Pediatric Fecal Incontinence

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Introduction

Fecal incontinence represents a devastating problem for all those who suffer from it. It often prevents a person from becoming socially accepted, which in turn provokes serious psychological sequelae. It is a problem that impacts more children than previously thought, affecting those born with anorectal malformations and Hirschsprung's disease as well as children with spinal cord problems or spinal injuries.

True fecal incontinence must be distinguished from overflow pseudoincontinence. Pediatric patients with true fecal incontinence include some surgical patients with anorectal malformations and Hirschsprung's disease and patient with spinal problems, either congenital or acquired. Those with pseudoincontinence are patients with potential for bowel control but who suffer from overflow or encopresis related to severe constipation.

Most patients who undergo an anorectal malformation repair suffer from some degree of functional defecation disorder, and all suffer from an abnormality in their fecal continence mechanism. Approximately 25% of patients are deficient enough in these mechanisms that they are fecally incontinent and cannot have a voluntary bowel movement. The others are capable of having voluntary bowel movements but may require treatment of an underlying dysmotility disorder, which manifests as constipation [1]. A small yet significant number of patients with Hirschsprung's disease (<5%) suffer from fecal incontinence. Patients with spinal problems or injuries can lack the capacity for voluntary bowel movements or have this ability only to varying degrees.

Patients with true fecal incontinence require an artificial method to keep them clean and in normal underwear, a regimen termed bowel management. Patients with pseudoincontinence require proper treatment of constipation. Understanding this major differentiation is the key to deciding on correct management.

Continence Mechanism

Fecal continence depends on three main factors: voluntary sphincter muscles, anal canal sensation, and colonic motility [1].

Voluntary Muscle Structures

In the normal patient, voluntary muscle structures are represented by the levators, the muscle complex, and the external sphincter. Normally, they are used only for brief periods when the rectal fecal mass reaches the anorectal area, pushed by the involuntary peristaltic contraction of rectosigmoid motility. This voluntary contraction occurs only in the minutes prior to defecation, and these muscles are used only occasionally during the rest of the day and night.

Patients with anorectal malformations have abnormal voluntary striated muscles with different degrees of hypodevelopment. Patients with spinal problems or injuries can have varying degrees of sphincter dysfunction. Voluntary muscles can be used only when the patient has the sensation that it is necessary to use them. To appreciate that sensation, the patient needs information that can only be derived from an intact anal sensory mechanism, a mechanism that many patients with anorectal malformations and spinal problems lack.

Anal Canal

Exquisite sensation in normal individuals resides in the anal canal. Except for patients with rectal atresia, most patients with anorectal malformations are born without an anal canal; therefore, sensation does not exist or is rudimentary. Patients with Hirschsprung's disease are born with a normal anal canal, but this can be injured if not meticulously preserved at the time of their colonic pull through. Patients with perineal trauma may have an injured or destroyed anal canal.

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It seems that patients can perceive distention of the rectum, but this requires a rectum that has been properly located within the muscle structures, a surgical point quite important for patients undergoing pull-through procedures for an imperforate anus. This sensation seems to be a consequence of voluntary muscle stretching (proprioception). The most important clinical implication of this situation is that the patient may not feel liquid stool or soft fecal material, as it does not distend the rectum. Thus, to achieve some degree of sensation and bowel control, the patient must have the capacity to form solid stool. This point is quite relevant in children with ulcerative colitis who have undergone an ileoanal pull through. They may suffer from varying degrees of incontinence due to the incapacity to form solid stool. In the majority of cases, normal sphincter muscles and anal canal allow them to overcome this problem.

Bowel Motility

Perhaps the most important factor in fecal continence is bowel motility; however, its impact has been largely underestimated. In a normal individual, the rectosigmoid remains quiet for variable periods (1 to several days), depending on specific defecation habits. During that time, sensation and voluntary muscle structures are almost not necessary because the stool, if it is solid, remains inside the colon. The patient feels the peristaltic contraction of the rectosigmoid that occurs prior to defecation. The normal individual can voluntarily relax the striated muscles, which allows the rectal contents to migrate down into the highly sensitive area of the anal canal. There, the anal canal provides accurate information concerning stool consistency and quality. The voluntary muscles are used to push the rectal contents back up into the rectosigmoid and to hold them until the appropriate time for evacuation. At the time of defecation, the voluntary muscle structures relax.

