

BOWEL MANAGEMENT FOR PATIENTS WITH FECAL INCONTINENCE

QUESTION #1: Under the best circumstances, the global results following the surgical treatment of anorectal malformations are:

- A) 75% chance of fecal incontinence.
- B) 50% chance of fecal incontinence.
- C) 25% chance of fecal incontinence.
- D) 5% chance of fecal incontinence.
- E) No chance of fecal incontinence

The correct answer is (C) 25% chance of fecal incontinence. This information is based on the followup of a large series of patients over a period of 20 years. (1) The 75% chance of bowel control and 25% of fecal incontinence represents a global figure. Anorectal malformations are represented by a spectrum of defects. Each type of malformation has a different prognosis. For instance, patients with an anorectal malformation called a perineal fistula have 100% chance of bowel control when the patient receives a good operation. Vestibular fistula have a 95% chance of bowel control. Cloacas with common channels shorter than 3 cm, 85%. Cloacas with a common channel longer than 3 cm, approximately 60%. Male patients with rectourethral bulbar fistula, 85%. Imperforate anus with no fistula, 90%, when patients have no Down syndrome. In patients with Down syndrome, the chances are 80% (2). Male patients with rectourethral prostatic fistula, 60% and those with rectobladder neck fistula, 20% chances of bowel control. These percentages also vary depending on the characteristics of the sacrum. We have never seen a patient with bowel control with a sacral ratio lower than 0.4.

All pediatric surgeons are morally obligated to follow all patients operated on with anorectal malformations and take care of their functional sequelae. We believe that all patients born with anorectal malformations, once they are three years old must remain completely clean in the underwear, either because they belong to the good prognosis types of malformations or because we keep them absolutely clean by the implementation of a bowel management program, which is an artificial way to keep them clean.

QUESTION #2: Which one of the following conditions represents a formal contraindication for a pull through or repair of an anorectal malformation:

- A) Myelomeningocele.
- B) Absent sacrum.
- C) Presacral mass.
- D) Absent colon.
- E) None of the above.

The answer is (D) absent colon. It is a common belief that when patients have no potential for bowel control (myelomeningocele, absent sacrum, multiple hemivertebra, absent sphincters) it is not indicated to do the pull through because the patient will suffer from fecal incontinence, which will give him/her a poor quality of life. We believe that all those patients without potential for bowel control can and must be subjected to a pull through provided the surgeons or the team that is taking care of the patient, provides and implements a good bowel management program, which will keep the patient completely clean 95% of the time and will give the patient a much better quality of life than a permanent colostomy. An absent colon or an almost absent colon, prevents the patient from forming solid stool. Incapacity to form solid stool represents a formal contraindication for a pull through, simply because the bowel management cannot be implemented in a patient with constant diarrhea.

That is the reason why the only formal contraindication for a pull through that we recognize at the present time is the absence of colon, almost absent colon, or incapacity to form solid stool from any cause.

The bowel management program, consists of finding the way to clean the colon of the patient once a day, and maintain the colon with minimal motility or paralyzed to prevent the patient from passing stool in between enemas. To clean the colon, we can use enemas with different volumes, different solutions, and different concentrations depending on the specific type of patient and type of colon. Patients who suffer from constipation, by definition suffer from hypomotility of the colon and therefore, we do not have to do anything to keep the colon quiet and/or paralyzed. The patient will stay clean once we find the enema that is capable of cleaning his or her colon. (3)

Patients who suffer from a tendency to diarrhea are more difficult to manage. To clean the colon in this group is an easy task, but to keep the colon paralyzed or moving very slowly is difficult. In order to slow down the motility of the colon, we use a constipating diet and medication such as Imodium

Enemas, constipating diet, laxatives and medications to decrease the colonic motility, have been used widely in the past to treat fecal incontinence, with poor results. The bowel management discussed here does not include any new therapeutic agents. Yet, we believe that the key for success resides in the use of the same therapeutic agents but following a well defined rational plan.

QUESTION #3: The best way to find out if the colon of the patient is clean after the administration of an enema is by:

- A) Measuring the amount of stool that came out.
- B) Palpating the abdomen.
- C) Rectal examination.
- D) Taking an abdominal x-ray film.
- E) None of the above.

