Recommendations for Anal Dilatations Following Repair of Imperforate Anus Repair

In children born with an imperforate anus, the anal canal, which would normally allow wastes to pass out of the body, fails to develop. When a young child has surgery to repair an imperforate anus, surgeons first determine the limits of the sphincter, a ring-like band of muscles that opens and closes the anus, and then accommodate the rectum and anus within the limits of the sphincter.

That means that sometimes for a little baby, the anus following surgery is smaller than normal for the child's age. In fact, in patients with good sphincters, the anus appears completely closed and it is not possible to see rectal mucosa, the lining of the rectum that produces mucus. The mucosa stays inside because the good muscle tone of the sphincter prevents the mucosa from coming out. If the patient is left in this condition, the anus would heal closed. Anal dilations are needed to gradually stretch the new anus without destroying the sphincter, until the anus reaches the size that is normal for the patient's age.

At the Colorectal Center at Cincinnati Children's Hospital Medical Center, we prefer to dilate the anus twice a day because every dilatation creates small lacerations or tears and we want to keep the rectum open before those lacerations heal. We are opposed to doing dilatations once a week (under anesthesia to avoid pain) because that method lets the laceration heal for an entire week only to create a new laceration during the next dilatation. This leads to severe fibrosis (scarring) and a narrowing of the anus that becomes impossible to dilate.

May Cause Discomfort, But Should Not Be Very Painful

Anal dilatations after a well done operation, in general should not be extremely painful. When the protocol for dilatation is followed properly, patients may experience uncomfortable dilatations but not real pain.

Severe pain can occur when the surgical procedure leaves that patient without adequate blood supply to the rectum, which therefore has a tendency to narrow. Pain can also occur when the protocol for dilatation is not followed properly and severe scarring develops in the anus, which is then subjected to further attempts at dilatation. If the anal dilatation stays at the same size for more than a week in one size, the anus will heal at that size and the large amount of scar tissue around the anus will make it impossible to dilate it further.

Recommended Dilatation Protocol

The protocol used at the Colorectal Center is based is based on experience and the size of the anal opening in children with normally developing anuses. Parents or other caregivers are encouraged to do the dilatation procedure themselves at home, but if they have difficulty, the procedure can be done at the hospital.

Dilatation is done with a set of Hegar dilators of increasing size.

- A normal newborn will take a #12 Hegar dilator with minimal discomfort
- A child 4 to 12 months old with take a #13 Hegar dilator

- A child 8 to 12 months old will take a #14 dilator
- A child 1 to 3 years old will take a #15 Hegar dilator
- A child 3 to 12 years old will take a #16 Hegar dilator
- A child more than 12 years old will take a #17 Hegar dilator

Major Improvements Over Previous Procedures

An operation to repair an imperforate anus should be done as early as possible since very little babies seem to tolerate dilatations better. The current surgical and dilatation procedures represent significant improvements over methods formerly used to repair an imperforate anus.

In years past, surgeons tended to create very large anal openings. The main goals of those operations were to avoid subjecting children to anal dilatations and possibly strictures, narrowing of the opening from improper dilatations. While these goals may have been worthy, those operations did not respect the limits of the sphincter mechanism. The newer surgical procedures and dilatation protocol do respect the limits of the sphincter mechanism, with better overall results.

Colorectal Center for Children
http://www.cincinnatichildrens.org/
Phone (800)344-2462 (513)636-3240
FAX (513)636 3248

DILATATION SCHEDULE
POST IMPERFORATE ANUS REPAIR FOR CHILDREN

PRIOR TO ARRIVAL IN CINCINNATI:

Order a set of Hagar Dilators from: **Jones Surgical Company, Inc.**
101-21 Metropolitan Avenue
Forest Hills, NY
(718) 261-9500.

Bring these dilators with you. Please pack them in your checked luggage since you will not be allowed to have them in carry-on luggage.

TWO WEEKS AFTER SURGERY

The doctor or nurse clinician will remove sutures, calibrate the anus and will teach you to dilate the anus. We will determine the caliber of the first dilator. EVERY DAY you will dilate the new anus TWICE A DAY.

Insert the dilator for 30 seconds twice in the morning and twice at night, always before meals. The baby must be held by another person, with the knees against the chest.

EVERY WEEK THE SIZE OF THE DILATOR SHOULD BE CHANGED TO THE NEXT SIZE. Dilations must continue TWICE A DAY UNTIL YOU REACH THE DESIRED SIZE. Once the desired size is reached (see below), the colostomy can be closed. However, the dilator must still be passed after the colostomy closure until the dilator passes easily without pain (usually 3-4 weeks after the last size dilator was reached).

Child's Age:	Hagar Dilator for Desired Size to reach for age at the time dilatations is started.
☐ 1-4 Months	#12
☐ 4-8 Months	#13
☐ 8-12 Months	#14
☐ 1-3 Years	#15
☐ 3-12 Years	#16
☐ More than 12 years	#17

Once you find that the dilator passes easily without pain, (twice a day), you MAY START TAPERING the frequency of the dilatations as follows.

- **Once a day for a month.**
- **Every other day for a month.**
- **Every third day for a month.**
- **Twice a week for a month.**
- **Once a week for a month.**
- **Once a month for three months.**

If the dilatation becomes difficult, painful, or bloody, at any time during the process of tapering the frequency of dilatations, dilate twice a day again and restart the process.

Usually the dilatations become painful during the passing of the last 2-3 sizes. At that particular time, the parents sometimes decide not to dilate everyday, in order to avoid pain. That is wrong! Dilatations must continue twice a day. Dilating the anus only once a week may produce a laceration which may heal and be reopened during dilatation the following week. This often can cause a severe scar that provokes a narrowing of the anus that will require an additional operation.

By staying in a single dilator size for a prolonged period of time (more than one week), the healing process may limit the anus to a smaller caliber than is desired. It will then be extremely difficult to dilate again with a larger dilator.

(Rev 4/9/07- G/colorectal/-forms/handouts for families-educational)

Purchase Instructions:

The quickest and most cost effective way to buy Calmoseptine is from a pharmacy. Talk ONLY to the Pharmacist and refer to information below. Calmoseptine is an over-the-counter product and usually takes 1 working day to order. Take this card with you and save it for future use.

If your pharmacy is not listed, chances are that they order from one of the 3 Wholesalers listed in this chart.
Questions call 1-800-800-3405

Pharmacy Item Numbers by Wholesaler Suggested Retail for 4 oz Tube: $7.50			
Amer/Bergen# 518449	Cardinal# 2144715	McKesson# 2143550	
Drug Emporium	CVS	Albertsons	
Longs	Eckerd Drug	Brooks	
Meijer	Fred Meyer	Costco Wholesale	
Pathmark	Giant Eagle	4 oz price about $5.55	
Publix	H-E-B	Hy-Vee	
Raley's	Hi-School	Rite Aid	
	K Mart	Safeway	
==========	King Soopers	Sav-On	
Walgreens	Kroger	ShopKo Stores	
	Medicine Shoppe	Target	
	Price Choppers	Vons	
	Schnucks	Wal-mart	
	Winn Dixie		(As of 9/2005)

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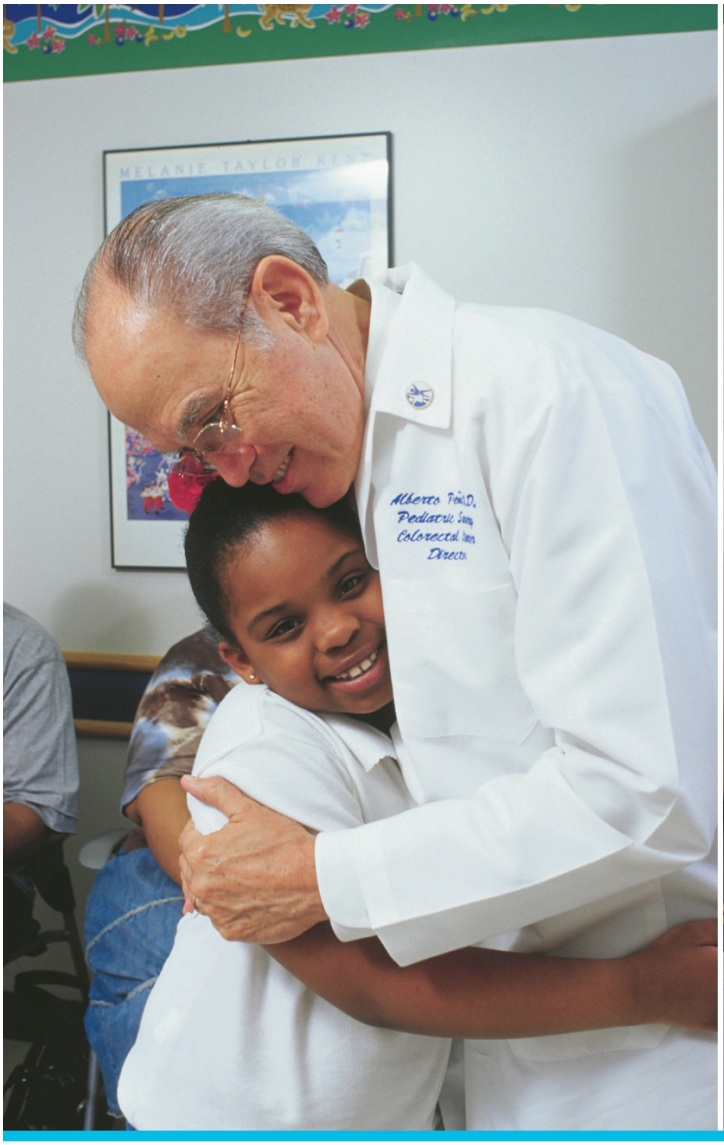
We also supply over 20 other wholesalers in the industry. Please ask the Pharmacist to check with the wholesaler if other than the 3 listed.



www.cincinnatichildrens.org/colorectal

Colorectal Center for Children

Cincinnati Children's Hospital Medical Center

The Bowel Management Program

Cincinnati Children's
change the outcome®

Contents

About the Center

The Colorectal Center for Children at Cincinnati Children's Hospital Medical Center was founded by Alberto Penã, MD, and Marc A. Levitt, MD. It is the first and only pediatric Colorectal Center in the world and thus provides care to children from all over the world with a unique, comprehensive and multidisciplinary approach.

