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ORIGINAL ARTICLE

Hirschsprung's disease in the laparoscopic transanal pull-through era: implications of age at surgery and technical aspects

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Abstract

Aim Detailed implications of age at laparoscopic transanal pull-through (LTAPT) on postoperative bowel function (POBF) in Hirschsprung's disease (HD) are somewhat obscure because of a spectrum of factors.

Methods Age at surgery was used to categorize 106 consecutive postoperative HD cases treated by our modified LTAPT (JLTPAT) between 1997 and 2015; group A: < 3 months old ($n=31$); group B: 3–11 months old ($n=44$); group C: 1–3 years old ($n=19$); and group D: ≥ 4 years old ($n=12$). POBF was assessed by reviewing outpatient records 1, 3, 5, 7, and 10 years after JLTAPT prospectively and scoring each of 5 criteria on a scale of 0–2; best score = 10.

Results Only operative time was statistically longer in group D versus groups A, B, and C. Differences in gender ratios, blood loss, duration of follow-up, and POBF scores were not statistically significant. Mean POBF scores over time were: group A: 6.8, 7.6, 8.4, 8.6, and 8.4; group B: 7.1, 7.8, 8.3, 8.5, and 9.0; group C: 6.9, 7.9, 8.1, 8.3, and 8.6; group D: 7.0, 7.4, 8.2, 8.1, and 8.5, respectively.

Conclusion Age at JLTAPT was not correlated with POBF in HD.

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Keywords Hirschsprung's disease · Laparoscopy · Transanal endorectal pull-through · Age at surgery · Postoperative bowel function

Introduction

Hirschsprung's disease (HD) is a relatively common cause of intestinal obstruction in the newborn, caused by an absence of ganglionic cells in the distal bowel. The main goal of surgical intervention in HD is to resect the aganglionic segment and pull down normoganglionic bowel to align it with the anus. There are three basic approaches to treatment: single-stage pull-through during the neonatal period; multistage pull-through characterized by a colostomy created during the neonatal period, followed by delayed coloanal reconstruction later in infancy; and cleansing enemas or colorectal irrigation during the neonatal period followed by pull-through later in infancy. The current trend is for early surgery soon after diagnosis [1], but there is a spectrum of POBF results that cannot be explained consistently [2–4], although the general consensus from the previous studies [5, 6] is that POBF improves with age. Whether or not the age at surgery has an influence on POBF, especially in laparoscopy-assisted transanal pull-though (LTAPT) era, remains unknown.

Thus, we conducted a prospective evaluation of the LTAPT procedure performed for HD at Juntendo (JLTAPT) by categorizing cases according to age at the time of surgery to enable a comparison of POBF as assessed by a standardized questionnaire which we developed and administered over an extended 10-year period. We also present the technical aspects of JLTAPT.

Methods

The subjects for this study were biopsy proven rectal, rectosigmoid, and sigmoid-type HD patients treated by JLTAPT between 1997 and 2015 with POBF followed-up for at least 12 months ($n = 106$). Patients with total colonic aganglionosis, an aganglionic segment proximal to the descending colon, chromosomal abnormalities, and incomplete POBF follow-up were excluded to minimize etiologic variables. Patient demographics, surgical outcome, and complications were compared.

Age at JLTAPT was used to categorize our subjects into four groups; group A: < 3 months old ($n = 31$); group B: 3–11 months old ($n = 44$); group C: 1–3 years old ($n = 19$); and group D: ≥ 4 years old ($n = 12$). POBF involved prospective review of questionnaire responses and care giver interview records about five standard criteria (frequency of motions, presence of soiling or staining, perianal erosions, anal appearance, and requirement for mediations to assist bowel function) obtained from outpatient records at 1, 3, 5, 7, and 10 years after JLTAPT, each scored on a scale of 0 to 2 to give a maximum score of 10 (Table 1).

