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Pediatric Fecal Incontinence: A Surgeon's Perspective

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Drs Levitt and Peña
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Objectives After completing this article, readers should be able to:

1. Describe the difference between true fecal incontinence and pseudo-incontinence.
2. Understand the patient groups that suffer from true fecal incontinence.
3. Discuss the physiologic mechanisms of continence.
4. Formulate the evaluation and treatment of a patient who presents with soiling.
5. Develop a treatment protocol for constipation with overflow pseudo-incontinence (encopresis).
6. Describe the primary differences between Hirschsprung disease and idiopathic constipation.

Introduction

Editor's Note: Fecal incontinence is a frustrating condition for patients, parents, and clinicians. This article discusses fecal incontinence caused by anatomic conditions as well as the overflow of stool that follows severe constipation. The perspective is that of the pediatric surgeon, and the discussion offers insights not always present in pediatric reviews. Some recommendations are derived from the extensive experience of the authors over many years of treating these patients.

All figures for this article are contained in the data supplement

Fecal soiling is a common problem that pediatricians are asked to evaluate. Fecal incontinence represents a devastating problem that may prevent a child from becoming socially accepted. More children are affected than previously believed, including those born with surgical conditions such as anorectal malformations (ARMs) and Hirschsprung disease (HD), as well as those who have spinal cord problems or injuries.

Patients can have true fecal incontinence or can suffer from overflow pseudo-incontinence. These two conditions have completely different treatments. Those who experience true incontinence include a percentage of surgical patients (who have ARMs and HD) as well as those who have congenital or acquired spinal problems. Pseudo-incontinence (encopresis) occurs in patients who have the potential for bowel control but whose constipation leads to overflow and soiling.

Of the surgical patients, approximately 25% of those undergoing surgery for ARMs have enough of a deficiency in their continence mechanism that they cannot have a voluntary bowel movement. The rest may be able to be continent but require treatment, usually for constipation but sometimes for loose stool. (1) A small number of patients born with HD (<5%) suffer from fecal incontinence postoperatively, (2)(3)(4) usually because of damage to the anal canal or sphincters. Spinal problems or injuries can manifest as a limited capacity for voluntary bowel movements. (5)(6)

Patients who have true fecal incontinence need an artificial (mechanical) mechanism to keep them clean and in normal underwear, that is, a tailored enema program. (7) Medical treatments with laxatives do not work for such patients and actually worsen the situation. In contrast, patients who have overflow pseudo-incontinence from severe constipation require adequate treatment of their constipation. This distinction is the key to determining the correct management.

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Physiologic Mechanisms of Fecal Continence

Fecal continence depends on voluntary sphincter muscles, anal canal sensation, and motility of the colon. (1)

Voluntary Sphincters

The voluntary sphincters include the levator, the muscle complex, and the external sphincter and are used only when stool in the rectum reaches the anorectal area, propelled there by involuntary peristalsis of the rectosigmoid. Most individuals voluntarily hold the stool by contracting the sphincters only in the minutes prior to defecation. Otherwise, these muscles are used only occasionally throughout the day and night. They also relax voluntarily at the appropriate time to allow the stool to exit the rectum.

The voluntary muscles in patients born with ARMs have varying degrees of hypodevelopment. Similarly, patients who have spinal problems or injuries can have sphincter dysfunction. Voluntary sphincters can be called on only when the patient feels the need to use them, information that can be derived only from intact anal sensation.

Anal Canal

The anal canal provides exquisite sensory information. Except for patients born with rectal atresia (who have normal anal canals), most patients who have ARMs are born without this distal anatomy, and the sensation does not exist or is rudimentary. Patients who have HD are born with normal anal canals that can be injured during the pull-through procedure if the canals are not carefully preserved. Those who have spinal problems or pelvic trauma may have injured, destroyed, denervated, or non-functional anal canals.

For the patient to perceive rectal distention, the rectum must be located precisely in the center of the sphincteric mechanism—a vital aspect of the pull-through procedure for imperforate anus. Stretching of the sphincters by stool in the rectum is perceived by the patient (proprioception). Thus, to achieve bowel control, the patient must have the capacity to form solid stool. Loose stool causes no stretching of the rectum and, therefore, no proprioception. This point also is relevant for children who have ulcerative colitis, have undergone an ileoanal pull-through, and may suffer from periods of incontinence due to their inability to form solid stool. Usually, however, their normal sphincters and intact anal canals allow them to overcome the hypermotility and avoid incontinence.

