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ORIGINAL ARTICLE

Treatment of classic-type Hirschsprung's disease: rectoplasty with posterior triangular colonic flap versus transanal endorectal pull-through with rectoanal myotomy

Megumi Nakamura[1] · Motoshi Wada[1] · Taichi Fukuzawa[1] · Hiromu Tanaka[1] · Hironori Kudo[1] · Ryo Ando[1] · Satoshi Yamaki[1] · Tomohiko Watanabe[1] · Keisuke Tada[1] · Yudai Nakajima[1] · Masaki Nio[1]

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Abstract

Purpose Our institution employs rectoplasty with a posterior triangular colonic flap (RPTCF) for classic-type Hirschsprung's disease. Recently, we employed a modified Soave procedure: transanal endorectal pull-through with rectoanal myotomy (TEPTRAM). In both procedures, the internal sphincter muscle is completely divided vertically at the 6 o'clock position. Unlike RPTCF, TEPTRAM does not require abdominal manipulation. We aimed to verify the usefulness of TEPTRAM.

Methods 64 patients with classic-type Hirschsprung's disease who underwent surgery between 1970 and 2017 were divided into group R (RPTCF, 47 cases) and group T (TEPTRAM, 17 cases). We compared the defecation function of the groups.

Results No patient showed fecal incontinence (R: 0/41, T: 0/10, ns). Three patients (6%) in group R and four (24%) in group T developed postoperative enterocolitis ($P=0.16$). Three patients (10%) in group R and one (20%) in group T needed an enema ($P=0.36$).

Conclusion There was no adverse effect of rectoanal myotomy; incontinence was not observed in either procedure. Although there was no significant difference, the incidences of enterocolitis and constipation were slightly higher in group T, perhaps because of the residual muscle cuff; therefore, it is necessary to provide best care with attention to constipation immediately after surgery.

Keywords Hirschsprung's disease · Rectoanal myotomy · Transanal endorectal pull-through · Posterior triangular colonic flap · Soave procedure · Duhamel procedure

Introduction

In our institution, we employ rectoplasty with a posterior triangular colonic flap (RPTCF) to treat classic-type Hirschsprung's disease. The concept of this procedure is to completely release anal achalasia and to preserve the sensation of defecation. The former improves postoperative constipation and the latter prevents fecal incontinence; both are considered essential for the surgical treatment of Hirschsprung's disease. Based on the concept of RPTCF, we recently employed an additional rectoanal myotomy with the transanal Soave procedure, namely transanal endorectal pull-through with rectoanal myotomy (TEPTRAM). Unlike RPTCF, TEPTRAM does not require abdominal manipulation, and is minimally invasive and highly esthetic. It seems to be a useful procedure for classic-type or shorter aganglionosis, which can be accomplished by transanal manipulation. We compared the postoperative course of TEPTRAM with that of RPTCF and aimed to verify the usefulness of TEPTRAM.

Methods

64 patients with classic-type Hirschsprung's disease who underwent surgery at Tohoku University Hospital between 1970 and 2017, and had a postoperative follow-up period of 6 months or more were included in this study. Patient records were retrospectively reviewed for clinical details.

✉ Masaki Nio
mnio@ped-surg.med.tohoku.ac.jp

[1] Department of Pediatric Surgery, Tohoku University Graduate School of Medicine, 1-1 Seiryo-machi, Aoba-ku, Sendai, Miyagi 980-8574, Japan

They were divided into group R (RPTCF, 47 patients) and group T (TEPTRAM, 17 patients).