The correct answer is (D), taking an x-ray film of the abdomen. Measuring or weighing the amount of stool that comes out through the rectum does not guarantee that all the stool came out; in constipated patients the amount of stool that the patient carries inside, sometimes is much greater than predicted. Palpation of the abdomen frequently does not allow the observer to determine the amount of stool that is in the colon, and the rectal examination allows one to know you if the rectum is empty, but the clinician does not know if there is stool in the rest of the colon. An abdominal x-ray is the only definitive way of knowing whether or not the colon is empty.

An x-ray film is the only objective way to know if the colon is clean or not.

QUESTION #4: The best way to determine the volume of the enema required for a specific patient is:

- A) By looking at the contrast enema and calculating the area of the colon multiplied by the body weight in kilograms, the result should be interpreted in cc.
- B) Simply by administering 25 cc of solution per kg of body weight.
- C) By administering 50 cc of solution per kilo of body weight.
- D) By trial and error.
- E) None of the above.

We have found that most formulas fail. It is not easy to determine the volume of the enema that a patient requires; therefore, we believe that the best way to do it is by trial and error (d). Obviously, the clinician looks at the contrast enema and if the patient has a giant megasigmoid, he can assume that the patient will need a very large volume type of enema. Subsequently, over a period of five days seeing the patient everyday, evaluating the result of the prescribed enema, and taking x-ray films every day, one can finally determine which enema is capable of keeping the colon completely clean.

QUESTION #5: The concentration of salt and phosphate in the enemas used in bowel management should be determined by;

- A) The age of the patient.
- B) By trial and error depending on the patient's response and avoiding toxic levels
- C) The characteristics of the colon as seen in the contrast enema.
- D) The body surface
- E) None of the above.

The answer is (b). The concentration of sodium chloride and (or) phosphate (fleet) must be adjusted by trial and error, and being sure not to reach toxic levels. High concentration of sodium chloride and (or) phosphate will produce a faster response (bowel movement). Colicky pain as well as manifestations of a vagal response (paleness, nausea and vomiting) are manifestations of excessive concentration. Symptoms of dehydration and or lethargy, are also manifestations of too much salt and (or) phosphate. The safest concentration of sodium chloride, as expected, is that of normal saline (). Higher concentrations are rarely necessary and must be used carefully to avoid the risk of hypernatremia. The commercially available phosphate enema (fleet) is sold in a pediatric form (60 cc of%) and an adult presentation (120 cc of% solution). From two to four years we recommend no to administer more than ½ of the pediatric fleet (30 cc) in 24 hours. From 4-10 years the limit is one pediatric fleet (60 cc) per 24 hours. Over 10 years, the patient may receive an adult fleet (120 cc) every 24 hours. Higher amounts of phosphate exposes the patient to suffer hypocalcemia

QUESTION #6: When a patient suffers from fecal and urinary incontinence the best course of action is;

- A) To treat first the problem of urinary incontinence followed by the management of fecal incontinence.
- B) To take care of the fecal incontinence problem first and then evaluate the urinary tract.
- C) To take care of both problems at the same time.
- D) To offer the patient a permanent colostomy.
- E) To try a course of biofeedback prior to the bowel management.

Our recommendation is to first take care of the fecal incontinence problem and once the patient is completely clean of stool in the underwear, reevaluate the problem of urinary incontinence (b). Twenty patients complaining of fecal and urinary incontinence were treated first for the problem of fecal incontinence and 19 of them turned out to be urinary continent, only one suffered from real urinary incontinence. This occurs basically because when children grow up using a diaper, they become accustomed to passing urine in the diaper, and they do not feel pressure to become toilet trained.

QUESTION #7: A patient suffers from fecal incontinence, he comes for consultation and the decision is taken to implement the bowel management program. The patient heard about the possibility of administering the enemas through the umbilicus or through a little orifice in the abdominal wall connected to his appendix, a

procedure that is called continent appendicostomy, also known as MACE or Malone procedure. The best course of action is;

- A) To perform the operation and one month after that to start the bowel management.
- B) To perform the operation and three months later to start the bowel management.
- C) To start the bowel management and six months later to try the surgical procedure.
- D) To try the bowel management and only when we demonstrated that the bowel management works, to offer the patient the operation.
- E) None of the above.

