Pediatr Gastroenterol Hepatol Nutr. 2024 Nov;27(6):372-382
https://doi.org/10.5223/pghn.2024.27.6.372
pISSN 2234-8646·eISSN 2234-8840

Pediatric Gastroenterology, Hepatology & Nutrition
pghn

Original Article

Check for updates

Prevalence and Factors associated with Bowel Dysfunctions after Pull-Through Surgery in Children Diagnosed with Hirschsprung Disease

Maliwan Surasen,[1] **Palittiya Sintusek**,[1,2] **Nimmita Srisan**,[2,3] **Katawaetee Decharun**,[2,3] **and Paisarn Vejchapipat** [3]

[1]Department of Pediatrics, King Chulalongkorn Memorial Hospital, The Thai Red Cross Society, Faculty of Medicine, Chulalongkorn University, Bangkok, Thailand
[2]Center of Excellence in Thai Pediatric Gastroenterology, Hepatology and Immunology, Division of Gastroenterology, Department of Pediatrics, King Chulalongkorn Memorial Hospital, The Thai Red Cross Society, Faculty of Medicine, Chulalongkorn University, Bangkok, Thailand
[3]Department of Surgery, King Chulalongkorn Memorial Hospital, The Thai Red Cross Society, Faculty of Medicine, Chulalongkorn University, Bangkok, Thailand

OPEN ACCESS

Received: Mar 7, 2024
Revised: Jun 18, 2024
Accepted: Jul 12, 2024
Published online: Nov 5, 2024

Correspondence to
Palittiya Sintusek
Center of Excellence in Thai Pediatric Gastroenterology, Hepatology and Immunology, Division of Gastroenterology, Department of Pediatrics, King Chulalongkorn Memorial Hospital, The Thai Red Cross Society, Faculty of Medicine, Chulalongkorn University, Bangkok 10330, Thailand.
Email: Palittiya.s@chula.ac.th

ORCID iDs
Maliwan Surasen
https://orcid.org/0009-0003-8699-8368
Palittiya Sintusek
https://orcid.org/0000-0003-4441-0151
Nimmita Srisan
https://orcid.org/0000-0002-0722-3309
Katawaetee Decharun
https://orcid.org/0000-0003-4449-0323
Paisarn Vejchapipat
https://orcid.org/0000-0002-4388-6900

ABSTRACT

Purpose: This study investigated the prevalence of bowel dysfunction and associated factors after pull-through surgery.
Methods: The medical records of children under 18 years old diagnosed with Hirschsprung disease (HD) based on histopathology between 2004 and 2022 were reviewed. Bowel dysfunction after pull-through surgery was categorized into Hirschsprung-associated enterocolitis (HAEC), constipation, and fecal incontinence.
Results: Among 97 children diagnosed with HD, the median age at presentation was 3 (2-15) days (84.54% male). The clinical manifestations included abdominal distension (58.76%), constipation (17.52%), bilious vomiting (17.52%), nonbilious vomiting (14.43%), and enterocolitis (12.37%). HDs were classified by the location of aganglionosis: short segments (74.23%), long segments (8.25%), total colonic (12.37%), and small intestinal (5.15%). Excluding surgical complications, the prevalence of bowel dysfunction was 64.95% during an average follow-up of 8.33 years. HAEC was the most common issue (46.39%), followed by nonretentive incontinence (22.68%), constipation (20.62%), and retentive incontinence (15.46%). Preoperative HAEC was significantly associated with post-surgery HAEC (adjusted odds ratio [aOR] 18.31; 95% confidence interval [CI], 1.30-257.73; p=0.031). The Duhamel operation was associated with constipation and retentive incontinence (aOR 62.15; 95% CI, 1.64-2,349.13; p=0.026). Age under 6 months at pull-through surgery was associated with nonretentive fecal incontinence after 4 years (aOR 8.83; 95% CI, 1.11-70.39; p=0.040).
Conclusion: The prevalence of bowel dysfunction in children with HD remains high despite successful surgical correction. Preoperative HAEC, Duhamel operation, and pull-through surgery before the age of 6 months were found to be independent factors associated with bowel dysfunction after pull-through surgery.

Keywords: Children; Megacolon; Fecal soiling; Constipation; Hirschsprung disease; Enterocolitis

Funding

This study was supported by the Center of Excellence in Thai Pediatric Gastroenterology, Hepatology and Immunology, Faculty of Medicine, Chulalongkorn University, Thailand. This study was supported by Thailand Science Research and Innovation Fund Chulalongkorn University (No. HEA663000047).

Conflict of Interest

The authors have no financial conflicts of interest.

INTRODUCTION

Hirschsprung disease (HD) is a congenital disorder that affects the enteric nervous system of the bowel and occurs in approximately one in 5,000 live births [1]. The underlying pathology involves the disrupted migration of neural crest cells, resulting in the absence of ganglion cells in the submucosal and myenteric plexuses [2]. HD is typically diagnosed during the neonatal period, and the pull-through operation is a standard and widely used treatment. However, the choice of operation may depend on anatomical issues and surgeon preference. In general, a staged approach is the standard surgical method for long-segment or total colonic HD, while a primary pull-through can be performed with short-segment HD. To date, there has been insufficient evidence to support the recommendation of surgical repair for HD [2].

