

Constipation—Surgical Aspects, Investigations, and Therapies

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Abstract

Purpose of Review The surgical management of pediatric constipation has been modified in recent years, with a more focused approach to operative interventions and colonic resection. This review will highlight recent insights into the investigation and management of children with intractable constipation.

Recent Findings Chronic constipation remains the domain of pediatricians and pediatric gastroenterologists. However, more targeted and accurate investigations have led to a greater understanding of the pathophysiology underlying constipation. These, in turn, have resulted in a more individualized approach to the surgical management of constipation. Improvements have been made in the investigation of constipation, including greater use of nuclear transit studies and colonic manometry. Greater fidelity in these investigations has allowed surgeons to employ a more algorithmic approach to patient management, with specific use of colonic enemas and colonic resection.

Summary Chronic constipation remains a difficult condition to treat in many children. Surgeons have an important role to play in specific circumstances, following detailed investigation of the underlying causes.

Keywords Pediatric · Constipation · Colectomy · Enema · Sacral nerve stimulation

Introduction

Chronic constipation is a common condition in childhood. Approximately 3% of pediatric consultations and 10–45% of pediatric gastroenterology consultations are related to constipation [1, 2]. The majority of these children have a functional condition and will be managed appropriately with laxative therapy, thus not requiring a surgical review. However, for those patients that fail medical management, there needs to be a systematic approach to employing possible surgical interventions.

Many of the difficulties in diagnosing and managing chronic constipation are related to unwieldy definitions. The Rome III criteria are commonly used in the research setting but are less frequently adhered to in the clinical environment. Failure to use standardized definitions impairs the ability of clinicians to diagnose constipation and then accurately determine true failure of medical management [3••]. It is not unusual for a child to be referred for surgical investigation and intervention, in whom maximal medical management has not been obtained.

A thorough approach to investigation (including operative interventions) and appropriate surgical management is required for the small proportion of children that are unable to be managed by our pediatric colleagues.

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Diagnosing the Child with Chronic Constipation

The Rome III criteria provide an age-specific diagnosis tool for chronic functional constipation [2]. Whilst imperfect, the criteria have enabled clinicians and researchers to speak a common language. The authors of the Rome IV criteria (released in 2016) have aimed to simplify the language and diagnostic criteria for chronic constipation. A thorough review of the diagnosis of functional constipation in children is beyond the scope of this article and may be found in Tabbers et al. [4•].

Investigating the Child with Chronic Constipation

Radiological Investigations

The Abdominal Radiograph

The use of the abdominal radiograph in the investigation and management of chronic constipation remains controversial. Whilst Tabbers et al. and Berger et al. reported that radiological testing is of limited or no use in the diagnosis of constipation, other clinicians have demonstrated extensive experience with the use of abdominal radiographs in the management of fecal incontinence and constipation [4•, 5–7]. The utility of quantifying the rectal diameter in constipation remains uncertain, and radiographs have been shown to be inadequate for this assessment [8]. In this author's experience, the majority of patients will have had a number of abdominal radiographs prior to surgical referral.

Ultrasonography

The lack of ionizing radiation has made ultrasonography an attractive proposition in the assessment and management of chronic constipation [9]. However, the majority of clinicians either rarely, or never, use ultrasonography [10•]. Proponents of the technique highlight its ability to determine the degree of rectal filling and propose its use as a replacement for digital rectal examination [11]. However, other researchers have demonstrated that ultrasonography fails to correlate with patient-reported symptoms of constipation [12].

The Contrast Enema

The contrast enema provides excellent information regarding the anatomy of the rectum and colon, with a particular focus upon the degree of rectal distension and proximal colonic dilatation [13]. However, it is not recommended as an initial diagnostic tool in the evaluation of constipation [4••]. Utilization of the contrast enema varies between pediatricians and pediatric surgeons. Koppen et al. [10•] demonstrated that, whilst surgeons would more often routinely use the contrast

enema in their diagnostic work-up of intractable constipation, the majority of pediatricians would be willing to use the study only in selected cases.