It is our philosophy that the continent appendicostomy or any other surgical procedure designed to administer antegrade enemas, should never be done prior to the successful implementation of the bowel management. The implementation of the bowel management requires a significant amount of time, dedication, and an authentic interest in the benefit of the patient. In fact, it requires much more time and work than the operation. There is no demonstration that any of these operations makes the bowel management more efficient. All those surgical procedures are excellent ways to improve the quality of life of these patients they become more independent because they can administer the enema themselves while sitting in the toilet alone. In other words, these operations only represent the creation of another route of administration of an enema. If the bowel management implemented through the rectum does not work, the Malone procedure is going to be useless. There is a significant number of cases that have been operated on first and then they try to implement the bowel management unsuccessfully. Under those circumstances the operation actually was useless. In addition, it gives the impression that the surgeons are taking the easy route of rushing into an operation in order to give antegrade enemas, but they are not interested in implementing the bowel management, which is the most difficult part.

QUESTION #8: In dealing with a patient who suffers from fecal incontinence consecutive to an operation performed for the repair of an anorectal malformation, the most important step is to determine;

- A) If the problem of the patient resides mainly in a nerve injury.
- B) If the patient suffers in addition to the fecal incontinence from constipation or tendency to diarrhea.
- C) If the relaxation reflex is present as demonstrated by a rectal manometry.
- D) If the muscle complex is intact as shown by intraluminal ultrasound.
- E) If the latency period is prolonged in a pudendal nerve evaluation.

The most important step in the evaluation of these patients, prior to the initiation of the bowel management is to determine if the patient suffers from constipation or a tendency to diarrhea in addition to the fecal incontinence (b). The treatment in each one of these two groups is completely different. In the group of patients who suffer from constipation, the key for success is to find out, by trial and error, the volume and concentration of the enema capable of cleaning the entire very dilated megasigmoid that the patient has. Once this has been determined most likely the patient will remain completely clean, regardless of the diet that he follows and with no need of medications because the rectosigmoid of the patient, by definition, suffers from hypomotility.

On the other hand, if the patient belongs to the group of "tendency to diarrhea", most of the times it is because he was subjected to a resection of the rectosigmoid or resection of another big part of the colon which makes him sensitive to different kinds of food, and to suffer from a tendency to have diarrhea. The colon in these patients is not dilated. As a consequence, it is very easy to clean the colon with a relatively small volume, and low concentration type of enema. The main problem, however, is to keep that colon quiet in the period of time in between enemas. For that, the clinician has to order a very specific constipating diet as well as medication

to slow down the motility of the colon. The treatment of these patients with increased motility (tendency to diarrhea) is much more complicated and not as successful as in the group of constipated patients.

A simple contrast enema shows very clearly the difference between those two groups (constipated and tendency to diarrhea). The history, of course contributes to an idea as to which type of patient are we dealing with.

The presence or absence of a relaxation reflex is of no relevance, there is no real correlation between the presence or absence of reflex and the degree of bowel control that patients have. The intraluminal ultrasound does not help in the study of these patients or the study of the pudendal nerve function.

QUESTION #9: The combination of enemas and laxatives is indicated;

- A) In all patients who suffer from fecal incontinence.
- B) In the group of incontinent constipated patients.
- C) In cases of fecal incontinence consecutive to spina bifida.
- D) In patients who suffer from fecal incontinence consecutive to an operation for Hirschsprung's disease.
- E) None of the above.

We almost never recommend the combined use of enemas and laxatives. The answer therefore, is (e). The combination of enemas and laxatives does not make sense; the enema will clean the colon and the laxatives will promote the passing of stool in between enemas. Unfortunately, we have seen many patients who come to us after being subjected to failed attempts of bowel management in which the doctors were combining laxatives with enemas.

Very occasionally, we have patients who suffer from a very severe megasigmoid and a tendency to become fecally impacted with hard stool that is difficult to mobilize with an enema. In those cases sometimes we recommend the use of small amount of stool softeners associated to the enemas.

When patients with Hirschsprung's disease suffer from fecal incontinence, they most frequently suffer from tendency to diarrhea. It is unusual to see patients suffering from Hirschsprung's disease constipated and fecal incontinence, although everything is possible.

