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Redo pull-through in total colonic aganglionosis due to residual aganglionosis: a single center's experience

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Abstract

Background Reoperation for total colonic aganglionosis (TCA) may be required for residual aganglionosis after an initial radical operation. We aimed to investigate the symptoms, management, and outcomes of patients who required a redo pull-through (Redo PT).

Methods Nine TCA patients underwent Redo PT at our center between 2007 and 2017. Their medical records were reviewed. Parental telephone interviews that included disease-specific clinical outcomes were conducted, and post-operative complications and long-term outcomes (including height-for-age/weight-for-age and bowel-function score) were compared to those of single-pull-through (Single PT) patients ($n = 21$).

Results All the nine Redo PT patients suffered obstruction within 1 month after the initial operation that could not be alleviated by conservative treatment. All abdominal X-ray/contrast barium enemas showed proximal bowel dilatation, indicating residual aganglionosis. The median ages at the initial operation and Redo PT were 200 and 509 days, respectively. Reoperation consisted of an intraoperative frozen biopsy and a modified laparotomic Soave procedure in all patients. Post-operative complications included perianal excoriation ($n = 3$), intestinal obstruction ($n = 2$), enterocolitis ($n = 2$), and rectovestibular fistula ($n = 1$). Seven Redo PT patients were followed up for a mean time of 7.1 ± 2.3 years; six (85.7%) had good growth and four (57.1%) had good bowel-function recovery. Post-operative complications and long-term outcomes were almost equal between the Redo PT and Single PT groups (all $P > 0.05$).

Conclusion TCA patients with recurrent obstructive symptoms and dilated proximal bowel may have residual aganglionosis after an initial operation. Redo PT is effective and provides good long-term outcomes comparable to those of patients who benefited from Single PT.

Key words: total colonic aganglionosis; residual aganglionosis; redo pull-through

Introduction

Total colonic aganglionosis (TCA) is an uncommon and severe disease that occurs in ~3%–15% of patients with Hirschsprung's disease (HD) [1–4]. Many different surgical techniques, such as the Soave, Swenson, and Martin techniques, have been described for TCA, but the best procedure remains unclear [5]. Successful operative management of TCA leads to a satisfactory outcome [6, 7]. However, TCA patients have more post-operative complications than patients with short-segment aganglionosis because, for TCA patients, more time is needed to resolve issues with bowel control, night-time diarrhea, and defecation patterns [1, 3, 8]. In addition, some TCA patients require multiple surgeries due to failure to consider the diagnosis or the absence of enteric ganglion cells by performing intraoperative multiple full-thickness punch biopsies during the initial operation [2, 9, 10]. Thus, these patients suffer from obstructive symptoms, persistent constipation, and recurrent enterocolitis due to residual aganglionosis from the initial operation and ultimately require reoperation [5]. Some studies in HD patients have analysed the diagnosis and treatment of residual aganglionosis and evaluated outcomes after reoperation [10–13], but literature concerning TCA patients' long-term outcomes following reoperation is limited. The aim of this study was to evaluate the long-term outcomes of patients who underwent a redo pull-through (Redo PT) due to residual aganglionosis by comparing them with consecutive TCA patients who underwent a single pull-through (Single PT). This current study represents one of the largest series of reoperations in TCA patients who have undergone an initial operation.

Materials and methods

Study design and patients

A retrospective review was made of the patient databases at the neonatal and general surgery departments, Beijing Children's Hospital. Patients who were diagnosed with TCA with the involvement of ≤50 cm of the terminal ileum by intraoperative multiple full-thickness punch biopsies, experienced post-operative obstruction after initial radical operation that was not alleviated by conservative treatments, and underwent a Redo PT at our center between June 2007 and June 2017 were included. They were compared to Single PT-matched patients treated at our center. All patients' diagnoses were confirmed by post-operative pathological examination. The study was approved by the Ethics Committee of Beijing Children's Hospital (approval number 2018-k-144).

