



The morbidity of constipation in patients with anorectal malformations

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Abstract

Background: Constipation in anorectal malformations (ARM) is extremely common, particularly in the lower types. Failure to adequately treat it can lead to significant morbidity.

Methods: From our series of over 2000 patients with ARM, we reviewed 398 with good prognosis for bowel control and a tendency toward constipation; rectoperineal fistula (63), rectovestibular fistula (114), rectobulbar urethral fistula (104), imperforate anus with no fistula (46), rectal atresia or stenosis (9), and cloaca with a common channel below 3 cm (62). Those lost to follow-up, not yet toilet-trained (<3 years old), or with poor prognostic features were excluded. We compared morbidities in patients we operated on and managed primarily (group A, n = 268) to those managed at other institutions who suffered from constipation or incontinence and were referred to us for treatment (group B, n = 130). Those we managed primarily were subjected to an aggressive senna-based laxative program, started after their primary repair or after colostomy closure.

Results: Morbidities associated with constipation were higher in the referral group and included fecal impaction (7.8% vs 38.5%), overflow pseudo-incontinence (4.9% vs 33.8%), and megacolon (14.6% vs 54.6%). A loop or transverse colostomy (4.9% vs 9.2%), stoma or anorectal stricture, or a stenotic fistula (2.2% vs 28.5%) were contributing factors. Adequate laxative treatment with, in certain cases, resection of a megarectosigmoid (2.6% vs 23.1%) enabled many pseudo-incontinent children to achieve bowel control (reported previously). Unneeded colorectal biopsies (1.9% vs 16.2%), Hirschsprung's-type pullthroughs (0% vs 3.1%), and, in retrospect, unneeded antegrade continent enema procedures (0% vs 3.1%) were higher in Group B. Overall, 19.8% of Group A and 66.2% of Group B experienced constipation-related morbidities.

Conclusion: The morbidity of constipation in ARM includes fecal impaction, megacolon, incontinence, and performance of unneeded surgeries. Inadequate treatment, the type of the original colostomy, and postoperative anal or stomal stricture as well as stenotic fistulae were key contributing factors. Children with ARM and good prognosis for bowel control are at the greatest risk for severe constipation and its consequences. With recognition and aggressive, proactive treatment, we have found that these morbidities can be reduced.

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Constipation in anorectal malformations (ARM) is extremely common, particularly in the lower types with good prognosis for bowel control [1-5], which include imperforate anus with no fistula, rectal atresia or stenosis, rectoperineal fistula, males with rectourethral fistula at the bulbar level, and females with rectovestibular fistula or cloaca with a common channel of less than 3 cm. Constipation in its most serious forms can produce overflow pseudo-incontinence, frequently related to a dilated rectosigmoid [6-10]. Failure to recognize or adequately treat this associated hypomotility in ARM patients can lead to significant morbidity [4,6-10], which we believe is largely preventable.

Minor childhood constipation is often dismissed as diet-related or behavioral. Diet does have some impact on colonic motility, and treating constipation with stool softeners may be sufficient in mild cases, but their therapeutic value is negligible in the most serious forms of constipation. The rectosigmoid normally stores the stool, and active peristaltic waves occur every 24 to 48 hours indicating that it is time to empty. A normal individual, with an intact continence mechanism, feels this sensation, contracts their sphincter, and then decides when to relax the voluntary sphincter mechanism to allow evacuation. The ability to have a voluntary bowel movement depends on the combined integrity of the anal canal, sphincters, and motility [11]. Patients with anorectal malformations lack a normal anal canal, have variably deficient sphincters, and have an accompanying motility disorder, usually hypomotility [8,11]. Management for this depends on treatment of constipation with laxatives to provoke peristalsis and to overcome the dysmotility. A child with a correctly repaired good-prognosis anorectal defect, normal lumbosacral spine, good sphincters, and an intact rectosigmoid should be fecally continent [1], so that if such a patient is soiling, we can expect that they are not being adequately treated for constipation.

Hirschsprung's disease is sometimes suspected when a child with ARM has severe constipation, and some clinicians perform biopsies [12,13]. In fact, Hirschsprung's disease is no more common in patients with ARM than in the general population [8]. Some surgeons have even done Hirschsprung's type pullthroughs in these patients, but loss of the rectum (a common part of certain pullthroughs for ARM done mostly before 1985 [14]) leads to loose stool and requires treatments that slow down the colon. Unfortunately, most of these patients become fecally incontinent, because, in addition to the lack of a true anal canal and deficient sphincters, they now no longer have a rectal reservoir and, thus, cannot retain loose stool or rely on proprioception for help with a voluntary bowel movement [15]. Therefore, such a resection when constipation is misinterpreted for Hirschsprung's can lead to fecal incontinence.

For patients with true fecal incontinence, a bowel management program is an artificial way to keep them clean with daily enemas [11,15]. For most ARM patients (75%), with the potential for continence, medical manage-

Table 1 Indicators of prognosis for bowel control in anorectal malformation patients

Good prognostic features	Poor prognostic features
<ul style="list-style-type: none"> ● Normal sacrum / spine ● No presacral mass ● Good buttock crease ● Good anal dimple ● Some types of ARM <ul style="list-style-type: none"> ○ Rectoperineal fistula ○ Rectobulbar urethral fistula ○ Rectovestibular fistula ○ Cloaca < 3 cm common channel ○ Rectal atresia or stenosis ○ Imperforate anus without fistula 	<ul style="list-style-type: none"> ● Abnormal sacrum ● Myelomeningocele ● Some types of ARM <ul style="list-style-type: none"> ○ Rectoprostatic urethral fistula ○ Rectobladder neck fistula ○ Cloacal exstrophy cloaca ○ >3 cm common channel ○ Complex defects

ment consists of avoidance and treatment of constipation, as well as effective potty training strategies [11]. Therefore, only a small minority require washouts and, thus, would need a Malone or cecostomy.

In this review, we sought to objectively assess the consequences of constipation in ARM who have good potential for bowel control (Table 1). We hypothesized that there is a lower incidence of morbidity when constipation is managed early and effectively.

1. Methods

From our series of over 2000 patients with ARM, we reviewed the charts of 398 patients with good potential for fecal continence (Table 2) and a tendency toward constipation, including those with rectoperineal fistula (63), rectovestibular urethral fistula (114), rectobulbar urethral fistula (104), cloaca with common channel below 3 cm (62), imperforate anus with no fistula (46), and rectal atresia or stenosis (9). These patients historically have a good prognosis for bowel control. [1] Those lost to follow up, not yet toilet-trained (or <3 years old), or with poor prognostic features (Table 1) were excluded. We compared morbidities in patients we operated on and managed primarily with an aggressive constipation avoidance protocol (group A, n = 268) to those operated on, treated primarily or managed postoperatively at other institutions who suffered from constipation or incontinence and were referred to us for treatment, operation or reoperation (group B, n = 130) [25].

In group A, constipation was treated with the following protocol: a senna-based laxative was begun after primary repair or after colostomy closure. Stool softeners were avoided because they make the stool too soft, and they do not help to provoke a bowel movement. We found stimulant laxatives in this patient group more effective as they are designed to increase the motility of the colon. Although often

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