Case series with literature review: Surgical approach to megarectum and/or megasigmoid in children with unremitting constipation

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ABSTRACT

Background: The role of surgery in treating children with functional constipation (FC) is controversial, because of the efficacy of bowel management programs. This case series is comprised of failures: 43 children, spanning 25 years’ practice, who had megarectosigmoid (MRS) and unremitting constipation.

Purpose: To determine whether these children were helped by surgery, and to contribute to formulating a standard of care for children with megarectum (MR) and/or redundancy of the sigmoid colon (MS) who fail medical management.

Method: We describe our selection criteria and the procedures we utilized – mucosal proctectomy and endorectal pull-through (MP) or sigmoidectomy (SE) with colorectal anastomosis at the peritoneal reflection. The internet (social media) allowed us to contact most of these patients and obtain extremely long follow-up data.

Results: 30/43 patients had MP and 13/43 had SE. Follow-up was obtained in 83% MP and 70% SE patients. 60% of MP and 78% of SE patients reported regular evacuations and no soiling. 20% MP patients had occasional urgency or soiling or episodic constipation. 12% MP and 22% SE patients required antegrade continence enemas (ACE) or scheduled cathartics and/or stool softeners. 4% MP had no appreciable benefit, frequent loose stools and soiling, presumably from encopresis.

Conclusion: MR is characterized by diminished sensation, poor compliance and defective contractility. Patients with MR do better with MP, which effectively removes the entire rectum versus SE, where normal caliber colon is anastomosed to MR at the peritoneal reflection; furthermore, MP reliably preserves continence; whereas total proctectomy (trans-anal or trans-abdominal) may cause incontinence.

1. Introduction

Functional Constipation (FC) is a disorder of homeostasis. With less frequent defecation, stool accumulates and stagnates. Increased water absorption results in bulky, hard stools; and pain during defecation promotes stool retention. A vicious cycle ensues. FC patients have symptoms of intestinal obstruction: bloating, cramps, and diminished appetite. Ultimately, their health and wellbeing suffer.

1.1. Table 1 Characteristics of children with severe constipation [1]

Bowel management programs usually are effective in treating FC [2]; however, not all patients tolerate laxatives, especially the large doses that are prescribed; stool softeners may exacerbate soiling [3]; and many children simply refuse enemas [4].

FC is a heterogeneous disorder, as physiological investigations have demonstrated. More precise understanding of pathogenesis promotes therapeutic efficacy. Colonic manometry and radionuclear transit distinguish “slow colonic transit” (SCT) from “fecal retentive constipation” (FRC), which is usually associated with MR [5]. Anal sphincter resting pressure usually is normal in MR [6] but increased in anal sphincter achalasia and dyssynergia. The anorectal inhibitory reflex is present in MR but absent in patients with Hirschsprung’s Disease.

In our estimation, the contrast enema remains the most valuable tool in evaluating children with unremitting constipation. Static radiographs define anatomy; and post-evacuation films document adequacy of rectal emptying. Defecography is a more accurate study but has limited usefulness in children.
MR has distinguishing radiologic features, other than distension (Fig. 1). The normal rectum has an oval shape, and it is located in the pelvis. MR is conical in shape and extends above the pelvic brim. The diameter of MR is maximal at the sacral promontory; and usually, there is an abrupt transition between the hugely dilated rectum and normal caliber colon [7].

1.2. Fig. 1 Adolescent girl whose megarectum simulates a gravid uterus!

MR was first described in association with anorectal malformations [8,9]. Later, it was recognized in otherwise normal children with unremitting constipation. Various terms have been used to describe MR: atonic baggy rectum, terminal fecal reservoir, rectal ectasia and inertia [10]. It may present during infancy or later (Figs. 2 and 3). The postulated causal sequence is as follows:

1. An enteric nerve or muscle abnormality
2. Impaired peristalsis resulting in distension
3. Excess stretch weakening muscular contraction
4. Dysmotility leading to incomplete evacuation and stool retention
5. Fecal impaction and ultimately encopresis (overflow incontinence)

1.3. Figs. 2 and 3 Infant and adolescent with megarectum

Continence requires a compliant rectum with effective peristalsis and an anal sphincter that contracts, generating an adequate resting pressure. In patients with ARM and MR, it may be difficult to distinguish true incontinence (inadequate sphincter) from pseudocontinence (overflow). Recovery of continence following proctectomy may be predicted by assessment of anal sphincter function with manometry and anal sphincter musculature by endosonography [6].

