

Colonic Diversion for Intractable Constipation in Children: Colonic Manometry Helps Guide Clinical Decisions

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ABSTRACT

Background: Colonic manometry helps discriminate functional and behavioral causes for childhood constipation from colonic neuromuscular disease.

Methods: Of 375 colonic manometries performed for clinical indications, 12 could not be interpreted because of chronic colonic dilation. Based on colonic manometries that showed either no contractions or an absence of the gastrocolonic response or an absence of high-amplitude propagating contractions, the authors recommended diverting colostomies or ileostomies in 12 chronically constipated children (mean age, 4 years; range, 2–14 years, 5 boys). Before study, medical treatment was ineffective in all children. These children had persistently dilated colons with pathologic diagnoses of intestinal neuronal dysplasia (n = 4), hypoganglionosis (n = 2), hollow visceral myopathy (n = 1), and normal (n = 5).

Results: Six to 30 months after diversion, the authors restudied all the children. Eleven of 12 diverted colons were no longer dilated. In two patients, abnormal motility involving the entire colon was unchanged from the initial study, small bowel mo-

tility was abnormal, and we recommended no further surgery. In two cases, the colon remained abnormal but small bowel motility was normal, and we recommended subtotal colectomy and ileoproctostomy. In four cases, the left colon remained abnormal, but the right colon was normal, and we recommended reanastomosis after left hemicolectomy. In four cases, motility in the diverted colons was normal, including a gastrocolonic response and high-amplitude propagating contractions, and the authors recommended reanastomosis. Defecation problems resolved in 10 of 12 when followed up 5 to 30 months after treatment.

Conclusion: These data suggest that in some cases of intractable childhood constipation associated with colonic distention, temporary diversion improved colonic motility. Colonic manometry may be used to predict which patients will benefit from resection or reanastomosis. *JPGN* 33:588–591, 2001. **Key Words:** Motility—Defecation disorders—Colorectal surgery—Pediatric surgery. © 2001 Lippincott Williams & Wilkins, Inc.

Chronic constipation may be defined as fewer than three stools per week or difficulty and discomfort with defecation. Constipation is a common childhood complaint, accounting for 3% to 5% of pediatric visits and 10% to 25% of referrals to pediatric gastroenterologists (1). In children, constipation is most often due to functional constipation or functional fecal retention (2), but rare patients may have a colonic neuromuscular disease. Colonic manometry helps discriminate between functional disorders and neuromuscular diseases in children

with intractable constipation unresponsive to standard medical treatment and of uncertain cause (3). In the dilated colon, contractions measured by colonic manometry are sometimes very low in amplitude or absent. In some children with diffusely dilated colon, an accurate interpretation of the colonic manometry data may be impossible, because there are no contractions recorded by the current manometry methods. In this small subset of children with diffusely dilated colons and an uninterpretable colonic manometry who have reduced quality of life, we recommended a decompressing colostomy or ileostomy.

The purpose of this study was to review the influence of colonic diversion and decompression on colonic motility abnormalities in children with intractable constipation and a dilated colon. Then we studied the role of colonic manometry in predicting which patients benefit from reanastomosis.

Received November 14, 2000; revised March 27, 2001; accepted April 30, 2001.

Supported (M. S. and T. Z.) by research fellowships from Janssen Pharmaceutica, Titusville, New Jersey.

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METHODS

We retrospectively reviewed data collected from 1997 to 2000 at three different centers. Diagnostic criteria, colonic manometry testing protocols, equipment, data analysis, and interpretation were the same in all three centers.

During this period we performed 375 colonic manometries: 160 were normal, and final diagnoses were functional fecal retention ($n = 123$), irritable bowel syndrome ($n = 25$), functional constipation associated with cyclic vomiting syndrome ($n = 6$), or Munchausen by proxy ($n = 6$); 203 colon manometries were abnormal. Final diagnoses were colonic neuropathy ($n = 130$), myopathy ($n = 15$), anal achalasia ($n = 11$) (4), neuropathy proximal to the aganglionic segment or high-amplitude propagating contractions (HAPCs) through the neorectum in children with persistent symptoms after surgery for Hirschsprung disease ($n = 30$) (5) and chronic intestinal pseudo-obstruction ($n = 17$) (6).

