

Primary sigmoidectomy and appendicostomy for chronic idiopathic constipation

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Abstract

Purpose To present our experience in patients with chronic idiopathic constipation complicated by megarectosigmoid and fecal incontinence who underwent a primary sigmoidectomy and appendicostomy.

Methods We reviewed eight patients referred to the Colorectal Center from 2014 to 2016 with chronic idiopathic constipation and undergoing a sigmoidectomy and appendicostomy. We analyzed the previous medical treatment, indications for the surgical procedure, and outcomes.

Results Age at operation was 5–19 years. Time with constipation was 4–15 years. All patients received multiple laxatives, mainly polyethylene glycol, and all had severe social problems. Four patients have autism. The indication for surgery was an unsuccessful laxative trial, refusal to continue with rectal enemas or both, and social fear of continued fecal incontinence. Post-operatively, all patients were having daily bowel movements without fecal accidents.

Conclusion Selected patients with chronic idiopathic constipation complicated by megarectosigmoid and fecal incontinence can obtain great benefit from primary sigmoidectomy and appendicostomy.

Keywords Idiopathic constipation · Sigmoidectomy · Appendicostomy · Fecal incontinence · Megarectosigmoid · Autism

Introduction

Idiopathic constipation is frequent in children, as it accounts for 25 % of pediatric gastroenterology evaluations [1]. This disease presents as a broad spectrum, ranging from minimal and transitory constipation to chronic constipation complicated by dilatation and enlargement of the rectum and sigmoid. This megarectosigmoid becomes a low motility zone that facilitates the persistence or recurrence of fecal impaction and fecal incontinence. These patients are a challenge to treat, but many patients with megarectosigmoid can be managed with a successful bowel management program, tailored to the specific needs of each patient. The goal of this program is to produce an effective bowel movement every day that completely evacuates the rectum and sigmoid. The patient should remain clean in regular underwear, and should not have involuntary bowel movements. Such programs include the use of a stimulant laxative or enemas to evacuate the rectum and sigmoid once a day.

Patients with megarectosigmoid struggle to produce a daily bowel movement, stay clean and not have recurrence of fecal impaction. They need high doses of a stimulant laxative or a daily enema. The patient with idiopathic constipation complicated by megarectosigmoid and fecal

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incontinence becomes a surgical patient when a high dose of stimulant laxative is required (e.g. 14 tablets of sennosides daily), and/or has severe symptoms even with low doses of laxatives (e.g. abdominal pain, cramping or vomiting), and/or when the patient requires daily trans-rectal enemas.

The sigmoidectomy and the antegrade continent enema (ACE) procedures are the most performed operations for these patients. The purpose of the sigmoidectomy is to reduce the length of the bowel affected by hypomotility [2]. This procedure does not completely resolve the major problem, i.e. *megarectum*. Thus, the risk of recurrent rectal fecal impaction and involuntary bowel movements, or both, is always present. If these occur, the patient will require again enemas to reset the use of laxatives. The ACE procedure allows the enemas to be given in a more comfortable manner, via the abdomen instead of through the rectum. This alternative route to give the enema does not resolve the problem per se. It is strongly recommended to achieve successful bowel management based on trans-rectal enemas before deciding to place an ACE.

The purpose of this article is to share our experience with patients with persistent idiopathic constipation, megarectosigmoid and fecal incontinence undergoing primary sigmoidectomy and appendicostomy.

Methods

Between November 2013 and April 2016, 214 patients with chronic idiopathic constipation, complicated by recurrent fecal impaction and fecal incontinence were referred to the Colorectal Center at the Children's Hospital of Pittsburgh of UPMC. Patients with an anorectal malformation or Hirschsprung were excluded. Eight patients underwent a sigmoidectomy and appendicostomy and were included in this series. We reviewed their medical records to analyze previous medical treatments, indications for the surgical procedure, and outcomes. All patients had a contrast enema to determine the size and shape of the rectum and sigmoid. For this radiologic study, the patients discontinued all bowel medications 2 days before and did not receive bowel preparation. For the contrast enema we used iopamidol diluted 50 % and administered via hand-controlled injection. All patients had a negative workup for an organic etiology of the constipation, performed by the referring physician. An approval for this protocol was obtained from our Institutional Review Board (PRO 15030254).