Manometry in patients with MR requires inflation of the balloon to supernormal volumes. A normal reading is encouraging; these patients may respond to bowel management programs. If the rectum is stiff and noncompliant, contractility is impaired, proprioception (perception of distension) is diminished, and the anorectal inhibitory reflex is attenuated. Medical intervention is unlikely to be effective in such patients.

Patients with MR are outliers in the FC spectrum. Typically, these children have always had difficulty moving their bowels. They were never “toilet trained”; they are incontinent. MR’s huge capacity predisposes to fecal impactions, painful defecation, and encopresis. These children usually have seen multiple practitioners; their parents are frustrated, at their wits’ end; whereas the child may present a façade of insouciance - “la bell indifference.” Self-control is a developmental milestone; these children cannot even control their bowel movements, a situation made all the more devastating because it is misunderstood. People assume that they are incontinent by choice and that they choose not to exercise self-control. As an example of the suffering caused by this disorder, one young man was required to sit in a particular chair. He could not choose where he sat, because he was considered unclean, like a leper.

MR occurs infrequently, comprising only 11% of patients referred to tertiary colorectal center for treatment of refractory constipation [3]. This low incidence may account for the lag in acceptance of MR as an
indication for proctocolectomy.

2. Materials and methods

Patients were referred either because of an acute decompensation, the clinical constellation of impaction, abdominal distention, and feculent vomiting; or because of constipation that was unremitting despite therapeutic interventions.

Information from an unpublished study conducted by MUSC students in 1996 enabled us to contact several patients who had surgery twenty or more years ago; however, the radiographs of these (older) patients were not available. We utilized the PROCESS and SCARE guidelines in collating our results and in preparing this report [11,12]; and IRB approval (PH Pro 42654) was obtained.

Patient selection began with our (ACS) case log. This compilation was culled to eliminate Hirschsprung’s Disease, Familial Polyposis, Inflammatory Bowel Disease, Necrotizing Enterocolitis, Intussusception, and other conditions in which MP or SE are performed. A history of fecal disimpaction was helpful in identifying MR patients; and patient age and rectal biopsy provided exclusionary criteria. All available contrast enemas were consistent with MRS.

The hindgut anatomy, as visualized by a water soluble contrast enema, was the primary determinant whether a patient was offered surgery. Was the configuration of the rectum consistent with MR? Was there redundancy of the sigmoid colon? In some extreme instances, the sigmoid colon recapitulates the course of the more proximal colon.

In addition, our radiologists usually would estimate the amount of contrast and stool remaining on the post-evacuation radiograph, and this provided an assessment of contractility, albeit qualitative. If good evacuation was noted, > 50% emptying of stool and contrast, surgical intervention was delayed. If evacuation was poor and the anatomy was consistent with either MRS or MR, we offered resection or an ACE procedure.

Gastrografin enemas (GGE) were also utilized in bowel preps. Because of its detergent-like qualities, Gastrografin is an effective tool in treating babies with Meconium Plug Syndrome and Meconium Ileus. This efficacy was employed in our patients to achieve a clean colon and rectum preoperatively.

Prior to surgery, there was deliberate intensification of medical therapy, because of the difficulty in achieving adequate bowel prep. Despite these efforts, post-evacuation films obtained pre-operatively showed no improvement in the adequacy of evacuation.

