Why X-Rays Are Needed if Bowel Management Program Stops Working and Needs Readjustment

After several failed attempts to improve their quality of life, many children suffering from fecal incontinence (the inability to control the bowels voluntarily) are brought to the Colorectal Center at Cincinnati Children's Hospital Medical Center. The Colorectal Center is an international treatment center for colorectal problems, including <u>fecal incontinence</u> following the surgical repair of anorectal malformations.

A specific protocol, the week-long <u>Bowel Management Program</u>, is used at the Colorectal Center to teach parents and other caregivers how to clean the child's colon once a day. A key element of this program is determining by trial and error the ideal type of enema that is capable of cleaning the colon of each individual patient.

In addition, for those patients who suffer from increased colonic motility (rapid movement of food through the colon), we have to find the best possible way to decrease their colonic motility to ensure that the patient's colon stays completely clean 24 hours per day in between enemas. That sometimes involves prescribing a specific <u>constipating diet</u> and/or medications that slow down the colon.

The bowel management program is considered successful once the patient can maintain a clean colon and is able to wear normal underwear all the time. The family and patients continue the program when they return home. After days, weeks, months or years, however, the program may no longer work, either because the enema is no longer cleaning the colon or because the colon is moving too fast due to dietary or other problems.

To readjust the bowel management program without the family having to come back to the Colorectal Center, parents are asked to obtain an X-ray of the abdomen when the patient has accidents (passing stool in the underwear in between enemas). This helps to determine the specific reason why the bowel management is no longer working. For instance, if the X-ray shows a colon with a large amount of stool, we may recommend using a larger enema or one with a more concentrated fluid to better empty the colon. On the other hand, if we see an empty colon, it is conceivable that the patient is suffering from some sort of diarrhea, and we have to prescribe treatment for that condition.



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Welcome to Bowel Management Week at Cincinnati Children's Hospital Medical Center! We are pleased and honored to assist you and your child in achieving a successful bowel management program.

Our bowel management week is offered once a month and runs as an outpatient program from a Friday through the following Friday. Prior to the start of Bowel Management, your child will be scheduled for some radiological tests and possibly consultations with other specialty services if appropriate for your child. The length of stay in Cincinnati would be approximately 10 days. The success of your child's bowel management program will be dependent upon your willingness to focus on your child during this time and understand that the length of stay here can be variable. Please, when choosing a week for bowel management, select a week that will allow you to be free of the responsibilities that would distract you from your child's bowel management program. We can offer a 95% success rate if families are willing to dedicate time and effort needed.

The week of bowel management will begin with a classroom lecture given by one of the physicians. Following lecture one of the RNs will teach enema administration. There will be a formal consultation with Dr. Pena , Dr Levitt , or Dr. Falcone in our clinic. All of the radiological tests will be completed prior to seeing the physicians. With those results, one of our physicians will determine the best program for your child. The program will consist of enemas or laxatives, and/or diet counseling, all of which are based on your child's capacity for fecal continence.

Each day during Bowel Management week, your child will get an abdominal x-ray to determine how effective the bowel management is. You and your child will have the availability of the Colorectal nurses, Tracy Ashworth, Cathy Bauer, or Lyndsey Foley to discuss issues and the daily regime which has been ordered for your child. Your child will be followed by one nurse throughout your stay in Cincinnati. The nurses and physicians meet daily to discuss your child's progress and then make recommendations dictated by whether your child is soiling in the underwear or not. Your nurse will call you with recommendations every day after our team meeting. You will then see Dr. Pena , Dr. Levitt, or Dr. Falcone in clinic at the end of bowel management prior to going home. It is important that families are aware that you will not see the physicians every day while you are in Cincinnati due to their busy surgery schedule, but please keep in mind, that your child is being discussed everyday with your physician. They are always kept informed of your child's progress and actively participating in your child's care and bowel management program.

We look forward to meeting you and your child during Bowel Management week.

Tracy Ashwork, RN

Tracy Ashworth, RN

Cathy Bauer, RN

Lyndsey Foley RN

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Bowel Management Week Documentation and Assessment

BMP	□lax trial	Immodium trial	Notes:	Previous BMP
Tethered cord		T/C surgery		
Sacral Ratio:		Sacrum:	C/E:	Common Channel:
Diagnosis:		Туре:		

DAY	DATE	RADIOLOGY FINDING	CLINICAL FINDING/ VBM	NS	FLEET	GLYCERIN	CASTILE	DIET	LAX	AMT	FIBER	AMT	IMMOD	INITIALS

Date/Notes: Date/Notes: Date/Notes:

Successful

Unsuccessful

Relationship

Continuing at home

Phone #1:

Why Unsuccessful:

Staying at

Home location

Phone#2:

Parent/Guardian Name:

Signature:

Diagnosis Hirschprungs ARM Cloaca

Sacral agenesis

Spina Bifida

Idiopathic constipation

type Fistula- perineal, etc., Prostatic fistula, Rectourethral fistula, Bladderneck



ENEMA ADMINISTRATION FOR THE BOWEL MANAGEMENT PROGRAM

The purpose of Bowel Management for patients with anorectal malformations is to allow a child that is incontinent of stool to be in clean underwear.

Gather equipment needed:

1) Enema Bag (Feeding tube bag)

- 2) 22fr or 24fr Silicone Catheter with 30ml balloon
- 3) 30ml syringe
- 4) Surgi-lube packets

5) One liter bottle of normal saline (amt ordered for enema is determined by physicians ordering enema)

6) Hook to hang enema bag (shower curtain hook)

Test balloon:

Pull out the plunger of catheter to the 30ml mark on the syringe and twist to secure onto the small port on the silicone catheter. Push the plunger into the port to blow up the balloon on the end of the silicone catheter. Deflate the balloon by withdrawing the plunger back to the 30 ml mark. The balloon should be fully deflated. This "tests" the balloon to be sure it is functioning prior to insertion into the rectum. To remove the syringe, simply untwist the syringe.

Pour prescribed amount of normal saline into enema bag and allow the fluid to drip out of the end of the silicone catheter (priming the tubing.) The fluid will drip out quickly, be sure not to allow too much fluid to drip out as you will not have the prescribed amount of fluid for the enema administration.

Administering the enema:

Now that the administration set is ready, **position** the child on their knees with buttock up and face on a pillow or lying down on stomach with hips propped up on towels or pillows. The idea of allowing gravity to pull the solution into the lower bowel during administration will allow for greater success. Place the silicone **catheter into the rectum** approx. 4-5 inches. Place your 30ml syringe onto the small port with plunger out to the 30ml mark and push gently to **inflate the balloon**. Once the 30ml of air is instilled into the balloon, take the syringe off and **pull gently on the catheter until you meet resistance**. This resistance is the balloon around the rectal wall and will prevent leaking of the solution around the rectum during the administration of the enema.



Administer the enema over 5- 10minutes, keeping gentle tension on the catheter. To slow down the administration of the fluid, use the roller clamp on the tubing, kink the tubing with your fingers, or lower the level of the enema bag to decrease the gravity flow.

