ORIGINAL ARTICLE

Does the Type of Pull-Through Procedure Affect Quality of Life in Children with Hirschsprung Disease? A Comparison of Duhamel and Soave Procedures

Dian Aldira Indrajati[1], Krisna Dwiantama[1], Supriyati[2], Kristy Iskandar[3], Clara Wenda Aginta Sembiring1, Nadia Shafira Dahani[1], Oviani Rezanti[1], Farras Nafauldy[1], Gunadi[1]

[1] Pediatric Surgery Division, Department of Surgery, Faculty of Medicine, Public Health and Nursing, Universitas Gadjah Mada/ Dr. Sardjito Hospital, Yogyakarta, Indonesia
[2] Department of Health Behavior, Environment, and Social Medicine, Faculty of Medicine, Public Health, and Nursing, Universitas Gadjah Mada, Yogyakarta, Indonesia
[3] Department of Child Health, Faculty of Medicine, Nursing and Public Health, Universitas Gadjah Mada/UGM Academic Hospital, Yogyakarta, Indonesia

ABSTRACT

Introduction: Hirschsprung disease (HSCR) is characterized by the absence of ganglion cells in the Auerbach and Meissner plexuses of the colon, leading to functional obstruction. Most studies focus on the functional outcomes of surgical procedures, but quality of life remains under researched. We used the Pediatric Quality of Life Inventory (PedsQL) to assess patients' quality of life (QoL). We aimed to compare the quality of life in HSCR patients after Duhamel and Soave procedures at our institution. **Method:** Mixed-method explanatory study in HSCR patients post Duhamel and Soave at our hospital from January 2012 to December 2018 by accessing medical records. The study collected quantitative data using the PedsQL questionnaire and qualitative data from parents of patients. **Result:** Our study analysed the quality of life of nine Duhamel and five Soave subjects and found no difference in their scores. However, patients with high quality-of-life scores were influenced by family and environmental factors such as supervision, diet management, encouragement, and independence training. **Conclusion:** HSCR patients after Duhamel surgery had a quality of life similar to that of Soave patients, but the presence of family roles and support improved their quality of life after the Duhamel procedure.
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Corresponding Author:
Gunadi, PhD
Email: drgunadi@ugm.ac.id
Tel: +62 878-8574-4234

INTRODUCTION

Hirschsprung disease (HSCR) is a congenital neurointestinal disorder characterized by the absence of ganglion cells in the gut, leading to functional obstruction. Most patients are diagnosed neonatally, with the main symptoms being failure to pass meconium within 24 hours of birth, constipation, abdominal distension, and vomiting (1). The incidence of HSCR is approximately 1:5000 births, with a higher male-to-female ratio (2).

The primary management of HSCR is pull-through surgery, such as the Duhamel and Soave procedures, which aim to remove the aganglionic segment and connect the ganglionic bowel to the anus. Although surgery is effective, postoperative complications, including constipation, incontinence, and enterocolitis, are common and affect patients' quality of life (3).

Most HSCR research focuses on functional outcomes, while patient quality of life remains understudied. Hirschsprung's disease/Anorectal Malformation Quality of Life (HAQL) measures the impact of disease and treatment on patients' lives (4); while the Pediatric Quality of Life Inventory (PedsQL) is a practical questionnaire with 23 questions across four dimensions (physical, emotional, social, and school) that has been used in the HSCR population (5).

A previous study at our institution used the Hirschsprung's disease/Anorectal Malformation Quality of Life (HAQL) questionnaire, a 40-item instrument with 9 dimensions. This questionnaire is more complex than the PedsQL, which makes it difficult to maintain full focus when completing it (6,7). In contrast, a widely validated instrument such as the PedsQL may offer a more practical

and standardized way to assess quality of life in patients with HSCR. Therefore, our study aimed to compare the quality of life in HSCR patients after Duhamel and Soave procedures at our institution

MATERIALS AND METHODS

Study Design

This study utilized an explanatory mixed-methods design, combining a retrospective cohort study with qualitative data to explain the quantitative results. It compare the quality of life of HSCR patients after Duhamel and Soave procedures. The study was conducted at our institution from April to August 2019.

The subjects were patients with HSCR who underwent Duhamel and Soave procedures at our hospital from January 2012 to December 2018, aged 3-18 years at the time of assessment, and without a stoma. Patients with incomplete medical records, who refused to participate in the study, and who could not be contacted/reached were excluded.

