Operative Techniques

Transanal rectal mucosectomy and partial internal anal sphincterectomy for Hirschsprung’s disease

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Abstract

Purpose: Hirschsprung-associated enterocolitis (HAEC) is a serious complication of Hirschsprung’s disease (HD), with generalized sepsis and high mortality rate. Although the surgical correction of HD is mostly technically successful surgery, a condition referred as Hirschsprung-associated enterocolitis (HAEC). The etiology of HAEC is controversial. HAEC is thought to be caused by multiple factors including deficient submucosal immunity, abnormal peristalsis of the remaining intestine with ganglion cells and lower expression of Cdx genes in rectal mucosa [1]. The internal anal sphincter (IAS) is the lowermost part of the circular muscle coat of the alimentary canal and normally receives an innervation from the distal intramural ganglia. However, the reflex response of IAS is absent owing to the absence of ganglion cells in the myenteric and submucosalplexuses of the distal intestine in HD. This is thought to be one of the contributing factors of HAEC [2]. The most common theory is that stasis caused by functional obstruction by the aganglionic internal anal sphincter permits bacterial overgrowth with secondary infection. There are reports showing that those who underwent internal sphincter resection had a satisfactory result, with reduced rates of the postoperative enteritis and

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An abnormality in the development of the autonomic nervous system results in absent peristalsis in the affected bowel and the development of a functional intestinal obstruction. Treatment consists of a surgical pull-through operation resecting the aganglionic portion of colon. However, 20–38% of HD patients may develop enterocolitis despite...
constipation [3–8]. Therefore, we proposed that the removal of rectal mucosa and internal anal sphincter in the treatment of HAEC is necessary to minimize HAEC. To establish the effectiveness of transanal rectal mucosectomy and partial internal anal sphincterectomy (TRM-PIAS) in HD treatment and preventing HAEC, we reviewed our results in this study.

1. Materials and methods

Between July 2001 to December 2006, one hundred twenty-seven children with HD (eighty-nine boys and thirty-eight girls, age: 8 days–16 years, mean: 0.96 years) underwent TRM-PIAS in our hospital. All of patients with HD were diagnosed by barium enema preoperatively, anorectal manometries and biopsy pathologies. There were 21 children with short or ultrashort type of HD, 92 with rectosigmoid type and 14 with long segment type. Enterocolitis was defined as the occurrence of a clinical syndrome consisting of diarrhea, fever (>38 °C), abdominal distension, crampy abdominal pain, and lethargy, with or without positive stool culture findings for pathological bacteria [8]. In this study, patients with enterocolitis suffered from repeated episodes of enterocolitis. The primary operative procedures were performed in 116 patients (36 patients with enterocolitis), and 11 patients underwent two stage operative procedures (7 patients with enterocolitis).

1.1. Operative technique

The patient was placed in the lithotomy position. The anal retractors inserted allowed full visualization of the anal canal and lower rectum (Fig. 1). The anus was everted by stitches, and the anorectal line was brought to the level of the anal skin (Fig. 2). The mucosa and internal anal sphincter were incised circumferentially at the junction between rectal and anal mucosa (anorectal line). The plane between the internal and external anal sphincters was exposed by the incision of the anorectal line. The boundaries between the internal and external anal sphincters were identified by the following criteria: 1. The external anal sphincter contracts in response to the stimulation of electrocautery, and the internal anal sphincter does not have this response. 2. The color of external anal sphincter is pink, and the internal anal sphincter is white. 3. There is a layer of connective tissue between the external and internal anal sphincters.

Anterior dissection was conducted between the incision (anorectal line) and the peritoneal reflection. To minimize damaging of the urethra, vagina, prostate and nerves around the rectum, the anterior rectal wall was dissected along the submucosal layer and the rectal muscular sleeve remained intact (Fig. 3). The posterior dissection was performed along the cleavage between internal and external anal sphincters, progressed closely to the rectal muscular wall and dissection level reached to the pelvic (Fig. 4). The pathway of dissection during operation was shown in Fig. 5. Our operative principles are: (1) the areas of resection include aganglionic segment, rectal mucosa, posterior half muscular cuff and part of internal anal sphincter and (2) leaving the anal mucosa, external anal sphincter, pelvic muscle, longitudinal muscle complex and nerves around the rectum intact. At the end, the normal colon was pulled through and anastomosed to anal mucosa with 5-0 absorbable suture (Fig. 6).

The patients started with fluid diet on postoperative 1 day and normal diet from postoperative 3 days onwards. Anal dilatation was started from 14 days for 3 months postoperatively.

1.2. Follow-up

All patients were followed up for 6–12 years (median: 8.2 years). The median age at last follow-up was 12.2 years (7.2–20.1 years).

Fig. 1. The anus was retracted by stitches, and the anal canal and lower rectum were exposed.