Designations of neuropathy and myopathy imply that we studied the colon only or that small bowel manometry was normal. Neuropathy was defined by either absence of an increase in motility after a meal (the gastrocolonic response) or absence of HAPCs. Myopathies were defined by persistent low-amplitude or absent contractions in a colon without distention. In children with chronic intestinal pseudoobstruction colon and small bowel manometries were both abnormal.

In 12 children (age, 2–14 years; mean, 4 years; 7 boys) with long histories of intractable constipation, persistently dilated colons, and abnormal colonic manometry, we were unable to determine a final diagnosis based on physiology. For these 12 children we elected to decompress the dilated bowel with ileostomy ($n = 8$) or colostomy ($n = 4$).

All 12 children had full-thickness biopsies with histopathologic studies from individual hospital laboratories before the first manometry study. Four had neuronal intestinal dysplasia, one had hollow visceral myopathy, two had hypoganglionosis, and five were normal. One with normal full-thickness histologic findings had a solitary rectal ulcer on endoscopy. Neuronal intestinal dysplasia (NID) was a diagnosis applied in only one of the three clinical pathology laboratories. We are aware of the difficulties in attempting to diagnose NID (7,8). We made no attempt to have experts review the histologic findings, but instead compared the routine clinical histologic reports with manometry and outcome.

Before each manometry, informed consent was obtained from a parent of each patient. Laxatives and drugs affecting intestinal motility were discontinued at least 3 days before the study. One day before, we prescribed a clear liquid diet and infused a balanced electrolyte solution (Golytely, Braintree Laboratories, Braintree, MA) through a nasogastric tube. We used no enemas or suppositories, because they might cause inflammation and alter motility. All patients fasted for at least 8 hours before colonoscopy. We used colonoscopy to facilitate placement of a manometry catheter with eight recording sites 10 cm or 15 cm apart with inner diameter of 0.8 mm and outer diameter of 4.8 mm, so that the most proximal site recorded from the cecum or ascending colon and the most distal site recorded from the rectum. We confirmed the manometry catheter position with brief fluoroscopy. We completed the colon manometry on the same day as catheter placement after the patient recovered from propofol anesthesia (Diprivan, Astra Zenica Pharmaceuticals, Wilmington DE) and midazolam sedation (Versed, Roche Pharmaceuticals, Nutley, NJ). When

general anesthesia or narcotics were used for catheter placement, the children were studied on the day after. The patients engaged in quiet play during the test session. The catheter was perfused with distilled water using a pneumohydraulic infusion system. Pressures were transmitted to a transducer and recorded on a personal computer (Redtech, Calabasas, CA). We recorded fasting motility in all patients for 1 hour. Next, the patients ate a meal appropriate for age, with fat providing more than 30% of the calories, and recording was continued for another hour. Finally, we infused 0.2 mg/kg bisacodyl (Dulcolax, Ciba Self-Medication, Inc., Woodbridge, NJ) through the central lumen of the catheter into the cecum and continued recording for another 30 minutes (9).

Colon contraction patterns were identified visually. Movement artifacts elicited rapid fluctuations that were easily distinguished from colonic contractions and occurred simultaneously at all recording sites. High-amplitude propagating contractions were defined by contractions more than 60 mm Hg that did not overlap, with duration more than 10 seconds and less than 30 seconds and propagation aborally across 30 cm or more, over three or more recording sites (10). Nonpropagating contractions were defined as contractions that did not propagate across more than two recording sites. Propagation of contraction was defined as antegrade or retrograde on the basis of their aborad or orad propagation, respectively. The gastrocolonic response was defined by an increase in colon motility that occurred within 20 to 40 minutes after beginning a meal (9,10). A dilated colon was identified in plain abdominal films. In children with abnormal studies, we always obtained an abdominal x-ray at the end of the test session to confirm catheter position.

