

Update on Paediatric Faecal Incontinence

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Key words

- faecal incontinence
- anorectal malformations
- Hirschsprung's disease
- bowel management

Abstract



Purpose: Faecal incontinence represents a devastating problem; it is often a barrier to social acceptance. It can affect many children including those with prior surgery (for anorectal malformations and Hirschsprung's disease) as well as those with spinal problems or injuries. Management involves distinguishing between true and pseudoincontinence, and then determining the proper protocol of treatment.

Methods: An extensive review of the authors' series of over 500 patients who presented with soiling was undertaken with the goal of determining helpful algorithms of treatment.

Results: Treatment begins first with proper categorisation of patients. Pseudoincontinence (encopresis) can be treated with disimpaction followed by laxative therapy. True incontinence requires an enema programme, with treatment tailored to either hypo or hypermotile colons. Surgery for pseudoincontinence, rarely required, takes the form of colonic resection but only for patients with a demonstrated ability to have voluntary bowel movements, albeit with enormous laxative requirements. Removal of the rectosigmoid in this situation can reduce or eliminate the

need for laxatives. Surgery for true faecal incontinence involves changing the route for a successfully demonstrated enema programme to an antegrade, i.e., a Malone appendicostomy.

Conclusion: The keys to success in helping a faecally incontinent child are dedication and sensitivity on the part of the medical team. The basis of the bowel management programme is to clean the colon (either with medical treatment for patients with the potential for bowel control, or artificially with enemas for patients with true faecal incontinence), and then keep the colon quiet for 24 hours until the next treatment, thereby ensuring that the patient is clean and no longer soiling. The programme is an ongoing process of trial and error that responds to the individual patient and differs for each child. We carry out this programme over the course of one week with daily abdominal radiographs as we tailor the regimen. More than 95% of the children who follow this programme are clean and dry. The clinician must embrace the philosophy that it is unacceptable to send a child with faecal incontinence to school in diapers when their classmates are already toilet trained. Proper treatment to prevent this is perhaps more important than any surgical procedure.

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Introduction



Faecal incontinence represents a devastating problem; it is often a barrier to social acceptance and can have 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 faecal incontinence must be distinguished from overflow pseudoincontinence. Paediatric patients with true faecal incontinence include

some surgical patients with anorectal malformations and Hirschsprung's disease, as well as patients with spinal problems, either congenital or acquired. Those with pseudoincontinence are patients who are potentially capable of bowel control but suffer from overflow (encopresis) related to severe constipation. Much has been learned from the management of surgical patients with severe constipation that can be applied to the patient with idiopathic constipation.

Most patients who undergo repair of an anorectal malformation (ARM) suffer from a functional defecation disorder, and all suffer from an abnor-

mality in their faecal continence mechanism. The mechanism in approximately 25% of patients is so deficient that these patients are faecally incontinent and cannot have a voluntary bowel movement; the others are capable of voluntary bowel movements but may require treatment of an underlying motility disorder, which usually manifests as constipation but sometimes as loose stools [19]. A small yet significant number of patients with Hirschsprung's disease (<5%) suffer from faecal incontinence [1, 13, 25]. Patients with spinal problems or injuries can lack the capacity for voluntary bowel movements, or have this ability but to varying degrees [8, 26].

Patients with true faecal incontinence require an artificial (mechanical) way of keeping themselves clean, a regimen termed bowel management, essentially a tailored enema programme [21]. Medical treatments with laxatives do not work in such patients and can make the patient worse. Patients with overflow pseudoincontinence require proper treatment of their constipation. Understanding this vital distinction is the key to deciding on the correct management.

Continence Mechanism

Faecal continence depends on three main factors, voluntary sphincter muscles, anal canal sensation, and colonic motility [19].

Voluntary muscle structures

In the normal patient, the voluntary muscle structures are represented by the levators, the muscle complex, and the external sphincter. Normally, they are used only for brief periods, when stool in the rectum reaches the anorectal area, pushed by the involuntary peristaltic contraction of the rectosigmoid. A voluntary contraction that holds the stool in occurs only in the minutes prior to defecation, and these muscles are used only occasionally during the rest of the day and night. These muscles also voluntarily relax at the appropriate time to allow the stool to exit the rectum.

