

Outcomes of patients with Hirschsprung's disease post-pull-through procedure

T Thelele,¹ J Scribante,^{1,2} C Bebington,³ G Brisighelli^{1,3}

¹ Department of Paediatric Surgery, School of Clinical Medicine, Faculty of Health Sciences, University of the Witwatersrand, South Africa

² Surgeons for Little Lives, South Africa

³ Johannesburg Paediatric Colorectal Clinic, Chris Hani Baragwanath Academic Hospital, Department of Paediatric Surgery, University of the Witwatersrand, Johannesburg, South Africa

Corresponding author, email: letebz@icloud.com

Background: In low- to middle-income countries, diagnosing and managing Hirschsprung's disease (HD) presents challenges. This study aimed to determine the outcomes of patients with HD post-pull-through (PT) surgery in the Department of Paediatric Surgery at Chris Hani Baragwanath Academic Hospital (CHBAH) between 01 January 2016 and 30 June 2024.

Methods: In Section 1, demographics and complications (according to the Clavien-Dindo Classification System) were collected retrospectively. In Section 2, the functional bowel outcomes (using the Rintala Bowel Functional Score) were collected prospectively.

Results: There were 78 HD-diagnosed patients in the database; 43 (55%) were included in section 1, and 32 (74%) in section 2. Thirty-seven patients (86%) were male, and 21 (49%) had inside flushing toilets. Thirty-two (74%) patients had a transition zone at the rectosigmoid junction, and five (14%) had total colonic HD. The median (IQR) age at diagnosis was 3 (22) months, and only 13 (30%) were diagnosed before 1 month. The median (IQR) age at PT was 19 (28.8) months. Eighteen (42%) patients had no postoperative complications, and 21 (49%) needed surgery-PT. Nineteen patients (59%) had excellent continence, and 11 (34%) had good continence. There was no statistically significant difference in functional outcomes, age at first diagnosis ($p = 0.924$), available toilet facilities ($p = 0.791$), or the level of disease ($p = 0.624$).

Conclusion: The majority of HD patients at CHBAH had a delayed diagnosis and some postoperative complications following PT. However, most patients had excellent to good functional bowel outcomes.

Keywords: Hirschsprung's disease, low- to middle-income countries, functional bowel outcomes, complications

Introduction

Hirschsprung's disease (HD) is a congenital condition marked by the lack of ganglion cells in the enteric nervous system. HD affects approximately 1 in every 5000 live births.¹ It most commonly affects the rectosigmoid but can extend proximally to the small bowel, with variable lengths from the rectum.² The affected segment is narrowed and impedes peristalsis, causing functional obstruction.³ HD typically manifests as neonatal abdominal distension, bilious vomiting, and feeding intolerance. Meconium passage is delayed in approximately 90% of patients after the first 24 hours. In some cases, caecal or appendiceal perforation may occur as the first event. Hirschsprung-associated enterocolitis, which can be fatal in 10% of neonates with HD, causes fever, abdominal distention, and diarrhoea.¹

In low- to middle-income countries (LMIC), a high number of patients with HD present beyond the age of one year, which may negatively impact the management and outcomes of these patients. These patients tend to have a higher incidence of recurrent enterocolitis, malnutrition, or severely dilated proximal bowel and perforation.¹ In these settings, a two-stage operative repair is typically considered safer, with the first stage consisting of a defunctioning stoma followed by a PT procedure.³

Long-term challenges in children who underwent PT surgery for HD may include recurrent enterocolitis, soiling, and obstructive symptoms. The exact incidence of each of these challenges is not well described.¹ The methods used in the literature vary greatly. However, the Rintala Bowel Function Score has been previously validated and used to evaluate bowel function in patients with HD, and it has the advantage of being validated in a control group of healthy children.⁴

Moreover, the lack of a definition and classification of surgical complications after an HD PT surgery has made interpreting surgical outcomes difficult. Different authors used terms such as mild, moderate, and severe to describe surgical complications, but little clarity has been given to defining the terms used.⁵ The Clavien-Dindo Classification of Surgical Complications is an objective grading system used to grade postoperative complications in a reproducible manner.^{6,7}

Of note, the majority of studies on the short- and long-term outcomes of HD are conducted in high-income countries^{1,2,8} and very little is known about bowel function in LMICs⁹ where, due to a lack of resources, data collection, reporting, and poor follow-up, data are scarce regarding postoperative outcomes and bowel function of children with Hirschsprung's disease. This study aimed to determine the outcomes of

patients with HD post-PT surgery in the Department of Paediatric Surgery at Chris Hani Baragwanath Academic Hospital (CHBAH) from 1 January 2016 to 30 June 2024.

