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REVIEW ARTICLE

Guidelines for the management of postoperative soiling in children with Hirschsprung disease

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Abstract

Although most children with Hirschsprung disease ultimately achieve functional and comfortable stooling, some will experience a variety of problems after pull-through surgery. The most common problems include soiling, obstructive symptoms, enterocolitis, and failure to thrive. The purpose of this guideline is to present a rational approach to the management of postoperative soiling in children with Hirschsprung disease. The American Pediatric Surgical Association Hirschsprung Disease Interest Group engaged in a literature review and group discussions. Expert consensus was then used to summarize the current state of knowledge regarding causes, methods of diagnosis, and treatment approaches to children with soiling symptoms following pull-through for Hirschsprung disease. Causes of soiling after pull-through are broadly categorized as abnormalities in sensation, abnormalities in sphincter control, and "pseudo-incontinence." A stepwise algorithm for the diagnosis and management of soiling after a pull-through for Hirschsprung disease is presented; it is our hope that this rational approach will facilitate treatment and optimize outcomes.

Keywords Hirschsprung disease · Soiling · Incontinence · Pseudo-incontinence · Bowel management · Pull-through

Background

Although most children with Hirschsprung disease ultimately achieve functional and comfortable stooling, some may experience a variety of problems after pull-through surgery. The most common problems include soiling, obstructive symptoms, enterocolitis and failure to thrive [1]. The severity of these problems varies significantly, and an individual child may have more than one of these issues. The purpose of this guideline developed by the American Pediatric Surgical Association (APSA) Hirschsprung Disease Interest Group is to present a rational and step-wise

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approach to the management of post-operative soiling in children with Hirschsprung disease.

Defining continence, true incontinence, and pseudo-incontinence

For the purposes of this guideline, we define fecal continence as the ability to have voluntary bowel movements without soiling in the absence of an enema program. To achieve continence in patients with Hirschsprung disease, there must be the ability to feel distention of the distal pull-through segment (defined hereafter as "neo-rectum"), to perceive stool contact with the anal canal, adequate anal sphincter tone, and appropriate colonic motility [2]. Disruption of one or more of these factors may result in patients being partially or totally incontinent, which then leads to soiling. Young children or those children with developmental delay or intellectual impairment may be at further risk for soiling as the feedback mechanisms between the body and brain may not be fully mature.

Those patients with anatomic or physiologic disruption of their continence mechanisms are defined as having true fecal incontinence. Those patients who have intact physiological mechanisms needed for continence, but who exhibit persistent soiling after a pull-through operation, will be defined as having "pseudo-incontinence."

Pseudo-incontinence may be secondary to one of two underlying etiologies:

1. Obstruction/fecal impaction: This is a more classic definition of pseudo-incontinence and is caused by the inability to efficiently empty the neo-rectum or by obstruction at the level of the sphincters. Symptoms are manifested by abdominal distension, bloating, vomiting, infrequent stooling, leakage of small amounts of formed stool ("skid marks"), and/or the overflow of liquid stool [3];

2. Hypermotility characterized by the rapid transit of stool through the colon: In Hirschsprung disease, hypermotility is presumably related to the fact that the neo-rectum consists of proximal colon rather than the natural reservoir of the rectum. Soiling occurs because the anal sphincter and anorectal sensation mechanisms are not capable of preventing the egress of stool in the face of the high amplitude propagating contractions (HAPC) which are being propelled to the anus [4].

Causes of soiling after pull-through

The main causes of persistent soiling following a pull-through are listed in Table 1 [5]. These can be broadly categorized as abnormalities in sensation, inadequate sphincter control, or "pseudo-incontinence."