In addition to the quantitative assessment using the PedsQL questionnaire, qualitative data were obtained through semi-structured interviews with parents of selected patients. The interviews aimed to explore parental experiences regarding the child's postoperative condition, emotional responses, follow-up care, and family support in daily activities.

Statistical Analysis

According to PedsQL, this instrument uses a 5-point Likert scale, ranging from 0 (Never) to 4 (Almost always). All items have equal weighting, meaning no specific item receives additional weight in the scoring process. The scores are then transformed to a 0–100 scale to facilitate easier interpretation of the results. The scoring procedure is conducted in several steps. Step 1 is score transformation, where each item is reverse scored and linearly transformed to a 0–100 scale with the following conversion: 0 = 100, 1 = 75, 2 = 50, 3 = 25, and 4 = 0. Step 2 involves calculating scores by dimensions. questionnaires with 50% or more completed questions in each domain were considered valid for calculation. Higher scores indicate a good quality of life. The results are reported as the total score (average of answers to questions across all four domains), the physical score (average of answers to questions in the physical domain), and the psychosocial score (average of answers to questions in the emotional, social, and school domains). Statistical methods used were the nonparametric Mann-Whitney test to compare the two surgical procedures, and correlation tests were performed to examine the relationship between parent-reported and child-reported scores. Available qualitative data were used to support the quantitative findings.

RESULTS

Subject Characteristics

Data were collected from the medical records of postoperative HSCR patients who underwent Duhamel or Soave procedures at our hospital from 2012 to 2018, including 53 Duhamel patients and 179 Soave patients, including transanal procedures. Data collected included patient identity, type of surgery, and type of HSCR. After selection based on inclusion and exclusion criteria, each group consisted of 22 patients.

We contacted the patients' parents by phone or home visit. In the Duhamel group, one patient refused, and 12 could not be contacted, while in the Soave group, two patients had no contact in the medical record, and 15 could not be contacted. Finally, the study included only 14 patients, comprising nine Duhamel patients (64%) and five Soave patients (36%) (Table I).

Table I: Characteristics of subjects of this study.

	Duhamel n (%)	Soave n (%)
Age at interview		
3 – 4 years old	1 (11.1)	
5 – 7 years old	2 (22.2)	4 (75)
8 – 12 years old	3 (33.3)	1 (25)
13 – < 18 years old	3 (33.3)	
Age at pull-through procedure		
< 1 year old		1 (25)
1 – 3 years old	1 (11.1)	4 (75)
4 – 5 years old	2 (22.2)	
> 5 years old	6 (66.7)	
Sex		
Male	8 (88.9)	5 (100)
Female	1 (11.1)	
Time span from surgery to interview		
< 3 years	7 (77.8)	
≥ 3 years	2 (22.2)	5 (100)
Type of aganglionosis		
Short	8 (88.9)	5 (100)
Long	1 (11.1)	
Type of questionnaire		
Parent reports (toddlers, children, adolescents)	9 (100)	5 (100)
Child reports (children, adolescents)	7 (77.8)	4 (80)

Most of the study participants were male, 89% in the Duhamel group and 100% in the Soave group. Duhamel patients were mainly aged 8-12 years and 13-<18 years, while Soave patients were predominantly aged 5-7 years. The mean age at surgery was older in the Duhamel group (8 years 4 months) than in the Soave group (1 year 6 months). After surgery, the mean time before questionnaire completion was longer in

Soave patients (5 years) than in Duhamel (2 years and 2 months). The majority of patients in both groups had short-segment HSCR.

All parental questionnaires were completed and returned, while countable child questionnaires were 88% in the Duhamel group and 80% in the Soave group. A few children were unable to complete the questionnaire, either due to unsuccessful contact attempts or because they were considered unable to respond to the questions. Due to the small sample size, qualitative interviews were conducted with four parents from each group, selected based on their highest and lowest quality-of-life scores. The highest-scoring Duhamel patient was a boy of 7 years 5 months who was operated on at 6 years 2 months, while the highest-scoring Soave patient was a boy of 5 years 11 months who was operated on at 1 year 3 months. The lowest-scoring Duhamel patient was a girl of 15 years 8 months who was operated on at 12 years 10 months, while the lowest-scoring Soave patient was a boy of 8 years 11 months who was operated on at 2 years 5 months.

This interview was conducted to better understand the patient's quality of life after surgery, including soiling and obstruction, as well as treatment, including special diets.

