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## Long-term assessment of functional outcome in operated cases of Hirschsprung disease



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### ABSTRACT

**Background:** Hirschsprung disease (HD) is a rare congenital anomaly (1 in 5000 live births) characterized by varying levels of aganglionosis. The evolution of surgical options and outcomes for the successful management of HD has been driven by the aspiration of pediatric surgeons. Hirschsprung's disease postoperative results of pull-through procedure could be influenced by surgical and pathological proficiency and the quality of perioperative care. Controversies persist as to which technique offers the best outcome. The absence of incontinence and constipation remains the most important marker of a good outcome.

**Methods:** Medical records of HD children who underwent temporary colostomy procedures were analyzed retrospectively from July 2017 to June 2021. Risk factors were evaluated to determine the outcome of HD procedures.

**Results:** Ninety-two HD patients were included. Based on the univariate analysis, most patients in the study (58.7%) were between the ages of 2 and 12 months. The study population predominantly consisted of

males (68.5%) who had low vegetable intake (65.2%) and occasionally exhibited stool stains (66.3%), but their stool consistency was mainly normal (47.8%). Further examination through bivariate analysis revealed a significant negative correlation between stool stains and SOAVE (name of the condition). Similar findings were observed for stool consistency, indicating a significant correlation. Consequently, both variables were subjected to multivariate analysis. The multivariate analysis demonstrated a significant association between stool consistency and SOAVE among patients aged 2-12 months ( $p=0.000$ ,  $OR=2.044E-7$ ) and those aged 1-5 years ( $p=0.000$ ,  $OR=2.044E-7$ ). An odds ratio (OR) value of less than 1 implies that individuals with normal stool consistency have a lower likelihood of experiencing SOAVE at a young age.

**Conclusions:** This study showed that children with HD correlated with low daily vegetable intake. Normal feces were associated with higher SOAVE ages and a lower possibility of experiencing SOAVE at a young age.

**Keywords:** children, functional outcome, Hirschsprung disease, temporary colostomy.

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### BACKGROUND

Hirschsprung disease (HD) is a rare congenital anomaly (1 in 5000 live births) characterized by varying levels of aganglionosis.<sup>1,2</sup> The presence of a genetic basis for HD has been proposed for some time, evident from its familial occurrence in 5-20% of patients and its association with specific syndromes. Recent research has further confirmed this genetic link by identifying coding mutations in the RET gene, accounting for 7-25% of sporadic cases and 40-60% of familial cases of HD. Additionally, mutations in several other genes have also been associated with HD, highlighting the complex genetic factors contributing to the development of this condition.<sup>3</sup>

HD is influenced by multiple genes, with at least 20 identified as significant contributors to its pathogenesis. Among them are RET, GDNF, GFR $\alpha$ 1, NRTN, EDNRB, ET3, ZFH1B (or ZEB2), PHOX2B, SOX10, IHH, and SHH. These genes code for receptors, ligands, and transcription factors. RET has been recognized as the primary gene responsible for HD, while the other genes collectively account for approximately 30% of all HD cases.<sup>4</sup>

HD typically leads to neonatal bowel obstruction, characterized by delayed passage of meconium, a distended abdomen, and bile-stained vomiting. However, patients may also present later in life with intractable constipation or

experience episodes of Hirschsprung-associated enterocolitis (HAEC).<sup>3</sup>

The evolution of surgical options and outcomes for the successful management of HD has been driven by the aspiration of pediatric surgeons. Pull-through surgery is considered a definitive treatment for HD. Several pull-through surgical approaches have been utilized, including transabdominal endorectal (Soave), Swenson, Duhamel, transanal endorectal pull-through (TEPT), and laparoscopic (Georgeson) approaches. While many studies have reported on the functional outcomes of HD patients following these surgical procedures, there is a scarcity of studies that directly compare the long-term functional outcomes among the

different surgical approaches.<sup>3,5</sup>

TEPT is the most commonly used definitive surgical procedure for patients with HD. In many pediatric surgical centers, intraoperative evaluation to determine the aganglionosis segment during TEPT is commonly performed using frozen section methods. However, certain hospitals do not have access to frozen section facilities and, as a result, rely on contrast enemas to predict the aganglionosis segment preoperatively. This variation in approach is due to the differences in available resources and technologies in different healthcare settings.<sup>6</sup>