Our JLTAPT procedure is described in full elsewhere [7]. Briefly, colorectal dissection and transanal rectal dissection were modified (Fig. 1) and distinguish JLTAPT from other LTAPT performed elsewhere. Specifically, transanal dissection is commenced just above the anorectal line (ARL), i.e., at the squamo-columnar epithelial junction [8, 9], leaving the ARL intact. If there is any residual aganglionic rectal muscular cuff remaining, its posterior wall should be divided

Table 1 Postoperative bowel function (POBF) evaluation scores (Worst = 0; Best = 10)

	0	*1*	*2*
Frequency of motions	≥ 6/day	3–5/day	1–2/day
Staining/soiling	Soiling	Staining	None
Perianal erosions	Often	Occasionally	Nil
Anal appearance	Prolapse requiring repair	Visible mucosa	Normal
Medications required	Antidiarrheals/ enemas	Probiotics/ laxatives/ enemas	Nil

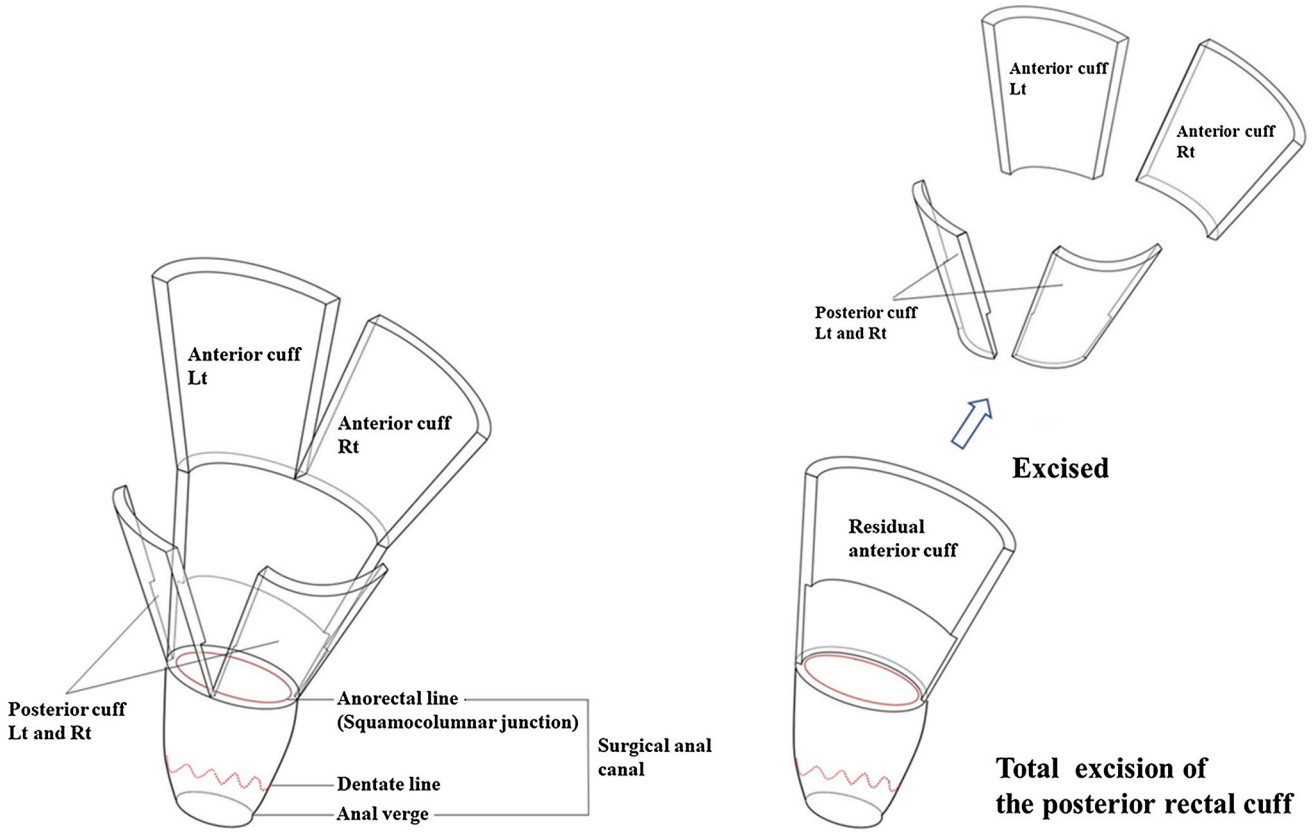

Fig. 1 Posterior aganglionic rectal muscular cuff has been transected in toto just above the anorectal line, leaving the anorectal line, dentate line, and surgical anal canal intact. Note that the first 10–15 mm of the proximal anterior and posterior cuffs are thinner, since the first 10–15 mm of transanal dissection of the rectum starting just above the intact anorectal line is near full-thickness and then switched to mucosectomy after 10–15 mm

