The Malone Antegrade Continence Enema: A Personal 15-Year Voyage

I was very fortunate to have had the privilege to perform, and hence describe the first MACE procedures done in North America. Indeed this was serendipitous. Philip Ransley, Tony Manzoni, along with Rick Hurwitz, David Diamond, and David Bloom were visitors in Denver in 1991. Philip and I were repairing a child with cloacal extrophy who had had her prior colostomy created with preservation of her rudimentary hindgut. During our operative procedure, he asked me if we were going to take down the colostomy in addition to the urological and vaginal reconstruction. Of course I thought that he was insane. He then went on to describe the work of his then registrar, Pat Malone, and Ed Kiely who were performing what they called the antegrade continence enema procedure. Pat Malone introduced this procedure over 10 years ago. Each of the variations of the technique, with the advent of more advanced technology, is addressed in this issue by experienced authors. It is likely that with time we will see further innovations on the basic theme that Pat Malone introduced. Most of us have witnessed a dramatic effect on the child and family. It has certainly stood the test of time.

I applaud the guest editors as well as the individual contributors for a wonderful update on the MACE procedure.
ema or ACE procedure for children with fecal incontinence. Indeed, I hazard a guess that few of us had read the preliminary report on this subject that this group had already published less than a year previously in Lancet. In reality, all of us had learned how to reconstruct the urinary tract in order to keep kids dry, yet they still remained in diapers due to fecal incontinence. Following this logic, Philip and I called in one of my pediatric surgical colleagues and explained the rationale behind the ACE to him. He left the room laughing, of course refusing to do a pull through. Philip then persuaded me to perform the pull through along with the remainder of the reconstruction, which I did with far less reluctance. Unfortunately Philip left Denver immediately after surgery, to follow yet another eclipse, and he didn’t let me know what one did to properly use the cutaneous enema site. Figuring an enema is an enema, we started administering increasing volumes of tap water until we had acceptable results. This became our standard instillate in Denver and indeed has been adopted as the most commonly used fluid in North America for MACE irrigations. That young lady has just passed her fifteenth year anniversary of having had her MACE created and continues to thrive, so far without a revision.

Over the next few years after our first MACE, a liaison developed between our center in Denver and Mr. Tim Terry and his associates in Leicester, UK. As a result of this relationship, a series of annual pediatric urological conferences and surgical theater sessions were organized by Mr. Terry. All the participants in the British pediatric urology scene were involved in these Leicester meetings and ultimately Tony Manzoni from Italy and Americans: Tony Caldamone, Rick Rink, Rick Hurwitz and I became frequent visitors as well. Mr. Malone ultimately became an attendee and more importantly, a colleague, collaborator and friend. Through these meetings and operative sessions, we all learned new tricks from another and conceived new ideas and challenged older concepts. Most importantly this led to the cementing of long term friendships and collaborative relationships that just like our original MACE patient thrive to this day.

Being in the right place at the right time was important for my first MACE patient and many others like her. In this DPU, Pat Malone and I have continued our now second decade of collaboration. We have asked some of our “younger” colleagues to contribute the material to this issue and independently, Pat and I then wrote editorial comments, which we hoped would provide additional insight and perhaps even controversy to these subjects. In addition, Pat has written an introduction on his current approaches to patients have undergone his procedure, the “Malone”.

It is important for all of us, in our ever changing and expanding field, to continue to be creative, think outside the box, and learn from one another.
are those developed in reconstructive urology it is the urologist who will end up managing the faecal incontinence1.

The MACE procedure combined three well established principles:
(a) The Mitrofanoff principle to afford a continent catheterizable conduit;
(b) Complete colonic emptying can produce faecal continence;
(c) Complete colonic emptying can be achieved by antegrade colonic irrigation.

Following a MACE, patients have continent intermittent catheter access to their colon through which they administer antegrade washouts, achieve colonic emptying and thus faecal continence.

The initial description of the MACE involved amputating the appendix from the caecum, reversing it and implanting it into a submucosal tunnel, to create a flap valve. The free end of the appendix was brought to a suitable site on the abdominal wall as a continent stoma. Stimulant enemata, high volume water washouts or a combination of both are administered daily or on alternate days. A large number of series including the results of over a thousand children have been published with success rates in the order of 80% reported. More recently significant improvements in quality of life scores have also been reported following the MACE2. Therefore, current evidence would dictate that it should always be considered when a patient with faecal incontinence is undergoing lower urinary tract reconstruction. The results of simultaneous combined bladder reconstruction, Mitrofanoff and MACE procedures have now been reported, with double continence rates of up to 80% described3.

Following the original description, a number of technical modifications were introduced. If the appendix is being used at the level of the caecum it is no longer necessary to disconnect it. It can simply be folded onto the caecum and a caecal wrap performed around it as a valve mechanism, the in-situ appendix4. This is the approach recommended when a combined Mitrofanoff procedure is also required, with the Mitrofanoff being constructed from either a split appendix or a Yang-Monti conduit.

There remains controversy as to whether any continence manoeuvre is required and some surgeons now advocate that the tip of the appendix is simply excised and brought out to the abdominal wall. Stomal incontinence rates are slightly higher using this approach but if a minimally invasive laparoscopic approach is being used to create the MACE the advantages of this would seem to offset the slightly increased risk of stomal leakage. Therefore, if an isolated MACE is only needed, a laparoscopic ACE (LACE) offers a simple and effective approach.