Motility

The rectosigmoid normally remains quiet for variable periods of time (one to several days) and when quiescent, anal sensation and sphincteric muscles are inactive be-

cause the stool (if solid) remains inside the colon. The patient often can feel the peristaltic contraction of the rectosigmoid occurring prior to defecation. Most individuals then voluntarily relax the striated muscles and allow the stool to migrate down to the rectum just above the highly sensitive anal canal. Information provided by the anal canal concerning the consistency and quality of the stool cues the patient to push the rectal contents voluntarily back up into the rectosigmoid and hold them until the appropriate time for evacuation. The voluntary muscles then relax at the time of defecation.

The key factor that provokes emptying of the rectosigmoid is an involuntary peristaltic contraction, helped by a Valsalva maneuver. Patients born with ARMs may suffer from a disturbance of this mechanism. Those who have undergone a posterior sagittal anorectoplasty or any other type of sacroperineal approach, in which the most distal part of the bowel is preserved, show evidence of an overefficient reservoir, usually associated with a megarectum that manifests as constipation (particularly evident in patients born with lower type anorectal defects). (8)

The physiologic cause for this same outcome in patients who were not born with anomalies but who develop severe constipation and encopresis remains unknown. It is clear that constipation that is not treated aggressively leads to more severe constipation. A vicious cycle develops, with worsening constipation leading to more rectosigmoid dilation, worse constipation, and ultimately soiling. We do not know why the enormously dilated rectosigmoid (equipped with normal ganglion cells) is hypomotile initially, and we do not know which comes first, constipation or loss of tone, leading to dilation. (1)

Patients born with ARMs who have had surgical procedures in which the most distal part of the colon is resected (9)(10) behave like patients who lack rectal reservoirs. Depending on the amount of colon removed, the patient may have loose stools. In such cases, medical management is needed, consisting of a constipating diet and medications to add bulk, such as pectin, and slow colonic motility, such as loperamide. (Warning: For children younger than 2 years of age, use of loperamide must be monitored carefully because of the risk of central nervous system depression and rare anaphylaxis.) Resection of the distal aganglionic bowel is precisely the operation performed for patients who have HD, but it is the normal anal canal and sphincters that allow most patients to remain continent despite the loss of the rectal reservoir. Interestingly, some patients who have injured anal canals and sphincters (pelvic trauma) can be continent if their motility is regular because the reliable contraction

of the rectosigmoid can be translated into a successful voluntary bowel movement.

Because fecal incontinence and constipation are associated so closely, clinicians should be aware of the many conditions that can cause constipation in a young child. All of the disorders listed in Table 1 can cause constipation to varying degrees. This article focuses on situations whereby the constipation leads to soiling and the management thereof.

When therapy is considered, it is important for the clinician to distinguish between laxatives, which enhance motility by provoking peristalsis, and stool softeners and lubricants, which soften the stool and make it easier to pass. Many children who have slow-moving colons, particularly those who have had surgery, need the provocative effect of laxatives and do not respond to stool softeners. With softeners, the colon remains full of stool, albeit softer stool, but the stool still does not come out.

Hirschsprung Disease

HD, in which a section of bowel of variable length proximal to the anal sphincter lacks neural innervation, deserves special attention because it is a major anomaly that can manifest as constipation. This relatively common colorectal condition may present in the newborn period, in the first several postnatal months, or later in childhood.

Babies born with HD usually become symptomatic during the first 24 to 48 hours after birth. Abdominal distention, delayed passage of meconium, and vomiting are the most frequent manifestations in the newborn. Spontaneous or induced explosive, massive, deflating passage of liquid bowel movement and gas occurs often and improves the baby's condition dramatically, followed by a period of hours or days of relative absence of symptoms, then recurrence of the same manifestations. Stools frequently are liquid and foul. When the abdomen is distended, the infant can become very ill from sepsis, hypovolemia, and shock. Enterocolitis is the most serious complication.

The differential diagnosis includes any condition that causes intestinal obstruction in the newborn, including meconium-plug syndrome, in which expulsion of a plug of meconium resolves symptoms. Meconium ileus causes a clinical picture consistent with that of intestinal obstruction, with the child frequently exhibiting respiratory symptoms. A family history of cystic fibrosis may be present. The absence of air-fluid levels in an upright abdominal radiograph and the "ground-glass" appearance of the lower abdomen are characteristic radiographic signs of this condition. Small left colon syndrome