Surgical treatment

Both Single PT and Redo PT at our center involve the modified laparotomy Soave procedure, which was suggested for HD patients based on the technique described by Soave [14] at the Institute G. Gaslini in 1964. All patients were treated in the neonatal or general surgery departments at our center according to their age at admission. The principles of the surgical strategy, including the Soave procedure and perioperative management, were the same in both departments. Anal-sphincter myotomy was not performed, but daily anal dilatation with metal dilators starting 2 weeks after pull-through was regularly performed for all patients for 3–6 months.

Follow-up and outcome measurement

All patients were followed for >2 years after the pull-through procedure. A questionnaire including the age at follow-up, post-operative complications, height, weight, and bowel-function score (BFS) was completed through parental telephone interviews in June 2019. Post-operative complications mainly included anastomotic fistula and stenosis, anal prolapse, intestinal obstruction, sepsis, enterocolitis, and perianal excoriation.

Growth was evaluated in terms of height-for-age (HFA) or weight-for-age (WFA). The patients were divided into three categories according to the median percentage of HFA or WFA: for HFA, 90%–110% indicated normal growth, 75%–89% indicated mild malnutrition, and <75% indicated moderate/severe malnutrition; for WFA, >95% indicated normal growth, 90%–95% indicated mild malnutrition, and <90% indicated moderate/severe malnutrition [15].

Bowel-function recovery was assessed by using BFS when the children were >4 years old. This score mainly evaluates the ability to hold back defecation, feeling/reporting the urge to defecate, and the frequencies of defecation, soiling, fecal accidents, constipation, and social problems. The patients were divided into three categories according to the total BFS: ≥17 indicated normal bowel function, 12–16 indicated mild bowel dysfunction, and <12 indicated severe bowel dysfunction [16].

Statistical analysis

Categorical variables were analysed with the chi-square test or Fisher's exact test. Continuous variables with normal distribution were presented as mean ± standard deviation (SD) and analysed using Student's t-test. Continuous variables with non-normal distribution were presented as median with range and analysed using the Mann–Whitney test. $P < 0.05$ (two-sided) was considered significant. Statistical calculations were performed using a software program (IBM SPSS Package, version 22.0; IBM Corporation).

Results

Overall TCA series ($n = 68$)

Sixty-eight patients with TCA were admitted to our center between June 2007 and June 2017. The male-to-female ratio was 50:18. Of these patients, 17 (25.0%) did not continue treatment after confirmation by laparotomy, 14 (20.1%) underwent a staged pull-through after undergoing initial enterostomy as neonates, 21 (30.9%) underwent Single PT, 4 patients had only undergone an enterostomy (5.9%) as of this study, and 12 (17.6%) had previously undergone surgery at another hospital.

Redo series due to residual aganglionosis ($n = 9$)

During the study period, nine patients (13.2%) underwent a Redo PT including three patients undergoing an initial Soave procedure at our center and all indications involved residual aganglionosis (Table 1). They all experienced obstructive symptoms (chronic abdominal distension and/or constipation) within 1 month after the initial operation and failed to respond to conservative treatments to alleviate symptoms before admission.

Abdominal X-ray was performed before reoperation in six patients and showed various degrees of proximal bowel dilatation (Figure 1). Contrast barium enema was performed in seven patients before reoperation and still indicated short or long

Table 1. Clinical features of patients with total colonic aganglionosis (TCA) requiring redo pull-through (Redo PT) surgery

Case	Age at initial operation (days)	Sex	Failed procedures before admission	Symptoms before Redo PT (days)	X-ray/enema before Redo PT	Treatment before Redo PT (days)	Indication for Redo PT	Type of reoperation	Age at Redo PT (days)
1	3	Male	Laparoscopic Soave, Out	Obstruction + recurrent HAEC	Yes/Yes	Anal dilatation + antibiotic therapy + colonic lavage	Residual aganglionosis	Soave	499
2	10	Male	Unknown, Out	Obstruction	Yes/Yes	Anal dilatation + antibiotic therapy	Residual aganglionosis	Soave	213
3	78	Female	Laparotomic Soave, In	Obstruction	No/No[a]	Anal canal indwelling	Residual aganglionosis	Soave	84
4	180	Male	Ileostomy / Martin, Out	Obstruction	Yes/Yes	Anal dilatation + anal indwelling + colonic lavage + ileostomy (In)	Residual aganglionosis	Soave	1,502
5	200	Male	Laparoscopic Soave, Out	Obstruction	No/Yes[b]	Changes in diet + laparoscopy (Out)	Residual aganglionosis	Soave	525
6	260	Male	Laparotomic Soave, In	Obstruction	Yes/Yes	Colonic lavage	Residual aganglionosis	Soave	386
7	270	Female	Laparotomy / unknown, Out	Obstruction	Yes/Yes	Colonic lavage	Residual aganglionosis	Soave	509
8	271	Male	Laparotomic Soave, In	Obstruction	No/No[c]	Colonic lavage + ileostomy (In)	Residual aganglionosis	Soave	522
9	317	Male	Unknown, Out	Obstruction + recurrent HAEC	Yes/Yes	Anal dilatation + colonic lavage + ileostomy (In)	Residual aganglionosis	Soave	959