After the primary pull-through operation or a staged procedure with a definitive pull-through operation, the prognosis of surgically treated HD is generally favorable, with most patients achieving near-normal fecal defecation [3]. However, there are potential long-term issues that may arise after surgical correction with respect to surgical complications or bowel dysfunction, including constipation, recurrent enterocolitis, stricture, stromal prolapse, perianal infection or abscess, and fecal incontinence [4-8].

The main objective of this study was to examine the prevalence of bowel dysfunction following surgical treatment in children diagnosed with HD. Additionally, factors associated with bowel dysfunction after pull-through surgery were assessed. By studying these factors, we can gain a better understanding of the potential underlying therapeutic challenges and determine strategies to improve long-term outcomes in patients with HD.

MATERIALS AND METHODS

Study design and participants

This retrospective descriptive study included patients who met the following inclusion criteria: (1) aged <18 years, (2) had HD definitively diagnosed using histopathological results of the aganglionic bowel segment, and (3) received medical care at King Chulalongkorn Memorial Hospital, a tertiary referral hospital in Bangkok, Thailand, between 2004 and 2022. The exclusion criteria included (1) children with HD who had a delayed stoma at the time of data extraction; (2) children with uncertain diagnoses or incomplete or missing data from the medical records; (3) children lost to follow-up; and (4) children diagnosed with ultrashort HD. Patient chart records were extensively reviewed for demographic data, clinical presentation, investigations, histopathology, type of surgical procedure, other management, and long-term follow-up outcomes. Because this study relied on an analysis of medical records, obtaining patient consent was not feasible. Approval was obtained from the hospital director for this study. Ethical approval was obtained from the Institutional Review Board of the Faculty of Medicine at Chulalongkorn University (IRB no. 183/65).

Definition

Bowel dysfunction is defined as a complication of gastrointestinal function after complete surgical correction or bowel anastomosis for HD and is categorized into four aspects as follows: 1) Nonretentive fecal incontinence is defined as the uncontrolled loss of feces in settings that are socially inappropriate, and it occurs without evidence of fecal retention. 2) Retentive fecal incontinence is defined as the uncontrolled loss of feces in settings that

are socially inappropriate, with evidence of fecal retention. It is observed in toilet-trained children or those aged >4 years. 3) Constipation is defined by the Rome IV criteria as <3 defecations per week, straining, or large or hard stools [9], in which anastomosis stricture was excluded. 4) Hirschsprung-associated enterocolitis (HAEC) is diagnosed based on the "HAEC score" by A.C. Pastor et al. [10]. The HAEC score comprises a history of diarrhea and enterocolitis, physical examination, radiologic examination and laboratory results. HAEC was diagnosed if the HAEC score was ≥4 [11].

According to the American Pediatric Surgical Association [12], the types of HD were categorized based on the extent of aganglionosis confirmed through histopathological study as follows: (1) short-segment HD: aganglionosis extending up to the sigmoid colon-descending colon junction; (2) long-segment HD: aganglionosis extending from the sigmoid colon to the descending colon but with ganglion cells present in some portion of the colon; (3) total colonic HD: aganglionosis of the entire colon and <5 cm of terminal ileum; and (4) small intestinal HD: aganglionosis extending proximal to >5 cm of the terminal ileum.

Statistical analysis

The collected data were analyzed using STATA version 15 (StataCorp). Continuous and categorical data are reported as medians (interquartile ranges, IQRs) and percentages or proportions, respectively. The relationship between patient characteristics, treatment, and bowel dysfunction was assessed using Pearson's chi-square test or Fisher's exact test, as appropriate. Univariate and multivariate logistic regression analyses were also conducted to identify factors associated with bowel dysfunction. The results are expressed as adjusted odds ratios (ORs) with corresponding 95% confidence intervals (CIs). A p-value <0.05 indicated statistical significance.

RESULTS

Study populations

Between 2004 and 2022, 142 children with HDs sought medical care at King Chulalongkorn Memorial Hospital. After the exclusion of patients who had incomplete medical records or were lost to follow-up (n=32), had delayed stroma closure (n=5), were diagnosed with ultrashort HD (n=4) or died from congenital heart disease (n=3) or sepsis (n=1), 97 children were included in the data analysis. Of these, the majority (74.23%) were referred from other hospitals in Thailand, and 92.78% were Thai. The median age at presentation was 3 (2–15) days, and 84.54% of the patients were male. The median follow-up time was 8.33 (4.24–12.36) years. Thirteen patients had syndromic anomalies. The syndromes identified in these patients were Down syndrome (n=11), Waardenburg syndrome (n=1), and DiGeorge syndrome (n=1) (**Table 1**).