3. Results

One pediatric surgeon operated upon 43 patients over a 25 year period; MP was performed in 30 patients and SE in 13. 90% (39/43) patients were treated at Palmetto Health Children’s Hospital in Columbia, SC; the remaining 10% were treated at USA Children’s and Women’s Hospital in Mobile, AL. All patients fulfilled the diagnostic criteria for FC (Table 1), and all had failed rigorous bowel management programs.

3.1. Table 2 Compendium of results

- Follow-up was obtained in 83% MP and 69% SE patients.
- The mean age for MP was 11 years and 7 for SE.
- The mean length of follow-up was 13½ years for MP, and 6 years for SE.
- One MR patient presented with toxic megacolon and a colostomy was performed prior to MP.
- Three MP patients (12%) developed cuff abscesses post-operatively. Two required temporary colostomies; simple drainage sufficed in the remaining patient.
- Patients who developed complications usually were ill pre-operatively. These patients typically required parenteral fluids, antibiotics, and nutritional support; in addition to the bowel prep. Primary MP was performed to avoid colostomy, but this was an unwise decision.

Primary MP was performed in 3 patients with borderline MR; 2 of these patients required subsequent ACE, because of unsatisfactory relief of constipation. Most probably, MP would have provided greater amelioration, since MR patients do best when the entire dysfunctional bowel is removed [13].

The third patient with borderline MR did well with SE. The post-evacuation film in this patient demonstrated good contractility (> 50% evacuation of stool and contrast). This study may prove helpful in sorting borderline MRS patients to either SE or MP.

Two patients from each group (MR, SE) required ACE. These patients are placed in those with "modest benefit"; however, resection of redundant sigmoid colon is sometimes necessary for antegrade enemas to be effective, especially in patients with colon inertia.

Our one "failure" has persistent overflow incontinence, whereas all patients had enecrosis preoperatively. One of three patients with ARM (caudal regression) wears a pad for occasional soiling and uses a plug during sexual intercourse. The other two patients with ARM (low) are

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1 One patient with a colostomy was lost to follow-up for 8 months. During this time, the developed disuse colitis and an anastomotic stricture, which required operative revision. Subsequently, there was intermittent bleeding from granulation tissue at the stricturoplasty sites; this resolved with ligation (End loop) of the granulation tissue pedicle.
The two patients with autism continue to be incontinent, but this is a behavioral issue.

3.3. Table 3 Duration of follow-up correlates with satisfactory outcome

A formal quality of life survey was not performed; however, according to Christison-Lagay et al. “quality of life scores demonstrate that physician assessment of a satisfactory clinical outcome correlates with patient and family satisfaction” [1]. Most responders (children or parents) were satisfied with their surgery. Many were extremely grateful. Even the autistic children seemed to derive benefit [14].

4. Discussion

Powell and Grosfeld compared rectal ectasia (MR) to the bulbous proximal intestine in jejunal atresia. In making comparisons, an unfamiliar entity is likened to one that is familiar. This is termed “analogical reasoning”. Pattern recognition is commonly utilized in science. Analytical chemists identify unknown compounds by comparisons with compounds whose properties are known. Analogical reasoning is
inductive, and its conclusions are probabilistic, not certain [19].

It is a generally agreed that dilated bowel has poor motility. If a baby with jejunal atresia has an adequate length of intestine distal to the obstruction, the bulbous proximal segment is resected. If there is a paucity of intestine distal to the obstruction (short gut syndrome), the bulbous segment is tapered, or elongated by the STEP procedure, or longitudinally divided as described by Bianchi. Experience with the Bianchi technique is highly illustrative, in an analogical way. Even if the blood functions to one of the divided halves is impaired and it dies, the viable half functions better. Decreasing the diameter of a bulbous segment of intestine improves its function.

