The Malone Antegrade Continence Enema: Single Institutional Review

Ahmad H. Bani-Hani,* Mark P. Cain, Martin Kaefer, Kirstan K. Meldrum, Shelly King, Cynthia S. Johnson† and Richard C. Rink

From the Division of Pediatric Urology, James Whitcomb Riley Hospital for Children, Indiana University School of Medicine, Indianapolis, Indiana

Purpose: We report the long-term outcome of the Malone antegrade continence enema procedure in the treatment of chronic constipation and fecal incontinence in children with neuropathic bowel.

Materials and Methods: We performed a retrospective chart review of 256 Malone antegrade continence enema procedures. Patient age at surgery, bowel segment used, location of stoma, number and type of revisions required, and fecal continence were documented and a database was created.

Results: A total of 236 patients with at least 6 months of followup are included in this report. There were 172 in situ appendicocecostomies, 23 split appendix channels, 9 appendicocecostomies with cecal extension, 22 Yang-Monti ileocecostomies and 10 colon flap channels performed. A total of 112 males (47%) and 124 females (53%) were included in the study. Median patient age at surgery was 9 years (range 2 to 36) and median followup for the entire cohort was 50 months (6 to 115). A total of 56 surgical revisions were performed in 39 patients (17%). Median time to first revision was 9.5 months (range 1 to 105). Stomal stenosis occurred in 32 patients (14%). Overall, surgical revisions were required in 14% of in situ appendix channels (24 of 172), 22% of split appendix channels (5 of 23), 33% of appendix channels with cecal extension (3 of 9), 18% of Yang-Monti Malone antegrade continence enema channels (4 of 22) and 30% of colon flap channels (3 of 10). Independent of channel technique, surgical revisions were required in 15% of umbilical stomas and 18% of lower quadrant stomas ($p = 0.516$). Two patients had minimal stomal leakage, and 94% (221 of 236) achieved fecal continence with irrigations.

Conclusions: The long-term results of the Malone antegrade continence enema channel in a large cohort of patients with neuropathic bowel and chronic constipation are encouraging. Of the patients 17% will require revision surgery, and patients/parents should be counseled accordingly. In our series in situ appendicocecostomy had the lowest revision rate on long-term followup, although the difference was not statistically significant ($p = 0.226$).

Key Words: appendix, constipation, enema, spina bifida cystica, surgical stomas

The MACE procedure has revolutionized the management and improved the quality of life of children with neuropathic bowel, and refractory constipation and fecal incontinence.[1] Since the initial description of the antegrade continence enema by Malone et al in 1990,[2] several modifications of the technique have been made, including the use of a variety of intestinal conduits in the absence of a suitable appendix. Success rates defined as partial or complete fecal continence of 57% to 100% have been reported from several centers of excellence across the Western hemisphere.[3–10] Despite this impressive outcome, the MACE procedure is not devoid of complications, requiring multiple surgical revisions in some cases. This fact underscores the importance of prolonged postoperative followup in these patients. Well-informed patients/parents, proper patient selection, meticulous surgical technique and compliance with channel irrigations are essential, and taken together will ensure minimal complication rates.

By far, stomal stenosis is the most commonly encountered complication with the MACE procedure, occurring in up to 30% of patients in one of the largest series reported to date.[4] We have previously reported our preliminary results with the MACE procedure using different intestinal segments separately.[11–13] Since we have accumulated an extensive experience with performing the MACE channels, we conducted this study with 3 objectives in mind. First, we updated our data, combining all intestinal conduits used to construct a MACE channel at our institution. In addition, we report the long-term success and complication outcomes of the MACE procedures in a large cohort of patients from a single institution. Finally, we use the outcomes of these data in patient/parent counseling regarding the likelihood of success and complications associated with the MACE procedure. To our knowledge this is the largest series in the literature to combine outcomes from different intestinal segments used to construct MACE channels.

MATERIALS AND METHODS

We retrospectively reviewed the records of 256 patients who had undergone a MACE procedure at our institution be-

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Study received institutional review board approval.
* Correspondence and requests for reprints: Division of Pediatric Urology, Indiana University School of Medicine, James Whitcomb Riley Hospital for Children, 702 Barnhill Dr., #4230, Indianapolis, Indiana 46202 (telephone: 317-312-1795; FAX: 317-274-7481; e-mail: banihana@iupui.edu).
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tween 1997 and 2006. Of these patients 236 with at least 6 months of followup are included in this study. Patient age at surgery, diagnosis, bowel segment used, location of stoma, number and type of revisions required, and fecal continence were documented and a database was created.