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. The voluntary muscles work only when the patient has the necessary sensation indicating when they need to be used. To experience 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

The anal canal is extremely sensitive. However, 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 it is not meticulously preserved at the time of their colonic pull-through. Patients suffering from spinal problems or perineal trauma may have an injured, destroyed, or dysfunctional anal canal.

It seems that patients can perceive distension of the rectum but this requires a rectum that has been properly located within the muscle structures, an important surgical point for patients undergoing pull-through procedures for imperforate anus. This

sensation seems to be a consequence of voluntary muscle stretching (proprioception). The most important clinical implication of this is that liquid or soft faecal material 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 stools. Usually their normal sphincter muscles and anal canal allow them to overcome this problem.

Bowel motility

Perhaps the most important factor in faecal continence is bowel motility; however, its impact has been largely underestimated. In a normal individual, the rectosigmoid remains quiet for variable periods of time (one to several days). During that time, sensation and voluntary muscle structures are almost unnecessary 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. Voluntarily, the normal individual can relax the striated muscles which allow the rectal contents to migrate down into the highly sensitive area of the anal canal. There, accurate information is provided by the anal canal concerning the consistency and quality of the stool. 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 emptying of the rectosigmoid is an involuntary peristaltic contraction helped sometimes by a Valsalva maneuver. Most patients with anorectal malformation suffer from a disturbance of this 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 over-efficient bowel reservoir (megarectum). The main clinical manifestation of this is constipation, which seems to be more severe in patients with lower defects [17]. It is unclear why patients with severe constipation (encopresis) have this problem as well, hence the term idiopathic. Clearly though, 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 and ultimately soiling. The enormously dilated rectosigmoid (● Fig. 1), with normal ganglion cells, behaves like a myopathic type of hypomotile colon [19].

Those patients with anorectal malformation treated with techniques in which the most distal part of the bowel was resected behave clinically like individuals without a rectal reservoir (● Fig. 2). This situation is 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 a constipating diet and medications to slow down colonic motility (such as loperamide and pectin) are indicated. Resection of the distal aganglionic colon is precisely the operation patients with Hirschsprung's disease have undergone, 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. The regular contraction of their rectosigmoid can be translated into a successful voluntary bowel movement.



Fig. 1 Contrast enema of megarectosigmoid. (Reprinted from [19] with permission from Elsevier.)



Fig. 2 Contrast enema in a patient who has had their rectosigmoid resected. Note the absence of the sigmoid loop and the haustrations in the pelvis. (Reprinted from Levitt M, Peña A. Treatment of Chronic Constipation and Resection of the Inert Rectum. In *Anorectal Malformations in Children*. Holschneider AM, Hudson JM, eds. With permission from Springer Media.)

True Faecal Incontinence

For patients with true faecal incontinence, the ideal approach is a bowel management programme which involves teaching the patient and his/her parents how to mechanically clean the colon once daily so as to stay completely clean for twenty-four hours. This is achieved by keeping the colon quiet in between enemas. These patients cannot have voluntary bowel movements and require an artificial mechanism to empty their colon, a daily enema. The programme, although simple, is adjusted through trial and error over a period of one week. The patient is seen every day, and an abdominal radiograph of the abdomen is taken, allowing the patient to be monitored with regard to the amount and location of any stool left in the colon, as well as the presence of stool in the underwear. This daily monitoring with X-rays is an essential key to success as it allows the clinician to make small adjustments to the regimen. The decision as to whether the type and/or quality of the enemas should be modified as well as changes in diet and/or medication can be made daily [21].

Which paediatric patients have true faecal incontinence?

In children with anorectal malformations (ARM), 75% who have had a correct and successful operation have voluntary bowel movements after the age of three [17]. About half of these patients soil their underwear on occasion. These episodes of soiling are usually related to constipation, and when the constipation is treated properly, the soiling frequently disappears. Thus, approximately 40% of all ARM children have voluntary bowel movements and no soiling, and behave like normal children. Children with good bowel control still may suffer from temporary episodes of faecal incontinence, especially when they experience diarrhoea.

Some 25% of all ARM children suffer from true faecal incontinence, and they are the patients who need bowel management (an enema programme) to keep them clean. Some patients with Hirschsprung's disease and patients with spinal problems can suffer from true faecal incontinence. For them, similar principles

Table 1 Prognostic signs for patients with anorectal malformations.

