

# Comparison of long-term outcome according to involved aganglionic segments of total colonic aganglionosis

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## Abstract

Total colonic aganglionosis (TCA) is a rare form of Hirschsprung disease, with more severe symptoms than rectosigmoid Hirschsprung disease. We aimed to evaluate the surgical outcomes according to the involved segments of TCA.

Patients with aganglionosis extending from the anus to at least the ileocecal valve were included. The medical records of 33 TCA patients from 1981 to 2014 were reviewed. Three groups were analyzed based on the involved segment (jejunum, jejunoleal junction, and distal ileum).

The median age at the pull-through operation was 6.2 (3.3–114) months. The median follow-up duration was 216 (21–411) months. Transition zone in the jejunum, jejunoleal junction, and distal ileum was identified in 3, 5, and 25 patients, respectively. The most common method of operation was Duhamel pull-through. Perianal excoriation and enterocolitis were the most common postoperative complications. The complication rates were 45% to 51% and not different among the groups. The defecation frequency normalized 3 years postoperatively, and body weight started to recover after 2 years irrespective of the involved segment.

Therefore, close monitoring with proper management of defecation and body weight for at least 2 to 3 years postoperatively is required.

**Abbreviations:** HD = Hirschsprung disease, PN = parenteral nutrition, TCA = total colonic aganglionosis.

**Keywords:** body weight change, defecation, postoperative complications, total colonic aganglionosis

## 1. Introduction

Total colonic aganglionosis (TCA) is a rare form of Hirschsprung disease (HD) which has been defined as aganglionosis extending from the anus to at least the ileocecal valve.<sup>[1,2]</sup> According to previous investigations, TCA accounts for 2% to 13% of all HD patients.<sup>[3]</sup>

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All data generated or analyzed during this study are included in this published article [and its supplementary information files].

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Patients with TCA show more severe symptoms, such as intestinal obstruction or abdominal distention, than those with rectosigmoid HD, and 80% of the patients experience these symptoms in the neonatal period.<sup>[4]</sup> TCA is difficult to correctly diagnose after finding the exact transition zone; thus, multiple operations are frequently performed with more long-term sequelae.<sup>[5,6]</sup> Therefore, perioperative mortality rate is higher than that of shorter-segment HD.<sup>[7–9]</sup>

Although various types of clinical studies have evaluated the clinical course, long-term outcomes, and results of different types of operations,<sup>[1,2,5,6,10–17]</sup> there are few studies on the clinical outcomes according to the involved segments of TCA. The purpose of this study was to evaluate and compare the surgical outcomes, postoperative complications, defecation frequency, and change of body weight according to the involved segments of TCA.

## 2. Materials and methods

### 2.1. Patient enrollment

Patients who underwent definitive operations for TCA at Seoul National University Children's Hospital from 1981 to 2014 and followed up at our center were included. We defined TCA as aganglionosis affecting the entire colon as described in our previous study.<sup>[12]</sup> Patients with or without involvement of variable length of the small intestine were included in the analysis.

The total number of HD patients identified during the period was 1298, including 41 (3.16%) patients with TCA. Of these, only 33 patients were included in the analysis, as 8 patients who

did not undergo pull-through surgery and currently have a permanent ostoma were excluded.

## 2.2. Division into 3 groups according to involved aganglionic segments

We divided our TCA patients into 3 groups according to the involved aganglionic segments, whose transition zones were pathologically confirmed. The confirmation of the transition zone was carried out in stages. First, rectal full-thickness biopsy was performed in the operating room. After confirming the presence of aganglionosis in the frozen rectal biopsy, further frozen biopsy was also performed in the sigmoid/descending, transverse, and ascending colon. If colonic aganglionosis was identified, then we proceeded to multiple small intestinal biopsy, and an ostoma was formed at the ganglionic location identified from the frozen biopsy result. The permanent biopsy report of these multiple intestinal frozen biopsy was checked to see if the results were corrected before the pull-through operation.