Quality of Life

Quality-of-life analysis was conducted by comparing total, physical, and psychosocial scores (including emotional, social, and school) between the Duhamel and Soave groups. Due to the small sample size, qualitative data were used to complement the quantitative results. In the main manuscript, PedsQL results are presented by domain.

Total and psychosocial scores, including emotional and social scores, were higher in the Duhamel group than in the Soave group, as reported by both parents and children. However, the difference was not statistically significant ($p > 0.05$) (Table II).

In the Duhamel group, there was a strong relationship between parent reports and child reports in the physical and school dimensions. Meanwhile, in the Soave group, the strongest correlation occurred in the emotion dimension. All correlations found were very strong and unidirectional (Table III).

Table II: PedsQL score between the Duhamel and Soave groups.

	Duhamel	Soave	*p*-value
	Median (IQR)	Median (IQR)	
Parent reports			
Total score	93.06 (89.13 – 95.65)	89.13 (88.04 – 89.13)	0.495
Physical score	93.75 (93,75– 100.00)	93.75 (93.75 – 96.88)	0.797
Psycho-social score	93.33 (85.00 – 96.67)	86.67 (81.67 – 86.67)	0.606
a. Emotion	90.00 (85.00 – 95.00)	80.00 (80.00 – 85.00)	0.240
b. Social	100.00 (90.00– 100.00)	95.00 (85.00– 100.00)	0.606
c. School	90.00 (77.50 – 97.50)	90.00 (80.00 – 95.00)	0.958
Child reports			
Total score	91.30 (88.32 – 95.65)	83.25 (78.14 – 89.13)	0.315
Physical score	93.75 (90.63– 100.00)	93.75 (87.50– 100.00)	1.000
Psycho-social score	93.33 (82.92 – 94.17)	77.50 (72.92 – 83.33)	0.230
a. Emotion	90.00 (72.50– 100.00)	72.50 (65.00 – 78.75)	0.214
b. Social	100.00 (85.00– 100.00)	80.00 (67.50 – 90.00)	0.109
c. School	82.50 (80.00 – 96.25)	85.00 (80.00 – 92.50)	0.706

The domain-level PedsQL scores in Table II suggested slightly better psychosocial functioning in the Duhamel group, particularly in the emotional and social domains, although these differences were small and did not reach statistically significance ($p > 0.05$).
Qualitative data showed that patients with a better quality of life tended to have higher spirits and more positive emotions. The Duhamel patient 1, for example, was described as obedient, cheerful, and healthy. In contrast, the Duhamel patient 2 patients with a lower quality of life still often complained of pain in the suture scar due to infection after surgery.

Table III: PedsQL Score Correlation between Parent and Child Reports.

Dimension	Duhamel				Soave			
	Parent reports (n = 9)	Child reports (n = 7)	r	p-value	Parent reports (n = 5)	Child reports (n = 4)	r	p-value
	Median	Median			Median	Median		
Physical	93.75	93.75	0.86	<0.01*	93.75	93.75	0.00	0.500
Emotion	90.00	90.00	0.57	0.093	80.00	72.50	0.99	<0.01*
Social	100.00	100.00	0.40	0.188	95.00	80.00	0.46	0.269
School	90.00	82.50	0.88	0.01*	90.00	85.00	0.32	0.340

In the Soave group, patient 2, who had a lower quality of life, experienced emotional changes, becoming more irritable and having difficulty expressing his feelings. He often cried when in pain or when his wishes were not granted. In contrast, patient 1, who had a better quality of life, never got angry and accepted their situation easily.

In terms of adherence to postoperative control, each patient had a different experience. The Duhamel patient 1 stopped control as instructed by the doctor. In contrast, patient 2 stopped on his own initiative after developing an infection due to improper care elsewhere, before returning to the hospital. The Soave patient 1 had regular controls for 5 months to remove the stitches and enlarge the anus using medical devices, but stopped on the parents' initiative, even though the doctor had not advised. The Soave patient 2 had five controls and stopped at the doctor's direction.

Family and environmental conditions play a significant role in patients' quality of life. Patients with better quality of life received close postoperative supervision, emotional support from family, and training in independence, such as managing bowel habits and proper diet. In contrast, patients with lower quality of life, especially in the Soave group, received less support and independence training, which impacted their emotional and social development.