QUESTION #10: The best way to administer an enema for the management of fecal incontinence is;

- A) With a large Foley catheter with the balloon inflated.
- B) With a large rubber tube introduced as much as possible.
- C) With a rubber tube introduced no more than 5 cm.
- D) With the patient in supine position.
- E) None of the above.

Each patient needs a different type of enema and each patient requires a different technique of application of the enema depending on the specific circumstances. Therefore, the answer is (e).

The technique for the administration of an enema also depends on the age of the patient. Very small children, for instance, can receive an enema while positioned on the mother's lap with the head towards the floor and the bottom up. School age children sometimes receive the enema being face down with the pelvis elevated in order to take advantage of the gravity to be sure that the enema fluid goes up into the colon. Older patients prefer the enema to be given while sitting on the toilet directly. However, for obvious reasons, the inconvenience of this technique is that the fluid of the enema (by gravity) comes out into the toilet and does not

work. In order to avoid that, sometimes it is necessary to use a large 30 cc balloon type of Foley catheter to inflate the balloon to avoid the leakage of fluid into the toilet.

When patients are fecally impacted, sometimes they require a high rectal irrigation. In other words, the fluid of the enema must be delivered in the colon, above the impacted fecal matter. For this, sometimes it is necessary to introduce a large tube, up to the descending colon. Most patients, however, do not need a large tube introduced in the rectum to deliver an enema.

Patients with spina bifida do not have a normal sphincter, they rather have a wide rectal opening; Therefore, they frequently need a catheter with a big balloon inflated in order to allow the introduction of the fluid administered under hydrostatic pressure to avoid leakage.

The technician, nurse, or doctor who is guiding the patient about the administration of the enema must be prepared to adapt the technique to the specific clinical circumstances.

QUESTION #11: It is important when indicating an enema;

- A) To provide written instructions to the mother to administer the enema correctly at home.
- B) To demonstrate practically, the way to give the enema as an outpatient.
- C) To admit the patient to the hospital.
- D) To sedate the patient.
- E) None of the above.

The answer is (b). It is extremely important for the nurse, the technician, or the doctor, to administer the enema to the patient with the parents. This is the only way that the doctor or the nurse can gain a moral authority. One of the most common errors in the implementation of the bowel management is to provide oral or written instructions to the parents and assume that they are going to do a good job at home. The parents frequently come to the clinic and complain about the fact that the child refuses to have the enema and that it is impossible to give it. Under those circumstances, the health provider may simply assume that the parents are ignorant or negligent. The only way to clarify this and again, to gain moral authority is by giving the enema himself or herself. The second day, the mother should give the enema in front of the health provider in order to be sure that she is doing it correctly.

It is amazing to observe how many parents do not know what an enema is. Many of them believe, for instance, that there is a single straight tube between the mouth and the rectum. Other parents give an enema while the fluid of the enema is leaking constantly during the administration. They may think that this is normal, while they are actually not giving an enema. All this means is that there is no substitute to the "hands on" way to instruct the parents.

If the health provider cannot convince the child to accept the enema or cannot give an efficient enema, he or she cannot expect that the parents will be able to do it. In some countries, it is possible to bring the patients to the hospital as inpatients, which allows the doctors and nurses to have a better control of the events. In the United States, on the other hand, because of the cost limitation

and pressure from the insurance companies, it is impossible to do that, and we have to do it as outpatients.

QUESTION #12: The continent appendicostomy or any other variant of procedure administer antegrade enemas are indicated:

- A) Between the ages of 3-6 years.

- B) Before 3 years of age.
- C) Between the ages of 6-9.
- D) After 12 years of age.
- E) When the patient asks for it.

The answer is (E). These procedures are indicated to improve the quality of life of the patient. We believe that the only person that can define quality of life, is the patient herself/himself. Sometimes, parents request one of these operations to be done when the patient is extremely young, for instance 4 years old. In that particular case, the parents are assuming that that is good for the patient, but the patient is not participating in that kind of decision. If the patient does not cooperate for the administration of an enema through the rectum (which should never be painful) he may not participate for the administration of the enema through an orifice in the abdomen.

Continent appendicostomies or other variants are particularly accepted and beneficial when the patients want to become independent, usually that happens after 10 years of age, when the patient starts feeling embarrassed while receiving an enema given by the parents.

