



International Foundation for
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Disorders of Defecation in Children: What is the Role of the Surgeon?

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INTRODUCTION

When a new baby is born, we assume that he or she will spend most of the first few months of life engaged in a small number of basic activities: sleeping, feeding, peeing, and pooping. The passage of waste is a basic function of all living organisms, which is so ingrained and routine that most of us do not even think twice about it. In particular, defecation (the passage of stool) is considered to be a normal part of being a human being.

Unfortunately, there are some children who may have difficulties with the normal passage of stool, either because of physical or physiological abnormalities. In severe cases, failure to pass feces may result in intestinal obstruction, which can be fatal if it is not recognized and treated. In milder cases, these disorders may become the source of embarrassment, discomfort, and stress due to lack of social acceptance. Despite this, and the fact that these conditions consume significant medical resources, there has been little research done about the basis and treatment of defecatory disorders in children.

Normal defecation requires several processes to work properly. First, both the internal and external anal sphincters [muscles in the pelvic floor] must function normally. The internal sphincter works automatically through reflexes, and is not under the individual's conscious control. The external sphincter, on the other hand, is under voluntary control, and is tightened at will. This is the mechanism that we use when we have the urge to move our bowels, but need to "hold it in" until we get to a bathroom. Second, the patient must have normal sensation. This involves being able to tell when the rectum is full, and also to differentiate gas from stool so that we can safely pass gas but not stool at the same time. Third, the patient must have normal "motility" in the colon (the movement of food through the digestive tract). Fourth, there must be no obstruction or blockage to the movement of stool. Defecatory problems can be broadly classified into inability to stool or inability to control stooling. Most defecatory problems arise from abnormalities of one or more of the mechanisms described above.

Children may suffer from a number of birth defects that interfere with defecation. In addition, the fact that control of defecation is a normal developmental process during childhood makes the diagnosis and management of these problems particularly difficult and challenging. Although the majority of children with defecatory disorders can be successfully treated with medical, dietary, and behavioral approaches, a surgeon can sometimes be extremely helpful or even provide a cure for the problem. This article will deal with surgical approaches to children who are unable to pass stool, and those who are unable to control the passage of stool.

THE INABILITY TO PASS STOOL

When There is Something Wrong with the Anatomy of the Colon, Rectum or Anus

Some children are born with abnormalities of the anus or rectum, which lead to obstruction of the normal flow of stool. Although anorectal malformations represent a wide spectrum of conditions, we generally think of them as either "low" or "high" varieties based on where the rectum ends. In low anorectal malformations, the rectum exits onto the skin, usually in front of the normal position of the anus. In some cases the location is normal, but the opening is narrowed or covered by a thin membrane. In children with a high anorectal malformation, the rectum either ends blindly in the pelvis, or forms a connection to the urethra (in a boy) or the vagina (in a girl). These types of anorectal malformations are often referred to as "imperforate anus."

Children with low anorectal malformations usually present with difficult passage of stool, although sometimes the opening is large enough that there is a delay in the recognition of the problem for weeks or even months. These children require surgical management, but there are a number of possible options. Children with mild forms can be managed using dilation of the anus, and often nothing further needs to be done. In more severe cases, the opening must be enlarged surgically, and positioned within the superficial anal sphincter mechanism. A number of options are available for this, including the cut-back anoplasty, anal transposition, and anal transplantation procedures. All three operations are designed to reposition the anal opening to its normal location at the center of the anal sphincter, and all three can be done from below without an abdominal incision or a colostomy. These operations can typically be done in the newborn period. Children with low anorectal anomalies usually have normal continence, although many of them develop long-term problems with constipation. The reason for the constipation is unclear, but it is important to ensure that there is no narrowing, which might be improved by surgery.

High anorectal anomalies present a more challenging problem. The goal of surgery for these children is to disconnect the rectum from the urethra or vagina, and to bring it down to the skin to create a new anus. In order to maximize the chance for normal continence, it is important to bring the rectum through the normal sphincter mechanism. Historically, these infants were treated by an abdominal operation, but more recently posterior sagittal anorectoplasty (PSARP), popularized by Dr. Alberto Pena, has become the most commonly performed operation. This operation is done from the back and does not involve any incisions in the abdomen.