Once the solution has been given, **keep the balloon inflated for 10 minutes, continue gentle tension on the catheter,** and allow fluid to sit in the lower colon. After 10 minutes, deflate the balloon with the 30ml syringe and have your child sit on the toilet. The time to expel the fecal contents may be as long as **45 minutes**. Expect for the **entire process to take approximately one hour**.

For younger children receiving enemas routinely, having special toys and books exclusively for the bathroom during enema administration will help distract the child. For the older child, their favorite book or homework could be used to help distract and keep the child occupied.

This time would be essential for the parent to be creative to assist in the successfulness of the bowel management program for their child. A time of year to begin bowel management should be when the child has no school, no physical distractions such as sports, practices, and or busy work schedule for the older teenager.

The administration of the enemas and the Bowel Management Program is trial and error. Complete cooperation by the parent, caregivers, and patients will be essential to be successful in keeping the child in clean underwear for 24 hours. The enema administration will be a process that will need to be done everyday.

Recipe for normal saline:

1 ¹/₂ teaspoon of table salt to 960 ml of warm tap water.



Supplies Needed for Daily Enema:

Kangaroo Bag (Kangaroo Pump Set) (Sherwood Medical # 713600)

Foley catheter 22fr. or 24fr. with 30 ml balloon (Kendall 100% Silicone Foley Catheter # 630245) or (Rochester Medical All Silicone Foley Catheter # 24222)

Syringe, 30 ml, slip tip (BD 30ml Syringe Luer Slip #309651)

Syringe, 60 ml, catheter tip (BD 60ml Syringe Catheter Tip #309620)

Supplies for Daily Enema through Malone:

Kangaroo Bag (Kangaroo Pump Set) (Sherwood Medical # 713600)

Feeding Tubes 8 fr (Kendall Feeding Tube 8 fr x 16 inch #260604) or (Kendall Feeding Tube 8 fr x 42 inch)

Coude Catheter 8 fr

(Mentor Self Cath Coude Olive Tip with Guide Strip #808)

Box of Water Soluble Lubricant

(K-Y or Surgi-lube)



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Enema Ingredients

Normal Saline:

To make at home:

- 1000 ml tap water and 1 ½ teaspoons of table salt
- After making this concentration pour into the enema bag only the amount ordered.
- *Do not change the concentration of water to salt

Glycerin:

Glycerin is a mild soap that can be added to the saline solution for the enema. This is sold over the counter

- May be purchased in our Outpatient Pharmacy
- Purchase the pint size bottle from our pharmacy
- May also be purchased in some pharmacies- call ahead and ask
- You can also find on the internet

Fleet (if prescribed by your physician):

This is a **phosphate** solution

- Fleet brand also makes plain saline enemas. Be sure to check the **active ingredient list. It should say phosphate**.
- This also comes in generic form
- It is sold over the counter
- 1 pediatric fleet is equivalent to ½ of an adult fleet (Buying the adult phosphate enema may save you money)

Castile soap:

This is a mild soap

- This is over the counter
- May be purchased from some pharmacies- call ahead and ask
- Can also be found on the internet
- We order in 'packet' amounts
- 1 packet is the equivalent of 9 ml

(cb 4/16/07 CR/RN/Pectin, glycerin, castile soap, normal saline)

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Castile Soap Sources for purchasing

You may be able to find it from a pharmacy

Online Sources:

Medex Supply: <u>http://www.medexsupply.com/products/pid-165/PDICastileSoap.htm</u>
Item#: P-L62550
 \$37.04
PDI Castile Soap

Click to enlarge

Packing info: Price per case (incl 10/boxes, 50 per each box)

Features:

- 0.30 fluid oz. packets (1 packet = 9 ml)
- Mild, gentle soap ideal for soft soap enemas
- CVS: Can order it. \$29 for 125 9 ml packets
- GNC: Dr. Bronners scented -- \$7.99/pint and \$12.99/quart
- The Soap Factory <u>http://www.alcasoft.com/soapfact/catalog/catagory.php?Catagory=liquid_soap</u>
- CCHMC DME -- \$4.26 for a box of 50 9 ml packets

NOTE: Walgreens and Kroger do not carry it.



Pectin (Fruit Pectin)

Soluble source of Fiber

- Also known as "Sure Jell", also a "Ball" product, or 'Certo'
- Can be found in grocery stores with canning supplies (used for making jam/ jelly)
- Comes in generic
- In general is sour in flavor but some unflavored products are available
- Available in powder and liquid
- Find a fook or drink that your child prefers- some examples are yogurt, jello jigglers, orange juice. *Be sure that as with all other medications that your child eats/ or drinks the entire serving or you will not know the dose that was taken.

Instructions for sending X-ray via e-mail

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- 4. Once the file is saved, you can now attach the file to an e-mail and send it.

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General Rules for the Administration of Laxatives

The Colorectal Center at Cincinnati Children's Hospital Medical Center recommends following these rules in order to become an expert in the management of your child's constipation.

- 1. The goal of laxatives is to prevent constipation. You do not want to have to treat constipation.
- 2. Your child should stay clean 24 hours a day.
- 3. The child must have at least one bowel movement every day.
- 4. Enemas are stopped and laxatives are to be administered once daily to provoke a bowel movement.
- 5. The amount of laxative is determined by trial and error.
- 6. Generally, the active ingredient used for the laxative trial is senna.
- 7. Senna is a laxative. It provokes a bowel movement. This is different than a stool softener, which just softens the stool but does not provoke a bowel movement.
- 8. Your child's stool should be soft and formed. Fiber will promote the right consistency of the stool.
- 9. Meals are to be at the same time every day. No snacks are to be given.
- 10. Stay close to a bathroom during this week.
- 11. Have your child sit on the toilet after meals as this is a time that movement of the colon occurs. This may provoke a bowel movement.
- 12. If one entire day goes by and your child does not have a bowel movement, it means that he/she did not receive enough laxative the previous day. Under those circumstances, you must give an enema to remove the stool from the rectum, but more importantly, you must increase the amount of laxatives and continue the new dose.
- 13. If your child develops diarrhea or very frequent, loose/watery stool it means that too much laxative was given. The dose needs to be decreased and you need to continue that dose consistently.
- 14. Within a week or two we should be able to determine the dose of laxative your child needs.
- 15. If your child has occasional voluntary bowel movements, but still soils significantly our recommendation is to go back to enemas for another year. Every year you will be dealing with a more mature child, who is more interested in becoming fecally continent, and therefore increasing the likelihood of success.

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Managing Fecal Incontinence in Children with Anorectal Malformations

Each year in the United States, an estimated 600 children, approximately one in every 4,000 babies, are born with malformations of the anus and/or rectum. The anus is the opening at the end of digestive tract where stool exits the body. The rectum is the final section of the large bowel leading to the anus. Children born with defects of the anus and rectum (anorectal malformations), can benefit from surgical and nonsurgical treatment approaches.

Colostomy

Most patients born with anorectal malformations receive a colostomy at birth and subsequently the malformation is repaired within the next eight months or so. A <u>colostomy</u> creates an artificial anus (stoma) to allow feces to pass out of the body and into a stoma bag.

