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Adjuncts to bowel management for fecal incontinence and constipation, the role of surgery; appendicostomy, cecostomy, neoappendicostomy, and colonic resection

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Abstract

Constipation and fecal incontinence are common problems in children after repair of an anorectal malformation (ARM). While many children can be effectively managed with an oral laxative regimen, others require a mechanical colonic washout to achieve social continence. Appendicostomy and cecostomy are two techniques which permit antegrade access to the colon for the purpose of enema delivery, which improves compliance and quality of life for patients and families. The purpose of this article is to review, using a case-based approach, the indications for placement of a channel for antegrade enema access, clinical scenarios in which one technique would be preferred over another, common complications following each procedure.

Case #1

You are seeing a 7 year old adopted male child in your clinic with fecal incontinence and a history of an anorectal malformation (ARM) repaired in his home country. Detailed information of the child's surgical repair is unavailable. His caregivers have been treating him with high-dose senna which causes severe abdominal cramping and little improvement in fecal continence. Rectal enemas were previously used with moderate success, however the child was extremely resistant to rectal catheter insertion and became quite anxious around the timing of his flush. You perform a contrast enema which shows a mildly dilated rectosigmoid. Examination under anesthesia demonstrates a well-centered anoplasty without stricture or prolapse. He is continent of urine and has no renal dysfunction.

What would you recommend?

A) Recommend medical therapy with miralax.

B) Recommend continued rectal enemas.

C) Recommend creation of an appendicostomy.

D) Recommend diverting sigmoid colostomy.

Constipation and fecal incontinence later in childhood are common in children born with anorectal malformations (ARMs). Children with more complex malformations, a poorly formed sacrum, or an associated spinal anomaly (e.g. tethered cord) are at an increased risk for incontinence most notable as children begin toilet training. [1] While some children

may be successfully managed with simple dietary modifications or laxative therapy, others require intensive bowel management programs (BMP) with enemas to achieve social continence.

For children who require enemas to achieve regular, predictable bowel movements, enemas can be administered into the colon either in retrograde (via the rectum) or antegrade (directly into the cecum) fashion. Data does not support a functional benefit from direction of enema administration; however, there are benefits afforded by antegrade colonic access. [2] Practically, a stoma on the abdomen is both easier and more comfortable to catheterize than inserting a rectal catheter. This is especially true in children with a history of painful anal dilations, as is common in children with an ARM. A combination of this anal defensiveness and the caregivers' important need to establish bonding with their adopted child makes rectal enema administration very difficult. Secondly, antegrade enemas allow the child to gain more independence in his or her bowel management as they grow. Children can be taught at a young age the process of catheterizing their appendicostomy channel and administering their flushes; children then gradually take over more responsibility for their flushes as they are developmentally ready.

Case #2

You are seeing a 3 year old adopted male child who had an ARM repaired in his home country. He has severe constipation which is minimally improved with

high-dose laxatives. The child is extremely resistant to rectal enemas. His sacral ratio is 0.4. He remains diapered for urine, however he has not had any urinary tract infections nor evidence for renal dysfunction. Examination under anesthesia does not identify any anatomic abnormalities.

What would you recommend?

A) Recommend laxative therapy.

B) Recommend continued rectal enemas.

C) Recommend creation of an appendicostomy.

D) Recommend laparoscopic-assisted placement of a cecostomy.

A cecostomy is a valuable tool in managing children with anorectal malformations with severe constipation or fecal incontinence. This involves placement, either surgically or percutaneously, of a tube directly into the cecum for the purpose of antegrade enema administration. [3,4] This can be useful in patients in whom the urologic status, or the need for possible future urologic reconstruction, is unknown, as it provides antegrade enema access while preserving the appendix.

If the patient requires appendicovesicostomy in the future, the cecostomy can be removed, and an appendicostomy and appendicovesicostomy can be created simultaneously using the split appendix technique, if amenable. [5] [Figure 1 a-

d] If the child does not require urologic reconstruction, then the cecostomy can either remain or be removed with conversion to in situ appendicostomy, depending on the preferences of the parents and patient.

