Journal of Pediatric Gastroenterology and Nutrition
41:S58–S60 © September 2005 Lippincott Williams & Wilkins, Philadelphia

Surgery and Constipation: When, How, Yes, or No?

Marc A. Levitt and Alberto Peña

Division of Pediatric Surgery, Cincinnati Children's Hospital Medical Center, Cincinnati, Ohio, USA

INTRODUCTION

Absorption of water from the stool and reservoir function of the colon are sophisticated processes that depend on colonic motility, and is an area of physiology that is not well understood, and for which treatments of problems are limited. It is clear that the rectosigmoid in normal individuals stores the stool, and every 24–48 hours develops active peristaltic waves indicating that it is time to empty. A normal individual feels this sensation, and decides when to relax the voluntary sphincter mechanism.

An absence of the colon results in multiple, liquid stools, since there is no water absorption or storage function. Decreased colonic motility is poorly defined, and is generally referred to as idiopathic constipation, which encompasses a broad spectrum, from mild constipation to intestinal pseudoobstruction. Treatments are empiric and must be adjusted to the magnitude of the symptoms. Similar decreased motility is also found in patients with anorectal malformations and Hirschsprung's disease, the motility disorders of which are beyond the scope of this review, but the treatments for these patients are applicable to patients with idiopathic constipation.

Idiopathic Constipation

Idiopathic constipation is the incapacity or difficulty to pass stool regularly, its etiology is unknown, and it is by far the most common defecation disorder in children which affects an enormous pediatric population. It is extremely incapacitating in its most serious forms, producing a form of fecal incontinence known as encopresis or overflow pseudo incontinence.

Even when the cause of this condition is unknown, the literature presents many potential causes, most without solid scientific basis. Diet impacts colonic motility; but its therapeutic value is negligible in the most serious forms of constipation. It is true that many patients with idiopathic constipation suffer from psychologic disorders, but a pure psychologic origin cannot explain the severe forms either, which can include a giant megacolon, megabladder, and serious nutritional and developmental disturbances. In addition, it is certainly not easy to voluntarily retain the stool when an otherwise autonomous rectosigmoid has normal peristalsis. Passage of large, hard pieces of stool may provoke pain, and make the patient behave like stool retainers. This may complicate the problem of constipation; but it is not the original cause.

An oversimplistic explanation is that there is a lack of relaxation of the internal sphincter, also known as ''achalasia''.(1) The concept is that incontinence means ''lack of sphincter,'' therefore, constipation means ''too much sphincter.'' However, the diagnosis of this achalasia of the internal sphincter is based mainly on manometric studies (2), which when analyzed carefully, are difficult to interpret. Manometry is performed by placing a balloon in the rectum while measuring the pressure of the anal canal. When the rectal balloon is inflated, under normal circumstances, there is a drop in the intra-anal canal pressure, the anorectal reflex. When the pressure does not drop in the anal canal, it is considered abnormal, and is a sign of lack of relaxation of the internal sphincter, potentially diagnostic for Hirschsprung's disease. If the patient's rectum has no ganglion cells on biopsy, the diagnosis of Hirschsprung's disease is confirmed. On the other hand, if there are ganglion cells, the patient receives the diagnosis of ''achalasia of the internal sphincter.'' In these cases the treatment proposed by many (3) is a myectomy or internal sphincterectomy, which is a controversial procedure, as descriptions of the surgical technique are not precise and are not easily reproducible. Also, the precise anatomic limits of the internal sphincter are not well documented and the results of such operations have not been uniformly good (4).

There are many publications that favor the idea that intestinal neuronal dysplasia may be the explanation for the abnormal colon motility observed in constipated patients, however there is no basic agreement among pathologists about how to make this specific histologic diagnosis (5). In order for a surgeon to propose a rational curative treatment for this condition, he or she must know the extent of the affected bowel which needs to be resected. In addition, the symptomatology of patients with intestinal neuronal dysplasia varies significantly. Described treatments vary widely from laxatives to enemas to different types of resections and the reported follow-up of patients has not been consistent. To confuse the issue further, some patients recover spontaneously. Intestinal

M.A. Levitt, MD (e-mail: marc.levitt@cchmc.org).

neuronal dysplasia is an entity that requires further scientific elaboration.

Hypoganglionosis has also been invoked as a potential explanation for patients with severe constipation, based on biopsies from specimens of dilated colon. If the colon becomes larger as the constipation problem gets worse, it is conceivable that a specimen taken from a giant megasigmoid will show relatively fewer ganglion cells, which could explain the "hypoganglionosis."

