27.1 Introduction

The surgical treatment of Hirschsprung’s disease has gone through several changes since Ehrenpreis in 1946 suggested that functional obstruction due to aganglionosis of the distal colon was the cause of the proximal colonic dilatation [1]. Swenson and Bill developed the first operation in 1948 that was successful in removing the aganglionic segment and establishing intestinal continuity [2]. Later, Duhamel developed the retrorectal anastomosis for Hirschsprung’s disease [3] and Soave developed the endorectal pull-through [4]. Prior to the development of these operations, the only successful surgical procedure was a diverting colostomy.

Until recently these operations were always performed in two or three stages, the first stage being the placement of a diverting colostomy or ileostomy. “Leveling” colonic biopsies were obtained to determine the location of the transition zone as part of this first operation. The second stage was performed later, usually between 3 months and 1 year of age. This operation involved removing the aganglionic segment and creating an anastomosis between the normally innervated small bowel or colon and the anus. Some surgeons preferred to protect the anastomosis with a proximal loop stoma, and close the stoma at a third operation.

Over the past two decades it has become increasingly recognized that the routine use of a colostomy is unnecessary, and an increasing number of pediatric surgeons perform the reconstruction as a single stage procedure at an early age.

In this chapter, the transanal endorectal pull-through operation is described. The development of this operation was the result of a number of advances in the treatment of Hirschsprung’s disease, including earlier diagnosis, better preoperative, perioperative and postoperative care, and the development and popularization of minimal access techniques in pediatric surgery.

27.2 Primary Pull-Through

In 1980 So et al. described the use of the Soave endorectal procedure as a single stage pull-through without a preliminary colostomy [5]. These and several other authors did the operation at several months age, managing the children with total parenteral nutrition (TPN) or elemental feeding and colonic irrigations to prevent retention of stool and secondary enterocolitis [5–7]. Since these initial reports there have been many single and multi-institution studies published that demonstrate the safety of single-stage repairs using each of the commonly performed operations [8–10]. In addition to minimizing the rate of complications due to the presence of a stoma [11] and decreasing the number of hospitalizations and cost [12], the avoidance of a colostomy has dramatically improved the quality of care to children with Hirschsprung’s disease in developing countries, where multiple visits to the hospital may be impractical and the presence of a stoma represents a significant social stigma [13].
27.3 Development of the Transanal Pull-Through

With the rapid development of laparoscopic techniques in the early 1990s, pediatric surgeons began to move toward less invasive surgical procedures for a wide variety of conditions. Georgeson et al. reported a technique utilizing laparoscopic dissection of the rectum combined with an anal mucosal dissection in 1995 [14] and subsequently laparoscopic approaches to both the Duhamel and Swenson procedures were described [15, 16]. These are discussed elsewhere in this volume.

The transanal pull-through was an extension of the procedure of Georgeson et al., but without the laparoscopic intraperitoneal dissection. This approach was concurrently described by De la Torre-Mondragon and Ortega-Salgado [17] and by Langer et al. [18] in 1998 and 1999. Since then the technique has evolved and a number of variations have been described. Despite the fact that this technique has now been widely adopted by surgeons all over the world, there remain a number of controversies regarding the optimal approach. The technique is described in this chapter, the data supporting its use is reviewed, and the ongoing controversies in its use are outlined.

27.4 Surgical Technique

The technique is illustrated in Fig. 27.1.

27.4.1 Preoperative Preparation

The diagnosis is confirmed by rectal biopsy. Prior to surgery, the colon must be decompressed and enterocolitis, if present, controlled. Nutritional status must also be evaluated and optimized. It has been shown that even patients presenting with intestinal obstruction or enterocolitis may respond well to aggressive nonsurgical management with antibiotics, decompression, and support [19]. In an older child with severe enterocolitis or massive colonic distension, a defunctioning stoma should be considered. We have found that routine preoperative mechanical bowel preparation is unnecessary and can cause significant distension and vomiting. Mechanical irrigation of the bowel can be accomplished with equal effectiveness from below once the child has undergone anesthesia. Intravenous prophylactic broad spectrum antibiotics are used in all patients.

27.4.2 Anesthesia

The operation is done under general anesthesia. In addition, a caudal block done at the beginning and at the end of the procedure provides excellent intraoperative anesthesia and postoperative analgesia.

27.4.3 Positioning

The patient is placed in the lithotomy position either transversely or longitudinally at the end of the operating table. The transverse position is particularly advantageous if laparoscopy is to be used for preliminary biopsies. The rectum and sigmoid colon are irrigated from below until clear. A urinary catheter is optional. We tend not to use one, choosing to intermittently empty the bladder with a Crede maneuver during the procedure. Some surgeons prefer the prone jackknife position for the transanal pull-through. Although this position provides excellent visualization, it makes concurrent use of laparotomy or laparoscopy, for biopsies or mobilization, impossible.

27.4.4 Submucosal Dissection

An anal retractor or retraction sutures are placed to expose the anus and distal rectal mucosa. Some authors recommend submucosal injection of a dilute epinephrine solution or air to enhance development of the submucosal plane [20]. The rectal mucosa is circumferentially incised using cautery approximately 3–5 mm from the dentate line, depending on the size of the child. Multiple fine sutures are placed in the proximal cut edge of the mucosal cuff, and traction is applied while the endorectal submucosal dissection is carried proximally. The optimal length of the submucosal dissection is a subject of controversy, and is addressed below.

27.4.5 Mobilization of the Rectum

When the submucosal dissection has been completed, the rectal muscle is divided circumferentially. Dissection then continues proximally, dividing all vessels as they enter the rectum, staying right on the rectal wall. When the peritoneal reflection is reached, the sigmoid is then mobilized in the same fashion and the rectum and sigmoid are delivered through the anus. Throughout this dissection, blood vessels are divided using cautery or ligated, depending on their size.

27.4.6 Anastomosis

The colonic dissection is completed when the transition zone is reached. The controversy about whether the pathological transition zone should be identified before the anal dissection or during mobilization of the rectum and sigmoid colon is addressed below. The colon is divided at least 2 cm above the most distal normal biopsy to prevent the possibility of a transition zone pull-through [21, 22]. The rectal muscular cuff is then split longitudinally, ei-