The Colorectal Center improves the lives of children born with:

- Anorectal malformations / imperforate anus
- Hirschprung's disease
- Inflammatory bowel disease
- Colonic dysmotility such as idiopathic constipation
- A variety of other colorectal disorders

The Colorectal Center evaluates and treats children by pairing corrective surgery (when necessary) with proven techniques for bowel management through a combination of routine and controlled enemas, medication and diet.

The goal of the Colorectal Center is to improve the quality of life for all children with colorectal problems. A team of health care professionals from across the medical center helps families learn to regulate and control their child's bowels. Successful bowel management means achieving predictable bowel emptying and a clean child without incontinence. It offers the chance for those with a serious disorder to live with a new sense of freedom and independence.

To request an appointment or to contact the Colorectal Center for Children at Cincinnati Children's:

Phone: 513-636-3240
Email: colorectalcenter@cchmc.org

You may also visit the Colorectal Center at **www.cincinnatichildrens.org/colorectal**

An Overview of the Bowel Management Program

Fecal continence, or control of the bowels, is an important achievement in a child's development. For children born with anomalies of the large bowel (anorectal anomalies and Hirschsprung's disease) or anomalies of the innervation of the pelvic organs (such as spina bifida), the process of toilet training is challenging and often requires medical intervention.

The goal of the bowel management program is to teach patients and their parents or guardians how to administer a daily enema in order to clean and quiet the child's colon to prevent accidents. This is an important step in establishing independence and freedom. Achieving bowel control makes going to school and participating in activities outside the home possible for children with even the most severe physical anomalies.

For a patient with true fecal incontinence, bowel control is possible through the use of daily enemas tailored specifically for each patient. Children begin the bowel management program at Cincinnati Children's by visiting the Colorectal Center for approximately one week. Each day the child is given an enema in a controlled environment. After passing stool, the child's abdomen is X-rayed in order to monitor the amount and location of any stool left in the colon.

Each afternoon, the clinical team meets to discuss and review the X-ray. Through trial and error, the medical staff can determine the correct amount and type of enema suitable for each patient and determine whether dietary restrictions or medications are needed to help the child control his or her bowels.

Before he was adopted, Deanna and Mike Anderson's son Shailesh underwent multiple surgeries in India that left him with a compromised bowel and fecal incontinence. The family spent three weeks in Cincinnati at the Colorectal Center where Shailesh received a Malone procedure allowing him to self-administer enemas through an opening in his belly button.

Mrs. Anderson describes the result as "Nothing short of a miracle. Nobody knows he has any special needs when they see him now. He can run, play and socialize all because of the care he received at Cincinnati Children's."

Communication is Key

At every visit, parents and guardians are given the opportunity to discuss any questions or concerns they have regarding the enema, diet, soiling issues and psychological aspects of the bowel management program. Nurses provide instruction to families and reply to questions so that they understand how to administer an effective enema.

A New Beginning

Today, there are treatment options and procedures available to children who suffer from fecal incontinence. An accurate evaluation along with an appropriate follow-up program can have a life-changing effect on a child. Treatment of incontinence is an on-going process. The goal is to help every child socially and physically adapt so that he or she can enjoy the same freedoms enjoyed by other children.

Bowel management is a treatment program that was originally conceived for children born with imperforate anus; however it can be utilized with all children who have fecal incontinence.

Understanding Fecal Incontinence

The medical definition of fecal incontinence is: The incapacity to voluntarily hold feces in the rectum. If you have a child who is unable to control his or her bowels then he or she may be afflicted with one of the following conditions:

- Real fecal incontinence
- Pseudoincontinence

A child with fecal incontinence must be evaluated so that the medical team can distinguish between real incontinence and pseudoincontinence. It may seem confusing, but in order to plan the best treatment the clinicians must understand the origin of the problem.

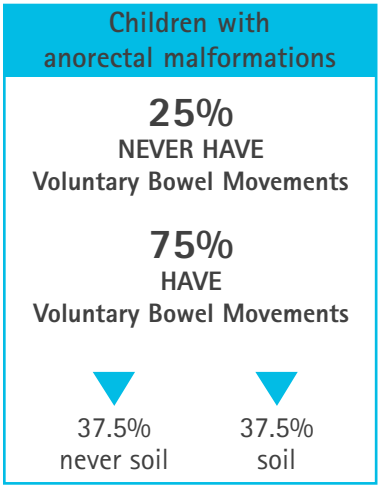
Real Fecal Incontinence

For a child with real fecal incontinence, the normal mechanism of bowel control is not working. An alteration of the muscles that surround the anorectal canal along with poor sphincters (those muscles which control the anus) are responsible for fecal incontinence in children operated on for anorectal malformations with a bad prognosis. Some patients operated on for Hirschsprung's disease have this anatomic problem as do those with spinal problems.

The innervation (supply of nerve connections) of these muscles is important for their correct function. A deficit of the innervation occurs in anorectal anomalies as well as in other conditions. For example, in cases of spina bifida, or following spinal cord injury, the contraction and relaxation of the muscles, as well as sensation, are both deficient. Thus, the presence and the passage of stool and the perception of the difference between solid and liquid stool and gas are limited.

Pseudoincontinence

In cases of pseudoincontinence, a child is believed to suffer from fecal incontinence. However, investigation shows that he or she suffers from severe constipation and fecal impaction. When the impaction is treated and the patient receives enough laxatives to pass stool, he or she becomes continent.



Physical Causes of Fecal Incontinence

Children born with anorectal malformations require surgery to correct their condition. Research demonstrates that 75 percent of all children with anorectal malformations who have undergone a correct and successful operation will have voluntary bowel movements after the age of 3. About half of these patients soil their underwear on occasion. Those episodes of soiling are usually related to constipation. When the constipation is treated properly, the soiling frequently disappears. Thus, approximately 40 percent of all children have voluntary bowel movements and no soiling. In other words, they behave like typical children.

But there is a long way to go and a lot to do to improve the quality of life for a significant number of children. Children with good bowel control still may suffer from temporary episodes of fecal incontinence. Some 25 percent of these children suffer from real fecal incontinence and they must undergo bowel management to stay clean.

It is important to reiterate that despite a good operation to correct a child's anorectal defect, there are many children who do not achieve bowel control or suffer from different degrees of fecal incontinence.

Reasons for Fecal Incontinence

There are three important physical elements to unaided bowel control. Children born with anorectal malformation often do not experience the following:

1. Sensation within the Rectum

Children born with anorectal malformations don't experience the sensation of stool or gas passing through their rectum. Therefore, many times the child may unknowingly soil. When this happens, it is typical for them to become accustomed to the smell of stool, which can upset the entire family and anyone around the incontinent child.

2. Motility of the Colon

The rectosigmoid is that portion of the bowel that acts as a natural reservoir for the feces. It is physiologically important to accumulate and "store" the feces between every bowel movement.

Normally, the rectosigmoid remains quiet for periods of 24 to 48 hours (time necessary to accumulate the feces); then a peristaltic wave (produced by bowel wall muscle contraction) allows the complete emptying of the rectosigmoid after which it remains quiet. If the rectosigmoid is slow the stool stays stagnant and constipation occurs. As a result, the child may suffer from overflow incontinence and will soil. On the other hand, if a child has no rectosigmoid (no reservoir) he or she passes stool constantly, which is called colonic hypermotility.

3. Muscles

Muscles or voluntary sphincters normally surround the rectum and anus and are considered fundamental for continence. Children with anorectal malformations lack some level of development of these muscles and therefore are incapable of holding in stool. Children with spinal problems have varying degrees of sphincter deficiency.

A surgical technique introduced by Dr. Alberto Peña in 1980 called a posterior sagittal anorectoplasty (PSARP or Pull-Through Procedure) greatly improved the outcome for children with anorectal malformation to achieve bowel control. The PSARP positions the rectum within the limits of the voluntary sphincters responsible for continence.

Unfortunately, the sphincter (a circular muscle) is frequently not normal in a child with anorectal malformation. The more complex the defect, the less developed the muscle. The surgeon is able to predict in advance which children may have good functional prognosis and which children may have poor prognosis. For many children, after the main repair and after the colostomy closure, it is possible to establish the functional prognosis.

The table below shows the most common indicators of good and poor prognosis for bowel control.

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 - Cloaca with a common channel <3 cm - Less complex malformations: perineal fistula	• Abnormal sacrum • Flat perineum (poor muscles) • Some types of anorectal malformations: - Rectal atresia - Rectobladderneck fistula - Cloacas with a common channel >3 cm - Complex malformations

The table shows prognosis signs for bowel control.

good prognosis signs	poor prognosis signs
• Good bowel movement patterns: 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

Success Rates for Bowel Management

At the Colorectal Center we understand the importance of providing families with a realistic picture of their child's chances for bowel control before any form of treatment begins. Although each child's condition will vary, the table below provides a reasonable picture regarding levels of success for children with anorectal malformations.

Type of Defect	Sex	Voluntary bowel mov.	Soiling	Voluntary bowel mov. never soiling	Constipation
Perineal fistula	F/M	100%	0%	100%	26%
Anal atresia or stenosis	F/M	100%	16%	84%	80%
Vestibular fistula	F	94%	38%	71%	64%
Bulbar fistula	M	88%	65%	32%	59%
ARM without fistula	F/M	85%	41%	71%	47%
Cloaca C. Ch. <3 cm.*	F	83%	78%	27%	32%
Prostatic fistula	M	76%	78%	28%	50%
Real vaginal fistula	F	75%	100%	0%	25%
Cloaca C. Ch. >3 cm.*	F	59%	89%	22%	53%
Bladder-neck fistula	M	28%	100%	0%	29%

C. Ch. = common channel

What Do the Numbers Mean for Your Child?