1. *Abnormal sensation* can stem from two components. The first is an inability to sense a full neo-rectum. Although all children with Hirschsprung disease have their native rectum removed at the time of pull-through, there is variability in the volume of distension of the neo-rectum that is sensed from one child to another after a pull-through procedure. A lack of distal sensation may be related to abnormal motility intrinsic to the underlying Hirschsprung disease, denervation at the time of pull-through, or may be secondary to chronic distension from untreated fecal impaction. Anorectal manometry can be helpful to measure the child's ability to sense distention as a potential etiology of soiling, although not all children will be adequate candidates for manometry depending on their age and development. A second component of abnormal sensation is an inability to differentiate between gas and stool. This differentiation has traditionally been thought to be dependent on an intact dentate line and the transitional epithelium of the anal canal, which may be lost if the anastomosis is performed too distally and the transitional epithelium is damaged during surgery [6]. The integrity of the dentate line may be determined by physical examination, which may require anesthesia for the older child (Fig. 1). We note that the role of the dentate line is controversial, and few investigations exist regarding its role in post pull-through soiling. While it is unclear whether either loss of sensation of distension or loss of the dentate line can independently lead to incontinence, it is likely that the loss of either can at least exacerbate incontinence that is due to other reasons.

2. *Inadequate sphincter control* may be caused by sphincter injury during the pull-through, most commonly due to excessive stretching of the sphincter muscles [7]. Sphincter function may also be altered due to a previous myectomy or sphincterotomy. Damage to the sphincters may be indirectly inferred on rectal examination, but anorectal manometry provides more objective evidence of both internal and external sphincter function [8–10]. It is important not to perform anorectal manometry under general anesthesia, which can cause an artificial lowering of sphincter pressures. Note that an absent rectoanal

Table 1 Causes of soiling after a pull-through for Hirschsprung Disease

Causes of soiling
Abnormal sensation
Inability to sense distension of the neo-rectum
Loss of transitional epithelium/dentate line
Inadequate sphincter control
Over stretch of sphincter mechanism during pull-through
Previous myectomy or sphincterotomy
"Pseudo-incontinence"
Obstruction/fecal impaction
Hypermotility

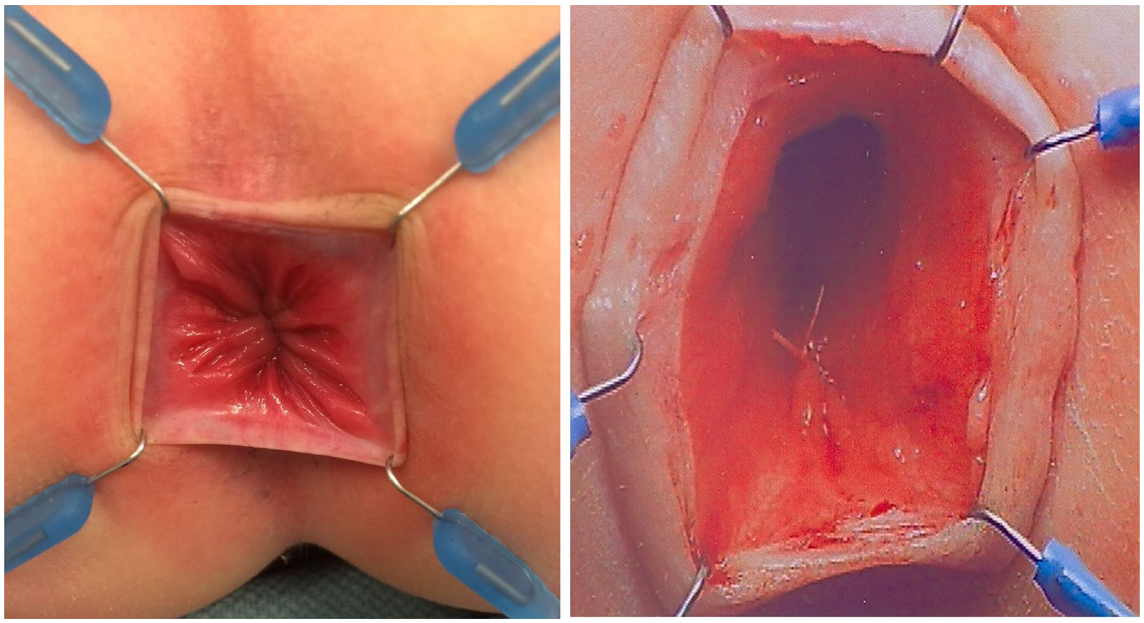

Fig. 1 Post-pull anatomy demonstrating an intact dentate line (left), and a dentate line which has been damaged (right) (images courtesy of M Levitt)

inhibitory reflex (RAIR) is expected even after pull-through surgery, and the value of anorectal manometry in this setting is to evaluate the adequacy of resting internal sphincter pressure.

