Malone antegrade continence enema in children with urinary and intestinal neurogenic disease

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ABSTRACT

Chronic constipation and fecal incontinence are complaints found in a large number of children. Diseases such as spina bifida, myelomeningocele, anorectal malformations, infantile cerebral palsy, etc., present these conditions and at the same time can include pathologies such as neurogenic bladder and urinary incontinence.

Objective: The purpose of the present study is to describe the progress of pediatric patients during 7-year follow-up who underwent Malone antegrade continence enema surgical procedure and its impact on their quality of life.

Results: From October 2001 to July 2008, six pediatric patients (3 boys and 3 girls) with myelomeningocele, chronic constipation, neurogenic bladder and fecal and urinary incontinence underwent Malone procedure, augmentation cystoplasty and Monti channel for the Mitrofanoff procedure. Complications were surgical wound dehiscence in one patient and stomal stenosis in another. Fecal continence was achieved in 5 patients and great improvement of chronic constipation in all patients.

RESUMEN

El estreñimiento crónico y la incontinencia fecal son padecimientos que aquejan a gran número de niños; los trastornos como la espina bífida, mielomeningocele, malformaciones anorrectales, parálisis cerebral infantil, y otros más, son causa de dichas entidades, junto con enfermedades como la vejiga neurógena y la incontinencia urinaria.

Objetivo: Describir la evolución en pacientes pediátricos sometidos al procedimiento quirúrgico de Malone para el enema anterógrado continente y su efecto sobre la calidad de vida en un periodo de siete años de seguimiento.

Resultados: El protocolo se extendió de octubre de 2001 a julio de 2008, con seis pacientes pediátricos, tres varones y tres mujeres, con mielomeningocele, estreñimiento crónico, vejiga neurógena e incontinencia fecal y urinaria, sometidos al procedimiento de Malone, cistoplastia de aumento y un canal de Monti según el principio de Mitrofanoff. Las complicaciones que se presentaron fueron dehiscencia de la herida quirúrgica en uno de ellos y estenosis del estoma en otro. Consiguieron la continencia fecal cinco de ellos y se observó una gran mejoría del estreñimiento crónico en todos.
Conclusions: Because of the positive results that have been observed in regard to patient quality of life, Malone procedure is a highly recommendable surgical intervention in the selected patient.

Key words: Malone procedure, antegrade continence enema, chronic constipation, fecal incontinence, Mexico.

INTRODUCTION

Twenty-five percent of clinical problems in pediatric urology are the result of lesions affecting the lower urinary tract and these include neurogenic bladder dysfunction in children. The development of surgical modalities for treating urinary and fecal incontinence spectacularly modified the way in which this group of children was traditionally treated.1

A large number of neurological diseases in pediatric patients that result in neurogenic bladder are accompanied by multiple disorders in other apparatuses and systems. Intestinal function is specifically affected and consequent motility problems make chronic constipation a common disorder in these children resulting in frequent visits to the emergency room for abdominal pain and fecal impaction.1-4

Abnormal defecation is very disruptive for patients and their families. Children with constipation are a source of continuous frustration for themselves, their families and their doctors. Unresolved constipation can lead to fecal retention and impaction and finally turn into incontinence. In the large majority of patients the cause of constipation is unclear and so is called functional constipation, unlike constipation that can be attributed to an underlying neurological problem. Functional constipation in children is defined as constipation not associated with congenital abnormalities and it can be looked at as an inability to complete the defecation process.5-7 There can be many organic causes of chronic constipation including those of neurologic origin. Medullar abnormalities such as myelomeningocele, sacral agenesia and imperforated anus can lead to chronic constipation. However, the mechanism by which it is produced is unknown.