Table 1. Causes of Constipation

Functional: Dietary factors (lack of dietary fiber, excessive calcium intake, dehydration, malnutrition), motility disturbance (slow transit), stool withholding, lack of exercise, and immobility
Structural Abnormalities: Anal disorders (imperforate anus, fissures, anal stenosis), colonic strictures, pelvic tumors (sacral teratoma), postsurgical repair of anorectal malformation, or Hirschsprung disease
Endocrine, Metabolic, and Immunologic Conditions: Celiac disease, cystic fibrosis, hypercalcemia, hyperparathyroidism, hypokalemia, hypothyroidism, uremia
Neurogenic Conditions: Cerebral palsy, hypotonia, spinal cord abnormalities (spina bifida, spinal tumors, tethered cord)
Neuromuscular Conditions: Hirschsprung disease, infant botulism
Connective Tissue Disorders: Scleroderma, systemic lupus erythematosus
Drugs: Antacids, anticholinergics, tricyclic antidepressants, bismuth, opiates, phenobarbital, sympathomimetics

might be confused with HD. Contrast enema demonstrates a rather narrow left colon to the level of the splenic flexure. Symptoms usually improve following this study and resolve after several weeks. The mother frequently has diabetes.

Other nonsurgical conditions that may be confused with HD that cause abdominal distention include hypothyroidism, adrenal insufficiency, and cerebral injury. A baby suffering constipation and painful passage of stool with distention can have simple idiopathic constipation or HD. The lack of watery stool and good growth point the clinician toward simple constipation. If mild measures such as gentle laxatives or dietary changes help, the patient likely does not have HD.

To evaluate for the possibility of HD, a contrast enema with water-soluble contrast material is the most valuable radiologic study. This study may reveal the presence of a distended proximal colon, the transition zone, and a "contracted" distal rectosigmoid. The older the patient, the more obvious the size difference between the normal ganglionic intestine and the abnormal aganglionic intestine. Therefore, the typical changes sometimes are not obvious during the neonatal period. Generally, however, the transition zone is recognized in most newborns. In instances of total colonic aganglionosis, the contrast enema may reveal a short colon, with retraction of the hepatic and splenic flexures and straightening of the sigmoid.

Confirmation of the diagnosis is based on the absence of ganglion cells and presence of hypertrophic nerves in a rectal biopsy. Acetylcholinesterase staining also is abnormal. The specimen must be taken at least 1.5 cm above the pectinate line.

Bowel irrigation with saline solution is a valuable procedure for the emergency management of enterocolitis. By decompressing the bowel, the procedure may improve a very ill infant dramatically until surgery can be performed. The definitive pull-through procedure can be performed in the newborn period in a single stage without a protective colostomy and sometimes by way of a transanal approach alone.

Occasionally, a child may have minimal or absent clinical manifestations during the first days or weeks after birth and exhibit moderate, intermittent symptoms later in life. In the older child who has constipation, the clinician may wonder if the symptoms are caused by HD. It is important to distinguish between HD and idiopathic constipation because the treatment and consequences of the two conditions are so different. If the patient has no episodes of enterocolitis and has thrived, HD is less likely. A contrast study provides confirmation.

True Fecal Incontinence

The term true fecal incontinence refers to situations in which an underlying structural abnormality leads to fecal soiling, as opposed to pseudoincontinence, in which the fecal loss results from constipation and overflow of liquid stool.

Which Patients Have True Fecal Incontinence?

A 5-year-old boy born with an ARM that was repaired during his first postnatal year has daily fecal soiling. His original malformation was a fistula between his rectum and bladder neck (the highest type of ARM in males). His buttock crease is flat, with minimal sphincteric musculature, he has a short sacrum, and he had a tethered cord released.

Approximately 75% of children who have repaired ARMs have voluntary bowel movements after the age of 3 years. (8) About 50% of such patients soil their underwear on occasion, usually due to constipation, but when the constipation is treated properly, the soiling disappears. Thus, most patients have voluntary bowel movements and no soiling and behave like unaffected children. Those who have bowel control may suffer from temporary episodes of fecal incontinence if they experience diarrhea.

About 25% of children who have ARMs suffer from true fecal incontinence. Some patients who have HD as well as those afflicted with spinal problems also suffer

from true fecal incontinence. For these children, the clinician can apply similar principles of bowel management learned from the treatment of patients who have ARMs. (7)

The prognosis for bowel control among children born with ARMs is largely predictable (Table 2). The prognosis for continence can be established after surgical reconstruction and colostomy closure. This determination sometimes is possible even in the newborn period and is very important because it avoids false expectations for the parents at the age of toilet training.