3. We have defined *pseudo-incontinence* as soiling in the presence of normal sensation and normal sphincter function. Pseudo-incontinence is most frequently due to fecal impaction and resultant overflow or leakage of stool. This phenomenon may be due to mechanical obstruction such as anastomotic stricture, a transition zone pull-through, internal sphincter achalasia, or most commonly stool-holding behavior. An algorithm for the investigation of a child with obstructive symptoms has been published previously and includes a contrast enema, rectal examination, repeat rectal biopsy, a trial of intrasphincteric botulinum toxin, and assessment of colonic motility [3]. The neo-rectum in a child with fecal impaction becomes hypo-motile over time through chronic distension. In other children, pseudo-incontinence may be secondary to colonic hypermotility, which results from the pulled-through bowel having more active peristalsis than a normal rectum. Hypermotility results in the neo-rectum not functioning well as a reservoir for stool, and thus leads to the child having difficulty holding the stool between bowel movements. These phenomena are likely mediated by changes in high amplitude propagated contractions in the remaining colon [4]. Hypomotility may be differentiated from hypermotility based on the caliber of the neo-rectum on contrast enema (Fig. 2). Additionally, colonic manometry can provide detailed measurement of the motility of the pulled-through bowel [4, 11].

Guidelines for investigation and management of post pull-through soiling

A proposed algorithm for the investigation and management of soiling after a pull-through is shown in Fig. 3. Generally, an investigation for an underlying cause of soiling should not begin until the time the child is expected to be toilet trained, most commonly around school-age. The ability to successfully achieve urinary continence may be a good proxy for whether the child is developmentally ready for fecal continence. Successful management depends on an understanding of the underlying basis for the soiling, which initially requires a complete history and physical examination. Evaluation of the integrity of the dentate line and anal canal is mandatory through a formal examination (usually under anesthesia, especially in older children). Evaluation of anal sphincter function and sensation of the neo-rectum requires anorectal manometry [10].

Neither sphincter weakness nor poor sensation is generally amenable to a surgical solution. These symptoms are best managed using a bowel management program that encourages the maintenance of solid stools and promotes proper rectal emptying. The former can often be achieved through dietary modification and occasionally through use of bulking agents, while the latter may be accomplished using stimulant laxatives such as senna or bisacodyl in patients who are neurodevelopmentally normal, or an enema program (retrograde or antegrade via appendicostomy or cecostomy) in those who are not

