The Malone Antegrade Continence Enema: Single Institutional Review

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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.
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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