Little data exists directly comparing cecostomy with appendicostomy. Cecostomy is a technically easier procedure than appendicostomy and can be performed percutaneously using only sedation. Additionally, cecostomy requires permanent placement of a Chait or other tube directly into the cecum, which prevents the need for daily catheterization of a channel, which may be of benefit in patients with coexisting sensory disorders who would resist catheterization of an appendicostomy channel. [Figure 2 a,b] Downsides include the need for routine tube exchanges, in addition to unplanned exchanges for clogged or dislodged tubes. Exchanges require a medical visit and often radiation to radiographically confirm tube placement. Cecostomy is also associated with a high rate of stool leakage around the tube, as they lack the valve that is created during plication of the cecum around the appendix during creation of an appendicostomy. [6] [Figure 3 a,b] Lastly, intraabdominal leakage of stool into the abdominal cavity is a major complication that can result from dislodgement of a percutaneous cecostomy tube in the early post-procedure period. This complication is virtually not seen after appendicostomy, even after the appendicostomy falls from the abdominal wall, because of the valve mechanisms preventing stool spillage.

Case #3

An 11 year old child with a previously repaired repaired anorectal malformation with recto-bladder neck fistula comes to your clinic for evaluation. He has been managed successfully on a retrograde enema regimen, however the parents are seeking an antegrade option to allow the child to take a more active role in his bowel management. On further investigation, you learn that he is incontinent of urine and requires intermittent catheterization to remain dry. He has frequent urinary tract infections and is followed by a nephrologist for renal dysfunction.

What would you recommend?

A) Continue rectal enemas and clean intermittent catheterization (CIC), reassess in 12 months for need for colorectal and urologic reconstruction.

B) Offer appendicostomy only, continue CIC.

C) Offer appendicostomy and appendicovesicostomy using the split appendix technique.

D) Offer cecostomy placement, reassess in 12 months for need for colorectal and urologic reconstruction.

Case #4

The patient in the above case agrees to appendicostomy and appendicovesicostomy. Intraoperatively, you find that the appendix measures 4 cm, and is not of adequate length to create a catheterizable channel to both the colon and the bladder.

What is your next step?

A) Perform appendectomy and abort colorectal and urologic reconstruction.

B) Perform appendicostomy and ileovesicostomy.

C) Perform appendicostomy and colovesicostomy.

D) Perform neoappendicostomy and appendicovesicostomy.

There are multiple techniques by which a surgeon can create a catheterizable channel to the colon for the purpose of administering enema flushes. Two common procedures are the appendicostomy and neoappendicostomy. The appendicostomy for the purpose of creating a catheterizable channel for the purpose of enema administration was first described by Dr. P.S. Malone in his seminal paper in The Lancet in 1990. [7] In the original description of the procedure, the appendix was divided at its base, reversed, and then reimplanted submucosally into the cecum. A later technical revision of the procedure described by Dr. Malone eliminated reversal of the appendix. [8] There have since been several

modifications of this procedure, but the most commonly performed technique and that for which we advocate is the in situ appendicostomy with 360° cecal plication. [6] [Figure 3 a,b] In patients without an appendix, those with an appendix that is unusable for appendicostomy, or those for whom the appendix is needed for urologic reconstruction, neoappendicostomy using a cecal flap is preferred. [9] [Figures 4 a, b - 5] Patients with a future need for an appendicostomy, who have malrotation and undergo a Ladd's procedure, should have their appendix preserved.

The techniques for both procedures are briefly described below.

Appendicostomy technique:

The cecum and right colon are mobilized from their retroperitoneal attachments in order to bring the appendix and cecum through an umbilical incision. The tip of the appendix is opened and a 10 Fr. feeding tube placed through the lumen. The cecum is plicated around the appendix using a non-absorbable suture, thereby creating a valve mechanism. A V-Y anastomoses of the appendix to the umbilical skin is performed with 5-0 absorbable suture. The 10 Fr. feeding tube/catheter is placed into the lumen and secured to the skin.