Clinical presentations, investigations, and treatments

The predominant clinical manifestation of HD was abdominal distension (58.76%), followed by constipation (17.52%), bilious vomiting (17.52%), nonbilious vomiting (14.43%), and HAEC (12.37%). Among the 49 children with available data on meconium passage, 33 (67.35%) exhibited delayed passage beyond 24 hours of age. According to the diagnostic investigations, among the 83 children with relevant data, the majority (96.4%) underwent barium enema, while anorectal manometry (ARM) was performed in 4.7% of the patients. Among the anatomical aganglionic segments of HD, 74.23% were short segments, 8.25%

Pediatric Gastroenterology, Hepatology & Nutrition pghn

Table 1. Demographic data, baseline characteristics, clinical presentations, treatments, and disease outcomes of the participants (n=97)

Demographic data	Number (%)
Sex, male	82 (84.54)
Age at the end of follow-up (yr)	9.78 (4.82–13.35)
Preterm (≤37 wk) (n=89)	14 (14.43)
Nationality: Thai	90 (92.78)
Referral cases	72 (74.23)
Family history of Hirschsprung disease	5 (5.15)
Associated anomalies	30 (30.93)
Syndromic	13
Down syndrome	11
Waardenburg syndrome	1
DiGeorge syndrome	1
Congenital heart disease	17
Patient characteristics and clinical presentations	
Age at presentation (d)	3 (2–15)
Clinical presentations	
Abdominal distension	57 (58.76)
Nonbilious vomiting	14 (14.43)
Bilious vomiting	17 (17.52)
Constipation	17 (17.52)
Hirschsprung associated enterocolitis	12 (12.37)
Delayed passage of meconium (>24 hr) (n=49)	33/49 (67.35)
Anatomical classification of Hirschsprung disease	
Short segment	72 (74.23)
Long segment	8 (8.25)
Total colonic segment	12 (12.37)
Small bowel Hirschsprung disease	5 (5.15)
Treatment and disease outcome	
Age at the pull-through operation (d)	97 (35–304)
Age at the pull-through operation <6 months	88 (90.72)
One-stage pull-through operation	77 (79.38)
The pull-through techniques	
Swenson	56 (57.73)
Yancey-Soave	9 (9.28)
Duhamel	4 (4.12)
Not otherwise specified	28 (28.87)
Follow-up periods (yr)	8.33 (4.24–12.36)
Complications after surgery until the end of follow-up	
Bowel dysfunctions	63 (64.95)
Non retentive incontinence	22 (22.68)
Retentive incontinence	15 (15.46)
Constipation	20 (20.62)
Hirschsprung associated enterocolitis	45 (46.39)
<2 weeks after pull-through surgery	8
>2 weeks after pull-through surgery	43
Other complications	
Retained aganglionic segment	9 (9.28)
Anastomosis stricture	12 (12.37)
Perianal infection	8 (8.25)

Data are presented as the number (%) or median (interquartile range).

were long segments, 12.37% were total colonic segments and 5.15% were small bowel HD. Surgical correction was performed in 79.38% of the patients who underwent one-stage of pull-through surgery, while an ostomy was chosen for the remaining patients, followed by pull-through surgery. The operative pull-through techniques performed were Swenson (57.73%), Yancey-Soave (9.28%), Duhamel (4.12%) and not otherwise specified (28.87%) (**Table 1**).

https://doi.org/10.5223/pghn.2024.27.6.372

Pediatric Gastroenterology, Hepatology & Nutrition pghn

Prevalence of bowel dysfunction in HD patients after pull-through surgery

In total, 64.95% of the children experienced bowel dysfunction. The most prevalent diagnosis was HAEC, which was observed in 46.39% of the patients. Constipation was identified in 20.62% of the children, while nonretentive incontinence and retentive incontinence were both present in 22.68% and 15.46%, respectively (**Table 1**).

Comparison of the short-segment group with the others (long segment, total colonic segment, and small bowel HD)

There were no differences in the demographic data or patient characteristics between the children in the short segment and those in the other segments, except that the clinical presentation of bilious vomiting was more prevalent in the others (p=0.040). In terms of treatment and patient outcomes during the long-term follow-up period, the others had longer delays in ostomy closure (p=0.009) and perianal infection after surgical anastomosis (p=0.025) than did the patients in the short-segment group (**Table 2**).