With knowledge of the specific type of ARM, the clinician can predict the functional prognosis. If the child's defect is one associated with a good prognosis (rectovestibular fistula, rectoperineal fistula, rectal atresia, rectourethral bulbar fistula, imperforate anus with no fistula or low cloaca), the child can be expected to have voluntary bowel movements by the age of 3 years. Such

Table 2. Prognosis for Bowel Control With Anorectal Malformations

Anatomic Indicators of Good Prognosis

- Normal sacrum
- Prominent midline groove (good muscles)
- Obvious anal dimple
- Some types of anorectal malformations
 - Rectal atresia
 - Rectoperineal fistula
 - Imperforate anus without fistula
 - Cloacae with a common channel <3 cm
 - Rectourethral bulbar fistula

Clinical Signs Associated With Good Prognosis

- Good bowel movement patterns: 1 to 2 bowel movements per day, no soiling in between
- Evidence of sensation when passing stool

Anatomic Indicators of Poor Prognosis

- Abnormal sacrum
- Flat perineum (poor muscles)
- Some types of anorectal malformations:
 - Recto-bladder neck fistula
 - About 50% of patients who have rectoprostatic fistula
 - Cloacae with a common channel >3 cm
 - Complex malformations

Clinical Signs Associated With Poor Prognosis

- Constant soiling and passing stool
- No sensation (no pushing)
- Urinary incontinence, dribbling of urine

children need careful supervision to avoid fecal impaction and constipation, which could lead to overflow pseudo-incontinence.

If the child has a type of defect that carries a poor prognosis (high cloaca with a common channel longer than 3 cm, recto-bladder neck fistula), poor sacral development, or an associated spinal problem such as myelomeningocele, he or she most likely will need a bowel management program with enemas to remain clean. This regimen should be implemented when the child is 3 to 4 years old, before starting school.

A child born with an ARM and a rectoprostatic fistula has about a 50% chance of being capable of having voluntary bowel movements. (8) An attempt should be made to toilet train such children by age 3 years. If unsuccessful, bowel management with enemas should be implemented. Each summer, after the school year, attempts can be made to assess the child's ability to train.

Patients who have undergone surgery for ARM and have fecal incontinence may have rectums that were not placed precisely in the center of the sphincter mechanism. For those who have the potential for bowel control (good sacrum, good sphincter mechanism, malformation with good functional prognosis), reoperation to relocate the rectum may improve their potential for bowel control. About 50% of the children operated on for these specific circumstances have a significant improvement in bowel control. (11)

The ideal approach for treating patients who have true fecal incontinence is an enema program that involves teaching the patient and parents how to clean the colon mechanically once daily to stay completely clean between enemas. This result is achieved by choosing the right enema and making sure that the colon remains quiet between enemas. Because patients are not continent and cannot have voluntary bowel movements, they require the artificial mechanism of a daily enema to empty their colons.

The program is implemented by trial and error over a period of 1 week, with the patient seen each day, and each day an abdominal radiograph is obtained to monitor for the amount and location of stool in the colon as well as for any episodes of soiling. The daily radiographs are essential because they allow the clinician to make small adjustments in the regimen, such as modification of the type and volume of the enema, doses of any medications, and the diet. (7) Enemas and laxatives never are combined because the enema will clean the colon, but the laxative will provoke a bowel movement (and, thus, an accident) before the next enema washout.

Two well-defined groups of patients who have ARMs with fecal incontinence can be identified. The first and

larger group includes patients who have a tendency toward constipation. The second group has a tendency toward loose stool. Patients who manifest fecal incontinence after operations for HD and those who have spinal disorders usually fall into the constipated group, but some patients who have HD pass multiple loose stools and need treatment for hypermotility. (4)

True Fecal Incontinence and Constipation Associated With a Slow Colon

An 8-year-old girl born with spina bifida and a myelomeningocele has suffered from lifelong severe constipation and soiling. She has been treated with laxatives but tends to have loose stool four to five times per day. She does not feel when she needs to stool and must remain in diaper pants. A contrast enema demonstrates a mildly dilated rectosigmoid, and plain radiograph shows a fecal impaction, with stool present throughout the colon.

Because patients who have fecal incontinence and constipation associated with a slow colon lack the ability to have voluntary bowel movements, the basis of their bowel management program is to teach the parents how to clean the child's colon once a day with an enema. No specific diets or medications are necessary. The constipation (hypomotility) helps patients because the slow-moving colon remains quiet between enemas. If the enema is adequate, no stool passes until the next enema 24 hours later. The challenge is to find the ideal enema capable of cleaning the rectosigmoid, which is accomplished with a process of trial and error. Accidents or soiling episodes occur if the colon is not cleaned by the enema and stool passes thereafter. In such a case, a more potent enema is required. The enema itself may irritate the colon (eg, phosphate enemas), causing the patient to pass stool between enemas, although the radiograph appears clean. In this case, a gentler enema is needed.

Patients who have fecal incontinence and firm stool do not respond well to laxatives, which make their soiling worse. They need bowel management with a daily enema and no laxatives. Many children are categorized incorrectly and are treated with laxatives for years, continuing to soil. Such children typically have ARMs plus severe anomalies (abnormal sacrum, poor muscles) that carry a poor prognosis for continence.

