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ORIGINAL ARTICLE

The mid-term outcomes of TRM–PIAS, proctocolectomy and ileoanal anastomosis for total colonic aganglionosis

Qi Li[1] · Long Li[1] · Qian Jiang[2] · Zhen Zhang[1] · Ping Xiao[3]

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Abstract
Aims The present study aimed to evaluate the mid-term outcomes of total colonic aganglionosis (TCA) after transanal rectal mucosectomy and partial internal anal sphincterectomy (TRM–PIAS), proctocolectomy and ileoanal anastomosis.
Patients and methods From 2012 to 2014, 12 patients (7 boys; 58.3 %) diagnosed with TCA and treated with the TRM–PIAS, proctocolectomy and ileoanal anastomosis. Seven TCA patients who underwent laparotomy-assisted endorectal pull-through (LEPT) between 2010 and 2012 were used as control group. Demographic features and complication of the two groups were evaluated. The functional outcomes were assessed by using a score system.
Results The procedure was successfully performed in all patients. The incidence of postoperative HAEC in the TRM–PIAS group was significantly lower (25.0 vs 85.7 %; $p < 0.05$) than control group within the second postoperative year. The number of bowel movement after 3, 12 and 24 months postoperatively, was 8.5 ± 3.5, 5.3 ± 2.9 and 3.1 ± 1.4 ($p < 0.05$) per day, respectively, in the TRM–PIAS group. The soiling was noted in 50.0 % ($n = 6$) of the patients in the 6th postoperative month, and 25.0 % ($n = 3$) in the 24th postoperative month in the TRM–PIAS group. There was no significant difference in overall functional outcome between two groups, but the TRM–PIAS group was better in terms of bowel movement and soiling.
Conclusion TRM–PIAS, proctocolectomy and ileoanal anastomosis might be an effective treatment for TCA. More prospective studies evaluating the TRM–PIAS technique over longer period and with greater sample size are needed to confirm the findings in this study.

Keywords Total colonic aganglionosis · Partial internal anal sphincterectomy

Introduction

Total colonic aganglionosis (TCA) represents a major challenge for pediatric surgeons. Comparing to short segment Hirschsprung's disease (HD), TCA shows the relatively higher morbidity and mortality rates and the poorer postoperative functional outcomes [1]. After surgery, it is quite common that patients may suffer from several postoperative complications, including recurrent Hirschsprung-associated enterocolitis (HAEC), frequent bowel movement, severe perineal excoriation and so. Many techniques and theirs modifications have been described for treating TCA, but none had been proven superior to others [2]. Transanal rectal mucosectomy and partial internal anal sphincterectomy (TRM–PIAS) with fewer complications and good functional outcomes have been introduced for treating other types of HD by Zhang [3]. Thus, TRM–PIAS, proctocolectomy and ileoanal anastomosis may be considered as a strategy of treating TCA.

The present study aims to evaluate the postoperative complications and mid-term clinical outcomes of the TCA patients undergone TRM–PIAS, proctocolectomy and ileoanal anastomosis.

✉ Qi Li
l817@sina.com

[1] Department of General Surgery, Capital Institute of Pediatrics, No. 2 Yabao Rd, 100020 Beijing, China

[2] Department of Medical Genetics, Capital Institute of Pediatrics, No. 2 Yabao Rd, 100020 Beijing, China

[3] Department of Pathology, Capital Institute of Pediatrics, No. 2 Yabao Rd, 100020 Beijing, China

Materials and methods

This retrospective controlled clinical trial was approved by the board of the Capital Institute of Pediatrics (China). Ethics approval from the Ethics Committee of Capital Institute of Pediatrics was obtained. Written consents were obtained from the parents prior to surgery.

In this study, the definition of total colonic aganglionosis was total colonic aganglionosis with or without distal ileum (<50 cm) involvement.