Overall, 172 in situ appendicocecostomies, 23 split appendix channels, 9 appendicocecostomies with cecal extension, 22 Yang-Monti ileocecostomies and 10 colon flap channels were performed. Among the patients 112 (47%) were male and 124 (53%) were female.

Channel revisions were classified as either skin level, requiring redo, wide base V-flap at the level of the skin or subfascial, requiring more extensive revision below the level of the fascia. The latter may include cases revised secondary to an extensive stenosis extending beyond the fascial level or revision of the entire channel, including the use of a different intestinal conduit to construct a new MACE channel in case of complete channel obliteration.

We start our MACE washouts in the postoperative period with the return of bowel activity and tolerance of oral intake. Tap water is our preferred washout solution, and typically we start it at 50 ml nightly. This volume is then increased by 50 ml every third night until the continence goal is achieved. Continence was defined stringently as the absence of stool accidents during a 1-year interval. Cases of stool incontinence due to gastroenteritis or medications were excluded from study.

To evaluate the association between stomal location and revision, a chi-square test was done. To assess test association between type of channel and revision, stenosis, leakage and obliteration, Fisher's exact tests were performed. A p value of less than 0.05 was considered statistically significant.

RESULTS

This study included 236 patients with at least 6 months of followup. Median patient age at surgery was 9 years (range 2 to 36), and median followup for the entire cohort was 50 months (6 months to 9.6 years). There were 112 males (47%) and 124 females (53%). Etiology of refractory constipation and types of MACE channel used are outlined in tables 1 and 2, respectively. Data on the status of fecal continence with channel irrigations at last followup were available in all patients. Median volume of tap water colonic flushes was 642 ml (range 100 to 1,000). A total of 221 patients (94%) were completely continent.

TABLE 1. *Etiology of chronic constipation*

Diagnosis	No. Pts (%)
Neurogenic:	
Myelomeningocele	199
Sacral agenesis	8
Lipomeningocele	7
Spinal cord injury	6
Cerebral palsy	3
Tethered spinal cord	2
Sacrococcygeal teratoma	1
Transverse myelitis	1
Total	227 (96)
Anorectal malformation:	
Imperforate anus	3
Persistent cloaca	1
Total	4 (2)
Other:	
Posterior urethral valves	2
Prune belly syndrome	1
Hinman syndrome	1
Functional constipation	1
Total	5 (2)

TABLE 2. *Summary of all surgical revisions*

Channel	No. Skin Level	No. Sound Dilation	No. Subfascial	No. Stomal Leakage	No. Prolapse	Total No.
In situ	20	4	6	4	1	35
Split appendix	3	1	2	1	0	7
Appendix with cecal extension	1	0	1	0	0	2
Yang-Monti	6	0	2	1	0	9
Colon flap	0	0	1	2	0	3
Totals	30	5	12	8	1	56

Channel Related Revisions

A total of 56 surgical revisions were performed in 39 patients (17%). Median time to surgical revision was 9.5 months (range 1 to 105). Two patients from the in situ appendicocecostomy group had periodic difficulty catheterizing the channels but did not require surgical intervention. Table 2 summarizes the channel related revisions.

Subfascial Revisions

Subfascial revision was required in 12 patients (5%). Complete channel obliteration was responsible for 4 revisions, with 1 patient requiring an end colostomy, 1 receiving a new Monti ileocecostomy and 2 awaiting redo MACE channels. Channel stenosis below the fascia occurred in 4 patients, and in all the channel was salvageable by continuing the dissection until a healthy mucosa was encountered. Two patients required open revision for constant stomal leakage in the early postoperative period where dehiscence of the cecal wrap was noted. One patient suffered recurrent channel stenosis and underwent a successful redo Yang-Monti MACE. One patient underwent end colostomy for failure to achieve fecal continence with MACE irrigations.

Stomal Stenosis

Among the entire cohort stomal stenosis developed in 32 patients (14%, table 3). There was not a statistically significant association between channel type and the development of stenosis ($p = 0.709$). Of the surgical revisions for stomal stenosis 30 were at the skin level, 4 were subfascial and 5 required dilation of the stoma over a guidewire. Stomal revision was successful without recurrence in all but 3 patients, of whom 2 in the in situ group had complete obliteration of the MACE channel (1 eventually was treated with an end colostomy and 1 underwent complete revision with a Yang-Monti ileocecostomy) and 1 in the Yang-Monti group underwent a second Monti MACE channel reconstruction.