Factors associated with bowel dysfunction in children diagnosed with HD

Bowel dysfunction was classified as nonretentive fecal incontinence, constipation, and/or retentive fecal incontinence and HAEC. Univariate and multivariate analyses were performed for potential factors (sex, gestational age, age at surgical pull-through operation less than 6 months, primary pull-through operation, operative pull-through technique, anatomical classification of HD, and history of HAEC before pull-through operation). Age at pull-through surgery less than 6 months was the only independent factor associated with

Table 2. Demographics, characteristics, treatment, and outcomes in children with Hirschsprung disease (n=97)

Demographic data	Short segment (n=72)	Others (n=25)	p-value
Age at presentation (d)	3 (1.5–14.5)	2 (2–30)	0.880
Sex, male	61 (84.72)	21 (84.54)	>0.999
Preterm (≤37 wk)	10 (13.89)	4 (16.00)	0.751
Referral cases	52 (72.22)	20 (80.00)	0.597
Family history of Hirschsprung disease	2 (2.78)	3 (12.00)	0.106
Associated anomalies			
Syndromic	9 (12.50)	4 (16.00)	0.735
Congenital heart disease	11 (15.28)	6 (24.00)	0.365
Patient characteristics and clinical presentation			
Abdominal distension	47 (65.28)	10 (40.00)	0.405
Nonbilious vomiting	12 (16.67)	2 (8.00)	0.725
Bilious vomiting	10 (13.89)	7 (28.00)	0.040
Constipation	14 (15.28)	3 (12.00)	>0.999
Enterocolitis	8 (11.11)	4 (16.00)	0.500
Delayed passage of meconium (>24 hr) (n=53)	25/40 (62.50)	8/13 (61.54)	>0.999
Treatment and disease outcome			
Age at the pull-through operation <6 months	67 (93.05)	21 (84.00)	0.230
One-stage pull-through operation	62 (86.11)	15 (60.00)	0.009
Follow-up period (yr)	8.96 (4.36–12.96)	7.52 (4.18–10.90)	0.627
Complication after surgery until the end of follow-up			
Other complications			
Retained aganglionic segment	7 (9.72)	2 (8.00)	>0.999
Anastomosis stricture	9 (12.50)	3 (12.00)	>0.999
Perianal infection	3 (4.17)	5 (20.00)	0.025
Bowel dysfunctions	45 (62.50)	18 (72.00)	0.471
Non retentive incontinence	13 (18.05)	7 (28.00)	0.389
Retentive incontinence	12 (16.67)	3 (12.00)	0.753
Constipation	16 (22.22)	4 (16.00)	0.580
Hirschsprung associated enterocolitis	32 (44.44)	13 (52.00)	0.642

Present as number (%) or median (interquartile range).
Others: long segment, total colonic segment and small bowel Hirschsprung disease.

https://doi.org/10.5223/pghn.2024.27.6.372

nonretentive fecal incontinence after pull-through surgery (adjusted OR 8.83; 95% CI, 1.11–70.39; *p*=0.040), while the Duhamel procedure was the only independent factor associated with constipation and/or retentive fecal incontinence after pull-through surgery (adjusted OR 62.15; 95% CI, 1.64–2,349.13; *p*=0.026). In addition, pre-HAEC was strongly associated with HAEC after the pull-through operation (adjusted OR 18.31; 95% CI, 1.30–257.73; *p*=0.031). A gestational age less than 37 weeks tended to be associated with HAEC after the pull-through operation (adjusted OR 6.57; 95% CI, 0.99–43.65; *p*=0.051) (**Table 3**).

DISCUSSION

This study revealed a high prevalence of bowel dysfunction after pull-through surgery in children diagnosed with HD. The most common issue was HAEC, followed by nonretentive incontinence, constipation, and retentive incontinence. An independent factor associated with postoperative HAEC was a history of enterocolitis before the pull-through operation, whereas the Duhamel procedure was associated with constipation and retentive incontinence. Additionally, a pull-through operation before 6 months of age was significantly associated with nonretentive fecal incontinence.

Long-term functional outcomes of the bowel after pull-through surgery in HD patients have been well described in studies of adult and pediatric populations [6-8,13,14]. Although some functional outcomes of the bowel usually improve with time, they affect the quality of life of patients. Niramis et al. [7] studied long-term functional outcomes after pull-through surgery for HD and reported that fecal incontinence was a common problem in some patients that subsided after more than 15 years of follow-up. However, obstructive symptoms, including constipation and fecal incontinence, are other concerns because they can persist in a substantial number of adults with HD [6,13,14]. In this study, the prevalence of bowel dysfunction after the pull-through operation was high compared with that in previous studies [6,8,15]. The different methodology of this study (the extensive assessment of many aspects of bowel dysfunction and the definition of bowel dysfunction, for example, using the Rome IV criteria for the diagnosis of functional constipation) might be the reason for the difference. In addition to these prevalence studies, further studies on the quality of life of children diagnosed with HD who have bowel dysfunction are necessary to determine the importance of these conditions following further investigations for specific treatment plans.