Twelve patients (7 boys; 58.3 %) diagnosed with TCA and treated with the TRM–PIAS, proctocolectomy and ileoanal anastomosis at Department of Pediatric Surgery, Capital Institute of Pediatrics, China, between 2012 and 2014 were reviewed. Because of the intestinal obstruction, abdominal distention or intestinal perforation, 11 of them underwent initial diverting ileostomy at the age between 2 day and 13 months. One of them underwent primary pull-through without stoma. The definitive pull-through procedure was performed at the mean age of 8.0 ± 3.9 months (range 3–16 months). Between 2010 and 2012, seven boys who were treated for TCA with laparotomy-assisted endorectal pull-through (LEPT) procedure were used as the control group. All of them underwent initial diverting ileostomy at the age between 15 and 40 days. The definitive pull-through procedure was performed at the mean age of 10.4 ± 5.1 months (range 5–19 months). All the surgeries were performed by the same group of clinicians.

Surgery

Surgical technique for TRM–PIAS group (Fig. 1)

1. The diagnosis of TCA was made by ileocolic multipoint seromuscular layer frozen-section biopsy at laparotomy.
2. Transabdominal colectomy and resection of the aganglionic small intestine were performed.
3. After placing a anal retractor, the rectal mucosa was circumferentially incised using cautery at the junction between anal and rectal mucosa (anorectal line).
4. The anterior rectal wall was dissected along the submucosal layer and the anterior rectal muscular sleeve remained intact. 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.
5. After delivered all aganglionic bowel, the normal innervated ileum was pulled through and anastomosed to anal mucosa with 5–0 absorbable suture.
6. Ileostomy was closed at the same time of pull-through.

Surgical technique for control group

After confirmed diagnosis by ileocolic multipoint seromuscular layer frozen-section biopsy, the patients of control group were treated with LEPT procedure. This procedure consists of transabdominal colectomy and resection of the aganglionic small intestine, transanal rectal mucosectomy leaving a split muscle cuff and ileoanal anastomosis.

Postoperative management

The patients could feed once bowel motion recovered. If the enteral nutrition was intolerant after the third postoperative day, they would get parenteral nutrition (PN) to maintain the nutritional input. Parenteral broad-spectrum antibiotic was administrated for 3–7 days. Local clean and care were required once needed. On the postoperative day 14, anal dilatation was initiated. Parents were instructed regarding perianal skin care and the symptoms of HAEC before discharge.

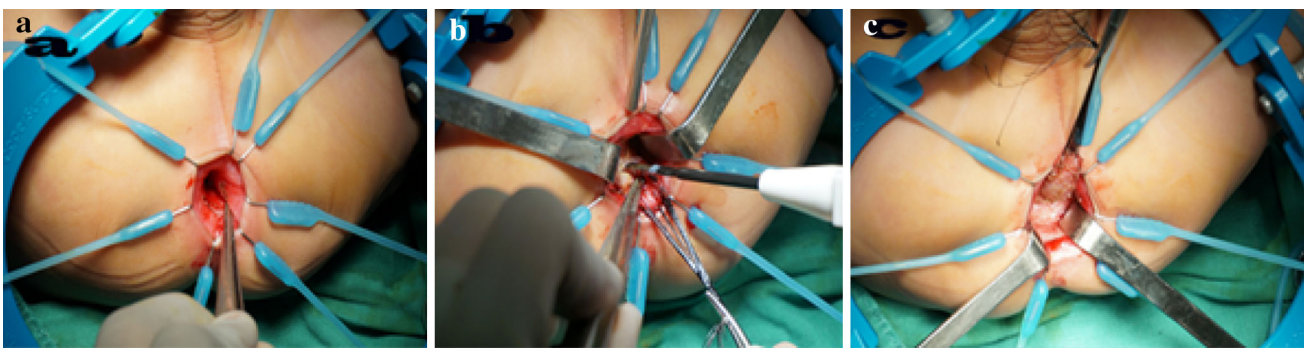

Fig. 1 **a** The rectal mucosa was circumferentially incised at the junction between anal and rectal mucosa. **b** The anterior rectal wall was dissected along the submucosal layer. **c** The posterior dissection was performed along the cleavage between internal and external anal sphincters

Outcome measures

The data were obtained by review of medical records (inpatient, outpatient, and follow-up investigations). Also, interviews were conducted via telephone and internet by one of the authors for both groups. According to the previously study, the objective bowel function was assessed by using a published score system [5]. 11–16 points were considered as good, 6–10 was fair and 0–5 was poor, respectively. The latest records of weight and height had been assessed according to the 2006 WHO Child Growth Standards. Underweight was defined as weight for age <-2 standard deviations (SD). Stunting was defined as height for age <-2 SD.