If your child's defect is of the type associated with good prognosis – such as a vestibular fistula, perineal fistula, rectal atresia, rectourethral bulbar fistula, or imperforate anus with no fistula – there is a strong likelihood that he or she will have voluntary bowel movements by age 3. However he or she will still need supervision to avoid fecal impaction, constipation and soiling.

If, instead, your 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 or a recto-bladder-neck fistula – then he or she will most likely need the bowel management program to remain clean and socially acceptable. This should begin when the child is 3-4 years old.

For children with rectoprostatic fistulas, there is a 50 percent chance that they will have voluntary bowel movements. We recommend that families try toilet training with their child by the age of 3. If this proves to be unsuccessful, bowel management should be implemented so that the child can remain clean and avoid social discomfort. Summer time can be the ideal time for a child to attempt toilet training as families can stay close to home without the additional pressure of school and peers.

Urinary Incontinence

Sometimes children with malformations experience urinary incontinence. Urinary incontinence occurs in male children with anorectal malformations only when they have an extremely defective or absent sacrum (a large, triangular bone at the base of the spine and at the upper and back part of the pelvic cavity) or if important nerves were effected during the operation. Thus, the overwhelming majority of male children with anorectal malformations who are properly treated have urinary control.

This is also true for female children with all anorectal malformations except patients with cloaca (a single opening to release urine and feces). A significant number of children who have undergone repair of a cloaca require intermittent catheterization in order to empty the bladder. This happens in nearly 70 percent of children with a high cloaca (defined as a common channel longer than 3 cm) and 20 percent of children with a low cloaca (defined as a common channel shorter than 3 cm).

The bladder neck in most girls with cloaca is competent and therefore they remain completely dry when treated with intermittent catheterization. If catheterization is not performed, then urinary overflow incontinence will occur (they will leak urine) once the bladder is full. Urinary tract infections are a common result.

Treatment Options

The impressive success rates for treating fecal incontinence can help inspire families struggling with the daily realities of their child's condition. In 95 percent of cases, the bowel management program is successful.

Some children, previously operated on for an imperforate anus who suffer from fecal incontinence, may be candidates for a second operation if the child was born with a good sacrum, good sphincter mechanism and a malformation with good functional prognosis and there is evidence that the rectum is mislocated; the length of the colon is intact and they have the capacity to form solid stool.

A PSARP can be performed and the rectum can be relocated within the limits of the sphincter mechanism. Approximately 50 percent of the children operated on under these circumstances have a significant improvement in bowel control and do not require bowel management following surgery.

Treatment for Pseudoincontinence

Children with pseudoincontinence suffer from different degrees of dilatation of the rectum and sigmoid, a condition defined as megarectosigmoid. These children suffer from a hypomotility disorder that interferes with complete emptying of the rectosigmoid. A child born with a good prognosis type of defect who underwent a technically correct operation but did not receive appropriate treatment for constipation, he or she can develop fecal impaction and overflow pseudoincontinence.

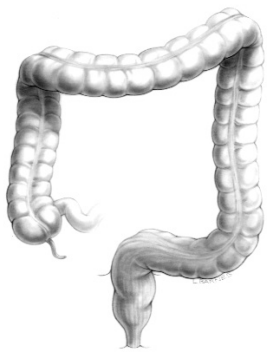
In this case, enemas and colonic irrigations (an aggressive program of enemas) are used to clean the megarectosigmoid and remove the impaction. Then the constipation is treated through administration of laxatives. The dosage of the laxative is increased daily until the proper amount is reached in order to completely empty the colon every day.

If a child cannot control his or her bowels once the constipation is treated adequately, then the diagnosis changes to real incontinence and constipation, and he or she needs bowel management with a daily enema.

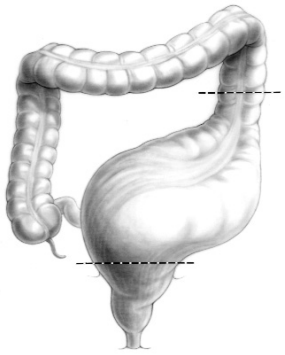
If after the constipation is treated adequately it becomes evident that the child is actually continent, then it is clear that the patient suffers from overflow pseudoincontinence. If medical treatment proves to be extremely difficult because the child has a severe megasigmoid and requires an enormous amount of laxatives, a surgical alternative, consisting of a resection (partial removal) of the sigmoid colon, is an option.

At the Colorectal Center, before a sigmoid resection is performed, work is done to confirm that the child is suffering from overflow pseudoincontinence rather than real fecal incontinence with constipation. Failure to make this distinction may lead to an unnecessary operation; a fecally incontinent constipated child subjected to this operation would be changed to one with looser stool which is much more difficult to manage.

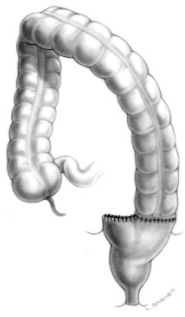
After the sigmoid resection, the amount of laxatives required to treat these children can be significantly reduced or even eliminated.



normal colon



megarectosigmoid colon

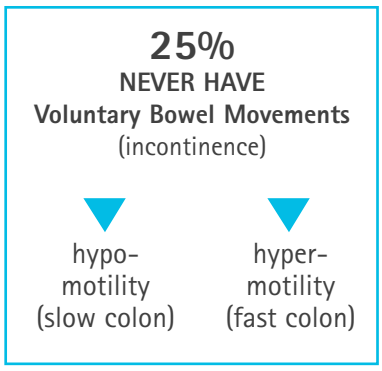


resected colon

Children Who Are Candidates for Bowel Management

Children who suffer from fecal incontinence after the repair of an imperforate anus are usually those born with a bad prognosis type of defect and severe associated defects (defect of the sacrum, poor muscle complex). However, children who were born with a poor prognosis type of defect can still achieve a good quality of life when treated with the bowel management program.

Children operated on for imperforate anus and who suffer from fecal incontinence can be divided into two well-defined groups that require individualized treatment plans:



Children with Constipation (Colonic Hypomotility)

The basis of the bowel management program is to teach parents to clean their child's colon once a day with an enema or a colonic irrigation. No special diet or medications are necessary for children with colonic hypomotility or constipation. Their tendency towards constipation helps them to remain clean between enemas. The real challenge is to find an enema capable of cleaning the colon completely. Soiling episodes or "accidents" occur when there is an incomplete cleaning of the bowel.

Children With Loose Stools and Diarrhea (Colonic Hypermotility)

The great majority of children who suffer from loose stool and diarrhea following surgery were operated on before 1980, before the introduction of the PSARP technique. Previous techniques frequently included resection of the rectosigmoid reservoir. Therefore, this group of children has an overactive colon. Rapid transit of stool results in frequent episodes of diarrhea. This means that even when an enema cleans the colon rather easily, stool keeps on passing fairly quickly from the cecum to the descending colon and the 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 to slow down.

If your child experiences hypermotility, our medical team provides your family with a list of constipating types of foods to be given and a list of laxative foods to be avoided. The diet is rigid and includes food such as banana, apple, baked bread, white pasta with no sauce, boiled meat, etc. Fried foods and dairy products are avoided.

Use to Constipate Your Child

Food groups	Food recommended	Food to avoid
Milk and milk products	Rice milk	All others
Vegetables	None	All others
Fruits	Applesauce, apples without skin, bananas	All others
Starches, bread and grain	Bread, crackers and cereals made from refined flours, pasta and noodles made from white flours, white rice, pretzels, white potatoes without skin, dry cereals such as: Rice Krispies, Rice or Corn Chex, Corn Flakes, Kixx.	All others
Meat or meat substitutes	Baked, broiled, boiled or grilled meat, poultry or fish	All others
Fats and oils	Limit amounts of butter, margarine and oils in food preparation during this phase, non-stick spray is allowed	All others
Sweets and desserts	Made from allowed ingredients, plain cake, gelatin or popsicles, Rice Dream Frozen Dessert and limited amounts of concentrated sweets such as jelly and marshmallows	All others

Some additional tips when beginning to add foods into your child's diet

- Avoid extremely hot or cold foods. Foods served warm or at room temperature may be better tolerated.
- All vegetables should be well cooked
- Avoid raw fruits and vegetables
- Avoid foods that may produce gas or cause cramps, carbonated drinks, chewing gum, beans, cabbage, highly spiced foods, and swallowing air while talking and eating, or using a straw.

More Nutrition Information: For more diet and nutrition information, go to www.cincinnatichildrens.org/colorectal, and look under the Bowel Management Program.

Through experience most parents know which meals provoke diarrhea and which constipate their child. To determine the right combination, the treatment starts with enemas, a very strict diet and loperamide (Imodium®). Most children respond to aggressive management within a few days. The child should remain on a strict diet until clean for 24 hours for two to three days in a row.

Once clean the child can choose one new food every two to three days, observing the effect on his or her colonic activity. If the child soils after eating a newly introduced food, parents are advised to eliminate that food from the diet on a permanent basis. The most liberal diet possible should be sought. If the child remains clean with a liberal diet, then the dose of the medication can gradually be reduced to the lowest dose effective.

Putting the Bowel Management Program into Practice

Almost 60 percent of children operated on at birth for an anorectal malformation will suffer from functional bowel problems of different types.

The main goal of the bowel management program is to improve the quality of life for children with anorectal malformation before and after surgery. There are a range of options for families with a fecally incompetent child to consider.

Children without bowel control can:

- Remain incontinent and use diapers permanently.
- Try the bowel management program.
- Have a permanent colostomy.
- It is the Colorectal Center's belief that successful bowel management provides the best quality of life of these options.

The bowel management program is most successful when families and health professionals work together. At Cincinnati Children's, every member of the Colorectal Center understands that sensitivity, authentic interest and dedication are critical to achieving a successful outcome. There must be communication between caregivers and family members about what's working and what's not going well in order to achieve a routine that works for everyone.

