Chapter 25

Soave's Extramucosal Endorectal Pull-Through Procedure

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25.1 History of the Endorectal Pull-Through Procedure

The use of the abdominal extramucosal dissection of the rectal pouch was first proposed by Romualdi at the Roman Society of Surgery on 15 May 1955. The technique was first published in 1960 [1]. During the next few years, Rehbein [2] and Kiesewetter and Turner [3] also popularized this operation. In 1957, Soave started using Romualdi's procedure for the treatment of ano-rectal malformations with urethral fistula. Since his initial experience with Romualdi's technique, he thought that this principle could be applied for the treatment of Hirschsprung's disease (HSCR). So, in 1961, he performed his first operation on a 2-year-old boy with the classic form of HSCR. After treating six children with endorectal pull-through, Soave reported his first results [4, 5]. The technique of separating the mucous coat from the muscular layer of the rectum for the treatment of HSCR was experimented with during about the same years by Soave and Boley. However, Soave's first report [4] on endorectal pull-through without anastomosis dates back to 1963, and other internationally more well-known reports [5, 6] to 1963 and 1964. In the same 1964 issue of Surgery, Boley [7] first presented a technique using the same endorectal approach but with a primary anastomosis of the pulled-through colon. In a short time, a large number of pediatric surgeons from all over the world were experimenting with the endorectal pull-through, and the indication for Soave's procedure was extended to other conditions including multiple juvenile polyposis, familial polyposis, and ulcerative colitis.

25.2 Preparation for Soave's Procedure

Up to the 1980s, the endorectal pull-through was generally performed in patients older than 5 months and weighing more than 8 kg. In those days, it was necessary to have a clear clinical and radiological picture to confirm the diagnosis of HSCR. Today, the histochemical preoperative diagnosis based on acetylcholinesterase activity [9, 10] can be made in the neonatal period without anesthesia using a suction rectal biopsy tool, the Solo-RBT, which was developed in 2000 at the Gaslini Institute [11]. The availability of an early and reliable diagnosis has led pediatric surgeons to perform, when possible, radical surgery within the first 3 months of life. In this way, the conservative period of nursing can be reduced to a minimum, decreasing the risk of complications and avoiding chronic mucositis of the rectum and colon due to the prolonged daily evacuating enemas. These alterations are well known to surgeons who have performed Soave's procedure in older children (more than 6–10 years of age) undergoing prolonged daily nursing maneuvers. In these subjects, endorectal dissection is very difficult due to
the tenacious adhesions on the submucous layer caused by chronic proctitis. If Soave's procedure is performed within the first 3 months of life—a very common approach today among pediatric surgeons—chronic inflammatory processes of the rectum are generally avoided and endorectal dissection between the internal submucous layer and the external muscular cylinder is straightforward and rapid.

Soave's procedure does not require any protective colostomy. The aim is to achieve radical treatment without contaminating the operative field at all. Therefore, colostomy is only indicated for the subgroup of HSCR patients presenting with acute enterocolitis or intestinal obstruction in the first weeks of life. When necessary, colostomy should be performed in the most distal portion of the ganglionic colon (level colostomy). In order to achieve this, it is necessary to perform multiple intraoperative seromuscular biopsies of the colon, using, if possible, histochemical techniques to evaluate the length of the aganglionic and hypoganglionic segments. At least four good intraoperative histochemical techniques are available today: succinic dehydrogenase [12] (SDH), lactic dehydrogenase [13] (LDH), alpha-naphthylesterase [14–18] (ANE), and rapid acetylcholinesterase [19, 20] (rapid-AChE). The level colostomy has to be terminal and part of the aganglionic distal colon has to be resected up to the rectosigmoid junction, where it is sutured and plunged. We choose a level terminal colostomy for the following reasons: none of the normoganglionic colon is resected and lost; the colostomy is not formed in an aganglionic or hypoganglionic segment, thus the risk of a second pull-through operation using these segments with abnormal innervation is avoided; and the number of operations is reduced from three to two, since the radical treatment is performed by mobilizing the level colostomy avoiding a subsequent operation for colostomy closure.

The preparation of the bowel the week before Soave's radical treatment is essential to reduce complications. The colon has to be cleaned with special enema preparations (we use a mixture of sorbitol enema or phosphate saline enema, saline solution and Vaseline oil). During the days preceding surgery, rectal probing should be repeated and alternated with evacuating enemas. Perioperative intravenous antibiotic prophylaxis is started 1 hour before surgery.

### 25.3 Operative Technique

#### 25.3.1 Positioning of the Patient

When the patient is under general anesthesia and a catheter has been inserted into the bladder, before the definitive positioning of the patient for the operation, the surgeon has to perform a wide dilatation of the anal canal. This procedure is performed using the two forefingers: the anorectal canal is dilated by traction in opposite directions. This maneuver is always essential for a successful pull-through procedure, especially in patients under 1 year of age. The patient should be positioned supine with the buttocks lying at the extreme edge of the operating table and the legs hanging freely, wrapped in drapes, and fixed to the table to prevent slipping of the patient's pelvis (Fig. 25.1a).

#### 25.3.2 Laparotomy

Different incisions may be performed for Soave's procedure. A good alternative to the classic paramedian left incision is the Pfannenstiel incision, which leads to better cosmetic results. It is performed above the pubis and is sufficient for the treatment of rectal and rectosigmoid forms of aganglionosis. In patients in whom the disease involves the descending colon, the Pfannenstiel incision can be extended in the left pararectal direction. In patients with level ileostomy for total colonic aganglionosis, a xiphopubic median incision is required in order to perform an endorectal ileal pull-through procedure. The operating field is exposed with malleable retractors, and all mesocolic adhesions to the left parietal peritoneum are dissected up to the splenic flexure (classic form of HSCR). Before starting endorectal dissection, it is essential to perform multiple seromuscular biopsies of the rectum and colon in order to evaluate the length of the aganglionic and associated hypoganglionic segments. A better evaluation of the segment to be resected is possible thanks to specific intraoperative enzymohistochemical techniques [12–20].

#### 25.3.3 Separation of the Seromuscular from the Mucosal Layer of the Rectum

This is the most technically difficult and peculiar step of Soave's procedure. In order to facilitate the initial separation of the seromuscular from the mucosal layer of the rectum, mepivacaina 2% with epinephrine 1:100,000 (10 μg) in 10 ml of normal saline solution is injected between the layers (Fig. 25.1b).

A longitudinal seromuscular incision is made on the previously infiltrated anterior wall of the rectum (Fig. 25.1c). It is important not to start dissection at a more proximal point to avoid a too-large dissection surface. The incision (Fig. 25.1c) is widened with blunt dissection, first on the long axis of the bowel and then progressively laterally and posteriorly (Fig. 25.1d). The edges of the seromuscular layers are held by atraumatic Williams forceps to allow traction (Fig. 25.1d). The mucosal tube is now freed completely (Fig. 25.2) and the blunt dissection of the seromuscular cuff is progressively carried downward, taking great care not to tear the mus-