The main factor that provokes rectosigmoid emptying is a massive involuntary peristaltic contraction sometimes helped by a Valsalva maneuver. Most patients with an anorectal malformation suffer from a disturbance of this sophisticated bowel motility mechanism. Patients who have undergone a posterior sagittal anorectoplasty or any other type of sacroperineal approach, in which the most distal part of the bowel was preserved, show evidence of an overefficient bowel reservoir (megarectum). The main clinical manifestation of this is constipation, which seems to be more severe in patients with lower defects [2]. Constipation that is not aggressively treated, in combination with an ectatic distended colon, eventually leads to severe constipation, and a vicious cycle ensues, with worsening constipation leading to more rectosigmoid dilation, leading to worse constipation. The enormously dilated rectosigmoid, with normal ganglion cells, behaves like a myopathic type of hypomotile colon [1].

Patients with anorectal malformation treated with techniques in which the most distal part of the bowel was resected behave clinically as individuals without a rectal reservoir. This is a situation equivalent to a perineal colostomy. Depending on the amount of colon resected, the patient may have loose stools. In these cases, medical management consisting of enemas plus a constipating diet and medications to slow down colonic motility is indicated. Patients with Hirschsprung's disease have undergone distal aganglionic colon resection, but it is their normal anal canal and sphincter mechanism that allows the vast majority of them to be continent despite the lack of a rectal reservoir. Amazingly, some patients with an injured anal canal and sphincters (perineal trauma) can be continent if their motility is normal, and the regular contraction of the rectosigmoid can be translated into a successful voluntary bowel movement.

True Fecal Incontinence

For patients with true fecal incontinence, the ideal treatment approach is a bowel management program consisting of teaching the patient and his or her parents how to clean the colon once daily so it stays completely clean for 24 h. This is achieved by keeping the colon quiet between enemas. These patients cannot have voluntary bowel movements and require an artificial mechanism to empty their colon: a daily enema. The program, although simplistic, is implemented by trial and error over a period of 1 week. The patient is seen by the physician each day, and an abdominal X-ray is taken so that the patient can be monitored on a daily basis for the amount and location of any stool left in the colon. Presence of stool in the underwear is also noted. The decision as to whether enema type and/or quality should be modified, as well as changes in diet and/or medication, can be made daily [3].

Which Pediatric Patients have True Fecal Incontinence?

In children with anorectal malformations, 75% who have undergone a correct and successful operation have voluntary bowel movements after the age of 3 years [2]. About half of these patients soil their underwear on occasion. Those episodes of soiling are

usually related to constipation. When the constipation is properly treated, soiling frequently disappears. Thus, approximately 40% of all children with anorectal malformations have voluntary bowel movements and no soiling. In other words, they behave like normal children. Children with good bowel control still may suffer from temporary episodes of fecal incontinence, especially when they experience severe diarrhea.

Some 25% of all children suffer from real fecal incontinence, and they are the patients who need bowel management to keep them clean. As noted, certain patients with Hirschsprung's disease and those with spinal problems can suffer from true fecal incontinence. For these patients, similar principles of bowel management that have proven effective in treating patients with anorectal malformations [3] can be applied.

For children with anorectal malformations, the surgeon should be able to predict which ones may have a good functional prognosis and which ones may have a poor prognosis. Table 1 shows the most common indicators of good and poor prognoses. After the main repair and colostomy closure, it is possible to establish the functional prognosis (Table 2). Parents must be informed of their child's realistic chances for bowel control, thus avoiding needless frustration later. It is imperative to establish the functional prognosis of each child as early as possible, which sometimes is possible even in the newborn period, to avoid creating false expectations for the parents.

Once diagnosis of the specific anorectal defect is established, functional prognosis can be predicted. If the child's defect is of a type associated with good prognosis–such as a vestibular fistula, perineal fistula, rectal atresia, rectourethral bulbar fistula, or imperforate anus with no fistula–the child can be expected to have voluntary bowel movements by the age of 3 years. These children will still need supervision to avoid fecal impaction, constipation, and soiling.

If the child's defect is of the type associated with a poor prognosis–for example, a very high cloaca with a common channel longer than 3 cm, a rectobladder-neck fistula, or if they have a very hypodeveloped sacrum-parents must understand that their child will most likely need a bowel management program to remain clean. This program should be implemented when the child is 3–4 years of age, before starting school. Children with rectoprostatic fistulas have an almost 50-50 chance of having voluntary bowel movements or of being incontinent. In these children, an attempt should be made to achieve toilet training by the age of 3 years. If this proves unsuccessful, bowel management should be implemented. Each summer, after school is finished, reattempts can be made to assess the child's ability to potty train.