Surgical technique

Two days before surgery, all patients received a bowel preparation with a clear liquid diet, 17 grams of polyethylene glycol orally twice a day, and one daily enema with saline, glycerin, and castile soap. They were admitted 1 day before, and an abdominal X-ray was obtained to evaluate the fecal load in the colon, and continue the bowel preparation accordingly. In the operating room, patients received intravenous antibiotics and colorectal irrigations with a warm saline solution until clear. Patients received general and regional anesthesia. A Foley catheter was placed in the urethra. Six patients underwent a laparoscopic-assisted technique, which was performed with three 5-mm ports to dissect the left and right gutters, cecum and appendix. A 37-cm Covidien LigaSure Maryland Jaw (Covidien, USA) was used to dissect, coagulate and cut the mesentery of the megasigmoid to the level of the peritoneal reflection and 5 cm of the normal-caliber proximal colon. A low Pfannenstiel incision with spreading of the muscles in the midline allowed us to place an Alexis retractor (Applied Medical Resources Corporation, USA) in the abdominal cavity. The proximal colon was transected with a 60 mm GIA stapler (Covidien, USA). The sigmoid was transected 1 cm above the peritoneal reflection using electrocautery, and the specimen was sent to pathology. An end-to-end 2-layer anastomosis was performed between the non-dilated proximal colon and the left side of the dilated rectum. The remaining rectal stump was sutured in two layers with interrupted stitches similar to a Hartmann's pouch. The cecum was exteriorized to create a continent appendicostomy, performing a cecal plication around the native appendix [3]. When the anatomy of the appendix did not allow such a cecal plication, continence was created by invaginating the appendix at the base with four equally spaced stitches of 5-0 silk (Fig. 1). Then, an inverted "V" incision was made in the umbilicus and a skin flap was created. The fascia was opened widely to allow the appendix to pass easily. The appendix was exteriorized through the deepest point of the umbilicus, and the cecum fixed to the anterior abdominal wall. The tip of the appendix was spatulated and anastomosed to the right arm of the V-shaped incision using 6-0 Vicryl sutures. The skin of the left arm of the V-shaped incision was sutured with interrupted stitches using 6-0 Vicryl. A 10Fr feeding tube was left in the stoma and remained there for 2 weeks. On postoperative day 2, patients began a clear liquid diet and flushes through the appendicostomy. An abdominal X-ray was used to confirm a successful clean out of the rectum before discharge. Two patients underwent a purely open

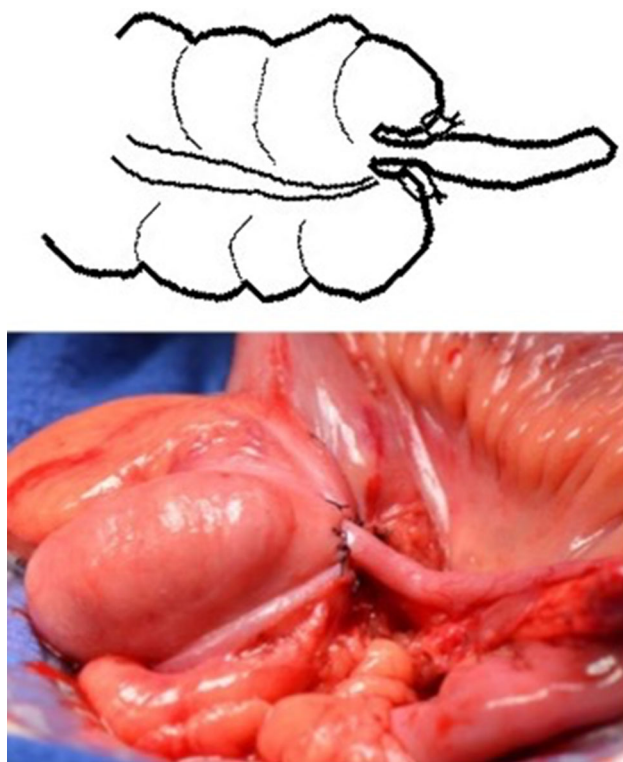


Fig. 1 **a** Diagram and **b** operative image of the continence mechanism at the base of the appendix. This was made by invaginating the base with 5-0 silk stitches

procedure. The open technique was performed through a Pfannenstiel incision, with the rest of the procedure being the same.