In: at our center; Out: at other hospital; HAEC: Hirschsprung's-associated enterocolitis.
[a] Post-operative pathology of initial operation showing residual aganglionosis.
[b] Abdominal ultrasound instead of X-ray.
[c] Diagnosed with TCA by intraoperative frozen biopsies during the initial operation, but the ascending colon was preserved at the insistence of the parents.
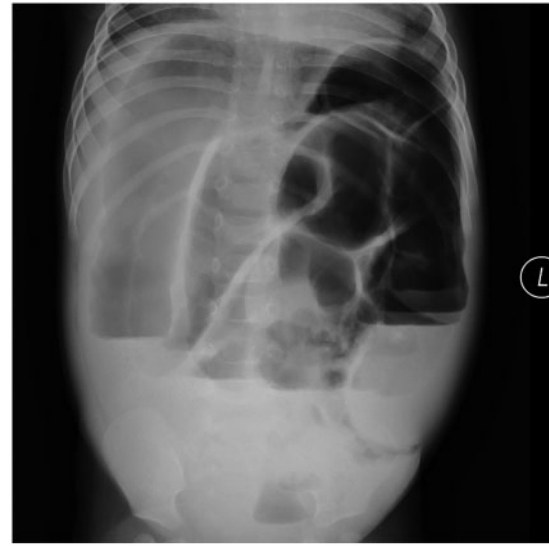

Figure 1. Abdominal X-ray showed proximal bowel dilatation

segment of aganglionosis (Figure 2). Two patients (Cases 3 and 6) underwent a Redo PT due to differences in the results of the intra- and post-operative pathological examination during initial operation at our center (Figure 3). One patient (Case 8) was diagnosed with TCA by intraoperative frozen biopsy during the initial operation at our center, but the parent insisted on preserving the ascending colon. Two patients (Cases 4 and 9) received an ileostomy before Redo PT over 1 year after the initial operation due to severe bowel dilation. All patients underwent a laparotomic Soave procedure as a Redo PT and were confirmed with residual aganglionosis by post-operative pathological examination.

Control group (n = 21)

A total of 21 consecutive patients who underwent Single PT during the same study period were used as the control group. The patient characteristics and perioperative data for the Single PT and Redo PT groups are shown in Table 2. The median ages at Single PT and Redo PT were 55 (9–1,509) and 509 (84–1,502) days, respectively (P = 0.01). There were no significant differences in sex, birth weight, family history, operative time, and intraoperative blood loss between the two groups (all P > 0.05). Three patients (14.3%) in the Single PT group died during the perioperative period; one died of septic shock and two died of dehydration and infection.