In patients previously operated on for an imperforate anus with fecal incontinence, a reoperation to

Table 1. Prognostic signs for patients with anorectal malformations. From [4]

Good prognosis signs	Poor prognosis signs
- Good bowel movement pattern: 1–2 bowel movement per day – no soiling in between - Evidence of sensation when passing stool (pushing, making faces) - Urinary control	- Constant soiling and passing of stool - No sensation (no pushing) - Urinary incontinence, dribbling of urine

Table 2. Predictors of prognosis in patients with anorectal malformations. From [4]

Indicators of good prognosis for bowel control	Indicators of poor prognosis for bowel control
- Normal sacrum - Prominent midline groove (good muscles) - Some types of anorectal malformations: • Rectal atresia • Vestibular fistula • Imperforate anus without a fistula • Cloacas with a common channel <3 cm • Less complex malformations: perineal fistula	- Abnormal sacrum - Flat perineum (poor muscles) - Some types of anorectal malformations: • Rectobladder-neck fistula • Cloacas with a common channel >3 cm • Complex malformations

relocate a misplaced rectum with the hope of obtaining good bowel control should be considered if the child was born with a good sacrum, good sphincter mechanism, and a malformation with good functional prognosis. A redo posterior sagittal anorectoplasty can be performed, and the rectum can be relocated within the limits of the sphincter mechanism. Approximately 50% of children operated on under these very specific circumstances have significant improvement in bowel control [5].

Patients with fecal incontinence and a tendency toward constipation cannot be treated with laxatives but need bowel management. In fact, laxatives in such patients make their soiling worse. Such children are usually those born with a poor prognosis type of defect and severe associated defects (defect of the sacrum, poor muscle complex).

Children operated on for imperforate anus who suffer from fecal incontinence can be divided into two well-defined groups, each requiring individualized treatment plans. The first and larger group includes patients with fecal incontinence and a tendency toward constipation. The second group comprises fecally incontinent patients with a tendency toward loose stool. Patients with fecal incontinence after operations for Hirschsprung's disease and those with spinal disorders usually fall into the first group: those with a tendency toward constipation.

Children with Constipation (Colonic Hypomotility)

In these children, colon motility is significantly reduced. The basis of the bowel management program in these patients is to teach parents to clean the child's colon once a day with a suppository, an enema, or colonic irrigation. No special diet or medications are necessary. The fact that these children suffer from constipation (hypomotility) is helpful, as it helps them remain clean between enemas. The real challenge is to find an enema capable of completely cleaning the colon. Definitive evidence that the colon is truly empty following an enema requires a plain abdominal radiograph. Soiling episodes or "accidents" occur when there is incomplete bowel cleaning and feces that progressively accumulates.

Children with Loose Stools and Diarrhea

The great majority of children with anorectal malformations who suffer from this kind of problem were repaired before 1980 and the introduction of the posterior sagittal technique. During those years, the procedures frequently included rectosigmoid resection [6, 7], Therefore, this group of children has an overactive colon because they lack a rectal reservoir. Rapid stool transit results in frequent diarrhea episodes. This means that even when an enema cleans their colon rather easily, stool keeps passing fairly quickly from the cecum to the descending colon and anus. To prevent this, a constipating diet and/or medications to slow down the colon are necessary. Eliminating foods that further loosen bowel movements will help the colon slow down. A small subset of patients with Hirschsprung's disease behaves as though they have hypermotility and can be managed similarly.

The keys to success of this bowel management program are dedication and sensitivity from the medical team. The basis of the program is to clean the colon and keep it quiet, thus keeping the patient clean for the 24 h after the enema.

The program is an ongoing process that is responsive to the individual patient and differs for each child. It is usually successful within a week, during which time family, patient, physician, and nurse undergo a process of trial and error, tailoring the regimen to the specific patient. More than 95% of children who follow this program are artificially clean and dry for the whole day and can have a completely normal life. One should embrace the philosophy that it is unacceptable to send a child with fecal incontinence to school in diapers when his classmates are already toilet trained. Proper treatment to prevent this is perhaps more important than the surgical procedure itself.

The first step in the program is to perform a contrast enema study with hydrosoluble material. The study should never be done with barium, and it is also important to obtain a picture after contrast material evacuation. This study shows the type of colon: dilated (constipated) Figure 1 or nondilated (tendency toward loose stool) (Fig. 2).

The bowel management program is then implemented according to the patient's type of colon, and results are evaluated daily. Changes in enema volume and content are made until the colon is successfully cleaned. To achieve this, a daily abdominal X-ray is invaluable in determining whether the colon is empty.