In the Duhamel group, patients' quality of life generally improved with age, except in the 13-<18 years age group. Meanwhile, in the Soave group, there was no consistent pattern of improvement in quality of life. Duhamel patients who underwent surgery after 5 years of age tended to have a better quality of life, while Soave patients showed the best quality of life if surgery was performed before 1 year of age. In addition, the longer the interval from surgery to questionnaire or interview completion, the better the quality of life of Duhamel patients, whereas no similar trend was observed in Soave patients.

DISCUSSION

A previous study at our hospital using the HAQL instrument found that HSCR patients who underwent the Soave procedure had a better quality of life than those who underwent the Duhamel procedure (7). In this study, we evaluated the quality of life of HSCR patients after Duhamel and Soave procedures using the PedsQL questionnaire, a widely validated instrument for assessing patient-reported quality of life. Unlike Saysoo et al.'s findings (7), our findings indicated that Duhamel patients exhibited higher QoL scores than Soave patients, although the difference was not statistically significant. This could be due to psychosocial factors influenced by postoperative functional issues. Qualitative data from Soave patients with a lower quality of life indicated that they sometimes cried due to pain from constipation.

Qualitative interviews with parents suggested that family supervision, emotional support, and environmental factors may influence the postoperative experiences of children with Hirschsprung disease. However, these psychosocial factors were not quantitatively measured in this study and should therefore be interpreted cautiously. These observations are presented as contextual qualitative findings that may help explain the quantitative results rather than as definitive determinants of quality of life.

In a previous study at our hospital comparing functional outcomes in HSCR patients, we found that constipation was more common in Soave patients than in Duhamel patients, with an 8.5-fold higher risk (8). This discrepancy might stem from anastomotic stricture of the rectal muscle sheath in the Soave procedure. Soiling in these cases might be a secondary effect of constipation (pseudoincontinence), but true incontinence could also occur due to rectal damage during the Soave procedure. Conversely, Duhamel patients experienced diarrhea more frequently than those in the Soave group. Another study at our institution by Parahita et al. (9) found that HAEC incidence was higher in Duhamel patients. These functional issues can affect confidence, school attendance, emotional stability, and social relationships. This study also found a strong correlation between parent and child reports on quality of life, but this correlation was inconsistent across different domains and surgical procedures. The agreement level between parent and child reports on the PedsQL was low, with intraclass correlation coefficients ranging from 0.02 to 0.23. The child's age could influence differences across these reports, the specific quality-of-life domain, and even the parents' well-being (10).

Regarding age progression, quality of life in Duhamel patients generally improved with age, except in the 13–<18-year-old group. However, this trend was not observed in Soave patients, whose quality of life tended to decline over time. Due to missing data in the statistical calculations, these findings could not be statistically tested. Research by Gunnarsdyttir and Wester noted that adolescents often experience psychosocial challenges. Even in healthy adolescents, self-confidence and school functioning tend to decline, possibly due to developmental changes or issues related to sexuality (11). On the other hand, functional disturbances tend to improve with age (4). Hartman et al. suggested that by adolescence, patients may have developed better coping strategies to manage previous functional difficulties (12). In this study, neither age at surgery nor the time interval between surgery and questionnaire completion could be analysed statistically due to missing data. However, a previous study reported that the average age at definitive surgery was 50.8 ± 47.5 months for Duhamel patients and 29.9 ± 45.2 months for Soave patients

(9). The Duhamel procedure is generally performed in older patients because the pelvic size is better suited to staplers. In contrast, the Soave procedure can be more challenging in older patients, as the process of separating the mucosal and muscle layers is often more difficult in this age group (13).

A key strength of this study is the use of the Pediatric Quality of Life Inventory (PedsQL), a widely validated and standardized instrument for assessing quality of life in pediatric populations. The use of this questionnaire allowed a more systematic and reliable evaluation of patient-reported outcomes, and contributes to the existing literature, as many previous studies have mainly focused on functional outcomes or used assessment tools that were less specific for this population.

Nevertheless, several limitations of this study should be considered. The final sample size was relatively small (n = 14) due to loss to follow-up from the initial cohort, which may have reduced the statistical power and introduced potential selection bias. Therefore, future studies with larger sample sizes and more complete follow-up are needed to further confirm and strengthen these findings.

CONCLUSION

This study found no statistically significant difference in the quality of life between HSCR patients who underwent Duhamel and Soave procedures. While qualitative data suggests that family support and environmental factors may play a role in improving patient outcomes, particularly in the Duhamel group, these findings are limited by the small sample size. Further large-scale prospective studies are warranted to more definitively assess the impact of surgical techniques and psychosocial support on the long-term quality of life in this population.

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