The primary surgical approach for HD is to bring the normally innervated bowel down to the anus while ensuring the preservation of sphincter function. However, it is crucial to address functional issues like constipation or fecal incontinence adequately, as these problems can have significant effects on the child's overall development and quality of life.<sup>7</sup>

The postoperative results of Hirschsprung's pull-through procedure could be influenced by surgical and pathological proficiency and the quality of perioperative care. As a result, these outcomes may be associated with the surgeon's or the hospital's experience with the procedure. Controversies persist as to which technique offers the best outcome. Numerous studies have reported on short-term bowel function outcomes following various surgical procedures for HD. The overall consensus among these studies is that short-term outcomes do not significantly differ between the surgical procedures employed. The absence of incontinence and constipation remains the most important marker of a good outcome. After transanal or transabdominal endorectal pull-through surgery for HD, similar rates of long-term bowel dysfunction, such as constipation and incontinence, occur for various reasons.<sup>2,3,8,9</sup>

This study aims to assess long-term functional outcomes in operated cases of HD.

## METHODS

This retrospective observational study involved medical records of HD children who underwent temporary colostomy procedures and were analyzed retrospectively from July 2017 to June 2021. Risk factors were evaluated to determine the outcome of HD procedures.

Inclusion criteria were pathologically proven HD with a maximum age of 16 years and had undergone temporary colostomy procedures. The following information was retrieved from the medical records and via a telephone survey: age, gender, age of colostomy, infant milk, defecation frequency, feces consistency, meconium discharge, stool stain, and weekly vegetable and fruit intake. Patients who were ineligible to participate (deceased, living abroad, had a permanent stoma, or an intellectual disability) were excluded.

The Chi-square Mann – Whitney U test and Kruskal Wallis test were employed. All analyses were performed Bivariate Analysis and Multivariate Analysis with  $P < 0.05$  considered significant.

## RESULTS

Table 1 shows the characteristics of patients included in the study. Ninety-two patients were involved in the study. The study population predominantly consisted of males (68.5%) who had low vegetable

intake (65.2%) and occasionally exhibited stool stains (66.3%), but their stool consistency was mainly normal (47.8%).

Table 2 shows the bivariate analysis of the study. The bivariate analysis using Spearman correlation revealed that gender ( $r = -0.032$ ,  $p = 0.766$ ) and low vegetable intake ( $r = 0.017$ ,  $p = 0.875$ ) were not significantly associated with SOAVE age. Among patients with clean stains, the majority of SOAVE cases occurred in the age group of 2-12 months (46.7%). Similarly, in the stool stain category, most SOAVE cases were also observed in the age group of 2-12 months (52.5%), followed by 31.1% of cases in the >1-5 years age group, 13.1% in the >5-11 years age group, and 3.3% in the >11-16 years age group. In contrast, the permanent category of fecal stains showed that most cases had a SOAVE age of 2-12 months (93.8%). Correlation analysis indicated a significant relationship ( $r = -0.252$ ,  $p = 0.015$ ) with a weak negative correlation between age and permanent fecal stains, suggesting that younger ages were associated with more permanent fecal stains. A significant correlation ( $r = 0.505$ ,  $p = 0.000$ ) was also observed between stool consistency and age. All respondents with abnormal feces had a SOAVE age of 2-12 months (100%), while those with normal feces had a SOAVE age of >1-5 years (38.6%), and some were even >5-11 years old (20.5%) and >11-

**Table 1. Patient Characteristics**

Patient Characteristics	n (%), n = 92
<b>Age</b>	
0-30 days	0 (0%)
2-12 months	54 (58.7%)
>1-5 years	25 (27.2%)
>5-11 years	11 (12%)
>11 – 16 years	2 (2.2%)
<b>Gender</b>	
Male	63 (68.5%)
Female	29 (31.5%)
<b>Low Vegetable Intake</b>	
Low Intake	60 (65.2%)
High Intake	32 (34.8%)
<b>Fecal stain</b>	
Clean	15 (16.3%)
Occasionally	61 (66.3%)
Permanent	16 (17.4%)
<b>Faecal Consistency</b>	
Abnormal	21 (22.8%)
Normal	27 (29.3%)
Frequent	44 (47.8%)