In terms of HAEC as an obstructive symptom after pull-through surgery in patients with HD, a recent meta-analysis demonstrated that anastomotic stenosis or fistula, preoperative enterocolitis, malnutrition, respiratory infection, long aganglionic segment, Down syndrome, and postoperative ileus were significantly associated factors [16]. In the present study, a history of HAEC before pull-through surgery was the only independent factor associated with HAEC; this observation aligns with the findings of a previous meta-analysis. The etiology of HAEC has been elusive thus far. There are many theories of HAEC, including stasis due to partial obstruction, loss of normal intestinal mucosal defense and mucin composition, and altered microbiota [6,17]. Another possibility for recurrent HAEC after pull-through surgery is a motility disorder of the internal anal sphincter and/or ganglionic bowel, which was previously described using ARM, transit study, and colonic manometry as investigative tools [4,18-20]. A recent paper by the North American Society of Paediatric Gastroenterology, Hepatology, and Nutrition [21] encouraged the integration

Pediatric Gastroenterology, Hepatology & Nutrition
pghn

Table 3. Factors associated with bowel dysfunctions after pull-through surgery in children diagnosed with Hirschsprung disease after long-term follow-up

Parameters	Number	Bowel dysfunctions											
		Nonretentive incontinence				Retentive incontinence/constipation				Hirschsprung associated enterocolitis			
		Crude OR (95% CI)	*p*-value	Adjusted OR (95% CI)	*p*-value	Crude OR (95% CI)	*p*-value	Adjusted OR (95% CI)	*p*-value	Crude OR (95% CI)	*p*-value	Adjusted OR (95% CI)	*p*-value
Male sex	97	0.83 (0.38–8.86)	0.454	2.18 (0.14–34.57)	0.578	0.67 (0.19–2.37)	0.531	0.43 (0.04–5.21)	0.511	1.36 (0.44–4.17)	0.590	0.93 (0.12–7.05)	0.940
Gestational age ≤37 wk	97	1.66 (0.34–8.11)	0.530	1.52 (0.19–21.17)	0.693	1.66 (0.34–8.11)	0.530	0.86 (0.15–5.15)	0.872	3.76 (0.98–14.45)	0.054	6.57 (0.99–43.65)	0.051
Age at the pull-through operation ≤6 months	97	6.08 (1.46–25.35)	0.013	8.83 (1.11–70.39)	0.040	2.09 (0.47–9.21)	0.331	4.53 (0.66–31.21)	0.125	1.5 (0.38–5.96)	0.565	0.36 (0.05–2.82)	0.333
The one-stage pull-through surgery	97	2.65 (0.89–7.93)	0.081	3.29 (0.64–17.08)	0.156	0.62 (0.16–2.38)	0.489	0.12 (0.01–1.79)	0.125	0.73 (0.27–1.96)	0.521	0.42 (0.07–2.61)	0.356
The pull-through operation techniques													
Swenson	56	Ref		Ref		Ref		Ref				Ref	
Yancey-Soave	9	2.04 (0.44–9.49)	0.361	1.57 (0.19–13.07)	0.678	0.46 (0.05–4.03)	0.482	0.25 (0.01–5.60)	0.379	2.67 (0.61–11.76)	0.195	1.86 (0.25–14.14)	0.547
Duhamel	4	1.36 (0.13–14.40)	0.260	0.59 (0.02–18.59)	0.761	11 (1.05–115.51)	0.046	62.15 (1.64–2,349.13)	0.026	1	-	1	-
Anatomical classification of Hirschsprung disease													
Short segment	72	Ref		Ref		Ref		Ref		Ref		Ref	
Long segment	8	1.51 (0.27–8.36)	0.635	0.17 (0.01–4.64)	0.292	0.17 (0.22–6.35)	0.858	0.92 (0.10–8.55)	0.938	1.25 (0.28–5.39)	0.765	1.53 (0.18–12.69)	0.696
Total colonic segment	12	1.51 (0.36–6.37)	0.573	0.43 (0.06–3.21)	0.413	0.32 (0.04–2.65)	0.290	0.36 (0.03–3.71)	0.388	1.75 (0.51–6.04)	0.376	1.59 (0.33–7.72)	0.564
Small bowel Hirschsprung disease	5	3.03 (0.46–19.97)	0.122	9.06 (0.56–147.60)	0.122	0.88 (0.09–8.39)	0.908	1.23 (0.04–33.90)	0.905	0.83 (0.13–5.30)	0.847	4.39 (0.11–171.16)	0.429
Enterocolitis before the pull-through operations	97	2.16 (0.58–8.05)	0.253	3.48 (0.53–22.93)	0.196	1.34 (0.27–6.69)	0.719	0.22 (0.02–3.00)	0.255	7.14 (1.47–34.62)	0.015	18.31 (1.30–257.73)	0.031

OR: odds ratio, CI: confidence interval, Ref: reference.

https://pghn.org
https://doi.org/10.5223/pghn.2024.27.6.372

of motility studies as part of working on the functional problems of the bowel after surgical management for HD, as motility disorders guide targeted management of these patients [21].