Sometimes, of course, the patient requests himself or herself the operation at a much younger age, and they understand what it entails. We allow the patients to have a dialogue with other children of the same age who have had the same operation so they can ask questions and learn what they are getting into when they receive this operation.

QUESTION #13: A 10-year-old male patient comes to the clinic suffering from fecal and urinary incontinence as well as severe constipation. Taking the history of the patient one learns that he was subjected to some sort of perineal or sacral operation when he was a newborn or in the first month of life. The parents cannot be specific as to what type of operation the patient was subjected to and they are unable to obtain medical records from that procedure.

A set of diagnostic studies are indicated for the initial evaluation, which show that he has a completely normal sacrum, normal lumbar spine and normal kidneys. A contrast enema done with hydrosoluble material shows that he has a very severe megarectosigmoid. The examination of the patient shows that he has what is known as "good looking perineum", meaning that he has a midline groove in a well located rectum within the limits of what seems to be a good sphincter. Rectal examination shows no evidence of stricture, but it provides evidence that the patient suffers from fecal impaction. For this patient, the best course of action is;

- A) To offer the patient a gracilis muscle operation to improve his bowel control.
- B) To subject the patient to a biofeedback training for three months and ask him to come back to the clinic.
- C) To rule out overflow pseudo-incontinence.
- D) To initiate the bowel management program with the administration of large enemas and no specific diet.
- E) To start the implementation of the bowel management with the use of large enemas and administration of laxatives.

The answer in this case is (C), to rule out overflow pseudo-incontinence. A small proportion of patients that were subjected to an operation for the treatment of an anorectal malformation come to the clinic suffering from what the doctors believe is fecal incontinence and when they are studied meticulously one actually finds out that they actually suffer from what we call overflow pseudo-incontinence. This is a group that may be improved dramatically without an operation and without bowel management actually.

Overflow pseudo-incontinence must be suspected in patients that have a series of characteristics that makes us believe that they were born with a type of malformation that is rather benign, meaning a malformation with a good functional prognosis.

The fact that this patient has a normal sacrum, normal lumbar spine, normal kidneys, good looking perineum, and a well-located rectum means that most likely he belongs to the "good side" of the spectrum of anorectal malformations. In addition, we can assume that he was subjected to a technically correct operation and therefore one would expect that this patient should have bowel control and yet he behaves like incontinent. The contrast enema shows that the patient suffers from severe megarectosigmoid and fecal impaction.

These patients behave like the encopretic (otherwise) normal child suffering from severe constipation that soils the underwear day and night. The treatment in both groups of patients is the same. We follow a specific protocol that includes:

1. To disimpact the patient with enemas
2. To administer what we call a "laxative test." This consists in the administration of increasing amount of laxatives until we reach the amount that is capable of making the patient empty the colon completely everyday as demonstrated by abdominal x-ray film. At that point, we observe the patient and see if he is really incontinent or has bowel control. If the patient recovers bowel control under this management, means that he actually suffered from overflow pseudo-incontinence. It is very rewarding and satisfactory to deal with this particular group of patients because the doctor can really make an enormous difference in the quality of life of the patient. Unfortunately, many of these patients are subjected to inadequate treatments including operations to "improve" bowel control. Obviously, those operations worsen the patient's condition.

When we reach the indicated amount of laxatives, as determined by an x-ray film of the abdomen that shows a clean colon, and if the patient is fecally incontinent, then she/he must be subjected to the implementation of the bowel management program, enema and no specific diet.

On the other hand if we demonstrate that the patient is continent and suffered only from overflow pseudo incontinence, the management will then consist of the administration of laxatives. The amount of laxatives that these patients frequently require is much higher than what most clinicians believe and certainly we should not be guided by the books, because the patients require much higher amounts of laxatives.