The jejunum group was defined as patients with involvement of up to the proximal one-third of the small intestine. The jejunoileal junction group included patients with involvement of up to the middle one-third of the small intestine. The distal ileum group comprised patients with involvement up to the distal one-third of the small intestine.

Two surgeons performed the Duhamel, Martin modification, and Soave procedures as the pull-through methods at our institution. Duhamel procedure is retrorectal approach with a segment of aganglionic rectum retained and anastomosed side-to-side with ganglionic bowel. Martin operation is modification of Duhamel procedure with longer aganglionic bowel remained. In the Soave procedure, or endorectal pull-through, ganglionic bowel is pulled through the aganglionic muscular cuff of the rectum after rectal mucosa and submucosa resection. One surgeon performed open Duhamel operation mainly in the earlier study period. The other surgeons preferred laparoscopic Soave procedure in the later study period, and chose Duhamel operation as the re-pull through method. Both performed Duhamel or Martin procedures if jejunum was involved.

## 2.3. Review of data and patient interview

Patient demographics, operation-related data, and postoperative complications were reviewed through retrospective chart reviews and telephone interviews. Complications after pull-through surgery were classified into early and late complications based on whether they developed within 4 weeks postoperatively or later. The surgical outcomes were confirmed using the difference between preoperative and postoperative body weight measurements and the postoperative defecation frequency including incontinence. The median follow-up period was 216 months (range, 21–411 months).

## 2.4. Statistical analysis

Standard score, or z-score is a dimensionless number that statistically creates a normal distribution and shows where individual cases occupy the standard deviation. Z-score was used to check the weight change.

SPSS version 21.0 (SPSS Inc., Chicago, IL) was used for statistical analysis. Categorical variables were compared using the chi-square or Fisher exact test, and continuous variables using

the Mann–Whitney *U* test. A *P*-value of  $<.05$  was considered significant. Median with range or mean values were used for comparison of continuous variables such as age and body weight.

## 3. Results

### 3.1. Data related to patient demographics and involved segments

Of the 33 patients, 3, 5, and 25 were classified into the jejunum group, jejunoileal junction group, and distal ileum group, respectively. The first 2 groups can also be termed as TCA with small bowel extension, accounting for 24.2% of all patients. There were 24 (72.7%) male patients, and the median weight at birth was 3.40 kg (range, 2.52–4.31 kg). There were 2 patients of cardiac and 3 pulmonary anomalies. The median age at diverting ostoma formation was 1 month (range, 0.1–108.0 months). The median age at the pull-through procedure was 6.2 months (range, 3.3–114 months). The Duhamel, Martin, and Soave procedures were performed in 19, 10, and 4 patients, respectively (Table 1).

We compared the demographics and surgical data of the 3 groups according to the transition zone. Sex, gestational age, body weight at birth, and underlying disease were not different among the 3 groups. All patients were diagnosed with TCA within the first 6 months of life, except for 1 patient in the distal ileum group who was diagnosed at age 9 years. In 23 patients, TCA was suspected and diagnosed during the neonatal period. There was no difference in operative method among groups; however, the Soave operation was performed in the distal ileum group only (Table 1).

### 3.2. Postoperative complications

Perianal excoriation was the most common early complication and was observed in 10 patients. Three patients were hospitalized with enterocolitis, and 2 patients had postoperative wound problems. All patients with early complications showed improved symptoms after supportive management. Late complications included enteritis, ileus, septum adhesion after Duhamel or Martin modification procedures, anorectal relaxation difficulty, and re-pull-through operation. Pathologic review and rectal biopsy were performed in 2 patients from distal ileum group, who underwent repeated hospitalization for abdominal distension and diarrhea, and re-pull-through was performed after acquired secondary aganglionosis was suspected. Two patients with septum adhesion underwent septum division with a stapler, under general anesthesia. Postoperative anorectal relaxation difficulty was present in 2 patients in the distal ileum group. For these patients, Hegar dilatation and Botox injection were performed, respectively; however, as the symptoms did not improve, posterior myectomy was performed. All other late complications improved with conservative treatment. No anastomosis leak was observed in all patients. The frequency of early and late complications did not differ among groups (Table 2).