Materials and methods

A retrospective and prospective descriptive research design was followed, and approval to conduct this study was obtained from the Human Research Ethics Committee of the University of the Witwatersrand (M230431).

The study was conducted in the Department of Paediatric Surgery at CHBAH. The Department admits, on average, 2400 patients and performs over 1500 surgeries per annum. The Department has a dedicated colorectal unit, the Johannesburg Paediatric Colorectal Clinic (JPCC), which manages, on average, 2100 patients per annum. The unit has performed more than 1200 surgeries since its inception.

The study population consisted of children undergoing PT surgery for HD at CHBAH. The sample size was realised by the number of children in the HD database, and a consecutive convenience sampling method was used. Children > 3 years of age who had a histological diagnosis of HD and had had PT surgery at least six months prior were included in the study. Children with developmental delays and those whose caregivers declined consent were excluded.

The authors compiled a data collection instrument comprising two sections. Section 1 consists of the following demographic variables: sex, age, type of toilet facility, age at HD diagnosis, level of disease, age at first procedure, and age at PT surgery and classifying postoperative complications using Clavien-Dindo Classification of Surgical Complications.¹⁰ Section 2 uses the Rintala Bowel Function Score⁶ to assess functional outcomes.

Post-surgery patients were followed up at the JPCC and were never discharged from the clinic; the follow-up period varied with each patient, depending on their condition. One author (TT) collected Section 1 data from the JPCC database and Section 2 data during the patient's follow-up visit, or by telephone. He is fluent in most of the languages spoken by patients attending CHBAH and was able to assist them with any problems they had.

At JPCC, the Swenson PT procedure is performed exclusively; CHBAH is a teaching hospital, so multiple surgeons could have performed the PT, but all used the same technique (Swenson). Preoperative contrast enema was done in all patients with a positive rectal biopsy for HD before an elective colostomy was performed. In the case of an emergency colostomy for bowel obstruction, a contrast enema was not performed. Diagnosis was confirmed histologically in all patients before PT surgery. At JPCC, colostomy is the procedure of choice in patients with colonic HD, as ileostomies carry a high risk of dehydration and mortality. Frozen section is not available at our centre; therefore, the stoma doughnut is sent out for identification of ganglion cells before PT surgery. In this study, enterocolitis was defined as intestinal inflammation characterised by fever, abdominal distension, diarrhoea, and sepsis,¹¹ requiring hospital admission. A caregiver referred to the patient's legal guardian. A patient was defined as a child aged 3 to 18 years. According to the age at diagnosis, the patients were divided into three groups:

- Neonatal group (0–28 days)
- Delayed diagnosis group (> 28 days–1 year)
- Late diagnosis group (> one year–18 years)

REDCap was used to capture data. Data analysis was performed using GraphPad Prism 2024. Categorical data are described using frequencies and percentages, and continuous data are described using means and standard deviations. Comparisons between age at first diagnosis and toilet type with regard to functional outcome were performed using the Kruskal-Wallis test. The comparison of disease severity and functional outcome was performed using the Mann-Whitney U test. A *p*-value of 0.05 was considered statistically significant. Charts were designed using the DATAtab Team (2024).

Results

Seventy-eight patients were captured on the database. Forty-three patients (55%) were included in Section 1, and 32 (41%) in Section 2. The sample realisation is shown in Figure 1. The demographic profile of the patients is shown in Table I. Determining the presentation of HD was beyond the scope of the study. However, the presentation differed and depended on the age of the presentation; older patients presented with constipation and a distended abdomen, as well as failure to thrive and recurrent need for washouts; younger patients presented with a distended abdomen, delayed meconium passage, and sometimes enterocolitis.