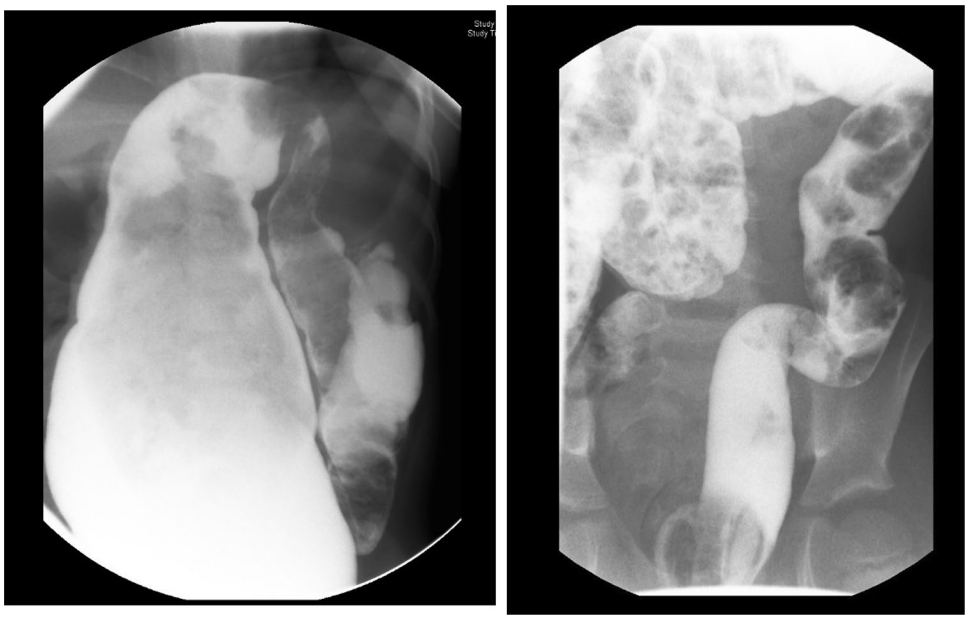

Fig. 2 Contrast enema demonstrating a hypomotile colon (left), and a hypermotile colon (right). A child with fecal impaction and hypomotility has a dilated neo-rectum filled with stool. A follow-up abdominal radiograph the following day will often demonstrate retention of contrast. In contrast, a hypermotile colon is nondilated, and a follow-up abdominal radiograph will usually demonstrate complete emptying (images courtesy of J Langer)

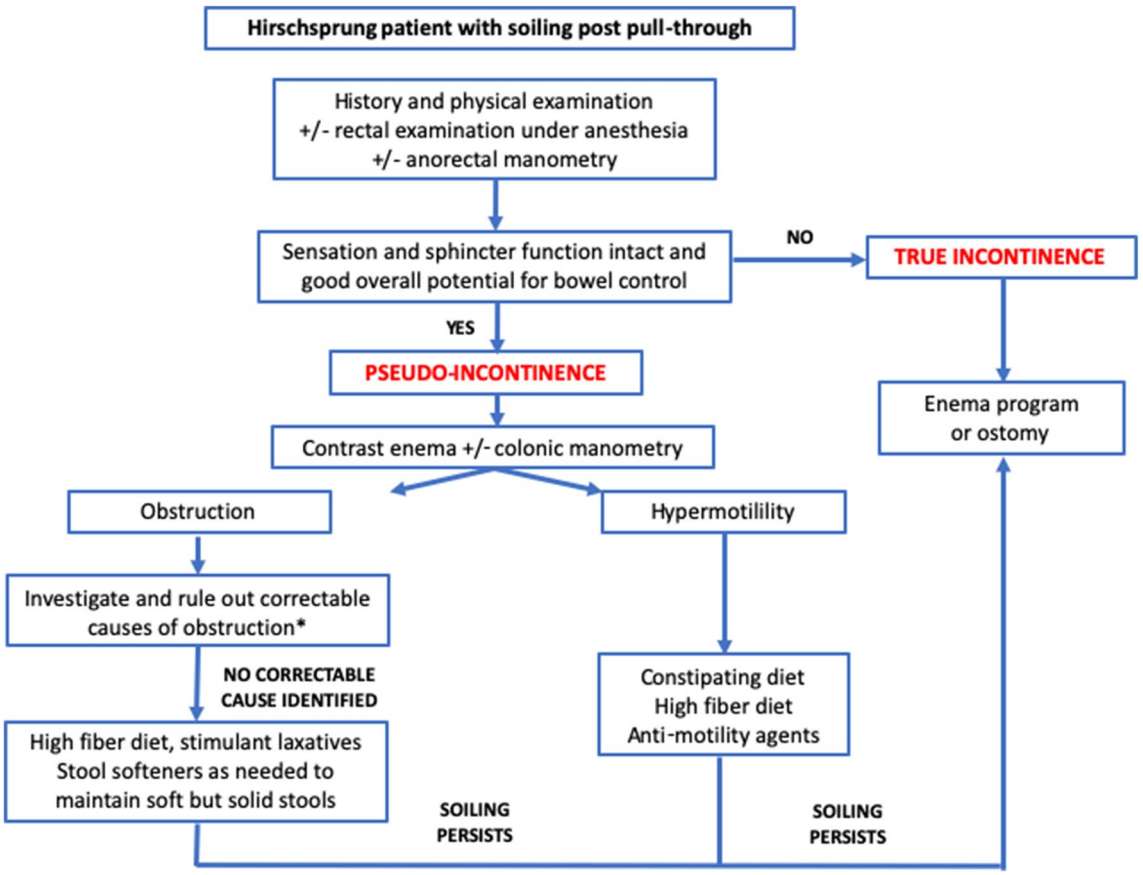

Fig. 3 Algorithm for the diagnosis and management of the child with soiling after a pull-through for Hirschsprung disease. *[3]