QUESTION #14) The same patient suffering from overflow pseudo-incontinence mentioned in our previous question is now very happy as well as the family, because of the results of the management. The only problem is that the patient has to take an enormous amount of laxative every day, which is extremely troublesome, gives him nausea and sometimes vomiting. What we have to offer this patient is:

- A) To tell him that there is no other choice except to take those laxatives for life.
- B) To subject the patient to a rectal myectomy to eliminate the action of the internal sphincter.
- C) To administer laxatives for six months followed by a course of biofeedback and by doing that most likely he would not need laxatives anymore.
- D) To explain to the family that another alternative to the administration of laxatives is an operation called sigmoid resection consisting in removing the most dilated part of the colon and anastomosing the descending colon to the rectum.
- E) To offer the family an abdominoperineal resection and a pull-through of the non-dilated part of colon as when we deal with a patient with Hirschsprung's disease.

The answer is (D). The constipation that these patients suffer from is usually incurable, but manageable. The patients must take the laxatives for life with a few exceptions. Sometimes, when these patients reach adolescence, they develop some sensitivity to specific types of food and the administration of that kind of food allows them to avoid laxatives. However, that is rather unusual.

The myectomy is an operation that is frequently performed with no scientific basis and certainly with a significant number of complications.

There is no evidence that biofeedback helps these patients.

The sigmoid resection performed in a series of patients certainly does not cure this condition, but improves the quality of life, because it allows the patient to reduce significantly the amount of laxatives that they have to take everyday. However, it is extremely important to offer this operation only after we have demonstrated that the patient has bowel control. To perform the sigmoid resection without having demonstrated this, may change the situation of a patient who suffers from constipation and fecal incontinence into a patient with fecal incontinence and tendency to diarrhea, which is much more difficult to manage.

Unfortunately, we have seen patients that have been subjected to abdominoperineal pull-through type of operations with or without demonstration of aganglionosis. Certainly, an abdominoperineal resection and pull-through, radically cures the problem of constipation, but leaves these patients completely incontinent. An abdominoperineal resection and pull-through in a patient with Hirschsprung's disease without an anorectal malformation does not suppose to provoke fecal incontinence because those individuals have a normal sphincter mechanism, normal anal canal, therefore, normal sensation; and if the surgeon performs a technically correct operation preserving the anal canal and the sphincter mechanism, the patients preserve bowel control. It is important to remember that patients with anorectal malformations are born without anal canal and with different degrees of deficient sphincter mechanisms and therefore that kind of pull-through will make the patient totally incontinent.

It is true that Hirschsprung's and anorectal malformation may occur together but we do not have evidence that is more common for a patient with an anorectal malformation to suffer from Hirschsprung's disease as compared to the general population. It is also true that patients suffering from anorectal malformation, after a pull-through, may have absent ganglion cells in some parts of the rectum that was pulled down, but that does not mean that the patient suffers from Hirschsprung's disease. Patients with Hirschsprung's disease suffer from many other problems besides constipation. They also have other histologic abnormalities in addition to absent ganglion cells.

QUESTION #15) The same patient that we suspect that suffers from pseudo-incontinence described in the previous two questions, accepts the management, which must consist in:

- A) Administration of GoLYTELY for 24 hours followed by the administration of laxatives.
- B) Administration of GoLYTELY for 4 hours followed by an enema and laxatives the following days.
- C) Disimpact with enemas with radiologic demonstration followed by the administration of laxatives.
- D) Fecal disimpaction under anesthesia followed by the administration of laxatives.
- E) None of the above.

We recommend that these patients should be disimpacted by the administration of three enemas everyday. One of the enemas contains a phosphate solution and saline solution. The other two enemas include only saline solution, because it is contraindicated to give more phosphate enema in a child everyday. Giving more phosphate enema exposes the patient to suffer severe hypocalcemia.

If after three days of the administration of three enemas for the treatment of fecal impaction, the x-ray film still shows the presence of severe impaction, the patient is brought to the hospital to continue with the administration of enemas, but in addition to the administration of GoLYTELY for 48 hours. If after that time, the patient is still impacted and only under those circumstances, we subject the patients toward fecal disimpaction under anesthesia. The administration of GoLYTELY and/or laxatives in a patient that is fecally impacted will provoke most likely a severe colicky pain and sometimes vomitus and manifestations of intestinal obstruction. We try not to administer laxatives unless we are certain, as demonstrated by an x-ray film, that the patient is already disimpacted.