Bowel Management

After the malformation has been repaired, the quality of life of children with anorectal malformations can be further improved through bowel management. The goal of a <u>bowel management program</u> is to help patients with fecal incontinence (the inability to voluntarily control the bowels) keep their bowels clean 24 hours per day and prevent problems that could lead to social isolation. The rate of success is high; 95% of patients who adhere to the Bowel Management Program are able to keep their bowels completely clean.

Developing Individualized Plans

To develop an individualized bowel management plan for each patient, the child is first evaluated to determine current colonic motility—how food moves through the colon. Patients with fecal incontinence can be classified into two major groups: those that suffer with <u>constipation</u> and those that tend to have diarrhea. Each group is managed completely differently.

For children with constipation, the treatment emphasis is on using large enemas capable of cleaning the entire colon every day. In general, these patients do not need any special diet or medication. The slow movement of waste through the colon guarantees that the patient will stay completely clean 24 and sometimes 48 hours in between enemas.

Children with a tendency to diarrhea receive a rather small enema because their colons are easy to clean. After the enema, these children then must follow a special constipating diet and medication to slow the motility of the colon and guarantee that they stay clean in between enemas.

The Bowel Management Program is implemented by trial and error over a period of one week with daily visits to the office of the Colorectal Center at Cincinnati Children's Hospital Medical Center. X-ray films are taken every day to see how clean the colon is maintained.

Optional Procedures Depend on Success of Management Plan

Once the bowel management program is shown to be successful, the patient can have an operation called a continent appendicostomy (Malone procedure). The operation connects the appendix (a small pouch attached to the large intestine) to the navel and creates a one-way valve mechanism. This allows a small catheter (a thin, flexible tube) to be passed through the navel so the child can receive an enema while sitting on the toilet, but the valve prevents leakage of stool at other times. About 10% of patients undergoing this procedure require a revision to make the opening a little larger to allow for the catheter, or to tighten the valve to avoid leakage of stool.

The purpose of this operation is to improve the quality of life of these patients, particularly later on when they become teenagers, by allowing them to become more independent. This operation is performed only if the bowel management program is successful. Bowel management is not successful in about 5% of patients. These patients are offered the option of a colostomy.



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BOWEL MANAGEMENT FOR FECAL INCONTINENCE IN PATIENTS WITH ANORECTAL MALFORMATIONS

Peña A, Guardino K, Tovilla JM, Levitt MA, Rodriguez G, Torres R

INTRODUCTION: Fecal incontinence is a serious problem that provokes social segregation and psychologic sequelae. Patients with anorectal malformations frequently suffer fecal incontinence despite the efforts of pediatric surgeons. Based on the estimated incidence of anorectal defects in newborns¹ and the population's vital statistics,² approximately 780 new children with imperforate anus are born in the United States every year. At least 25 to 30% of them will suffer from fecal incontinence.³ In addition, another 30% will suffer from other functional defecation disorders such as constipation, occasional soiling, and fecal incontinence during periods of diarrhea.³⁻⁵

Medical management with enemas, laxatives, and medications has been tried in the past with varying success.⁴⁻⁹ These treatments are often given without a specific rationale and in an indiscriminate manner. There are patients who were referred to a psychiatrist when the surgeon felt the treatment had been technically correct and that the incontinence must, therefore, have a psychologic basis.¹⁰

Our institution is a referral center for the treatment of patients with anorectal malformations. Over the last 11 years we have seen 348 patients in consultation for the treatment of fecal incontinence. These patients were all born with an anorectal malformation and were operated on at another institution. From this experience we have learned that there are different types of fecal incontinence that depend on the original malformation and the original operation. We have also learned how to evaluate these patients, to recognize the specific type of fecal incontinence that they suffer from, and to implement the best modality of treatment for them. Laxatives, medications to modify colonic motility, enemas, and colonic irrigations were found to play a role in the management of these patients, provided they are used with a specific rationale, which depends on the specific type of fecal incontinence. The impact on a patient's quality of life with bowel management is perhaps more significant than that of surgery itself.

MATERIALS AND METHODS: Between 1985 and 1996, 348 patients operated on at other institutions for imperforate anus were referred to our center with the diagnosis of fecal incontinence. They formed the basis of a retrospective review. All patients were evaluated with history, physical examination, spinal and sacral radiographs, contrast enema, and magnetic resonance imaging of the pelvis. On the basis of this evaluation we learned to distinguish three different groups of fecally incontinent patients.

Each group has enough characteristics in common to be separated one from the other, and each group benefits from a specific treatment. The determination of these groups was clarified in retrospect and proved to be the key element for success in the management.

Group I included 147 patients who were considered candidates for a reoperation. These were patients with potential for bowel control, good sphincters, normal or near-normal sacra, and evidence of being born with a defect that has a good functional prognosis.³ They also have a completely mislocated rectum and a wellpreserved sphincter. These patients underwent a secondary pull-through and will be the subject of a future report.

Group II, which included 172 patients, were those without potential for bowel control and, therefore, candidates for our Bowel Management Program. This group represents the essential part of this study. The characteristics of these children include a poor sacrum, poor sphincters, and evidence of being born with a defect that has a poor functional prognosis³ (i.e., rectobladder neck fistula or a long common channel cloaca). These patients may have the rectum well located (within the limits of the sphincter mechanism) or mislocated, but they were not considered candidates for a reoperation because they had no potential for voluntary bowel control based on their history and their anatomy. This group included two distinct types of fecally incontinent patients: (A) those with a tendency to constipation and (B) those with a tendency to suffer diarrhea. Each subgroup required a different type of bowel management. Designation of the patient to one of these groups turned out to be essential for success.

There were 44 patients with a tendency toward constipation (Group IIA). They suffered from different degrees of megasigmoid as shown by contrast enema (Figure 1). The operation these patients underwent at another institution to repair their original malformation included the preservation of their rectum (i.e., anoplasty, sacroperineal approach, or posterior sagittal anorectoplasty). The bowel management involved the use of large enemas or colonic irrigations, every 24 hours, to clean a large, floppy megacolon. They did not receive a special diet or medication. It was expected that they would remain completely clean in between enemas because of their problem of constipation, which was a manifestation of a hypomotility disorder of the rectosigmoid.

Figure 1. Contrast enema of megasigmoid.



There were 128 patients with a tendency to have diarrhea (Group IIB). They had all been

operated on previously with a surgical technique that included the resection of their rectum and at least part of their sigmoid colon (i.e. endorectal resections). Their contrast enema shows a characteristic image of absence of the sigmoid: a nondilatated colon running straight from the splenic flexure down to the anus. One can frequently see normal colonic haustra down near the anus (Figure 2).

Figure 2. Contrast enema of a patient with absent rectosigmoid. Note the straight trajectory of a nondilatated colon from the splenic flexure to the perineum.



The bowel management in these cases involved the use of daily small enemas to clean the colon. This turned out to be an easy task because these patients have a short colon. The main challenge in this group was to decrease the motility of the colon to avoid passing of stool between enemas. For this we used a strict constipating diet and/or administration of drugs to slow colonic motility. The agents we used include loperamide hydrochloride (1-2 mg orally three times per day) or diphenoxylate hydrochloride with atropine sulfate (0.3-0.4 mg per kilogram orally per day in four divided doses).