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

of bowel management learned from treatment of patients with anorectal malformations [21] can be applied.

When treating children with anorectal malformations, the surgeon should be able to predict in advance which children have a good functional prognosis and which have a poor prognosis. **Table 1** shows the most common indicators of good or poor prognoses. After the main repair and after closure of the colostomy it is possible to establish the functional prognosis (**Table 2**). It is very important to evaluate this, sometimes even during the newborn period, in order to avoid creating false expectations for parents when the children arrive at the age of toilet training.

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

If the child's defect is of the type associated with a poor prognosis – for example, a high cloaca with a common channel longer than three centimetres, or a recto-bladder neck fistula, or if they have a very hypo-developed sacrum, or an associated spinal problem such as myelomeningocele, the parents must understand that their child will most likely need a bowel management programme to remain clean. This should be implemented when the

Table 2 Predictors of prognosis in patients with anorectal malformations.

| Indicators of good prognosis for bowel control | Indicators of poor prognosis for bowel control |
|--|---|
| Normal sacrum | abnormal sacrum |
| Prominent midline groove (good muscles) | flat perineum (poor muscles) |
| Certain types of anorectal malformations | certain types of anorectal malformations |
| ▶ rectal atresia | ▶ recto-bladder neck fistula |
| ▶ rectoperineal fistula | ▶ about 50% of patients with rectoprostatic fistula |
| ▶ imperforate anus without a fistula | ▶ cloacas with a common channel > 3 cm |
| ▶ cloacas with a common channel < 3 cm | ▶ complex malformations |

child is three to four years of age, before starting school. Children with rectoprostatic fistulas have an almost 50–50 chance of having either voluntary bowel movements or of being incontinent [17]. In these children, an attempt should be made to achieve toilet training by the age of three. If this proves to be unsuccessful, bowel management with an enema programme should be implemented. Each summer, after school is finished, new attempts can be made to assess the child's ability to become toilet trained. In patients previously operated on for an imperforate anus who suffer from faecal incontinence, a reoperation to relocate a misplaced rectum within the sphincter mechanism, with the hope of obtaining good bowel control should be considered if the child was born with a good sacrum, a good sphincter mechanism and a malformation with a good functional prognosis [9]. Approximately 50% of the children operated on under these very specific circumstances show a significant improvement in bowel control [9].

Patients with faecal incontinence and a tendency toward constipation cannot be treated with laxatives but instead need bowel management. In fact, laxatives in such patients make their soiling worse. Many children are unfortunately miscategorised in this way. Such children are usually those born with a defect with a bad prognosis and severe associated anomalies (abnormal sacrum, poor muscles).

Children operated on for imperforate anus who suffer from faecal incontinence can be divided into two well differentiated groups requiring individualised treatment plans. The first and larger group includes patients with faecal incontinence and a tendency towards constipation. The second group have a tendency towards loose stools. Patients with faecal incontinence after operations for Hirschsprung's disease and those with spinal disorders usually fall into the first group, i.e., those with a tendency towards constipation. However, some Hirschsprung's patients have multiple loose stools, and need treatment for hypermotility [14].

Children with true faecal incontinence and constipation (colonic hypomotility)

In these children the motility of the colon is significantly reduced. For them, the basis of a bowel management programme consists of teaching the parents to clean the child's colon once a day with an enema. No special diet or medications are necessary. The fact that they suffer from constipation (hypomotility) is helpful, as it helps them to remain clean between enemas. The real challenge is to find the ideal enema capable of completely cleaning the colon. Definitive evidence that the colon is truly empty following an enema requires confirmation by a plain abdominal radiograph. Soiling episodes or "accidents" occur when there is incomplete cleaning of the bowel with faeces that progressively accumulate. Sometimes the enema itself irritates the

Table 3 Constipating diet.

| Foods to avoid | Foods to encourage |
|-----------------------|--|
| Milk or milk products | apple sauce |
| Fats | apple without Skin |
| Fried foods | rice |
| Fruits | white bread |
| Vegetables | bagels |
| Spices | boiled, broiled, baked meat, chicken or fish |
| Fruit juices | soft drinks |
| French fries | bananas |
| Chocolate | pasta |
| | pretzels |
| | tea |
| | potatoes |
| | jelly |

colon (phosphate enemas can do this) and the patient passes stool between enemas although the radiograph is clean. A gentler enema can solve this problem.