Results

From November 2013 to April 2016, 214 patients with chronic idiopathic constipation complicated by megarectum and fecal incontinence were referred and registered in the Colorectal Center Data Base. From this group, eight patients underwent a sigmoidectomy and appendicostomy. The age of the patients at the time of the procedure was between 5 and 19 years of age and they had suffered constipation for 4–15 years. Two were females and six males (Table 1). All patients presented with daily involuntary bowel movements and severe social problems. By contrast enema all patients had a megarectosigmoid with a normal proximal descending colon (typical image shown in Fig. 2).

All patients had received several clean-outs at home with polyethylene glycol mixed with electrolyte solutions. Also, several hospital admissions for clean outs, fecal disimpaction under anesthesia, or both. All had received polyethylene glycol as the primary laxative, and for several

years. Other ancillary laxatives used together with polyethylene glycol included bisacodyl, sennosides, docusate, and milk of magnesia (Table 1). Four patients had autism spectrum disorder, and were on various medications that can cause constipation (Table 2). One teenage patient presented with acute respiratory distress due to restricted breathing from severe abdominal distention, which had to be managed in the pediatric intensive care unit Fig. 3. She underwent fecal disimpaction under anesthesia, bowel management with enemas, and was scheduled for surgery 3 months after the acute event.

Six patients were able to develop a successful bowel management program with one daily enema, but had an unsuccessful laxative trial with sennosides. The enemas were tailored to the specific needs of each patient using a combination of normal saline, glycerin, and soap, and an abdominal X-ray was performed to confirm complete emptying of the rectum. The indication for surgery was an unsuccessful laxative trial, refusal to continue with rectal enemas, and fear of continued fecal incontinence due to severe social embarrassment. Two patients did not receive preoperative bowel management with enemas. In one of those case, the family refused enemas because there was an alleged history of sexual abuse. The second patient has autism with an aggressive component that would not allow for the administration of rectal enemas.

Age at sigmoidectomy/appendicostomy was 6–19 years. Six patients underwent a laparoscopic-assisted surgery, and two patients had open surgery. There were no immediate or late post-operative complications. Follow up was 2–10 months. After the sigmoidectomy/appendicostomy, all patients remain clean with one daily enema via appendicostomy. Our protocol is to continue with enemas via appendicostomy for 6 months and then attempt a laxative trial. One teenager requested to stop the use of enemas and we switched to sennosides 4 months after surgery. On follow up, he skipped two doses of sennosides but did not have any bowel movements; he then had to use the appendicostomy to give antegrade enemas in order to reset the use of laxatives.

Discussion

Persistent idiopathic constipation becomes a medical challenge when it is complicated by fecal incontinence and megarectosigmoid. It is critical to timely detect unsuccessful medical treatment with laxatives to prevent or stop the progression of the damage to the intestine. Unsuccessful medical treatment of chronic idiopathic constipation is characterized by: failure to produce a daily bowel movement, recurrent fecal impaction requiring clean-outs, uncontrolled fecal incontinence, progressive frustration of

Table 1 Clinical characteristics, previous treatments, and type of operation in eight children with idiopathic constipation complicated with megarectosigmoid and fecal incontinence

Therapeutic intervention	Patient							
	1	2	3	4	5	6	7	8
Sex (male or female)	f	m	m	f	m	m	m	m
Years of constipation	4	5	15	12	10	8	6	6
Age at onset of constipation	1	9	4	2	2	1	1	6
Age at surgery	5	14	19	15	13	9	9	12
Different laxatives tried (<i>n</i>)	3	5	2	1	5	4	3	2
Polyethylene glycol	+	+	+	+	+	+	+	+
Bisacodyl	+	+	+		+		+	+
Sennosides	+	+			+	+	+	
Docusate		+			+	+		
Milk of magnesia						+		
Fiber					+			
Lubiprostone		+						
Botox		+						
Clean out/disimpaction	2	2	1	1	1	5	1	3
Successful bowel management with enemas	+	+	+	+	+	+	NA ^a	NA ^b
Ganglion cells in rectum	+	+	+	+	+	+	+	+
Contrast enema	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes
Autism specter disorder	+	—	—	—	+	+	—	+
Laparoscopic-assisted	No	Yes	Yes	Yes	Yes	Yes	No	Yes

^a Step father was accused for sexual abuse and family refused enemas

^b Autism with severe aggressive behavior and chronic recurrent fecal impaction

Fig. 2 Contrast enema showing a megarectosigmoid with a normal proximal descending colon. **a** Preoperative, **b** after sigmoidectomy/appendicostomy