An Individualized Program

The bowel management program is tailored to individual patients and differs from child to child. A routine is usually achieved within a week while the family, patient, physician and nurse undergo a process of trial and error designing the program to the specific patient. This requires a great deal of dedication but the results are significant.

More than 95 percent of the children who follow the bowel management regimen remain artificially clean and dry for the entire day and can have a completely normal life.

How to Prepare the Enema

The type and amount of daily enema required to control the bowel varies from patient to patient. The formula that works best for your child is determined during their initial week-long evaluation at the Colorectal Center.

Equipment needed:

1 Enema bag (kangaroo feeding tube bag)

2 The determined ingredients for the enema (pre-packaged phosphate enema, saline, mild soap, glycerin, castile soap, etc.)

3 Foley catheter (22 or 24 French) with a 30 ml balloon

4 Water-soluble lubricant

5 30 ml syringe (slip tip)

There are different types of solutions to use for enemas. There are prepared solutions that can be bought in a drugstore or solutions that can be prepared at home based on water and salt.

The use of phosphate enemas (Fleet®) is convenient since it comes pre-prepared. However, pure saline enemas are often just as effective and some families find it easier and less expensive. Occasionally, children will complain of cramping with the Fleet® enema but have no complaints with saline. Phosphate enemas can be added to saline enemas (check the list of ingredients on prepared enemas to ensure it contains phosphate). Glycerin and castile soap are also ingredients that can be added to the enema solution. If you are unsure of what to use, consult your child's medical team at the Colorectal Center. Phosphate enemas are usually avoided in patients with associated renal (kidney) problems.

Children should never receive more than one phosphate enema a day because of the risk of phosphate intoxication. Children with impaired renal function should use Fleet® enemas with caution.

This recipe for normal saline solution should *not* be altered:

1000 ml tap water with 1 1/2 teaspoon table salt
OR
500 ml tap water with 3/4 teaspoon table salt

Since the bowel management program is an individualized, ongoing process, there is not a precise formula that fits all children. The "right" amount of the saline enema is the one that can empty the child's colon and allow him or her to stay clean for the following 24 hours. This can be achieved only by trial and error.

Remember:

- Generally, a larger volume of saline solution is necessary in those children who have hypomotility as compared to those who have hypermotility and tend to empty their colon more rapidly.
- The solution in the enema may be warmed to body temperature to reduce cramping.
- It's important to examine the consistency and the amount of stool obtained by the enema to determine if it was effective. Parents can learn when to repeat an enema with saline.

Administered on a regular basis, the enema should result in a bowel movement followed by a period of 24 hours of complete cleanliness. If the colon, as demonstrated by X-ray, isn't empty following the enema or, if the child keeps soiling, then the child requires a more aggressive treatment. If the saline enema produces inadequate results, then other additives such as Fleet®, liquid glycerin or castile soap can be added to increase the potency of the enema.

Administering the Enema

1 Gather all equipment.

2 Prepare the prescribed enema and additives.

3 Position the child (see page 15 on position options).

4 Lubricate a 22 or 24 French Foley catheter and gently introduce it into the anus approximately 4-6 inches.

5 Using a 30 ml syringe filled with air or warm water, attach the syringe to the small port and instill 15 ml of the water then pull back on the catheter. If the balloon comes out, push the catheter back and instill another 5 ml of water then pull back again. You may continue this process up to the full 30 ml. Remember: never inflate the balloon right at the anus.

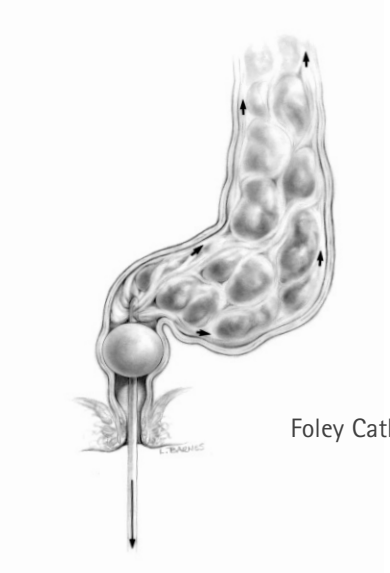
6 As soon as the balloon is inflated, take the syringe off the catheter to prevent the balloon from deflating.

7 While keeping tension on the catheter to avoid leaking, start the flow of enema fluid.

8 For optimal effect, the child must retain the solution for 5-10 minutes once the enema is complete.

9 When retention time is up, place the child on the toilet and then deflate the balloon, allowing the catheter to slip out.

10 The enema is most effective when the child can remain on the toilet for 45 minutes.



Foley Catheter

The Foley catheter is helpful in preventing leakage of the enema solution. The balloon acts as a plug during the enema process.

Tips

The higher the bag is held, the faster the flow; the lower the bag is held, the slower the flow. Giving the enema should take about 5-10 minutes. If there are cramps then slow down the flow by lowering the enema bag to help decrease cramping.

Check the results of the enema by looking into the toilet. If there is no stool or minimal stool, report these observations. Your child may require a different or larger enema.

Frequently Asked Questions About the Bowel Management Program

At what age should my child begin bowel management?

Children who suffer from fecal incontinence are basically oblivious to their condition when they are young and in diapers. Problems begin when their peers begin to wear underwear while they remain in diapers. This is the time when social discrimination may start.

Toilet training for stool is a long-term goal for children with anorectal malformations, although it is not always possible. Parents of children born with a good prognosis type of defect should be encouraged to use the same strategies for toilet training as those followed by families with typical children.

What are Recommended Tips for Toilet Training?

Keep it simple
Between ages 2-3 years old, parents are advised to sit their child on the toilet after every meal. Allowing him or her to play with toys while on the toilet is encouraged.

Keep it light
Just as in training any child, parents are encouraged to toilet train as a game and not as a punishment. Parents should sit with their child and not argue or force the child to remain seated. If the child gets up, the parents should put the toys away.

Keep it fun
The child should be rewarded for a bowel movement or urinating while on the toilet.

Can my child go to school if toilet training isn't working?

If the child is not successfully toilet trained by school age a family has two alternatives:

- Do not send the child to school for another year and continue attempts at toilet training.
- Begin the bowel management program of prescribed enemas to keep the child clean.

We recommend to most families that they begin the bowel management program by the time their child is 3-4 years old. At this age most children are no longer wearing diapers.

It is important to emphasize that the choice as to when and how to start bowel management is something that each family has to decide. The goal is to have the child in school wearing normal underwear without being ridiculed by his or her classmates who are already toilet trained.

What is the best time of day to administer an enema?

The timing of the enema plays a role in how efficiently it cleans the bowel. We recommend that you give an enema after the main meal of the day to take advantage of the gastrocolic reflex (this motion of the colon happens after each meal). Most families give the enema in the evening when there is more time.

Consider what time of day will work best for your family. It's important to give the enema at the same time every day in order to create a routine. Keep in mind that if the enema is given every other day that the child should expel the amount of stool for two days. No more than 48 hours should elapse between enemas.

Why not use a micro-enema?

Administration of a micro-enema to a child that is severely constipated only cleans the very last part of the rectum. This leaves the colon full of stool, now softened by the micro-enema, and the stool will leak more easily. If the goal of bowel control is reached and the child is clean, this regimen is acceptable.

Are there ways to make the enema less unpleasant?

Success of the enema depends on making the procedure as comfortable and efficient as possible. Children should be encouraged to take their time on the toilet. Reading books, watching a favorite television show or playing with toys are all good ways to help make a child feel at ease. Providing a foot stool for the child to rest his or her legs while on the toilet is often helpful.

How can I teach my child to be patient during the enema?

Teaching a child to be patient in order to give the enema time to work requires creativity. Distractions like books and television can help, depending on the age of the child. A special toy given only at toilet time can work as an incentive for staying put. It is also important to involve siblings and family members instead of isolating the child.

What if my child refuses to have an enema?

If your child does not want to have an enema administered, there is no point on insisting he or she comply. It is really important to transform a negative experience into a positive one, reinforcing all the positive aspects such as being clean, wearing regular underwear, no diapers, the use of a bathing suit, and the possibility to change garments in front of other children.

What is the best position for administering an enema?

The child's position is important in making the enema effective. The child should be in a position that facilitates delivery of the fluid as high up into the colon as possible. If the child is small, this can be done by placing him or her on the parent's lap with the head down and the buttocks on the lap. The older child may lie on a bed with his or her buttocks on the bed and a pillow under their abdomen. Another position for the older child or adolescent is in a knee-chest position with the buttocks in the air. Adolescents who are trying to achieve independence by self-administering enemas may also try the knee-chest position while lying on their side. If it's more comfortable, lying on the left or right side to administer the enema can be equally effective

How long until the enema is effective?

The enema fluid should be retained as long as possible and this depends on the child and the quantity of fluid introduced. A wait time of 5-10 minutes is recommended.

After administration of the enema, the child should sit on the toilet for as long as necessary (usually 45 minutes) to allow for complete emptying of the colon.

It is very important to be sure that the child empties his or her colon; otherwise, a more aggressive enema is necessary. Keep in mind that this process is learned by trial and error, which is the only way to obtain the maximum benefit.

What are the long-term effects of daily enemas?

The bowel management program has been in practice for over 20 years. There are no known negative effects that can be attributed to the use of enemas or colonic irrigations. Of course, it is impossible to predict any consequences decades from now.

It is important to remember that there are several kinds of enemas. A Fleet® enema is a phosphate enema and can be toxic when given in excess. If the specific recommendations of the manufacturer are followed, problems of phosphorus intoxication or hypocalcaemia (low calcium) have not been seen.

Many children receive enemas with saline solution which is not different from the liquid that exists throughout our body in terms of the concentration of electrolytes. No secondary effects from the use of these enemas have been seen. Too much salt in the solution, however, can lead to sweating, feelings of nausea, vomiting and hypernatremia (too much sodium). It is very important to follow the recipe on page 12 when making a saline enema.

Do daily enemas interfere with nutrition?

It's common to worry that enemas affect the absorption of nutrients. It is very important for families to remember that when enemas are given, it is the colon that is being washed. Only stool, which is waste, is being removed. The main absorption of nutrients occurs in the small bowel, and enemas do not wash that part of the intestine.