Children with true faecal incontinence and loose stools

The great majority of children with anorectal malformations who suffer from a fast moving colon underwent repair prior to 1980 and the introduction of the posterior sagittal technique. At the time, procedures frequently included resection of the rectosigmoid [6,22]. Therefore, this group of children have an overactive colon because they lack a rectal reservoir. Rapid transit of stool results in frequent episodes of diarrhoea. This means that even when an enema cleans their colon rather easily, stool keeps passing fairly quickly from the caecum to the descending colon and the anus. To prevent this, a constipating diet and/or medications to slow down the colon (loperamide and pectin) are necessary. Eliminating foods that further loosen bowel movements will help the colon to slow down (● **Table 3**). A small subset of patients with Hirschsprung's disease behaves as though they had hypermotility and can be managed similarly.

The key to the success of every bowel management programme is dedication and sensitivity on the part of the medical team. The focus of the programme is on cleaning the colon and keeping it quiet, thus keeping the patient clean for 24 hours after an enema. The programme is an ongoing process for each individual patient and differs for each child. It is usually successful within a week, during which time the family, patient, physician, and nurse learn to tailor the regimen through a process of trial and error. More than 95% of the children who follow this programme 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 faecal incontinence to school in diapers when their classmates are already toilet

trained. Proper treatment to prevent this is perhaps more important than any surgical procedure.

Implementation of the Programme

After a thorough history and exam, the next step is to perform a contrast enema with hydrosoluble material. The study should never be done with barium. It is also important to obtain a picture after evacuation of the contrast material. This study will show the type of colon, i.e., dilated/constipated (● Fig. 1) or non-dilated/tendency towards loose stools (● Fig. 2), helps to empty the colon, and helps the clinician select the type and volume of enema. We have not found manometry helpful for evaluation or planning treatments.

The bowel management programme is then implemented according to the patient's type of colon, and the results are evaluated every day. Changes in the volume and content of the enema are made until the colon is successfully cleaned. For this, an X-ray of the abdomen, taken every day, is vital to determine whether the colon is empty.

Different types of solutions can be used for enemas: some are ready-made and are available from pharmacies and some are prepared at home based on water and salt (a 0.9% saline solution can be made up by adding 2 teaspoons [10 cc] of salt to 960 ml of water). The use of a phosphate enema is convenient since it already comes in a prepared vial. However, saline enemas are often just as effective and some families find them easier and less expensive. Saline can be combined with phosphate or glycerine to make a more effective enema. Occasionally, children will complain of cramping with the phosphate enema but have no complaints with saline. Children older than 8 years of age or weighing more than 30 kg may receive one adult phosphate enema daily (133 ml). Children between the ages of 3 and 8 years or between 15 and 30 kg may receive one paediatric phosphate enema (70 ml) each day. Children should never receive more than one phosphate enema per day because of the risk of phosphate intoxication, and in other children with impaired renal function these enemas should be used with caution. Sometimes phosphate can cause colitis.

The enema, administered on a daily basis, should result in a bowel movement, within 30–45 minutes, followed by a period of 24 hours 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, and phosphate or glycerine can be added to the saline solution. The "right" enema is the one that will empty the child's colon and allow them to stay clean for the following 24 hours. This can be achieved only by trial and error and through learning from previous attempts.

Children with loose stools have an overactive colon and usually do not have a rectal reservoir. This means that even when an enema cleans their colon easily, new stool passes quickly from the caecum to the descending colon and the anus. To prevent this, a constipating diet, bulking agents, and/or medications (such as loperamide and pectin) to slow down the colon are used. Eliminating foods that loosen bowel movements will help the colon move more slowly.