Vagus nerve damage produces slow right colonic movement. Medullar damage above the sacral nerves is manifested in alterations of colonic motility of the left side and medullar injury at T-9 to S-2 (central sympathetic) and S-2 to S-4 (parasympathetic) produces slow rectosigmoid intestinal movement.9

Chronic constipation together with fecal incontinence or encopresis and urinary incontinence largely contribute to diminishing quality of life in these patients. Fecal incontinence can be more debilitating from a social viewpoint. Fecal soiling is a common characteristic of chronic constipation and leads to significant psychological disorders with repercussions in social and educational development and is also a source of great anxiety and worry for parents.1,7,10

Even if these diseases do not directly pertain to the genitourinary tract, the urological pediatrician is involved in making treatment decisions and in their integral management. Such decisions can involve indicating alternate surgical procedures for managing constipation and fecal incontinence that have been refractory to conservative treatment that can be performed during the same surgery in which bladder enlargement is carried out, when different canalizations for clean intermittent catheterization (CIC) are done or during continence surgeries. Such decisions take advantage of the correct moment for adding a beneficial procedure to the integral treatment of the patient with intestinal and genitourinary neurogenic disease.9

Since 1990 Malone Antegrade Continence Enema (MACE) procedure has proved to be an excellent alternative for quality of life improvement in children with neurogenic intestine, refractory constipation and fecal incontinence in spite of the reported complication percentage (17% in the most numerous series) principally involving stoma stenosis.

Surgical modifications such as the use of a variety of intestinal ducts and techniques for forming catheterizable channels when there is no appendix have been made over the past years for the purpose of improving results and minimizing complications.8,10,11,12
The present article describes a total of six cases with MACE procedure using the original Malone technique and evaluates patient progression and satisfaction compared with the results from studies reported in the international literature.

**MATERIALS AND METHODS**

From October 2001 to July 2008, six patients from the Hospital de Pediatría of the Centro Médico Nacional Occidente (CMNO) diagnosed with myelomeningocele, neurogenic bladder and intestine, severe chronic refractory constipation and fecal incontinence underwent Malone technique procedure together with augmentation cystoplasty and bladder catheterizable channel for neurogenic bladder. All patients had a history of abdominal pain, encopresis, fecal incontinence ending in daily diaper use, intestinal subocclusion and fecal impaction that caused them to be admitted into the emergency room at least four times for manual de-impaction.

Patients were 3 boys and 3 girls with a mean age of 12.6 years. All six patients had constipation refractory to conventional treatment and were diagnosed with constipation secondary to neurological disorder from myelomeningocele confirmed by magnetic resonance together with urinary incontinence disorders due to neurogenic bladder. Diet, defecation behavior, enemas, medication and laxatives were consistent among the six patients.

Surgical procedure was indicated based on clinical diagnosis criteria for chronic constipation and imaging studies related to urinary disease. Informed consent was obtained from the parents and patients and the surgery, MACE, advantages and disadvantages of clean intermittent catheterization as well as the most frequent complications were all thoroughly explained.

Appendicoccecostomy in situ was performed on all patients under the Malone principle through appendicular mobilization. Overlapping seromuscular Lambert stitches from one side of the tenia to the other were made using 00 vicryl as an antireflux mechanism and the appendix was wrapped. Mesenteric vasculature was preserved and exteriorization through the abdominal wall in the right iliac fossa was carried out with the creation of a splinted stoma with a Nelaton catheter that was attached to the skin and aponeurosis from the mucosa with simple 00 vicryl stitches (Images 1, 2 and 3).

All patients underwent intestinal resection for bladder enlargement (ileum was used in 4 patients and sigmoid colon in 2) and Monti channel elaboration from the ileum in order to reimplant in the bladder with exteriorization at the umbilical cicatrix utilizing the Mitrofanoff principle. Urinary continence surgery was also performed in one of the patients.

With 7-year follow-up, enema regimen was morning applications of 50-70 mL phosphate solution in one patient and natural water in the others. Patients were asked how they felt after MACE procedure, evaluating quality of life with the following qualifications: a) worse b) the same c) better d) very good and e) excellent.
RESULTS

The six patients had a mean 7.3 day postoperative hospital stay. One patient presented with surgical wound dehiscence and was re-operated on for surgical cleaning and closure. Another patient presented with stenosis at the mucocutaneous junction two years after surgery which was resolved by surgical dilatation with scalpel and did not present again.

After enema application, generally with 60 mL of natural water, intestinal voiding usually presented after 40 minutes in 4 children and 60 minutes in the other 2. All stomas remained continent and free from gas (Image 4).

Only one of the patients remained dry and clean after surgery. Four of the others had improvement in encopresis and incontinence after the first year of surgery, with a reduction of daily stool leakage to one or two times per week. Only one patient continued with fecal incontinence at least once a day.