Neoappendicostomy technique:

The cecum and right colon are mobilized from their retroperitoneal attachments in order to bring the appendix and cecum through an umbilical incision. A U-shaped flap of cecum is incised over a single vessel and tubularized using a 10 Fr. feeding tube/catheter over a single vessel. The cecal defect is closed in 2 layers with 4-0 absorbable sutures. The cecum is plicated around the neoappendix using a non-absorbable suture, thereby creating a valve mechanism. A V-Y anastomoses of the appendix to the umbilical skin is performed with 5-0 absorbable suture. A 10 Fr. feeding tube is placed into the lumen and secured to the skin.

The choice of conduit is determined by patient factors including anatomy, coexisting disease, and future reconstructive needs. For instance, patients with bladder dysfunction who may ultimately need urinary diversion can undergo a split appendix technique whereby the appendix is used to reconstruct both systems. [5] [Figure 3 a,b] We feel it is very important that Malone appendicostomies not be performed until the plan for urinary reconstruction is clear and advocate for a strong collaboration between urology and colorectal surgery. For patients in need of colorectal and urologic reconstruction, collaboration between both services allows for 1) potential sharing of the appendix using the split appendix technique, 2) minimization of the number of abdominal procedures, 3) minimization of general anesthetic events, and 4) overlay of postoperative recovery periods. [10]

Case #5:

A mother brings her 7 year old daughter to your clinic for a follow up appointment 6 months after undergoing creation of an appenicostomy for antegrade continence enema administration.

What do you advise in terms of long-term outcomes after appendicostomy?

A) Complications, although rare, always require major revision.

B) A catheter must remain in the channel at all times to prevent stenosis.

C) There is no difference in the rate of complications after appendicostomy or neoappendicostomy.

D) Stenosis is the most common complications and can be repaired, in most cases, with a minor skin-level procedure.

Relatively little data is available on the surgical outcomes of these two surgical procedures, owing mainly to small studies with small populations, especially in the neoappendicostomy group, as this is a more specialized procedure. [11, 12] Fairly well-described complication profiles follow these cases, including skin leakage, prolapse, stenosis, wound infection, as well as detachment from the abdominal wall. [11]

Stenosis is the most common complication after appendicostomy and may complicate up to 20% of cases. [13-15] The lead author of this report published a 13% rate of stenosis in a series of 150 consecutive patients undergoing appendicostomy at a single institution. [11] This lower rate may be attributed to the practice of leaving an ACE stopper in patients for 6 months postoperatively after appendicostomy. Stenosis is an expected complication as it is part of the natural healing process to attempt closure of appendiceal opening at the skin. Regular cannulation of the lumen is therefore required to ensure the lumen remains patent. In this same series, the rate of stenosis after neoappendicostomy was found to be nearly three times higher than that after appendicostomy, which was likely the result of the relatively poor blood supply to the cecal flap provided by the single vessel compared to the rich mesentery supplying the appendix. [11]

Case #6

A 12 year old female with history of an anorectal malformation repaired in the newborn period comes to your office for follow-up. She underwent Malone appendicostomy 6 months earlier; however flushes take 3 hours despite several attempts to adjust the volume and dose of stimulant, and she has several accidents throughout the day. The patient would like a solution before starting high school next year. A contrast enema demonstrates a markedly dilated rectosigmoid colon with significant stool burden, not significantly changed from the study obtained pre-appendicostomy.

What would you recommend?

A) Recommend initiating laxative therapy.

B) Recommend daily suppository.

C) Recommend appendicostomy only.

D) Recommend resection of the portion of dilated rectosigmoid.

Significant dilation necessitating segmental colonic resection is the end result of chronic, untreated or inadequately managed, constipation in children with ARMs. This is relatively infrequent, which speaks to the success of an effective bowel management program. However in children unresponsive to a bowel management program, megarectosigmoid should be ruled out. In these cases, when bowel management is unsuccessful in emptying the dilated colon of stool, a segmental colonic resection of the dilated portion of bowel may help the child achieve social continence, improving the reliability of their flush regimen. In these cases, it is important to consider the residual blood supply to the bowel. In patients after PSARP, the distal bowel is supplied primarily by the superior hemorrhoidal arteries, a branch of the inferior mesenteric artery, as the inferior and middle hemorrhoidal arteries are normally disrupted during the course of PSARP. It is therefore important to ligate vessels close to the bowel to preserve blood supply via the IMA.