Stomal Leakage

Among the entire cohort 7 patients had stomal leakage (3%). Two of these patients had minimal stomal leakage. There was not a statistically significant association between the channel and stomal leakage ($p = 0.263$). Eight revisions

TABLE 3. *Summary of required surgical revisions among different MACE channels*

	In Situ	Split	Cecal Extension	Monti	Colon Flap	Total No.	p Value (Fisher's exact test)
No. pts	172	23	9	22	10	236	
No. revisions needed (%)	24 (14)	5 (22)	3 (33)	4 (18)	3 (30)	39 (17)	0.266
No. revisions performed	34	6	3	9	4	56	
No. stenosis (%)	23 (13)	4 (17)	1 (11)	4 (18)	0 (0)	32 (14)	0.709
No. stomal leak (%)	4 (2)	1 (4)	0 (0)	1 (5)	1 (10)	7 (3)	0.263
No. channel obliteration (%)	2 (1)	1 (4)	2 (22)	0 (0)	2 (20)	7 (3)	0.002

were required in 7 patients who had stool leakage from the MACE stoma. A dehiscence of the cecal wrap was observed in 2 patients, and they had a successful outcome after open revision. Five patients underwent channel injections with dextranomer/hyaluronic acid copolymer, of whom only 1 had a successful outcome. Of the remaining 4 patients who underwent Dx/HA injection 2 continue to experience occasional minor stool leakage and 2 are awaiting surgical revision of the MACE channel.

Channel Obliteration

Complete MACE channel obliteration occurred in 7 patients (3%). There was a statistically significant association between type of channel and channel obliteration ($p = 0.002$). The incidence of channel obliteration was significantly less in the in situ channels compared to cecal extension and colon flap based MACE channels. Channel obliteration was secondary to noncompliance with catheterization in 4 patients, multiple false passages in 2, and channel necrosis from poor blood supply in 1 who underwent a colon flap MACE.

Stomal Prolapse

Of the entire cohort stomal prolapse developed in only 1 patient in the in situ appendicocecostomy group (0.4% overall). This patient underwent surgical revision at the skin level for cosmetic reasons.

Stomal Location

Abdominal stomas for the MACE channels were positioned in the umbilicus in 108 patients (46%) and in a lower quadrant site in 128 (54%). Surgical revisions were required in 15% of umbilical stomas and in 18% of lower quadrant stomas ($p = 0.516$).

Continence Outcome

Of the entire cohort 221 patients were continent per stoma and per rectum (94%). Of the remaining 15 patients with incontinence (6%) 4 continue to have stomal leak of stool or gas, of whom 2 have had some improvement with Dx/HA channel injections and 2 are currently being considered for MACE revision. The remaining 11 patients continue to have stool accidents per rectum.

DISCUSSION

In the last 3 decades significant strides have been made that have revolutionized the treatment of urinary and bowel incontinence in patients with neuropathic bladder and bowel. The Mitrofanoff principle for continent catheterizable urinary stomas described in the 1980s has been adapted for administration of a large volume antegrade enema through a continent channel. The initial description of the Malone antegrade continence enema in 1990 involved creation of a nonrefluxing channel by reversing and reimplanting the appendix into the cecum.[2] Since that time, several modifications in surgical techniques have been made. At our institution it has been our preference to leave the appendix in situ, and to create a cecal wrap through windows in the mesoappendix as described by Rink et al.[14]

The appendix continues to be the most widely chosen channel for the MACE procedure among pediatric surgeons. However, in the absence of the appendix other alternatives using the Mitrofanoff principle have been described to allow daily antegrade enema access to the colon. Such alternatives include use of a cecal stapling technique to extend the functional length of the appendix,[15] use of a reconfigured small piece of small intestine as described by Monti et al[16] and Yang,[17] and use of a tubularized colonic flap.[18] Moreover, open or percutaneously inserted cecostomy tubes have had results comparable to the original Malone procedure.[19] However, some fecal incontinence may be expected around the cecostomy device, and tract and cuff infections have been reported in percutaneously tunneled catheters in 12.5% of the patients.[20] In our cohort of 236 patients only 4 (2%) had postoperative wound infections, of which 2 were stomal abscesses drained at the bed site and 2 were intra-abdominal loculated abscesses treated successfully with percutaneous drain placement.

