ORIGINAL ARTICLE

G. Wang · J. Yuan · X. Zhou · B. Qi · D. H. Teitelbaum

A modified operation for Hirschsprung's disease:

Posterior longitudinal anorectal split with a "heart-shaped" anastomosis

Abstract A variety of complications can arise after the definitive pull-through operation for Hirschsprung's disease. Among these are constipation and soiling, which may be due to mismanagement of the internal anal sphincter. In order to prevent these complications, we developed an improved operative procedure based on preservation of the anterior anorectum that utilizes a posterior longitudinal split with an oblique anastomosis. Forty patients underwent this procedure over a 4-year period; adequate follow-up was obtained in 34 cases. Mean age at operation was 13 months (range 3 months to 7 years). A primary pull-through without a colostomy was performed in 38 of the 40 patients. The results showed excellent preservation of function, with a mean follow-up of 5 years and 7 months. There was no occurrence of spastic stenosis of the internal sphincter, no incontinence, and minimal constipation or soiling.

Key words Hirschsprung's disease · Soiling · Constipation · Surgical treatment

Introduction

A variety of definitive operations exist for Hirschsprung's disease (HD), including those described by Swenson, Duhamel, Soave, and Rehbein [1]. Each of these repairs is highly effective in the relief of functional obstruction, and many authors feel that the outcome is predominantly

G. Wang (✉) · J. Yuan · X. Zhou · B. Qi
Section of Pediatric Surgery, Affiliated Tongji Hospital,
Tongji Medical University, Wuhan Hubei, 430030,
People's Republic of China

D. H. Teitelbaum
The Section of Pediatric Surgery,
University of Michigan Medical School, Ann Arbor, MI, USA

dependent upon the experience of the surgeon rather than the performance of any particular procedure. Each of these operations has its own inherent flaws leading to both early and late complications. Among these are the occurrence of either constipation or soiling in approximately 8% to 12% of cases [1–6]. We felt that many of these complications occurred because of mismanagement of the internal sphincter, and have designed a new operative method in which we perform a posterior longitudinal anorectal split with an obliquely shaped (heart-shaped) anastomosis. We describe here the operative technique and present our current results with this method.

Materials and methods

All cases of HD were diagnosed on the basis of clinical history, radiologic studies, and rectal biopsy using standard hematoxylin and eosin as well as acetylcholinesterase stains. Isotonic saline was used to mechanically clean the colon for up to 10–14 days prior to surgery. Oral neomycin was given the day before surgery and IV metronidazole was used through the 3rd postoperative day. We found it unnecessary to place a urethral catheter. The bladder was compressed just after opening and before closing the abdomen; all patients resumed spontaneous micturition within 3 h after operation.

A low left transverse incision is made extending slightly to the right of the midline. The peritoneal reflection from the rectum is dissected on both left and right sides down to just above the level of the dentate line. To protect the pelvic autonomic nervous system, the anterior rectal wall is left undisturbed and the upper one-third of the lateral rectal ligaments are dissected as close as possible to the rectal wall. Hemostasis is achieved with a sponge pressed into the posterior cavity behind the rectum. The proximal ganglionic bowel is identified by operative biopsies and frozen sections. Ganglionic bowel is then dissected and widely mobilized up to the splenic flexure to allow for a tension-free anastomosis with an adequate blood supply.

Attention is now directed to the perineum. The anus is widely dilated. The operator inserts an esophageal dilator through the anus into the lumen of the rectosigmoid and fastens the aganglionic rectum to the dilator at the transition level. The bowel is then prolapsed out of the rectum, everting it (Fig. 1). The rectum is transected and the ganglionic bowel is pulled through the anal canal. The most dilated distal portion of ganglionic bowel is resected at this point. The posterior wall of the aganglionic anorectum is split longitudinally to the level of the dentate line (Fig. 2). The tips of the two halves are trimmed so that the remaining rectal wall, whose anterior aspect is