Parents are provided with a list of constipating foods to give and a list of laxative foods to avoid. The diet is strict: bananas, apples, toasted bread, white pasta with no sauce, boiled meat, etc. Fried foods and dairy products must be avoided (● Table 3). Parents

learn which meals provoke loose stools and which will constipate their child. To determine the right combination, the treatment starts with enemas, a very strict diet, loperamide and pectin. Most children respond to this aggressive management within several days. The child should remain on a strict diet until he/she is clean for 24 hours several days in a row. They can then choose one new food every two to three days, observing the effect on their colonic activity. If the child soils after eating a newly introduced food, that food must be eliminated from the diet. Over several months, the most liberal diet possible is sought. If the child remains clean with a liberal diet, the dose of the medication can gradually be reduced to the lowest effective dose to keep the child clean for 24 hours.

In children in whom a successful bowel management programme has been implemented, the parents frequently ask if this programme will be needed for life. The answer is "yes" for those patients born with no potential for bowel control. However, since there is a wide spectrum of defects, there are patients with some degree of bowel control. These patients undergo a bowel management programme so as not to be exposed to embarrassing accidents of uncontrolled bowel movements. However, as time goes by, the child will become more cooperative and more interested in the problem. It is conceivable that later in life, the child may be able to stop using enemas and remain clean, following a specific regimen of disciplined diet with regular meals (three meals per day and no snacks), often with laxatives added, to provoke bowel movements at a predictable time. Every summer, children with some potential for continence may try to find out how well they can control their bowel movements without the help of enemas. This is done during vacations to avoid accidents at school, during a time that they can stay home and try some of the toilet training strategies. We often have such patients return for another one week programme, a "laxative trial," again with daily radiographs and tailoring now of a laxative regimen with the goal of eliminating the enema.

Pull-through vs. permanent stoma

For patients with a colostomy and no potential for bowel control a key question is whether to perform a pull-through or leave a permanent stoma. We are of the opinion that if a patient has the capacity to form solid stools, a pull-through can be performed and a daily enema given to keep them clean. For these patients successful bowel management offers a better quality of life than a permanent stoma. In these patients we first try bowel management through the stoma (● Fig. 3). This is particularly relevant for patients with cloacal exstrophy, although there are many children who are told that because of their poor prognosis for continence, they should remain with a colostomy [12]. If their stoma remains quiet (usually with a constipated diet, pectin, and loperamide) in between enemas, then that stoma could be closed or pulled through with a daily antegrade enema continuing to keep the patient clean.

Antegrade enema

Most preschool and school-age children enjoy a good quality of life while undergoing the bowel management programme. However, when they reach puberty, many 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 [10,11,15], whereby the appendix is connected to the



Fig. 3 Picture showing bowel management administered through the colostomy.

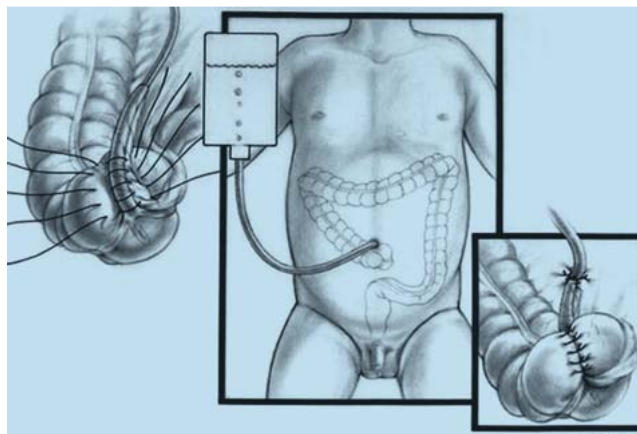


Fig. 4 Malone appendicostomy. (Reprinted from [11] with permission from Elsevier.)

umbilicus, allowing antegrade administration of the enema (● Fig. 4). A valve mechanism is created that allows catheterisation of the appendix for the enema fluid but avoids leakage of stool through it. If the child no longer has an appendix, it is possible to create a new one from a colonic flap (a continent neoappendicostomy) (● Fig. 5).

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

Pseudoincontinence (encopresis)

It is vital to differentiate real faecal incontinence from overflow pseudoincontinence. Pseudoincontinence occurs when a patient behaves as though he or she is faecally incontinent but in reality suffers from severe constipation and overflow soiling (encopresis). Once the disimpaction is treated and the patient receives sufficient laxatives to avoid constipation, they become continent.

The colon absorbs water from the stool and has a reservoir function. These processes depend on colonic motility, which is an area of physiology that is not well understood, and for which treatments are limited. In normal individuals the rectosigmoid 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.