Another major problem after pull-through surgery in children diagnosed with HD in this study is constipation and retentive fecal incontinence. Several studies have indicated that constipation is a functional outcome notably influenced by the chosen surgical approach. Although successful in many patients, Duhamel pull-through may cause significant symptoms, such as impaction and overflow incontinence [22,23]. In our study, the most frequently performed procedure was the Swenson pull-through in 57.73% of patients, while the Yancey-Soave and Duhamel procedures were performed in a few patients (9.28% and 4.12%, respectively). The Duhamel procedure is an independent factor associated with constipation or retentive fecal incontinence. Constipation following pull-through procedures may occur owing to anatomical, pathological, or functional problems [24,25]. After excluding anatomical strictures and retaining aganglionic segments from pull-through surgery [25], why some patients with Duhamel's pouch experience efficient emptying has remained unclear. A retained spur in the native rectum or a dilated Duhamel pouch may lead to stool impaction, obstructing ganglionated pull-through [21,22,25]. Moreover, obstructive symptoms, such as the absence of the rectoanal inhibitory reflex (RAIR) and abnormal proximal motility, have been attributed to functional causes and were not investigated in the present study. Although the RAIR is absent in every child with HD, it leads to obstructive symptoms in some cases [25,26]. The evaluation of anorectal function by ARM to identify candidates suitable for botulinum toxin injections might be helpful [25,27]. In addition, if obstructive symptoms persist despite multiple botulinum toxin injections, consideration of an associated motility disorder of the colon is the next step. Techniques for diagnosing motility disorders include radiological Sitz marker studies, radionuclide colon transit studies, and colonic manometry, which are the most accurate methods for characterizing the nature and location of dysmotility [25,28].

Previous studies have focused on age at the time of the pull-through surgery and functional bowel dysfunction and have yielded varied results. According to a national survey conducted in the UK and Ireland (2010), pediatric surgeons typically prefer to perform a pull-through before 3 months of age [29] to minimize the need for irrigation or stoma and reduce the risk of preoperative HAEC. Surgery can be advanced before 1 month of age. However, pull-through surgery may be postponed for reasons such as total colonic aganglionosis, delayed diagnosis, other urgent surgical procedures for cardiac comorbidities, or surgeon preference [30]. Onishi et al. [31] studied data from 65 children with HD and indicated that pull-through surgery performed at <6 months of age led to optimal bowel function. Bradnock and Walker [29] also concluded that age at pull-through surgery did not influence the probability or severity of constipation or fecal incontinence in the long term. In contrast, our study revealed that surgical correction before 6 months of age was significantly associated with nonretentive fecal incontinence. A recent study from Poland and China showed results similar to those of our study; Sosnowska et al. [32] reported that children who underwent radical surgery before 4 months of age had a greater incidence of complications, including fecal incontinence.

Zhu et al. [33] reported parallel outcomes, revealing that infants with short-segment HD at <3 months of age who underwent single-stage endorectal pull-through surgery had higher rates of anastomotic leakage in the short term and increased soiling in the midterm than did those who underwent surgery after 3 months of age. The preparation of fragile structures in the anal region is challenging and requires precision, but it may not always be feasible

when performing pull-through surgery in small children [34,35]. Hence, incontinence after the procedure can result from factors such as internal sphincter and pelvic nerve injury or sensory deficits in the anal skin during surgical dissection. These risks are greater in neonates than in adults, highlighting the need for careful neonatal surgical approaches [26,33]. To clarify this hypothesis, ultrasonography and magnetic resonance imaging are recommended for evaluating anorectal structures, if available [36]. Recently, three-dimensional high-resolution ARM has been used to identify functional problems such as sphincter hypotonia and asymmetric squeezing efforts after pull-through surgery in adult patients with HD [37], but these investigations are limited to infants and young children.

The strengths of this study include the use of histopathology results for diagnosis in children with HD, and the presence of bowel dysfunction was evaluated over a long follow-up period. Nevertheless, our findings must be contextualized with certain limitations. First, the retrospective nature of this review led to the omission of certain potential parameters related to bowel dysfunction. Another limitation was the loss to follow-up and the associated risk of selection bias, which are common in follow-up studies. Patients with good clinical outcomes are more likely to be lost to follow-up. Finally, the burden of bowel dysfunction and motility was not evaluated in any of the children with bowel dysfunction in the present study.

Conclusion

The prevalence of bowel dysfunction in children diagnosed with HD was high, even after successful pull-through surgery. Preoperative HAEC, Duhamel operation, and pull-through surgery before the age of 6 months were found to be independent factors associated with bowel dysfunction after pull-through surgery.

ACKNOWLEDGEMENTS

The authors are very grateful to Dusit Viravaidya, Pornchai Achatsachat, Pattamon Sutthattarn, and all the staff in the Department of Pediatrics and Department of Surgery, Faculty of Medicine, Chulalongkorn University, and King Chulalongkorn Memorial Hospital for their excellent patient care.