If Bowel Management Stops Working

Sometimes after a period of successful bowel management a child begins to soil again. This is an indication that something has changed and a thorough evaluation in a clinical setting is necessary.

Your pediatrician can order an abdominal X-ray. If the X-ray shows a large amount of stool in the colon after the enema, it means that the enema solution needs to be adjusted (increased volume and/or concentration) to the new needs of the child. It can take a week of enemas and X-rays to make this determination.

If the X-ray shows a clean colon then the "accidents" are due to increased motility and it may be necessary to introduce some medications to slow the colon as described previously, as well as to observe a more strict diet. Your child's medical team can advise you if this is indicated.

As Children Grow, Their Treatment Needs May Change

Any changes in routine play an important role in successful bowel management. No one exists in a controlled environment free from outside stress. Changes in the diet, or certain occasions like birthdays or holidays can have repercussions on the effectiveness of the program, especially for children with hypermotility. Other, more significant changes such as divorce, moving locations, or changing schools all play a role in bowel function.

Families can learn to anticipate stressful situations and, in many cases, it is possible to give a medication such as Imodium® on the day before a known event or an outing to slow down a child's bowel motility.

Independence

As children grow older they often feel their privacy is being infringed on when a parent continues to help them with bowel management. When a child begins to feel this way, and if they have been successful with bowel management, they benefit from a "continent appendicostomy" (also known as a Malone or ACE procedure, see page 19). This operation creates a small orifice in the umbilicus (belly button) through which the patient can pass a small catheter into their colon and administer the enemas themselves while sitting on the toilet. It allows the child to be more independent and can further improve his or her quality of life.

The Future

Parents of children who enter the bowel management program frequently ask if this program will be needed for life. For patients born with a poor prognosis type of defect, the answer is yes, they will continue to need bowel management for life. However, since we are dealing with a spectrum of defects, there are patients with some degree of bowel control. They are subjected to the bowel management program in order to prevent occasional embarrassing accidents of uncontrolled bowel movements. However, as time goes by children become more cooperative and more interested and concerned about their problem, and more likely to achieve success with potty training.

It is conceivable that later in life a child may 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.

If all three of the following conditions are met, the child might be a candidate for discontinuing daily enemas:

1 The child is completely clean with bowel management. This means no accidental soiling and regular, consistent bowel movements following every enema for a period of time.

2 The child is cooperative. The child has to be aware of his or her problem and motivated to experiment with new strategies to solve the problem.

3 The child understands there's a risk it won't work. If a family attempts to regulate bowel movements through diet and/or medication, then there must be a willingness to return to the bowel management program with a daily enema if bowel activity is not predictable and accidents reoccur.

Every summer, the children with some potential for bowel control can try, on an experimental basis, to find out how well they can control their bowel movements without the help of enemas. This is best done during summer vacation or prolonged time at home to avoid accidents at school. We call this process a "laxative trial."

Steps to Success

- Stay home and close to the toilet.
- Continue with a regular diet and routine schedule of meals.
- Have the child sit on the toilet after every meal to try and pass stool.
- Often, laxatives are introduced to help provide daily bowel movement.
- Remain vigilant: the child must remain alert to try and learn to discriminate the feeling of an imminent bowel movement.

For children who are diagnosed as constipated, a daily laxative given at the same time each day is recommended. The dose of the laxative is adjusted by trial and error. It is best to first try the less aggressive and natural types of laxative, and then, depending on the child's response, use medications with more active ingredients. The first choice is a laxative type of diet; the next choice is fiber or a bulk forming type of product. If this does not work, a laxative with an active ingredient is indicated and can be added.

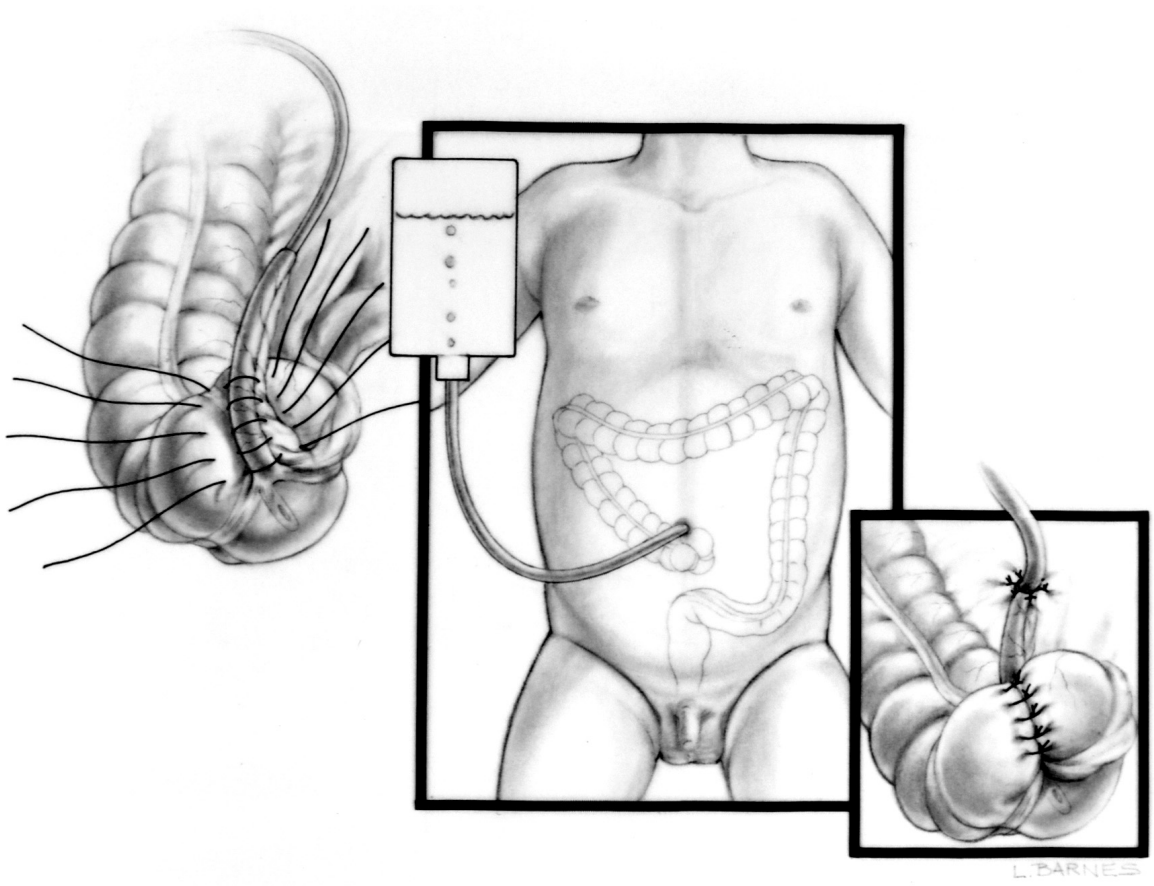
After a few days or weeks, the family and child can decide whether they want to continue with the new regimen or go back to the bowel management program. This decision is up to the family and the child and is based on the quality of life experienced with each method.

Additional Bowel Management Strategies for Children With Anorectal Malformations

The ACE or Malone Procedure

Most preschool and school-age 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 the 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.

It is important to stress that the Malone procedure is just another way to administer an enema. This procedure allows the child to administer an enema by inserting a small catheter into the orifice at the belly button while sitting on the toilet. The enema infusion enters the bowel pushing the stool forward. It is very easy and comfortable for any child to do.



Before implementing the Malone procedure some preliminary conditions need to be met:

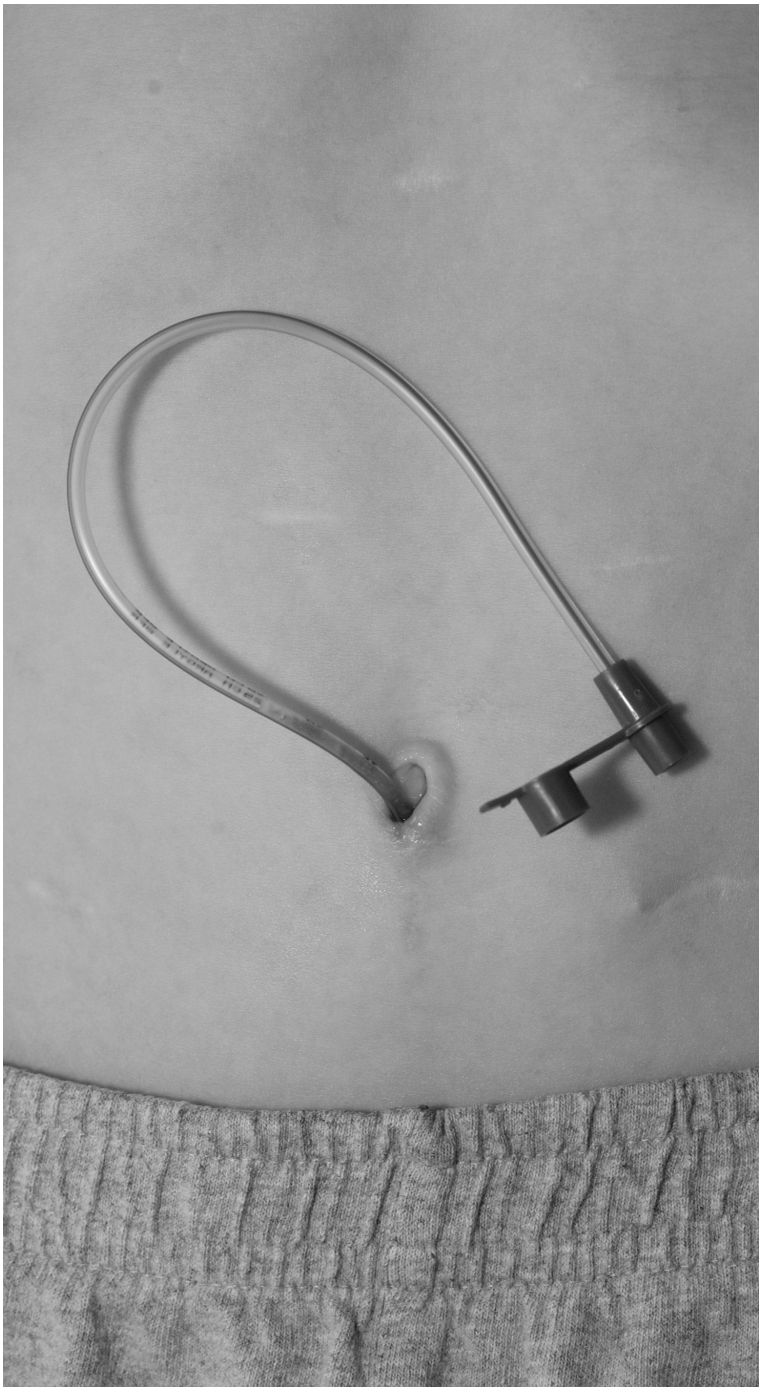
- The child has to be perfectly clean with his or her regular bowel management. It does not make sense to operate on a child who is not successful with the bowel management in order to create another route to administer the enema that will also be ineffective.
- The child has to be absolutely motivated, both in terms of the operation and the administration of the enemas as well as in his or her ability to evaluate the effects.