One of the problems encountered in some patients, particularly those with constipation, is the time it takes for the washout to work. In an attempt to overcome this problem some surgeons have placed a Monti-MACE conduit on the left side just proximal to the sigmoid colon to reduce the length of bowel that has to be washed through5. Preliminary results are encouraging and in the authors’ experience this approach has salvaged the procedure in 5 patients in whom an original caecal MACE failed. A left MACE should also be considered in patients with severe constipation.

Stomal stenosis is encountered in up to 30% of patients. In an attempt to overcome this, a minimally invasive technique for placing a caecostomy tube under radiological control was developed6. Once the tract matures the tube can then be replaced with a caecostomy button and this offers an alternative to a catheterizable conduit. For patients with recurrent stomal stenosis it is also possible to leave a button in the conduit following dilatation. Recently a development embracing the principles of minimally invasive surgery and the left sided MACE has occurred. Colonoscopy is performed and with the scope in the distal descending colon a colostomy tube is inserted, as one would insert a percutaneous endoscopic gastrostomy (PEG), the Percutaneous Endoscopic Colostomy (PEC). Washouts can commence the next day and once the tract has matured the patient has a choice of either keeping the tube, exchanging it for a button or having a formal conduit created, knowing that the MACE will work.

There are now a number of different approaches to the MACE. If the patient requires a synchronous bladder reconstruction a formal conduit is constructed using the in situ appendix or a left sided Monti-MACE if constipation is a severe problem. If the MACE is only required a LACE would be the procedure of choice for caecal placement or a PEC if the patient is constipated and a left sided placement is needed.

There is no single correct washout regimen and most patients adapt any regimen to their own needs, when they leave hospital and return home. There is a trans-Atlantic difference, with practitioners in the United States using high volume washouts without stimulants in the majority of their patients, whereas in Europe most patients use some stimulant such as phosphate, bisacodyl or liquirice to name a few. Despite this difference there would now seem to be universal agreement that saline is not necessary for the washouts and tap water is safe and effective. The volume of washout used, the infusion rate, the frequency of washouts and the time taken varies for all patients but on average the majority of patients will use the MACE on alternate days and the average time taken is approximately 40 minutes.

Even with these technical developments it is important to remember that the original principles that contribute to a successful MACE still apply. In summary these are patients who are over 5 years of age with a neuropathic bowel or an anorectal malformation and who are motivated to be continent do best. Pre-operative counselling and continued post-operative support, provided by nurse specialists, is essential.

References

The initial description of the MACE involved amputating the appendix from the caecum, reversing it and implanting it into a submucosal tunnel, to create a flap valve. The free end of the appendix was brought to a suitable site on the abdominal wall as a continent stoma.
What Is the Recipe for Success?

Beyond careful patient selection and preoperative counseling regarding appropriate expectations for improvement in bowel management, choosing and fine-tuning the enema regimen is critical to the success of the antegrade enema program. Persistence and flexibility can play an important role. Multiple solutions, each with its own advantages and potential side effects, have been used successfully. Fortunately we can draw from the different approaches in several larger series to troubleshoot the more challenging patients and optimize the clinical outcome.

Hypertonic, isotonic and hypotonic solutions are all used as the primary enema with good results. Hypertonic solutions have the advantage of a stimulating or cathartic effect on the colon and typically result in more rapid washout at lower volumes. Painful colonic spasms with irrigation are likewise more frequently observed with hypertonic solutions, but may occur also with isotonic or hypotonic solutions at high infusion rates or volumes. Larger volumes and slower washout are more typical with normal saline and tap water, but there appear to be fewer electrolyte complications. Tap water is of course free and readily available, an advantage whether at home or on the road.

Due to the absorptive capabilities of the colon, no enema solution is completely safe, and administration of multiple enemas is potentially hazardous in any patient. Enemas must be used with more caution, however, in patients with altered gastrointestinal motility or impaired renal function. Unfortunately the patients who will benefit from an enema program, those with neuropathic bowel or anorectal malformations, are also the most likely to have some degree of acquired megacolon and/or renal impairment. Failure to evacuate the enema due to fecal impaction or anal stenosis can also increase the risk of water or electrolyte absorption.

Phosphate toxicity has been reported after serial retrograde phosphate-based enemas and in several patients receiving antegrade phosphate-based enemas. Dehydration, mental status changes, cardiopulmonary instability and carpopedal spasm accompany the hyperphosphatemia with secondary hypocalcemia, hypernatremia and metabolic acidosis. Treatment is directed toward aggressive fluid resuscitation for systemic perfusion and renal clearance of the phosphate. The retained enemas should be evacuated by manual disimpaction with or without colonic lavage or endoscopic intervention. Calcium replenishment is guided by symptomatology rather than the absolute value.

One case of fatal hypernatremia occurred after use of a homemade saline preparation for antegrade washout in a child with a repaired anorectal malformation and recurrent anal stenosis. The enema was prepared by a less experienced caregiver and was possibly more hypertonic than usual. An isotonic saline solution is likely safer than a hypertonic solution and is prepared with 1.5 teaspoons of table salt (100 mEq each of sodium and chloride) per liter of plain tap water.