The surgical techniques of the two procedures are as follows. RPTCF is a modification of the Duhamel procedure [1]. In this procedure, the anal canal is circumferentially preserved, except at the 6 o'clock position, where a vertical incision is made across the dentate line (Fig. 1a). In addition, the internal sphincter muscle, which was halfway incised as given in the original report [2], is currently totally divided vertically. The external anal sphincter that is exposed in the anal canal is covered by the anal end of the pulled-through intestine, which is sutured to the anoderm and forms a triangular flap (Fig. 1b, c). TEPTRAM is approximately the same as the transanal Soave procedure [3]. Moreover, myotomy of the internal sphincter is performed in the same manner as RPTCF. The internal sphincter muscle is vertically divided until the external sphincter muscle is exposed. After completing myotomy of the internal sphincter, the surgeon completely divides the muscular cuff of the rectum at the 6 o'clock position in the pelvic cavity.

The two- or three-step surgery was performed in 42 cases and 1 case in groups R and T, respectively, and the one-step surgery was performed in 5 cases and 16 cases in groups R and T, respectively. We compared the defecation function of the groups, such as fecal incontinence, postoperative enterocolitis, and constipation. Patients with complications, such as ileus, related to the abdominal operation, including stoma, were excluded from this study. The data obtained in this study were assessed by Wilcoxon rank sum test or chi-squared test for statistical comparison between groups R and T. All statistical analyses in this study were performed using JMP® Pro 14 (SAS Institute Japan Ltd.). Statistical significance was determined as less than 0.05 level of probability ($P < 0.05$). The present study was approved by the Clinical Research and Ethics Committee of Tohoku University Graduate School of Medicine in 2017 (Approval No. 2017-1-939).

Results

The patient demographic data are shown in Table 1. The age at the time of operation was from 118 to 5162 (median 486) and 19 to 576 (median 69) days in groups R and T, respectively ($P < 0.001$). The postoperative follow-up period was from 0.5 to 20.1 (median 5.9) years and from 0.7 to 14.8 (median 4.5) years in groups R and T, respectively ($P = 0.083$). No patient above 4 years of age showed fecal incontinence (R: 0/41, T: 0/10, ns). Three patients (3/47, 6%) in group R and four (4/17, 24%) in group T developed postoperative enterocolitis ($P = 0.052$) (Fig. 2). The patients developed postoperative enterocolitis at 14–225 (median 27) days after the surgery in group R and at 147–520 (median 337) days after the surgery in group T ($P = 0.053$). Enterocolitis improved with dietary restriction and bowel irrigation

Table 1 Characteristics of the groups

	Group R	Group T	*P* value
Number	47	17	
Gender (M:F)	38:9	14:3	0.892
Age at operation (days)	486 (118–5162)	69 (19–576)	< 0.001
POFP[a] (years)	5.9 (0.5–20.1)	4.5 (0.7–14.8)	0.083
Over all survival (%)	97.9	100	0.544