transanally at the 6 o'clock position taking great care not to injure the pelvic floor muscles. Then, the entire posterior cuff should be excised. The end point of division/excision of the posterior rectal cuff is also just above the ARL. The anterior rectal cuff is also excised transanally to the point where laparoscopic rectal dissection was ceased.

All management was standardized according to our preoperative, operative, and postoperative protocols. Preoperatively, all subjects were fed normally, and those subjects who did not have a colostomy had colon decompression and saline irrigations using a rectal tube. In patients older than 12 months or patients with hugely dilated, elongated colons, a central venous catheter was inserted routinely and patients fasted for 1 week prior to surgery if they did not have ileostomies or colostomies.

Analysis of variance with the Bonferroni post-test correction was used for multiple comparisons of sample means. A p value of less than 0.05 was considered to be statistically significant.

This study was approved by the Juntendo University School of Medicine Ethics Committee.

Results

Gender ratios were (group A: 20M/11F; group B: 32M/12F; group C: 10M/9F; and group D: 6M/6F). Initially, the dentate line (DL) was used as the landmark for starting transanal rectal dissection, but now, we use the ARL. The overall DL:ARL ratio was (41:65) which was similar for each group, as well (p = ns).

Mean weights at surgery were (group A: 3.8 kg; group B: 7.6 kg; group C: 10.9 kg; and group D: 22.6 kg). Mean operative times were (group A: 157.2 min; group B: 163.6 min; group C: 167.3 min; and group D: 244.2 min). Mean operative time was significantly longer in group D ($p < 0.05$). Differences in blood loss were not statistically significant (group A: 6.7 mL; group B: 7.5 mL; group C: 7.9 mL; and group D: 8.3 mL; p = ns).

There were no intraoperative complications. Postoperative complications included one bowel obstruction in group A caused by residual rectal cuff that had become folded during pull-through that was repaired through a posterior sagittal approach; three anastomotic strictures, one each in groups A, B, and C, each of which resolved within 1 month after daily dilatation (p = ns); one perineal abscess in group D that resolved after daily irrigations without requiring a stoma (p = ns); and five episodes of Grade-1 Hirschsprung-associated enterocolitis (group A: $n = 2$; group B: $n = 2$; group C: $n = 1$; group D: $n = 0$) (p = ns) [10].

The number of subjects who had POBF follow-up for 1, 3, 5, 7, and 10 years in each group was: group A: 2, 3, 3, 5, and 18; group B: 3, 3, 10, 10, and 18; group C: 1, 1, 2, 6, and 9; group D: 1, 2, 2, 2, and 5, respectively (Fig. 2). Mean POBF scores after each duration of follow-up were: group A: 6.8, 7.6, 8.4, 8.6, and 8.4; group B: 7.1, 7.8, 8.3, 8.5, and 9.0; group C: 6.9, 7.9, 8.1, 8.3, and 8.6; and group D: 7.0, 7.4, 8.2, 8.1, and 8.5, respectively. Differences were not statistically significant (p = ns).

Discussion

Our POBF assessment is a standardized easily administrated questionnaire that accurately records POBF at a point in time and can be filed in patients' medical records for future reference. We previously reported that POBF can also be assessed in smaller children less than 4 years old, or even in infants using our POBF scoring system [11], because we are not assessing social fecal continence, but assessing signs of poor POBF that can be assessed at any age. By conducting regular POBF assessment over time, we have a continuous record of POBF status that in itself is valuable, because it illustrates how POBF changes. POBF data used in this study were obtained prospectively by reviewing outpatient medical records, which we believe ensures the reliability of our POBF data. There is a report that found fecal control following TAPT for HD to be initially inferior to controls during childhood, but that any problems were largely expected to resolve by adulthood [12] which is consistent with a previous report of our experience [13] and this series. The majority of their patients and 95% of adult patients were actually socially continent, with only a small minority of cognitively normal TAPT cases likely to require later secondary intervention.