There are different types of enema solutions: some are ready-made and can be bought in a drugstore, and some can be prepared at home based on water and salt (0.9% saline can be made by adding 2 teaspoons of salt to 960 ml of water). The use of phosphate enemas is most convenient, as they are available in a prepared vial. However, saline enemas are often just as effective, and some families find them easier and less expensive. Occasionally, children will complain of cramping with the phosphate enema but

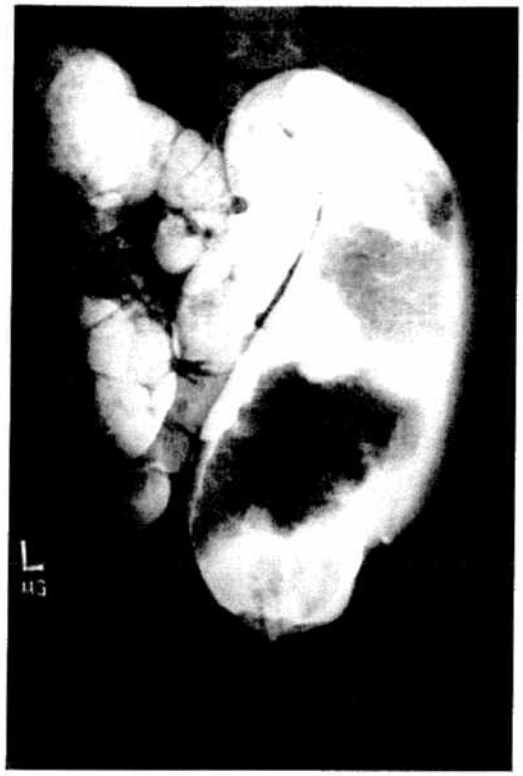

Fig. 1. Contrast enema of megarectosigmoid. Reprinted with permission from [1]

have no complaints with the saline one. Children older than 8 years or heavier than 30 kg may receive one adult phosphate enema daily (133 ml). Children between 3 and 8 years or between 15 and 30 kg may receive one pediatric phosphate enema (70 ml) daily. Children should never receive more than one phosphate enema a day because of the risk of phosphate intoxication, and others with impaired renal function should use these enemas with caution.

The enema administered on a regular basis should result in a bowel movement within 30–45 min, followed by a period of 24 h of complete cleanliness. If one enema is not enough to clean the colon (as demonstrated by an X-ray, or if the child keeps soiling), then the child requires a more aggressive treatment program, and saline solution is added to the phosphate one. If addition of the saline enema still produces inadequate results, then glycerin can be added. Administering the enema with a balloon catheter may help prevent enema leakage. The "right" saline enema is the one that can empty the child's colon and allow him or her to stay clean for the following 24 h. This can be achieved only by trial and error and learning from previous attempts.

Children with loose stool have an overactive colon and most often have no rectal reservoir. This means that even when an enema cleans their colon rather easily, new stool passes quickly from the cecum to the descending colon and anus. To prevent this, a constipating diet, bulking agents, and/or medications (such as loperamide) to slow down the colon are recommended. Eliminating foods that loosen bowel movements will help the colon move more slowly.

Parents are provided with a list of constipating foods to be given and a list of laxative foods to be avoided. The diet is rigid: banana, apple, baked bread, white pasta with no sauce, boiled meat, etc. Fried foods and dairy products must be avoided (Table 3). Most parents know which meals provoke loose stools and which ones constipate their child. To determine the right combination, the treatment starts with enemas, a very strict diet, and loperamide. Most children respond to this aggressive management within several days. The child should remain on a strict diet until clean for 24 h for 2–3 consecutive days. They can then choose one new food every 2–3 days, and the parent observes the effect on his or her colonic activity. If the child soils after eating a newly introduced food, that food must be eliminated. Over several months, the most liberal diet possible should be sought. If the child remains clean with a liberal

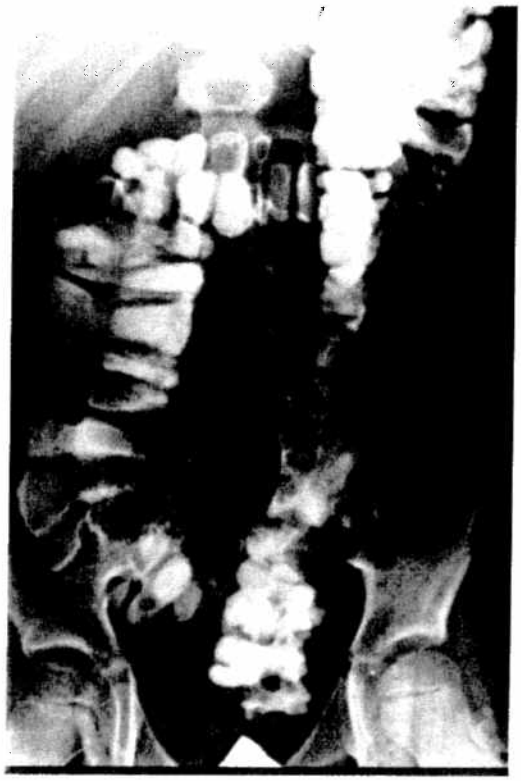

Fig. 2. Contrast enema in a patient with resected rectosigmoid. Reprinted with permission from [8]