**Table 2. Bivariate Analysis**

Factor	Category	AGE					r (p-value)	p-value
		0-30 days, n (%)	2-12 mo, n (%)	>1-5 yr, n (%)	>5-11 yr, n (%)	>11-16 yr, n (%)		
Gender	Boys	0 (0%)	36 (57.1%)	18 (28.6%)	9 (14.3%)	0 (0%)	-0.032	0.766
	Girl	0 (0%)	18 (62.1%)	7 (24.1%)	2 (6.9%)	2 (6.9%)		
Fecal stain	Clean	0 (0%)	7 (46.7%)	6 (40%)	2 (13.3%)	0 (0%)	-0.252	0.015*
	Occasionally	0 (0%)	32 (52.5%)	19 (31.1%)	8 (13.1%)	2 (3.3%)		
	Permanent	0 (0%)	15 (93.8%)	0 (0%)	1 (6.3%)	0 (0%)		
Low Vegetable Intake	Low Intake	0 (0%)	36 (60%)	15 (25%)	7 (11.7%)	2 (3.3%)	0.017	0.875
	High Intake	0 (0%)	18 (56.3%)	10 (31.3%)	4 (12.5%)	0 (0%)		
Fecal Consistency	Abnormal	0 (0%)	21 (100%)	0 (0%)	0 (0%)	0 (0%)	0.505	<0.001
	Frequent	0 (0%)	17 (63%)	8 (29.6%)	2 (7.4%)	0 (0%)		
	Normal	0 (0%)	16 (36.4%)	17 (38.6%)	9 (20.5%)	2 (4.5%)		

16 years old (4.5%). The correlation was positive and quite strong, indicating that more abnormal feces were associated with lower SOAVE ages, whereas more normal feces were associated with higher SOAVE ages.

Table 3 shows the multivariate analysis conducted in this study.

The stool stain and stool consistency variables underwent multinomial regression analysis (dependent variable >2 categories), with results presented in Table 3. The analysis revealed that fecal stain did not have a significant effect on SOAVE age in the categories of 2-12 years (p=0.770), 1-5 years (p=0.575), and age >5-11 years (p=0.840). However, stool consistency significantly correlated with SOAVE age in 2-12 months (p=0.000, OR =2.044E-7) and 1-5 years (p=0.000, OR =2.044E-7). An odds ratio (OR) value of less than 1 indicates that respondents with normal stool consistency had a reduced likelihood of experiencing SOAVE at a young age.

**DISCUSSION**

The progression of surgical choices to effectively manage HD has been motivated by pediatric surgeons' desire to achieve optimal functional results. Ever since Swenson introduced recto-sigmoidectomy and colo-anal anastomosis as a treatment for HD in 1948, several new techniques have emerged.<sup>10</sup> During the initial three years following surgery, children who undergo new surgical approaches exhibit a reduced occurrence of postoperative complications. Although these definitive surgeries have shown promising short-term outcomes, some HD patients still experience complications such as constipation, fecal incontinence, and enterocolitis, among others, which can negatively impact their quality of life (QoL).<sup>11</sup>

Over the past 17 years since its initial description, the one-stage transanal endorectal pull-through (TERPT) has gained popularity as a surgical option for treating HD. This procedure is preferred due to its less invasive nature, resulting in better cosmetic outcomes, reduced postoperative pain, and fewer complications compared to traditional laparotomy. TERPT is particularly suitable for cases with short-segment aganglionosis

occurring distal to the sigmoid colon and may be performed without laparoscopic assistance or laparotomy. However, despite its advantages, there remains a need for adequate assessment of the postoperative outcomes of TERPT, particularly concerning postoperative anorectal manometry (AM) and electromyography (EMG). Studies have not yet fully explored the incidence of constipation or fecal incontinence after this procedure, and the outcomes in these areas are not always entirely satisfactory.<sup>12</sup>