Although water intoxication has been observed in children and in the elderly after multiple high colonic enemas, hyponatremia or suspicious associated symptoms have not been found in patients who use tap water for daily antegrade enemas. Tap water enemas should be used with increased caution, however, in patients with the aforementioned underlying conditions and in those already at risk for increased free water retention (excessive water intake, SIADH, nocturnal use of DDAVP). Asymptomatic hypernatremia has been detected when the tap water was treated with a sodium-based home water softener, and the abnormality resolved after the softener was disconnected. Due to this potential risk and probable day-to-day variation in the sodium content of treated water, families should be advised to use water from untreated faucets or an alternative source.

Trial and error with the enema regimen is part of the normal postoperative course, as each child and each colon is somewhat different. Frustration and dissatisfaction are likely if the patient and parents do not anticipate weeks to months of finetuning. Typically the antegrade irrigations are initiated on postoperative day three to four when bowel function begins to return. When using tap water or saline, the initial modest daily volume of 50 milliliters is increased by 50 to 100 milliliters every 3 days until reliable results are obtained. Rectal output may not occur with the lower enema volumes. If good results are not obtained by the time the enema volume reaches 500-600 milliliters, changes in or additions to the regimen should be considered to prevent undue increases in washout time. Changes in the volume, rate or frequency of the enemas may improve the result, but each adjustment deserves a reasonable trial for 4-5 days before making further adjustments. If results remain suboptimal, Gos-Lytele or phosphate-based enema may be used to modify the regimen.

In the United Kingdom, phosphate-based enema solution is typically used, immediately followed by a variable volume of normal saline to complete the cleanout. Initially 50 milliliters of phosphate-based enema is diluted with an equal volume of normal saline. This initial irrigant is followed by 500 milliliters of normal saline. By changing the volume and concentration of the initial phosphate enema the result can be optimized, but the volume of undiluted phosphate enema should not exceed 100 milliliters. The risk of phosphate absorption increases with dwell time. Should there be failure to evacuate the enema after one hour, the saline lavage is repeated. The phosphate enema should never be repeated, however. If the second saline lavage does not produce efflux, the patient should be admitted for retrograde evacuation and observation.

Recurrent constipation can impede normal washout and result in increased soiling. In this situation, 10-15 milliliters of mineral oil can be instilled after the regular enema to lubricate the colonic contents. Alternatively in the absence of a peanut allergy, a Fletcher’s arachis oil retention enema (Pharmax, UK) can be given 12 hours before the usual enema. Oily rectal seepage may require adjustment of timing or volume of instillation. If severe constipation recurs, a formal cleanout from above and below may be required before an enhanced regimen can be reinstated.

Patience, persistence and a healthy respect for the potential electrolyte complications of any given regimen are the foundation for finding an effective antegrade enema regimen for each patient. The above tricks and modifications can help troubleshoot the most challenging patients to find an acceptable result.
Although typically less of a problem after the first few months, pain with instillation of the enema can limit long-term compliance and success with the antegrade enema program in some patients. These spasms can be addressed by decreasing the rate of enema infusion, by reducing the relative concentration of irritant (phosphate) or by pre-treating with an antispasmodic agent (Colofac®, Solvay, UK; 5-15 milliliters of 50 mg/5 ml preparation). Finally a liquorice root solution can be used in lieu of the phosphate-based enema, as a less irritating but effective colonic stimulant. The solution is prepared by boiling 7.5 cm of liquorice root or 5 milliliters of granules in 100 ml of saline and diluting further with 200 milliliters of saline for instillation at room temperature.

Patience, persistence and a healthy respect for the potential electrolyte complications of any given regimen are the foundation for finding an effective antegrade enema regimen for each patient. The above tricks and modifications can help troubleshoot the most challenging patients to find an acceptable result.

References

Guest Editors’ Comments
This contribution clearly explores the different enema regimens in current use and emphasizes the trial and error approach until an appropriate one is found. It is correct to say that this is upsetting for the patient and care givers and it is at this stage that the input of a dedicated nurse specialist is vital. There can be daily support and if necessary home and hospital visits to troubleshoot problems. I do not believe it would be possible to establish a successful ACE program without such dedicated nurse specialist support.

The importance of trial and error enema administration must be emphasized in the preoperative counseling program with the patient and his caretakers. This is a family affair. We have learned a lot from families who have taught us more than we have taught them. Although we initially initiate our program with tap water instillation alone, various families independently tried additional “instillates” such as mineral oil, soap suds and split irrigations in the morning and evening. We prefer that any addition or change to the regimen be done via our nurse practitioners as the unsupervised addition of other constituents, such as salt can be disastrous in the wrong patient. Both Mr. Malone and I cannot emphasize how important the role of nurse practitioner or nurse specialist in assuring the success of any regimen. These are complicated cases, often with even more complicated social situations where constant support and guidance are mandatory.