Since we have accumulated significant experience with the MACE channel in a large cohort of patients with neuropathic bladder and chronic constipation, we conducted this study with 3 main objectives in mind. First, we wished to update and combine all MACE procedures performed at our institution using different intestinal segments. In addition, we wanted to report the long-term outcomes and complications associated with the MACE procedure in a large cohort of patients from a single institution. Finally, we wished to provide suitable data for patient/parent counseling regarding the likelihood of channel related complications and the need for revision surgery.

The fecal continence rate in our series of 94% compares favorably to rates reported in the literature, which range from 57% to 100%. In our study continence was defined stringently as the absence of stool accidents during a 1-year interval.

Overall, 56 channel related surgical revisions were required in 37 patients (16%), with stomal stenosis being the most commonly encountered reason for surgical intervention. A stomal stenosis rate of 30% was reported in one of the largest collective series to date.[4] Our low stomal stenosis rate of 14% compares favorably with what has been reported in the literature. We believe that this outcome is related to 2 main factors. First, we routinely create wide based V-shaped skin flaps when maturing MACE channels, and al-

low externalization of some of the bowel mucosa. Also, by performing an in situ technique coupled with cecal imbrication through windows in the mesoappendix, preservation of the cecal blood supply to the base and apex of the appendix, respectively, is achieved.

Stomal leakage occurred in 3% of our cohort. In 2 patients stool leakage was identified in the immediate postoperative period, and it was apparent that the cecal plication had come undone at the time of open intraperitoneal revision. Following cecal reimbrication, both patients had a successful outcome and are completely continent. Among the remaining 5 patients Dx/HA injections of the channel in an attempt to control stool or gas leakage were successful in only 1.

We currently do not believe that injecting the MACE channel with bulking agents is effective in patients with stomal leakage. It is noteworthy that although injecting a foreign body in a contaminated field such as the MACE channel carries the risk of infection in the submucosal space, we did not experience any complication related to the Dx/HA injection. Our practice has limited this option to patients with stool or gas leakage who are unwilling to proceed with open MACE revision. Furthermore, meticulous surgical technique using permanent sutures and ensuring good seromuscular bites during cecal wrapping is critical during the creation of MACE channels.

Total channel obliteration occurred in 7 patients (3%), among whom noncompliance with channel catheterization was the main etiological factor in 3 (43%). Multiple false passages were responsible for channel obliteration in 3 patients, and channel necrosis from poor blood supply occurred in 1 patient who underwent a colon flap MACE. Our preferred technique for the colon flap is a transverse, laterally based pedicle that uses the posterior cecal artery for the cecal flap, the middle colic artery for the transverse colon flap or the left colic artery for the descending colonic flap.[11] Mucosal prolapse occurred in 1 patient who underwent revision for cosmetic reasons.

We recognize that our study is limited because the majority of our patients underwent in situ appendicocecostomy for MACE channel construction, making interpretation of outcomes between different intestinal segments a challenging task. Nonetheless, the appendix continues to be our segment of choice for MACE construction even if a concomitant continent catheterizable channel is needed for bladder drainage. In the latter case the options are to split a long, well vascularized appendix and use the distal end as an appendicovesicostomy, or to use the appendix for MACE construction and a Yang-Monti tube as an ileovesicostomy for the bladder portion of the procedure. When the distal portion of the appendix is used as an appendicovesicostomy the presence of a reliable blood vessel to the proximal appendix and a body habitus of the patient that allows the appendix to be brought to the skin level rather than the length of the remaining appendix determine whether a cecal extension is a reasonable option. In this scenario a stapled cecal extension using a stapling device as described by Sheldon et al can be used.[15] In the absence of the appendix a MACE channel constructed from a Yang-Monti small bowel segment is preferable to a colon flap, since the former has more of a defined and predictable blood supply.

CONCLUSIONS

In our experience the long-term outcome of the MACE procedure in a select cohort of patients with chronic constipation and fecal incontinence is encouraging. Preoperative patient/parent counseling is of paramount importance, and should include expected outcomes and the potential need for additional surgery. This preoperative education should stress the vital role of compliance with catheterization, as well as the potential need for revision surgery. In our series in situ appendicocecostomy had the lowest revision rate at long-term followup, and it continues to be our segment of choice for the MACE procedure when available.

Abbreviations and Acronyms

Dx/HA = dextranomer/hyaluronic acid
MACE = Malone antegrade continence enema

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