The surgical outcomes post-PT surgery (using the Clavien-Dindo Classification of Surgical Complications) are shown in Table II.¹² Of the 43 patients, 5 (12%) died. One of the 5 (20%) died before the PT surgery as parents denied consent for diversion and withheld irrigations. The remaining 4 (80%) patients who demised post-surgery died of dehydration – 3 (60%) with ileostomy and 1 (20%) demised from unknown causes.

The mean (SD) Rintala Bowel Function Score¹³ score was 17 (3) with a range of 11–20. Table III shows the functional outcomes post-PT surgery using the Rintala Bowel Function Score¹³ and Table IV shows the classification according to continence.

Of the 32 patients included in section 2, 27 (84%) were male, 27 (84%) had rectosigmoid HD, and 5 (16%) had total colonic disease.

As shown in Figure 2, there was no statistically significant difference between functional outcomes and age at first diagnosis (*p* = 0.924), available toilet facilities (*p* = 0.791), and the level of disease (*p* = 0.624)

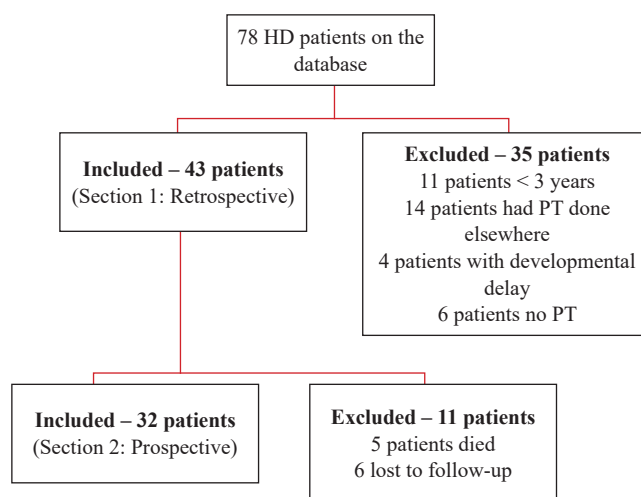


Figure 1: Sample realisation

Table I: Demographic profile of patients

Variables	n	%
Sex		
Male	37	86
Female	6	14
Type of toilet		
Inside toilet	21	49
Outside flushing toilet	15	35
Outside pit latrine	6	14
Community toilet	1	2
Age at first diagnosis		
1–12 months	17	40
> 12 months	13	30
< 1 month	13	30
Level of disease		
Rectosigmoid	32	74
Total colonic	5	12
Descending colon	2	5
Transverse colon	4	10
	Median	IQR
Age at diagnosis (months)	3	22
Age at first pull-through (months)	19	29
Age at questionnaire (months)	60	60
Time between pull-through and questionnaire (months)	36	48.5

Discussion

In this study, a male-female ratio of 6:1 is reported; this is higher than the range of 3.3:1–4:1 reported in the United States of America,¹⁴ a 3.4:1 ratio noted in the United Kingdom and Ireland,¹⁵ and a 3.3:1 in Burkina Faso.¹⁶ In some LMICs, male patients have better access to health care than female patients,¹⁷ however, at JPCC, there are no gender-preferential treatments, and this higher ratio of male patients cannot be explained.

The late diagnosis of HD may negatively impact the management and outcomes of patients.¹⁸ Only 30% of patients in this study were diagnosed in the first month of life. This is lower than the 90% described in high-income countries.^{9,15,19} However, it is higher than the 5% reported in Tanzania.²⁰ A mean age of presentation of 2.4 years was noted in Malawi.²¹ The delayed diagnosis observed in this study may be attributed to several factors, including limited awareness of HD among healthcare workers and the persistence of traditional beliefs among parents and caregivers, who often rely on remedies such as traditional enemas instead of seeking timely medical intervention. Traditional enemas are made with herbs and homemade mixtures and are widely used among the black population in South Africa, especially among children.^{22,23}

A single-stage transanal Swenson PT surgery is typically performed when patients present with HD within the first month of life.⁸ However, in this study, the median age at diagnosis was three months, and patients generally present with poor nutritional and general health status. Therefore, a colostomy is performed to optimise the patient's nutritional status and allow the dilated colon to decompress before a

Table II: Surgical outcomes of post-pull-through using the Clavien-Dindo Classification of Surgical Complications¹⁰