The Button Cecostomy for Management of Fecal Incontinence
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Introduction
Fecal incontinence can be a devastating problem for children both physically and emotionally. In the pediatric population, it is frequently seen in patients with neurological abnormalities such as myelomeningocele or sacral agenesis as well and children with ano-rectal malformations such as imperforate anus or cloacal extrophy. In the past, one was often forced to resort to a colostomy. In 1990 Malone and associates revolutionized the bowel management of these patients with the introduction of the continent appendico-cecostomy for administration of an antegrade enema1. This allowed administration of an enema capable of washing out the entire colon and was not dependent on an intact sphincter. This method of enema administration was felt to be more efficient than retrograde enemas, especially in patients with sphincter incompetence. In addition, the antegrade enema can be self-administered by the patient, allowing a greater degree of independence. The initial report by Malone et al was followed by a larger series, in which they reported high patient satisfaction and an acceptable complication rate. In 1995, they also reported the feasibility of simultaneous lower urinary reconstruction2. The antegrade enema concept and Malone procedure found wide acceptance in patients with neurogenic bowel disease, anal sphincter incompetence or chronic constipation.

However for the pediatric urologist, the majority of patients with fecal incontinence are due to a neurological etiology and often would benefit from complex urologic reconstruction with a Mitrofanoff continent appendicovesicostomy3,4, rendering the appendix unavailable for a Malone procedure. It was for those patients without an appendix that the idea of using a gastrostomy button as a continent cecostomy for administration of antegrade enemas was first reported by Fukunaga in 19965. In 1999 Duel and Gonzalez reported their experience with the open placement of a standard gastrostomy button into the cecum in five patients with neurogenic incontinence and compared them to two patients who underwent a conventional Malone procedure during the same period6. In their initial experience four of the five cecostomy button patients and 1 Malone patient had postoperative complications. Three button patients developed infections at the implantation site. Two of these were responsive to local wound care, but one required a brief course of intravenous antibiotics. The fourth patient returned one month
postoperatively with abdominal pain, and was found to have a periappendiceal phlegmon, without involvement of the button site. All seven patients were free of fecal soiling, and all patients were pleased with the results. This high complication rate led to a loss of enthusiasm for the open cecostomy button procedure despite the excellent patient satisfaction with the later functioning of the appliance.

Since that time, there has developed multiple methods for providing access for antegrade enemas that address the various anatomical challenges. The difficult task today is not whether to enable antegrade enemas but how to select the best option for each patient.

The options can be first divided into continent stomas (Malone) versus appliances (cecostomy). The Malone conduit can be preformed with the appendix, reconfigured ileum or a colonic tube. The appendix, if long enough, can also be split if needed for an appendicovesicostomy. Only a short stump is necessary as the appendix can be functionally lengthened by tubularizing the adjacent cecum with a staple line and then imbricating the extension. These procedures can be preformed open or laparoscopically and can be safely combined with bladder reconstruction procedures. Caution should be used if considering the simultaneous placement of an artificial urinary sphincter due to an increased risk of infection. The appendix has been suggested to be the superior material though the alternative conduits have been successfully used. Complications include stomal stenosis, stricture, leakage, difficulty with catheterization and perforation. The most frequent complication is stomal stenosis with reoperation rates of 15-50%.

Numerous appliances can be used as cecostomy tubes. The initial appliances included gastrostomy tubes, both long g-tubes as well as low profile tubes such as the Bard gastrostomy button which is held in place with a Pezzer like tip (Figure 1) or the low profile MIC-Key button which is held with a Foley type balloon (Figure 2). The gastrostomy devices were typically placed through an open incision though there have been case reports of colonoscopic and laparoscopic placement. The devices require regular exchanges every 3-6 months which are generally well tolerated in the office setting. Frequent complications include wound infection, peristomal granulation tissue, and accidental dislodgement. Rarely there can be migration in the terminal ileum, bowel injury or occlusion.

Broviac tunneled central venous catheters have been used as a cecostomy. The tunnel helps reduce site infections but excludes simple exchange and the narrow lumen is not infrequently plagued by obstruction with inspissated stool.

Chait reported the percutaneous placement of a cecostomy tube using a novel self-retaining catheter, Trapdoor catheter (Figure 3). The 10 F catheter has a low profile external end with a low profile trapdoor cap. The internal portion is multi-coiled, like a double pig-tailed catheter, with a distal extension. The multiple coils serve to hold the cecum against the abdominal wall, yet adjust for subsequent changes in abdominal wall thickness. The catheter is placed in a two step manner with the initial percutaneous placement under fluoroscopic guidance of an 8.5 F locking pigtail catheter. Once a tract is formed, the catheter is exchanged over a guidewire. Complications include granulation tissue and accidental dislodgement.

All methods of tubed cecostomies report excellent results in terms of managing the fecal incontinence. Great care is necessary in choosing the location for the appliance as it can interfere with the comfortable wearing of clothes if it is in the path of the waistband.

Given the success reported with many of these alternatives it is clear that there is no single best answer and the choice of procedure needs to be individualized to the patient. Unfortunately the choice is most commonly based on the physician’s experience and preference. However, the concerns and values of the children and their parents have are often far different than those of their surgeon. A review of the parent’s internet bulletin board of the Pull-Through Network, a support group for parents of children with imperforate anus and similar disorders, re-
veals the kinds of concerns that parents have but may be inhibited from sharing with their caregivers. In general, parents report significant complaints with stomal stenosis and difficulty in catheterization of the Malone. Many resort to leaving a catheter or plug in the stoma when not in use to keep it open. A company in Great Britain, Medicina, even makes a silicone device called the ACE plug specifically for this use. Even if the stenosis is not significant enough to warrant revision it can often result in discomfort with catheterization. Parents with the Malone do like the appearance though parents with the cecostomy tube report that the appearance of the appliance is not of concern to the children.