True Fecal Incontinence and Loose Stools Associated With a Fast Colon

In the years before the introduction of the posterior sagittal anorectoplasty for the repair of anorectal malformations, procedures frequently included resection of the rectosigmoid. (9)(10)

Children who have a tendency toward loose stool have hyperactive colons, and they often lack rectal reservoirs.

Even when an enema cleans the colon rather easily, new stool passes quickly through the colon. To prevent this effect, a constipating diet, which we have developed over time through personal experience, and agents such as loperamide and pectin that slow the colon and add bulk are needed. Foods that loosen bowel movements should be avoided. Table 3 lists constipating foods and laxative foods to avoid. The diet is strictly regulated to food such as banana, apple, baked bread, white pasta without sauce, and boiled meats. Fried and oily foods as well as dairy products must be avoided. Some patients who have HD behave as if they have hypermotility and can be managed similarly.

A successful bowel management program requires dedication and sensitivity from the medical team. The goals are to clean the colon once a day, keep it quiet, and allow the patient to remain clean for the 24 hours after the enema. The regimen is an ongoing process of trial and error that is individualized and usually is successful within 1 week. (7) During that time, the family, patient, physician, and nurse learn to tailor the enema routine.

Implementing the Enema Program for True Fecal Incontinence

After obtaining a history and physical examination, the next step is to perform a contrast enema using water-

soluble material. Barium never should be used because it can lead to impaction. The postevacuation radiograph is important in showing the type of colon: dilated (constipated) (Fig. 1) or nondilated (tendency toward loose stool) (Fig. 2). The water-soluble contrast material also helps empty the colon. The enema type and volume can be estimated by the size of the colon on the study. We do not use manometry at our center because we have not found it helpful in evaluating or planning treatments or providing information that we cannot glean from the contrast study.

The results of the enema program are evaluated daily. Changes in the enema volume or content are made until achieving the goal of a clean colon between enemas. A daily radiograph of the abdomen is vital in determining whether the colon is empty.

There are different types of enema solutions. We use 0.9% saline (from a pharmacy or homemade using the recipe of 1½ teaspoons [7.5 mL] of salt added to 1,000 mL of water), usually in a volume of 400 to 750 mL. Phosphate enemas are convenient because they are available in a prepared vial, but they sometimes irritate the colon and cannot be used in patients who have renal insufficiency. The saline enema can be combined with glycerine (10 to 30 mL) or Castile soap (9 to 27 mL) to make a more effective enema.

The enema should result in a bowel movement within 30 to 45 minutes, followed by a period of 24 hours of complete cleanliness (Fig. 3). If that enema does not clean the colon completely (as demonstrated by radiography or if the child continues to soil), more aggressive treatment is required, and glycerine, Castile soap, or phosphate should be added to the saline solution. By learning from previous attempts and through a process of trial and error, the ideal enema can be determined.

For patients who have fast-moving colons, parents learn over time which foods provoke loose stools and which help in constipation. The treatment starts with enemas (usually 250 mL to 400 mL of saline), a very strict diet, loperamide, and pectin. Most children respond within 1 to 2 weeks. They stay on the strict diet until clean for 24 hours several days in a row and then are allowed to choose one new food every 2 to 3 days, observing the new food's effect on colonic motility. If the child soils after eating a newly introduced food, that food should be excluded from the diet. The most liberal diet possible is sought over several months, and the doses of the medications can be reduced gradually to the lowest effective dose to keep the child clean for 24 hours.

A 5-year-old child suffers from daily soiling and wears diaper pants. She was born with a rectoperineal fistula and

Table 3. **Constipating Diet**

Foods to Discourage (Foods That Loosen Bowels)

- Milk or milk products
- Fats
- Fried foods
- Fruits
- Vegetables
- Spices
- Fruit juices
- French fries
- Chocolate

Foods to Encourage (Foods That Bulk Stool)

- Applesauce
- Apples with skin
- Rice
- White bread
- Bagels
- Soft drinks (artificially sweetened)
- Bananas
- Pastas
- Pretzels
- Tea
- Potatoes
- Jelly (no jam)
- Broiled, boiled, baked meat, chicken, or fish

has a normal sacrum and spine. A radiograph shows colonic impaction with stool. An enema program is successful. Given her excellent potential for bowel control, she is started on a daily laxative and demonstrates the capacity for voluntary bowel movements.