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Figure 1: (a) Normal anatomy of the appendix and cecum. To perform the split-appendix technique for creation of an appendicostomy and appendicovesicostomy, (b-c) the appendix is sharply divided, with the proximal portion serving as the appendicostomy and the distal end as the appendicovesicostomy. (d) After division of the appendix.

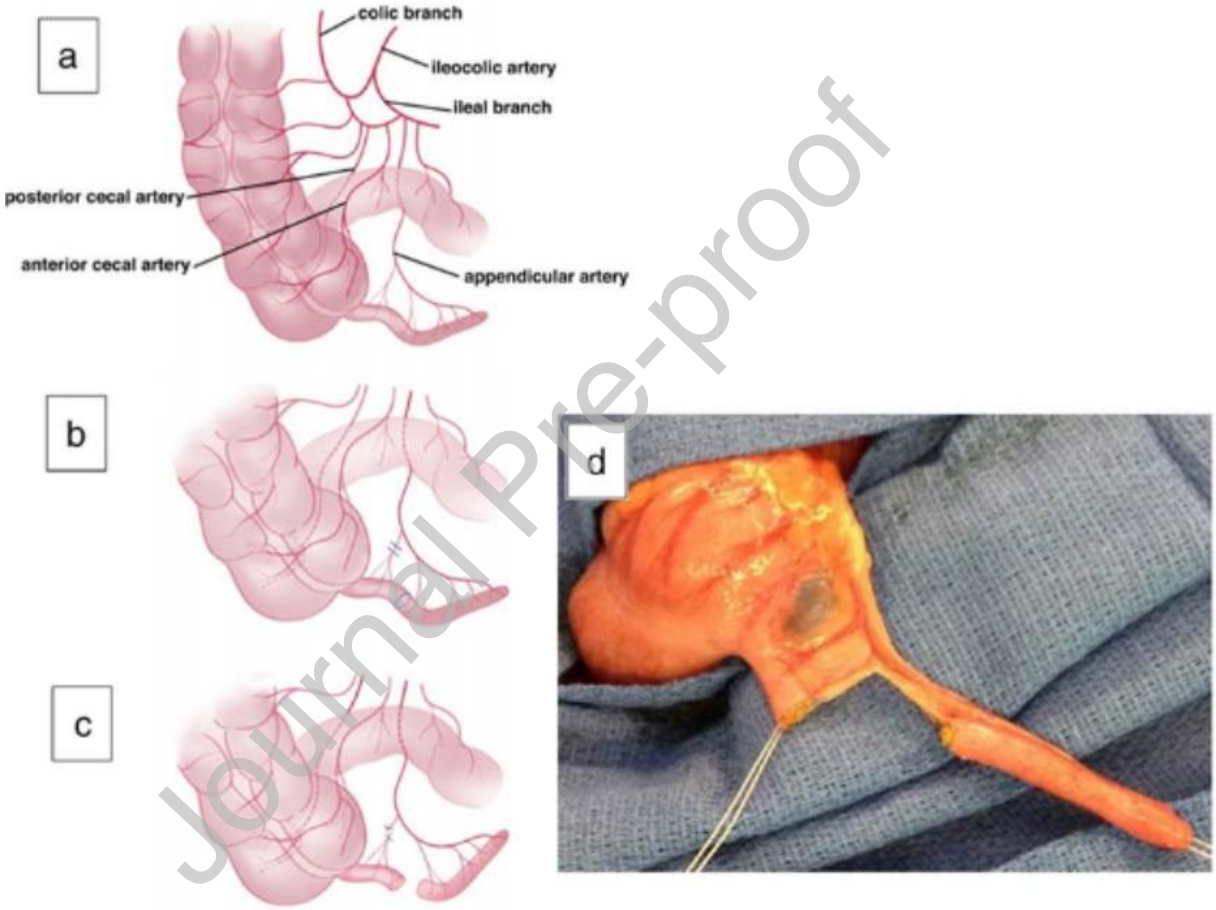

Figure 2: Postoperative, external view of the (a) Malone appendicostomy in the umbilicus and a (b) cecostomy (Chait) device in the right lower quadrant.