Complication classification	n	%
0 No complications	18	42
I Any deviation from the normal postoperative course without the need for pharmacological treatment or surgical, endoscopic, or radiological interventions	6	14
Dietary changes	6	14
II Requiring pharmacological treatment with drugs other than such allowed for grade I complications	10	23
Enterocolitis requiring admission	10	23
X Intervention not under general anaesthesia	0	0
IIIB Intervention under general anaesthesia	21	49
Botox in theatre	11	26
Examination under anaesthesia and full-thickness rectal biopsy	2	4
Anastomotic stricture requiring dilatation	2	4
Severe abdominal distension requiring laparotomy or diversion	2	4
Twist of the pull-through segment	1	2
Recto vaginal fistula requiring repair and diversion	1	2
Stricture of ileoanal anastomosis requiring a re-do	1	2
Hemoperitoneum requiring a relook laparotomy	1	2
Wound sepsis requiring secondary wound closure	1	2
Disruption of the dentate line with faecal incontinence	1	2
Adhesive bowel obstruction	1	2
IV Life-threatening complications requiring intermediate or intensive care management	0	0
V Death of the patient	5	12
Before pull-through	1	20
After pull-through	4	80

Table III: The functional outcomes post-pull-through surgery using the Rintala Bowel Function Score⁴⁹

Description	n	%
Ability to hold back defecation		
Always	19	59
Problems less than once a week	11	34
Weekly problems	2	6
No voluntary control	0	0
Feels/reports the urge to defecate		
Always	19	59
Most of the time	13	40
Uncertain	0	0
Absent	0	0
Frequency of defecation		
Every other day – twice a day	30	94
More often	2	6
Less often	0	0
Soiling		
Never	25	78
Staining < once a week, no change of underwear required	5	16
Frequent staining < change of underwear often required	2	6
Daily, requires protective aids	0	0
Accidents		
Never	13	41
Less than once a week	10	31
Weekly accidents, often requiring protective aids	5	16
Daily, requires protective aids during the day and night	6	19
Constipation		
No constipation	26	81
Manageable with diet	4	13
Manageable with laxatives	2	6
Manageable with enemas	0	0
Social problems		
No social problems	32	100
Sometimes (foul odours)	0	0
Problems causing restrictions to social life	0	0
Severe social and/or psychic problems	0	0

Table IV: Outcomes of Rintala Bowel function score⁴⁹

Score	Description	n	%
18–20	Excellent continence	19	59
11–16	Good continence	11	34
9–11	Fair continence	2	6
6–9	Poor continence	0	0

Swenson PT of the colostomy is performed at a median age of 19 months in this study.

Rectosigmoid HD predominance of more than 80% has been reported in high-income countries and LMICs.^{1,24-27} A systematic review and meta-analysis²⁸ found that total colonic HD is rare, occurring in fewer than 10% of cases. Although rectosigmoid disease accounted for 74% of the population and total colonic HD for 12% in this study, this