Question #16) An 11-year-old male child comes to the clinic with a history of anorectal malformation operated through a sacroperineal procedure in a good institution by a good surgeon. Reading the operative note, makes us believe that the patient received a good operation. The evaluation of the patient shows that his sacrum has a couple of hemivertebrae. In addition, there are hemivertebrae in the lumbar spine. The left kidney is absent. The MRI of the pelvis shows that the rectum is well located. The patient suffers from important megarectosigmoid and physical examination shows that there is no evidence of anal stricture and the patient has a moderately flat perineum. In other words, there is no significant midline groove and no evidence of a good sphincter. The best course of action in this particular case is:

- A) Initiate the implementation of the bowel management with the administration of small enemas.
- B) Implement the bowel management with the administration of large enemas and no diet.
- C) Rule out the possibility of overflow pseudo-incontinence.
- D) To perform a rectal biopsy to rule out the possibility of Hirschsprung's disease.
- E) To take care of the urinary incontinence problem first and then to continue with the bowel management.

This patient most likely suffers from real fecal incontinence and constipation. We can say that based on the fact that he has a very abnormal sacrum. Sacral hemivertebrae are not a good sign in terms of functional prognosis. In addition, he has lumbar hemivertebrae and an absent kidney. Absent kidneys are the most common urologic associated defects in anorectal malformations, but in general is much more common in patients that are born with bad malformations. The patient has a mega rectosigmoid, which indicates that he suffers from constipation in addition to the fecal incontinence. The flat perineum confirms our suspicion that this patient was born with a bad defect. Therefore, we do not think that this is justified to suspect that the patient may suffer from overflow pseudo-incontinence. Therefore, there is no indication for a rectal biopsy. The patient must be initiated right away into a bowel management program with the administration of large enemas, since the patient has mega sigmoid, and no need for the administration of diet because the patient suffers from constipation. As we have mentioned before, we should always take care first of the fecal incontinence problem and chances are that after that the patient finds out that he has urinary control, except that he has been peeing in the diaper all his life because it was convenient for him.

Question #17) A 13-year-old boy comes to the clinic with a history of being born with an anorectal malformation. At birth, he underwent the opening of a colostomy and, when he was 1-year-old, he was subjected to an abdominoperineal endorectal pull-through. He suffers from fecal and urinary incontinence and comes for consultation.

Evaluation of the patient shows that he has a sacral ratio of 0.4. His lumbar spine is normal. He has normal kidneys and a contrast enema shows that there is no dilatation of the colon; in fact, the colon runs in a straight line from the splenic flexure all the way down to the pelvis, showing colonic haustrations down in the pelvis. The best course of action in this patient is:

- A) Implementation of a bowel management with the use of small enemas.

- B) Implementation of the bowel management with the administration of small saline enemas plus constipating diet and the administration of a drug to decrease the colonic motility.
- C) Implementation of the bowel management with the administration of large enemas and no diet.
- D) Initiation of a constipating diet plus Imodium only.
- E) Administration of large volume enemas with saline solution plus phosphate.

This patient obviously was born with a bad type of malformation, since he has a very short sacrum (sacral ratio is 0.4). That means that he is really incontinent, most likely. In addition, it is obvious that he was subjected to an abdominoperineal endorectal pull-through, which by definition means that he lost his rectosigmoid. Endorectal dissection sacrifices the rectosigmoid and that makes it mandatory to pull the descending colon all the way down to the perineum. This is demonstrated because the contrast enema shows us a non-dilated colon running straight from the splenic flexure down to the pelvis and also because we can see haustrations of the colon down in the pelvis. The colon in these patients can be cleaned relatively easily with a saline solution. In general, we do not recommend the use of phosphate enemas in these kinds of patients because the phosphate will make the hypermotility of the colon worse and would make it more difficult for the patient to stop passing stools in between enemas. So, since we are limited by time, it is better for these patients to receive small saline enemas, which most likely will clean the colon completely, and the patients should receive a constipating diet and a drug to decrease the motility of the colon such as Imodium. Once we demonstrate that we have been successful with this kind of management, we can gradually liberalize the constipating diet, reintroducing the type of food that the patient misses the most in order to detect the real offenders from the list of the prohibited-type of food for these patients. Eventually, if the patient remains clean, we can gradually decrease the amount of Imodium to see what is the minimal amount of Imodium that the patient needs to remain clean. Sometimes we find that actually the patient does not need Imodium and the patient can do well even with a normal diet.

