# RESEARCH



# Surgical outcomes for patients with rectosigmoid hirschsprung disease who underwent transanal endorectal pull-through after 1 year of age

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# Abstract

**Purpose** This study aimed to compare the differences in postoperative complications and long-term bowel function outcomes between patients with rectosigmoid Hirschsprung disease (HD) who underwent transanal endorectal pull-through (TEPT) beyond infancy (age> 1 year of age) and those during infancy (≤ 1 year of age).

**Methods** All patients with rectosigmoid HD at Beijing Children's Hospital between January 2011 and December 2020 were analyzed retrospectively. They were divided into two groups based on age at TEPT: group A was defined as patients who performed TEPT beyond infancy (age>1 year of age), and group B as patients who performed TEPT during infancy (age  $\leq$  1 year of age). Clinical details were collected from medical records. Bowel function outcomes were assessed by the Rintala questionnaire (age  $\geq$  4 years).

**Results** A total of 339 patients were included: 216 (63.7%) who operated with TEPT beyond infancy (group A) and 123 (36.3%) during infancy (group B). Regarding postoperative complications, all patients suffering anastomosis leakage following TEPT (7/216, 3.2%) occurred in group A, and the rate of anastomosis leakage in group A was significantly higher than in group B (3.2% vs. 0.0%, p = 0.044). 228 patients (228/327, 69.7%) completed the Rintala questionnaire. There was no significant difference in long-term bowel function outcomes between the two groups.

**Conclusion** Compared with patients who performed TEPT during infancy, those beyond infancy are more likely to suffer anastomosis leakage. **however**, **the long-term bowel function outcomes seem comparable.** 

Type of Study A retrospective single-center study.

Level of evidence III.

**Keywords** Hirschsprung disease, Transanal endorectal pull-through, Infancy, Complications, Bowel function outcomes

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# Introduction

Hirschsprung disease (HD) is characterized by the lack of ganglion in the submucosal and myenteric neural plexuses, requiring resection of the aganglionic segment as treatment [1]. With the heightened awareness of the disease and improvement in earlier diagnosis techniques, 80-90% of HD patients were diagnosed in the neonatal period in developed countries [2]. A survey of pediatric surgeons in Europe revealed that 87.5% of HD patients performed transanal endorectal pull-through (TEPT) before three months of age [3]. However, in developing countries, the majority of HD patients presented late to tertiary children's hospitals due to the poor awareness of HD in primary hospitals and the general public [4, 5]. Unfortunately, the available literature on the surgical outcomes of patients who operated with TEPT beyond infancy is sparse, at best, and consists mainly of case reports [4, 6]. Ekenze et al. [4] suggested that the majority of HD patients after 1 year of age would require colostomy before radical surgery to reduce the incidence of postoperative complications. However, Samir et al. [7] revealed that one-stage TEPT was feasible and safe for adolescents and adults, which could avoid the morbidity of a stoma. Due to this controversy and lack of understanding of the treatment and prognosis of TEPT in HD patients after 1 year of age, we conducted a retrospective study with the largest sample size in China to investigate the clinical features of HD patients beyond infancy and compare the difference in postoperative complications and long-term bowel function outcomes between HD patients beyond infancy and those during infancy. Given the limited research available in this area, this study is essential to provide insights into the treatment and prognosis for patients with rectosigmoid HD beyond infancy following TEPT.

# **2. Materials and methods** Ethical approval

This study adheres to the ethical principles of the Declaration of Helsinki. It was approved by the Ethics Committee of Beijing Children's Hospital (**2021-E-132-R**). Written informed consent was obtained from the patient's parents.

# Patient selection

Approved by the Ethics Committee of Beijing Children's Hospital, we reviewed the medical records of 339 consecutive patients with rectosigmoid HD who underwent TEPT at Beijing Children's Hospital, National Center for Children's Health, between January 2011 and December 2020. Patients who underwent radical surgery in other hospitals or patients without histopathological confirmation of HD were excluded. **Patients diagnosed with Down syndrome or other identified genetic syndromes**  **were excluded.** All patients enrolled in our study underwent Soave pull-through by the same surgeon team.