Parents of children with cecostomies like the fact that its placement can be minimally invasive and the catheter can be used right away. They are bothered by the buildup of granulation tissue but have discovered many treatments including hydrogen peroxide, mercurochrome, and silver nitrate. For many parents they and the children prefer catheter insertion into the device rather than “sticking” the catheter into the child. For many, especially those with imperforate anus, the experience of rectal dilation and rectal enemas was so traumatic that they are forever wary of having to insert any tubes into their child. They also like that it is painless to use as opposed to the discomfort some children report with catheterization of their Malone. Many parents also worry about using the appendix for the Malone due to concern that a Mitrofanoff may be needed it the future.

Therefore, when considering a treatment for antegrade enema access one should consider whether this is a stand alone procedure or part of a more significant reconstruction, is there a suitable appendix in an appropriate location, will the appendix be put to better uses elsewhere, is the family willing and able to catheterize a stoma, how burdensome to the family will it be to maintain the appliance, and most importantly what is the risk tolerance and wishes of the family. Malones have more upfront surgical risks and need for revisions. Cecostomies have more risk for chronic local but non-surgical complications.

In summary, antegrade enemas constitute a significant advance in the treatment of childhood fecal incontinence due to anatomic or neurological abnormalities. There are numerous means to provide access for the administration of antegrade enemas. The Malone procedure is of particular benefit in a child undergoing an open reconstructive procedure in which the appendix, or a part of it, is available for reconstruction. The percutaneous cecostomy is of particular benefit in a child who does not otherwise require an open procedure. The open cecostomy button may have a role in the patient undergoing an open procedure though without a suitable appendix or a patient requesting a cecostomy in a location where percutaneous placement is not available. However at the heart of the decision must be the wishes of a well informed family.

References

Guest Editors’ Comments

Drs. Duel and Freedman have elegantly demonstrated the use and advantages of the cecostomy button in this contribution. There are other broader uses that are worthy of consideration. In patients with recurrent stomal stenosis some surgeons are inserting a button into the conduit following dilation and leaving it there on a permanent basis. This works well and there is no problem with granulation or leakage around the button site. John Hutson from Melbourne has actually adopted this technique when creating an appendico-cecostomy for all patients (personal communication). The use of the left sided button is also worth mentioning and this is dealt with elsewhere in this issue.

Using external devices can also be considered as “training wheels” in those patients where there is worry about the MACE procedure’s potential for success. We have also used temporary external devices in the complex patient who we might consider an alternative site to the appendiceal-cecal unit for permanent MACE placement and a more distal location in the left colon might be advantageous. Clearly a minimally invasive option is preferable in the totally dependent patient where the antegrade enema is always administered by a caretaker.

PSM
Laparoscopic and Minimally Invasive Approaches to the MACE Procedure

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Introduction

Neurogenic bowel conditions are often part of severe congenital malformations such as spina bifida, cloaca or imperforate anus. Affected patients frequently require aggressive daily bowel management to prevent constipation and continuous stool leakage. The regular application of retrograde enemas often in combination with manual disimpaction is necessary to ensure proper emptying of the rectal vault. Accessing the cecum in an antegrade fashion via a continence flap valve mechanism using the appendix as catheterizable channel has been popularized by Malone. The creation of such an antegrade continence enema (ACE) stoma involves mobilization of the caecum, ascending colon and appendix. The cecum is then imbricated over the appendix and the free end is brought to the skin. The standard open technique requires a midline incision from a few centimeter above the pubis beyond the umbilicus to allow proper bowel mobilization and stoma positioning. In most cases the ACE can be created using a three port minimal invasive technique, encompassing all the advantages of laparoscopy. In the following we review the technical aspects and complication possibilities of the laparoscopic ACE procedure.

Indications and preoperative preparation

The laparoscopic ACE is indicated for patients with chronic constipation or fecal incontinence due to neurogenic malfunction of the bowel. As with all laparoscopic approaches previous abdominal surgery with extensive adhesions or space limitations due to co-existing malformations of the spine can complicate the minimal invasive technique.

Preoperatively the patients and caretakers are counseled, that regular and correct use of the newly created stoma is mandatory to ensure long-lasting proper function. The future stoma location is discussed with the most common sites being in the right lower abdominal quadrant or preferably concealed in the umbilicus. The procedure as well as the complication possibilities, including possible open conversion are explained in detail. A preoperative bowel preparation with oral Go-Lytely, antibiotics and enemas is initiated.

Imbrication versus Non-Imbrication

Three different minimal invasive techniques have been described. Laparoscopic ACE without imbrication of the conduit; laparoscopic ACE with intracorporeal imbrication and laparoscopic ACE with extracorporeal imbrication of the appendix. Similar to all techniques is the laparoscopic mobilization of the cecum, appendix and ascending colon.