Table 3. Constipating foods. From [4]

Constipating diet	
Avoid	**Encourage**
Milk or milk products	Apple Sauce
Fats	Apples without skin
Fried foods	Rice
Fruits	White bread
Vegetables	Bagels
Spices	Soft drinks
Fruit juices	Banana
French fries	Pasta
Chocolate	Pretzels
	Tea
	Potato
	Jelly (no jam)
	Boiled, broiled, baked
	Meat, chicken or fish

diet, medication can gradually be reduced to the lowest effective dose necessary to keep the child clean for 24 h.

In children in whom a successful bowel management program has been implemented, parents frequently ask if this program will be needed for life. The answer is "yes" for those patients born with no potential for bowel control. However, because we are dealing with a spectrum of defects, there are patients with some degree of bowel control. These patients are subjected to the bowel management program in order not to be exposed to embarrassing accidents of uncontrolled bowel movements. However, as time goes by, the child becomes more cooperative and more interested in his or her problem. It is conceivable that later in life, a child may be able to stop using enemas and remain clean, following a specific regimen of a disciplined diet with regular meals (three meals per day and no snacks) to provoke bowel movements at a predictable time. Every summer, children with some potential for bowel control can try to determine how well they can control their bowel movements without the help of enemas. This is done during vacations to avoid accidents at school, a time when they can stay home and try some potty training strategies.

For patients with a colostomy and no potential for bowel control, a key question is whether to perform a pull through or to leave the permanent stoma. We feel that if patients have the capacity to form solid stool, a pull through can be performed, with a daily enema to keep them clean. We believe that for these patients, successful bowel management gives a better quality of life than does a permanent stoma.

Most preschool and school-aged children enjoy a good quality of life while undergoing the bowel management program. However, when they reach puberty, many express a high degree of dissatisfaction. They feel that their parents are intruding on their privacy by giving them enemas. It is feasible but rather difficult for them to administer the enema themselves. For this specific group of children, an operation called a continent appendicostomy or a Malone procedure has been designed [9] whereby the appendix is connected to the umbilicus and through which the enema can be administered (Fig. 3). A valve mechanism is created that allows catheterization of the appendix for the enema fluid but avoids leakage of stool through it. If the child has lost his or her appendix, it is possible to create a new one from the colon. This procedure is known as a continent neoappendicostomy.

It is important to stress that the Malone procedure is just another way of administering an enema, and therefore, before it is performed, the child must be perfectly clean with a bowel management regimen.

Pseudoincontinence

It is vital to differentiate real fecal incontinence from overflow pseudoincontinence. As in patients with real fecal incontinence, the normal bowel control mechanism is deficient. Pseudoincontinence occurs when a patient behaves as if they are fecally incontinent, but they really have severe constipation and overflow soiling. Once the disimpaction is treated and the patient receives enough laxatives to avoid constipation, he or she becomes continent.

The colon absorbs water from the stool and serves a reservoir function. These processes depend on colonic motility, an area of physiology not well understood and for which treatments of problems are limited. In normal individuals, the rectosigmoid stores the stool and every 24–48 h 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.

If a child is fecally continent, then pseudoincontinence management involves treatment of constipation using laxatives, which help provoke peristalsis and overcome the dysmotility disorder. Patients who have undergone successful surgery for Hirschsprung's disease and for anorectal malformations (with a good prognosis type of anorectal defect) and have normal spines should be fecally continent.

Constipation in anorectal malformations is extremely common, particularly in the more benign types [10]. It is also common in patients following successful surgery for Hirschsprung's disease and occurs in a large group of patients considered to have idiopathic constipation [1]. When left untreated,