In this study, the diagnostic criteria of constipation are the voluntary bowel movement two or less times per week for at least 2 weeks. Soiling is defined as involuntary leaking of small amounts of stool or liquid, requiring changing underwear or diapers. The diagnostic criteria of soiling are suffering from soiling once or more per week for at least 1 month. Enterocolitis is defined as the occurrence of clinical symptoms including abdominal distention, explosive diarrhea, fever (>38 °C) and lethargy with or without vomiting, the passage of blood stained stool, increased leukocytes in the blood or stool and positive stool culture [3, 4].

Statistical analysis

The data were analyzed by SPSS software version 19.0. For between-group comparisons of non-normally distributed variables, Mann–Whitney *U* test was used. The Fischer exact test was used to compare frequencies between groups and Kruskal–Wallis ANOVA was used in multiple comparisons. In all analyses, a significance level was set at 0.05.

Results

Descriptive data and early postoperative complications

In the TRM–PIAS group, mean operative time was 220.8 ± 35.0 min (range 160–280 min). Blood loss was 15–40 ml. Involved distal ileum was 22 ± 7.0 cm (range 10–35 cm). One of the 12 patients that had abdominal infection required anti-infective therapy and abdominal drainage, whereas the others recovered uneventfully without anastomotic leak, cuff abscess, dehiscence, retraction, incorrect leveling of pull-through or bowel obstruction. All the patients were discharged 11.6 ± 3.7 days (range 7–21 days) after pull-through without PN dependent.

In the LEPT group, mean operative time was 207.1 ± 18.7 min (range 185–240 min). Blood loss was 25–45 ml. Involved distal ileum was 21.1 ± 9.9 cm (range 5–31 cm). One of them suffered from postoperative wound dehiscence. They were discharged 14.1 ± 4.8 days after surgery (range 7–23 days). The demographic features of the two groups were illustrated in Table 1. There was no statistically significant difference between the two groups.

Late postoperative complications (Table 2)

In the TRM–PIAS group, the incidence of postoperative HAEC was decreased from 66.7 % ($n = 8$) within the first postoperative year to 25 % ($n = 3$) ($p = 0.10$) within the

Table 1 Demographic data of two groups

	TRM–PIAS group	LEPT group	*p* value
Age of definitive pull-through (months)	3–16 (8.0 ± 3.9)	5–19 (10.4 ± 5.1)	0.372
Operative time (min)	160–280 (220.8 ± 35.0)	185–240 (207.1 ± 18.7)	0.329
Involved distal ileum (cm)	10–35 (22 ± 7.0)	5–31 (21.1 ± 9.9)	0.832
Early postoperative complication	8.33 % ($n = 1$) abdominal infection	14.3 % ($n = 1$) wound dehiscence	1
Postoperative stay (days)	7–21 (11.6 ± 3.7)	7–23 (14.1 ± 4.8)	0.126

Table 2 Late postoperative complication

	TRM–PIAS group	LEPT group	*p* value
Enterocolitis within the 1st postoperative year	66.7 % ($n = 8$)	100 % ($n = 7$)	0.25
Enterocolitis within the 2nd postoperative year	25 % ($n = 3$)	85.7 % ($n = 6$)	0.02
Osteomyelitis	8.3 % ($n = 1$)	0	1
Recurrent perianal excoriation	0	28.5 % ($n = 2$)	0.12
Abnormal growth	16.7 % ($n = 2$)	28.5 % ($n = 2$)	0.60
PN dependent	0	0	N/A