Studies of Intestinal Transit

Colonic Transit Time

Measurements of colonic transit time (CTT), typically using Sitz markers, may be employed as a proxy-marker for the assessment of colonic motility patterns (e.g., slow transit constipation versus functional fecal retention). These studies are well-described and may be matched with pediatric control data [14, 15]. However, there is evidence that pediatric surgeons remain unconvinced as to the utility of this study [10•].

Nuclear Transit Study

Nuclear transit scintigraphy is a superior investigation of colonic motility, when compared with CTT [14]. However, this modality remains rarely used, except in specialist institutions, and is even less utilized than CTT [4•, 10•, 16, 17]. This author's preference is to use scintigraphy in those patients in whom Hirschsprung disease has been previously excluded and who have failed maximal medical therapy.

Manometry

Anorectal Manometry

High-resolution anorectal manometry is steadily replacing the use of water-perfused catheter systems in the pediatric setting [18, 19]. In expert hands, anorectal manometry may be used to discriminate between Hirschsprung disease, external sphincter dyssynergia and internal anal sphincter achalasia [20•]. The majority of clinicians (and researchers) will focus upon the presence or absence of the rectoanal-inhibitory reflex (RAIR). It is thought that the presence of the RAIR excludes Hirschsprung disease; however, false positive results have been reported [4••]. As with many investigations in pediatric practice, the utility of anorectal manometry as a diagnostic tool is limited by a paucity of control data.

Colonic Manometry

Colonic manometry, even more than anorectal manometry, remains an investigation largely limited to specialist referral centers and academic institutions [10•, 21]. Children with slow transit constipation have been shown to have clear manometric anomalies, consistent with failure in antegrade colonic propagation [21, 22]. Colonic manometry has been used to predict the potential for success with antegrade colonic enemas [23]. In addition, more than two-thirds of those patients

reported to have undergone a colonic resection for chronic constipation had pre-operative colonic manometry [20•]. However, recent advances in catheter sensitivity and accuracy have led to a more nuanced understanding of manometry, and many of the inferences from earlier manometric findings will require reassessment [2, 18, 24].

The Rectal Biopsy

The rectal biopsy remains the gold standard for the diagnosis of Hirschsprung disease [4•, 25]. However, its role in the investigation of chronic constipation remains controversial. Whilst some authors have argued that the rectal biopsy is mandatory in the diagnostic work-up, the recent study by Koppen et al. did not include the rectal biopsy as one of the available investigations [3•, 10•]. There is also a lack of standardization with regards to the technical aspects of the biopsy and the available pathological investigations [26]. This author uses a selective approach to the rectal biopsy, with an emphasis upon family history and the neonatal presentation of stooling.

Non-operative Approaches to Chronic Constipation

Retrograde Continence Enema

The retrograde continence enema (RCE), despite centuries of use, was first shown to be effective in spina bifida children by Shandling and Gilmour at the Hospital for Sick Children, Toronto [27]. The technique is now regularly employed for children affected by fecal incontinence and/or constipation in the setting of spina bifida, Hirschsprung disease, anorectal malformations, or functional constipation [28–31]. When managed effectively, with long-term nursing intervention to maintain compliance, RCE has excellent results in patients with constipation, with more than 80% achieving pseudo-continence [32].

Transcutaneous Electrical Stimulation

Transcutaneous electrical stimulation (TES) was first demonstrated to have a positive effect upon children with slow transit constipation in 2005 [33]. In patients with chronic constipation, it has led to improvements in quality of life, symptom scores, as well as reduced requirements for appendicostomy formation [34–36]. The technique is able to be performed at home, following minimal training [17]. It is likely the effects of TES result from improved colonic motility, as demonstrated by Clarke et al. in their study of colonic manometry performed prior to, and following, a course of TES [37]. However, the technique remains limited to academic centers and has not yet reached levels of acceptance comparable to sacral nerve stimulation.

Operative Approaches to Chronic Constipation

Local Interventions

Botox

The injection of botulinum toxin (Botox) into the internal and external anal sphincters remains controversial, despite two decades of experience in pediatric surgery. First described by Langer and Birnbaum for the management of persistent constipation following pull-through for Hirschsprung disease, Botox has now been used in patients with chronic anal fissure, internal anal achalasia, and chronic constipation [3•, 38–41]. Botox injection has largely replaced the technique of sphincterotomy in the pediatric population, and may be used in the presence of abnormal anal sphincter pressures, as measured on anorectal manometry [10•].