# Study design

The patient characteristics and clinical details were recorded retrospectively from medical records, including sex, gestational age, birth weight, congenital malformation, history of ostomy, age at TEPT, weight at TEPT, preoperative enterocolitis, surgical approach, and postoperative complications (anastomosis leakage, postoperative enterocolitis, and residual transitional zone). The choice of surgical approaches mainly depended on the extent of the lesion bowel and anastomotic tension. If the scope of the lesion bowel did not exceed the splenic flexure, we usually gave priority to performing total transanal endorectal pull-through (TERPT). However, if preoperative angiography indicated the lesion bowel involved more than the splenic flexure or intraoperative mesenteric tension was too high, we often chose laparoscopic or laparotomy-assisted colonic mobilization (LERPT). All patients with rectosigmoid HD enrolled in our study were divided into two groups according to their age at TEPT: group A was defined as patients who performed TEPT beyond infancy (age>1 year of age), and group B as patients who performed TEPT during infancy (age  $\leq 1$ year of age) [4]. A comparative study was conducted to analyze the difference in postoperative complications and long-term bowel function outcomes between the two groups.

Bowel function outcomes were assessed by a bowel function score (BFS) questionnaire established by the 7-item scoring system with a maximum score of 20 [8]. Patients' parents were systematically followed up by telephone and filled out the questionnaires. The BFS was only evaluated when patients were not less than 4 years old. Based on the total score of BFS, we divided them into three categories:  $\geq 17$  indicating good bowel function; 12-16 indicating moderate bowel function; <12 indicating poor bowel function. Soiling refers to fecal staining of underwear, fecal accidents to involuntary loss of feces needing a change of underwear or use of protective aids, and constipation is defined as the need for a special diet, laxatives, or enemas [8]. Enterocolitis was diagnosed when patients presented the typical clinical signs of bowel inflammation, such as abdominal distension, diarrhea, fever, or lethargy [9].

## Statistical analysis

Statistical analysis was conducted using IBM SPSS Statistics for Statistics ver. 26.0 Software. Data were presented as frequency (percentage) for qualitative variables and median and interquartile range (IQR) for continuous variables. All the statistical tests were two-sided, with a significant level of p <0.05. Continuous Chi-squared tests

## Table 1 Patient characteristics

Characteristics	Group A ( <i>n</i> =216)	Group B ( <i>n</i> = 123)	<i>p</i> -Value
Sex, n (%)			
Male	177 (81.9)	96 (78.0)	0.384
Female	39 (18.1)	27 (22.0)	
Gestational age			
Preterm (<37 weeks)	7 (3.2)	5 (4.1)	0.693
Term (≥ 37 weeks)	209 (96.8)	118 (95.9)	
Birthweight, kilograms	3.4 [3.0, 3.7]	3.4 [3.1, 3.8]	0.364
Congenital malformation, n (%)	12 (5.6)	7 (5.7)	0.958
History of ostomy, <i>n</i> (%)	4 (1.9)	4 (3.3)	0.414
Age at radical surgery, month	36.0 [19.0, 63.0]	7.0 [4.9, 9.0]	< 0.001
Weight at radical surgery, kilogram	14.0 [11.0, 18.0]	7.5 [6.5, 9.0]	< 0.001
Surgical approach, <i>n</i> (%)			
Transanal only	177 (81.9)	110 (89.4)	0.156
Open surgery + transanal	20 (9.3)	8 (6.5)	
Laparoscopic + transanal	19 (8.8)	5 (4.1)	
Preoperative enterocolitis, n (%)	25 (11.6)	14 (11.4)	>0.999
Data are precented as median [IOP interguartile	rangel and frequency (%)		

Data are presented as median [IQR, interquartile range] and frequency (%)

#### Table 2 Postoperative complications

	Group A	Group B	p-
	( <i>n</i> =216)	( <i>n</i> = 123)	Value
Mean follow-up time, years	7.4 [5.1, 9.7]	7.7 [5.8, 9.8]	0.261
Anastomosis leakage, n (%)	7 (3.2)	0 (0.0)	0.044
Postoperative enterocolitis, n (%)	20 (9.3)	9 (7.3)	0.539
Residual transitional zone, n (%)	2 (0.9)	3 (2.4)	0.357
Data are presented as median [IOR inte	rausrtile range	and from one	

Data are presented as median [IQR, interquartile range] and frequency (%)

or Fisher's exact tests (Fisher's exact if >25% of cells have expected counts less than 5)were applied for categorical variables. Independent sample t-test or Mann-Whitney U test (Mann-Whitney if the data did not meet normal distribution) for continuous variables.