The operation consists of connecting the appendix, which is attached to the cecum, at the very beginning of the colon, to the abdominal wall (usually at the belly button) and creating a valve mechanism that allows catheterization of the appendix but avoids leakage of stool.

If the child does not have his or her appendix, it is possible to create a new one from the colon. This is called a continent neo-appendicostomy.

Recipe for Success

Bowel management is not as simple as giving an enema or following a prescribed diet or taking a medication. There is no miraculous recipe. However, a methodical combination of enemas, colonic irrigations, diet and medication can help those children with fetal incontience to remain artificially clean.



Remember:

- The success of any of these strategies depends upon collaboration among the family, the child and the health care team.
- There are many variations depending on the child's needs: it takes dedication, determination, consistency and love by everyone involved.
- Children who have completed the bowel management program and remain clean for 24 hours experience a new sense of confidence based on an improved quality of life.
- The health professionals at the Colorectal Center of Cincinnati Children's are dedicated to helping families find the best regimen for you and your child.

www.cincinnatichildrens.org/colorectal

To request an appointment or to contact the Colorectal Center for Children at Cincinnati Children's:

Phone: 513-636-3240
Email: colorectalcenter@cchmc.org

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Appendicostomy (Malone)

by
Marc A. Levitt MD., Richard Falcone Jr. MD., and Alberto Peña, MD

Most patients who undergo repair of an anorectal malformation suffer from a degree of functional defecating disorder, and all suffer from at least some 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. Some patients with Hirschsprung's disease suffer from fecal incontinence as do some with spinal problems such as spina bifida or myelomeningocele, or following spinal trauma.

Fecal incontinence represents a devastating problem which can prevent a person from becoming socially accepted and lead to serious psychological sequelae. These patients require an artificial way to keep them clean and in normal underwear, a regimen termed Bowel Management.

The Bowel Management Program consists of teaching the patient or his/her parents how to empty the colon once daily so as to stay completely clean for 24 hours. This is achieved by keeping the colon quiet in between enemas. The program is implemented by trial and error over a period of one week. The patient is seen each day and an x-ray film of the abdomen is taken to monitor on a daily basis the amount and location of any stool left in the colon as well as the presence of stool in the underwear. The decision as to whether the type and/or quality of the enemas should be modified, as well as changes in the diet and/or medication, can be made.

The keys to success of the 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, and thus the patient clean for the 24 hours after the enema that is given once a day. Sometimes manipulation of diet and medication are utilized for patients with a hypermotile colon. Most patients have a hypomotile or slow colon.

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 family, patient, physician, and nurse undergo a process of trial and error, tailoring the program to the specific patient. More than 95% of the children who follow this program are artificially clean and dry for the whole day and can have a completely normal life. We believe that it is unacceptable to send a child with fecal incontinence to school in diapers when his classmates are already toilet trained, and that proper treatment to prevent this is perhaps more important than any surgical procedure with regard to its impact on the patient's quality of life.

The enema administered on a regular basis should result in a bowel movement 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 may be added to the saline enema. If the addition of the phosphate still results in inadequate results, then glycerin can be added, or an enema with a balloon catheter may help. The "right" saline enema is the one that can empty the child's colon and allow him to stay clean for the following 24 hours. This can be achieved only by trial and error and learning from previous attempts.

In children in whom a successful bowel management program has been implemented, the parents frequently ask if this program will be needed for life. The answer is usually "yes" for those patients born with a poor prognosis for bowel

control. However, since 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/her problem. It is conceivable that later in life, a child may stop using enemas and remain clean, following a specific regimen of a disciplined diet with regular meals (3 meals per day and no snacks) to provoke bowel movements at a predictable time. Every summer, the children with some potential for bowel control can 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 potty training strategies.

If we conclude that the patient needs a daily enema in order to remain clean, then at the appropriate age we discuss with them an operation called an appendicostomy, or Malone procedure.

Most preschool and school-age children enjoy a good quality of life while undergoing the bowel management program. However, when they get older, 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 has been designed.

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

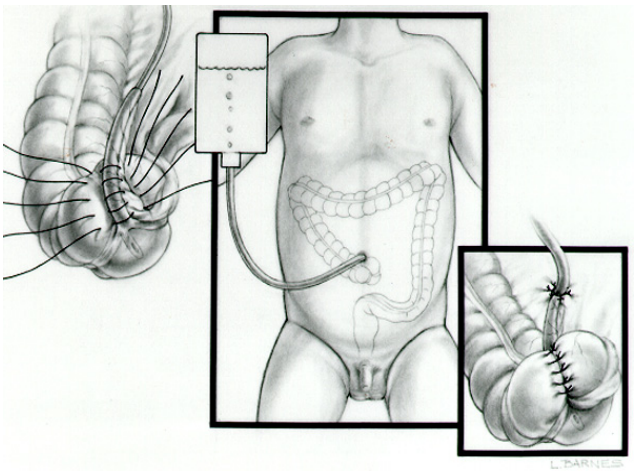
The operation consists of connecting the appendix to the umbilicus, and creating a valve mechanism that allows catheterization of the appendix for the enema fluid, but avoids leakage of stool through it. The operation involves a small incision below the belly button and sometimes can be done with the help of laparoscopy to minimize the incision. It takes approximately 2 hours to complete. We prefer the use of the appendix rather than an artificial device because it is cosmetically more hidden, and it avoids problems with the device caused by irritation of the surrounding skin.

At the conclusion of the operation a tube is left through the appendix coming out of the belly button, and is used for the enemas. At 2-3 weeks following surgery, the tube is removed and the patient and parents are taught how to pass the tube through the umbilicus once a day for the enema administration. Once the tube is out, since the orifice is hidden in the belly button, no one except for the patient, family, and doctor know it is there. The child can participate in all activities including swimming.

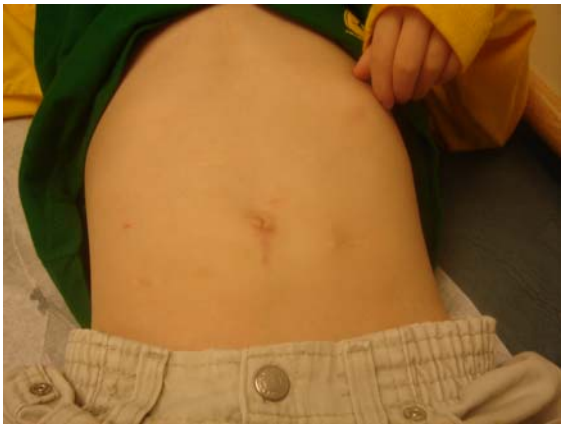
If the child has had his or her appendix removed, it is possible to create a new one from the colon. This is called a continent neo-appendicostomy. The appendix is made from a flap of colon which is fashioned into a tube. In such a case we do not start using the neoappendix for 4 weeks.

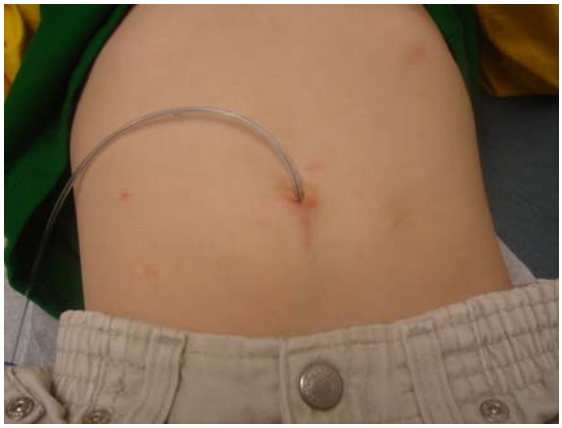
With an appendicostomy the enema remains the same, but the route of administration is changed. Some families note that the enema runs through more efficiently when it is administered through the Malone site, and there is a great deal of satisfaction with this procedure as it gives the child significantly more independence with their bowel management regimen.

Appendicostomy illustration: The appendix is connected to the belly button so a tube can be passed through it for the enema administration.



Appendicostomy procedure: *Top left:* Appendix and cecum *Top right:* Cecum wrapped around appendix to make a valve mechanism *Bottom:* Appendix connected to the belly button





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Bowel Management

Marc A. Levitt, MD and Alberto Peña, MD

Cincinnati Children's Hospital Medical Center

Colorectal Center for Children

Fecal incontinence represents a devastating problem for all who suffer from it. It often prevents one from becoming socially accepted which in turn provokes serious psychological consequences. This is a problem that affects more children than previously thought.

Operations designed to treat fecal incontinence are still controversial and render variable results. That is why at our institution, we have designed a bowel management program specifically for individuals with this problem.