Dentate Line
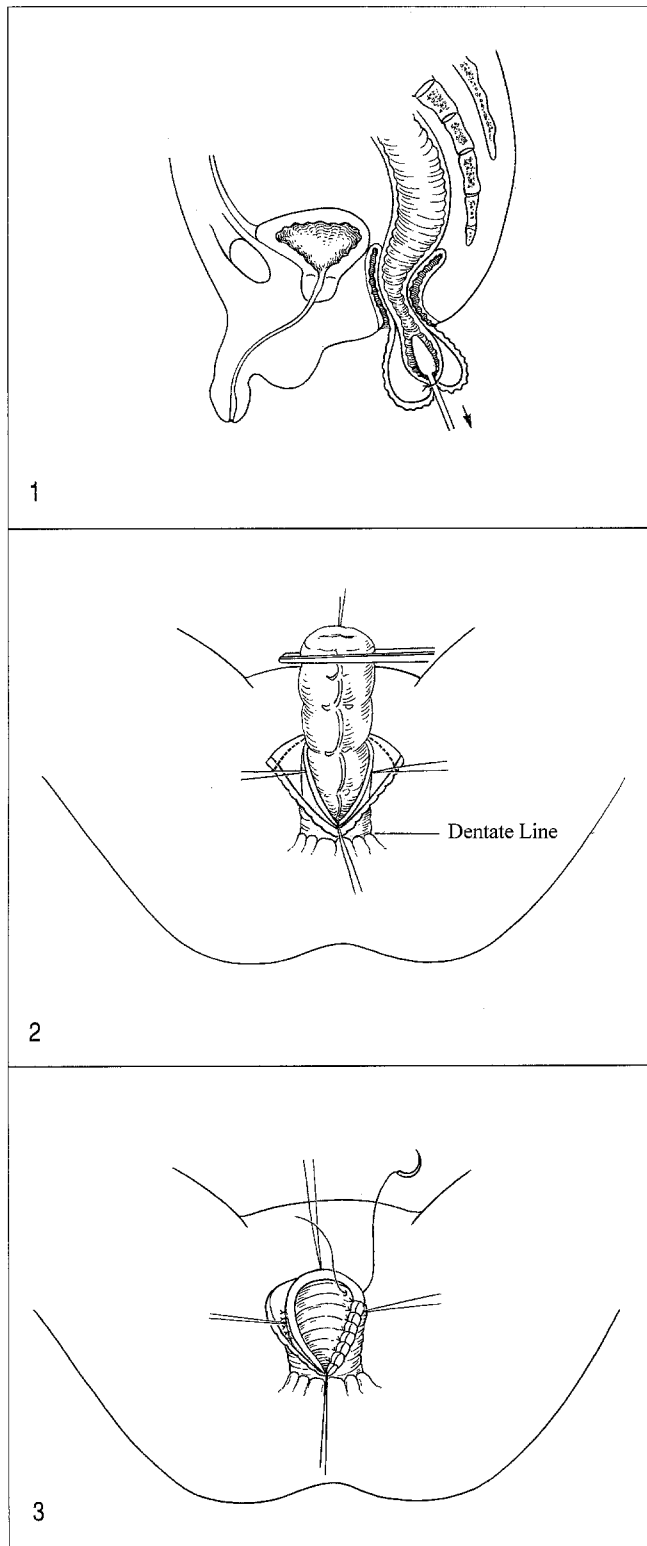
Fig. 1 Transrectal dilator, which has been tied around ganglionic bowel at transition zone, is pulled through rectum. Bowel is everted until mucocutaneous line is exposed posteriorly, but not anteriorly

Fig. 2 The most dilated bowel has been resected, exposing ganglionic bowel. Note longitudinal split in posterior wall of anorectal canal to dentate line

Fig. 3 Suturing of seromuscular coats of rectum and colon, facilitated by bowel being everted and exteriorized out of anus

longer than the posterior one, has the shape of a heart. Interrupted sutures are placed circumferentially at each quadrant through the seromuscular coats of the proximal bowel and the full-thickness edge of the transected rectum. Each suture is tied and grasped with clamps to prevent retraction (Fig. 3). Subsequent sutures are added to each quadrant as full-thickness bites to complete the anastomosis. In order to prevent leaking, the posterior wall must be meticulously sutured. A cigarette-type drain is placed in the pelvis and removed on the 3rd postoperative day. At the completion of the procedure, the anterior anastomosis is 4 cm above the anal verge and the posterior anastomosis 1.5 to 2.0 cm above the anodermal junction (Fig. 4 A and B).