The mean age at operation in group T was younger than in group R

Continuous parameters are expressed as median and range

[a]Postoperative follow-up period

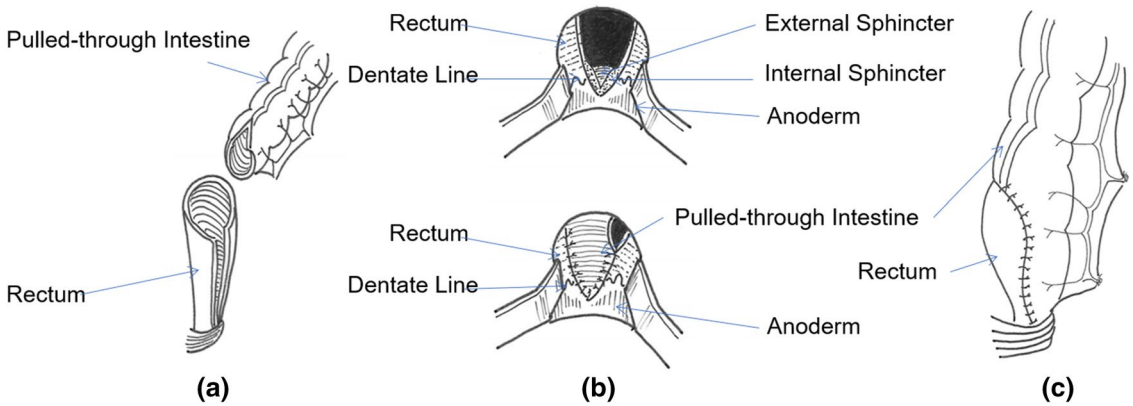

Fig. 1 Rectoplasty with a posterior triangular colonic flap (RPTCF) procedure. **a** The posterior wall of the rectum is split in the midline. The colon is split along its taenia libera. The anastomosis is performed between them. **b** The incision of the posterior wall of the rectum is extended through the dentate line. The triangular colonic flap is pulled down through the rectum and the colorectal anastomosis is performed. **c** Completion of colorectal anastomosis

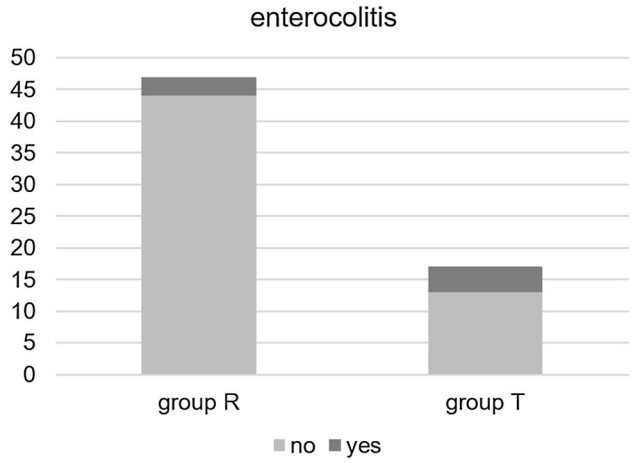

Fig. 2 The frequency of postoperative enterocolitis. There is no significant difference in the incidence of enterocolitis

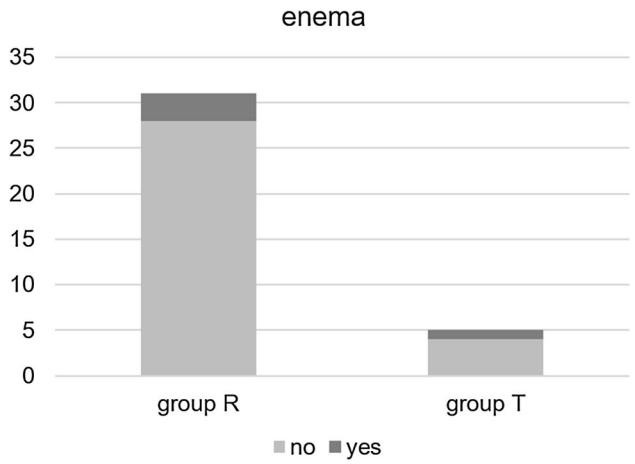

Fig. 3 The number of patients who needed an enema. There is no significant difference in the patients who needed an enema

in all cases. It took 16–44 (median 30) days in group R and 4–21 (median 8) days in group T for enterocolitis to improve ($P = 0.052$). In the patients above 7 years of age, three patients (3/31, 10%) in group R and one (1/5, 20%) in group T needed enemas or suppositories ($P = 0.496$) (Fig. 3). There were no complications related to urination or anal prolapse in either group. Independent of surgery, one patient died of measles at 2 years of age.

Discussion

The surgical outcome in terms of postoperative survival has been generally good; however, postoperative morbidities such as constipation and incontinence are still occasionally encountered. In such cases, morbidity is due to a residual aganglionic segment that causes anal sphincter achalasia or damage to the sensation of defecation. Kasai et al. reported that RPTCF can be used to solve these problems. These authors reported performing rectal myotomy with colectomy [4] based on the idea that rectal myotomy is useful in preventing damage to rectal sensation. Furthermore, they modified this surgery and reported an RPTCF procedure [2] that could completely release anal achalasia and protect better the sensation of defecation. We recently employed an additional rectoanal myotomy with the transanal Soave procedure, transanal endorectal pull-through with rectoanal myotomy (TEPTRAM) for classic-type Hirschsprung's disease.