The bowel management program consists of teaching the patient or his/her parents how to clean the colon once daily so as to stay completely clean in the underwear for 24 hours We do this by keeping the colon quiet in between enemas. The program, although simplistic, is implemented by trial and error over a period of one week. The patient is seen each day and an x-ray film of the abdomen is taken so that we may monitor, on a daily basis, the amount and location of any stool left in the colon as well as the presence of stool in the underwear. We then decide whether the type and/or quality of the treatment should be modified as well as diet and/or medication.

As children grow older, they want to be more independent and do not like their privacy infringed on by adults helping them with their bowel management. When a child begins to feel this way and they have been successful with bowel management, we offer them a "continent appendicostomy" (also known as a Malone or ACE procedure). This is a small stoma in the umbilicus (belly button) through which the patient can pass a small catheter into their colon and administer enemas while sitting on the toilet. It allows the child to be more independent and further improves his/her quality of life.

We have treated over 500 patients and have been successful 95 percent of the time. As you can imagine, it is a rewarding experience to witness the dramatic change in the quality of life for the children in the program.

Introduction

Almost 60 percent of children operated on at birth for an anorectal malformation, even when they received a technically correct operation, will suffer from functional bowel problems of different types.

The main goal of a bowel management program is to improve quality of life. The parents and child need to define quality of life for their situation. Therefore, every child must be free to choose from the alternatives offered. For instance, a child that is completely fecally incontinent has the following options:

1. To remain incontinent and use diapers permanently
2. To try the bowel management program
3. To have a permanent colostomy

The key to a successful bowel management program is dedication, timing, sensitivity, and for the medical team to be authentically interested in the patient. If the elements of sensitivity, authentic interest, and dedication are missing, the chance of success is very low.

What is a bowel management program?

The basis is to clean the colon and keep the colon quiet and thus clean for the 24 hours after the enema, colonic irrigation,or suppository, that is given once a day. Sometimes manipulation of diet and medication are utilized.

- Bowel Cleaning (Enemas, Suppositories, Micro-Enemas)
- Modification of the Diet (If necessary)
- Medications Of necessary)

There are some medications that are able to stow down the colon's motility. The use of these specific medications such as Lomotil or lmodium must be decided on with the physician.

The program is an ongoing process that is responsive to the individual patient and differs from child to child. Success is usually achieved within a week during which family, patient, physician and nurse employ trial and error to tailor the program to the specific patient. This requires a lot of dedication. More than 90 percent of the children who follow this program are artificially clean and dry for the whole day and can have a completely normal life. They develop a new sense of self esteem and confidence based on an improved quality of life.

It is unacceptable to send a child with fecal incontinence to school in diapers when his classmates are already toilet trained. Children who require diapers or who have accidents while in school because of fecal incontinence are exposed to ridicule from their peers, which can lead to adverse psychological consequences.

The first step is to perform a contrast enema study. The contrast enema in children with ARM is performed with hydro-soluble material (Hypaque) and without a previous bowel preparation. The study should never be done with barium; it is also important to obtain a picture after the evacuation of the contrast material. This study allows the

physician to learn about the patient's type of colonic motility (hypomotility – constipated or hypermotility diarrhea). The bowel management program 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 enemas are made until the colon is successfully cleaned. For this, one should take an x-ray film of the abdomen every day to see whether the colon is empty.

Enemas

There are different types of solutions to use for enemas. There are some ready-made solutions that can he bought in a drugstore or solutions that can be prepared at home based on water and salt.

The use of phosphate enemas (Fleet) is most convenient since it is already a prepared vial. However, pure saline enemas are often just as effective and some families find it easier and less expensive. Occasionally, children will complain of cramping with a Fleet enema but have no complaints with saline. Children older than 8 or heavier than 65 lbs (30kg) may receive one adult phosphate enema daily. Children between 1 - 8 years old or between 35 - 65 lbs (15 and 30 kg) may receive one pediatric phosphate enema each day. Children should never receive more than one phosphate enema a day because of the risk of phosphate intoxication. Children with impaired renal function should use Fleet enemas with caution.

The phosphate enema administered on a regular basis should result in a bowel movement 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 a saline enema is added to the phosphate one. If the addition of the saline enema still results in inadequate results, then high colonic washings are indicated with a Foley catheter attached to the tip of the bottle of the Fleet enema (see section an enemas using a Foley catheter).

A bowel management program is an individualized, ongoing process and there is no "magic formula." A larger volume of saline solution is necessary in those children who have hypo-motility as compared to those who have hypermotility and tend to empty their colon more rapidly.

The "right" saline enema is the one that can empty a child's colon and allow them to stay clean for the following 24 hours. This can be achieved only by trial and error and learning from previous lack of success.

Since the program is very personalized, the parents and children learn to look at the consistency and the amount of stool obtained after the enema to determine whether it was effective. After a period of time, parents will know when the enema was not effective and when they need to repeat it with a saline solution. Suppositories are only occasionally sufficient to stimulate a full bowel movement.

To make sure you are administering the enema in the right way, a nurse or physician should administer the first enema with the parent present. This will clarify any questions the parent may have regarding the technique, positioning or amount of solution to be given.

Implementation

A child who suffers from fecal incontinence is basically a happy child during the first part of his life when he/she is still in diapers because he/she is not different from other children. The problem begins when they start to socialize by themselves in an environment where all their classmates are already wearing normal underwear and they are still in diapers. That is the moment when the real problems (discrimination and ostracizing) start, potentially with serious psychological effects.

Toilet training for stool is a long-term goal for children with anorectal malformations, although this is not always possible. In children born with a good prognosis type of defect, parents should be encouraged to use the same strategies for toilet training as in children with normal anatomy. Children between 2 and 3 are instructed to sit on the toilet after every meal.

The parents are encourages to do this as a game and not as a punishment. The child can sit in front of a little table and play with his/her favorite toys.

The parents should sit with the child and not argue or force the child to remain seated. However, if the child gets up, the parents should put the toys away. The child should be rewarded for a bowel movement or voiding while on the toilet. If the child is not successfully toilet trained by school age, there are two alternatives: (1) do not send the child to school for one more year and continue attempts at toilet training or (2) try a bowel management program.

Therefore, it is advisable to start a bowel management at approximately 3 years of age. At this, age most of the children do not wear diapers.

It is important to emphasize that the choice as to

Bowel Management

Marc A. Levitt, MD and Alberto Peña, MD

when and how to start a bowel management program is something that each family has to decide. The goal is to have the child in school wearing normal underwear without being ridiculed by his/her classmates who are already toilet trained.

Microenema

Administration of a micro-enema to a child that is severely constipated only cleans the very last part of the rectum. This leaves the colon full of stool, now softened by the micro-enema, and the stool will leak more easily leading to episodes of soiling.

Obviously, there are some exceptions as there are some children who benefit from the use of microenemas and suppositories. If the goal is reached and the child is clean, this regimen is acceptable.

Long-Term Effects

Bowel management programs have been implemented for the last 15 years. We are not aware of any negative effects that can be attributed to the use of enemas or colonic irrigations. However we do not know what the consequence could be 20-30 year's from now.

It is important to remember that there are several kinds of enemas that can be administered. A Fleet enema is a phosphate enema and can be toxic when given in excess. If the specific recommendations of the manufacturer are followed, problems of phosphorus intoxication or hypocalcemia (low calcium) have not been seen. Many children receive colonic irrigations or enemas with saline solution, which is not different from the liquid that exists throughout our body in terms of the concentration of electrolytes, arid therefore any secondary effects from the use of these enemas have not been seen. Too much salt in the solution can lead to sweating, feelings of nausea and hypernatremia (too much sodium).

Many parents express a common misconception. They think that giving enemas may interfere with nutrition and absorption of nutrients. Enemas only wash stool from the colon, not nutrients. The main absorption of nutrients occurs in the small bowel and enemas do not wash that part of the intestines. Only the waste is being washed out.

Loose Stool and Diarrhea

Children with diarrhea have an overactive colon and most of the time they do not have a reservoir. This means that even when an enema cleans their colon rather easily; new stool passes fairly quickly from the cecum to the descending colon and the anus. To prevent this, a constipating diet and/or medications to slow down the colon (such as Loperamide) is recommended. Eliminating foods that further loosen bowel movements will help the colon to move slowly.

Parents are provided with a list of constipating type of foods to be given and a list of laxative foods to be avoided. The diet is very rigid: banana, apple, baked bread, white pasta with no sauce, boiled meat etc. Fried foods and dairy products must be avoided. Most parents know which meals provoke diarrhea and which constipate their child. To determine the right combination, the treatment starts with enemas, a very strict diet and loperamide (Imodium). Most children respond to this aggressive management within 24 hours. The child should remain on a strict diet until clean for 24 hours for 2 - 3 days in a row.

Then the child can choose one new food every 2 - 3 days to observe the effect on his/her colonic activity. If the child soils after eating a newly introduced food, eliminate that food from the diet on a permanent basis. The most liberal diet possible should be sought for the child. If he or she remains clean with a liberal diet, the dose of the medication can gradually be reduced to the lowest dose effective to keep the child clean for 24 hours.

Again, this is found by trial and error. This strict diet does not need to last forever. After about two months, in which the child has remained clean for 24 hours, they may have one of their "black list" foods that they have been craving. If the child soils after eating that food, the children know they must stay away from it. They must only introduce one new food a week and observe the effect on the bowel movement pattern.

Failure

If incontinence reappears, it means that something has changed in the child's habits and a meticulous evaluation is needed. The first thing to question is whether the enemas are still effective. The questions are, "Is my child emptying himself/ herself properly?" "Is she/he emptying the colon less successfully than before?"

To understand this, it is necessary to have an X-ray of the abdomen taken and analyze the quantity of stool present in the colon. If the X-ray shows a large amount of stool

in the colon after the enema, the enema needs to be adjusted (increase volume and/or concentration). A week of trial and error with daily X-rays to determine if the child is clean follows. On the other hand, if the X-ray shows a clean colon, the 'accidents" had to be due to increased motility and therefore it is necessary to introduce some medications to slow the

An important issue is to evaluate the consistency of the stool. Feces which remains in the colon for a long time become hard and sticky colon as described previously, as well as to observe a more strict diet. Therefore, it is necessary to carefully evaluate both the stool quantity and consistency.