Long-term outcomes of the Single PT and Redo PT groups

There were 15 patients in the Single PT group and 7 patients in the Redo PT group who completed the follow-up questionnaire. A comparison of long-term outcomes between the two groups, including post-operative complications, growth, and bowel-function recovery, is shown in Table 3. The most prevalent uncontrolled complication was perianal excoriation, which occurred in both groups (66.6% vs 42.9%, P = 0.38), followed by enterocolitis (33.3% vs 28.6%, P = 0.99) and intestinal obstruction (13.3% vs 28.6%, P = 0.57). In addition, one patient in the Single PT group exhibited rectal prolapse and sepsis, whereas one patient in the Redo PT group had an acquired persistent rectovestibular fistula for >11 years.
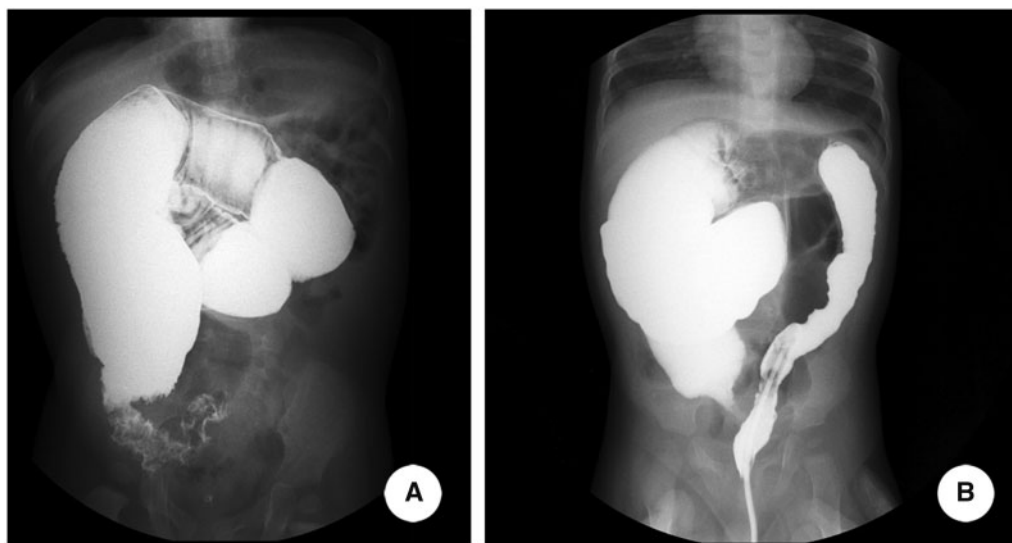
Figure 2. Contrast barium enema performed before reoperation indicates a short (A) or long (B) segment of aganglionosis
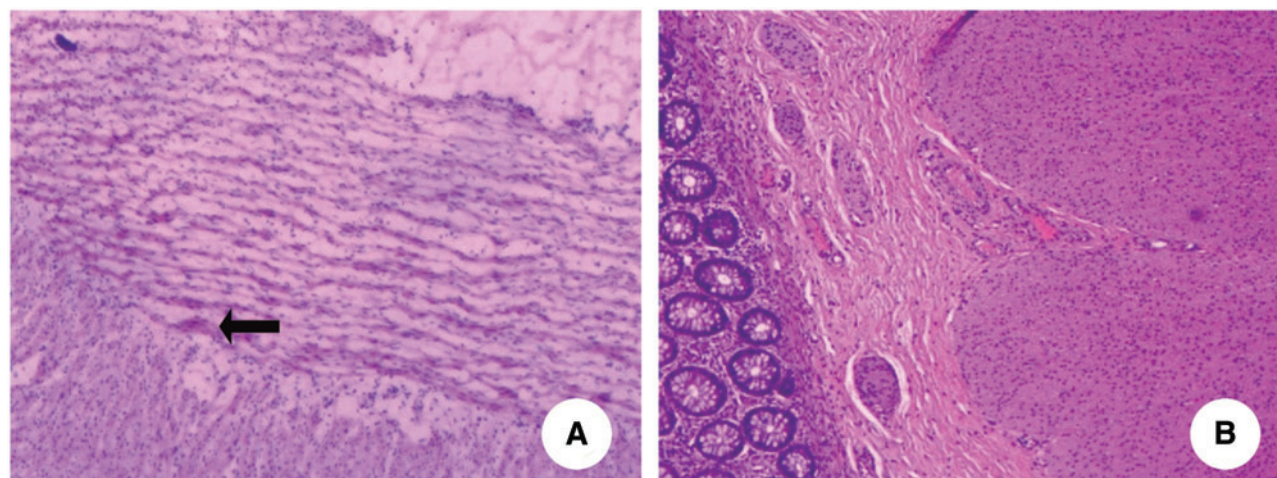

Figure 3. Intra- and post-operative pathological examination of Case 6. (A) Intraoperative seromuscular biopsies showing atypical, immature ganglion cells (arrow) in the proximal margin. (B) Post-operative pathology showing no ganglion cells in the whole resected colon.