In this study, we compared the postoperative course of TEPTRAM with RPTCF and aimed to verify the usefulness of TEPTRAM.

The median age at operation was 16 months and 2 months in groups R and T, respectively, ($P < 0.001$). The age at operation in group T was significantly lower than in group R. Because a surgical stapler is not used in TEPTRAM unlike in RPTCF, the size of the anal canal does not matter, and earlier surgery might be possible. As an exception, three patients received surgery at above 6 months of age because of delayed diagnosis.

The postoperative follow-up period was slightly shorter in group T because the patients in group T were recent cases, though there was no significant difference.

Fecal incontinence significantly reduces quality of life. Stensrud et al. [5] reported that eight patients (29%) who underwent transanal endorectal pull-through (TEPT) procedure had constant soiling, leading to social problem, and that the soiling tended to be more severe after TEPT than with laparotomy-assisted endorectal pull-through. Neuvonen et al. [6] reported that fecal soiling was generally occasional and 14 patients (12%) over 3 years of age had frequent soiling. In general, most children will complete toilet training by 4 years of age. In our study, no patient above 4 years of age demonstrated fecal incontinence or severe soiling with either procedure. There are two important elements to preventing fecal incontinence. The first is to maintain the anal sensation. The mucosa of the transitional zone 2.5–7.5 mm above the dentate line has abundant sensory receptors. To maintain anal sensation, this area should not be damaged. Because we perform the mucosectomy from 5 mm above the upper margin of the surgical anal canal, the so-called Herrmann line (Fig. 4), the mucosa of the transitional zone is preserved and the anal sensation is maintained in TEPTRAM. The other element is to not damage the external anal sphincter. It is necessary to distinguish the internal anal sphincter and the external anal sphincter during rectoanal myotomy to release anal achalasia. We always check muscle contractions repeatedly during rectoanal myotomy using a nerve stimulator to safeguard the external sphincter.

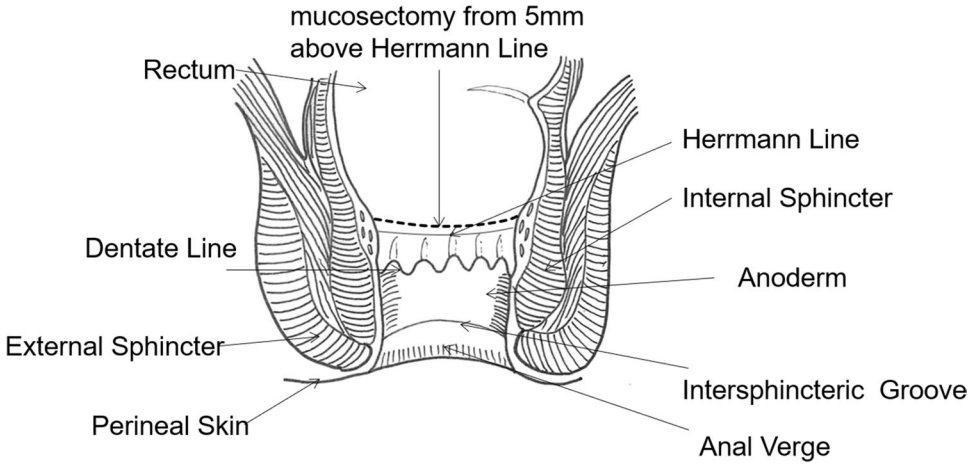

Fig. 4 The line to start mucosectomy. We perform mucosectomy from 5 mm above the Herrmann line. The dashed line indicates the start position

Regarding constipation, we actively encourage defecation with enemas or suppositories until bowel habits are established. Most patients can quit enemas or suppositories before entry into primary schools. In this study, three patients in group R and one patient in group T needed enemas or suppositories for constipation though they were more than 7 years of age. There are some reports that 0.9–18% of the patients who underwent the transanal Soave procedure had constipation, requiring enemas [6–8]. Our results may be comparable or somewhat worse than the results of a few other recent reports because we do not hesitate to use enemas or suppositories.