REFERENCES

1. Kliegman RM, St Geme JW, Blum NJ, Shah SS, Tasker RC, Wilson KM. Motility disorders and hirschsprung disease. In: Kliegman RM, St Geme JW, Blum NJ, Shah SS, Tasker RC, Wilson KM, eds. Nelson textbook of pediatrics. 21th ed. Philadelphia: Elsevier, 2020:1955-65.e1.
2. Kawaguchi AL, Guner YS, Sømme S, Quesenberry AC, Arthur LG, Sola JE, et al.; American Pediatric Surgical Association Outcomes and Evidence-Based Practice (OEBP) Committee. Management and outcomes for long-segment Hirschsprung disease: a systematic review from the APSA Outcomes and Evidence Based Practice Committee. J Pediatr Surg 2021;56:1513-23. PUBMED | CROSSREF
3. Verkuijl SJ, Meinds RJ, van der Steeg AFW, van Gemert WG, de Blaauw I, Witvliet MJ, et al. Functional outcomes after surgery for total colonic, long-segment, versus rectosigmoid segment Hirschsprung disease. J Pediatr Gastroenterol Nutr 2022;74:348-54. PUBMED | CROSSREF
4. Miele E, Tozzi A, Staiano A, Toraldo C, Esposito C, Clouse RE. Persistence of abnormal gastrointestinal motility after operation for Hirschsprung's disease. Am J Gastroenterol 2000;95:1226-30. PUBMED | CROSSREF
5. Neves Romaneli MT, Ribeiro AF, Bustorff-Silva JM, Carvalho RB, Lomazi EA. Hirschsprung's disease - Postsurgical intestinal dysmotility. Rev Paul Pediatr 2016;34:388-92. PUBMED | CROSSREF

6. Meinds RJ, van der Steeg AFW, Sloots CEJ, Witvliet MJ, de Blaauw I, van Gemert WG, et al. Long-term functional outcomes and quality of life in patients with Hirschsprung's disease. Br J Surg 2019;106:499-507. PUBMED | CROSSREF
7. Niramis R, Watanatittan S, Anuntkosol M, Buranakijcharoen V, Rattanasuwan T, Tongsin A, et al. Quality of life of patients with Hirschsprung's disease at 5 - 20 years post pull-through operations. Eur J Pediatr Surg 2008;18:38-43. PUBMED | CROSSREF
8. Chantakhow S, Tepmalai K, Singhavejsakul J, Tantraworasin A, Khorana J. Prognostic factors of postoperative Hirschsprung-associated enterocolitis: a cohort study. Pediatr Surg Int 2023;39:77. PUBMED | CROSSREF
9. Hyams JS, Di Lorenzo C, Saps M, Shulman RJ, Staiano A, van Tilburg M. Functional Disorders: Children and Adolescents. Gastroenterology 2016;150:1456-68.E2. PUBMED | CROSSREF
10. Pastor AC, Osman F, Teitelbaum DH, Caty MG, Langer JC. Development of a standardized definition for Hirschsprung's-associated enterocolitis: a Delphi analysis. J Pediatr Surg 2009;44:251-6. PUBMED | CROSSREF
11. Gunadi , Luzman RA, Kencana SMS, Arthana BD, Ahmad F, Sulaksmono G, et al. Comparison of two different cut-off values of scoring system for diagnosis of Hirschsprung-associated enterocolitis after transanal endorectal pull-through. Front Pediatr 2021;9:705663. PUBMED | CROSSREF
12. Veras LV, Arnold M, Avansino JR, Bove K, Cowles RA, Durham MM, et al.; American Pediatric Surgical Association Hirschsprung Disease Interest Group. Guidelines for synoptic reporting of surgery and pathology in Hirschsprung disease. J Pediatr Surg 2019;54:2017-23. PUBMED | CROSSREF
13. Zimmer J, Tomuschat C, Puri P. Long-term results of transanal pull-through for Hirschsprung's disease: a meta-analysis. Pediatr Surg Int 2016;32:743-9. PUBMED | CROSSREF
14. Granström AL, Danielson J, Husberg B, Nordenskjöld A, Wester T. Adult outcomes after surgery for Hirschsprung's disease: evaluation of bowel function and quality of life. J Pediatr Surg 2015;50:1865-9. PUBMED | CROSSREF
15. Dai Y, Deng Y, Lin Y, Ouyang R, Li L. Long-term outcomes and quality of life of patients with Hirschsprung disease: a systematic review and meta-analysis. BMC Gastroenterol 2020;20:67. PUBMED | CROSSREF
16. Zhang X, Sun D, Xu Q, Liu H, Li Y, Wang D, et al. Risk factors for Hirschsprung disease-associated enterocolitis: a systematic review and meta-analysis. Int J Surg 2023;109:2509-24. PUBMED | CROSSREF
17. Heuckeroth RO. Hirschsprung disease - integrating basic science and clinical medicine to improve outcomes. Nat Rev Gastroenterol Hepatol 2018;15:152-67. PUBMED | CROSSREF
18. Trinh TH, Nguyen NM, Lam KT, Pham TN, Vu NT, Truong LNU, et al. Anorectal manometry findings in relation with long-term functional outcomes of the patients operated on for Hirschsprung's disease compared to the reference-based population. Pediatr Surg Int 2023;39:131. PUBMED | CROSSREF
19. Zhang SC, Bai YZ, Wang W, Wang WL. Stooling patterns and colonic motility after transanal one-stage pull-through operation for Hirschsprung's disease in children. J Pediatr Surg 2005;40:1766-72. PUBMED | CROSSREF
20. Tran VQ, Mahler T, Bontems P, Truong DQ, Robert A, Goyens P, et al. Interest of anorectal manometry during long-term follow-up of patients operated on for Hirschsprung's disease. J Neurogastroenterol Motil 2018;24:70-8. PUBMED | CROSSREF
21. Ambartsumyan L, Patel D, Kapavarapu P, Medina-Centeno RA, El-Chammas K, Khlevner J, et al. Evaluation and management of postsurgical patient With Hirschsprung disease Neurogastroenterology & Motility Committee: position paper of North American Society of Pediatric Gastroenterology, Hepatology, and Nutrition (NASPGHAN). J Pediatr Gastroenterol Nutr 2023;76:533-46. PUBMED | CROSSREF
22. Chatoorgoon K, Pena A, Lawal TA, Levitt M. The problematic Duhamel pouch in Hirschsprung's disease: manifestations and treatment. Eur J Pediatr Surg 2011;21:366-9. PUBMED | CROSSREF
23. Fortuna RS, Weber TR, Tracy TF Jr, Silen ML, Cradock TV. Critical analysis of the operative treatment of Hirschsprung's disease. Arch Surg 1996;131:520-4. PUBMED | CROSSREF
24. Levitt MA, Dickie B, Peña A. Evaluation and treatment of the patient with Hirschsprung disease who is not doing well after a pull-through procedure. Semin Pediatr Surg 2010;19:146-53. PUBMED | CROSSREF
25. Ahmad H, Levitt MA, Yacob D, Halleran DR, Gasior AC, Di Lorenzo C, et al. Evaluation and management of persistent problems after surgery for Hirschsprung disease in a child. Curr Gastroenterol Rep 2021;23:18. PUBMED | CROSSREF
26. 26Levitt MA, Dickie B, Peña A. The Hirschsprungs patient who is soiling after what was considered a "successful" pull-through. Semin Pediatr Surg 2012;21:344-53. PUBMED | CROSSREF