Table 2. Comparison of patient characteristics and perioperative data between the two groups

Characteristic	Single PT ($n=21$)	Redo PT ($n=9$)	P-value
Male-to-female ratio	13/8	7/2	0.68
Birth weight, g, mean ± SD	3,530 ± 344	3,467 ± 327	0.65
Family history, n (%)	2 (9.5)	1 (11.1)	0.99
Age at pull-through, days, median (range)	55 (9–1,509)	509 (84–1,502)	0.01
Operation time, min, mean ± SD	196 ± 48	188 ± 42	0.69
Blood loss, mL, median (range)	5 (2–20)	5 (2–50)	0.28
Perioperative death, n (%)	3 (14.3)	0 (0.0)	0.53

PT, pull-through procedure; SD, standard deviation.

The median HFA or WFA percentage was applied to assess the patients' growth. The proportions of patients with good growth in the Single PT and Redo PT groups were similar based on both HFA (93.3% vs 85.7%, $P=0.57$) and WFA (86.7% vs 85.7%, $P=0.95$). Eighteen patients >4 years of age completed the BFS questionnaire; 7 (63.6%) in the Single PT group and 4 (57.1) in the Redo PT group had good bowel-function recovery ($P=0.49$). The patient with acquired persistent rectovestibular fistula presented with mild malnutrition based on HFA and severe based on WFA. The patient with rectovestibular fistula had severe malnutrition (WFA < 90%) and severe bowel dysfunction (BFS = 11).

Table 3. Comparison of post-operative complications and long-term outcomes between the two groups

Outcome	Single PT ($n = 15$)	Redo PT ($n = 7$)	P-value
Age at follow-up, years, mean $\pm$ SD	5.6 $\pm$ 2.0	8.4 $\pm$ 2.5	0.01
Length of follow-up, years, mean $\pm$ SD	4.8 $\pm$ 1.8	7.1 $\pm$ 2.3	0.02
Post-operative complications, n (%)			
Enterocolitis	5 (33.3)	2 (28.6)	0.99
Perianal excoriation	10 (66.7)	3 (42.9)	0.38
Intestinal obstruction	2 (13.3)	2 (28.6)	0.57
Rectovestibular fistula	0 (0.0)	1 (14.3)	0.32
Rectal prolapse	1 (6.7)	0 (0.0)	0.99
Sepsis	1 (6.7)	0 (0.0)	0.99
Height-for-age, n (%)			0.57
Normal (90%–110%)	14 (93.3)	6 (85.7)	
Mild malnutrition (75%–89%)	1 (6.7)	1 (14.3)	
Moderate/severe malnutrition (<75%)	0 (0.0)	0 (0.0)	
Weight-for-age, n (%)			0.95
Normal (>95%)	13 (86.7)	6 (85.7)	
Mild malnutrition (90%–95%)	0 (0.0)	0 (0.0)	
Moderate/severe malnutrition (<90%)	2 (13.3)	1 (14.3)	
Bowel-function score, n (%)[a]			0.49
Normal (≥17)	7/11 (63.6)	4/7 (57.1)	
Mild bowel dysfunction (12–16)	4/11 (36.4)	2/7 (28.6)	
Severe bowel dysfunction (<12)	0/11 (0.0)	1/7 (14.3)	

PT, pull-through procedure; SD, standard deviation.

[a] Bowel-function score was evaluated only in patients >4 years of age.