Finally, many surgeons believe that many patients with idiopathic constipation suffer from what is called "ultra short Hirschsprung's disease" (6). The concept is that some patients have a short area above the anal canal with absent ganglion cells. This attractive explanation has its flaws because normal individuals have a zone of aganglionosis above the pectinate line, the length of which has not been accurately described. The proposed treatment include botulinim toxin injection and/or a posterior rectal myectomy. There is no explanation as to why such procedures improve the symptoms in these patients, and the results of these interventions are debatable.

More recently, new potential explanations for colonic hypomotility have been proposed including a deficiency of substance P immuno reactivity in the colonic nerve fibers (7), abnormalities in colonic monoclonal anti-neurofilament antibodies (8), increased plasma level of pancreatic polypeptide, and decreased plasma levels of motilin (9). All these new contributions, represent very promising perspectives and deserve further investigation.

The etiology of idiopathic constipation is unknown. These patients suffer from a colonic hypomotility disorder, presenting with different degrees of severity and creating a wide spectrum of symptoms. On the benign side of the spectrum, one can see patients that suffer from a minor form of constipation that is treatable with diet. On the other extreme, patients have severe constipation that may overlap clinically with a serious condition known as intestinal pseudo-obstruction. Most of the proposed treatments for constipation do not take into consideration the spectrum concept, but rather they are standard therapeutic protocols that render good results in a percentage of cases, but always leave a group of patients that do not respond. The medically intractable group represents the most serious side of the spectrum, and it is these patients that may benefit from surgical intervention.

CLINICAL MANIFESTATIONS

Idiopathic constipation is a self-perpetuating disease. A patient that suffers from a certain degree of constipation that is not treated adequately, goes through life only partially emptying the colon, leaving larger and larger amounts of stool inside the rectosigmoid, which results in greater degrees of megasigmoid.

Most surgeons accept the clinical fact that dilatation of a hollow viscus produces poor peristalsis which explains the fact that constipation leads to fecal retention, which produces megacolon, that exacerbates the constipation. In addition, the passage of large, hard pieces of stool may produce anal fissures that result in a reluctance by the patient to have bowel movements.

The clinician must accept the fact that this condition is essentially incurable. It is manageable, but requires careful follow-up for life. Treatments are frequently given on a temporary basis; they then are tapered or interrupted followed by a subsequent recurrence. Sometimes, colostomies or colonic washouts via a catherizable stoma or button device are performed, and the patients are followed with contrast studies to monitor the degree of colonic dilatation. Once the distal colon regains a normal caliber, the physician assumes that the patient is cured, the colostomy is closed or the washouts are discontinued with the predictable return of symptoms.

Fecal impaction is a stressful event defined as a condition of retained stool for several days or weeks, crampy abdominal pain, and sometimes tenesmus. When laxatives are prescribed to a patient that suffers from fecal impaction, the result is exacerbation of the crampy abdominal pain and sometimes vomiting. This is a consequence of an increased colonic peristalsis (produced by the laxative) acting against a colonic obstruction due to the fecal impaction.

Soiling of the underwear is an ominous sign of bad constipation. A patient who at an age of bowel control soils the underwear day and night and basically does not have spontaneous bowel movements has "encopresis" or "overflow pseudoincontinence." These patients behave as fecally incontinent individuals. When the constipation is treated adequately, the great majority of these pseudoincontinent children regain bowel control.

SURGICAL TREATMENTS

Our protocol of treatment of patients with severe forms of idiopathic constipation includes a trial of medical management. If the patients do not respond to this treatment, then a specific type of operation is considered. The regimenn uses the same medications (laxatives) as have likely been previously tried, but the protocol is different in that the dosage is adapted to the patient's response. Almost always, the patient previously had received less laxative than they required. The patient's response should be monitored radiologically with the laxative dose adjusted daily.

SPHINCTER MYOTOMY

This procedure has many advocates, but as discussed, is not one that we perform.

SIGMOID RESECTION AND OTHER TYPES OF COLECTOMY

For the last 14 years, we have been performing a sigmoid resection for the treatment of these conditions (10,11). The very dilated megarectosigmoid is resected and the descening colon is anastomosed to the rectum. During this time we have followed 237 patients suffering from idiopathic constipation. Seventeen of them elected to have the operation. We also treated 315 patients suffering from constipation and anorectal malformations. From this last group, 53 underwent a sigmoid resection.