The bowel management program in both groups of patients was implemented by trial and error over a period of several days. There is no way to predict in a specific patient the exact type and volume of enema that will succeed in cleaning the colon. The family was encouraged to stay at our Ronald McDonald House, which is located within the premises of the Hospital.

The medical records and available films of the patients were sent well in advance to be reviewed by the senior author. A diagnostic plan was developed, the patient was interviewed and examined on the first day, and the necessary diagnostic tests were performed. The patient was classified through use of the above- mentioned criteria (Figure 3). A specific type of enema was administered to the patient, depending on the type of colon seen in the contrast enema. A very large megacolon was expected to require a large enema usually with a combination of phosphate enema plus saline solution. Modifications were made in the subsequent days depending on the patient's response. A nurse clinician was responsible for instructing the parents how to administer the enemas. This included the administration of the first enema, which was given by the nurse in the presence of the parents. Subsequent enemas were given by the parents supervised by the nurse.

Figure 3. Decision-making algorithm followed in patients with anorectal malformations suffering from fecal incontinence.



The patient was seen every day by the surgeon and the nurse, and the parents reported the results of the management, specifically whether or not the patient had been clean and, if not, when the patient had soiled.

Episodes of soiling the underwear or large unexpected bowel movements were called "accidents." The presence of "accidents" in between enemas was interpreted in a different way, depending on whether or not the patient belonged to the constipation group (A) or to the diarrhea group (B). In the constipation group, an accident represented a failure to clean a very large colon. The treatment was modified to administer a larger enema or to increase the concentration of salt. The family was also specifically asked about possible mishaps or errors in the method of giving the enema. Sometimes, for instance, it was found that the enema fluid was leaking during the process of giving the enema. (Figure 4) To avoid this problem, the parents or the patient (when older than 12 years old) were instructed to pass a welllubricated rectal rubber tube (cal. 20-24 F) as high as possible to deliver the enema fluid into the left colon. Occasionally leakage occurred with this technique, resulting from kinking of the tube (Figure 5). If this method was unsuccessful, the patient or the parents were instructed to use a large Foley catheter to avoid the leakage of the enema fluid during the administration, a procedure similar to the one described by Shandling in cases of spina bifida.⁶ The catheter was introduced into the rectum (approximately 8 cm), the balloon was inflated with 10 cc of water, and traction was exerted on the catheter. Sometimes 20 or 30 cc were needed to be sure the balloon was large enough to remain inside the rectum even with pulling on the catheter. The enema was given while traction was applied on the catheter with the balloon inflated. The balloon thus served as a plug to avoid leakage, creating a more efficient enema (Figure 6).

Figure 4. Enema administration in a rectum with impacted stool, which leads to leakage.



Figure 5. The rubber tube used for enema administration can kink and lead to leakage.



Figure 6. Foley catheter technique with inflated balloon and traction employed to avoid leakage.



The presence of "accidents" in the patients suffering from diarrhea was interpreted as failure to slow down the motility of the colon between enemas. In that case, the diet was made more strict and the medication doses were increased.

If it was unclear why the patient was suffering "accidents," a plain radiograph of the abdomen was taken, which allowed us to determine whether or not the colon was clean of stool. When we found a colon that was not clean, we administered a larger enema. If the patient did not tolerate more volume, we doubled the concentration of sodium chloride in the water or added soap or bisacodyl (dulcolax). If the plain radiograph showed a clean colon, the "accidents" had to be due to increased motility, which was treated as described previously.

When the patients continued having "accidents" in spite of our efforts over a period of 7-10 days of trial and error, we considered that the program had failed, and we offered the patient a permanent colostomy.

When the patients remained completely clean for three consecutive days we considered the program successful. The family was then informed about another therapeutic alternative, continent namely. the creation of а appendicostomy (Malone procedure).¹¹⁻¹³ This technique allows the patient to administer the enema in an antegrade manner and is found highly convenient by most patients. Since this is an operation done with the specific purpose of improving the quality of life of the patient, it was considered optional and the opinion of the patient was fundamental in making the decision. If the patient had undergone an appendectomy in the past, a new appendix is created from a flap of cecum with a one-way valve mechanism. The appendicostomy or neoappendicostomy allows for catheterization and enema administration and also avoids leakage of stool. The specific details of this operation and its results in this group of patients are the subject of a future publication.

Group III included 29 patients who were

found serendipitously. These patients all were born with a benign defect, for which one would expect a good result in terms of bowel control. They also

had a normal or near-normal sacrum, good sphincters, and a technically correct operation as demonstrated on MRI by the presence of the rectum located within the limits of the sphincter mechanism. Despite this, the patients presented complaining of fecal incontinence. They also suffered from severe constipation and the contrast enema showed a severe megasigmoid.

The first of these 29 patients was seen in the emergency room frequently for fecal impaction in spite of an aggressive treatment that included high doses of laxatives and enemas. In desperation, we offered the family a sigmoid resection with preservation of the rectum¹⁴ if not to cure his constipation, at least to facilitate his management. The operation was performed and to our surprise the patient not only had no further symptoms of constipation but also became fecally continent. In retrospect, we can say that the patient was not suffering from real fecal incontinence but rather from overflow pseudoincontinence.

After that case, 28 subsequent patients were detected with similar characteristics. Before offering these patients a sigmoid resection it was mandatory to confirm that they are suffering from overflow pseudoincontinence rather than real fecal incontinence with constipation. Failure to make this differentiation may lead to an unnecessary operation; a fecally incontinent constipated patient would be changed to one with diarrhea, who is much more difficult to manage.

The differentiation is carried out by disimpacting the patient first with enemas. During the following days we prescribe increasing dosages of laxatives to find out the amount of medication necessary to provoke a bowel movement without an enema or a suppository. Once we reach that dosage, we observe the patient's bowel control. If continent, then it is obvious that the patient was suffering from overflow pseudoincontinence and will benefit from the sigmoid resection. If the patient has no bowel control while on laxatives, it means that the patient suffers from real fecal incontinence. In this case the sigmoid resection is contraindicated. The patient must be managed as an incontinent patient with constipation (large enemas, no diet, and no medication).

RESULTS: Group I, 147 patients, were subjected to a reoperation and will be discussed in a separate publication.

Group II, 172 patients, were subjected to our Bowel Management Program. Of 44 patients with a tendency toward constipation (group IIA), the management was successful in 41 (93%) and it took an average of 4.4 days (range 2-15 days) to achieve success. Of 128 patients with a tendency to suffer diarrhea (group IIB), the management was successful in 112 (88%) and it took an average of 3.6 days (range 1-15 days) to achieve success.

Group III, 29 patients, were considered to have pseudoincontinence. Of these, 28 patients (97%) had voluntary bowel movements with varying doses of laxatives. Nineteen patients began having voluntary bowel movements with appropriate doses of laxatives alone. Nine patients underwent sigmoid resection¹³ which led to voluntary bowel movements and a substantial reduction of their laxative requirement.

Overall success was achieved in 181 patients (90%).

Of the three patients who suffered incontinence and constipation and in whom bowel management was unsuccessful, two were noncompliant with the bowel management regimen, and one patient (0.2%) underwent a permanent colostomy.