# Results

# **Patient characteristics**

A total of 339 patients were included in our study, including 216 patients in group A and 123 patients in group B. The baseline characteristics of all patients are presented in Table 1. Except for age at TEPT and weight at TEPT, there was no significant difference between the two groups in terms of patient demographics (age at TEPT, 36.0 [19.0. 63.0] vs. 7.0 [4.9, 9.0] months, p < 0.001; age at weight, 14.0 [11.0, 18.0] vs. 7.5 [6.5, 9.0] kilograms, p<0.001). Eighteen patients (18/339, 5.3%) had congenital malformation: rectal perineal fistula (10/339, 2.9%), congenital heart malformation (5/339, 1.5%), spina bifida (3/339, 0.9%).

# Postoperative complications

Table 2 shows postoperative complications between the two groups. All patients were followed up for more than two years with a median follow-up time of 7.5 [5.5, 9.7] years, and there was no significant difference in the median follow-up time between the two groups (7.4 [5.1, 9.7] vs. 7.7 [5.8, 9.8] years, p=0.261). Postoperative complications included anastomotic leakage, postoperative enterocolitis, and residual transitional zone. All patients who suffered anastomosis leakage following TEPT (7/216, 3.2%) occurred in group A, and the rate of anastomosis leakage in group A was significantly higher than in group B (3.2% vs. 0.0%, p=0.044). No significant difference in the two groups was observed regarding postoperative enterocolitis and residual transitional zone (9.3% vs. 7.3%, p=0.539; 0.9% vs. 2.4%, p=0.357, respectively).

# Long-term bowel function outcomes

Twelve patients were excluded due to the follow-up age of less than 4 years old, and ninety-nine patients dropped out. Therefore, a total of 228 HD patients (median age 9.4 [6.8, 11.3] years) were included in the study of long-term bowel function outcomes with a response rate of 69.7% (228/327). A dropped-out analysis revealed the baseline characteristics were comparable between respondents and nonrespondents (Table 3). The results of bowel function outcomes were presented in Table 4. There was no statistical difference between the two groups regarding the median follow-up interval (6.8 [4.7, 9.4] vs. 7.5 [5.6, 9.8] years, p=0.238), while the median follow-up age in group A was significantly higher than group B (10.0 [8.1, 12.4] vs. 7.9 [6.0, 9.9] years, p<0.001). No significant difference between the two groups was observed in terms of the ability to hold back defecation (p=0.337), feeling to urge to defecate (p=0.338), frequency of defecation (p=0.526), soiling (p=0.886), fecal accidents (p=0.754), constipation (p=0.422), and social problems (p=0.409). As shown in Fig. 1, the median total bowel function score

Tabl	e 3	Baseline c	haracteristics of	<sup>-</sup> respond	lents anc	l nonrespond	lents
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Characteristics	Respondents (n = 228)	Nonrespondents (n = 99)	<i>p</i> -Value	
Sex, n (%)				
Male	181 (79.4)	80 (80.8)	0.881	
Female	47 (20.6)	19 (19.2)		
Gestational age, n (%)				
Preterm (<37 weeks)	9 (3.9)	2 (2.0)	0.515	
Term (≥ 37 weeks)	219 (96.1)	97 (98.0)		
Birthweight, kilograms	3.4 [3.1, 3.8]	3.4 [3.0, 3.8]	0.842	
Congenital malformation, n (%)	14 (6.0)	5 (4.7)	0.801	
History of ostomy, <i>n</i> (%)	8 (3.5)	0 (0.0)	0.112	
Age at radical surgery, month	20.0 [8.6,46.0]	16.0 [8.0, 43.4]	0.462	
Age at radical surgery, <i>n</i> (%)			0.382	
>1 year old	150 (65.8)	60 (60.6)		
≤1 year old	78 (34.2)	39 (39.4)		
Weight at radical surgery, kilogram	11.0 [8.0, 15.0]	10.5 [8.0, 15.0]	0.591	
Surgical approach, n (%)				
Transanal only	191 (83.8)	90 (90.9)	0.227	
Open surgery + transanal	22 (9.6)	6 (6.1)		
Laparoscopic + transanal	15 (6.6)	3 (3.0)		
Re-operation, n (%)	7 (3.1)	4 (4.0)	0.740	

Data are presented as median [IQR, interquartile range] and frequency (%), and 12 patients were excluded due to the follow-up age less than 4 year old

in group A was similar to group B (19 [16, 20] vs. 18 [14, 20], *p*=0.308).