Antegrade Colonic Enema

The antegrade colonic enema (ACE) has greatly altered the management of chronic constipation and fecal incontinence, following its description by Malone et al. in 1990 [42]. By providing pseudo-continence, and thus independence, the ACE has been shown to significantly improve the quality of life in patients with slow transit constipation [43]. Recent evidence suggests that use of the ACE may actually facilitate improvements in colonic motility, which raises the possibility that this technique could be employed earlier in many patients [23]. In addition, the improvement in colonic motility may lead to symptom resolution in selected patients [44].

The ideal surgical approach to the ACE remains undecided. Variation in practice includes the segment of bowel utilized (appendix, cecum), the approach (laparoscopic, open, percutaneous, colonoscopic), and the need for a valve mechanism to prevent reflux of effluent. There are no studies that adequately address these issues.

Stimulation

Sacral Nerve Stimulation

Sacral nerve stimulation (SNS) has been used in adult patients with chronic constipation and fecal incontinence, with variable efficacy [45•, 46]. Its role in the pediatric population remains uncertain, despite some early promising results [47, 48].

Stoma Formation

Ileostomy

Formation of an ileostomy is rarely required in patients with chronic constipation. In the majority, the ACE should be able to

provide adequate management. However, in a selected group of patients (pan-colonic dysmotility, failure to thrive, failure of ACE therapy), there is a role for temporary diversion [3••, 20•].

Colostomy

The indication for colostomy formation is, perhaps, even more obscure than that for ileostomy formation. Koppen et al. [10•] demonstrated that neither pediatric surgeons nor pediatric gastroenterologists believed that the colostomy played a significant role in the management of chronic constipation.

Selective Resection

Partial Colectomy

Historically, colonic resection was performed with some regularity in both adult and pediatric patients with chronic constipation. With improvements in medical management, and the introduction of the ACE, the requirement for extensive resection has diminished [13]. However, there remain children in whom their clinical condition and quality of life are significantly impaired, despite maximal medical therapy and the use of ACE.

Targeted resection has replaced subtotal colectomy. In specialist centers, contrast enemas, colonic transit studies (Sitz marker, scintigraphy), and/or colonic manometry have been used to guide the appropriate extent of resection [3••]. The mode of resection (laparotomy, laparoscopically assisted, transanal) remains controversial. Levitt et al. [13] described early success with a modified transanal Swenson technique. However, the long-term effects of that approach are unknown.

Partial Colectomy Plus Appendicostomy Formation

A recent adjunct to colonic resection has been the addition of an appendicostomy during the same procedure [20•, 49]. Whilst the number of reported cases remains small, the early results are promising for conditions including anorectal malformations and spina bifida [50]. Similar results have now been demonstrated for chronic constipation [49, 51]. The presumed advantages of this technique include retention of the rectal reservoir with an ability to remain fecally clean, and flexibility with the ongoing requirements for ACE.

Novel Therapies for Chronic Constipation

Experimental

Stem Cell Therapy

Stem cell therapy for enteric neuropathies has become a feasible proposition in the last 5 years [52]. Advances in the

understanding of Hirschsprung disease, esophageal achalasia, and chronic intestinal pseudo-obstruction have led to an increased focus upon potential cures for intestinal conditions [53••]. However, the etiologies underlying chronic constipation remain elusive, thus hampering efforts for stem cell-based therapies.

Colonic Pacing

The use of intramuscular electrodes to aid with colonic pacing has been described in adults with slow transit constipation [54]. This invasive technique, whilst effective in highly selected patients, is yet to be demonstrated in the pediatric population.

Conclusions

The investigation and management of chronic constipation remain complex, with many patients referred to pediatric surgeons. In the majority of patients, appropriate and focused investigations will lead to minimal surgical input. However, there remains a minority of patients that will require operative interventions, ranging from rectal biopsy to partial colectomy. Only with improved physiological investigations and systematic long-term follow-up will the implications of these interventions be truly appreciated.

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Compliance with Ethical Standards

Conflict of Interest Sebastian King has no conflict of interest to disclose.

Human and Animal Rights and Informed Consent This article does not contain any studies with human or animal subjects performed by the author.

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