Of 16 patients who suffered from diarrhea

and incontinence and had unsuccessful bowel management, failure was attributed to noncompliance in six, an extremely short colon in one, and failure to control the severe diarrhea in two; in seven a cause of failure could not be determined. Six of these patients (0.5%) underwent a permanent colostomy.

In 17 patients it took more than seven days to achieve a successful bowel management. Seven of these patients were fecally impacted at the beginning and it took several days to disimpact them. Two patients were noncompliant with the regimen, 2 suffered from diarrhea, and 2 had the flu during the regimen; in 4 we do not have enough information to explain the delay.

There were seven patients (3 from the constipation group, 4 from the diarrhea group) who underwent a continent appendicostomy for administration of enemas.

Twenty of our patients also complained of urinary incontinence on the first consultation. Subsequently, 19 of them developed urinary control once the bowel management was successful.

From the entire series, 103 patients were determined to have an original defect with good prognosis³ based on their history and/or radiologic or anatomic evidence. These included patients with rectoperineal fistula, rectovestibular fistula, imperforate anus without fistula, and rectourethral bulbar fistula. Of these, 24 had constipation, 52 had diarrhea, and 27 had pseudoincontinence. In retrospect we found that these patients had some form of mismanagement that rendered them fecally incontinent, even when they were born with potential for bowel control.

DISCUSSION: Most of our patients had previous treatment for incontinence, using enemas, laxatives, or medications. The major difference, however, was that these therapeutic measures were frequently given in an indiscriminate manner. Not only did they fail to keep the patient clean but many times they actually worsened the patient's situation. Examples of these include:

• Administration of an enema to a constipated patient without supervision often gave the patient the impression of making him worse. This is because the enema fluid only softened the impacted stool, provoking a more serious problem of stool leakage.

The indiscriminate administration of loperamide to a patient with fecal incontinence gave the initial impression of improvement by provoking obstipation, but that was frequently followed by a bout of explosive diarrhea.

• The administration of laxatives to patients with fecal impaction usually provoked severe cramps, vomiting, and worsening of the leakage of stool.

• The simultaneous administration of laxatives and enemas provoked passing of stool in between enemas, despite the success of the enema in cleaning the colon.

We commonly heard the parents of our patients express disappointment when they learned that they have travelled a long distance with a great expectation of improving their condition, only to find out that our Bowel Management Program was nothing more than enemas, diet, and/or medication, which most of them had tried before without success. It was necessary, therefore, to go through a complete explanation to the parents to convince them that even when we were using the same therapeutic armamentarium, the results should be better because we were following a specific rationale.

It was not uncommon to hear that their local physicians made pejorative remarks when they learned that the patients were travelling far away "just to learn how to give an enema." This also influenced insurance companies, who had

problems understanding the basis of this treatment and refused to pay for the amount of time employed by our team to improve these patients.

In our literature review we found that the management of fecal incontinence can be divided into surgical and nonsurgical approaches. Surgical measures that have been employed include attempts to reconstruct the sphincter mechanism by using smooth muscle,¹⁵⁻²⁰ voluntary muscle (gluteus,²¹⁻²³ gracilis,²⁴⁻³⁵ internal obturator,³⁶ levatorplasties³⁷⁻³⁹), or synthetic material⁴⁰⁻⁴⁵ around the rectum in an effort to form an active sphincter.

Nonsurgical management has included biofeedback techniques,⁴⁶⁻⁵⁰ behavior modification,⁵¹⁻⁵³ psychotherapy,⁵⁴ medications,⁹ behavior and enemas or high colonic irrigations.⁶ There has been recent enthusiasm for the gracilis muscle reconstruction with electrical stimulation,³⁵ but long term results of this method are still not available.

Of all the modalities of treatment of fecal incontinence that we have used and have read about, our Bowel Management Program has rendered the best results and has significantly improved the quality of life of many families.

The results were slightly better in patients with a tendency to constipation than in patients with a tendency to diarrhea, and there were more failures and permanent colostomies in the diarrhea group. Noncompliance with specific dietary recommendations was the most common cause of failure. The constipation group did take slightly longer to achieve successful bowel management, but this was due to the need for fecal disimpaction. Cleaning the colon in the diarrhea group was relatively easy; more difficult was keeping the colon quiet between enemas.

Essentially the entire pseudoincontinence group was represented by patients with an

original anorectal defect with good prognosis. It cannot be overemphasized that in these cases the original mismanagement was the failure to prevent constipation and its sequelae.

Interestingly, in retrospect, 19 patients who presented complaining of both urinary and fecal incontinence never really had urinary incontinence. When successful bowel management was achieved and the patients were clean and able to wear normal underwear, they became aware of their capacity to control urine.

A truly unfortunate group of patients were those who were born with an anorectal defect with good prognosis but who became fecally incontinent as a consequence of some form of surgical mismanagement at the original operation. To avoid this, more emphasis should be placed in the training of pediatric surgeons on the correct indications and performance of the operations to repair anorectal malformations with good prognosis. In a benign type of defect, a mismanagment is totally unacceptable.

The group of patients who underwent an endorectal pull-through during their original operation were all fecally incontinent with a tendency toward diarrhea. The endorectal approach was originally created to avoid damage to important pelvic structures (mainly nerves) during the dissection of the rectum. However, the endorectal dissection by itself, by definition, sacrifices the rectum and part of the sigmoid, which represents the patient's fecal reservoir. The consequence of this is a tendency to suffer diarrhea and to constantly pass stool, no different than a perineal colostomy. In our experience these are the patients who are most difficult to manage. When the endorectal dissection is performed in patients with a benign malformation (for which one would expect otherwise excellent results in terms of bowel function) the patients always suffer from fecal incontinence in spite of the fact that they were born with a benign condition

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Constipation in Imperforate Anus

Constipation is a problem in the great majority of patients who are born with an <u>imperforate anus</u> (a birth defect in which the anal canal fails to develop) and have an operation to repair the condition that includes the preservation of their original rectum.

This constipation seems to be the clinical manifestation of a hypomotility disorder (a slower than normal movement of food) through the rectosigmoid colon (the lower section of the colon that attaches to the rectum, where stool is usually stored until it is expelled from the body).

Further analysis of our cases shows that this problem of constipation is worse in lower defects. ¹ We have found the reverse is also true: the higher the malformation, the more chances of suffering from <u>fecal</u> <u>incontinence</u> (the inability to control bowel movements voluntarily) and the less chance of having <u>constipation</u>.

"High" and "low" refer to the relationship of anorectal defects to the levator muscle complex, which elevates the organs and structures in the area. In "low" anorectal defects, the rectum descends through the levator muscle complex, while in high anorectal defects, the rectum fails to descend through the levator muscles.

Site of Colostomy Affects Severity of Constipation

The other primary factor affecting the degree of constipation appears to be the site of <u>colostomy</u>. A colostomy creates an artificial anus (stoma) to allow feces to pass out of the body and into a stoma bag. Patients who have had colostomies in the transverse section of the colon have a greater chance of having constipation than those who have had colostomies in the descending section of the colon.

In patients with transverse colostomies, distal colostograms, the most accurate diagnostic studies of the area, typically show severe dilatation of the rectosigmoid, especially if closure of the colostomy was delayed for a long period of time.