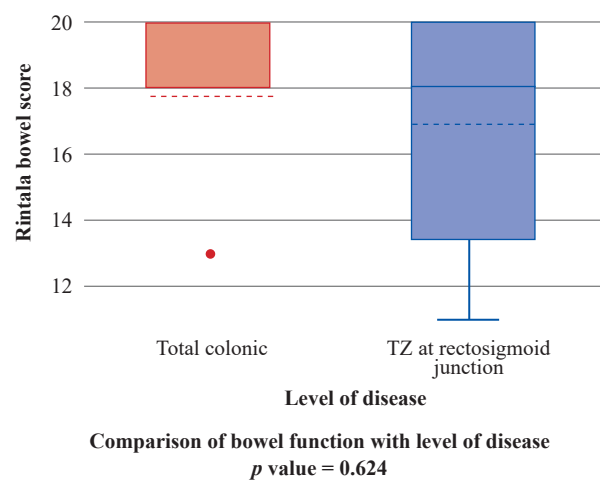
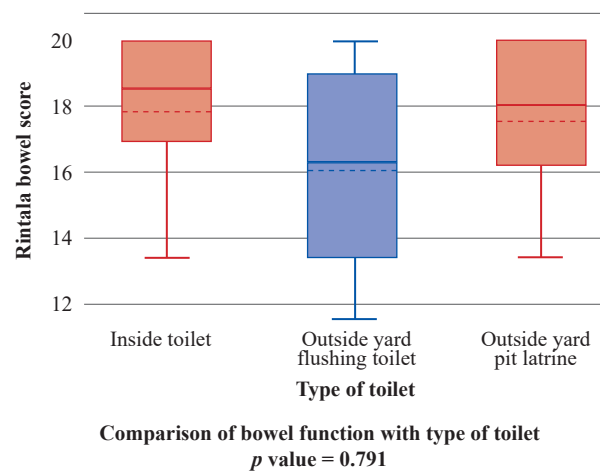
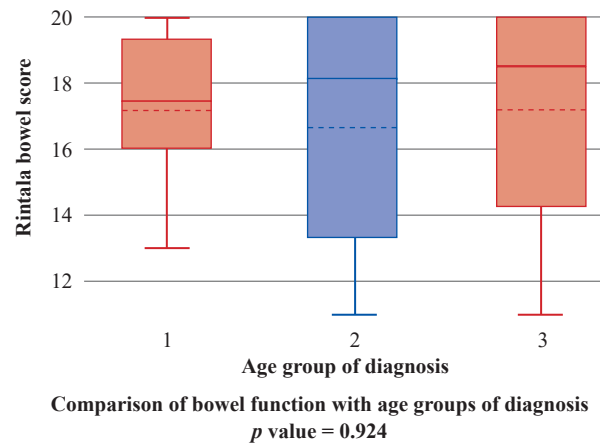


Figure 2: Comparison of bowel function with age group, type of toilet, and level of disease

may not reflect the population served by the JPCC. Due to the widespread use of traditional enemas, the prevalence of rectosigmoid HD is concealed, and the prevalence of total colonic HD may be exaggerated.

This study reported a 49% complication rate; 28% of these occurred in patients who underwent examination under anaesthesia in theatre. As a routine at JPCC, patients who present with features of obstruction that may occur post-PT surgery undergo a contrast enema and an examination under anaesthesia, combined with a Botox injection or a full-thickness rectal biopsy. General anaesthesia is considered a complication under the Clavien-Dindo Classification

of Surgical Complications. Excluding these patients, our complication rate was 21%. The 30-day postoperative complication rate reported in Sweden²⁹ and the Netherlands¹² was 21%. Our study included complications until the study was done, well beyond 30 days. Tanzania³⁰ had a 47% short-term postoperative complication rate, and Nigeria³¹ had a 92.7% complication rate in patients who presented beyond the age of one year. However, neither study utilised the Clavien-Dindo Classification of Surgical Outcomes. Two important factors contributing to the complication rate at JPCC are the delayed presentation of HD and immunosuppressive disorders such as human immunodeficiency virus (HIV) and AIDS. In 2016, it was reported that 15.2% of the global HIV-infected children were from South Africa.³² This may have a potential impact on our results and complication rate.

The mortality rate in this study was 12%. This is higher than reported in Sweden (3%),³³ in Japan (2.7%),³⁴ and the United States of America (< 2%).³⁵ However, it is similar to Uganda's (9.1%)²² and lower than Tanzania's (22%).³⁰ There may be multiple reasons for the JPCC's demise. The delayed presentation of HD, the exposure to HIV, as well as the social conditions, with many patients living in rural areas and with long distances to travel to reach the hospital. Having an ileostomy is a risk factor for mortality in this study, and this is secondary to the dehydration that patients can develop secondary to increased losses; this is true in both high-income countries (HICs)³⁶ and LMICs.³⁷

Postoperative enterocolitis represents severe morbidity and mortality in patients with HD.³⁸ Incidence rates of 40%³⁹ and 25–37%⁴⁰ have been reported in two American studies, respectively. A Chinese systematic review showed an incidence of 23.1%,⁴¹ similar to the incidence (23%) in our study; postoperative enterocolitis may be underreported in our patient population due to a lack of knowledge of patients and healthcare workers, as described above, and may also represent a significant risk factor for mortality in HD patients.