If a child is faecally continent, management involves the treatment of constipation, using laxatives, which help provoke peristalsis to overcome the dysmotility disorder. Patients who have undergone successful surgery for Hirschsprung's disease and for anorectal malformations (with an anorectal defect with a good prognosis) and have a normal spine should be faecally continent. Patients with idiopathic constipation (encopresis) also have an intact continence mechanism.

Severe constipation is common with anorectal malformations, particularly in the more benign types [20]. It is also common in patients following successful surgery for Hirschsprung's disease, and of course occurs in a large group of patients considered to have idiopathic constipation.



Fig. 5 Neoappendicostomy. (Reprinted from [10] with permission from Elsevier.)

Constipation, when left untreated, can be extremely incapacitating, and in its most serious forms can produce a form of faecal incontinence known as overflow pseudoincontinence (encopresis). Diet impacts colonic motility; but the therapeutic value of diet is negligible for the most serious forms of constipation. It is true that many patients with severe constipation can suffer from a psychological disorder, but a psychological origin cannot explain the severe forms, as it is not easy to voluntarily retain stool when an otherwise autonomous rectosigmoid has normal peristalsis. The passage of large, hard pieces of stool may be painful and make the patient try to retain the stool. This may complicate the problem of constipation; but it is not the original cause.

The clinician must decide which type of patient they are dealing with. Patients with a good prognosis for bowel control (those with benign anorectal malformations are the ones more likely to have constipation) and patients with severe idiopathic constipation need an aggressive, proactive treatment of their constipation. The child must be deemed capable of being faecally continent, and have the capacity for voluntary bowel movements before initiating treatment for constipation, otherwise laxatives will just make them soil more.

Most of these patients suffer from different degrees of dilation of the rectum and sigmoid, a condition termed megarectosigmoid (● Fig. 1), due to a hypomotility disorder that interferes with complete emptying of the rectosigmoid [19]. This can occur in

children born with an anorectal defect with a good prognosis, who underwent a technically correct operation but did not receive appropriate treatment for constipation and therefore developed faecal impaction and overflow pseudoincontinence. This can also happen in children with severe idiopathic constipation (encopresis) who have a very dilated rectosigmoid.

Treatment of Severe Idiopathic Constipation with Pseudoincontinence (Encopresis)

First the 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. Laxatives started before disimpaction cause cramping. The dosage of the laxative is increased daily until the right amount of laxative is reached that completely empties the colon every day.

If medical treatment proves to be extremely difficult because the child has a severe megasigmoid requiring an enormous amount of laxatives to empty it, the surgeon can propose segmental resection of the colon [10,18] (● Fig. 6a and b). After the rectosigmoidal resection, the amount of laxatives required to treat these children will 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 faecal incontinence with constipation. Failure to make this distinction may lead to an operation in which a faecally incontinent constipated child becomes a child with a tendency to loose stools, which will make them much more difficult to manage.

When children with anorectal malformations, Hirschsprung's disease and a tendency to constipation are managed from early on in life with aggressive treatment of constipation, children with good prognosis should toilet train without difficulty. When constipation is not managed properly and a patient presents after many years, they behave very much like children with idiopathic constipation and have overflow pseudoincontinence (encopresis).

Constipation is a self-perpetuating disease. A patient suffering from a certain degree of constipation that is not treated adequately will only partially empty the colon, leaving larger and larger amounts of stool inside the rectosigmoid, which results in a greater degree of megasigmoid. It is clear that dilation of a hollow viscus produces poor peristalsis, which explains the fact that constipation leads to faecal retention, followed by megacolon, which exacerbates the constipation. In addition, the passage of large, hard pieces of stool may produce anal fissures leading to reluctance on the part of the patient to have bowel movements. The dysmotility of the colon that occurs in certain patients with Hirschsprung's disease, even after successful surgery to remove the aganglionic bowel, is not understood. These patients do benefit from a proactive medical treatment of their constipation. Likewise, the explanation for severe constipation and encopresis remains idiopathic. Proactive, aggressive laxative therapy helps these patients as well.

