Complex Pediatric Constipation: A Case Study of a Child With a Malone Appendicostomy

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KEYWORDS

Appendicostomy, case study, constipation, Malone appendicostomy, pediatrics

Constipation is the chief complaint at approximately 3% to 5% percent of pediatric outpatient clinic visits (Borowitz, 2016). This diagnosis is challenging at times and is defined by the North American Society of Gastroenterology, Hepatology, and Nutrition as "a delay or difficulty in defecation present for two weeks or more and sufficient to cause significant distress to the patient" (Borowitz, 2016, p.1). Additionally, up to 25% of all visits to pediatric gastroenterologists are for constipation-related complaints (Nurko & Zimmerman, 2014). The patient presented in this case study has a diagnosis of complex pediatric constipation.

CASE PRESENTATION

A 9-year-old White male with Trisomy 21 presented with the chief complaint of "chronic constipation and fecal incontinence." After years of conventional treatment modalities, the family presented to a pediatric colorectal service for consultation.

The child was born vaginally at 39 weeks to a gravida 1, para 1 mother. The child's gestation and birth were uncomplicated. At birth, he presented with clinical features consistent with Trisomy 21, and a non-mosaic diagnosis was later confirmed by genetic testing. The child's infancy and toddlerhood were negative for many of the associated diagnoses of Trisomy 21 including heart disease, thyroid disease, feeding intolerance, hearing loss, or abdominal distention (Ivan & Cromwell, 2014). He was exclusively breastfed for the first year of his life with no report of constipation. Milestones in gross motor function and speech were delayed, and hypotonia was significant. Pressure equalizer tubes inserted at 15 months were successful in reducing the number of recurrent acute otitis media episodes.

From a gastrointestinal perspective, the child received lactulose every 3 to 4 weeks for mild constipation with good success during the second year of life. This regimen resulted in the anticipated daily stool. The family history was negative for Crohn's disease, ulcerative colitis, irritable bowel, Hirschsprung's disease, and chronic constipation. The child was followed up at a Down syndrome clinic, where a plan of care was developed to manage his apraxia, significant cognitive delay, and global delays. During this period, he received regular occupational therapy, physical therapy, and speech therapy as recommended by his providers. Immunizations were administered on a predicted schedule.

Years 3 and 4 of life were clinically significant for the child's family and pediatrician because his constipation was frequent, lasting up to 3 days. Treatment for his constipation initially included increasing doses of oral

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Conflicts of interest: None to report.

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J Pediatr Health Care. (2017) ■, ■-■.

0891-5245/$36.00

http://dx.doi.org/10.1016/j.pedhc.2017.05.001

laxatives with subsequent enemas as needed. Although evacuation of the colon was successful with this regimen, the resulting fecal incontinence proved to be socially unacceptable and was dramatically interfering with the child and family's quality of life. The patient's constipation and fecal incontinence were further supported with medical management, pharmacologic therapies, and behavioral modifications recommended by his pediatrician. His constipation proved to be nonresponsive to these therapies, and the child was referred to a pediatric gastroenterologist when he was 5 years old.

The gastroenterologist performed intestinal and rectal biopsies, the results of which were negative for Hirschsprung's disease. Celiac disease was also excluded. At this time, it was determined by the gastroenterologist that the child's constipation was a result of his significant hypotonia. High-dose laxatives were recommended by his gastroenterologist, followed with serial abdominal x-ray images to evaluate complete evacuation of stool from the colon. The goal of the gastroenterology team was to increase laxative doses until daily evacuation of the colon was achieved. To accomplish this goal, daily aggressive doses of oral lactulose and polyethylene glycol were required. This bowel regimen resulted in the goal of a daily stool; however, the stools were of diarrhea consistency, occurred one to two times per day, and were unpredictable in timing. Despite parental efforts to toilet train, the child's cognitive delays coupled with unpredictable loose stools made continence an impossibility. His incontinence made social interactions difficult at times and made mainstream schooling a challenge. The adult diaper undergarments, which only partially contained the bowel movement, made it problematic to plan or participate in routine daily activities without considering where to change and clean up after an incontinent event in a child who was now 9 years old.

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Because of the boy's deteriorating quality of life, his family sought counsel from their pediatrician to discuss further options. The child also had a long history of urinary incontinence that had been attributed to his hypotonia and cognitive delays. In an effort to rule out other pathology that could be a contributing factor to both of these issues, magnetic resonance imaging was recommended and showed a tethered spinal cord. Because of the possibility that the tethered cord could be contributing to his urinary and fecal incontinence, a referral to a pediatric neurosurgeon was made.

A laminectomy was recommended by the neurosurgeon to provide a potential for improved urinary and fecal continence. A period of 12 months was allowed after laminectomy for healing and nerve regeneration. After surgery, the patient was still able to have daily loose bowel movements with pharmacologic intervention, yet despite the successful laminectomy, urinary and fecal incontinence persisted. The family described a deteriorating quality of life as he grew and his life expectations broadened.

At the age of 9 years, and after 6 years of aggressive medical management for his chronic constipation, a referral to a pediatric colorectal specialist was made to help with fecal continence. The colorectal specialist and colorectal team provided a multidisciplinary approach with an organized protocol to manage his pediatric incontinence, colonic dysfunction, and constipation. The goal of the colorectal team was *fecal continence* defined as "the ability to have voluntary bowel movements and to predictably evacuate stools when socially acceptable" (Eradi et al., 2013, p. 2296).

The colorectal team evaluated the boy's history and previous examination findings. An abdominal radiologic study with contrast of the abdomen was preformed, showing a segment of megacolon and confirming the diagnosis of functional constipation. Colonic manometry, a study whereby a catheter is placed through the rectum and into the colon, evaluated peristalsis and determined colonic motility. At the conclusion of testing and evaluation, the colorectal team determined that pharmacologic management alone would not achieve the desired results of fecal continence and made the recommendation for a Malone appendicostomy and excision of megacolon to enable the child to achieve predictable daily bowel movements without fecal incontinence between bowel movements.

SURGICAL MANAGEMENT

The Malone appendicostomy, developed by Dr. P. S. Malone in 1990, is a surgical procedure that provides a means of administering antegrade enemas for the treatment of functional bowel incontinence and chronic constipation. The Malone appendicostomy enables the patient in nearly all cases to achieve scheduled and consistent bowel movements (Chatoorgoon et al., 2011). In the pediatric population, this procedure has improved the quality of life for children with anorectal malformations, spinal abnormalities, pelvic tumors, severe chronic constipation, and other diagnoses that result in fecal incontinence (Chatoorgoon et al., 2011). The Malone appendicostomy's ease of use and potential for independent administration have been shown to significantly increase the quality of life for families and children afflicted with fecal incontinence (Chatoorgoon et al., 2011; Har, Rescorla, & Croffie, 2013; Joon-ha & Kurzrock, 2011).

The Malone appendicostomy procedure mobilizes the appendix to the abdominal skin surface, creating a fistula between the skin surface and, most often, the