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Original article

Correlation between quality of life and gender, age, aganglionosis level, and surgical technique in pediatric patients with Hirschsprung disease post pull-through at Dr. Wahidin Sudirohusodo Hospital, Makassar

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Abstract. *Background and Aim:* Hirschsprung disease (HD) is a congenital disorder characterized by the absence of ganglion cells in the Auerbach and Meissner plexuses, leading to functional bowel obstruction. The pull-through procedure, including the Duhamel, Swenson, and Soave techniques, is the standard surgical treatment. However, postoperative complication remain prevalent, significantly impacting patients' quality of life (QoL). This study aims to evaluate the impact of aganglionosis level and surgical techniques on the QoL of HD patients using the PedsQL Gastrointestinal Scale. *Methods:* This retrospective cohort study included 400 HD patients who underwent pull-through surgery from January 2018 to December 2022 at Wahidin Sudirohusodo Hospital, Makassar. Data were collected from medical records, and QoL was assessed using the PedsQL GI questionnaire and statistical analyses were performed Chi-square tests. *Results:* A total of 400 patients were analyzed. The majority were male (63.7%) and had ultra-short segment aganglionosis (52.5%). The Duhamel technique was the most commonly used (49.0%). QoL was significantly associatied with gender (p=0.023) and aganglionosis level (p=0.001), but not with age (p=0.118) of surgical technique (p=0.489). Patients with good QoL were predominantly male and had ultra-short segment aganglionosis. *Conclusions*: gender and aganglionosis level significantly impact QoL in HD patients post pull-through, while age and surgical technique do not. Regular QoL assessments using PedsQL GI may improve postoperative patient management. (www.actabiomedica.it)

Key words: Hirschsprung disease, pull-through surgery, quality of life, PedsQL gastrointestinal, surgical technique

Introduction

Hirschsprung disease (HD) is a hereditary condition characterized by the absence of ganglion cells in the enteric nervous system, which affects the lower gastrointestinal tract (1). This deficiency leads to functional bowel obstruction, which clinically presents as delayed passage of meconium, abdominal distension, and persistent constipation. HD occurs in approximately 1 in 5,000 live births, with a higher prevalence in males at a ratio of 4:1, and is associated with genetic conditions such as Down syndrome (2). The primary treatment for HD is surgical resection of the aganglionic segment, followed by pull-through anastomosis.

Common surgical techniques include Swenson, Duhamel, and Soave procedures, which aim to restore bowel continuity and functionality. However, postoperative outcomes vary significantly depending on factors such as the extent of aganglionosis and the surgical technique used (3). While advancements in surgical techniques have reduced morbidity, patients often experience persistent issues such as fecal incontinence, constipation, and enterocolitis, impacting their quality of life (QoL). The extent of ganglionosis, ranging from ultra-short to total colonic, may exacerbate these outcomes, with long-segment aganglionosis posing greater surgical and functional challenges (4). This study aims to evaluate the impact of aganglionosis levels and surgical techniques on the QoL of HD patients using the PedsQL gastrointestinal Scale. By analyzing data from a five-year cohort, we hope to provide insights into factors influencing postoperative recovery and long-term patient well-being.