Results

From January 1986 to June 1990, we performed this operation on 40 patients. A primary pull-through was achieved in 38 with the remaining 2 receiving a temporary diverting colostomy. The mean age at the time of the pull-through was 13 months (range 3 months to 7 years). In the postoperative period, there were no abdominal infections and no anastomotic leaks. Complete follow-up was obtained in 34 of these children, and ranged from 2 to 6.5 years (mean 5 years 7 months). In 20 cases follow-up was in our surgery clinic and in the remaining 14 by written correspondence from the patient's home hospital. No dilatations were performed in these patients other than rectal examinations on follow-up visits.

None of the patients developed incontinence, nor was any stenosis of the anastomosis noted. Two children had occasional soiling, which corrected itself with alterations in their diet. One child has intermittent constipation for which he is treated with enemas and laxatives. The remaining 33 have satisfactory stooling patterns. No incontinence or significant constipated episodes have been noted and no cases of enterocolitis have occurred.

Discussion

The primary goal in performing an endorectal pull-through for HD is to resect the maximum amount of aganglionic bowel while preserving a sufficient portion of the child's internal sphincter [1]. These components are difficult to balance. If too much of the internal sphincter is excised, incontinence or soiling can occur. In contrast, if too much aganglionic bowel is preserved, recurrent constipation may develop [2]. Each of the established pull-through procedures achieves these objectives in different ways. The Swenson procedure allows for resection of the maximal amount of aganglionic bowel, but risks greater injury to the internal sphincter as well as the deep pelvic nerves. The Soave endorectal pull-through leaves the aganglionic muscular cuff intact, but allows for maximal preservation of the internal sphincter, thus potentially leaving the patient at greater risk for internal sphincter spasm. The Duhamel procedure lies somewhere between: a large portion of the aganglionic bowel is preserved, as is most of the internal sphincter, which lies distal to the anastomosis. However, by
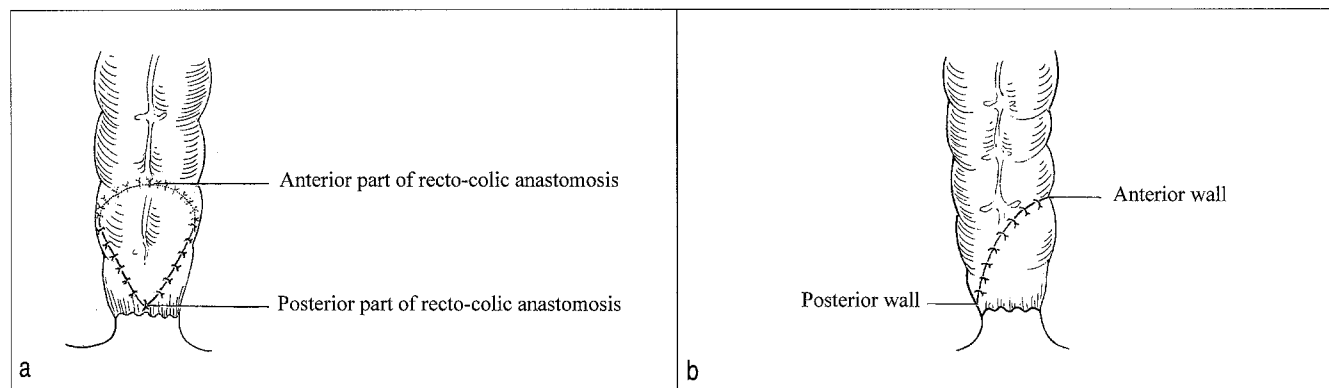
Fig. 4 a Cut-away view of anastomosis. Note that anastomotic ring does not lie in same plane: anterior portion is more proximal than posterior wall. b Lateral view of anastomosis. Anterior wall is 4 cm and posterior wall 1.5 to 2.0 cm above anal verge

performing a vertical incision posteriorly, the internal sphincter is less likely to become spastic [1].