In the laparoscopic approach without imbrication, the appendix is mobilized, brought to the skin and the stoma is usually created by a V-shaped skin flap in the umbilicus. The continence of the stoma is, therefore, dependent on the ceco-appendical sphincter and intraabdominal appendiceal coaptation alone. Several studies found no difference in the continence rates between imbricated and non-imbricated stomas. Lynch et al found the success of the procedure to be more dependent on patient selection with higher continence rates in patients without colonic dysmotility disorders. Curry et al. confirmed better continence rates in patients with spina bifida and found success also to be age dependent with children older than five years tolerating the ACE procedure better. Finally compliance with the washout regimen is crucial to achieve and maintain long-lasting continence.

Surgical Technique

Latex precautions are recommended. The patient is placed in the supine position and general anesthesia applied. A Foley catheter is inserted and the patient placed in a Trendelenburg position. A broad based triangular skin flap is outlined with the base pointing lateral and slightly cranial while the tip of the flap is located just above the umbilicus. The flap is incised with a scalpel avoiding electrocautery to prevent later necrosis. The flap is freed up and the underlying abdominal fascia dissected free. A 2-0 silk is placed through the center of the umbilicus and used for traction. The skin around the umbilicus is circumferentially excised with a scalpel and the umbilical vessels are dissected to the level of the fascia using electrocautery. Redundant fat tissue is excised. A Veress needle is attached to a 5mm Step sheath and placed through the incision into the peritoneal cavity at the base of the umbilical cord structures. The peritoneal cavity is insufflated to 12mmHg. The Step sheath is dilated to 5mm. A laparoscope is inserted through the umbilical port. Two additional 5mm ports are inserted in a similar fashion under direct vision. The first port is placed at McBurney’s point and the second in the midline between the symphysis pubis and the umbilicus. The table is placed at 45 degrees Trendelenburg with 45 degrees roll to the patient’s left. The small bowel is swept medially. The appendix is mobilized on its mesentery, avoiding injury to the appendiceal artery. The cecum is mobilized superiorly. The right colon is mobilized to the hepatic flexure. Mobilization can be stopped when the cecum reaches the abdominal wall at the umbilicus. The umbilical port is removed and the abdomen desufflated. The fascial incision is enlarged about 2-3 cm above and below the umbilical access site to allow delivery of the cecum with a Babcock clamp. Holding sutures are placed in four quadrants to stretch the ventral side with the tenia in the middle. The tip of the appendix is excised. An 10-12Fr catheter is passed down the appendix to ensure patency and ease of passage. For the imbrication, using needlepoint electrocautery the serosa is marked left and right of the ventral tenia to outline the later channel. The appendix is placed against the tenia and the cecum imbricated over the appendix and its mesentery with interrupted sutures of 4/0 Ethibond with the catheter in place. Alternatively the cecum may be approximated over the appendix through appendiceal mesenteric windows. Later continence of the stoma is dependent on the length and quality of the cecal tunnel and not on the fascial or skin site fixation. Therefore, continence and ease of catheter passing of the catheterizable channel should be tested before proceeding with the operation. The
stoma has to reach the selected skin site without tension in a straight fashion avoiding kinking of the channel. Tension can cause impairment of the blood supply leading to stricture or necrosis, while kinking can make CIC difficult by causing false passages or the inability to enter the reservoir. The imbricated cecum is released back into the abdominal cavity leaving the appendix at the skin level. To avoid kinking in a long channel, the conduit can be secured to the abdominal wall with four interrupted sutures placed in quadrants around the imbricated appendix. The appendix is spatulated on the antimesenteric side and the tip of the triangular based flap is secured to the spatulated channel using absorbable sutures. The rest of the stoma is secured with interrupted sutures to the skin. The stoma is catheterized several times with a 10 to 12 french catheter to ensure ease of catheterization before the wound is closed. A 10-12 French balloon catheter is placed and secured to skin and left to drainage. The fascia is closed with interrupted sutures. The fascial opening should be wide enough to allow the channel to pass with ease to avoid stenosis, but small enough to prevent parastomal herniation. As a rule of thumb, the fascial gap should be 1.5 times the diameter of the catheterizable channel. The abdomen is re-insufflated and inspected for occult injuries and correct position of the intraabdominal ACE portion. The ports are removed under direct vision.

The port sites are closed in the usual fashion. The bladder catheter is removed after the procedure.

Postoperative care

The patients receive intravenous ketorolac for pain control and morphine is avoided to allow rapid return of bowel function. The diet is advanced as tolerated and the patients can leave the hospital when tolerating diet and oral pain control achieved. The ACE is flushed on postop day one with 50-100ml of normal saline daily and slowly advanced to the maximum volume. The caretaker and the patient are instructed in the management of the stoma catheter and washout procedure and released home with detailed instructions, follow up appointments and telephone numbers to call in case of complications or emergencies. The catheter is removed 3 weeks postoperatively in the clinic and stoma catheterization and bowel call in case of complications or emergencies. The catheter is removed 3 weeks postoperatively in the clinic and stoma catheterization and bowel washout explained and actively instructed. Regular follow up is scheduled.

Complications

Stomal stenosis is the most common problem independent from technique, tissue or site used. It can be avoided by placing a broad based skin flap into a well vascularized, widely spatulated catheterizable conduit. Despite its frequency it can be mostly treated with increased frequency of catheterizations, serial dilations or minor surgical corrections. If stenosis appears to be a problem, the authors favor placing a 3-5 cm catheter plug in the ACE tube when not in use to act as a passive dilator.