27. Soh HJ, Nataraja RM, Pacilli M. Prevention and management of recurrent postoperative Hirschsprung's disease obstructive symptoms and enterocolitis: systematic review and meta-analysis. J Pediatr Surg 2018;53:2423-9. PUBMED | CROSSREF

28. Di Lorenzo C, Solzi GF, Flores AF, Schwankovsky L, Hyman PE. Colonic motility after surgery for Hirschsprung's disease. Am J Gastroenterol 2000;95:1759-64. PUBMED | CROSSREF

29. Bradnock TJ, Walker GM. Evolution in the management of Hirschsprung's disease in the UK and Ireland: a national survey of practice revisited. Ann R Coll Surg Engl 2011;93:34-8. PUBMED | CROSSREF

30. Kyrklund K, Sloots CEJ, de Blaauw I, Bjørnland K, Rolle U, Cavalieri D, et al. ERNICA guidelines for the management of rectosigmoid Hirschsprung's disease. Orphanet J Rare Dis 2020;15:164. PUBMED | CROSSREF

31. Onishi S, Kaji T, Nakame K, Yamada K, Murakami M, Sugita K, et al. Optimal timing of definitive surgery for Hirschsprung's disease to achieve better long-term bowel function. Surg Today 2022;52:92-7. PUBMED | CROSSREF

32. Sosnowska P, Błaszczyński M, Moryciński S, Mańkowski P. Definitive surgery for Hirschsprung's disease under 4 months of age is associated with long-term complications: A cohort study. Pediatr Pol 2017;92:548-52. CROSSREF

33. Zhu T, Sun X, Wei M, Yi B, Zhao X, Wang W, et al. Optimal time for single-stage pull-through colectomy in infants with short-segment Hirschsprung disease. Int J Colorectal Dis 2019;34:255-9. PUBMED | CROSSREF

34. Sosnowska P, Błaszczyński M. A 15-year experience with the one-stage surgery for treatment of Hirschsprung's disease in newborns, infants, and young children. Indian J Surg 2015;77(Suppl 3):1109-14. PUBMED | CROSSREF

35. Puri P. Hirschsprung's disease and allied disorders. Cham: Springer International Publishing, 2019.

36. Carrington EV, Scott SM, Bharucha A, Mion F, Remes-Troche JM, Malcolm A, et al.; International Anorectal Physiology Working Group and the International Working Group for Disorders of Gastrointestinal Motility and Function. Expert consensus document: advances in the evaluation of anorectal function. Nat Rev Gastroenterol Hepatol 2018;15:309-23. PUBMED | CROSSREF

37. Krois W, Reck CA, Darbari A, Badillo A, Levitt MA. A technique to reconstruct the anal sphincters following iatrogenic stretching related to a pull-through for Hirschsprung disease. J Pediatr Surg 2021;56:1242-6. PUBMED | CROSSREF