All patients were supplied with parenteral nutrition (PN) immediately after pull-through operation, and only 10 had PN supply for more than 30 days postoperatively. Currently, no patient is under PN at home.

### 3.3. Defecation frequency

The mean defecation frequency was 7.7 per day after 3 months from operation, with no significant difference among the groups.

**Table 1**  
Demographics and operation-related data according to involved segments.

	Total	Jejunum	Jejunoleal junction	Distal ileum	P value
Number of patients (%)	33 (100)	3 (9.1)	5 (15.2)	25 (75.7)	-
Male (%)	24 (72.7)	2 (66.7)	4 (80)	18 (72)	.246
Gestational age (wk)*	39.0 (35.3–41.4)	39.0 (39.0–40.1)	39.1 (38.0–40.4)	39.4 (35.3–41.4)	.511
Body weight at birth (kg)*	3.40 (2.52–4.31)	3.37 (2.91–3.78)	3.18 (2.78–4.23)	3.49 (2.52–4.31)	.434
Underlying conditions (%)	5 (15.2)	1 (33.3)	0	4 (16)	.311
Cardiac defects	2 (6.1)	1 (33.3)	0	1 (4)	
Respiratory disorders	3 (9.1)	0	0	3 (12)	
Renal agenesis	1 (3)	0	0	1 (4)	
Hypothyroidism	1 (3)	0	0	1 (4)	
MEN2 syndrome	1 (3)	0	0	1 (4)	
Age at diagnosis with ostoma formation (mo)*	1.0 (0.1–108.0)	0.3 (0.1–0.3)	1.0 (0.5–6.0)	1.0 (0.1–108.0)	.079
Age at pull-through (mo)*	6.2 (3.3–114)	6.5 (6.3–12)	5.8 (4.5–10.1)	6.0 (4.3–114)	.371
Body weight at pull-through (kg)*	8 (4.3–20.5)	7.5 (6.8–8.2)	7.2 (6.3–9.1)	8 (4.3–20.5)	.214
Methods of pull-through	–	–	–	–	.329
Duhamel (%)	19 (57.6)	3 (100)	3 (60)	13 (52)	
Martin (%)	10 (30.3%)	0	2 (40)	8 (32)	
Soave (%)	4 (12.1%)	0	0	4 (16)	
Follow-up (mo)*	216 (21–411)	154 (139–262)	248 (23–295)	214 (21–411)	.653

MEN2 = multiple endocrine neoplasia type 2.  
\*represents median values with range.

The defecation frequency decreased gradually; the mean number of defecations per day after 3 years from operation and at the current telephone interview (median 216 months after the operation) was 2.4 and 2.3, respectively. There were no differences among groups (Table 3, Fig. 1).

**3.4. Postoperative changes in body weight**

The z-score of body weight at pull-through surgery were –1.05, which were lower than the averages of the same age group and showed no differences among groups. The mean z-score of body weight decreased by 2 years postoperatively, then it increased to –0.81 at the current time point being –0.81. There was no difference in body weight among the 3 groups currently (Table 3, Fig. 2).

**3.5. Outcomes according to the operational methods in the distal ileum group**

Since all 3 surgical methods were performed only in the distal ileum group among the 3 groups, the outcomes according to the surgical methods in the distal ileum group were evaluated and compared. None of the demographic data showed differences

according to the surgical method (Table 4). Early and late complication also did not differ according to the surgical methods (Table 5).

When comparing the defecation frequency, there was a difference in the number of bowel movements at 3 months after the operation. But there was no difference in the number of bowel movements at the next follow-up and thereafter. Postoperative weight was not statistically different between groups at all time points. (Table 6).