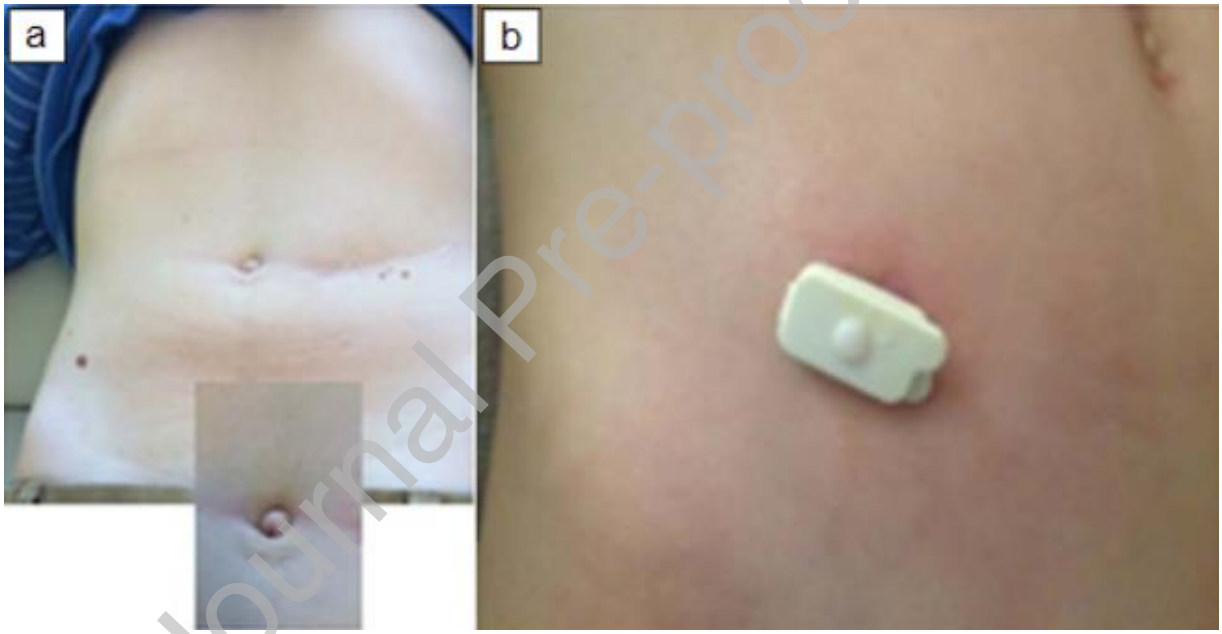

Figure 3: (a) To create the valve mechanism, a 360° cecal plication is performed around the base of the appendix with interrupted suture which avoids interrupting the blood supply to the appendix. (b) After plication of the cecum and transection of the appendix to its desired length.