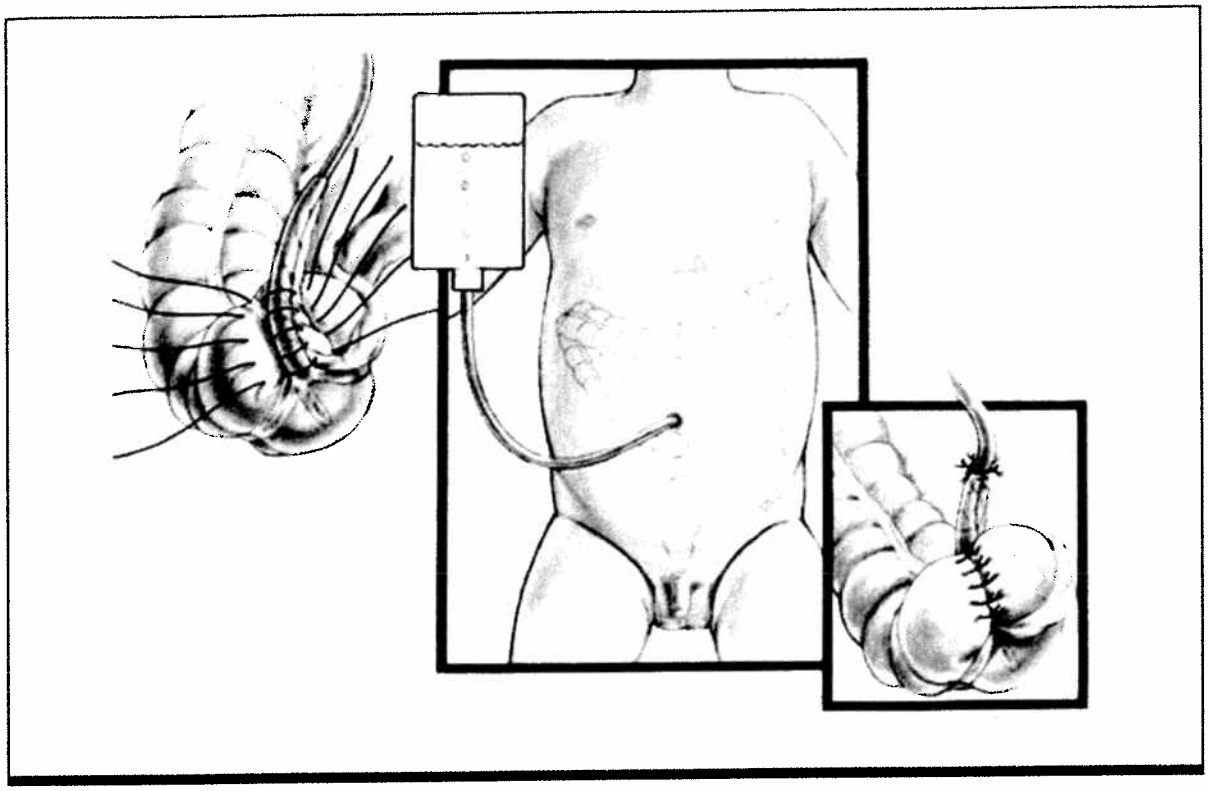

Fig. 3. Malone appendicostomy. Reprinted with permission from [9]

constipation can be extremely incapacitating and in its most serious forms can produce a form of fecal incontinence known as overflow pseudoincontinence. Diet impacts colonic motility, but its therapeutic value is negligible in the most serious forms of constipation. It is true that many patients with severe constipation suffer from psychological disorders, but a psychological origin cannot explain severe forms, as it is not easy to voluntarily retain the stool when an otherwise autonomous rectosigmoid peristalses. Passage of large, hard pieces of stool may provoke pain and make the patient behave as if they are stool retainers. This may complicate the problem, but it is not the original cause.

The clinician must decide which type of patient he or she is dealing with. Patients with good prognosis for bowel control are more likely to have constipation, and aggressive, proactive treatment of their constipation is the best approach. The child must be deemed capable of being fecally continent and have the capacity for voluntary bowel movements before initiating treatment for constipation.

Most of these patients suffer from different degrees of dilation of the rectum and sigmoid, a condition defined as megarectosigmoid (Fig. 1), due to a hypomotility disorder that interferes with complete rectosigmoid emptying [1]. These patients may be children born with a good prognosis type of anorectal defect and who underwent a technically correct operation but did not receive appropriate treatment for constipation. They therefore developed fecal impaction and overflow pseudoincontinence. These may also be children with severe idiopathic constipation who have a very dilated rectosigmoid.

Impaction needs to be removed with enemas and colonic irrigations to clean the megarectosigmoid. Subsequently, the constipation is treated with the administration of large doses of laxatives. Laxative dosage is increased daily until the right amount of laxative is reached to completely empty the colon every day. If medical treatment proves to be extremely difficult because the child has a severe megasigmoid and requires an enormous amount of laxatives to empty, the surgeon can offer a segmental resection of the colon. After the sigmoid resection, the amount of laxatives required to treat these children can be significantly reduced or even eliminated. Before performing this operation, it is mandatory to confirm that the child is definitely suffering from overflow pseudoincontinence rather than true fecal incontinence with constipation. Failure to make this distinction may lead to an operation in which a fecally incontinent constipated child is

changed to one with a tendency to have loose stool, which will make them much more difficult to manage.