**4. Discussion**

TCA is a rare form of HD, for which it is crucial to perform a pull-through operation by accurately identifying the involved segment through a preoperative contrast study and multiple intestinal biopsies under general anesthesia. Current advances in management, including early diagnosis, immune-staining, innovative surgical methods, and the provision of PN before and after surgery, have resulted in a better outcome compared with in the past.

The proportion of male patients was 72.7%. Although no difference in the male-to-female ratio was observed in some individual studies,<sup>[6,18]</sup> a review paper by McLaughlin et al<sup>[19]</sup> reported that the prevalence of male patients among 969 patients

**Table 2**  
Postoperative complications according to involved segments.

	Total	Jejunum	Jejunoleal junction	Distal ileum	P value
Early complications (<4 wk)	15 (45.5%)	2 (66.7%)	3 (60%)	10 (40%)	.530
Perianal excoriation	10	2	2	6	
Enterocolitis	3	0	1	2	
Wound complications	2	0	0	2	
Late complications (≥4 wk)	17 (51.5%)	1 (33.3%)	3 (60%)	13 (52%)	.762
Enterocolitis	8	1	2	5	
Ileus	3	0	0	3	
Septum adhesion	2	0	1	1	
Anorectal relaxation difficulty	2	0	0	2	
Re-pull-through	2	0	0	2	

**Table 3**  
**Postoperative frequency of defecation and peri-operative body weight change in the mean z-scores according to involved segments.**

	Total	Jejunum	Jejunoleal junction	Distal ileum	P value
Defecation frequency					
At 3-month follow-up	7.7	10.0	7.4	7.9	.126
At 6-month follow-up	5.5	7.0	5.4	5.6	.336
At 1-year follow-up	4.5	4.5	4.8	4.1	.890
At 2-year follow-up	3.2	4.5	3.3	2.8	.425
At 3-year follow-up	2.4	3.0	2.6	2.1	.527
At survey	2.3	2.7	2.5	2.0	.613
Body weight (z-score)					
At operation	-1.05	-1.39	-0.99	-1.09	.801
At 1-year follow-up	-1.37	-3.03	-1.08	-1.19	.230
At 2-year follow-up	-1.48	-2.46	-1.34	-1.17	.150
At 3-year follow-up	-1.42	-2.04	-1.47	-1.02	.325
At survey	-0.81	-0.98	-1.10	-0.56	.316

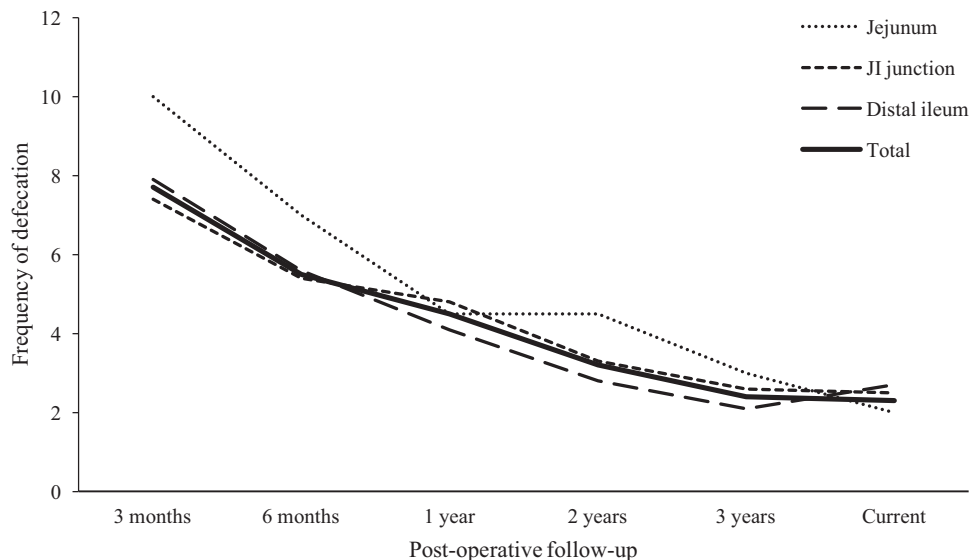
was 1.8 times higher than that of female patients, and our data showed the same pattern.