neurodevelopmentally normal or those who continue to soil with diet and stimulant laxative therapy. Stool softeners such as PEG3350 are usually contraindicated in children with true incontinence, as looser stools are more difficult to control. Biofeedback training and pelvic physical therapy have been advocated, especially for those

children with sphincter weakness [7]. In severe cases, and in those where bowel management is not successful, the child may be best served by a diverting ostomy.

If sensation and sphincter function are intact, then it is reasonable to assume that the patient has the potential for voluntary bowel movements without soiling. At this stage, it is useful to distinguish between soiling due to fecal impaction and soiling due to hypermotility. Radiographic imaging with a contrast enema may demonstrate a dilated colon and retention of contrast on a subsequent follow-up film in children with hypomotility. If the colon is non-dilated, then the problem may be hypermotility. If the etiology of soiling is still unclear after the contrast study, colonic manometry may be helpful.

Children with soiling due to fecal impaction are best managed long-term with dietary modification including a high-fiber diet and stimulant laxatives. Stool softeners such as PEG3350 should be used only to help create stools that are soft enough to pass easily since overly soft or liquid stools may exacerbate soiling. A complete workup for causes of constipation after a pull-through should be done to rule out mechanical obstruction, transition zone pull-through, internal sphincter achalasia, and colonic dysmotility [3]. Stool-holding behavior may be the primary cause of soiling or may exacerbate fecal impaction in many children. Patients with Hirschsprung disease are especially prone to developing this problem because of persistent internal sphincter non-relaxation. The anal discomfort associated with repeated use of retrograde enemas and dilations can also contribute to the development of stool holding behavior, and, if possible, the use of these techniques should be limited. If soiling persists despite medical therapy, a large volume antegrade enema program may be needed to achieve long-term continence. Patients with a very dilated colon from longstanding impaction may be especially difficult to manage initially with laxatives, thus a treatment option in this group is to initiate large volume enemas for 6–12 months and then perform a laxative trial once the colon has been adequately decompressed. Some have advocated posterior myectomy to overcome the non-relaxing anal sphincter [12], but it carries the risk of permanent injury to the sphincter resulting in true fecal incontinence. Intrasphincteric botulinum toxin injection is a useful alternative to myectomy since it reversibly relaxes the sphincter and can be repeated if successful in relieving constipation. Over time most children will eventually learn to overcome their non-relaxing internal sphincter [13].

If imaging or colonic manometry suggests hypermotility, the child should initially be managed with a constipating diet and bulking agents such as fiber. If anti-motility agents are needed, anti-diarrheals such as loperamide or atropine/diphenoxylate can be added. Amitryptyline may also be useful as it reduces the frequency of high amplitude contractions [14]. If soiling persists despite dietary modification and medical therapy, a small volume enema program can be considered alone or in conjunction with medical therapy.

Most children with pseudo-incontinence will ultimately improve as they get older, with the majority being clean by the time they are five or six years old. A notable exception to this is the population with trisomy 21 and other forms of developmental delay. For most children, an optimistic approach should be taken with the family, which helps them to be motivated in managing the soiling issues.

Summary

This guideline presents expert-based consensus recommendations for the management of a patient with soiling after pull-through surgery for Hirschsprung disease. A clear understanding of the underlying etiology and a stepwise, logical approach to the diagnosis and management of patients experiencing soiling following pull-through for Hirschsprung disease will facilitate effective treatment and render the majority of such patients clean and socially continent.

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Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

Ethical approval This article does not contain any studies with human participants or animals performed by any of the authors.

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