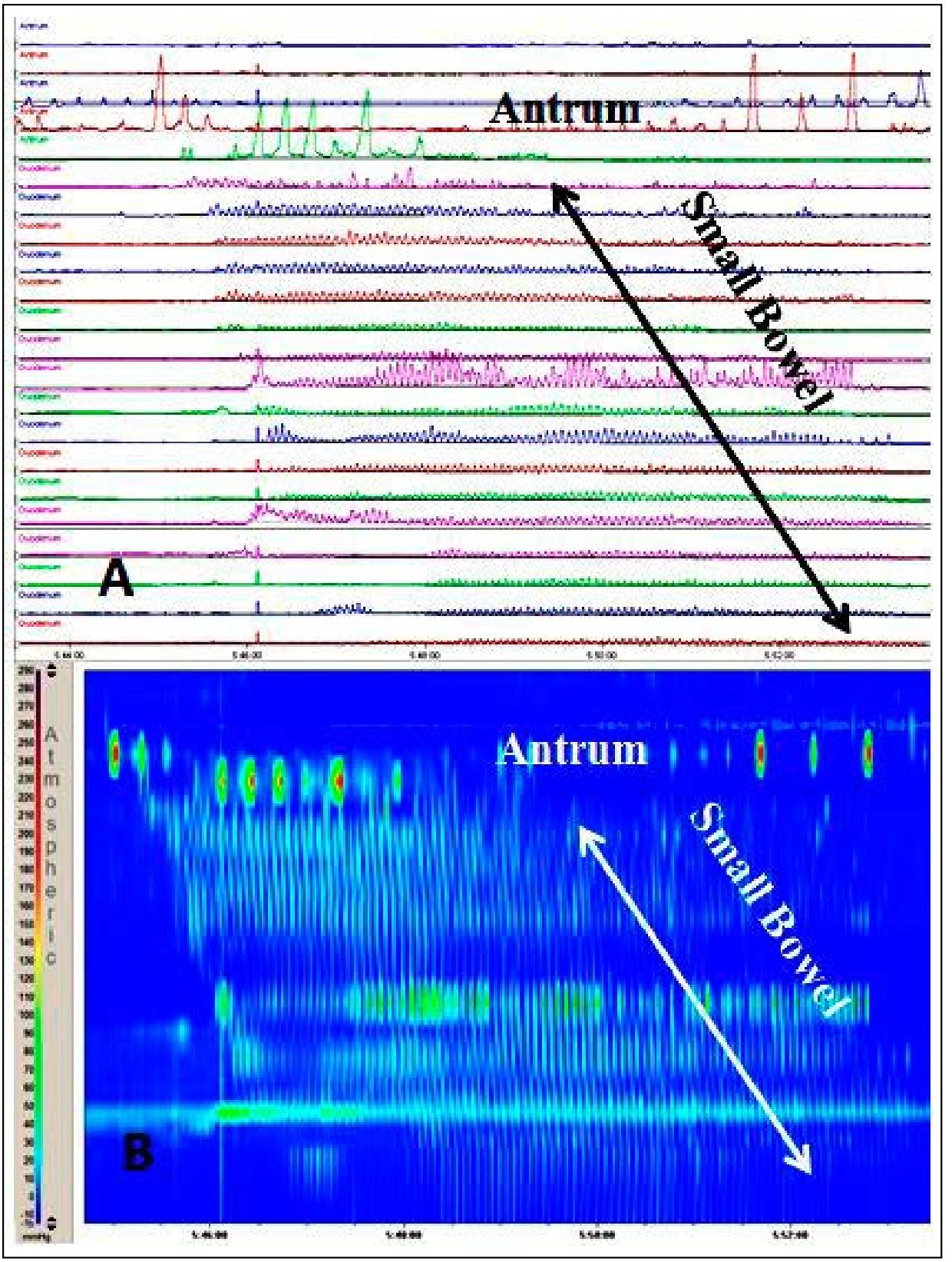

Figure 2. Tracings of a normal high-resolution antroduodenal motility study in a 6-year-old boy with feeding intolerance. **A:** A conventional tracing. **B:** The same study in a 2-dimensional colored topographic tracing. Note that phase 3 of the migrating motor complex starts in the antrum and migrates to the distal duodenum in both tracings.

resistance with proximal diversion of the fecal stream[90] and to potentially minimize bacterial translocation from increased intraluminal pressure.

Intestinal transplantation has emerged as a potentially curative intervention for intestinal failure. An early intestinal transplantation evaluation is recommended for patients with the PN complications mentioned above.[79,91] Multivisceral transplantation survival rates of 66.7% at 1 year and 50% at 3 years have been reported in children with CIPO.[92]

Outcome

Most children will require some form of nutritional support, and the overall mortality rate has been reported to be between 10% and 32%.[75-77]

Intractable Constipation

Constipation is among the most common complaints in children, with a worldwide prevalence of 0.7% to 29.6%.[93] Constipation accounts for 3% of pediatrician visits and 25% of referrals to pediatric gastroenterologists.[94] Intractable constipation (IC) refers to constipation that is refractory to conventional treatment, such as stool softeners and laxatives.