Changes in diet on certain occasions like birthdays, holidays etc. may have repercussions on children with hypermotility, in the same way, moving from one's house, divorces, changes of school, etc. can play a role on the bowel function.

In some adolescents, the circumstances are predictable in which changes may influence the bowel function: exams, stress, etc. In this case it is possible to give a medication such as Imodium on the day before an exam, to slow down his/her bowel motility.

Duration of Treatment

In successful cases, the parents frequently ask if bowel management will be needed for life. The answer is yes for those patients born with a poor prognosis type of defect. It is conceivable that later in life, a child with a good prognosis type of defect may stop using enemas and remain clean by following a disciplined diet with regular meals (3 meals per day and no snacks) to provoke bowel movements at a predictable time.

However, there are some preliminary conditions to be met before trying this change: 1. The child has to be completely clean with a bowel management program. The child or the parents have to be able to evaluate both the quantity and the consistency of the stool. 2. The child is cooperative. The child has to be aware of his/her problem and motivated to experiment with new strategies to solve the problem. 3. The main goal of bowel management is to improve the quality of life of our children and insert them in a social context without "accidents," especially at school. 4. To understand that if the experiment is unsuccessful, the family can fall back on a bowel management program to ensure that the child is clean.

How to Proceed

Every summer, the children with some potential for bowel control can try, on an experimental basis, (by trial and error) to find out how well they can control their bowel movements without the help of enemas. This is done during the summer vacations when they can stay home and try some of the strategies. Some of the fundamental points include:

A. Reduced socialization. – It is expected that parents and child will stay at home and socialize very little.

B. Regular diet with a regular schedule – it is very important that the child have regular meals at a regular time, in order to take advantage of the gastrocolic reflex.

C. The child must try to pass stool after every meal.

D. He/She must remain alert all day while trying to learn to discriminate the feeling of imminent bowel movement.

E. If the child belongs to the "constipated" group, give him/her A laxative at a specific time everyday to provoke a single bowel movement per day. The dose of the laxative is adjusted by trial and error. It is best to first try the less aggressive and natural types of laxative, and then, depending on the child's response, use medications with more active ingredients. The first choice should be a laxative type of diet; the second a bulking agent or a stool softener. The third option is a laxative with an active ingredient and can be added to the stool softener. After a few days or weeks, the family can decide whether they want to continue the new regimen or go back to the bowel management program. This decision is up to the family and the child and is based on the quality of life experienced with each method.

A.C.E. or Malone Procedure

Most preschool and school-age children enjoy a good quality of life with 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

appendicostorny or a Malone procedure has been designed.

It is important to stress that the Malone procedure is just another way to administer an enema. Before implementing the Malone procedure, some preliminary conditions need to be met:

- The child has to be perfectly clean with his/her regular bowel management.
- The child has to be absolutely motivated - both in terms of the operation and the administration of the enemas as well as for his/her ability to evaluate the effects. The operation consists of connecting the appendix to the abdominal wall (usually at the belly button) and creating a valve mechanism that allows catheterization of the appendix but avoids leakage of stool. If the child has lost his/her appendix, it is possible to create a new one from the colon. This is called a continent neo-appendicostomy.

This procedure allows the child to administer an enema via inserting a small catheter into the stoma at the belly button while sitting on the toilet. The enema enters the bowel "above" stool. It is very easy and comfortable for any child to do.

Conclusions

Bowel management isn't just enema, a prescribed diet or taking medication. There is no miraculous recipe. The success of any of these strategies requires collaboration among the family, the child and the health care team. There are many variations depending on the child's needs: it takes dedication, determination, consistency and love by everyone involved.

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Pediatric Fecal Incontinence

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Marc A. Levitt, Richard A. Falcone Jr., Alberto Peña

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

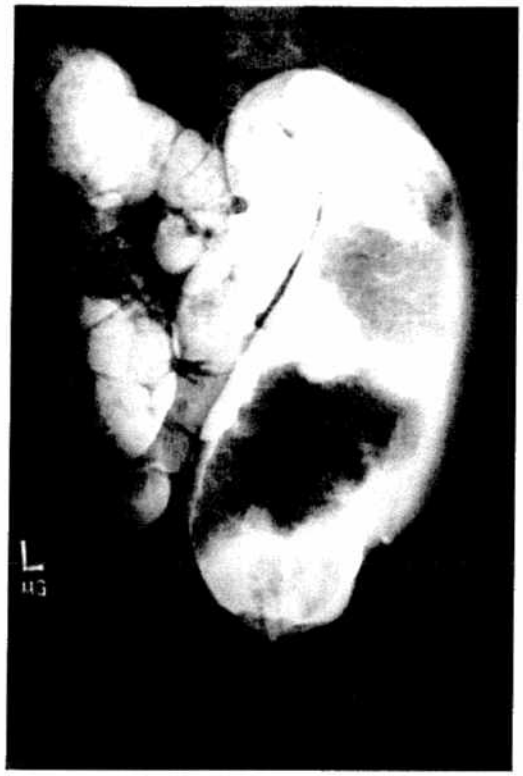

Fig. 1. Contrast enema of megarectosigmoid. Reprinted with permission from [1]

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

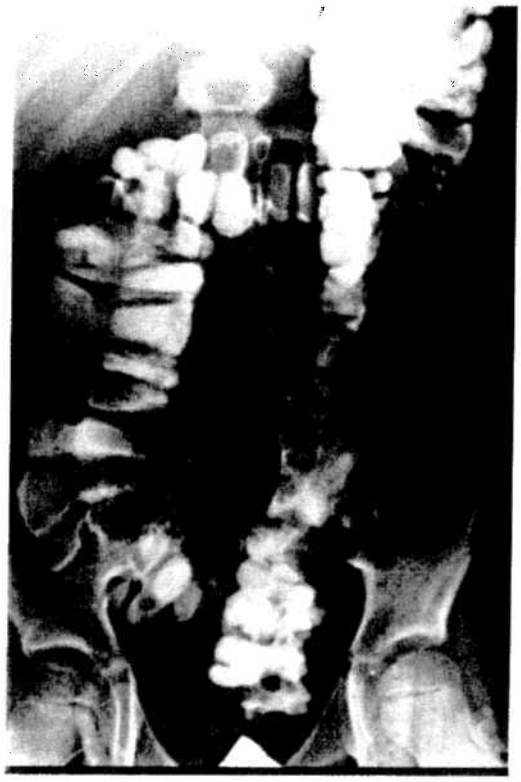

Fig. 2. Contrast enema in a patient with resected rectosigmoid. Reprinted with permission from [8]

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,

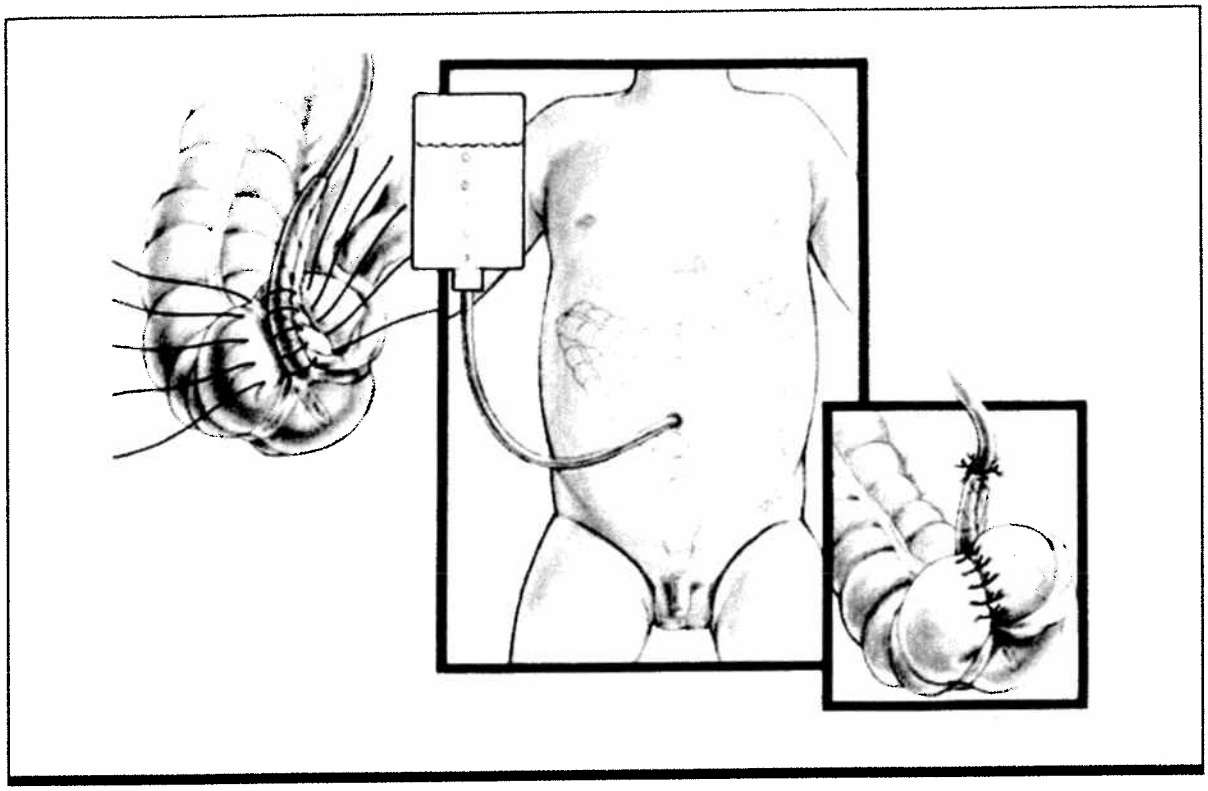

Fig. 3. Malone appendicostomy. Reprinted with permission from [9]

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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