Of the patients' symptoms, 80% are known to be present in the neonatal period.<sup>[4]</sup> In our data, 69.7% of the patients received the diagnosis and underwent intestinal diversion within 1 month after birth, and the timing did not differ according to the involved segment. Other studies reported that diverting ostoma was performed within a median of 1 month after birth before the definitive pull-through operation<sup>[6,20]</sup>; however, no published study has reported differences according to the involved segment. In some studies, small-bowel perforation has been reported with total obstruction, but bowel perforation before ostoma formation was not observed in our data.<sup>[21]</sup>

Perianal excoriation was the most common postoperative complication in a number of published papers, with a rate of 10% to 77%.<sup>[6,18,21,22]</sup> In our data, perianal excoriation also had the highest frequency among the immediate complications. Our patients, like those in other reports, mostly showed improvement with supportive management. Postoperative enterocolitis, de-

finied as persistent diarrhea, weight loss, poor oral intake, and abdominal pain after the operation, was found in 12% to 76% in most studies.<sup>[6,10,21,23]</sup> In this report, enterocolitis was observed in 9.7% within 4 weeks and in 24.2% thereafter. In a study by Hukkinen et al,<sup>[23]</sup> there were no differences in the enterocolitis episodes among the 3 groups: distal ileum, mid-small bowel, and duodenojejunal flexure. Similarly, there was also no difference in the incidence of enterocolitis according to the length of the involved segment in our data.

In general, postoperative bowel function in HD has been reported to improve over time.<sup>[24]</sup> The same pattern has been reported for TCA, although the ages of patients showing normal bowel function were different among reports. Anupama et al<sup>[21]</sup> reported that 7 of 10 patients after 12 to 18 months of surgery had 1 to 3 bowel movements per day in a single-institution study that followed up 25 patients. In another study on 43 TCA patients with a median follow-up of 3.6 years, 23 of 31 patients had normal stool frequency.<sup>[25]</sup> A report with 58 patients followed up for a median of 12.5 years confirmed that normal