Surgical incontinence problems are always caused at the level of the imbricated tunnel. Minor leaks can be successfully treated by endoscopic injection of a bulking substance into the submucosal tunnel. In case of failure or larger leaks, open revision of the flap valve mechanism might be necessary. Other complications include false passage, stricture, stomal breakdown and prolapse. A false passage may develop secondary to kinking of an excessively long conduit or poor catheterizing angles and can be treated by a properly placed balloon catheter which is left for 10-14 days to allow the false passage to heal.

Conclusion

The laparoscopic ACE procedure can be performed in a select patient population in need for regular, aggressive bowel management who do not require concomitant open urinary reconstruction. The procedure is well tolerated with minimal intraoperative risks. Complications are common but usually easily manageable. Overall patient satisfaction remains high (>85%) indicating that patients are willing to accept a high rate of easily managed complications for an improvement or protection of lifestyle.

References


Guest Editors’ Comments

I think Drs. Frimberger and Kropp are correct to advocate the use of a valve mechanism when performing the laparoscopic ACE as in my experience leakage occurs slightly more commonly when no valve is created. The creation of this valve extracorporally is a great step forward and is to be recommended. The authors also allude to an ACE stopper and I am including a figure of the Medicina ACE Stopper (Figure 1) as this is now freely available in a number of sizes and in my experience it has significantly reduced the incidence of stoma stenosis.

Laparoscopy is a valuable asset in children who undergo urological reconstructive surgery. Steve Docimo really deserves kudos for spearheading this effort. I have been sadly disappointed when I tried to perform a LACE without imbricating the appendix in order to prevent incontinence. I perform my laparoscopic MACEs similarly to the Oklahoma group, except that I use only the umbilical port. Instead of additional port sites, I insert working instruments, only as needed, through small stab sites which leave less of a scar and to date have been complication free, using only Dermabond to oppose the skin. One valuable trick that I have incorporated in the situation where the right colon and appendix are not scarred in is to increase my umbilical port to a 10 mm STEP and then pass a large cystoscope with grasping forceps into the abdomen after initial laparoscopic inspection. I then grasp the distal appendix and gently advance it into the port and assure that I have a firm grasp on the appendix. Then the port and scope are both slowly removed as a unit which brings the appendix into the wound. After the fascia is opened further, an extracorporal antireflux mechanism is created and colon is tacked to the posterior abdominal wall.

Figure 1 - The Medicina ACE Stopper.
ACE Complications

Introduction
Since the first description of the ACE procedure in 1990 by Malone and colleagues, more than a dozen pediatric series have been published in the literature. Most investigators report a highly successful procedure that improves fecal continence and enhances the quality of life in most patients. Reports of complications have been low. These include short-term post-operative complications inherent in abdominal surgery such as wound infections and adhesive small bowel obstruction. Long term, chronic problems mainly involve stomal and catheterization difficulties. Isolated case reports of major morbidity and mortality secondary to metabolic abnormalities have been published but are considered quite rare.

Stomal Complications
The most commonly reported long-term complication in published series has been stenosis of the conduit at the level of the skin. A review of the experience of 12 institutions as well as the results of a United Kingdom questionnaire (Table 1) revealed a range of 6 to 50% (mean 22%). The etiology of stomal stenosis is most likely multi-factorial. Factors such as obesity and vascular compromise have been suggested. The rate is higher than for similarly constructed Mitrofanoff neo-urethral conduits perhaps due to the fact that it is cannulated much less often. Older patients with presumably less parental supervision were felt to be at a higher risk in a review from Barqawi et al. A significant difference in stenosis rates by type of channel (appendiceal versus re-configured ileum) has not been shown.

Table 1. ACE Stomal Stenosis Rates

<table>
<thead>
<tr>
<th>Series</th>
<th>Year</th>
<th>Patients N</th>
<th>Follow-up (years)</th>
<th>Stomal Stenosis N(%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Barqawi et al.</td>
<td>2004</td>
<td>53</td>
<td>4.0</td>
<td>14 (26)</td>
</tr>
<tr>
<td>Cascio et al.</td>
<td>2004</td>
<td>37</td>
<td>NR</td>
<td>4 (11)</td>
</tr>
<tr>
<td>Herndon et al.</td>
<td>2004</td>
<td>168</td>
<td>2.3</td>
<td>10 (6)</td>
</tr>
<tr>
<td>Tackett et al.</td>
<td>2002</td>
<td>45</td>
<td>2.2</td>
<td>10 (22)</td>
</tr>
<tr>
<td>Marshall et al.</td>
<td>2001</td>
<td>32</td>
<td>1.5</td>
<td>16 (50)</td>
</tr>
<tr>
<td>Curry et al.*</td>
<td>1999</td>
<td>273</td>
<td>2.4</td>
<td>82 (30)</td>
</tr>
<tr>
<td>Driver et al.</td>
<td>1998</td>
<td>29</td>
<td>2.3</td>
<td>11 (38)</td>
</tr>
<tr>
<td>Hensle et al.</td>
<td>1998</td>
<td>27</td>
<td>NR</td>
<td>5 (19)</td>
</tr>
<tr>
<td>Wilcox et al.</td>
<td>1998</td>
<td>36</td>
<td>3.3</td>
<td>8 (22)</td>
</tr>
<tr>
<td>Levitt et al.</td>
<td>1997</td>
<td>20</td>
<td>NR</td>
<td>2 (10)</td>
</tr>
<tr>
<td>Elsworth et al.</td>
<td>1996</td>
<td>18</td>
<td>0.5</td>
<td>2 (11)</td>
</tr>
<tr>
<td>Griffiths et al.</td>
<td>1995</td>
<td>21</td>
<td>NR</td>
<td>5 (24)</td>
</tr>
<tr>
<td>Koyle et al.</td>
<td>1995</td>
<td>22</td>
<td>NR</td>
<td>3 (14)</td>
</tr>
<tr>
<td>Squire et al.</td>
<td>1993</td>
<td>25</td>
<td>1.1</td>
<td>5 (20)</td>
</tr>
<tr>
<td>Total</td>
<td></td>
<td>806</td>
<td></td>
<td>177 (22)</td>
</tr>
</tbody>
</table>