In most cases, the child requires a colostomy in the newborn period as soon as the diagnosis is made. This operation permits the baby to pass stool into a bag on the

front of the abdomen, so that he or she can eat normally and grow to a size that is more favorable for the definitive reconstruction (usually several months later). In nearly all cases the colostomy is then closed as a third stage. Recently a number of authors have reported a definitive single stage repair without colostomy in newborns. Another innovative approach has been the recent application of laparoscopic surgery to infants with high anorectal anomalies.

Most children with high anorectal anomalies do not have normal continence. The reasons for this include poorly developed pelvic muscles as a part of the birth defect, placement of the muscle outside the sphincter during the surgery, inadequate sensation, and associated abnormalities of the nerve supply to the rectum. Many also have significant constipation following surgery, often associated with massive dilatation of the sigmoid colon. In patients with this problem who have some degree of continence, surgically removing the dilated bowel may be beneficial.

When There is Something Wrong with the Function of the Colon, Rectum or Anus

Hirschsprung's Disease. Children with Hirschsprung's disease are missing the nerve cells ("ganglion cells") within the wall of their colon or rectum. These cells are responsible for the normal wave-like motion of the bowel (peristalsis), and when they are missing the stool stops and an obstruction occurs. The length of affected bowel varies, but the most common transition point is in the upper rectum or the sigmoid colon. Children with Hirschsprung's disease present to the doctor in one of three ways: 1) vomiting and abdominal distention as a newborn, 2) chronic constipation, or 3) more rarely diarrhea, fever, and distention (symptoms associated with enterocolitis—inflammation of the small intestine and colon). The diagnosis is made by a combination of barium x-rays and rectal biopsy.

The treatment of Hirschsprung's disease is primarily surgical. The goal of surgery is to remove the abnormal bowel and attach the normal bowel to the anus just above the sphincter. The three commonly performed operations, known as pullthrough procedures, are called the Swenson, Duhamel, and Soave operations. The long-term results appear to be similar whichever operation is used.

Similar to high anorectal malformations, children with Hirschsprung's disease used to routinely have a colostomy done at the time of diagnosis, with definitive reconstruction done at a later stage. In the past 10–15 years, an increasing number of surgeons have been performing a single-stage repair in most cases. More recently, less invasive operations using laparoscopy or a transanal "incisionless" approach have been used by many surgeons, with shorter hospital stays, less pain, and less scarring.

Although most children have excellent results following surgery for Hirschsprung's disease, approximately 10–20% experience continued defecatory problems in the form of enterocolitis, incontinence, and persistent constipation. Incontinence may be caused by sphincter damage at the time of the operation, abnormal sensation, or overflow associated with chronic constipation. Anorectal manometry, a test in which the pressures in the anal sphincter are measured, may

be very helpful in determining the cause in an individual patient.

Persistent constipation can be a frustrating problem after a pullthrough. Some patients may have mechanical causes such as a stricture, and rare patients may have recurrence of the Hirschsprung's disease. Although many of these children can be managed without repeat pullthrough surgery, some may require an additional operation. In the majority of children with persistent constipation, the problem is that the internal anal sphincter doesn't relax normally making it difficult to push stool past it. Most of these children can successfully be managed using laxatives or enemas, but if problems continue many surgeons recommend cutting the sphincter to "relax" it (sphincterotomy or myectomy). [A possible complication is incontinence.] In an attempt to avoid the potential incontinence associated with a sphincter cutting procedure, we have been advocating the use of botulinum toxin (Botox) in these patients. Although not all children improve with this approach, those who do can be treated by repeated injections of botulinum toxin, and in most cases the problem resolves by itself by the age of five or six.

Colonic Motility Disorders. There are a small number of children who present with many of the same symptoms consistent with a diagnosis of Hirschsprung's disease, but when the biopsy is done there are ganglion cells present. In some of these children, the ganglion cells are completely normal, and others may exhibit microscopic abnormalities known as intestinal neuronal dysplasia. Still other children may have abnormal motility as a result of abnormalities of the intestinal muscle, or for other unknown reasons.

Colonic motility disorders may involve a small area of the bowel, or the entire colon. Specialized pressure studies of the colon (colonic manometry) may help in differentiating between the two, and in some cases a surgeon may need to do biopsies. These can be done using a regular abdominal incision, although the laparoscopic approach causes less pain and scarring. If only a small part of the colon is abnormal, an operation to remove it may be very helpful.