Table 3 Functional outcomes

	TRM–PIAS group	LEPT group	*p* value
Recurrent abdominal distension	1.80 ± 0.42	1.86 ± 0.378	0.77
Frequency of defecation	1.20 ± 0.63	0.57 ± 0.53	0.05
Stool consistency	1.80 ± 0.42	1.57 ± 0.53	0.32
Soiling	1.70 ± 0.48	0.71 ± 0.95	0.028
Urgency period	1.50 ± 0.71	1.43 ± 0.79	0.87
Diapers required	1.40 ± 0.70	1.29 ± 0.95	0.91
Long-term use of medication	1.80 ± 0.42	1.71 ± 0.49	0.69
Diet	2.00 ± 0	2.00 ± 0	1
Total score	13.20 ± 2.49	11.14 ± 2.79	0.09

second postoperative year, which was mild and occasional except one suffering from the recurrent HAEC for half year in the first postoperative year. The incidence of postoperative HAEC in the TRM–PIAS group was not significantly different from the LEPT group (66.7 vs 100 %; $p = 0.25$) within the first postoperative year, and had become significantly lower (25.0 vs 85.7 %; $p < 0.05$) within the second year. They were treated with rectal decompression, enema and broad-spectrum antibiotics. The perianal excoriation was cured successfully with local care within 2.2 ± 2.1 months in all the TRM–PIAS group patients. In the LEPT group, two of the patients (28.6 %, $p = 0.12$) complained the recurrent perianal excoriation. One of the patients suffered from osteomyelitis for 4 months in the TRM–PIAS group. In the TRM–PIAS group, one of patients was both stunting and underweight, another of them was underweight. In the LEPT group, one of them was both stunting and underweight, another of them was stunting. All the other patients grew normal. None reported anastomotic stricture formation, constipation, rectal prolapse, adhesive ileus, PN dependence, redo, rediversion or death in both groups.

Outcomes of function

The median follow-up period was 33 months (30–43 m) in the TRM–PIAS group and 52 months (68–43 m) in the LEPT group.

The frequency of the bowel movement (100 %), liquid or loose stool (100 %) were observed in every patient in the TRM–PIAS group within the first 3 months after pull-through. The number of bowel movement after 3, 12 and 24 months postoperatively, were 8.5 ± 3.5, 5.3 ± 2.9 and 3.1 ± 1.4 ($p < 0.05$) per day, respectively, in the TRM–PIAS group. At the last follow-up, pasty stool consistency was noted in 83.3 % ($n = 10$) of the TRM–PIAS group patients. The soiling was noted in 50.0 % ($n = 6$) of the patients in the 6th postoperative month, and 25.0 % ($n = 3$) occasionally in the 24th postoperative month in the TRM–PIAS group.

Bowel function was assessed by the score system for the patients who were older than 3 years at the last follow-up in the TRM–PIAS group ($n = 10$). For comparison, the follow-up records after 36 months postoperatively were reviewed in the LEPT group ($n = 7$) (Table 3).

There was significant difference in the soiling item between two groups. The TRM–PIAS group had higher score in frequency of defecation but did not reach statistically significant level (1.20 ± 0.63 vs 0.57 ± 0.53; $p = 0.05$). There was no significant difference between two groups in other outcomes and total score. In the TRM–PIAS group, eight patients (80.0 %) showed good overall functional outcomes (score of 11–16) and two patients (20.0 %) were fair (score of 6–10). Five patients (71.4 %) showed good outcomes and two patients (28.6 %) were fair in the LEPT group.

Discussion

Total colonic aganglionosis involving the total colon with or without the small intestine is a rare and severe form of HD. The incidence is between 3 and 12 % among all forms of HD [5, 6]. It is associated with higher morbidity and mortality rates and lower quality of life than short segment HD. With the improvement of medicine, the survival rate has been elevated remarkably over the decades.

However, the recurrent HAEC is still the most common and intractable problem to the TCA patients. There are numerous theories to try to explain the etiology of HAEC. However, the exact pathogenesis is still unclear. The functional obstruction by aganglionic internal anal sphincter is considered to be a major contributing factor [7]. Zhang firstly introduced the TRM–PIAS technique, which had shown the satisfied results to reduce the incidence of postoperative HAEC and constipation [3]. In this study, no constipation was reported but similar incidence of postoperative HAEC was comparable to other reports in the TRM–PIAS group [2, 6, 8, 9]. This finding might due to the intensive awareness of HAEC in the hospital, with early

recognition and prompt treatment. Thus, the home care of mild HAEC were recorded in this study. The incidences of postoperative HAEC in both groups were decreased over the time. However, the incidence of the HAEC in the TRM–PIAS group was significantly lower than the LEPT group in the second year after surgery.