Early postoperative complications are of significant concern as they can adversely affect long-term functional outcomes. For example, anastomotic leaks and severe strictures can have negative consequences. Studies have indicated that approximately 11% and 14% of patients experience at least one early complication after transanal and transabdominal Soave procedures, respectively. Another study by Thakkar et al. reported early complications in 15% of patients.<sup>3</sup>

Perianal excoriations are a common issue that can typically be managed with ointments. Anastomotic leaks are relatively rare, occurring in 0–5.6% of patients, but can lead to scarring, which negatively impacts long-term function. Strictures have been reported in 4–9% of patients and may require dilation or, in some cases, a redo pull-through. Redo surgery may also be needed to narrow the muscular cuff, twisted pull-through, residual aganglionosis, or transition zone pull-through. Persistent obstructive symptoms in about one-third of patients requiring a redo pull-through can be attributed to residual aganglionosis or transition zone pull-through. The functional outcomes of redo pull-through are somewhat debated, with some reports indicating satisfactory results, while others negatively impact stooling scores and fecal continence. Adhesive small bowel obstruction has also been recently reported in up to 29% of patients with Hirschsprung's disease.<sup>3</sup>

The current study aims to assess long-term functional outcomes after surgery on HD. The characteristics of the study population predominantly consisted of males (68.5%) who had low vegetable intake (65.2%) and occasionally exhibited stool stains (66.3%), but their stool

**Table 3. Multivariate Analysis**

		Parameter Estimates					95% Confidence Interval for Exp(B)		
Age <sup>a</sup>		B	Std. Error	Wald	df	Sig.	Exp(B)	Lower Bound	Upper Bound
2-12 mo	Intercept	47.337	3.671	166.293	1	0.000			
	faecal Stain	0.423	1.445	0.086	1	0.770	1.527	.090	25.909
	faecal Consistency	-15.403	0.754	417.832	1	0.000	2.044E-7	4.667E-8	8.951E-7
1-5 y	Intercept	45.428	3.765	145.587	1	0.000			
	faecal Stain	-0.809	1.443	0.314	1	0.575	.445	.026	7.537
	faecal Consistency	-13.899	0.797	304.090	1	0.000	9.202E-7	1.930E-7	4.389E-6
>5-11 y	Intercept	42.152	3.080	187.274	1	0.000			
	faecal Stain	-0.302	1.499	0.041	1	0.840	.739	.039	13.956
	faecal Consistency	-13.345	0.000	.	1	.	1.601E-6	1.601E-6	1.601E-6

a. The reference category is >11-16 years old.

consistency was mainly normal (47.8%). Gender and vegetable intake did not achieve a significant association with SOAVE age. Similarly, a study conducted by Gunadi found no significant difference in gender.<sup>13</sup>

Regarding the effect of gender on the long-term outcomes of HD, Gunnarsdóttir et al. conducted a study. They found that female sex was associated with lower scores on the Gastrointestinal Quality of Life Index (GIQLI) after performing a multivariate analysis ( $p < 0.05$ ).<sup>14</sup>

Fecal incontinence is a condition that can significantly impact one's quality of life. Patients often experience fear and embarrassment, leading them to avoid social activities and even stay home to avoid accidental bowel leakage. The complexity of the disease's epidemiology arises from varying definitions and reliance on self-reported data. Additionally, fecal incontinence can stem from diverse underlying causes. The initial treatment involves medical and physical therapy, but surgical management may also be necessary. Fecal incontinence can arise from various iatrogenic factors, with anorectal surgery being one of them.<sup>15</sup>

Based on bivariate analysis, correlation analysis indicated a significant relationship ( $r = -0.252$ ,  $p = 0.015$ ) with a weak negative correlation between age and permanent fecal stains, suggesting that younger ages were associated with more permanent fecal stains. The impact of HD on the QoL of affected children compared to healthy children remains uncertain. Collins et al. revealed that children with

HD experienced lower psychosocial QoL scores when compared to their healthy counterparts. These reduced QoL scores were influenced by poorer functional outcomes, particularly concerning fecal incontinence. However, no significant difference was observed in physical and overall QoL outcomes. Interestingly, in children aged between 5 and 7 years, there was an improvement in physical QoL for those with HD, although the reason behind this improvement remains unknown. Numerous studies have supported the finding of reduced psychosocial QoL in HD patients.<sup>16</sup>