Question #18) A 15-year-old boy comes to the clinic complaining of fecal and urinary incontinence. He was born with an anorectal malformation and was operated when he was 3 months old with an abdominoperineal-type of operation. He does not know the details of the procedure, and there is no operative report available. The sacral ratio in this patient is 0.5. The contrast enema shows a normal looking sigmoid. The kidneys are normal, as well as the lumbar spine. Physical examination shows a moderate tendency to a flat bottom. The main course of action in this particular patient is to:

- A) Implement a bowel management program with the administration of very large enemas plus phosphate.
- B) To implement a bowel management consisting in the administration of enemas plus a constipating diet.
- C) To suspect and rule out overflow pseudo-incontinence.
- D) The implementation of bowel management with the use of moderate-sized saline enemas nor diet and no Imodium.
- E) None of the above.

This patient most likely will benefit with the administration of a moderate-sized saline enema and no need for diet nor Imodium. A phosphate enema is rather an irritant solution that is necessary to be used in cases of significant sigmoid and megasigmoid.

There is no need for large enemas in this patient, because his colon is not dilated. There is no need for a constipating diet, because he does not suffer from absent rectosigmoid, and we do not see haustrations of the colon in the pelvis.

There is no justification to suspect overflow pseudo-incontinence, because the patient seems to have been born with a type of malformation with some chances of bowel control but not very good, as demonstrated by the sacral ratio of 0.5 and a moderate flat bottom.

Question #19) The same patient that we described in the previous question, one week later comes back very happy because now he is completely clean of stool in the underwear following the bowel management that we prescribed. We then decided that to evaluate the urinary incontinence that he suffers from. We specifically suggest to the family and the patient to stop using diapers, to urinate every three hours, and report to us in five days. The patient comes back and states that he has been able to urinate voluntarily every three hours. However, in between voiding episodes, he dribbled urine and sometimes he passes significant amount of mucus through the urethra. Under these circumstances, we must suspect that the patient suffers from:

- A) Overflow due to incapacity to empty the bladder.
- B) A urethrocele interfering with the closure of the bladder neck.
- C) A posterior urethral diverticulum.
- D) A hypotonic detrusor with uninhibited contractions.
- E) None of the above.

The answer is that we must suspect the presence of a posterior urethral diverticulum. This actually represents a piece of rectum that was originally left attached to the urethra. This patient most likely was born with a rectourethral vulva fistula. In that particular type of case, the rectum is originally and congenitally connected to the lowest part of the posterior urethra, what is known as vulva urethra. The patient was subjected to an abdominoperineal procedure when he was a baby. In those years, many surgeons divided the anorectal malformations into only two categories (low and high). The low malformations were operated perineally and the so-called "high malformations" were operated abdominoperineally.

When the patients had a rectourethral vulva fistula, it was frequently impossible for the surgeon to reach the end of the rectum through the abdomen, and this resulted in the amputation of the rectum, leaving a piece of rectum attached to the posterior urethra. The patients then grow up relatively asymptomatic. However, when they pass urine through the urethra, part of the urine comes out of the urethra and the other part goes into the diverticulum that has a tendency to grow. When the patient finishes voiding, walks around dribbling urine that comes from the diverticulum. The diverticulum is usually connected to the urethra below is so-called external urinary sphincter and that is why it dribbles urine. The fact that the patient passes mucus in between voiding episodes, justifies our suspicion.

Overflow due to incapacity to empty the bladder may occur particularly in patients that have a very abnormal sacrum and had an inadequate operation that denervated the bladder.

A urethrocele, this is an extremely unusual malformation associated to anorectal malformations.

A hypertonic detrusor with uninhibited contractions may occasionally occur in patients with anorectal malformations, but it is extremely unusual. Patients with anorectal malformations who suffer from neurogenic bladder most frequently have rather smooth bladder, but not a hypotonic bladder.

To confirm the presence of the posterior urethral diverticulum, one may perform a voiding cystourethrogram, which may or may not note the diverticulum. Then, a magnetic resonance imaging study of the pelvis will confirm the presence of the diverticulum. We have seen diverticula even larger than the urinary bladder. These patients require an operation to resect the diverticulum and the patients dramatically recover total urinary control when the operation is done correctly.