These patients suffer more severe constipation after repair of the anorectal malformation and closure of the colostomy. In fact, there is a direct relationship between the degree of rectosigmoid dilatation prior to colostomy closure and the degree of constipation the patient will suffer.

Loop Colostomies

Loop colostomies, particularly those that allow the passing of stool from proximal to distal stoma, will provoke the worse fecal impaction in the distal rectum, the worse megasigmoid syndrome (swelling in the sigmoid colon), and therefore, the worse constipation.

Tapering or not tapering the rectum in our series of cases, did not have any relationship with the degree of constipation that the patient suffers from.

Colostomy Closure

After <u>colostomy closure</u>, proper treatment of constipation is imperative. When constipation is not treated properly, the megasigmoid worsens.

More dilatation of the rectum leads to greater constipation and constipation becomes a self perpetuating and self aggravating condition. The lessons then seem to be very clear; we must try to keep the rectum as empty as possible to avoid rectosigmoid dilatation.

Constipation is a very serious problem because eventually it provokes fecal incontinence, even in patients who were born with a potential for bowel control. This is what we call over-flow pseudo-incontinence.

Treatment for Constipation

The treatment method for constipation depends on whether or not the patient has the potential for bowel control. We are very thorough about classifying patients according to their potential for bowel control and try to determine this from the very beginning of treatment.

A poorly formed sacrum (the lower part of the spine that forms part of the pelvis), a very high malformation (recto-bladderneck fistula or a long cloaca, the fusion of rectal, vaginal, and urinary channels into a single common channel), or the presence of a mass in front of the sacrum, usually means that the patient has no potential for bowel control.¹

We define a poor sacrum as one with more than three vertebrae missing, or with a sacral ratio of less than 0.4. The sacral ratio is defined as the ratio of the distance from the bottom of the sacro-iliac joint to the tip of the sacrum divided by the distance from the top of the iliac bone to the bottom of the sacro-iliac joint.

Patients with Good Potential for Bowel Control

Conversely, patients with good potential for bowel control include those with a well formed sacrum, or a "benign" malformation, such as rectal atresia (lack of a normal opening in the rectum), perineal fistula (an abnormal anal opening), vestibular fistula, imperforate anus with no fistula, or rectourethal-bulbar fistula.¹

Patients with Poor Potential for Bowel Control and Constipation

Patients with poor potential for bowel control and constipation should be treated with an <u>enema</u> every day. In these cases, hypomotility of the rectosigmoid represents an advantage because it makes the bowel management easier. Emphasis should be on cleaning the colon every day. This can be done with aggressive, large volume, salt water enemas.²

The program should be implemented over a period of one week by trial and error. Daily abdominal X-rays allow reliable assessment of effectiveness of the enemas.

The volume and salt concentration of the enemas is increased until the patient's colon is effectively cleaned. This regimen is then continued on a daily basis, with the goal of keeping patients completely clean for 24 hours a day between enemas.

Patients with Good Potential for Bowel Control and Constipation

Patients with good potential for bowel control and constipation should be treated with <u>laxatives</u>. Trial and error determines the amount of laxatives that each patient needs. The amount of laxative that each patient needs is frequently very large.

The goal is to empty the rectum every day. Once bowel control has been achieved with laxatives, the patient is maintained on this regimen. If the amount required is enormous, a sigmoid resection may be offered, which will allow a reduction in the amount of laxative requirements.^{2, 3}

Additional Conditions and Considerations

During surgery to treat imperforate anus, the rectosigmoid should never be surgically removed because this leads to fecal incontinence. This occurred in many patients treated in the past with a surgical procedure that is no longer used.

Tissue samples from the most distal part (the end) of the rectum and/or fistula (an abnormal passageway between two areas of the digestive tract) in patients with anorectal malformations may show evidence of nerve abnormalities, including neuronal intestinal dysplasia (NID) and/or aganglionosis, the absence of certain nerve cells known as ganglion cells. There is no way, however, to know the relevance of those findings. While it may be tempting to use these findings to try to explain the constipation, the cause of constipation in these patients is more likely to be related to a very distended rectosigmoid and therefore, impaired peristalsis, a wavelike movement of muscles that moves food and liquid through the gastrointestinal tract. Any hollow internal organ of the body that becomes overdistended for a long period of time loses its peristaltic ability.

Hirschsprung's Disease

We are very skeptical about the diagnosis of imperforate anus and <u>Hirschpsrung's disease</u>, a birth defect in which some nerve cells are lacking in the large intestine and the intestine cannot move stool through. As a result, the intestine gets blocked and the abdomen becomes swollen. The reason for this skepticism is that we have never seen a patient with imperforate anus develop enterocolitis, inflammation of both the large and small intestine. The correct diagnosis is necessary to implement proper treatment strategies and avoid surgery that could cure constipation but make the patient fecally incontinent.

Tethered Cord

<u>Tethered cord</u>, the abnormal attachment of the spinal cord to the bones of the spine, has occurred in 24% of patients in our study of anorectal malformations.⁴ The more complex and high the malformation, the higher the chances of tethered cord. Forty-three percent of our group of complex anorectal malformations have tethered cord, but it is present in only 11% of our patients with rectovestibular fistulas.

Tethered cord is clearly more common in patients with imperforate anus who suffer from fecal incontinence, but our study could not clarify whether or not the fecal incontinence was due to the presence of the tethered cord. This is because most of those patients had a very complex malformation, which alone could be responsible for the fecal incontinence. Eighteen of our patients underwent surgical untethering of the cord and none had any significant change in bowel or urinary function postoperatively. We could not find a single patient with tethered cord who experienced incontinence which could be attributed to the cord defect alone. Surgical untethering may be indicated for other reasons, but at the present time, there is no good evidence to demonstrate that surgical untethering improves the prognosis of these patients.

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Toilet Training for Patients Previously Treated With a Bowel Management Program

Children with fecal incontinence, meaning they are unable to control their bowels, can be treated with a <u>bowel management program</u> that uses daily enemas to keep their colon quiet and clean 95% of the time. <u>Anorectal malformations</u> are defects of the anus, the opening at the end of digestive tract where stool exits the body, and the rectum, the final section of the large bowel leading to the anus.

When a patient reaches 3 years of age and is still incontinent and the parents are considering sending the child to school, we talk to the parents about the necessity of implementing a bowel management program to keep the child's bowel clean. This will allow the child to go to school with normal underwear and nobody at school that does not have to know should be aware of the child's problem.

Parents frequently ask the question, "Is this treatment for life?" The answer is, not necessarily. Some of those patients were born with anorectal malformations with a very bad prognosis or chance of gaining control of their bowels and most likely they will continue with the bowel management for many years. Other patients may have partial fecal incontinence, anatomical features such as well constructed sacrum (the lower part of the spine that forms part of the pelvis), and other factors that may indicate these patients could achieve bowel control in the future. For these individuals, bowel control trials should be conducted periodically.

Each year during summer vacations, when the child is not attending school, we can conduct a trial with laxatives. For this, we stop the enemas and see how much bowel control the child has. Within one or two weeks, the parents would be able to determine whether or not the patient is ready to continue without enemas.