In this study, a faecal soiling rate of 22% was reported. This is lower than the 29.41% in India⁴² and higher than the 11% reported in Ethiopia.⁴³ Of the patients in this study, 19% reported constipation that required dietary changes or laxatives. This was the same as the rate reported in India (19%)⁴⁴ and lower than the 53% in Ireland.⁴⁵ Soiling and constipation are very well-described long-term complications of patients with HD, also in HICs.⁴⁶ A possible explanation for faecal soiling and constipation in our study was that 51% of patients lacked access to an indoor flushing toilet. It has been noted that if patients are not comfortable with their toilet facilities, they present with avoidance behaviours, which may result in overflow diarrhoea.⁴⁷ Another possible explanation may be the patient's diet. In South Africa, the staple diet for lower-income families is maize meal, which is not fiber-rich.⁴⁸

While none of the children in this study reported social problems according to parental responses on a single questionnaire item, this finding should be interpreted with caution. The reliance on parental proxy reporting and the limited scope of the questionnaire's social issues question prevent definitive conclusions regarding the absence of social problems within this population. A comprehensive assessment of quality of life (QOL) employing validated instruments that encompass diverse aspects of social functioning is necessary before asserting that the participating

children lack social difficulties. Davidson et al.⁴⁶ conducted a comprehensive review of patients diagnosed with HD, employing a QOL assessment specifically validated for individuals with HD, alongside the Rintala Bowel Function Score. The findings of their study revealed that patients with HD showed a markedly heightened awareness of their bowel health in contrast to the general healthy population. Notably, the authors highlighted that 42% of patients assessed had impaired QOL scores, with a substantial proportion of these cases occurring in children younger than eight years.

In this study, 70% of patients presented with a delayed diagnosis. Despite this, 59% of patients reported excellent and 34% good continence. The continence scores observed in our study are similar to those reported in Ethiopia (57% excellent and 32% good continence); however, the authors reported a lower incidence of delayed diagnosis (57.5%).⁴³ Our continence score was higher than the 52% excellent continence rate reported in the United Kingdom⁴⁶ and the 47% of Finnish patients who obtained a score of more than 17 in patients presenting timeously.⁴⁹

The favourable functional outcomes observed in our study can be attributed, in part, to the proactive approach during the establishment of JPCC. At the time, it was known that patients presenting to JPCC presented late, predominantly had malnutrition, and came from economically disadvantaged backgrounds; this influenced us to tailor surgeries to patients' specific needs. A critical component of our intervention strategy involved implementing a staged surgical procedure, which facilitated necessary nutritional rehabilitation before definitive surgical intervention. This approach included integrating multidisciplinary support and engaging dietitians and psychologists as essential contributors to the patient's routine clinical assessments. Furthermore, establishing a dedicated caregiver follow-up communication platform that enables direct interactions with the surgical team and fosters caregiver support groups was instrumental in enhancing patient outcomes. Collectively, these factors show the importance of a comprehensive, team-oriented approach to managing the health of this vulnerable population, ultimately leading to improved clinical results.

This study has some limitations. It was done contextually at CHBAH, and the results of this study may not be generalisable to other hospitals. However, the results from this study provide important information regarding patients treated at CHBAH. Due to the study's scope, functional outcomes were assessed during the study period rather than at standardised times throughout patients' postoperative journey.

It is recommended that a longitudinal study assessing patients' improvement in bowel function and a QOL study using a validated instrument to describe patients' social problems be conducted at JPCC. More studies utilising validated instruments, such as the Rintala Bowel Function Score⁵⁰ and the Clavien-Dindo Classification of Surgical Complications,¹⁰ should be considered in LMICs.

Conclusion

The majority of HD patients at CHBAH had a delayed diagnosis and some postoperative complications following PT surgery. However, most patients had excellent to good functional bowel outcomes.

Conflict of interest

The authors declare no conflict of interest.

Funding source

There is no funding to declare.

Ethical approval


Ethical approval obtained from the Human Research Ethics Committee of the University of the Witwatersrand, ethics number M230431.

ORCID

T Thelele  <https://orcid.org/0000-0002-9159-7425>

J Scribante  <https://orcid.org/0000-0002-2221-5024>

C Bebington  <https://orcid.org/0000-0002-0234-323X>

G Brisighelli  <https://orcid.org/0000-0002-5449-7820>

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