We believe that the good POBF achieved in our HD patients is because during JLTAPT, transanal dissection is commenced just above the ARL irrespective of the age of the

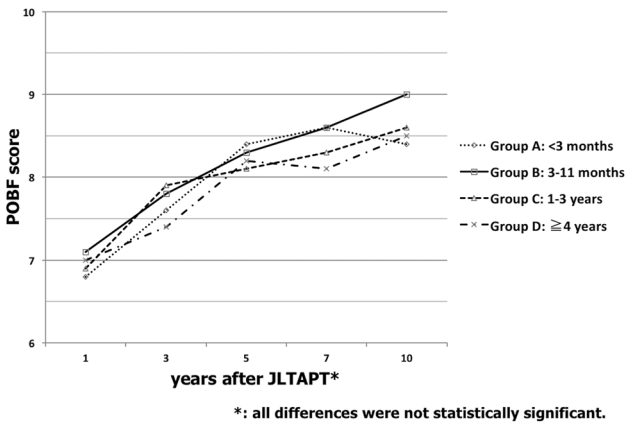

Fig. 2 Postoperative bowel function (POBF) after Juntendo transanal endorectal pull-through (JLTAPT). POBF scores between groups A–D were not statistically significant

patient, leaving the ARL, DL, and surgical anal canal intact (Fig. 1), while ensuring complete excision of the aganglionic rectum. In addition, we believe the reason why there were no age group differences in POBF in our series was because transanal rectal dissection started at exactly the same level in all cases. In other words, the starting point, the ARL, was completely reproducible in every case irrespective of age. This is not the case elsewhere, unfortunately where a certain distance proximal to the DL is the norm for determining the landmark for where transanal rectal dissection should start, e.g., 5, 10, or 15 mm about the DL based loosely on body size and experience. Obviously 10 mm in an infant is different to 10 mm in an older child which could be a reasonable cause for differences in POBF observed between age groups at other centers. If transanal rectal dissection is commenced using the DL as a landmark, dissection could start anywhere from 5 to 15 mm above the ARL resulting in a risk for postoperative constipation because of residual achalasia, especially if the cuff is also not divided (as we advocate), and if lower, i.e., between the ARL and the DL, postoperative incontinence will be universal. We also perform biopsies at four points circumferentially (at 3, 6, 9, and 12 o'clock) around the end of the pulled-through colon routinely to double-check the innervation of the pulled-through colon [14]. Biopsy specimens are examined immediately before the pulled-through colon anastomosis is commenced, to prevent bowel with patchy innervation [14, 15] from being anastomosed, as such bowel may be the cause of poor POBF.

A survey of members of the European Pediatric Surgeons' Association [16] found that 2/3 of respondents would delay pull-through surgery in neonates and that 27% would create a stoma. By implication, single-stage repair would thus be performed by 71% of respondents, a finding similar to the results of an American Pediatric Surgical Association survey [17]. The majority of members of the European Association who responded they would delay pull-through surgery recommended waiting until the infant was either 3 months old or weighed more than 5 kg which would vary according to nutrition. The final consensus was that surgery for should be performed between 1 and 6 months of age. This is consistent with the timing of pull-through surgery reported after other surveys [18, 19]. However, there are no established guidelines for the timing or ideal age for surgery for HD reported in the literature.