It is quite obvious that the frequency of bowel movement, fecal soiling and perianal excoriation considerably impair quality of postoperative life. Several authors [2, 9–12] reported the techniques of Martin, Kimura or stapled J-pouch provided a lack of peristalsis aganglionic patch or reservoir that would help to form solid stool and decrease bowel movements. However, the outcomes such as anastomosis leak, recurrent HAEC, persistent diarrhea and pouchitis were also noted. Therefore, according to some authors [6, 13–15] suggested, a standard-length pull-through and direct ileoanal anastomosis were adopted in this study. Andrea Bischoff [16] suggested a strategy consisting of resection of the colon, ileo-recto anastomosis and ileostomy until the child was totally toilet trained. However, staging operation is a tremendous burden and hard to bear for the family in China.

After performing the TRM–PIAS technique and ileoanal anastomosis, the outcomes of bowel function are concerned predominantly. Liquid stool, stooling frequencies, fecal soiling, and perianal excoriation were noted in almost all patients within the early postoperative period. It is worth noted that the clinical outcomes in present study showed the anal function had been gradually improved over time [3, 6, 17]. With the decreased peristalsis, enhanced absorption of ileum and developed contraction of external anal sphincter, most of the patients in the TRM–PIAS group had decent quality of life in regard of their anal function 24 months postoperatively.

In this study, overall bowel functional outcomes demonstrated no significant difference between two groups. But the better outcomes in terms of bowel movement and soiling were noted in the TRM–PIAS group. Considering the similar overall bowel function scores and that the surgeries were performed by the same group of experienced clinicians, the loss of rectal sensation and the damage of anal sphincter might not be the trigger of the discrepant bowel function. Some studies demonstrated the internal sphincter spasm caused functional obstruction or the so called outlet obstruction after pull-through [2]. And other authors reported residual muscle cuff, whether split or not, also could lead to functional obstruction with recurrent HAEC, constipation or overflow incontinence in some of postoperative patients [18–21]. Therefore, some LEPT group patients might suffer from varying degrees of obstructive symptoms. Obstructive defecation with incomplete evacuation, frequent overflow incontinence and chronic HAEC with diarrhea, which were caused by obstructive symptom, might manifest increased bowel movement and soiling and might lead to recurrent perianal excoriation in some patients. In the procedure of TRM–PIAS, we resected including posterior half muscular cuff and part of internal anal sphincter to relief the obstructive symptoms. Moreover, Zhang [3] demonstrated anorectal resting pressure restored to the same level of healthy children 6 months after performing TRM–PIAS technique. These were possible explanations of improvement in bowel movement and soiling in the TRM–PIAS group.

Other complications, such as anastomosis leak, cuff abscess, dehiscence or retraction are preventable by meticulous surgical skill, strict sterilization and hemostasis, decent blood supply of the pull-through segment and tension-free anastomosis.

Primary pull-through without a stoma is still controversial. In this study, one patient was observed with severe perianal excoriation and obstinate HACE for half year in the TRM–PIAS group. Clearly, the comparison is difficult, as most patients had undergone an ileostomy.

There were some limitations in this study. Firstly, the present study was retrospective study. The follow-up period is relatively short. The sample size is small, particularly in the control group. Finally, we only evaluate the clinical parameters, but no other objective approaches for evaluating the functional outcome such as manometry. In the future, manometry study is required to obtain the objective evidence to support the improvement of the postoperative bowel function.

Conclusion

Within the limitations related to sample size and follow-up duration, this retrospective study demonstrated that: TRM–PIAS, proctocolectomy, and ileoanal anastomosis might be effective to treat the TCA. Moreover, the quality of life after surgery using the TRM–PIAS technique might be better than the LEPT procedure. We consider HACE as non-preventable complication in the early time after surgery among the TCA patients. Therefore, prompt diagnosis and treatment of postoperative HAEC are also important to the improvement of the outcomes. However, more prospective studies evaluating the TRM–PIAS technique over longer period and with greater sample size are needed to confirm the findings in this study.

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