The degree of improvement in these patients varied. Following sigmoid resection, 10% of patients did not require any more laxatives, have bowel movements every day and no soiling. Thirty percent of patients decreased the laxative requirement by 80%. The remaining 60% of patients decreased the laxative requirement by 40%. These patients must be followed closely because the condition is not being cured by the operation. The remaining rectum is most likely abnormal, and without careful observation and treatment of constipation, the colon can redilate.

An alternative could be to resect the rectosigmoid including the rectum, down to the pectinate line in a similar manner as for patients with Hirschsprung's disease, and anastomose the non-dilated colon (that is assumed to have normal motility) to the rectum above the pectinate line. This treatment of course, is something to be considered, however requires a colo-anal anastomosis, which carries a higher morbidity, and eliminates the rectal reservoir, which may impact continence.

The most dilated part of the colon is resected because it is most seriously affected. The non-dilated part of the colon is assumed to have a more normal motility. Clearly, there must be a more scientific way to assess the dysmotile anatomy. The patients who improve the most, are the patients that have a more localized form of megarectosigmoid. Patients with more generalized dilation of the colon do not respond as well.

The administration of antegrade enemas through a continent appendicostomy or a button cecostomy is becoming popular (12). This is particularly useful in patients who are fecally incontinent who require a daily enema and seek more independence for their bowel management program (13). This modality of treatment represents a useful alternative for patients that are treated with enemas only, since those antegrade enemas are only a different route of administration of enemas. However, the overwhelming majority of patients with idiopathic constipation can be treated with laxatives, with or without a sigmoid resection and therefore do not need washouts at all. Distinguishing which patients require washouts because they cannot empty on their own from patients that could empty if their constipation was adequately managed with laxatives is a key challenge for the clinician.

The future will continue to bring a more serious, scientific approach to the knowledge of constipation. It is likely that modern techniques will identify histologic abnormalities that currently escape our eye. Motility studies of the colon will likely reach a degree of sophistication that will make them more reliable, and more clinically useful. Eventually, genetics could play a role in the prevention of this condition, and finally, development of pharmacologic control of motility will greatly enhance the clinical armementarium.

REFERENCES

1. Fadda B, Welskop J, Muntefering H, et al. Achalasia of the anal sphincter. *Pediatr Surg Int* 1987;2:81–5.
2. Keren S, Wagner Y, Heldenberg D, Golan M. Studies of manometric abnormalities of the rectoanal region during defication in constipated and soiling children: Modification through biofeedback therapy. *Am Coll of Gastroenterology* 1988;83:827–31.
3. Ecaluwe D, Yoneda A, Akl U, Puri P. Internal anal sphincter achalasia: Outcome after internal sphincter myectomy. *J Pediatr Surg* 2001;36:736–8.
4. Pinho M, Yoshioka K, Keighley MRB. Long term results of anorectal myectomy for chronic constipation. *Br J Surg* 1989;7: 1163–4.
5. Csury L, Peña A. Intestinal neuronal dysplasia: Myth or reality? *Pediatr Surg Int* 1995;10:441–6.
6. Neilson IR, Yazbeck S. Ultrashort Hirschsprung's disease. Myth or reality? *J Pediatr Surg* 1990;25:1135–8.
7. Wheatley JM, Hutson JM, Chow CW, et al. Slow-transit constipation in childhood. *J Pediatr Surg* 1999;34:829–33.
8. Kluck P, Tibboel D, Leendertse-Verloop K, et al. Diagnosis of congenital neurogenic abnormalities of the bowel with monoclonal anti-neurofilament antibodies. *J Pediatr Surg* 1986;21:132–5.
9. Stern HP, Stroh SE, Fiedorek SC, et al. Increased plasma levels of pancreatic polypeptide and decreased plasma levels of motilin in encopretic children. *Pediatrics* 1995;96:111–6.
10. Peña A, El-Behery M. Megasigmoid – A source of pseudoincontinence in children with repaired anorectal malformations. *J Pediatr Surg* 1993;28:1–5.
11. Levitt MA, Carney DE, Powers CJ, et al. Laparoscopically Assisted Colon Resection for severe Idiopathic Constipation with Megarectosigmoid. *J Pediatr Endosurgery and Innovative Techniques* 2003;7:285–9.
12. Marshall J, Hutson JM, Anticich N, Stanton MP. Antegrade continence enemas in the treatment of slow-transit constipation. *J Pediatr Surg* 2001;36:1227–30.
13. Levitt MA, Soffer SZ, Peña A. The continent appendicostomy in the bowel management of fecally incontinent children. *J Pediatr Surgery* 1997;32:1630–3.