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Reoperative surgery for Hirschsprung disease

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Despite most children undergoing a successful pull through for Hirschsprung disease, a small portion of children are left with persistent stooling issues. Most of these stooling issues can be addressed by nonoperative approaches. However, in a small group of remaining children, a reoperation may be necessary. Most children who may need a redo pull-through procedure may have a persistent area of aganglionosis, unremitting enterocolitis, or a torsion or stricture of the pull-through segment. Each of these influences the approach the surgeon must take to correct the presenting problem. The chapter details the diagnostic approach as well as the operative techniques, which best deal with each of these complications.

Congenital aganglionosis was named after Härold Hirschsprung who described the clinical features in 1886; however, there is new evidence which suggests there is recognition of the disease process dating several centuries before this.[1] Since Swenson and Bill[2] described successful treatment of Hirschsprung disease (HD) in 1948, there have been many modifications and advances in technique leading to improved quality of life for those affected by HD. Presently, the most common techniques are a transanal approach with or without laparoscopic assistance, a modified Soave pull-through and a Duhamel's retrorectal pull-through procedures. Many studies have attempted to tease out which of these is superior without consensus.[3-5] Each of these approaches can ultimately have very good long-term results. However, regardless of operative technique, many patients have long-term postoperative complications; and in some of these children, the type and extent of the morbidity may require reoperation to adequately address the child's problems.

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Long-term postoperative complications following pull though

A significant number of children whom undergo a successful pull through will develop postoperative complications. The most common and serious long-term complications after definitive treatment for HD can be divided into 3 groups: soiling/incontinence, persistent problems with the passage of stool (eg, constipation), and recurrent Hirschsprung-associated enterocolitis (HAEC).[6] The contributing factors resulting in these problems can be described as either pathologic or anatomic. The pathologic causes include residual aganglionosis or transitional zone pathology. The anatomic causes comprise stricture, retained dilated segment, obstructing Duhamel pouch or Soave (aganglionic) muscular cuff, and finally a twisted pull through. Lastly, immunologic and or physiological causes may contribute to the development of recurrent episodes of HAEC.

Recent reviews have described a wide range in the incidence of incontinence, which may range from 3% to 30%. Although most of these episodes can be treated with medical care, some have required an appendicostomy and, at times, a permanent stoma.[7] Postoperative HAEC ranges from 10% in some groups up to 45% in our own group of patients. However, the differences between series maybe more related to differences in definitions between centers.

Despite conservative treatment working in most cases, such approaches prove unsuccessful in nearly 20% of children. Many reports indicate that patients will outgrow most of these complications by the age of 5 years for unexplained reasons, without the need for further treatment. Unfortunately, this is not always the case; some children may develop treatment-resistant symptoms or recurrent episodes. In these latter cases, further intervention is required. Patients presenting with stooling problems require thorough and thoughtful workup. These complications are described brief. The reader should refer to earlier chapters on complications following pull-through procedures for further detail.

Incontinence

Soiling and incontinence are theoretically preventable by use of proper surgical techniques; however, many patients are affected even in the most experienced hands. Incontinence has been reported in variable percentages and degrees. Rates range from as little as none in several series[8-10] to as high as 76% in 1 series.[11] Soiling may simply be due to a loss of rectal sensation. Sensory rectal mucosa is critical for differentiating between gas, liquid, and solid stool, and it can be compromised after a very low anastomosis. Incontinence may also be due to scaring or iatrogenic damage to the anal sphincters. The surgeon must be cognizant of the fact that the level of the internal anal sphincters (IAS, the major contributor to continence) can be identified as the level of the dentate line, and thus all anastomoses must be performed at least 5 mm or more above this level.

However, the most common cause of soiling after a pull through is actually because of chronic constipation with encopresis. The treatment of incontinence ranges from a simple bowl management program to the placement of a permanent stoma depending on the underlying cause. A critical challenge for the surgeon evaluating a child with soiling is differentiating true incontinence, which is typically managed nonoperatively, from encopresis or HAEC. The latter 2 warrant continued workup and possible operative intervention.

Persistent stooling problems

Persistent stooling problems include recurrent distension, bloating, and constipation. Constipation is a common complaint after any pull-through procedure, although typically it does not develop for weeks to months post-operatively. Recent reports have recognized that obstructive symptoms are occurring in 9%[12,13]-40% of children after what appears to be a technically excellent operation. A common theme in the literature is that symptoms improve with greater follow-up, being reported as normal in 58% of those at <5 years of follow-up and 88% of those at greater than 15 years of follow-up.[14] The fact that, many children have persistent problems with their IAS or more proximal tissue even after removal of all aganglionic tissue suggest that our understanding of the pathophysiology of HD is still incomplete.

There are several causes of persistent stooling after a pull through. Anatomic causes including an anastomotic stricture or retained spur after Duhamel's procedure are common.[15] Other contributing processes, such as retained or acquired aganglionosis,[16] rarely hypoganglionosis[17] or transition zone pathology, which can be associated with either focal or general motility disorders are also frequently encountered. Less commonly, IAS achalasia (IASA), and "functional megacolon," a chronically dilated distal rectum secondary to chronic difficulty with passing bowel movements, can lead to persistent stooling problems.[18]

In some children there is no identifiable cause for their symptoms. Responses to the use of botulinum toxin injections for relaxation of the sphincters are widely variable.[19-22] Many of these children suffer from stool-holding behavior and are best treated by also using a bowel management regimen, consisting of laxatives, enemas, and behavior modification, including support for the child and family.[23] The approach to address all above causes varies, and one should be aware of all of these various possibilities to be able to successfully manage a complicated HD case.

Hirschsprung disease-associated enterocolitis

HAEC remains the most common cause of morbidity and death in HD.[24] The exact etiology and pathogenesis are still unclear.[24,25,26] There is no clinical factor or test that is available to predict the development of this disorder and no single strategy for prophylaxis from this major complication. In a multicenter review of primary endorectal pull through (ERPT), logistic regression analysis of risk factors for the development of enterocolitis showed that stricture development was a significant risk factor.[15] Based on this, it is suggested that in such cases, a redo pull through may, at times, be beneficial in controlling recurrent HAEC in some patients once they undergo a complete workup.

Principles for workup of postoperative complications following a pull-though

HD cases should be assessed thoroughly by experienced physicians at an institution that has the infrastructure to evaluate, diagnose, and treat these complex problems. Meticulous documentation should be maintained in any HD case as these records will prove useful for routine follow-up and essential for those requiring reoperation. In the case of referral, documentation of the neonatal period, associated anomalies, type of presentation, time and method of diagnosis, preoperative course, type of procedure, pathologic material, and detailed postoperative management should be carefully reviewed. An algorithm should be tailored for each case according to the type and nature of the complication (Figure 1).