**Figure 1.** Postoperative frequency of defecation. JI = jejunoleal.

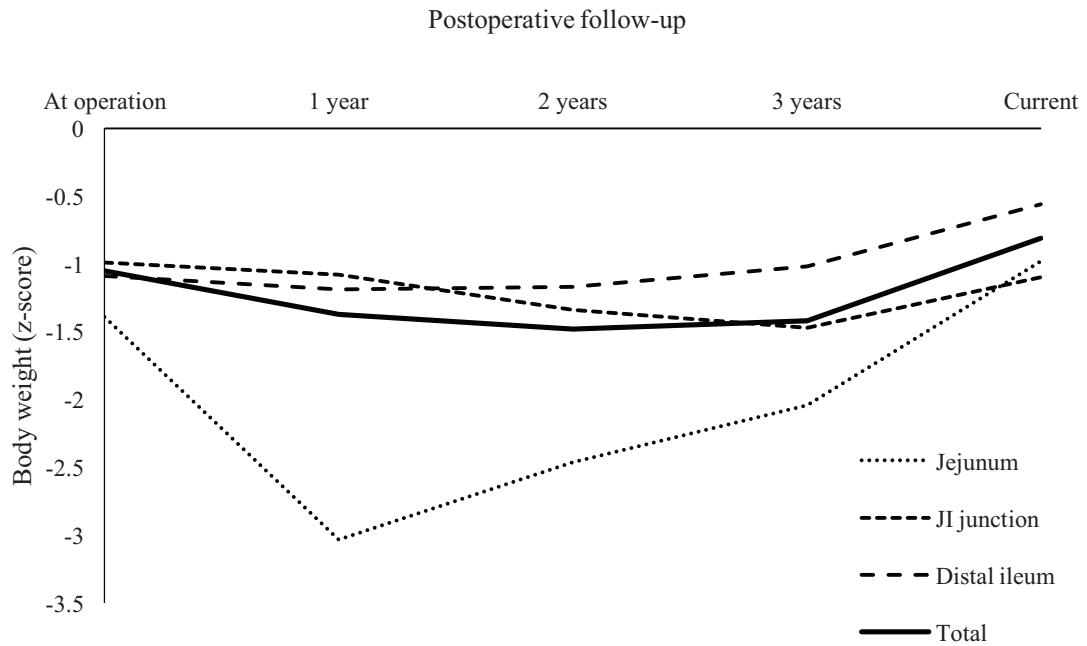


Figure 2. Postoperative body weight (z-score). JI = jejunioleal.

**Table 4**  
Demographics of the distal ileum group according to operational methods.

	Total	Duhamel	Martin	Soave	P value
Number of patients (%)	25	13 (52.0)	8 (32.0)	4 (16.0)	-
Male (%)	18 (72)	10 (76.9)	5 (62.5)	3 (75.0)	.244
Gestational age (wk)	39.4 (35.3–41.4)	39.6 (35.3–41.1)	39.1 (36.3–40.8)	39.2 (37.8–41.4)	.733
Body weight at birth (kg)	3.49 (2.52–4.31)	3.47 (2.88–3.90)	3.32 (2.52–4.20)	3.94 (3.53–4.31)	.051
Age at ostoma formation (mo)	1.0 (0.1–108.0)	1.0 (0.1–108.0)	0.5 (0.2–26.0)	0.6 (0.1–1.0)	.690
Age at pull-through (mo)	6.0 (4.3–114)	6.1 (4.3–114)	6.0 (4.5–30)	9.6 (7.0–15.1)	.743
Body weight at pull-through (kg)	8 (4.3–20.5)	8.1 (4.3–20.5)	7.6 (4.9–12.1)	8.0 (5.1–9.2)	.714
Follow-up (mo)	214 (21–411)	229 (21–354)	171 (43–261)	247 (119–411)	.178

bowel function was observed at age 7.6 years.<sup>[26]</sup> In this study, the defecation frequency immediately after surgery was 7.7 times daily and that at 3 years after surgery was 2.4 times daily, with no difference among the 3 groups. At the current telephone survey, the average number of daily defecations was 2.3, and there was no difference among the groups according to the length of the aganglionic segments. In a study by Hoehner et al<sup>[2]</sup> on the

satisfaction with functional outcome according to the length of aganglionic segments, patient satisfaction did not differ according to the length of the involved segment.

The nutritional status after surgery varied across different studies. In the study by Menezes et al,<sup>[26]</sup> body weight change was observed at a median follow-up of 12.5 years postoperatively, and failure to thrive (defined as <3%) was observed in <5% of

**Table 5**  
Postoperative complications in the distal ileum group according to operational methods.

	Total	Duhamel	Martin	Soave	P value
Early complications (<4 wk)	10 (40.0)	5 (38.5)	3 (37.5)	2 (50.0)	.905
Perianal excoriation	6	3	2	1	
Enterocolitis	2	1	-	1	
Wound complications	2	1	1	-	
Late complications (≥4 wk)	13 (52.0)	7 (53.8)	4 (50.0)	2 (50.0)	.982
Enterocolitis	5	2	2	1	
Ileus	3	2	-	1	
Septum adhesion	1	1	-	-	
Anorectal relaxation difficulty	2	-	2	-	
Re-pull-through	2	2	-	-	