When children with anorectal malformations and Hirschsprung's disease are managed from the beginning with aggressive treatment of constipation, children with good prognosis should potty train without difficulty. When constipation is not managed properly and a patient presents after many years, they behave much like children with idiopathic constipation and have overflow pseudoincontinence.

Constipation in anorectal malformations is a self-perpetuating disease. A patient suffering from a certain degree of constipation that is not treated adequately only partially empties the colon, leaving larger and larger amounts of stool inside the rectosigmoid, which results in greater degrees of megasigmoid. It is clear that dilatation of a hollow viscus produces poor peristalsis, which explains the fact that constipation leads to fecal retention and thereafter megacolon, which exacerbates constipation. In addition, the passage of large, hard pieces of stool may produce anal fissures that result in reluctance by the patient to have bowel movements.

Colon dysmotility in patients with Hirschsprung's disease, even after successful surgery to remove the aganglionic bowel, is not understood. These patients do, however, benefit from proactive medical treatment of their constipation. The clinician must accept the fact that dysmotility is essentially incurable. It is, however, manageable, but requires careful life-long follow-up. Treatments cannot be given on a temporary basis, as once they are tapered or interrupted, constipation recurs.

Some clinicians treat such patients with colostomies or colonic washouts via a catheterizable stoma or button device and monitor the degree of colonic dilatation with contrast studies [11]. Once the distal colon regains a normal caliber, the physician assumes that the patient is cured, and the colostomy is closed or the washouts are discontinued. Unfortunately, symptoms quickly recur. We believe that washouts are really only for patients with true fecal incontinence who are incapable of having voluntary bowel movements and thus require a daily irrigation to empty. Patients with pseudoincontinence are capable of emptying their colon with the help of adequate doses of laxatives and thus do not need washouts.

Determining with which patient the clinician is dealing is the challenge. If the patient is incontinent, washouts with a bowel management regimen are appropriate. If the patient is continent, then aggressive constipation management after ensuring disimpaction is the treatment choice.

Fecal impaction is a stressful event of retained stool for several days or weeks, crampy abdominal pain, and sometimes tenesmus. When laxatives are prescribed to such a patient, the result is exacerbation of the crampy abdominal pain and sometimes vomiting. This is a consequence of increased colonic peristalsis (produced by the laxative) acting against a fecally impacted colon. Therefore, disimpaction, proven by X-ray, must precede initiation of laxative therapy.

Underwear soiling is an ominous sign of bad constipation. A patient who at an age of bowel control soils their underwear day and night and basically does not have spontaneous bowel movements may have overflow pseudoincontinence. These patients behave similarly to fecally incontinent individuals. When the constipation is treated adequately, the great majority of these pseudoincontinent children regain bowel control. Of course, this clinical presentation may also occur in a patient with true fecal incontinence. When uncertain, the physician can start a three-and-a-half to four-year-old child having trouble with potty training on a daily enema. Once the child is clean with this regimen and if he or she has the potential for bowel control, then a laxative program can be attempted.

A contrast enema with a hydrosoluble material (never barium) is the most valuable study, which in the constipated patient usually shows a megarectosigmoid with colon dilatation all the way down to the level of the levator mechanism (Fig. 1). There is usually a dramatic size discrepancy between a normal transverse and descending colon and the very dilated megarectosigmoid. Colon size guides laxative dosing, and it seems that the more localized the rectosigmoid dilation, the better the results of a sigmoid resection in reducing or eliminating the need for laxatives.

Rectal and colonic manometry may help in the evaluation of these patients; however, techniques that are more objective are required. Manometry is performed by placing balloons at different levels of the colon and recording contraction waves [12] or electrical activity [13]. Scintigraphy, a nuclear medicine study, is also being used to assess colonic motility [14]. These are sophisticated studies that do not yet help guide therapeutic decisions. The key information the surgeon needs to know is whether and where a colonic resection would provide benefit to the patient who requires enormous doses of laxatives to empty. Histologic studies of the colon in these patients mainly show hypertrophic smooth muscle and normal ganglion cells in the area of the dilated colon, but more sophisticated histopathologic investigations will hopefully soon yield more valuable results. Further investigations in this area will enhance our knowledge about colonic dysmotility in these patients and thereby guide therapy.