Parents should learn about the specific type of malformation that their child was born with, since each defect has a different prognosis. This knowledge will enable parents to have realistic expectations about whether their child may achieve bowel control.

Basic Principles of Bowel Control Trial

A bowel control trial is carried out at home for a period of one or two weeks and must follow certain basic principles and procedures.

- Talk to your child about the trial and make sure your child understands the trial's purpose and the general process.
- Explain that during the trial, the child must avoid social gathering because of the risk of having unexpected embarrassing bowel movements.
- Motivate your child to keep a clean colon. Establish some incentive, such as buying new underwear, and provide rewards for every day "clean day."
- Stop the enemas.

- Give three meals per day with no snacks. The purpose of this is to try to condition the colon to empty periodically at the same time. It is much easier to toilet train a child that has one, or even two or three bowel movements every day at predictable times, than a child that has very irregular bouts of diarrhea and constipation. Regularity is very important, but not always possible to achieve because of the difficulty in regulating colonic motility (the movement of food through the digestive tract).
- Try to include the same type of food in every meal. The type of food largely depends on whether the child has been classified as belonging to the constipated group or the diarrhea group. Most patients with anorectal malformations suffer from constipation and therefore need a <u>laxative type of food</u>. Most parents know what type of food has a laxative effect in their children, but if you need help determining types of foods that are laxative or constipating, <u>call the Colorectal Center</u> at Cincinnati Children's Hospital Medical Center.
- Make sure the child stays close to the bathroom.

Laxatives Are Often Needed

Because most of these children suffer from <u>constipation</u>, they commonly need some form of laxative to have bowel movements without enemas. The type and amount of laxative that a specific patient needs must be determined on an individual basis by trial and error. Most children with anorectal malformations do not respond to the usual recommended dosage of laxative. In determining how much laxative your child needs, try to remember if your child previously took <u>laxatives</u> and if so, how the child responded.

The laxative must be given once a day (not three times per day) in order to try, again, to provoke one or two bowel movements per day. Start by giving 1 teaspoon of a determined laxative at night. Keep in mind the effect can be expected the day after the laxative was administered. If the entire day goes by without the child having a bowel movement, that means that that not enough laxative was given and the amount should be doubled that night. It also means that the child needs an enema to remove the stool that has been there for 24 hours in order to avoid fecal impaction.

If the child does not have bowel movement the following day, the amount of laxative should be increased further and you should continue that way every day, while also giving enemas to avoid impaction. If the child suddenly develops diarrhea, that means that the amount of laxative given the day before was excessive and therefore should be reduced, but not eliminated completely. What we are describing here is a process of trial and error to try to find the right amount of laxative—the dose that provokes bowel movements but not diarrhea.

Most of the time we suggest parents also administer some form of fiber product in addition to the laxative. The fiber will provide bulk in the stool so the child can have formed stool rather than liquid stool. If the child's stools are too liquid, increase the amount of fiber and decrease the amount of laxative. Conversely, if the child has formed stool, but can't have bowel movements easily, increase the amount of laxative.

Sometimes the child is having bowel movements in the toilet, but the parents are not sure whether the bowels are being completely emptied. Under those circumstances, we recommend getting an X-ray film of the abdomen so we can see how well the child is emptying the colon. To promote complete emptying of the colon, the child is asked to sit on the toilet or potty three times per day, particularly after breakfast, lunch

and dinner, or at another time the parents think the child is most likely to have a bowel movement. Sometimes suppositories can be used to provoke and help regulate the bowel movements.

Using Results To Determine Future Course of Action

With this trial and error approach, within a week or two, parents should be able to determine the type and amount of laxative their child needs, how much bowel control the child has, and whether or not the child is able to maintain a clean colon. If the test shows the child actually has bowel control, that means the child does not need any more enemas but must continue with the same <u>bowel management program</u>. If the test shows that the child has some bowel control, with occasional voluntary bowel movements, but still soils significantly, the child may need to go back to being treated with enemas. If the child is still soiling significantly, our recommendation is to go back to the bowel management for another year, and not to take unnecessary risks of embarrassing accidents at school.

Every summer, when the child is not attending school, the parents may try again. Every year, they will be dealing with an older patient, more interested in becoming fecally continent, and therefore, with more possibilities of success.



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Constipation in Imperforate Anus Dr. Alberto Peña

The great majority of patients born with imperforate anus who undergo a repair which includes the preservation of their original rectum will suffer constipation. This constipation seems to be the clinical manifestation of a hypomotility disorder of the rectosigmoid colon. Further analysis of our cases show that this problem of constipation is worse in lower defects. We have found that the reverse is also true, the higher the malformation, the more chance of suffering from fecal incontinence and the less chance of having constipation. The other primary factor affecting the degree of constipation appears to be the site of colostomy formation. Patients subjected to transverse colostomies have a greater chance of having constipation than those with a descending colostomy. The distal colostograms of patients with transverse colostomies typically show severe dilatation of the rectosigmoid, especially if closure of the colostomy was delayed for a long period of time. These patients suffer more severe constipation after repair. In fact, there is a direct relationship between the degree of redo sigmoid dilatation prior to colostomy closure and the degree of constipation the patient will suffer. Loop colostomies, particularly those that allow the passing of stool from the proximal to distal stoma, will provoke the worst fecal impaction in the distal rectum, the worst megasigmoid, and therefore, the worst constipation. In our series, tapering or not tapering the rectum

did not have any relationship with the degree of constipation that the patient suffers from.

After colostomy closure, proper treatment of the constipation is imperative. When constipation is treated not properly (meaning, to empty the rectum every day), the megasigmoid worsens. More dilatation of the rectum leads to greater constipation and therefore, constipation is a selfperpetuating and self-aggravating condition. The lessons then seem to be very clear; we must try to keep the rectum as empty as possible to avoid rectosigmoid dilatation. Constipation is a very serious problem because in the end, it provokes fecal incontinence, even in patients who were born with a potential for bowel control. This is what I call over-flow pseudoincontinence, The question is open for the future whether or not we should decompress the dilated rectum in utero in those patients with a prenatal diagnosis of imperforate anus.

On the other hand, someone may come up with the apparently brilliant idea of resecting the rectosigmoid during the main treatment of the imperforate anus malforniaticin. This should never be done because it results in fecal incontinence. This occurred in many patients when we were doing abdominoperineal pullthroughs of the endorectal type in which we eliminated the

Specimens from the most distal pan of the rectum and/or fistula in patients with anorectal malformations may show evidence of NID and/or agangliosis. The problem is that there is no way to know the relevance of those findings. We do now know the extension of those abnormalities and the clinical significance of them. Of course, it is very simple for someone to appeal to these findings to try to explain the constipation. To me, it is more likely that the cause of the constipation in these patients is related to a very distended rectosigmoid and therefore, impaired peristalsis. We pediatric surgeons have known for many years that any hollow viscus that becomes over-distended for a long period of time loses its peristaltic ability.

I am very skeptical about the diagnosis of Hirschsprung's disease and imperforate anus. The reason for this is that I have never seen a patient with imperforate anus develop entercolitis, I suspect that many surgeons have been over-diagnosing Hirschsprung's disease and imperforate anus. The concern is not only academic. Many patients are subjected abdominal perineal to pullthroughs simply because the surgeons found one biopsy that was reported as aganglionic. The abdomino-perineal pullthrough will certainly cure the constipation but will also make the patient fecally incontinent 100% of the time, even if he was born with a benign condition.