*results of questionnaire sent to surgeons in the United Kingdom
Metabolic Abnormalities

Iatrogenic metabolic complications of enema administration in children have been well-described. Most are associated with hypertonic phosphate enemas. The majority seem to be recognized and treated without subsequent major morbidity. Risk factors include children on long-term therapy due to atonic or neurogenic colonic abnormalities, as well as those with chronic renal insufficiency, although complications have been seen in otherwise normal children. Water toxicity from high colonic tap water enemas has been reported, although a large series from Indiana has been published regarding the safety of tap water for ACE irrigations (Yerkes et al). The authors warned that periodic electrolyte evaluation is warranted and that patients using a home water softening system should be alerted to only utilize untreated water. A case of fatal hypernatremia was reported in a 4 year old boy with VATER syndrome by Schreiber and Stone (1999) that was felt to be due to variations in the amount of table salt used for the enema solution combined with recurrent anal stenosis.

Our practice has been to use isotonic saline (approximately 1.5 teaspoons table salt in 1 liter of tap water) for irrigations with an initial volume of 30 ml/kg up to a maximum of 1 liter. The regimen is then adjusted individually based on clinical response. Patients with VATER syndrome and a history of imperfect anus repair should be monitored closely for anal stenosis. Acute viral illnesses such as gastroenteritis with dehydration should prompt clinical and electrolyte evaluation in patients using an ACE with deformation of the irrigations while ill.

Rare Complications

Isolated reports in the literature have presented patients with rare but devastating complications. Tackett et al. described a patient with anal stenosis who developed peritonitis and lower extremity vascular compromise from severe constipation and required emergent total colectomy due to colonic vascular congestion. A cecal volvulus in one patient requiring a hemi-colectomy was reported by Herndon et al. Other investigators have reported cecal-flap necrosis and gangrenous channels. Perforation of the channel with intra-abdominal instillation of irrigant has been reported as a rare complication with potentially morbid sequelae (DeFoor et al.) Seven cases were identified out of 187 consecutive ACE procedures. Most were seen within the first few months after the procedure, and endoscopic management with placement of a catheter over a guide wire was a successful treatment if recognized early. Once endoscopic access is obtained, a contrast study is helpful to evaluate for bowel perforation. Wide spectrum antibiotics and inpatient observation with bowel rest is recommended until the patient is clinically asymptomatic but immediate exploration may be warranted if peritoneal signs are present.

Conclusion

ACE procedures are successful procedures with low morbidity. The main complications seen are stomal-related problems and difficulty with catheterization. However, major life-threatening issues can arise and must be recognized and promptly treated. At our institution, experienced nurse practitioners play a vital role in helping us maintain close contact with these patients while providing families with ready access for questions and problems. At each clinic visit, reinforcement of proper technique and documentation of the irrigation solution and catheterization schedule should be performed. It should not need to be stressed that these patients require lifelong close follow-up with continuous re-evaluation of the home routine and the clinical results.

References


References

Guest Editors’ Comments

This contribution covers all of the common complications of the ACE and the only one I would add is incontinence of the conduit which occurs in approximately 5% of cases even when a valve mechanism is created. I would advise caution in the use of a Lofric catheter in an ACE conduit. As when the catheter may be in place for up to an hour it can dry out leaving it stuck in the conduit and very difficult to remove.

PSM

It is important for families to understand that complications are omnipresent once a catheterizable channel of any kind is constructed. Fortunately, most MACE problems are minor, stoma related issues. Still, the author has published supporting evidence that even major problems such as perforation, can be managed with less aggressive, endoscopic approaches. We must always remember, however, that the majority of those patients with neurogenic involvement may not present in similar manner to those with sensation in the face of abdominal catastrophes. Moreover, the presence of a ventriculoperitoneal shunt adds another risk factor that requires consideration. Hence, like patients with suspected spontaneous ruptured neobladders, the involved caretakers and medical staff must be vigilant.

MAK
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      Martin A. Koyle, M.D., FAAP, FACS
      Padraig S.J. Malone, MCh, FRCSI, FRCS

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♦ ACE Complications