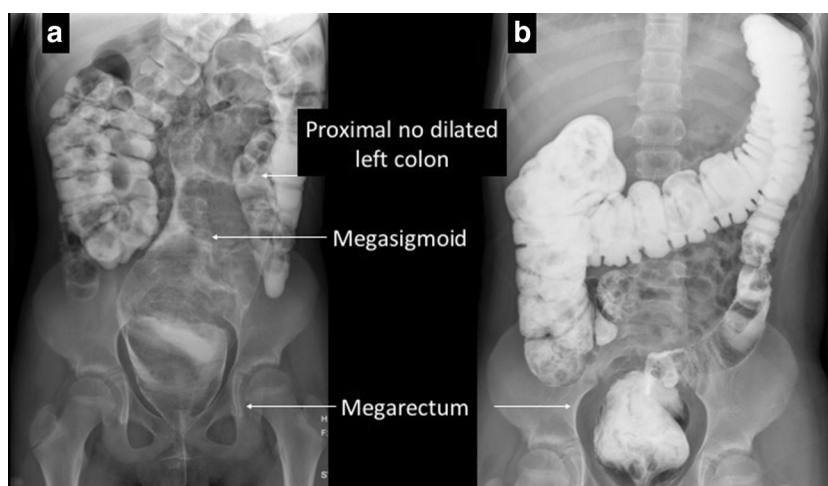
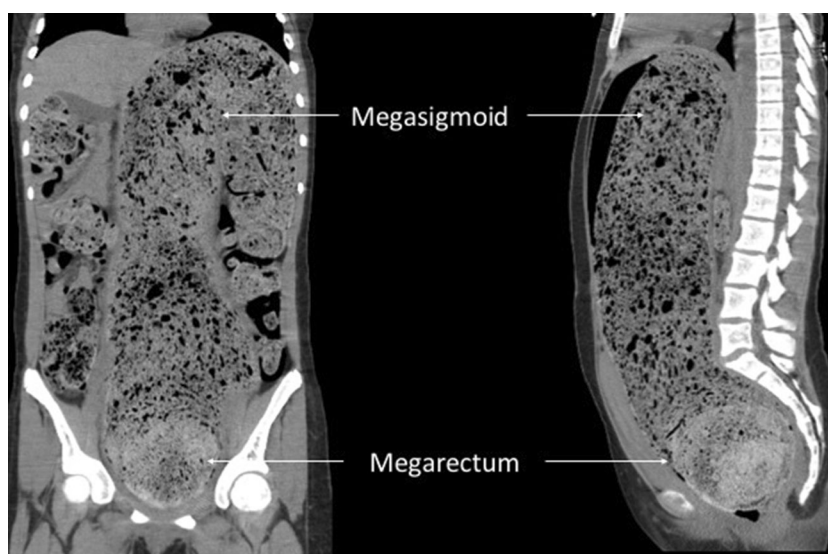


Table 2 Patients with autism spectrum disorder and medication they received

Patient	Medication for autism
1	Clonidine
5	Atomoxetine, Risperidone
6	Atomoxetine, Mirtazapine, Lamotrigine
8	Quetiapine, Guanfacine, Amphetamine/Dextroamphetamine

the patients and family, and loss of self-esteem with often severe social difficulties. We found that all patients in our series had previously received polyethylene glycol and other medications for several years, and they all had been having chronic fecal impaction and daily involuntary bowel movements that caused severe social problems. It has been reported that as many as 30 % of treated constipated children remain symptomatic after puberty [4], an incidence we consider unacceptable. We believe that

Fig. 3 Abdominal CT images of a patient with megarectosigmoid. She had acute respiratory distress and restricted breathing for severe abdominal distention due to chronic fecal impaction



children with severe chronic idiopathic constipation and megarectosigmoid do not benefit from treatment with polyethylene glycol, they should be referred to a specialized service in a timely manner.

Antegrade enemas and bowel resections are colorectal surgical procedures having an important role in the treatment of different functional colorectal problems. These two operations have evolved since the first use of antegrade continence enema (ACE) in 1990 to treat patients with fecal incontinence associated with myelomeningocele and anorectal malformations [5], and since colorectal resections in 1982 were used to treat megarectum in patients with anorectal malformations [6–8]. Colonic resections or ACE procedures have also been used in children with chronic idiopathic constipation [1, 3, 9–11].