None of the patients complained of pain during enema application which have been continued up to the present time as have the clean intermittent catheters. All six patients showed significant improvement in relation to constipation and remission in episodes of abdominal pain. No further emergency room treatment for pain or fecal impaction has been necessary.

In response to the quality of life question one patient said she felt good and the other five said very good.

One of the patients presented with problems of urinary incontinence and malfunction of the Heyer-Pudenz valve unrelated to the Malone procedure for which she was admitted to the hospital on various occasions.

DISCUSSION

Since its design and performance in 1990, Malone antegrade continence enema surgery has become an important therapeutic recourse, perhaps comparable to the revolutionary Mitrofanoff in 1980 for management of neurogenic intestine and neurogenic bladder.\(^{13}\)

Much has been written about this procedure and it has been performed in various countries, including Mexico. Even though the present study has a small number of patients, it contributes to furthering knowledge about the technique and its results add to the experience acquired not only in the area of genitourinary surgery but also in fecal incontinence and chronic constipation therapies, all of which together contributes to improving the quality of life of these patients.

MACE was originally developed to improve fecal incontinence management but it is being used more and more in severe constipation management when medical therapy has failed. Because of the positive results seen with its use, especially in patients with neurogenic intestine, the procedure has been indicated and applied in diseases such as anorectal malformation, Hirschsprung’s disease, spina bifida and myelomeningocele, spinal cord damage, severe perineal damage, neurogenic disorders, cerebral palsy and idiopathic or functional constipation.\(^{12,13}\)
In addition, there have been numerous modifications to the Malone surgical procedure over the past years in relation to anatomical resources available in each patient and his or her necessities. Specifically one resource is the availability or absence of a natural anatomical channel such as the appendix. New Yang-Monti- catheterizable channel reconstruction techniques from the digestive tract have been developed. These techniques have also been attempted towards the intestinal and bladder cavities, reproducing Malone and Mitrofanoff procedures respectively, dividing the appendix as indicated by Kajbafzadeh and Chubak in 2001. 14–16

Of all the techniques available, in the present cases it was necessary to make a new channel since the cecal appendix was reserved for the Malone technique. As indicated in the original technique, the simplest and easiest option was to modify its implantation in situ, plus it was the technique the surgeon was most familiar with. In the most numerous MACE series reported by Bani-Hani in 2008, the surgical modality used in the present study (appendicocoeostomy in situ) had the lowest incidence of complications and thus is considered to be the technique of choice, when available, for MACE procedure.

Relation to and congruency with the most numerous evaluated studies has been observed. Mucocutaneous stenosis of the stoma, as referred to by Castellán in 2004, appears to be the most common late postoperative complication (up to 21%) and the main factor resulting in additional surgery for dilatation or surgical revision (up to 17%) as published by Bani-Hani et al. in 2008.11,17

In relation to the material used for the enemas, there was no defined pattern or protocol for determining required quantity or the number of enemas. Application was managed according to dose response of the six patients. Only one patient was given enemas with saline solution at 0.9% and the other patients were given natural water once a day in the mornings. Response was good with evacuations 40 and 60 minutes after enema. There was no need to increase volume and water was considered the ideal material for its low cost and effectiveness.

In relation to fecal continence, results were positive and satisfactory and concurred with scientific reports referring to better response with MACE procedure in cases of neurogenic intestine compared with patients with idiopathic constipation. In the present study the greatest successful impact of the procedure was seen in the big improvement in continence, reduction of abdominal pain and remission of fecal impaction events and reflected in the opinion of the patients when asked to rate quality of life post-procedure.12

The authors agree with Cascio et al. in that part of the success of the MACE procedure has to do with the age of the patient. Age above 12 years is the ideal age in the select patient with a certain level of maturity, conviction and willingness. For there to be positive results and quality of life improvement, it is fundamental for the patient, parents and family to thoroughly understand the procedure.9

In conclusion, surgical procedure using the Malone technique is highly recommendable in the select patient taking into account patient maturity, conviction and willingness to go through with the procedure. Recent articles have made reference to good results in fecal continence even in patients with functional constipation, further broadening the horizon for its application.9

BIBLIOGRAFÍA