# Discussion

This study represents the first attempt to compare the surgical outcomes between rectosigmoid HD patients who underwent TEPT beyond infancy and those who underwent the procedure during infancy. The key finding from this study is that patients who underwent TEPT beyond infancy are more susceptible to anastomosis leakage compared to those who had the procedure during infancy, however, the long-term bowel function outcomes seem comparable.

Based on the findings of our study, there is a significant difference in the age at TEPT compared to previous literature [10]. Our study found that over 60% of HD patients operated with TEPT beyond infancy, which is later than what has been reported in previous literature where more than 90% of patients received TEPT during infancy [4, 10]. This disparity can be attributed to two reasons. Firstly, HD patients included in our study aganglionosis limited to the rectosigmoid. As a result, many of them did not exhibit severe clinical symptoms and could be treated conservatively with glycerine enema at local hospitals [11]. Most of them would not be transferred to our center for radical surgery until presenting more severe symptoms [11]. This may also explain why the incidence of preoperative enterocolitis was relatively lower in our study than in most previous literature [12, 13]. Secondly, the delayed referral of HD patients from primary or secondary healthcare facilities to tertiary health facilities is responsible for the later age at radical surgery. This could be due to a lack of recognition of rectosigmoid HD by physicians from primary or secondary healthcare facilities [4, 5]. To address this issue, it is recommended that enlightenment programs be conducted for primary care physicians and public health education to shorten the diagnosis time of HD, enabling them to receive prompt and standardized treatment [4].

Regarding postoperative complications, the most striking difference between group A and group B was the rate of anastomosis leakage. All patients (7/216, 3.2%) who suffered anastomosis leakage occurred in patients operated TEPT beyond infancy. A plausible explanation for a higher anastomosis leakage rate in the older age group might be related to the problems of a severely dilated colon [6, 11, 14, 15]. Especially for patients with rectosigmoid HD, it could significantly contribute to the difficulty in dissection above the dentate line when performing TEPT [11]. Furthermore, older patients who performed TEPT tend to have a deeper pelvis and more abundant mesenteric blood vessels, making the **dissection more difficult.** These factors might eventually result in a hemodynamic disorder of anastomosis, leading to a high incidence of anastomosis leakage [6, 15]. To reduce the incidence of anastomosis leakage, some scholars recommend that HD patients beyond infancy require a stoma before radical surgery to decompress the distal colon [4, 6, 16]. However, specific stoma has many complications, such as prolapse, stenosis, and wound infection [17]. The requirement of multiple admissions and operations also placed a heavy burden on families and

#### Table 4 Bowel function score (≥4 years old)

Evaluation of bowel control	Score	Group A ( <i>N</i> =210)	Group B (N=117)	<i>p</i> -Value
Patient answering the Rintala questionnaire		N=150 (71.4%)	N=78 (66.7%)	
Median follow-up interval, years		6.8 [4.7, 9.4]	7.5 [5.6, 9.8]	0.238
Median follow-up age, years		10.0 [8.1, 12.4]	7.9 [6.0, 9.9]	< 0.001
Ability to hold back defecation, n (%)				
Always	3	112 (74.7)	50 (64.1)	0.337
Problems less than 1/week	2	17 (11.3)	10 (12.8)	
Weekly problems	1	12 (8.0)	11 (14.1)	
No voluntary control	0	9 (5.9)	7 (9.0)	
Feels/reports the urge to defecate, <i>n</i> (%)				
Always	3	108 (72.0)	47 (60.3)	0.338
Most of the time	2	26 (17.3)	20 (25.6)	
Uncertain	1	11 (7.3)	7 (9.0)	
Absent	0	5 (3.3)	4 (5.1)	
Frequency of defecation, n (%)				
Every other day to twice a day	2	114 (76.0)	55 (71.4)	0.526
More than	1	17 (11.3)	13 (16.9)	
Less than	1	19 (12.7)	9 (11.7)	
Soiling, n (%)				
Never	3	67 (44.7)	31 (39.7)	0.886
Staining<1/week, no change of underwear required	2	32 (21.3)	17 (21.8)	
Frequent staining, change of underwear often required	1	36 (24.0)	22 (28.2)	
Daily soiling, requires protective aids	0	15 (10.0)	8 (10.3)	
Fecal accidents, n (%)				
Never	3	116 (77.3)	59 (75.6)	0.754
Fewer 1/week	2	13 (8.7)	10 (12.8)	
Weekly, requires protective aids	1	15 (10.0)	7 (9.0)	
Daily, requires protective aid day and night	0	6 (4.0)	2 (2.6)	
Constipation, n (%)				
No constipation	3	133 (88.7)	64 (82.1)	0.422
Manageable with diet	2	11 (7.3)	9 (11.5)	
Manageable with laxatives	1	6 (4.0)	5 (6.4)	
Manageable with enemas	0	0 (0.0)	0 (0.0)	
Social problems, n (%)				
No social problems	3	125 (83.3)	67 (85.9)	0.409
Sometimes	2	17 (11.3)	7 (9.0)	
Problems restricting social life	1	6 (4.0)	1 (1.3)	
Severe social/psychosocial problems	0	2 (1.3)	3 (3.8)	
Total BFS, n (%)				
Good bowel function	≥17	105 (70.0)	47 (60.3)	0.311
Moderate bowel function	12–16	29 (19.3)	21 (26.9)	
Poor bowel function	<12	16 (10.7)	10 (12.8)	