Evaluation

A detailed history and physical examination should guide the evaluation of IC for underlying organic diseases and should direct the appropriate treatment. Organic causes (neurologic, anatomic, metabolic, neuroenteric, gastrointestinal, and toxic)[95] are responsible for less than 10% of childhood constipation.[93] Delayed passage of meconium (>24-48 hours of life), history of enterocolitis, and acute intestinal obstruction are suggestive of Hirschsprung disease (HD). In addition, up to 30% of children with chromosomal abnormalities (eg, trisomy 21), anorectal malformations, and Waardenburg syndrome have been associated with HD.[96] ARM is used primarily to evaluate the presence of the rectoanal inhibitory reflex. When absent, a rectal suction biopsy should be performed to rule out HD by confirming the presence of ganglion cells and normal acetylcholinesterase staining in the lamina propria. Equivocal results must be confirmed with a surgical full-thickness rectal biopsy. Barium enema may be used to delineate the transition zone; however, it is not diagnostic, and normal findings do not exclude HD. A nonrelaxing internal anal sphincter (IAS) on ARM with normal rectal biopsies is diagnostic of IAS achalasia.

Neurologic lesions, such as spinal dysraphism, spinal cord lesions, and tethered cord have been reported in up to 9% of pediatric patients with IC.[97,98] Progressive neuromuscular deficits, abnormal gait, back pain, and new onset of fecal and urinary incontinence may be among the presenting symptoms. ARM may show abnormal sphincter tone, prolonged IAS relaxation and/or abnormal recovery with sustained balloon inflation, and anal spasms.[99,100] The presence of anal spasms has been shown to be predictive of spinal abnormalities in 60% of children with IC.[100]

The radiopaque marker (ROM) study is the simplest, most readily available method to evaluate colonic transit. Its use in children is limited by a lack of protocol standardization and normative pediatric data. Pediatric patients should be screened with a ROM study before more invasive studies, such as CM, are undertaken. A normal ROM study correlates with normal CM findings, whereas an abnormal ROM study does not correlate with abnormal CM findings. Therefore, CM can be avoided in patients with a normal ROM study.[101] CM evaluates gastrocolonic response to a meal and the presence of fasting, a meal, or bisacodyl-induced high-amplitude peristaltic contractions (HAPC). Abnormalities in CM include abnormal gastrocolonic and/or abnormal HAPC amplitude or propagation. Colonic dysfunction may be segmental or may involve the entire colon. A lack of gastrocolonic response and absence of HAPC are indicative of colonic inertia[81] (Figure 3).

Treatment

A diet rich in fiber is recommended, although dietary fiber has a limited role in the therapy of IC. Lubiprostone (Amitiza, Takeda) and stimulant laxatives such as bisacodyl are among the medications used in IC. Lubiprostone, a chloride channel-2 agonist, enhances intestinal fluid secretion, thereby facilitating intestinal motility. It is approved for chronic idiopathic constipation and irritable bowel syndrome with constipation in adults. Information of its use in children is lacking. In our experience, lubiprostone has been useful as an adjunct to other stool softeners and stimulant laxatives and for patients with fecal incontinence due to spinal or anorectal abnormalities. Bisacodyl has been reported to increase colonic emptying/transit in healthy adults[102] as well as stool frequency and quality of life at 4 weeks compared with placebo in adults with constipation,[103] although there are reports of association with ischemic colitis. No information of long-term use is available in children.

IAS BT injection has been reported to be safe and effective and is the treatment of choice for IAS achalasia in children,[104-106] with a short-term clinical efficacy rate of 88.3%, long-term efficacy rate of 65.1%, and a mean sustained response of 17 months.[106] Transient fecal incontinence is reported in 9.5% to 21% of patients.[105,106] A recent meta-analysis found myectomy to be superior to BT in increasing the frequency of bowel movements in adults with IAS achalasia, with no difference in medication use and complications.[107] However, a high incidence

