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Transanal endorectal pull-through for Hirschsprung disease: technique, controversies, pearls, pitfalls, and an organized approach to the management of postoperative obstructive symptoms

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The transanal endorectal pull-through emerged in the late 1990s as the most recent step in the evolution of the surgical correction of Hirschsprung disease. This operation provides the advantages of a minimal access approach with shorter hospital stay, shorter time to full feeding, less pain, and improved cosmesis with excellent outcomes. This article will review the technical principles of the transanal endorectal pull-through, and will address ongoing controversies in the application of this technique. We will also discuss an organized approach to the problem of obstructive symptoms that may affect a subgroup of patients after the transanal pull-through.

Swenson first described definitive surgical management of neonates and infants with Hirschsprung disease in the late 1940s. Because these children often presented with severe malnutrition or enterocolitis, a preliminary colostomy was usually done, followed by a pull-through procedure many months later. Earlier recognition and diagnosis of the disease led a number of surgeons in the 1980s to report series of single-stage pull-through procedures even in small infants, using each of the 3 common operations (Swenson, Duhamel, and Soave). Since then, one-stage operations have become increasingly popular and many reports have suggested that this approach is safe, cost-effective, and avoids the morbidity of stomas in infants.[1]

In the early 1990s, Georgeson et al[2] described a minimal access approach, consisting of a laparoscopic biopsy to identify the transition zone, laparoscopic mobilization of the rectum below the peritoneal reflection, and a short endorectal mucosal dissection from below. The anastomosis was done from below after prolapsing and excising the rectum. Multiple reports documented a short time in the hospital, and early results were equivalent to those reported for the open procedures. Subsequently, laparoscopic approaches have been described for the Duhamel and Swenson operations,[3,4] with excellent short-term results reported.

The transanal Soave procedure represented a natural evolution from the laparoscopic operation. Transanal resection of the rectum was shown to be possible in an animal model, and initial series of children with Hirschsprung disease were

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published by de la Torre and Ortega-Salgado[5] and Langer et al[6] in the late 1990s. The transanal approach has the principal benefit of avoiding the need for intra-abdominal mobilization of the rectum through either laparotomy or laparoscopy. Several studies have demonstrated that the transanal approach is associated with less pain, shorter hospital stay, and a better cosmetic result than open surgery.[7-9] There have not been any studies comparing the transanal to the laparoscopic approach. However, the transanal pull-through can be done by any pediatric surgeon, including those without laparoscopic skills, and by pediatric surgeons in parts of the world where access to appropriately miniaturized laparoscopic equipment is limited.[10]

This article will review preoperative considerations before performing the transanal pull-through, technical aspects of the operation, pitfalls and pearls the authors have learned, and ongoing controversies. We will also present an organized approach to the common problem of obstructive symptoms after the transanal pull-through.

Preoperative considerations

Hirschsprung disease may present as neonatal intestinal obstruction, as constipation in an older child, or less commonly with primary enterocolitis. In the neonate, Hirschsprung disease must be differentiated from other causes of intestinal obstruction, including meconium ileus, intestinal atresia, anorectal malformation, malrotation, and congenital bands. Careful history and physical examination, abdominal radiogarph, and water-soluble contrast enema are the initial diagnostic maneuvers in most cases. The definitive diagnosis of Hirschsprung disease is either made or excluded on the basis of a suction rectal biopsy, looking for the presence or absence of ganglion cells and of hypertrophic nerves. Some pathologists also use cholinesterase staining to complement the standard histologic evaluation. The biopsy must not be taken too close to the pectinate line, because there is normally a paucity of ganglion cells in this location.[11] It is also important to initiate early resuscitation in infants with intestinal obstruction or enterocolitis, including the administration of intravenous fluids and antibiotics, and a nasogastric tube. Early decompression of the colon using digital rectal stimulation and/or irrigations through a rectal tube is important to prevent and treat enterocolitis and to decrease the diameter of the colon. Children with associated abnormalities, such as cardiac disease or congenital central hypoventilation syndrome, must have these problems dealt with before definitive surgical repair. After the child has been stabilized, the definitive surgical procedure can be done semielectively. During the waiting period, most children can be fed breast milk or an elemental formula, in combination with rectal stimulations or irrigations. Those who cannot tolerate oral or nasogastric feeding can be nourished with parenteral nutrition.

In the older child who presents with severe constipation, a suction biopsy is less likely to give the pathologist enough submucosa to make the diagnosis. In these children a deep or full-thickness biopsy must be done, usually under general anesthesia. In some centers, anorectal manometry is also used as a screening test, because the presence of a rectoanal inhibitory reflex effectively rules out the diagnosis of Hirschsprung disease. Absence of a recto-anal inhibitory reflex must be followed by a rectal biopsy to confirm the diagnosis, as there is a significant false-positive rate for this test. Older children who present with an extremely dilated colon may require weeks or months of irrigations to bring the colon to a more normal size before the definitive surgical procedure, and in some children with particularly dilated or thickened proximal colon, a preliminary colostomy may be necessary to achieve adequate decompression.

Most children with Hirschsprung disease have a contrast enema done as part of the diagnostic workup, looking for the presence and location of a radiological transition zone. However, many studies have documented that a maximum of 10% of neonates with Hirschsprung disease will not have a transition zone on contrast enema.[12] In addition, older children with a very short aganglionic segment may not demonstrate a transition zone on contrast enema, particularly if the catheter has been placed above the transition zone in the rectum. Finally, the contrast study is not always completely accurate in identifying the location of the pathologic transition zone, with 12% of cases having a pathologic transition zone which is different from the radiological transition zone.[13] It may be possible to increase the accuracy of the contrast enema in identifying the transition zone in older children by waiting until histopathological confirmation of the disease is available, and discontinuing the rectal irrigations for 1-3 days before performing the contrast enema. It is also important to recognize that the lateral projection is better than the anteroposterior view in identifying a rectal or rectosigmoid transition zone (Figure 1).

Surgical technique

Basic principles of the operation

After induction of anesthesia, before beginning the operation, a single shot caudal block should be placed to minimize the need for anesthetic agents. We routinely repeat the block again at the end of the procedure to provide early postoperative pain relief. All patients should receive prophylactic antibiotics, such as cefoxitin to cover Gram-negative bacilli and colonic anaerobes.

The operation starts with a mucosal incision above the dentate line. The distance above the dentate line depends on the surgeon and the size of the child, but it is crucial that the incision be high enough above the dentate line so that the transitional epithelium is not damaged. This is important to prevent loss of sensation, which may predispose the child to long-term problems with incontinence.

Fine silk sutures are placed in the mucosa, either before or after the mucosal incision, to provide traction on the