The treatment method of constipation in these patients depends whether or not the patient has the potential for bowel control. In our Division, we are very obsessive about classifying our patients as to their potential for bowel control, and try to determine this from the very beginning. A poor sacrum, a very high malformation (rectobladderneck fistula or a long common channel cloaca), a

flat bottom, or the presence of a presacral mass usually means that the patient has no potential for bowel control. On the other hand, patients with potential for bowel control include those with a good sacrum, or a benign malformation, such as rectal atresia, perineal fistula, vestibular uistula. imperforate anus with no fistula, or rectourethral-bulbar fistula. We define a poor sacrum as one with more than 3 vertebrae missing, or with a sacral ratio of less than 0.4. The sacral ratio is defined as the ratio of the distance from the bottom of the sacro-iliac joint to the tip of the sacrum divided by the distance from the top of the iliac bone to the bottom of the sacro-iliac ioint.

Patients with poor potential for bowel control and constipation should be treated with an enema every day. The fact that they suffer from hypomotility of the rectosigmoid represents an advantage because it makes the bowel management easier. The emphasis should be on cleaning the colon every day. In order to achieve this, aggressive, large volume, salt water enemas are employed. The program should be implemented over a period of one week by trial and error. Daily abdominal x-rays allow reliable assessment of effectiveness of the enemas. The volume and salt concentration is increased until the patient is effectively cleaned out. This regimen is then continued on a daily basis, and the patients are kept completely clean for 24 hours a day.

Patients with a good potential for bowel control and constipation should be treated with laxatives. We just have to find out the amount of laxatives that each patient needs. We never combine laxatives and enemas because it is illogical. The amount of laxative that each patient needs is frequently very large. The goal once again is to empty the rectum every day. Once bowel control has been achieved with laxatives the patient is maintained on this regimen. If the amount required is enormous, a sigmoid resection may be offered, which will allow a reduction in the amount of laxative requirements.

Tethered cord has occurred in 24% of our patients with anorectal malformations. The more complex and high the malformation, the higher the chances of tethered cord. Forty three percent of our group of complex anorectal malformations have tethered cord, but it is present in only 11% of our patients with rectovestibular fistula. There is no question that the presence of tethered cord is more common in patients with imperforate anus who suffer from fecal incontinence. However, our study could not clarify whether or not the fecal incontinence was due to the presence of the tethered cord. This is because most of those patients had a very complex malformation which alone be responsible for could the fecal incontinence. Eighteen of our patients underwent surgical untethering of the cord and none had any significant change in bowel or urinary function postoperatively. We could not find a single patient with tethered cord who experienced incontinence which could be attributed to the cord defect alone. Surgical untethering may be indicated for other reasons, but at the present time there is no good evidence to demonstrate that it improved the prognosis for bowel control of these patients.



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Presidents Letter

Hello, As always our sincere thanks go out to those of you that are regular contributors to the Pull-thru Network.

We know how hard it is to make time for writing an article because every few months, Karen and I start wondering how we are going to fill another issue of the Pull-thru Network News. Contrary to what some people may think, the PTN is run by the two of us, Dee and Paul Schur, and Cathy and Dave Tague. No staff. No offices. We earn no money for our efforts. And we alt have families and jobs and lives outside of the Pull- thnu Network. So when someone sends us an unsolicited story and/or some photos of their kids, we jump for joy.

Our fear is that in the absence of submissions, we will start spinning out endless "Sammy stories" (our daughter) for lack of other stories. (Besides... we've already bored you with enough Sammy stories already!) We have a pretty steady flow of medical information that constitutes a large segment of our issues but the feedback we get is that the personal stories are what most most people want to read. That means that you have to a little time and put your thoughts down on paper, pop it in an envelope, or even email them directly. The other possibility is that we can cut back the number of issues we do a year. We don't want to do this, but we don't want to mail out newsletters devoid of useful information either. So, Please send up your stories and

help keep the Pull-Thru Network regular.

Scott and Karen Brownlow

Our child was born with impertorate anus, absent sacrum and a neurogenic <u>bladder</u>, and is fecally Incontinent. We have tried to do enemas, but our son hates them and we are unable to find out if they will work. At this time, we are considering a continent appendicostomy. How do we go about determining if this will be a working solution for him?

A patient with no sacrum and a neurogenic bladder, if seen by us, would receive the bowel management to prove that most likely it will work. After that, we will give the patient the choice between continuing the rectal enemas or performing a continent appendicostomy.

We are in favor of improving the quality of life of all children with fecal incontinence. Quality of life should be defined by the patient and his parents. Most patients prefer to receive an enema through the umbilicus while sitting on the toilet rather than through the rectum. However, there is a sub-group of patients that do not want to hear about surgery and, in addition, they are happy with the rectal enemas. Most patients do prefer the continent appendicostomy. On the other hand, our results with bowel management showed that it has been successful in approximately 95% of cases.

My main concern is the fact that continent appendicostomy is а very appealing procedure. Surgeons like to operate. "We should never ask a barber if we need a haircut. It is easier to perform the continent appendicostomy than to implement the management program. Most bowel importantly, the continent appendicostomy does not preclude the necessity to implement the bowel management program. The bowel management program, as published by us, takes a significant amount of time,

dedication and trial and error to find out the best enema for each type of patient. Doing or not doing a cecostomy or a continent appendicostomy does not avoid going through the bowel management.

Therefore, my concern is that a patient might be subjected to an operation only to find out that he is still soiling, If you look at the original publication of Malone presented last year in Turkey, 3896 of his patients were not clean after the Malone procedure (continent appendicostomy). During that meeting, I explained to the audience that it was not the operation that failed but rather the fact that those patients did not receive a successful preoperative bowel management. It was the bowel management (i.e. patient selection) that was failing, not the operation itself.

In addition, the age of the patient is another very important factor. Take for instance the case of a three year old child who is fecally incontinent. He will start attending school and he needs the bowel management. Again, once we proved that the bowel management is successful, we let the family decide about the appendicostomy. However, the fact is that at that age, children have a much greater tolerance for enemas than older children. children benefit from the Most appendicostomy at an age when they want to become independent.

To summarize, I am in favor of the operation. I just want to warn everybody that the operation represents only a different route of administration of an enema. The operation itself does not do anything else except make the administration of the enema easier for the patient.

Cloaca Project

My name is Jenny Halper and I am a 17 year old high school senior attending the Packer Collegiate Institute in Brooklyn, New York. I was born with a cloacal malformation. As my senior thesis project I am working on a book dealing with the experiences of children growing up with cloaca anomalies. I am interested in speaking to parents and children born with this birth defect. My book will address issues regarding experiences in school, camp and hospital situations. Information regarding identity will be kept confidential. I hope the information that I compile will be helpful and informative to other children growing up with this anomaly.

I can be contacted at (718) 768-0414. or send email to HalperCaol.com.

Thanks for your help!

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We would like to thank the following people for their generous support of the Pull-Thru Network.

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