THE INABILITY TO CONTROL DEFECATION

Fecal incontinence represents an enormous problem for affected patients and their families. There are many causes of incontinence in children, including abnormalities of sensation, poor sphincter function, and chronic constipation with overflow. Surgical approaches to this problem can be broadly categorized as those designed to improve the function of the anal sphincter, and those designed to achieve social continence through a bowel management routine.

Techniques to Improve Sphincter Function

Sphincter function may be abnormal due to abnormal nerve supply (e.g., myelomeningocele, spinal cord injury), abnormal development (e.g., high anorectal malformation), or injury (e.g., surgical, traumatic). Most patients can be successfully managed using non-surgical approaches, including dietary manipulation, biofeedback, medications, and enemas. Surgery is only considered if these fail.

The ideal surgical approach is to repair the primary problem, if possible. This is often the case for patients who have a sphincter injury. In adult females, most sphincter injuries occur during childbirth, but children may have the same type of damage caused by injury. Repair of these injuries can be done either immediately, if the injury is recognized, or after a delay of several months. In experienced hands, improvement in continence can be achieved.

In patients for whom a simple repair of the sphincter is impossible, or those in whom the sphincter is missing or non-functional, several techniques have been designed to enhance the function of the sphincter. In the gracilis muscle flap procedure, a long thin muscle from the inner thigh is brought up and wrapped around the anus to create a new sphincter. Most surgeons doing this procedure place a continuous pulse generator to the muscle, which means that the patient does not have to actively tighten the muscle to maintain continence. The majority of the reported patients undergoing this procedure have been adults, however some older children were also included. Although approximately 60% of the patients achieved an improvement in continence, a significant number had infections from the surgery, as well as constipation long term. Application of this technique to a large group of children has not yet been reported.

The other technique that has been developed in recent years is the artificial anal sphincter. This technique is based on a device similar to one that has been used in the treatment of urinary incontinence, and involves an inflatable ring that is implanted around the anus. A significant proportion of adults having this device placed experience complications. It remains to be seen whether this technique will ever play a significant role in the management of incontinence in children.

Surgical Aids to Bowel Management

Social continence, which is really the goal we all have when we are out in the everyday world, can be defined as “defecating when you want to, and not defecating when you don’t want to.” The standard method for achieving social continence involves keeping the rectum and the left side of the colon empty, so that the patient can be active in society without fear of accidental soiling. This is often accomplished using enemas. Enema use, however, may be accompanied by leakage of fluid and discomfort. In addition, some children have difficulty giving themselves enemas. In order to address these problems, a number of antegrade enema techniques have been developed and widely adopted. These procedures involve placing a tube into the cecum (the top part of the colon), and instilling fluid so that the entire colon can be washed out without placing a tube into the rectum. Examples of antegrade enema techniques include the Malone appendicocostomy enema (ACE), the button cecostomy, the use of a tunneled Broviac catheter into the cecum, and the percutaneous cecostomy placed by an interventional radiologist. Although these techniques are quite effective, allowing 90–100% of the children to achieve social continence, they may be complicated in some cases by leakage, infection, dislodgement, or obstruction.

Some children have such severe cases of incontinence that they do better with a colostomy or ileostomy. This may be necessary for a number of reasons, including severe skin breakdown around the anus, failure of other surgical techniques, or unsuitability of the patient for other surgical approaches due to age, anatomy, or family support. Stomas may either be permanent or temporary, depending on the specific child and family, and the reason for creating the stoma.

SUMMARY

The inability to defecate in children is usually due either to a problem with formation of the anus (anorectal malformations) or with the inability of the colon to push the stool from one end to the other (Hirschsprung’s disease and other motility disorders). The surgeon can help children with these problems by reconstructing the anatomy so that there is no mechanical obstruction to the flow of stool, and so that the colon can function normally. In addition, children may develop complications of these conditions, which may be treated by additional surgical procedures.

The inability to control the passage of stool may be due to inadequate muscle, impaired sensation, or severe constipation with “overflow” incontinence. Surgical approaches to these problems include repair of a damaged sphincter, creation of a new sphincter using muscle transfer techniques, or the use of an artificial sphincter, although there is little experience with the last two operations in children. In many cases, adequate sphincter function cannot be regained, and social continence may be achieved using colonic irrigation through a cecostomy tube, which can be placed using a number of different techniques. Occasionally, the best option for an individual child may be the creation of an ileostomy or colostomy.

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