Discussion

With advancements in perinatal management and improvements in operative management, TCA cases have gradually achieved favorable outcomes, with lower misdiagnosis and death rates [3, 17]. Considering more complex treatment and unpredictable post-operative complications in these cases than in cases of short or classic segment HD, TCA is still a challenging disease for pediatric surgeons in terms of diagnosis and treatment [17–19]. Many surgical procedures and approaches (from staged to primary) have been described for TCA and no technique has been proven to be superior to others with regard to post-operative complications and functional outcomes [6, 7, 20]. Unlike HD cases, there are few studies concerning individuals with TCA undergoing a reoperation while experiencing persistent complications after the initial operation due to the limited number of TCA patients [10, 11, 21–23]. The reoperation for TCA is more technically difficult and risky, and the long-term prognosis is unclear. Our study described long-term follow-up of nine Redo PT patients and showed that such patients experienced post-operative relief from symptoms caused by residual aganglionosis and achieved good long-term outcomes.

Despite the accumulation of surgical experience and the development of conservative treatments, post-operative complications remain a serious issue for TCA patients [5]. Some complications, such as enterocolitis, may be successfully treated with conservative treatment and perianal excoriation can be improved by the recovery of bowel function [1–3, 24]. However, a small fraction of patients still require reoperation because of anatomical or histopathological problems after the initial operation [21]. The most frequent indication for reoperation is residual aganglionosis, which accounts for more than half of redo patients [23]. The use of full-thickness intraoperative biopsies rather than seromuscular biopsies (without submucosa) is far more important for avoiding residual aganglionosis, which also depends on the experience level of the pathologists [10, 21, 25, 33]. In our study, all patients were confirmed to have residual aganglionosis by full-thickness intraoperative biopsies during Redo PT.

Obstruction was shown to be the most relevant symptom in TCA patients requiring reoperation. Many studies have shown that adhesion is the most common etiology of obstruction after surgery [2, 20, 26, 27]. However, residual aganglionic bowel remains a primary etiology of concern in TCA patients with persistent obstruction that cannot be alleviated by conservative treatment and surgical management by Redo PT has been generally recommended to address this concern [12, 13]. We make the diagnosis of residual aganglionosis based on the combination of abdominal X-ray, barium enema, and even rectal-suction biopsy. Intraoperative multiple full-thickness punch biopsy is still the gold standard [28]. The need for a stoma is controversial and it depends upon the initial pull-through technique used, the availability of conservative treatment options, and the overall condition of each patient. All patients who were confirmed to have residual aganglionosis underwent a Soave procedure in our center because of the simple steps and acceptable outcomes.

The median age at which the patients in the Redo PT group underwent the Soave procedure was much older than that of the Single PT group because every patient in the Redo PT group received an initial radical operation followed by conservative treatment before undergoing Redo PT. Although there was no significant difference between the two groups in terms of operative time and intraoperative blood loss, we found that the perioperative mortality of the Redo PT group was slightly lower than that of the Single PT group. This finding cannot be fully explained, but it may be caused by the younger age of the patients when they underwent Single PT, which agrees with our previous findings that an early radical operation yielded a negative outcome [29].

A short-term clinical study found that the outcome after a Redo PT for HD is complicated by a relatively high rate of soiling

and fecal incontinence [22]. In the present study, no significant difference in post-operative complications and clinical outcomes was found between the Redo PT and Single PT groups. In the literature review, enterocolitis and perianal excoriation were the most common and challenging post-operative complications in TCA patients and our results concur with this finding [3, 4, 28]. Most of the reported complications were observed to improve over time (months or years) with bowel-function recovery. However, a successful pull-through procedure performed by an experienced pediatric surgeon remains a challenge in TCA patients because Redo PT is technically more difficult and has a higher risk of some serious complications, such as rectovestibular fistula [5, 21]. In our study, a female patient suffered from an acquired rectovestibular fistula after Redo PT because of an intraoperative injury. Due to the poor recovery of bowel function and repeated infection through the persistent fistula, the patient's growth and mental health were affected. At present, we are actively communicating with her parents and planning to repair the rectovestibular fistula with anterior perineal fistulectomy and another pull-through because refistulization may be prevented by pulling an intestinal wall sleeve through the original site of the fistula [30, 31].