Recurring constipation and fecal incontinence can have significant impacts on various aspects of an individual's life, including personal, social, and professional spheres, leading to long-term consequences. In a particular study, patients with HD exhibited lower psychosocial QoL scores compared to healthy children, primarily because of poorer functional outcomes. Interestingly, the children with HD scored lower on QoL than healthy children, but adult patients scored significantly higher than healthy adults. This difference might be attributed to the adult patients having developed better-coping strategies to manage their symptoms compared to the children with HD. Despite experiencing poor functional outcomes, the patients showed comparable overall QoL scores with healthy individuals.<sup>17</sup>

Numerous studies have extensively examined the long-term outcomes of patients who underwent TERPT surgery

for Hirschsprung's disease. These studies have consistently identified high rates of fecal soiling, constipation, and increased stooling frequency compared to control groups, leading to significant social morbidity and impact on the patient's quality of life.<sup>18</sup>

In a review of 2,430 patients who underwent a Duhamel operation, Bourdelat et al. found that the majority of patients achieved good or excellent anal function at a follow-up period of 15–30 years. Only 8.07% of patients experienced constipation, and 5.3% had soiling issues.<sup>19</sup> However, Baillie et al. reported residual postoperative problems in a broader range, affecting 10% to 80% of patients. These problems included soiling, constipation, enterocolitis (EC), and anastomotic strictures. Additionally, other studies have reported incidences of soiling and perineal excoriations ranging from 32% to 35%. Prolonged perineal excoriation indicates persistent soiling and incontinence and may also indicate anastomotic stenosis.<sup>7</sup>

Recent publications have delved into factors that influence functional outcomes after TERPT surgery, with age at the time of surgery being one of the critical factors under investigation. While some studies reported that age at TERPT was not associated with inferior outcomes, others found that patients who underwent TERPT surgery as neonates had worse postoperative functional results compared to those operated on later in life. For instance, Huang et al. identified age less than 2 months at the time of TERPT as a risk factor for early incontinence, late

incontinence, and constipation, but not for Hirschsprung-associated enterocolitis (HAEC) or anastomotic stricture. Another study by Sun et al. compared outcomes between patients younger and older than 3 months at the time of TERPT and found that younger patients had higher rates of perianal erosion, enterocolitis, and fecal soiling.<sup>18</sup>

Interestingly, despite these findings, a survey of pediatric surgeons in Europe revealed that the majority of respondents (87.5%) still performed TERPT surgery before 3 months of age. The highlight need for further research and consensus among medical professionals regarding the optimal timing of TERPT surgery to improve long-term functional outcomes and quality of life for Hirschsprung's disease patients.<sup>18</sup>

According to a study by Aworanti, following an endorectal pull-through procedure, incontinence scores were found to have a positive correlation with the duration of follow-up. In contrast, constipation scores did not show a significant correlation with the duration of follow-up. Clinically, this suggests that improved continence can be expected to improve over time, while constipation tends to persist as an ongoing issue even after the procedure.<sup>10</sup>

Incontinence after a pull-through procedure can be attributed to various factors. These include damage to the internal sphincter and pelvic nerves, sensory impairments in the anal skin, diminished rectal reservoir for stool storage, heightened frequency of muscular contractions in the colon, and colonic contractions extending to the anal canal due to the absence of the rectum.<sup>20</sup>

The observed phenomenon can be explained by the fact that specific mechanisms responsible for maintaining continence, such as autonomic and somatic anal canal innervations for adequate resting anal tone, reflex and voluntary tightening of the external sphincter, and the ability to distinguish between solid, liquid, and gaseous rectal contents, might have been disrupted or compromised during the surgical procedure. However, these mechanisms are not destroyed, allowing for some degree of continence. Due to these disruptions, toilet training

may not be successful at the expected age, and additional supportive measures are required to maintain continence. As these children grow older and become more socially aware, their desire and motivation to remain continent increase. This motivation leads to better control over the external sphincter muscle and an improved ability to differentiate between different types of fecal matter, resulting in a reduced frequency of soiling accidents. Therefore, the study proposes that any damage to the continence mechanisms that may have occurred during the pull-through surgery in infancy is often reversible and might not become apparent until toilet training begins. At this point, supportive measures should be introduced to help these incompletely continent children achieve continence and prevent problems from escalating. By doing so, their overall long-term quality of life can be improved.<sup>10</sup>