The clinician must accept the fact that the dysmotility observed is essentially incurable. It is, however, manageable, but requires careful follow-up for life. Treatment cannot be given on a temporary basis, once tapered or interrupted, constipation recurs. Some clinicians treat such "intractable" patients with colostomies or colonic washouts via a catheterisable stoma or button de-

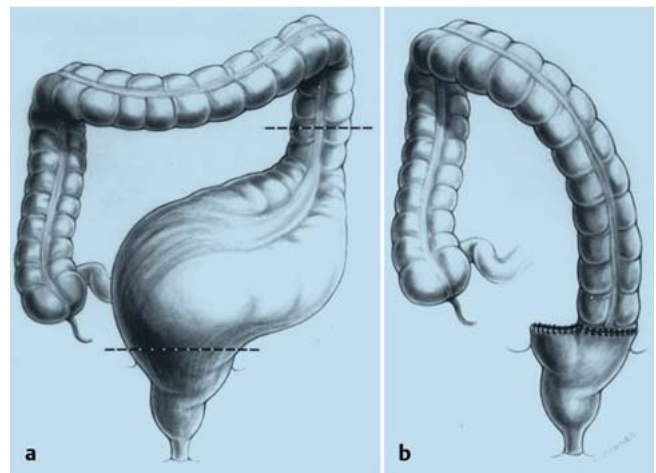


Fig. 6a and b Diagram of a sigmoid resection. (Reprinted from [18] with permission from Elsevier.)

vice [2,5,7,16,24]. Once the distal colon regains a normal calibre, the physician assumes that the patient is cured and may be tempted to close the colostomy or discontinue the washouts. Unfortunately these patients' symptoms quickly recur. We believe that washouts are really only for patients with true faecal incontinence who are incapable of having voluntary bowel movements and thus require a daily enema to empty. The patients with pseudoincontinence (encopresis) are capable of emptying their colon with the help of adequate doses of laxatives and thus do not need washouts at all.

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

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

Soiling of the underwear is an ominous sign for bad constipation. A patient who at an age of bowel control soils his/her underwear day and night and basically does not have spontaneous bowel movements may have "overflow pseudoincontinence." These patients behave similar to faecally incontinent individuals. When the constipation is adequately treated, the great majority of these pseudoincontinent children regain bowel control.

A contrast enema with a hydrosoluble material (never barium) is the most valuable study which, in the constipated patient, usually shows a megarectosigmoid with dilatation of the colon 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. This is obviously the exact opposite pattern of Hirschsprung's disease. The size of the colon must serve as a guide for dosing of the laxatives, and it appears that the more localised the dilation of the rectosigmoid, the better the results of a rectosigmoid resection in reducing or eliminating the need for laxatives.

Rectal and colonic manometry are often used in the evaluation of these patients; however we are of the opinion that more objective techniques are required. Manometry is performed by placing balloons at different levels of the colon and recording the waves of contraction [4] or the electrical activity [23]. Scintigraphy, a nuclear medicine study, is also used to assess colonic motility [3]. 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 benefit a patient who requires enormous doses of laxatives to empty. Manometry does not consistently answer this question. Histological studies of the colon in these patients mainly show hypertrophic smooth muscle in the area of the dilated colon and normal ganglion cells, but more sophisticated histopathological investigations will hopefully soon yield more valuable results. These patients do not have Hirschsprung's disease and the concept of "very low segment Hirschsprung's" or "internal sphincter achalasia" is confusing. Further investigations in this area will expand our knowledge of colonic dysmotility in these patients, and thereby guide therapy. For now, we treat these patients empirically, based on the contrast study indicating dysmotility in the dilated segment of the colon.

Patients with anorectal malformations with the potential for bowel control and severe constipation as well as those with severe idiopathic constipation in whom dietary measures or gentle laxatives do not work require a more aggressive regimen. Drugs that are designed to increase the motility of the colon are best (senna-based laxatives are ideal) as opposed to medications that are only stool softeners. Softening of the stool without improving colonic motility will likely make the patient worse, because they no longer have control with soft stool and will soil more, whereas they do reasonably well with solid stools that allow them to feel the distension of the rectum. Essentially, stool softeners soften the stool but in cases of severe hypermotility do not help it come out.

The laxative regimen we describe uses the same medications that may have been tried previously, but the protocol is different, in that disimpaction is done first, the dosage is then adapted to the patient's response, and the response is monitored daily with an abdominal radiograph with the laxative dose adjusted if necessary. Almost always, the patient had previously received a lower dose of laxatives than they ultimately needed.