In addition, in the Redo PT group, growth and BFS recovered gradually with increasing post-operative time and bowel function was normal ($\geq$17) $\sim$8.5 years after Redo PT. Compared with other studies, the long-term follow-up for TCA patients after Single PT or Redo PT in our study showed more favorable growth and bowel-functional outcomes [3, 32]. On the one hand, this finding might be related to the fact that none of the patients included in this study had severe congenital malformations such as Down syndrome [1]. On the other hand, careful management and lifelong follow-up by specialized teams, including surgeons, nurses, and students, improves the prognosis of patients [32]. However, during the follow-up, we found that, after the pull-through procedure, TCA patients were unable to tolerate cold foods and hypertonic foods such as juice and some fruits, which would lead to diarrhea. Additionally, there were no objective indicators to evaluate the psychological status of these patients.

The present study has some strengths and limitations. One strength is that it includes one of the largest cohorts of TCA patients treated by Redo PT at a single center with substantial long-term follow-up. Although aspects of the Redo PT were previously reported for cases of HD, the present study included more recent TCA patients, a high follow-up rate, and new information regarding post-operative complications. The limitations of this study are the relatively low number of patients and the fact that detailed information about outcomes was only acquired from conversations with the patients' parents. Selection bias toward patients with a good outcome is possible.

In conclusion, TCA patients with recurrent obstructive symptoms and a dilated proximal bowel may have residual aganglionosis after an initial operation. Redo PT is effective for them and offers good long-term outcomes comparable to those of patients who required only Single PT.

Authors' contributions

J.Y.Y. contributed to data collections, data analysis, drafting the manuscript, and writing the paper; C.L.D. contributed to data collections; W.B.P. contributed to data analysis; C.H.P. participated in the writing of the paper; Y.W.C. contributed to study design, data collections, data analysis, and writing the paper;

Y.J.C. contributed to study design, data analysis, and writing the paper. All authors read and confirmed the final manuscript.

Conflicts of interest

None declared.

References

1. Escobar MA, Grosfeld JL, West KW *et al.* Long-term outcomes in total colonic aganglionosis: a 32-year experience. *J Pediatr Surg* 2005;**40**:955–61.
2. Wildhaber BE, Teitelbaum DH, Coran AG. Total colonic Hirschsprung's disease: a 28-year experience. *J Pediatr Surg* 2005;**40**:206–7.
3. Menezes M, Pini PA, Jasonni V *et al.* Long-term clinical outcome in patients with total colonic aganglionosis: a 31-year review. *J Pediatr Surg* 2008;**43**:1696–9.
4. Moore SW. Total colonic aganglionosis and Hirschsprung's disease: a review. *Pediatr Surg Int* 2015;**31**:1–9.
5. Bischoff A, Levitt MA, Peña A. Total colonic aganglionosis: a surgical challenge. how to avoid complications? *Pediatr Surg Int* 2011;**27**:1047–52.
6. Shen C, Song Z, Zheng S *et al.* A comparison of the effectiveness of the Soave and Martin procedures for the treatment of total colonic aganglionosis. *J Pediatr Surg* 2009;**44**:2355–8.
7. Cheung ST, Tam YH, Chong HM *et al.* An 18-year experience in total colonic aganglionosis: from staged operations to primary laparoscopic endorectal pull-through. *J Pediatr Surg* 2009;**44**:2352–4.
8. Ieiri S, Suita S, Nakatsuji T *et al.* Total colonic aganglionosis with or without small bowel involvement: a 30-year retrospective nationwide survey in Japan. *J Pediatr Surg* 2008;**43**:2226–30.
9. Tsuji H, Spitz L, Kiely EM *et al.* Management and long-term follow up of infants with total colonic aganglionosis. *J Pediatr Surg* 1999;**34**:158–61.
10. Lawal TA, Chatoorgoon K, Collins MH *et al.* Redo pull-through in Hirschsprung's disease for obstructive symptoms due to residual aganglionosis and transition zone bowel. *J Pediatr Surg* 2011;**46**:342–7.
11. Leeuwen KV, Teitelbaum DH, Elhalaby EA *et al.* Long-term follow-up of redo pull-through procedures for Hirschsprung's disease: efficacy of the endorectal pull-through. *J Pediatr Surg* 2000;**35**:829–33.
12. Gobran TA, Ezzat A, Hassan ME *et al.* Redo transanal endorectal pullthrough: a preliminary study. *Pediatr Surg Int* 2007;**23**:189–93.
13. Ghose SI, Squire BR, Stringer MD *et al.* Hirschsprung's disease: problems with transition-zone pull-through. *J Pediatr Surg* 2000;**35**:1805–9.
14. Soave F. Hirschsprung's disease: a new surgical technique. *Arch Dis Child* 1964;**39**:116–24.
15. World Health Organization & United Nations Children's Fund. WHO child growth standards and the identification of severe acute malnutrition in infants and children. 2009.