Once the enema program is successful, parents often ask if the enemas will be needed for life. For patients born with no potential for bowel control, the answer is yes. However, because the spectrum of defects is wide, many patients have some potential. Our routine is to subject patients to the bowel management program with enemas first, so they are not exposed to embarrassing accidents. Often, with time, the child becomes more cooperative and more interested in his or her problem. After a period of a successful enema program, he or she may be able to control bowel movements successfully, following a regimen of a disciplined diet with regular meals, often with laxatives added to provoke bowel movements at predictable times. Having had time in clean underwear, albeit artificially with enemas, is advantageous to the child's future success with laxatives once he or she tries again for control. Each summer, the children who have some potential for continence can attempt to control their bowel movements without the help of enemas. They can try some training strategies during vacations, a time that they can stay home, and thereby avoid having an accident at school. This experimentation can be undertaken during a 1-week program, called a "laxative trial," again with daily radiographs and tailoring of a laxative regimen with the goal of eliminating the enema.

Pull-through Versus Permanent Stoma

An 8-year-old girl born with cloacal exstrophy had a colostomy performed at birth. She has been told that because of her lack of potential for continence, she must have a permanent stoma.

For patients who have colostomies and lack the potential for bowel control, a key question is whether a pull-through operation should be performed or whether they should be left with a permanent stoma. Many clinicians believe that, given the lack of continence common in these children, a permanent stoma provides a better quality of life. However, because of the success with the bowel management program (enema regimen), if a daily enema cleans the colon and no stool passes between enemas, the patient conceivably could be clean and wear normal underwear, despite a lack of potential for continence. We believe that this is a better option than a permanent stoma. (12)

The key factor, therefore, is whether the patient has the capacity to form solid stool. An option for these patients is to perform bowel management through the

stoma (Fig. 4). If the stoma remains quiet (usually with the help of a constipating diet, pectin, and loperamide) between enemas, it could be closed or pulled through and can be kept clean with a daily antegrade enema (via a Malone appendicostomy).

Antegrade Enema

When children receiving enemas are young (younger than school age), they do not mind the enemas, but many older children feel that their parents are intruding on their privacy by giving them enemas, and it is difficult for them to administer an enema themselves. A continent appendicostomy (Malone procedure) (13)(14)(15) can be performed whereby the appendix is connected to the umbilicus, through which the antegrade enema can be administered (Fig. 5). If the child has no appendix, a new one can be fashioned from a colonic flap. This opening is called a continent neoappendicostomy (Fig. 6). Because the Malone procedure only changes the route of enema administration, the child must be perfectly clean with a bowel management regimen before the procedure. This is a particularly useful option for patients who have spinal problems and are in wheelchairs.

Pseudoincontinence (Encopresis)

An 8-year-old boy who has had no prior surgery has encopresis with daily soiling and wears diaper pants to school. He has had severe constipation since starting on regular foods as a toddler. He has a large bowel movement approximately once weekly and has suffered from several anal fissures.

True fecal incontinence must be distinguished from overflow pseudoincontinence, which occurs when a patient behaves as if fecally incontinent, but really has severe constipation with overflow soiling (encopresis). Once the child is disimpacted and given sufficient laxatives to empty the colon on a daily basis, the soiling stops.

Normally, the rectosigmoid stores the stool and an active peristaltic wave occurs approximately every day that indicates the need to empty. Most individuals feel the wave, tighten the muscles surrounding the anus, and decide when to relax the sphincter mechanism. For many children, this process of relaxation is slow, leading to severe constipation over time.

If a child is fecally continent, constipation must be treated with laxatives, which are needed to provoke peristalsis and overcome the hypomotility. Patients who have undergone successful surgery for ARMs (associated with a good prognosis type of anorectal defect and a normal spine) or HD should be fecally continent. Patients who suffer idiopathic constipation or encopresis also have intact conti-

nence mechanisms and should be capable of having voluntary bowel movements. These patients, in our opinion, do not need enemas and, therefore, appendicostomies (16)(17) and stomas. (18) They need only the appropriate medical therapy with stimulant laxatives.

Severe constipation is common in patients who have ARMs (particularly the more benign types), (8) in patients following successful surgery for HD, and in a large group of patients considered to have idiopathic constipation. Untreated constipation can be extremely incapacitating and, in its most serious form, can produce soiling that really is overflow pseudoincontinence (encopresis).

The therapeutic value of dietary changes is minimal for this group of patients. Passage of large, hard pieces of stool may provoke pain and make patients behave as if they are stool retainers, ultimately leading to soiling, which may have a psychological impact.

Clinicians must determine the type of patient with whom they are dealing. Pseudoincontinence can be associated with enuresis and urinary tract infection. Once the stool problem is fixed, urinary control often follows. Those whose prognosis for bowel control is good need aggressive, proactive treatment of their constipation.