Treatment

Patients with anorectal malformations with potential for bowel control and severe constipation as well as patients with severe idiopathic constipation in whom dietary measures or gentle laxatives do not work require a more aggressive regimen. Drugs designed to increase colon motility are best, as opposed to medications that are only stool softeners. Softening the stool without improving colonic motility will likely make the patient worse, because with soft stool, patients no longer have control, whereas they do reasonably well with solid stool that allows them to feel rectal distension.

In many cases, the laxative regimen uses the same medications that have been tried previously, but the protocol is different in that the dosage is adapted to the patient's response. Response is monitored daily with an abdominal radiograph, and the laxative dose is adjusted if necessary. Almost always, the patient had previously received a lower dose than they need. Severe constipation is treated as follows:

Disimpaction

The disimpaction process is a vital and often neglected step. The routine includes administration of enemas three times a day until the patient is disimpacted. This is confirmed radiologically. If the patient remains impacted after 3 days, then he or she is given a balanced electrolyte solution via nasogastric tube in the hospital, and the enema regimen is continued. If this is unsuccessful, manual disimpaction under anesthesia may be necessary. It is important to remember not to prescribe laxatives to a fecally impacted patient. To do so may provoke vomiting and crampy abdominal pain. In addition, the patient will become reluctant to take laxatives because they are afraid of those symptoms.

Determining Laxative Requirement in a Disimpacted Patient

Once the patient has been disimpacted, an arbitrary amount of laxative is started, usually a senna derivative. The initial amount is based on information the parent gives about previous response to laxatives and the subjective evaluation of the megasigmoid on the contrast enema. The empiric dose is given, and the patient is observed for the next 24 h. If the patient does not have a bowel movement in the 24 h after receiving the laxative, it means the laxative dose was not enough and must be increased. An enema is also required to remove the stool produced during the previous 24 h. Stool in these extremely constipated patients should never remain in the rectosigmoid for more than 24 h.

The routine of increasing the amount of laxatives and giving an enema, if needed, is continued every night until the child has a voluntary bowel movement and completely empties the colon. The day that the patient has a bowel movement (which is usually with diarrhea), a radiograph should confirm that the bowel movement was effective, meaning that the patient completely emptied the rectosigmoid. If the patient passed stool but did not empty completely, the laxative dose must be increased.

As this condition covers a wide spectrum, patients may have laxative requirements much larger than the manufacturer's recommendation. Occasionally, in the process of increasing the amount of laxatives, patients throw up before reaching any positive effect. In these patients, a different medication can be tried. Some patients vomit all types of laxatives and are unable to reach the amount of laxative that produces a bowel movement that empties the colon. Such a patient is considered intractable and therefore a candidate for surgical intervention. Most of the time, however, the dosage that the patient needs in order to empty the colon completely, as demonstrated radiologically, can be achieved. At that dose, the patient should stop soiling because they are successfully emptying their colon each day, and because the colon is empty, they remain clean until the next voluntary bowel movement.

At this point, the patient and the parents have the opportunity to evaluate the quality of life attained with the treatment, understanding that this treatment will most likely be for life. For many of these patients, a sigmoid or rectosigmoid resection can provide symptomatic improvement leading to significant reduction in or complete elimination of laxatives.

Rectosigmoid Resection

For the last 14 years, we have been performing a sigmoid resection to treat select patients with severe constipation [15, 16]. The very dilated megarectosigmoid is resected, and the descending colon is anastomosed to the rectum. In a recent review of patients with anorectal malformations, 315 suffered from severe constipation, were fecally continent, but required significant laxative doses to empty their colon. Of these, 53 underwent a sigmoid resection. The degree of improvement varied. Following sigmoid resection, 10% of patients no longer required laxatives, had daily bowel movements, and no longer soiled; 30% decreased their laxative requirement by 80%; and the remaining 60% decreased their laxative requirement by 40%. These

patients must be followed closely because the condition is not cured by the operation. The remaining rectum is most likely abnormal, and without careful observation and treatment of constipation, the colon can redilate.

A possible alternative could be to resect the rectosigmoid, including the rectum, down to the pectinate line in a similar manner used for patients with Hirschsprung's disease and anastomose the nondilated colon (which is assumed to have normal motility) to the rectum above the pectinate line. This is particularly applicable to the patient with idiopathic constipation who has normal sphincters and a normal anal canal. It should not be performed in patients with anorectal malformations, because this treatment eliminates the rectal reservoir, which may impact continence in some patients.

The most dilated part of the colon is resected because it is the most seriously affected. The nondilated 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. Perhaps with the emergence of new colonic motility techniques, these studies will help with surgical planning. It does seem that patients who improve the most are those who have a more localized form of megarectosigmoid. Patients with 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 can be better predicted by noninvasive modalities.

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