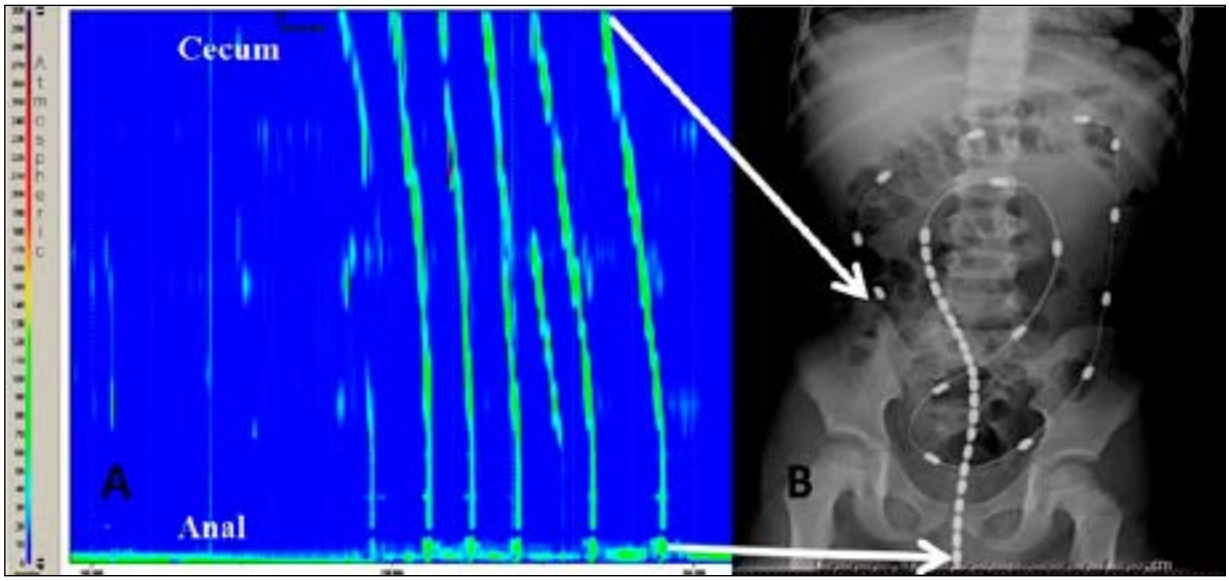

Figure 3. A normal high-resolution colonic motility study in an 8-year-old boy with constipation. **A:** A 2-dimensional colored topographic tracing. **B:** An abdominal radiograph depicting the location of the motility catheter. Note the normal migration of colonic contractions from the cecum to the rectum.

of fecal incontinence has been reported in long-term follow-up of children after myectomy.[108] Myectomy is recommended in those who have failed or have become dependent on BT.

The use of anal BT injections also has been reported to be successful in the management of children with symptoms of colonic obstruction after surgical repair of HD,[106] typically guided by an ARM showing elevated resting pressure that impairs normal evacuation of stools with a nonrelaxing sphincter. Patients who do not respond should be further evaluated for colonic dysmotility as the cause of their symptoms.

The use of an antegrade colonic enema (ACE) has been associated with an improvement in bowel movement frequency, fecal incontinence,[109,110] quality of life, and global health of children and their families.[111-113] Its long-term use is associated with improvement and normalization of CM in up to 83% of children.[114,115] A successful ACE response can be predicted by the presence of bisacodyl-induced HAPC on CM.[116] The rate of long-term successful bowel management ranges from 69% to 91%,[117,118] and complete symptom resolution and successful ACE discontinuation have been reported in 6% to 25% of children.[111,115,117-119] Complications have been reported in 60% to 63% of patients,[117,118,120] ranging from minor site infection in 4% to 29%, stoma stenosis and narrowing in 14% to 50%, leakage in 3% to 43%, and formation of granulation tissue[111,117,118,120-127] to significant morbidity from peritonitis, stoma revision, abscess formation, intestinal obstruction, and volvulus.[118,119,121,122,125]

Surgery beyond ACE has a limited role in children. Segmental colonic resections are controversial, with early studies supporting resection guided by CM[128,129] and recent reports demonstrating poor long-term outcomes with such an approach.[130] There is a consensus, however, regarding the role of diverting ileostomy or colostomy guided by CM in patients with colonic inertia and severe colonic distention.[129-131] The goal of diversion is to permit effective colonic decompression, thereby allowing a partial or total return of colonic function. Villarreal and colleagues reported resolution of distention in 11 of 12 patients with diverting ostomies and normalization of CM in 4 patients who subsequently underwent successful reanastomosis.[131] Those with persistent segmental abnormalities on repeat CM after diversion or ACE may benefit from segmental resection.[128-131] CIPO should be suspected in patients who fail surgical diversion. Although subtotal colectomy is commonly performed in adults with colonic inertia,[132] it is rarely performed in children. Subtotal colectomy is recommended only for those patients who do not recover colonic function and desire to close the ostomy and/or have significant ostomy complications.

Outcome

Despite proper management, 25% to 30% of children with constipation continue to have symptoms into adulthood.[133,134] Factors associated with poor outcomes when these children become adults include older age at onset, delay in diagnosis, and decreased defecation frequency.[134]

Summary

Gastrointestinal motility disorders in children are common and challenging, with limited epidemiologic information, diagnostic techniques, and therapeutic interventions. Further studies are needed to better diagnose and treat these conditions in children.

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