Usually these patients suffer from a megarectosigmoid (Fig. 1), which results from a vicious cycle of hypomotility, constipation, inadequate colonic emptying, and colon dilatation. (1) This cycle is observed in children born with a good prognosis type of anorectal defect who underwent technically correct operations but did not receive adequate treatment for constipation. Over time, they develop fecal impaction and overflow pseudoincontinence. This outcome also can occur in children who have severe idiopathic constipation (encopresis), who tend to have a similarly appearing dilated rectosigmoid.

Diagnosis

A contrast enema with a hydrosoluble material is the most valuable diagnostic study. In the severely constipated patient, this study usually shows a megarectosigmoid, with dilatation of the colon all the way down to the levator mechanism (Fig. 1). A dramatic size discrepancy usually is evident between a normal descending colon and a dilated megarectosigmoid. This is the exact opposite pattern from that seen in patients who have HD. The size of the colon guides the dosing of the laxatives.

Rectal and colonic manometry have been used in patient evaluation, (19)(20) although we have not found these modalities helpful. Attempts to assess colonic motility objectively include manometry, which records the waves of contraction (19) and electrical activity, (21) or scintigraphy, a nuclear medicine study. (22) The key information for the

surgeon is if and where a colonic resection would benefit the patient who requires enormous doses of laxative to empty. Unfortunately, these modalities do not answer this question consistently. We are hopeful that more objective techniques will be developed.

Histologic investigation of these colons reveals only hypertrophic smooth muscle in the area of the dilated colon and normal ganglion cells. We clearly need more sophisticated histopathologic techniques to help understand what is wrong with these colons. These patients do not have HD, but their dysmotility is not yet understood. We find the concepts of “very low segment HD” or “internal sphincter achalasia” (23) confusing and are hopeful that further investigations will enhance our knowledge about colonic dysmotility and, thus, guide therapy.

Treatment

At our center, we treat patients empirically based on the contrast study, inferring dysmotility in the dilated segment of the colon. The mandatory first step, which often is neglected, is to remove the child’s fecal impaction, which is accomplished with enemas and sometimes a bowel clean-out. Once the colon is clean, the constipation is treated with the administration of large doses of stimulant laxatives, not stool softeners. Stimulant laxatives provoke the peristaltic wave; stool softeners only soften the stool and do not help the motility problem. If the patient is not disimpacted first, laxatives cause cramping. In these cases, the laxatives we use often are the same as those that have been tried previously, but the protocol is more aggressive. The laxative dose is adapted to the patient’s response, which is monitored daily with an abdominal radiograph, and the laxative dose is adjusted until the amount that successfully empties the colon every day is reached. Almost always, the new dose of laxative is much more than had been tried previously.

The colonic dysmotility seen in certain patients who have HD, even after successful surgery that removed the aganglionic bowel, is not understood. These patients, like those who have ARMs, benefit from early and proactive medical treatment of their constipation. The cause of severe constipation and encopresis in many children cannot be determined, and proactive and aggressive laxative therapy can be helpful. The dysmotility is incurable, but it is manageable. Treatments must be consistent, and patients require close observation. Once treatments are tapered or interrupted, constipation recurs.

Some clinicians treat such “intractable” patients with colostomies or with antegrade enemas through a catheter in a stoma. (17)(18) We feel that these patients should be treated instead with stimulant laxatives. We reserve wash-outs for patients who have true fecal incontinence and are

incapable of having voluntary bowel movements. We definitely do not feel that any such patient needs a colostomy.

If the child is incontinent, enemas as part of a bowel management regimen are appropriate. If continent, maintenance treatment of constipation with laxatives is the treatment of choice.

Determination of the Laxative Requirement

Once the colon is disimpacted, a dose of stimulant laxative is started empirically based on the contrast enema results. A senna derivative is ideal. After administration, the patient is observed for 24 hours. If the child does not have a bowel movement in that time, the laxative dose must be increased. An enema should be given to remove the stool produced during the previous 24 hours. If the patient stools multiple times and the radiograph shows clean bowel, the laxative dose can be decreased. The routine of choosing the laxative dose and giving an enema if needed is continued every night, until the dose of laxative is found that provokes a voluntary bowel movement and empties the colon completely (as confirmed by radiograph). If the laxative empties the colon effectively but the stool is too loose, adding pectin provides bulk and makes the same laxative dose more effective.

Patients may have laxative requirements much larger than the manufacturer's recommendation. Usually the dose needed to empty the colon completely can be achieved. At that dose, the patient should stop soiling because he or she is emptying the colon effectively each day. Because the colon is empty, the patient remains clean until the next voluntary bowel movement. Occasionally, during the process of increasing the amount of laxatives, patients vomit before reaching an adequate effect or do not tolerate such a high dose because it causes nausea and cramping. Such patients are candidates for surgical intervention.