Patients and methods

Study design and settings

This retrospective cohort study included 400 pediatric patients with Hirschsprung disease (HD) who underwent pull-through surgery at Dr. Wahidin Sudirohusodo Hospital between January 2018 and December 2022. The inclusion criteria for the sample were pediatric patients with Hirschsprung disease (HD) who had undergone pull-through surgery, had no additional congenital anomalies, and were willing to participate in the study. Diagnosis of HD in this study relies on a range of clinical symptoms, including abdominal distension, vomiting, delayed passage of meconium (occurring beyond the first 24-48 hours of life), explosive passage of stool or gas, constipation, inadequate growth, abdominal X-rays and the findings from rectal biopsy, as recorded in the medical records. Age, gender, and surgical technique data were obtained from medical records, while patient quality of life was assessed through interviews using the PedsQL Gastrointestinal Scale questionnaire. The researchers obtained permission to use the questionnaire from MAPI Research Trust as the license holder of the PedsQL Gastrointestinal Scale questionnaire. The questionnaire was then adapted into Indonesian by the research team and tested on 30 individuals with characteristics similar to those of the research sample. The adapted questionnaire was completed independently by the patients, accompanied by their parents. The Cronbach coefficient test showed a result of 0.957, indicating that the questionnaire used has good validity and reliability. This study and use of the PedsQL Gastrointestinal Scale Indonesia version have been approved by the Ethics Commission of the Faculty of Medicine, University of Hasanuddin-Dr. Wahidin Sudirohusodo Hospital Makassar (protocol no.: UH24040230; approval no.: 300/UN4.6.4.5.31/PP36/2024) on March 3, 2023.

Data analysis

All data were analyzed using SPSS software (version 22.0; IBM Corp., Armonk, NY, USA). The frequency distribution and Chi-square statistical tests were used as statistical methods. The Chi-square test used to determine whether two categorical variables are correlated (nominal or ordinal scale). If the P value of the test was <0.05, the results were statistically significant.

Results

A total of 400 patients were analyzed. The majority were male (63.7%) and had ultra-short segment aganglionosis (52.5%). The Duhamel technique was the most commonly used (49.0%). were included in this study. The study participants' characteristics are shown in detail in Table 1.

Analysis of factors influencing quality of life

The analysis examined the relationship between patient characteristics and quality of life outcomes, categorized as 'Good' (Excellent and Good) or 'Not Good' (Fair and Poor). Statistical analysis indicated that age was not significantly associated with the quality of life of Hirschsprung disease patients post pull-through ($p = 0.118$). However, most patients with a

good quality of life were under one year old, while those with a poor quality of life were predominantly between 6 and 12 years old, as shown in Table 2.

Male patients were more likely to have a good quality of life (65%) compared to female patients (35%). Among those with a 'Not Good' quality of life, 60% were female, while 40% were male. The relationship between gender and quality of life was statistically significant (p = 0.023), as shown in Table 3.

Patients with ultra-short segment aganglionosis had the highest percentage of good quality of life (51.3%). Patients with long-segment and total colonic aganglionosis had the highest proportions of "Not Good" quality of life (27.5% and 25%, respectively). A significant relationship was observed between aganglionosis level and quality of life (p = 0.001). as shown in Table 4.

The most commonly used pull-through techniques at Dr. Wahidin Sudirohusodo Hospital are the Swenson and Duhamel techniques, while the Soave technique was performed in only a small percentage of patients (3.2%). Statistical analysis revealed no significant relationship between surgical technique and quality of life (p = 0.489). as shown in Table 5.

Table 1. Participants Characteristics.

Variable	n	%
Age		
< 1 year old	156	38,0
1-5 years old	115	28,7
6-12 years old	111	27,8
13-18 years old	18	4,5
Gender		
Male	255	63,7
Female	145	36,3
Aganglionosis Level		
Ultra-short Segment	210	52,5
Short Segment	165	41,3
Long Segment	14	3,5
Total colonic	11	2,8
Pull-through techniques		
Swenson	192	48,0
Duhamel	196	49,0
Soave	12	3,0
Quality of Life		
Excellent	334	83,5
Good	26	6,5
Fair	27	6,7
Poor	13	3,3

Discussion

This study highlights the critical role of aganglionosis levels in determining QoL among HD patients post pull-through surgery. Patients with ultra-short and short-segment aganglionosis exhibited significantly better QoL scores compared to those with long-segment or total colonic aganglionosis. These findings align with previous studies suggesting that longer aganglionosis levels are associated with more extensive surgical interventions and greater postoperative complications.

Table 2. Age and Quality of Life.