16. Jarvi K, Laitakari EM, Koivusalo A *et al*. Bowel function and gastrointestinal quality of life among adults operated for Hirschsprung disease during childhood: a population-based study. *Ann Surg* 2010;**252**:977–81.

17. Laughlin DM, Friedmacher F, Puri P. Total colonic aganglionosis: a systematic review and meta-analysis of long-term clinical outcome. *Pediatr Surg Int* 2012;**28**:773–9.

18. Jasonni V, Martucciello G. Total colonic aganglionosis. *Semin Pediatr Surg* 1998;**7**:174–80.

19. Marquez TT, Acton RD, Hess DJ *et al*. Comprehensive review of procedure for total colonic aganglionosis. *J Pediatr Surg* 2009;**44**:257–65.

20. Hukkinen M, Koivusalo A, Merras-Salmio L *et al*. Postoperative outcome and survival in relation to small intestinal involvement of total colonic aganglionosis. *J Pediatr Surg* 2015;**50**:1859–64.

21. Ralls MW, Freeman JJ, Rabah R *et al*. Redo pullthrough for Hirschsprung disease: a single surgical group's experience. *J Pediatr Surg* 2014;**49**:1394–9.

22. Dingemans AJM, van der Steeg HJJ, Rassouli-Kirchmeier R *et al*. Redo pull-through surgery in Hirschsprung's disease: short term clinical outcome. *J Pediatr Surg* 2017;**52**:1446–50.

23. Pini-Prato A, Mattioli G, Giunta C *et al*. Redo surgery in Hirschsprung disease: what did we learn? Unicentric experience on 70 patients. *J Pediatr Surg* 2010;**45**:747–54.

24. Granéli C, Dahlin E, Börjesson A *et al*. Diagnosis, symptoms, and outcomes of Hirschsprung's disease from the perspective of gender. *Surg Res Pract* 2017;**2017**:1–8.

25. Joseph OT, Michael MD, Maureen O *et al*. A retrospective cohort study of total colonic aganglionosis: is the appendix a reliable diagnostic tool? *J Neonatal Surg* 2016;**5**:44.

26. Reddy SRR, Cappell MS. A systematic review of the clinical presentation, diagnosis, and treatment of small bowel obstruction. *Curr Gastroenterol Rep* 2017;**19**:28.

27. Choi HK, Law WL, Ho WC *et al*. Value of gastrografin in adhesive small bowel obstruction after unsuccessful conservative treatment: a prospective evaluation. *World J Gastroenterol* 2005;**11**:3742–5.

28. Cinel L, Ceyran B, Güçlüer B. Calretinin immunohistochemistry for the diagnosis of Hirschprung disease in rectal biopsies. *Pathol Res Pract* 2015;**211**:50–4.

29. Yan JY, Chen YJ, Ding CL *et al*. Clinical outcomes after staged and primary laparotomy Soave procedure for total colonic aganglionosis: a single-center experience from 2007 to 2017. *J Gastrointest Surg* 2020;**24**:1673–81.

30. Kubota A, Kawahara H, Okuyama H *et al*. Endorectal pull-through with posterior sagittal approach to the repair of postoperative rectourethral and rectovaginal fistula. *J Pediatr Surg* 2003;**38**:1775–7.

31. Wang ZM, Chen YJ, Zhang TC *et al*. Anterior perineal fistulectomy in repairing H-type rectovestibular fistula with a normal anus in female children. *J Pediatr Surg* 2015;**50**:1425–8.

32. Stenström P, Brautigam M, Borg H *et al*. Patient-reported Swedish nationwide outcomes of children and adolescents with total colonic aganglionosis. *J Pediatr Surg* 2017;**52**:1302–7.