Surgical Options: Rectosigmoid Resection

A sigmoid resection can be performed in select patients who have repaired ARMs and severe constipation. (15)(24) The very dilated megarectosigmoid is resected, and the descending colon is anastomosed to the rectum (Fig. 7). Because the rectum plays a vital reservoir role that allows the patient to perceive proprioception of rectal distention, it is preserved. The remaining rectum is abnormal, and without careful observation and treatment of constipation, the colon proximal to it can redilate.

For patients who have intractable idiopathic constipation but normal sphincters and a normal anal canal, the sigmoid likewise can be resected. In cases of a huge rectum, sometimes the rectosigmoid down to the pectinate line can be resected, in a similar manner used for

patients who have HD. The nondilated colon (which is assumed to have normal motility) is anastomosed to the rectum above the pectinate line (Fig. 8). (15) This procedure is a good alternative to traditional recommendations such as colostomy or antegrade stomas for the most severely affected, intractable patients. (17)(18)

The most dilated part of the colon is resected because it is considered empirically to be affected most seriously. The nondilated part of the colon is assumed to have a more normal motility, which we infer from the contrast enema. Perhaps emerging colonic motility diagnostic techniques will help with surgical planning. We have observed that the patients who improve the most are those who have a more localized form of megarectosigmoid. Patients who have more generalized dilation of the colon do not respond as well and may require a more extensive resection. Perhaps in the future, these observations can be corroborated and results of resection predicted better by noninvasive modalities.

Summary

- The goal for treatment of a child who has fecal incontinence is to have the child clean and wearing normal underwear, which is achieved either with medical treatment for patients who have the potential for bowel control or artificially with enemas for patients who have true fecal incontinence.
- The program is an ongoing process of trial and error, is responsive to the individual patient, and differs for each child. The laxatives or the enema empty the colon, and if the colon remains quiet for the next 24 hours until the next treatment, the child stays clean.
- Our routine is to employ either the laxative trial or the enema regimen during a 1-week outpatient program, with a daily abdominal radiograph checked as the regimen is tailored. Based on strong clinical evidence reviewing hundreds of children, more than 95% of children who follow this program become clean and dry. (7)
- Clinicians should embrace the philosophy that no child who has fecal incontinence should go to school in diapers when classmates are already toilet trained. Successful anatomic reconstruction does not do much for a patient who continues to soil. Proper treatment to keep a patient clean and dry is perhaps more important than any surgical procedure itself.

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problems described here and which seeks a practical and organized approach to helping them.

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PIR Quiz

Quiz also available online at <http://pedsinreview.aappublications.org>.

1. A 5-year-old girl who has severe constipation and soiling and who was born with a myelomeningocele and has sensory and motor dysfunction of the lower extremities can be described as having:
 - A. Aganglionsis.
 - B. Capability of having voluntary bowel movements.
 - C. Overflow fecal incontinence with constipation.
 - D. True fecal incontinence with constipation.
 - E. True fecal incontinence with hypermotility.
2. The treatment for the previously described 5-year-old girl should include:
 - A. Daily enema.
 - B. Laxatives.
 - C. Loperamide and pectin.
 - D. Potty training strategies.
 - E. Surgical correction.
3. An 8-year-old boy who has had no prior surgery experiences soiling and encopresis. Management should include:
 - A. An enema if no stool in any 24-hour period.
 - B. Appropriate laxative dosing.
 - C. Disimpaction first.
 - D. Regular radiographs to ensure emptying.
 - E. All of the above.
4. An 8-year-old boy, a recent immigrant, has had constipation since infancy. He stooled normally at birth. He now soils daily despite retrograde enemas once a week. His general physical examination, except for a malleable mass in the left lower abdomen, yields normal results. Rectal examination reveals normal tone. Stool is palpated on the examining finger. The *most* likely finding on radiography with water-soluble contrast material is:
 - A. Dilated colon with normal rectosigmoid.
 - B. Dilated proximal colon, transition zone, contracted distal rectosigmoid.
 - C. Narrow left colon with megarectosigmoid.
 - D. Normal colon and rectosigmoid.
 - E. Normal colon except for megarectosigmoid.
5. You have diagnosed severe idiopathic constipation with soiling in the previously described 8-year-old patient. Of the following, the *most* appropriate initial therapy is:
 - A. Aggressive use of stimulant laxatives.
 - B. Daily loperamide.
 - C. Fecal disimpaction.
 - D. Increased intake of bananas, apples, and pasta.
 - E. Stool softener twice daily.

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