Regarding the functional outcomes of Hirschsprung's disease (HD) in adult patients, constipation (either obstructed defecation or slow transit constipation) and fecal incontinence are the most common long-term complications, as reported by Heikkinen et al. In their study of 102 patients, impaired continence was observed in 45% of cases, while constipation affected 5% of patients.<sup>21</sup> A Swedish study examined the bowel function and gastrointestinal quality of life (GI QoL) in 92 individuals who had undergone HD surgery during childhood, comparing them to matched controls. The patients with HD exhibited a significantly deteriorated bowel function score (17.1 vs. 19.1,  $p > 0.0001$ ) with increasing difficulties in controlling defecation, fecal soiling, constipation, and social problems related to bowel function. The fecal incontinence rate was 14%. While gastrointestinal QoL was impacted, it was not significantly lower than controls (121 vs. 125,  $p = 0.0578$ ). Age was associated with poorer bowel function (OR, 1.07;  $p = 0.049$ ), with the total HAQL scores deteriorating by 4 points per year in another series.<sup>22</sup> A recent study investigating bowel function from childhood to adulthood after transanal endorectal pull-through observed a significant improvement in fecal control with increasing age in their

young population (median age, 15). However, adult patients still experienced abnormal stooling frequency (mainly increased) compared to controls.<sup>14</sup>

A study by Davidson et al. observed more favorable functional outcomes in adult patients with HD than children within the same cohort. This observation aligns with previous studies that have indicated improved bowel functional outcomes and better quality of life with age in HD patients. However, it is essential to note that none of these studies reported on bowel and urological function and quality of life in the same patients. Additionally, there is a lack of research reporting data on HD patients from childhood to adulthood within a single study. Further investigation is needed to comprehensively understand the long-term functional outcomes and quality of life in individuals with HD across different age groups.<sup>23</sup>

The TERPT operation is an excellent option for treating most classic cases of Hirschsprung's disease (HD) with favorable results, particularly in younger children. Although it is feasible in older children, the follow-up fecal continence outcomes for these older patients are statistically lower compared to those who undergo the operation at a younger age. Given these findings, it is advisable to initiate the diagnosis and consider surgery for HD patients at a younger age, especially when TERPT is deemed to be the suitable surgical option. Starting treatment at a younger age can help minimize the incidence of preoperative recurrent enterocolitis and chronic constipation, leading to smoother surgery with fewer complications. Early intervention can improve long-term outcomes and overall quality of life for patients with Hirschsprung's disease.<sup>12</sup>

The limitation of this study was the cross-sectional design. A longitudinal design may be more suitable for analyzing long-term functional outcomes.

## CONCLUSION

To date, several surgical method options have emerged to manage HD effectively. Although these definitive surgeries have shown promising short-term outcomes, some HD patients still experience long term complications which negatively

impact their quality of life (QoL). In this study, we assess the long-term functional outcomes in operated HD cases. Mostly, the patients had low vegetable intake and occasionally exhibited stool stains, but their stool consistency was normal. Normal feces were associated with higher SOAVE ages and had a lower possibility of experiencing SOAVE at a young age. Pediatric surgeons should strive to make sure their patients have normal bowel movements. This is important because problems with bowel function can make it hard for children with HD to live an everyday life (impaired QoL). HD can affect the child's mental and emotional health (psychosocial well-being) as well as their physical health. The involvement of a psychologist or other mental health professional in the long-term care plan for children with HD may help improve their social and emotional well-being.

## AUTHORS' CONTRIBUTIONS

All authors contributed equally.

## ETHICAL APPROVAL

This research has been approved by the Ethical Committee, Research Center, Faculty of Medicine, Lambung Mangkurat University, Banjarmasin, Indonesia.

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## CONFLICT OF INTEREST

The authors declare no conflict of interest.

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