Grosfeld utilized proctectomy (low anterior resection) in two (older) patients with ARM and MR [10]. Pena applied this procedure to children with ARM (anorectal malformations) who had severe constipation overflow incontinence (encopresis). Pena, however, divided the rectum at the peritoneal reflection, because he believed that a rectal reservoir was necessary for proprioeception (discrimination of flatus from stool) and continence. The partially resected rectum was anastomosed to the sigmoid colon. Patients' laxative requirements were dramatically decreased; however, these children still required bowel management [8]. They were significantly helped, but not cured. Preservation of the dilated, dysfunctional rectum appeared illogical and likely responsible for relapse - constipation and fecal impaction [13]. When more complete rectal resections were performed, not only were laxative requirements dramatically decreased, but many children with encopresis became continent, as long as their anal sphincter mechanism was intact. Equally surprising was that manometry of pull-through colon was normal.

Complex problems are multifactorial, with interacting, mutually influencing components. Evaluation must consider the whole, as well as the individual parts. There may be competing goals that require nuanced compromise [19]. For instance, speed and durability are both the individual parts. There may be competing goals that require resection with preservation of the distal rectum. This procedure is the dentate line). Excellent amelioration of symptoms was obtained, but racing bike has desirable qualities, but they cannot be optimized simultaneously. A
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The rapid reduction in the need for cathartic agents after MR resection suggests that the principal determinant for ongoing fecal retention in this patient population is the megarectum itself... Once the diagnosis of megarectum is made, surgery should not be delayed; because conservative therapy not only fails, but it may have significant psychological and physical morbidity.” [4].

This review was prompted by skepticism, “Is it appropriate to operate upon children with FC?” Most people in our study insist that they are normal, which is remarkable considering that they were “failures of medical management”. It may be that surgery was one factor among many - lifestyle adjustments, dietary modifications, vigilance in maintaining regular bowel movements - that enabled them to cope with their disability; nevertheless, the majority credit surgery with their turn around, for which they are enormously grateful.

Ethical approval

IRB approval obtained, PH Pro 42654.

Sources of funding

None.

Author contribution

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5. Conclusion

Functional constipation (FC) causes considerable morbidity in children. Bowel management programs have revolutionized the care of these children. MRS is an entity as distinct as Hirschsprung’s Disease. Children with MRS are infrequently encountered; they consistently fail medical management, and practitioners usually do not know how to treat them. The efficacy of surgery for these children is not widely appreciated; otherwise, why is it controversial?

Indications for surgery in patients with MR are:

1. Characteristics of FC as listed above (Table 1)
2. Failure of bowel management programs
3. Characteristic appearance of MR on contrast enema
4. Post-evacuation film shows poor (<50%) evacuation of stool and contrast
5. Manometry shows a stiff, non-compliant rectum

Despite the limitations of this retrospective study - small numbers, single surgeon and institution - it affirms the utility of surgery for MRS and clarifies the quandary regarding the ‘procedure of choice” for MRS. The conundrum is that the most effective operation (total proctectomy) may cause incontinence. To avoid this unintended consequence, SE or anterior resection, is utilized albeit with less efficacy. MP offers an advantage over these two alternatives - retention of the rectal muscular sleeve preserves continence, whilst accomplishing a functionally complete proctectomy.

In our patients, leaving a cuff of MR was associated with an unsatisfactory result; and usually, a subsequent ACE was required.

Incontinence, in our patients, was associated with autism or a severe anorectal anomaly (caudal regression syndrome) or recurrent encopresis.

If megarectum is responsible for FC in these patients, proctectomy should be curative. Our results are consistent with this supposition. “The rapid reduction in the need for cathartic agents after MR resection suggests that the principal determinant for ongoing fecal retention in this patient population is the megarectum itself... Once the diagnosis of megarectum is made, surgery should not be delayed; because conservative therapy not only fails, but it may have significant psychological and physical morbidity.” [4].

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2 "I no longer do a transanal resection as removal of rectum induces incontinence in many patients; many recover but it may take 6-12 months. So therefore I now would do a laparoscopic low anterior resection and leave intact 6-8 cm of rectum.” [20].
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Conflicts of interest

None.

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Guarantor

James G. Glasser, MD.

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