All patients had a diverting colostomy or ileostomy after the first manometry. Five to 30 months after colonic decompression, we restudied all the children. We used the same protocol for colonic manometry to study the diverted colons from the anus to the ostomy and the ostomy to the cecum in patients with sigmoid or transverse colostomies. We analyzed the colonic contractions using the same criteria. On the basis of the second manometry, we developed a treatment plan for each patient. Outcome was assessed by clinical follow-up averaging 15 months (range, 5–30 months) after the treatment.

RESULTS

All patients had dilated colons at the time of the first colonic manometry. In that initial manometry, no one had spontaneous HAPCs, and gastrocolonic responses were absent. Six of 12 children had no response to bisacodyl. One had a partial response, with contractions ending at the splenic flexure. Four patients had simultaneous contractions. One patient had retrograde contractions.

After the first study all patients underwent elective colostomy ($n = 4$) or ileostomy ($n = 8$) for decompression. We performed a second manometry 6 to 30 months (mean, 15 months) after decompression (Table 1). In 11 patients, the colon was no longer dilated. The gastrocolonic response and HAPCs were present in four patients. Another four patients had a gastrocolonic response, but the HAPCs were present only in the right colon and

TABLE 1. Individual patient summaries

Age (years)	Sex	Disease	Gastrocolonic response	HAPCs	Gastrocolonic response	HAPCs	Intervention
4	F	NID	Absent	No contractions	(11) Present	R colon: HAPCs L colon: absent	Hemicolectomy and reanastomosis
2	F	NID	Absent	No contractions	(12) Present	Present	Reanastomosis
5	M	Normal	Absent	No contractions	(5) Present	Present	Reanastomosis
9	F	Rectal ulcer	Absent	No contractions	(6) Present	Present	Reanastomosis
1	M	Visceral myopathy	Absent	No contractions	(24) Present	Absent	Maintain ileostomy
14	F	Normal	Absent	No contractions	(24) Present	Absent	Subtotal colectomy and ileoproctostomy
8	M	Normal	Absent	R colon: HAPCs L colon: absent	(9) Present	R colon: HAPCs L colon: absent	Hemicolectomy and reanastomosis
2	F	NID	Absent	Simultaneous contractions	(30) Present	R colon: HAPCs L colon: absent	Hemicolectomy and reanastomosis
3	M	Hypoganglionosis	Absent	Simultaneous contractions	(24) Present	Present	Reanastomosis
1	F	Hypoganglionosis	Absent	Simultaneous contractions	(12) Absent	Absent	Maintain ileostomy
6.5	M	NID	Absent	Retrograde contractions	(9) Absent	R colon: HAPCs L colon: absent	Hemicolectomy and reanastomosis
13	F	Normal	Absent	Simultaneous contractions	(8) Absent	Absent	Subtotal colectomy and ileoproctostomy

Intervals between the manometry studies in months are in parentheses. HAPCs, high-amplitude propagated contractions; NID, neuronal intestinal dysplasia.

absent in the left colon. One patient had no gastrocolonic response, and the HAPCs were present in the right colon and absent in the left colon. Three patients had no contractions.

In four patients who had gastrocolonic response and HAPCs, we recommended reanastomosis without colon resection. These four patients now have normal evacuation.

In the 12- and 13-year-old patients, there were no gastrocolonic responses or HAPCs, but they had normal small bowel manometries. We recommended subtotal colectomy and ileoproctostomy. One has normal evacuation, and the other still requires daily oral polyethylene glycol to ensure daily defecation.

In four patients who had a gastrocolonic response but HAPCs only at the right colon, we recommended a left hemicolectomy and colorectal anastomosis. Three patients have no current defecation complaints. In one patient who had no gastrocolonic response and HAPCs only at the right colon, left hemicolectomy and colorectal anastomosis has not cured constipation. In two patients with no contractions after diversion and decompression and abnormal upper gastrointestinal motility, the ileostomy was maintained.

A pathologic diagnosis of NID was not associated with discrete manometric features, and did not predict outcome.