Sigmoidectomy reduces the length of the dysfunctional hypomotile segment in patients with megarectosigmoid. Unfortunately, this operation still leaves in the distal dysfunctional hypomotile megarectum. Thus, these patients require close follow-up to prevent rectal fecal impaction and possibly the development of new enlargement of the proximal segment. The resection of both problematic segments: the rectum and the sigmoid has been reported [12]. The risk of this procedure is that it can lead to fecal incontinence, since it eliminates the main fecal reservoir [13]. An ACE procedure allows for a more comfortable trans-abdominal enema instead of the uncomfortable trans-rectal enema, mainly in older children, adolescents and patients with comorbidities.

In 2002, Lee et al. placed a cecostomy button and performed the resection of the megarectum in four patients with fecal incontinence associated with idiopathic constipation [14]. In 2013 Eradi et al. created an appendicostomy and performed a colonic resection in 18 patients with

anorectal malformations, spina bifida and sacrococcygeal teratoma [15].

Ours is the largest series of patients with idiopathic constipation complicated with megarectosigmoid and fecal incontinence surgically treated with this combined procedure. The eight patients all showed an excellent outcome. All have an effective daily bowel movement, no involuntary bowel movements and all are able to wear regular underwear. This combined procedure reduced the length of the dysfunctional bowel segment and, because we are leaving the megarectum, which increases the risk of fecal impaction, we decided to also perform the appendicostomy to supplement the bowel management of these particular patients as needed.

Patients with autism spectrum disorder receive medications that can produce hypomotility, thus making the medical management of the constipation in these patients more difficult. Even more, the administration of rectal enemas in these patients with neurologic comorbidity is challenging. So, this combined procedure offers benefits to these particular patients. In our series, the indication for these procedures was comorbidity of autism spectrum disorder in four patients.

Children with chronic constipation and megarectosigmoid need high doses of stimulant laxatives to produce an effective daily bowel movement to completely clean the rectum and sigmoid. Some patients receiving this high dose of laxative have discomfort and unpleasant side effects such as cramping, vomiting, poor appetite or bloating, even when they have only one effective bowel movement daily. These patients are considered to have failed medical treatment of constipation [10] and are suitable for a Bowel Management Program based on daily enemas. These patients benefit from an appendicostomy [11] providing a

safe and comfortable route to administer the enema. In our series three patients had severe fecal incontinence due to massive dilation of the rectosigmoid. All three stayed clean pre-operatively with high volume enemas for several months. In these patients the primary sigmoidectomy and appendicostomy resulted in an excellent outcome. Two are clean with a daily enema with less volume and one was able to switch to oral laxatives 4 months after the procedure.

Social problems related to constipation and fecal incontinence have a negative impact on children and their families [1, 16]. Children who wear pull-ups out of the potty training age, or with constant soiling, suffer severe social and psychological consequences. Bongers et al. found 60 % of 136 children with constipation and secondary fecal incontinence were afraid of having feces in their underwear, 78 % abhorred this situation, and 23 % had been bullied by other children [16]. All of the patients in our series had presented with involuntary bowel movements for many years before they were referred to the Colorectal Center. Once they began on a bowel management and remained clean wearing regular underwear, three patients were afraid of returning to fecal incontinence again. Knowing that surgery would leave behind a problematic megarectum, we created the appendicostomy in combination with the sigmoidectomy to safeguard against involuntary bowel movements secondary to recurrent fecal impaction in the megarectum. This surgical plan was particularly useful in the one patient who had fecal impaction due to missing 2 days of sennosides.

Conclusion

Sigmoidectomy is enough to reduce the dose of laxatives in most patients with chronic idiopathic constipation complicated by megarectosigmoid and fecal incontinence. It alone is ideal for those patients who can tolerate rectal enemas in the event of a rectal fecal impaction to reset the use of laxatives.

Appendicostomy is useful in patients with chronic idiopathic constipation who have an unsuccessful laxative trial and have a good response to a Bowel Management Program based on trans-rectal enemas.

Primary sigmoidectomy and appendicostomy together represent an excellent surgical approach for patients with chronic idiopathic constipation complicated with megarectosigmoid and fecal incontinence who have a comorbidity with autism spectrum disorder or when patient and family refuse the use of enemas per rectum. The

appendicostomy safeguards against recurrent fecal impaction and involuntary bowel movements in these cases.

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