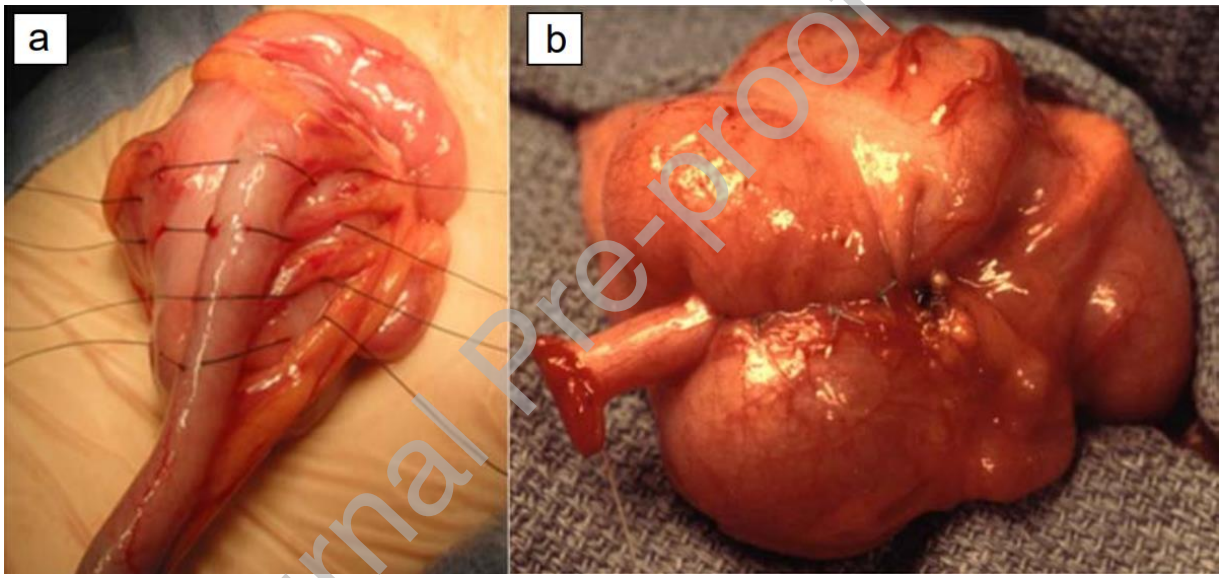

Figure 4: (a) To create a neoappendicostomy with a cecal flap, a U-shaped flap of bowel is planned around a single feeding vessel and (b) tubularized over a 10 Fr. feeding tube.

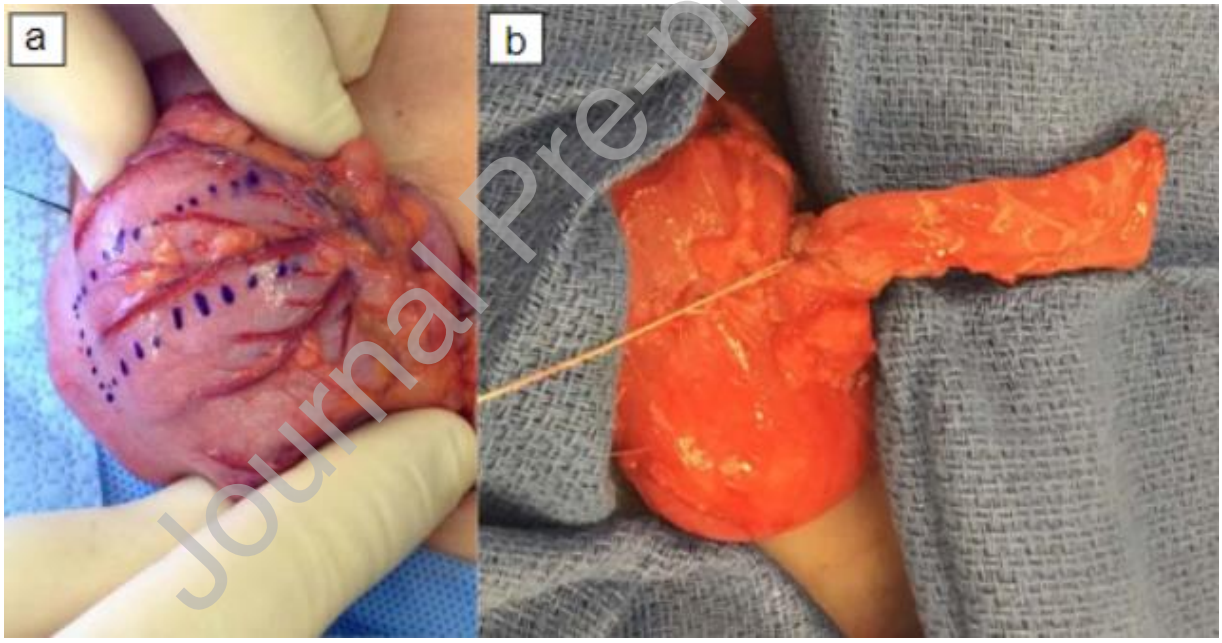

Figure 5: Intraoperative decision making for utilization of the appendix depends, in large part, on the length of the appendix. In cases where the appendix is short (≤5 cm), the appendix should be used preferentially for creation of an appendicostomy, and an appendicovesicostomy can be created using small intestine. When the length of the appendix is between 5 and 7 cm, the appendix should be preferentially utilized for appendicovesicostomy and a U-shaped flap of cecum used for neoappendicostomy. A split appendix appendicostomy and appendicovesicostomy is feasible in most cases when the appendix length is greater than 7 cm.