Data are presented as median [IQR, interquartile range] and frequency (%)0.12 HD patients were excluded due to the follow-up age of less than 4 years old, and 99 HD patients dropped out

the medical system [7, 17]. Therefore, the desire to avoid stroma creation and adopt one-stage TEPT was established. In our study, all the patients beyond infancy performed one-stage TEPT when the general condition was permitted. However, the incidence of postoperative anastomosis leakage in the older age group was much lower than in previous literature [4, 6]. The possible reason might be as follows. On the one hand, we routinely perform careful bowel preparation before radical surgery to reduce the tension of the distal colon [7, 18]. The duration of bowel preparation depended on the degree of fecal impaction and the effectiveness of bowel irrigation. Most patients routinely underwent bowel irrigations for 1–2 weeks before radical surgery. However, some HD patients who performed TEPT beyond infancy might have longer bowel preparation due to fecal impaction and severe bowel dilation. On the other hand, we are prone to remove all the lesion bowel





**Fig. 1** Bowel function scores (BFS) between group **A** (n = 150) and group **B** (n = 78)

(including spasm segment, transitional and proximal dilated zones) to make the proximal pull-through bowel normal in terms of pathology and morphology, leading to a good coloanal anastomosis [19]. Based on our experience, despite of higher incidence of postoperative anastomotic leakage in HD patients beyond infancy, the one-stage operation is feasible for them if patients are in good condition [7]. However, it was still recommended that radical surgery should be performed during infancy to reduce the incidence of postoperative complications [6].

Our study found that long-term bowel function outcomes of HD patients who performed TEPT beyond infancy were comparable to those who underwent the procedure during infancy. Considering the discrepancy in the follow-up age, further research needs to confirm it. In addition, the long-term bowel function outcomes were also affected by other factors, such as the level of anastomosis above the dentate line, motility of the remnant ganglionic colon, and the integrity of the anal sphincter [20, 21]. Scholars generally believed that damage to the anal sphincter and too low anastomosis could lead to fecal incontinence [8, 22]. In our center, the TEPT procedure started rectal mucosectomy from 0.5 to 1 cm proximal to the dentate line, ensuring the integrity of the dentate line [23]. The study has some limitations. Firstly, despite the comparable baseline characteristics of respondents and nonrespondents, it remains unknown whether the postoperative bowel function differed in those who responded and those who did not, which might result in potential selection bias. Secondly, the median follow-up age in group A was significantly higher than in group B, which could result in certain deviations in long-term bowel function outcomes. Thirdly, our study is retrospective and single-center, leading to a particular deviation.

To conclude, the study found that rectosigmoid HD patients who underwent TEPT beyond are more likely to suffer anastomosis leakage. However, the long-term bowel function outcomes seem comparable and further investigation is required.

#### Supplementary Information

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Supplementary Material 1

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#### Author contributions

All authors read and confirmed the final manuscript. Chuanping Xie, Jiayu Yan, Kexin Wang participated in the data collection, data analysis, and drafting of the article; Chuanping Xie and Kexin Wang contributed to the data collection; Jiayu Yan, Wenbo Pang, Dan Zhang, and Kai Wang contributed to revising the article. Yajun Chen was the major surgeon who conducted the surgery and contributed to reviewing and drafting the article.

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#### Data availability

No datasets were generated or analysed during the current study.

#### Declarations

#### **Consent for publication**

Not applicable.

#### **Competing interests**

The authors declare no competing interests.

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