The technique described here utilizes Several of the approaches from these standard techniques in order to maximize the resection of aganglionic bowel while preserving a major portion of the internal sphincter. Similar to the Swenson procedure, our "heart-shaped" technique requires dissection of the aganglionic bowel down into the pelvis, preserving the distal anterior rectal wall. Unlike the former, however, our technique preserves the majority of the internal sphincter and has additional similarities to the Duhamel technique. In both procedures the anterior, aganglionic rectum is left alone and the posterior wall of the aganglionic rectum is transected vertically. Thus, only a small portion of the internal sphincter is transected, with interposition of ganglionic bowel. Our procedure differs from Duhamel's in the large amount of aganglionic bowel that is left with the latter operation. The total length of aganglionic bowel is only 3 to 4 cm in our procedure, but may be up to twice that in Duhamel's [1]. Our anastomosis is also performed completely outside of the abdominal cavity, which may account for the low incidence of postoperative infections.

We have found that our present technique not only prevents the development of stenosis or spasm of the internal sphincter, but the long-term results are excellent in terms of continence and soiling. We have also noticed a very low incidence of postoperative enterocolitis compared to other series [6–8], and speculate that the low occurrence may be related to the minimal degree of internal sphincter manipulation. Only 1 child has persistent constipation; his father and brother both suffer from congenital deafness and other abnormalities, and it is suspected that this child is neurologically impaired, which may well contribute to his constipation [9].

The advantages of this technique are numerous. First, it can be accomplished in one stage without the need for a colostomy. Even a young infant can tolerate the operation. Second, the rectocolic anastomosis is performed end-to-end, and therefore a blind rectal pouch or septum, which may be encountered in the Duhamel technique, cannot occur. In addition, the configuration of the anastomosis is heart-shaped, which, similar to the oblique anastomosis of a

Swenson procedure, makes it less likely that an anastomotic stricture will develop. Because of this large anastomosis, there was no need to dilate any of our patients. The technique is not difficult and no special clamps or stapling devices are required; the complications resulting from these clamps are thus avoided. Finally, not only is the internal sphincter preserved to a maximum degree, but by making a posterior incision the syndrome of internal sphincter spasm is avoided, thereby reducing the incidence of both soiling and constipation.

In conclusion, we describe a new pull-through technique for HD. The results have been excellent to date. Further follow-up of all 100 children will hopefully continue to show excellent long-term results.

References

1. Holschnelider AM (1982) Hirschsprung's disease. Thieme-Stratton, New York, pp 230–231
2. Wang G (1978) Long-term complications after operations for Hirschsprung's disease. Wuhan J N Med 3: 31
3. Polley TZ Jr, Coran AG, Wesley JR (1985) A ten-year experience with ninety-two cases of Hirschsprung's disease including sixty-seven consecutive endorectal pull-through procedures. Ann Surg 202: 349–355
4. Zhang S (1988) Complications of Duhamel operation. Chin J Pediatr Surg 9: 107
5. Swenson O, Sherman JO, Fisher JH, et al (1975) The treatment and postoperative complications of congenital megacolon: A 25 year follow-up. Ann Surg 182: 266
6. Kleinhaus S, Boley SJ, Sheran M, et al (1979) Hirschsprung's disease; a survey of the members of the Surgical Section of the American Academy of Pediatrics. J Pediatr Surg 14: 588
7. Ikeda K, Goto S (1984) Diagnosis and treatment of Hirschsprung's disease in Japan. Ann Surg 199: 400
8. Teitelbaum DH, Qualman SJ, Caniano DA (1988) Hirschsprung's disease: identification of risk factors for enterocolitis. Ann Surg 207: 240–244
9. Molander ML (1988) Hirschsprung's disease in mentally retarded patients: a bad prognostic combination. Pediatr Surg Int 5: 339