Protocol for Treatment of Severe Constipation in Patients with Pseudoincontinence



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 three days, they are then given a balanced electrolyte solution via a nasogastric tube in the hospital, and the enema regimen is continued. If this is unsuccessful, a manual disimpaction under anaesthesia may be necessary.

Determination of the laxative requirement in a now disimpacted patient

Once the patient has been disimpacted, a dose of laxative is started, usually a senna derivative. The initial amount is based on the information that the parents give on the patient's previous response to laxatives, and subjective evaluation of the

megasigmoid on the contrast enema. An empirical dose is given and the patient is observed for the next 24 hours. If the patient does not have a bowel movement within 24 hours after receiving the laxative, this means the laxative dose was not enough and must be increased. An enema is also required in order to remove the stool produced during the previous 24 hours. Stool in these extremely constipated patients should never remain in the rectosigmoid for more than 24 hours. If they stool multiple times and the radiograph is clean, the laxative dose can be decreased. This routine of choosing the amount of laxatives and giving an enema, if needed, is continued every night, until the child has a voluntary bowel movement, and empties the colon completely. On the day the patient has a bowel movement, a radiograph should confirm that the bowel movement was effective, meaning that the patient has completely emptied the rectosigmoid. If the patient passed stool but did not empty completely, the dose of laxative must be increased. Sometimes the laxative effectively empties the colon but the stool is too loose. Adding pectin provides a little bulk in this situation and makes the same laxative dose more effective.

Since 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 an adequate effect is obtained or do not tolerate such a high dose because it causes nausea and cramping. Some patients vomit all kinds of laxatives and are unable to reach the amount of laxative that would produce a bowel movement that empties the colon. Such patients are considered to have failed medical management and are therefore candidates 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. 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 that they have with this treatment, and must understand that it will most likely be for life. For some of these patients a rectosigmoid resection can provide symptomatic improvement leading to a significant reduction or complete elimination of their laxative requirement.

Rectosigmoid resection

A sigmoid resection (◉ Fig. 6) for the treatment of selected patients with a repaired anorectal malformation and severe constipation can be considered [10,18]. The very dilated megarectosigmoid is resected and the descending colon is anastomosed to the rectum (◉ Fig. 6). The rectum is left intact in view of its contribution to bowel control, as such patients need the reservoir and depend on proprioception of rectal distension as a cue. 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.

An alternative is to resect the rectosigmoid including the rectum, down to the pectinate line in a similar manner used for patients with Hirschsprung's disease, and to anastomose the non-dilated colon (that is assumed to have normal motility) to the rectum above the pectinate line [10] (◉ Fig. 7). This is ideal for a patient with idiopathic constipation who has normal sphincters and a normal anal canal and is a great alternative to traditional recom-



Fig. 7 Transanal resection of a megarectosigmoid. (Reprinted from [10] with permission from Elsevier.)

mendations such as colostomy or antegrade stomas [2,5,7,10,16,24].

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. We infer this from the contrast enema but, clearly, there must be a more scientific way to assess the dysmotile anatomy. Perhaps with emerging colonic motility techniques, these studies will help with surgical planning. It does seem that the patients who improve most are those that have a more localised form of megarectosigmoid. Patients with a more generalised 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 the results of resection better predicted by noninvasive modalities.

Conclusion

The key to success in helping a faecally incontinent child is dedication and sensitivity on the part of the medical team. The basis of the bowel management programme is to clean the colon (either using medical treatment for patients with the potential for bowel control, or artificially with enemas for patients with true faecal incontinence) and then keep the colon quiet for 24 hours until the next treatment, thereby keeping the patient clean and eliminating soiling. The programme is an ongoing process of trial and error that differs for each individual child. We carry out this programme over the course of one week and perform a daily abdominal radiograph as we tailor the regimen. More than 95% of the children who follow this programme are clean and dry. One should embrace the philosophy that it is unacceptable to send a child with faecal incontinence to school in diapers when their classmates are already toilet trained. A beautiful anatomic reconstruction in a patient who functionally continues to soil does not accomplish much. Proper treatment to prevent this is perhaps more important than any surgical procedure.

Conflict of Interest: None

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