Variable	Good QoL n (%)	Not Good QoL n (%)	P-Value
Age			
< 1 year old	151 (39,7)	5 (25,0)	
1-5 years old	110 (28,9)	5 (25,0)	0,118*
6-12 years old	101 (26,6)	10 (50,0)	
13-18 years old	18 (4,7)	0 (0,0)	

*Statistically significant at p < 0.05

Table 3. Gender and Quality of Life.

Variable	Good QoL n (%)	Not Good QoL n (%)	P-Value
Gender			
Male	247 (65,0)	8 (40,0)	0,023*
Female	133 (35,0)	12 (60,0)	

*Statistically significant at $p < 0.05$

Table 4. Level of Aganglionosis and Quality of Life.

Variable	Good QoL n (%)	Not Good QoL n (%)	P-Value
Aganglionosis Level			
Ultra-short Segment	195 (51,3)	15 (75,0)	
Short Segment	161 (42,4)	4 (20,0)	0,001*
Long Segment	3 (3,4)	11 (5,0)	
Total colonic	1 (2,9)	10 (0,0)	

*Statistically significant at $p < 0.05$

Table 5. Surgical Technique and Quality of Life.

Variable	Good QoL n (%)	Not Good QoL n (%)	P-Value
Pull-through techniques			
Swenson	184 (48,4)	8 (40,0)	
Duhamel	184 (48,4)	12 (60,0)	0,489
Soave	12 (3,2)	0 (0,0)	

*Statistically significant at $p < 0.05$

Impact of aganglionosis level:

Patients with long-segment and total colonic aganglionosis often require extensive bowel resections, which can lead to reduced bowel length and impaired absorption. These outcomes may result in chronic diarrhea, malnutrition, and electrolyte imbalances, contributing to poorer QoL. Additionally, the likelihood of postoperative enterocolitis increases with longer aganglionosis segments, further affecting recovery and QoL. Conversely, patients with ultra-short and short-segment aganglionosis generally have better preserved bowel function, leading to improved long-term outcomes (5).

Surgical techniques and qol:

The lack of significant differences in QoL outcomes between Swenson, Duhamel, and Soave procedures in this study indicates that surgical expertise

and postoperative care may play a more critical role than the specific technique used. While the Duhamel procedure is often preferred for its simplicity and reduced risk of pelvic nerve injury, the Swenson and Soave techniques also yield comparable outcomes when performed by experienced surgeons (6). However, complications such as anastomotic strictures, fecal incontinence, and constipation remain prevalent across all techniques, highlighting the need for tailored postoperative management (7).

Psychosocial and functional aspect

HD significantly impacts psychosocial well-being, particularly in school-aged children and adolescents who face stigma related to fecal incontinence or frequent bowel movements. QoL is influenced not only by surgical outcomes but also by social support, access to healthcare, and patient education on bowel management (8).

Conclusion

This study demonstrates that the level of aganglionosis significantly influences QoL in HD patients post pull-through surgery, with poorer outcomes observed in patients with long-segment or total colonic aganglionosis. In contrast, surgical technique does not appear to significantly affect QoL, underscoring the importance of skilled surgical intervention and comprehensive postoperative care. To optimize outcomes, early diagnosis and intervention for HD are crucial. Tailored management plans addressing both surgical and psychosocial needs should be implemented. Future research should focus on long-term functional outcomes, the role of multidisciplinary care, and the psychosocial impact of HD on patients and their families.

Ethic approval: This study received ethical approval from the Research Ethics Committee of Hasanuddin University with approval number 300/UN4.6.4.5.31/PP36/2024.

Conflict of interest: Each author declares that he or she has no commercial associations (e.g. consultancies, stock ownership, equity interest, patent/licensing arrangement etc.) that might pose a conflict of interest in connection with the submitted article.

Authors Contribution: All authors contributed significantly to the study design, data collection, analysis, and manuscript preparation. AN: contributed to the study design and data collection. S, TRH, NM, FH, MM and SRL: guided the manuscript writing. AW and FN: were responsible for data analysis.

Declaration on the Use of AI: None

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