If constipation is not well controlled, congestive enterocolitis will occur. In this study, three patients in group R and four in group T developed postoperative enterocolitis. Although there is no significant difference between the two groups, the incidences of constipation and enterocolitis were slightly higher in group T. To prevent postoperative constipation and enterocolitis, it is important that there is no residual aganglionic segment and that anal achalasia is released. In our institution, rectoanal myotomy is performed to release anal achalasia. In some institutions, the muscle cuff is resected into a strip at the posterior wall [3, 5]. We do not cut off the muscular cuff, but only incise in the 6 o'clock direction. It is important that the internal anal sphincter is incised completely across the dentate line to the intersphincteric groove in addition to incision of the muscular cuff of the rectum (Fig. 5). This procedure will release anal achalasia and result in the prevention of constipation and congestive enterocolitis. However, patients who undergo TEPTRAM require more careful postoperative management than those who undergo RPTCF. In this procedure, the muscular cuff remains; thus, postoperative contraction always occurs because of scarring around the muscular cuff. The surgeon performs bougie using his or her index finger, starting at 2 weeks postoperatively. It is important to sufficiently dilate the full range of the muscular cuff. There was no statistically significant difference, but postoperative enterocolitis occurred later in group T. This may be due to non-adherence over time to dilation, although it was strictly managed at the early stage. The patient's family needs to continue performing the bougie technique daily for at least 1 year after TEPTRAM. In cases where there was careful management, no patient

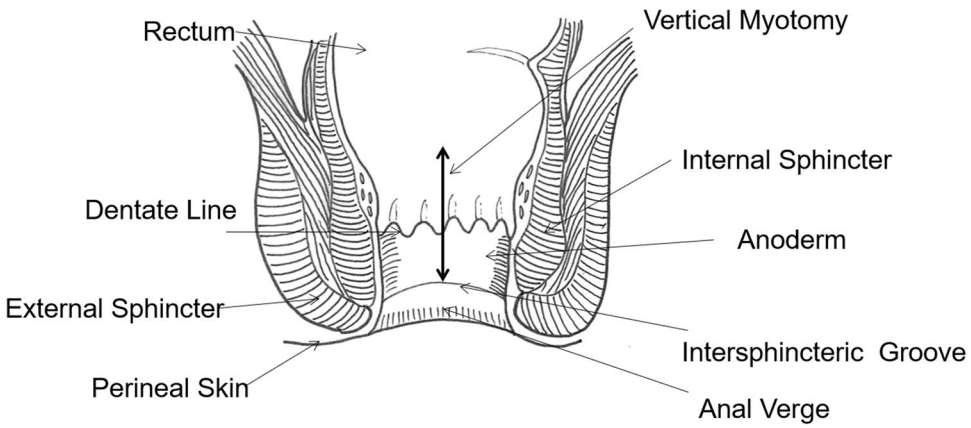

Fig. 5 Rectoanal myotomy in the transanal endorectal pull-through with rectoanal myotomy (TEPTRAM) procedure. The internal sphincter muscle is completely divided vertically at the 6 o'clock position (bidirectional arrows)

developed enterocolitis in either group one and a half years postoperatively.

Conclusion

TEPTRAM is a modified transanal Soave procedure, and rectoanal myotomy is added following the concept of RPTCF. The advantage of this procedure is that abdominal surgery is not required; however, more careful postoperative management is required due to a residual muscular cuff. With correct surgical techniques and best postoperative care, the functional outcome of TEPTRAM is expected to be similar to that of RPTCF.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

Ethical approval This study was approved by the ethics committee of Tohoku University Graduate School of Medicine and has, therefore, been performed in accordance with the ethical standards laid down in the Declaration of Helsinki, 1964, and its later amendments.

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