DISCUSSION

Constipation is a symptom, not a disease. As a symptom, constipation is influenced by functional, behavioral, dietary, and organic conditions (11). Clinical features, such as onset at birth, extrarectal fecal masses (fecal

masses palpable in the colon proximal to the rectosigmoid, but an empty rectum), and an absence of retentive posturing, encopresis, and painful defecation suggest colonic neuromuscular disease. Conversely, a history of painful defecation, retentive posturing, and infrequent passage of enormous hard stool are diagnostic for functional fecal retention (2). When there is a symptom-based diagnosis of functional fecal retention, no testing is necessary or desirable. For the unusual patient whose symptoms are consistent with colonic neuromuscular disease, or who has symptoms that overlap disease and a functional disorder and who has not responded to medical management, colonic manometry provides documentation of the physiology associated with the symptoms. Most of the children referred for colonic manometry had constipation of uncertain cause.

In all but 12 of 375 patients, colonic manometry was interpretable and clarified the cause of constipation. In many children with functional fecal retention, the colon was dilated, but colonic manometry was normal. In 12 children (3%) with intractable symptoms, a chronically dilated colon interfered with a conclusive interpretation of data. In these 12 carefully selected children who failed medical management, had radiologic documentation of persistently dilated colons, and colon manometries that showed either no contractions or absent gastrocolonic response and HAPCs, we recommended diverting colostomies or ileostomies. We elected to divert the colon in an attempt to reduce wall tension and so allow recovery of normal function. In those with disease, the dilation was secondary to failed motility. In those with massive dilation secondary to functional fecal retention, the dilation was due to outlet obstruction at the pelvic floor. Decompressing the dilated colon improved the diagnosis-

tic accuracy of the second colonic manometry in the 12 children.

After diversion, the dilated bowel resolved in 11 of 12 children. For several patients, the amplitude of the colon contractions improved as the lumen diameter narrowed. These patients had histories of stimulant laxative use, and one possibility is that chronic use of stimulant laxatives may have reversibly impaired normal colonic functions (12). After colostomy or ileostomy, no stimulant laxatives were required. Repeating the colonic manometry study after diversion would have allowed time for recovery from the possible effect of stimulant laxatives.

Four patients had normal motility in the diverted colon at the time of the second manometry, and their bowel continuity was re-established. The normal colonic manometry gave the clinicians, parents, and child confidence that there would be a successful outcome to a reanastomosis. All four of these children did well. Presumably, their severely dilated, nonfunctioning colons were a consequence of functional fecal retention with failed management.

In our study, full-thickness biopsy results did not seem to correlate either with manometry results before or after diversion or with outcome. Pathologists did not come to consensus on histologic findings from our patients, and only one pathologist from one center reported NID. Therefore, there must be cautious in interpreting the histopathologic findings. However, our experience is similar to Cord-Udy et al. (7), who found that NID in rectal suction biopsy specimens did not correlate with outcome, and with Simpson et al. (13), who found that NID in full-thickness biopsy specimens did not predict outcome in 75 patients undergoing surgical procedures for childhood constipation. In contrast to the poor correlation of histologic findings with outcome, we found that manometry helped in the choice of procedure(s) for our patients.

Specifically, we used HAPCs as a marker for neuromuscular function. We directed surgeons to resect those colon segments proximal to the rectosigmoid that failed to demonstrate HAPCs.

One patient has not done well after surgery. We suspect that chronic unresolved family problems and non-compliance are the cause of continuing constipation and fecal soiling. However, other possible explanations include 1) functional fecal retention, and 2) a progressive colonic neuromuscular disease. Children who have had colonic neuromuscular disorders may be predisposed to functional fecal retention, because those children expe-

rienced chronic defecation failure and possibly repeated painful defecation. Functional fecal retention is a common disorder. In contrast, progressive colonic neuromuscular disease is rare. If a trial of assuring painless defecation with oral polyethylene glycol fails to alleviate the symptoms in a compliant child and family, colonic manometry would be appropriate.

We conclude that colonic diversion and subsequent colonic manometry aids the evaluation and management of patients with intractable constipation, dilated colons, and an abnormal colonic manometry.

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