**Table 6**  
**Postoperative frequency of defecation and body weight change in the mean z-scores in the distal ileum group according to operational methods.**

	Total	Duhamel	Martin	Soave	P value
Defecation frequency					
At 3-month follow-up	7.9	9.4	5.6	4.5	.021
At 6-month follow-up	5.6	5.9	4.8	2.8	.372
At 1-year follow-up	4.1	4.0	4.8	2.5	.243
At 2-year follow-up	2.8	2.5	3.5	2.0	.267
At 3-year follow-up	2.1	2.2	2.8	1.5	.338
At survey	2.0	1.7	1.9	1.5	.772
Body weight (Z-score)					
At operation	-1.09	-0.89	-1.26	-0.99	.892
At 1-year follow-up	-1.19	-0.53	-1.27	-1.03	.301
At 2-year follow-up	-1.17	-0.46	-0.97	-0.73	.468
At 3-year follow-up	-1.02	-0.91	-1.22	-0.72	.467
At survey	-0.56	-0.88	-1.30	-0.66	.497

all patients. In another report, 80% of patients had a body weight >50%,<sup>[21]</sup> and this was because even if the terminal ileum was surgically removed, the remaining small-bowel function is sufficient to maintain adequate absorption and growth.<sup>[10]</sup> However, in a small study with a follow-up of 9 years in 7 patients, 4 patients had a body weight <15%.<sup>[27]</sup>

In this study, the mean weight at the time of surgery was -1.05 when compared with the z-score. This was attributed to the difficulty in dietary progression due to defecation difficulty and abdominal distension before surgery. Moreover, after diverting ostoma formation, the patients did not recover well owing to fluid loss.

Body weight did not significantly increase immediately after surgery. The difference from the mean increased, but catch-up growth began to occur 1 to 2 years after surgery and it was found to recover to -0.81 through the telephone survey. These trends did not differ according to the length of the involved segment. The increase in body weight in this report is related to the reduced number of bowel movements and the steady diet with the start of school age. This is similar to the results of Hukkinen et al,<sup>[23]</sup> in which the majority of the patients' body weights were normal or near-normal, and no difference was observed in the subgroups, but none of the patients had a z-score > 0. In the same study, all patients with distal ileum involvement who required postoperative PN supply were weaned from PN after a median of 2.1 years postoperatively. Among patients with longer involved segments, 78% remained with PN support at the last follow-up.<sup>[23]</sup>

We wanted to see the difference according to the surgical method for each group, but the only group that was treated by all 3 surgical methods was the distal ileum group. There was no difference in complication according to the 3 surgical methods. The number of bowel movements was different immediately after surgery, but there was no difference over time. Weight change was not also different among surgical methods. This is because small bowel is almost preserved at the distal ileum group, so it was not a short-bowel status and it may not show differences according to the surgical methods.

This was similar to the results of Barrena study, and there was no difference in complication and growth when comparing between Martin modification and straight endorectal pull-through (Soave and Swenson) in TCA patients with limited ileal involvement.<sup>[10]</sup> Since there are not many other studies, if there

are more patients, the difference according to each group's surgical method can be checked, and it can be confirmed through studies such as nationwide survey.

The limitation of this study lies in its retrospective nature with a small number of patients (33 in total and only 3 in the jejunum group). Long-term clinical evaluation of each involved segment in TCA is required through prospective cohort or multicenter studies in the future.

In conclusion, the transition zone in most TCA patients was in the distal ileum and the total rate of postoperative complications was 45% to 51%. The postoperative defecation frequency normalized after 3 years from the operation, and body weight started to recover after 2 years without differences according to the length of the involved segment. Therefore, close monitoring of defecation frequency and body weight for 2 to 3 years after surgery and active evaluation with PN support for adequate growth and nutrition are required.

### Author contributions

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**Writing – review & editing:** Joong Kee Youn, Hyun-Young Kim, Dayoung Ko, Kwi-Won Park.

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