Neonates who have undergone TAPT are at risk for adverse short-term outcomes. Huang et al. [20] reported that neonates took longer to recover after surgery and bowel motions were more frequent 3 months after surgery. They also reported a higher incidence of anastomotic stricture and anastomotic leakage in neonatal cases, which is consistent with the findings of another report [3] and was explained as being a consequence of having lower immunity or poorer tolerance to infection or surgical stress [2]. Technically, pull-through surgery in neonates can be problematic if there is insufficient sigmoid colon to ensure adequate resection of aganglionic bowel, resulting in an anastomosis under tension which can cause stricture or leakage [21]. In this series, we found no differences in the incidence of complications, including stricture and infection related to age at TAPT, even in young infants including neonates. Laparoscopic colorectal dissection during LTAPT probably improves the maneuverability of dissected bowel compared with TAPT without laparoscopic assistance, because laparoscopic vessel dissection can protect marginal arteries in the colon under direct (laparoscopic) view, ensuring that the distal end of the pulled-through colon has an adequate blood supply resulting in a good coloanal anastomosis. Without laparoscopic assistance, the likelihood of injuring marginal vessels in the pulled-through colon is high, since colonic vessels are divided along the wall of the colon and the blood supply to the distal end of the pulled-through colon is almost only from intramural vessels in the colon wall resulting in a potentially poorly perfused coloanal anastomosis.

A higher incidence of postoperative enterocolitis has also been reported in neonatal cases because of increased risk for sphincter spasm and anastomotic stricture that can contribute to intestinal obstruction [22, 23]. Neonates also have a tendency to be constipated [21], but there are a myriad of causes for constipation not specific to the neonatal age group that may include long muscular sheath or Soave cuff and erroneous interpretation of intraoperative biopsies [24, 25]. To help alleviate postoperative enterocolitis, we believe excising the entire posterior rectal muscle cuff (Fig. 1) is effective and the low incidence of enterocolitis in our series reflects this. In addition, there were no differences in the incidence of enterocolitis in our series indicating that our surgical technique is probably effective for preventing enterocolitis. Similarly, we believe that the low incidence of postoperative constipation in our series, even in neonates and young infants, is also because of the total excision of the posterior rectal cuff caudally, almost down to the ARL, that we perform routinely in JLTAPT. There are many reports about short residual cuff remnants not being associated with POBF sequelae in TAPT or LTAPT cases, with some even claiming good POBF in cases with cuff remnants [26], but we do not recommend leaving any aganglionic cuff, let alone a short one, because there may be a subgroup of patients in whom the balance of peristalsis in the pulled-through colon cannot overcome the achalasia in the cuff remnant and cause some degree of residual constipation that would otherwise not be an issue if there was no cuff. We believe that the good POBF results achieved in our JLTAPT cases are due entirely to total excision of the posterior rectal cuff. In fact, the increasing number of reports of poor POBF after TAPT with or without laparoscopy compared with the conventional

transabdominal pull-through [27] are most likely to be in cases with cuff remnants.

In older patients with HD, compared with younger patients, such as neonates or infants, the chronic accumulation of stools causes inflammation in the colon wall, causing greatly thickened muscle layers, ulceration of the mucosa, and repeated episodes of subclinical enterocolitis can cause fibrous adhesions to develop between the mucosa and the submucosa. The mesenteric vessels in this age group are also larger and may bleed in spite of electrocoagulation [28, 29]. The difficulty of displaying a very dilated colon, anastomotic incongruence that may require resection of dilated proximal colon or several plication sutures to allow near normal caliber colon to be anastomosed are problems typically seen in older patients [3, 30]. There is a report that operative time in older children was as much as two times longer than in newborns and infants, and that the most serious early operative complication was anastomotic leakage [31, 32]. In fact, while operative time in group D was significantly longer than in other groups in this study, consistent with other reports [33] in the literature, POBF in older children did not differ from other groups in this study probably because of our routine use of central venous catheters preoperatively or creation of a protective ileostomy.

There were limitations in this study. First, the LTAPT procedure performed was modified during the study period. Second, the number of patients who had POBF assessment in all groups fell from 1 to 10 years. Third, some group B patients were actually delayed group A patients or patients who were diagnosed later.

To summarize, by analyzing our data categorized according to age at LTAPT, there would appear to be no influence of age at LTAPT on POBF in children with HD and steady improvement in POBF over time, findings that support Bjornland et al.'s very recent report that includes both LTAPT and TAPT without laparoscopy [34]. Our JLTAPT technique may also provide fellow surgeons with a more reliable landmark for commencing dissection that may improve the reliability of surgery, thus assisting in improving outcome and the accuracy of prognosis.

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