Fig. 2. The relationship between the anorectal line and anal sphincters changed in transanal pullthrough.
Follow-up data were obtained by review of medical records (in-patient, outpatient, and follow-up investigations) and telephone interviews performed by one of the authors. The diagnostic criteria of constipation are at least two weeks of scybalous, pebble-like, hard stools for a majority of stools; or firm stools two or less times/week [9]. Soiling is defined as involuntary leaking of small amounts of stool, requiring change of underwear or diapers [10]. In this study, the diagnostic criterion of soiling is soiling three or more times/week, and lasted over 6 months. The defecation frequency per day, occurrence of enterocolitis, weight and nutritional status were recorded at each visit. In the study, 25 age-matched children without defecation dysfunction underwent anorectal manometry, and were used as the control group in anorectal manometry study. The preoperative and postoperative anorectal resting pressure was measured in all of children with HD and compared.

1.3. Statistical analysis

Data (the mean ± SD) were analyzed using paired-samples t test. Statistical analysis was carried out using the SPSS 13.0 (SPSS Inc, Chicago, IL) software program package. Results were considered statistically significant when P value was less than 0.05.

2. Results

TRM-PIAS was successfully performed in all patients. The mean operative time was 95 minutes. There was no one with anastomotic leakage postoperatively. The incidence of enterocolitis decreased from 33.9% (43/127) preoperatively to 1.6% (2/127) postoperatively (P < 0.01). The incidence of constipation decreased from 100% (127/127) preoperatively to 2.4% (3/127) postoperatively (P < 0.01).

Anorectal manometries showed that the anorectal resting pressure significantly reduced from 37.9 ± 12.5 mm Hg preoperatively to 20.2 ± 6.4 mm Hg on postoperative 1 month and 24.8 ± 9.9 mm Hg on postoperative 6 months, which were similar to age-matched normal controls (27.9 ± 9.6 mm Hg, P > 0.05). The anorectal resting pressure was in general leveling off after 6 months.
During the follow-up, except for 3 patients with constipation, the daily defecation frequency was increased postoperatively in all of patients. However, the defecation frequency decreased slowly from 3–17 times/day in the first week to 1–3 times/day sixth month later. The soiling incidence decreased gradually from 32.3% (41/127) 1 month postoperatively to 1.6% (2/127) 6 months later. No patient developed incontinence postoperatively.

3. Discussion

In 1971, Kasai et al [3] introduced the internal anal sphincterectomy and removal of the muscular layer in the posterior rectal wall for the treatment of HD, which had a good defecation results postoperatively. However, in this procedure, only the muscular layer of the posterior rectal wall was incised caudally as far as the internal anal sphincter, leaving the mucosal layer intact, and the friable mucosa of the myotomized rectum was then Anastomosed to the proximal colon. However, there is considerable risk of anastomotic leakage with this technique, which occurred in 2 of his 11 patients. In this study, we described a modified procedure in which patients with HD underwent transanal rectal mucosectomy and partial internal anal sphincterectomy (TRM-PIAS). In our study, the removal of rectal mucosa was performed, and the normal colon was pulled through and Anastomosed to anal mucosa, which eliminated the risk of anastomotic leakage reported by Kasai et al [3]. There was no one with anastomotic leakage postoperatively. In addition, our transanal approach has the advantage of eliminating the risks of intraabdominal pelvic dissection, such as bleeding, injury to other organs, and adhesion formation.

It has been suggested that postoperative enterocolitis is the most reliable indication of the successful or unsuccessful relief of the low intestinal obstruction of HD [11]. In our study, the postoperative incidence of enterocolitis was 1.6%, which was low. Although we do not have a control group in this retrospective study, the incidence of postoperative enterocolitis is lower than those of other reported procedures (10.8% by Rintala RJ [12], 22.2% by Langer JC [13], 56% by Van Leeuwen K [14] and 23.5% by Liu DC [15]).

In this study, partial internal anal sphincterectomy was performed. Naturally, one would be concerned with postoperative incontinence. There is a study which reports that defecation function is not hampered in patients who underwent the internal sphincterectomy [16]. In addition, reflex response of the IAS is absent and preserving IAS poses a risk of postoperative enterocolitis and constipation in HD. Iwai [17] reported that the anal resting pressure of patients undergoing Rehbein’s procedure and anorectal myectomy were significantly lower than those undergoing Rehbein’s procedure alone. It was demonstrated that anorectal myectomy is a more definitive procedure to restore the anal resting pressure to normal. In this study, the anorectal resting pressure decreased postoperatively, and returned to normal level at postoperative sixth month. Although, the early postoperative defecation frequency and soiling incidence are higher (which may be related to the reduced anal resting pressure secondary to the absence of internal anal sphincter), the daily defecation frequency returned to normal and soiling incidence decreased to 1.6% 6 months later. This may be related to the improved contraction of external anal sphincter, the decreased peristalsis frequency of new rectum and the restoration of internal anal sphincter function replaced by thicken muscular layer of distal rectum. Significantly, no patient suffered from incontinence postoperatively. These results suggested that the function of internal anal sphincter could be replaced by partial external anal sphincters. Admittedly, the drawback of this study is that it is a retrospective study, lacking proper control group. Patients with mild enterocolitis were not included in our definition of HAEC. This makes the comparison with some of published articles difficult. Further prospective randomized study is required to confirm our preliminary finding.

4. Conclusion

Our results showed that TRM-PIAS is effective in treatment of HD. This procedure appears to have low postoperative HD-related enterocolitis.

References