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Association between functional outcomes with quality of life in Hirschsprung patients after pull trough at Dr. M Djamil General Hospital

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Abstract

Background: Hirschsprung disease is a congenital disorder characterized by the absence of ganglion cells in the Meissner plexus (submucosa) and Auerbach plexus (muscularis) which extend for varying distances proximally. Hirschsprung disease can occur in 1:5000 births. The aim of this study was to determine the Association between functional outcomes and the quality of life in Hirschsprung patients after pullthrough at Dr. M. Djamil General Hospital.

Methods: This study was an analytical descriptive study with a cross sectional approach in patients Hirschsprung's disease after pullthrough in 2023, whose functional outcomes were evaluated using the Krickenbeck classification questionnaire and quality of life using the PedsQL questionnaire. The subjects selected for this study were patients diagnosed with Hirschsprung who underwent Pullthrough surgery at Dr. M. Djamil General Hospital, has sufficient data in medical records, is ≥ 3 years old, can still be contacted, and is willing to be included in the research.

Results: The research sample was 22 (55%) men and 18 (45%) women. With 87.5% of patients having voluntary bowel movements, 12.5% did not experience voluntary bowel movements. There are 2.5% of patients who cannot feel the urge to defecate (soiling) every day, 15% of patients who experience soiling once or twice a week and 82.5% of patients who do not soil. 2 patients (5%) had constipation that required laxatives, 6 patients (15%) had constipation that could be managed with diet and 32 patients (80%) did not experience constipation. There were 8 patients with physical function disorders (20%), 12 patients with emotional function disorders (30%), 7 patients (17.5%) with social function disorders and 0 patients (0%) with school function disorders.

Conclusion: There is a significant association between the functional outcomes of Hirschsprung patients and the quality of life patients after pull through.

Keywords: Hirschsprung, krickenbeck classification, quality of life, PEDSQL

Introduction

Hirschsprung Disease is a congenital disorder characterized by the absence of ganglion cells in the Meissner plexus (submucosa) and Auerbach plexus (muscularis) which extend for varying distances proximally ^[1]. It is caused by the cessation of craniocaudal migration of neural crest cells in the distal colon area in fifth to twelfth week of pregnancy to form the intestinal nervous system ^[2].

World Health Organization estimates that around 7% of all infant deaths in the world are caused by congenital abnormalities. Hirschsprung's disease can occur in 1:5000 births. The highest risk of Hirschsprung's disease is usually in patients who have a family history of Hirschsprung's disease and in patients with Down syndrome ^[1, 3].

Management of Hirschsprung disease consists of non-surgical and surgical procedures with almost all Hirschsprung cases requiring surgery. The surgical procedure can be carried out in one stage or two stages. Some of the pullthrough surgical procedures that are frequently performed are the Swenson's sigmoidectomy procedure, the Duhamel procedure, the Soave's Transanal Endorectal Pullthrough procedure ^[1-6]. Even though the pullthrough is considered effective, several post-operative functional outcomes can still arise which become social problems that can affect the patient's quality of life.

The Krickenbeck classification is considered the main postoperative parameter to evaluate the success of surgery based on spontaneous bowel movements, soiling and constipation ^[11, 14].

The Krickenbeck classification was developed based on a classification system by an international conference for standard development of management and assessment of functional outcomes in anorectal malformation patients, but its widespread use can be used to assess post-operative outcomes of Hirschsprung patients [4].

Quality of life is the subjective health status of a patient and measures the impact of an illness through a doctor's assessment to consider the patient's well-being and progress. In terms of assessing the quality of life of children with Hirschsprung who have undergone pull through, subjective perception which has an impact on their daily activities such as physical function and social function is an important goal to carry out [5,6].

Based on this background, the author is interested to evaluate association between functional outcomes based on the Krickenbeck classification and the quality of life in Hirschsprung patients after pullthrough using the PedsQL Gastrointestinal questionnaire.

Methods

This research is a descriptive analytical study with a cross-sectional design. It is used primary data from the Krickenbeck classification questionnaire, the PedsQL questionnaire and secondary data from medical records in Hirschsprung patients after pullthrough at Dr. M. Djamil Hospital in 2023. The subjects selected for this study were patients diagnosed with Hirschsprung who underwent Pullthrough surgery at Dr. M. Djamil General Hospital, has sufficient data in medical records, is ≥3 years old, can still be contacted, and is willing to be included in the research. Research data is collected and recorded in a formula. The data is presented in tabular form and data analysis is carried out.

Table 2: Functional Outcomes of Hirschsprung Patients after Pullthrough

Krickenbeck classification	Clinical	Frequency (n)	Percent (%)
Voluntary Bowel Movements	No	5	12,5
	Yes	35	87,5
Soiling	Never	33	82,5
	Grade I (Occasionally, once or twice per week)	6	15
	Grade II (Every Day, no social problems)	1	2,5
	Grade III (Constant/social problems)	0	0
Constipation	Never	32	80
	Grade I, Can be managed with diet	6	15
	Grade II Requires laxatives	2	5
	Grade III Resistant to diet and laxatives	0	0

In this study, it was found that 87.5% of patients had voluntary bowel movements, while 12.5% did not experience voluntary bowel movements. There are 2.5% of patients who cannot feel the urge to defecate/soil (soiling) every day, 15% of patients who experience soiling once or twice a week and 82.5% of patients who experience no soiling. Constipation requiring laxatives was found in 2 patients (5%), while 6 patients (15%) had constipation that could be managed with diet and 32 patients (80%) did not experience constipation. The characteristics of constipation patients based on gender table 3.

Table 3: Distribution of characteristics constipated patients according to gender

Gender	Frequency (n)	Percent (%)
Female	5	62,5
Male	3	37,5
Amount	8	100

Result

The results of the analysis are based on Hirschsprung's disease patients in 2023 from the medical records of pediatric surgery patients at Dr. M. Djamil General Hospital. The medical records found was 135 patients, and those included inclusion criteria were 40 patients.

The distribution of patient characteristics in table 1.

Table 1: Distribution of Patient Characteristics

Gender	Frequency (n)	Percent (%)
Male	22	55
Female	18	45
Amount	40	100
Age	Frequency (n)	Percent (%)
3 years	20	50
4 years	7	17,5
5 years	3	7,5
6 years	3	7,5
7 years	7	17,5
Amount	40	100

The distribution of patients based on gender found that there were more males than females. Of the 40 patients used as research samples, 22 samples were men (55%) compared to 18 women (45%).

Meanwhile, according to age category, the highest frequency was at the age of 3 years (50%) with a mean value of 4.25 and a median of 3.50.

The functional outcomes of patients after Pull through surgery based on the Krickenbeck classification in table 2.

The distribution of subjects according to the Krickenbeck classification results in table 4.

Table 4: Distribution of patients according to Krickenbeck classification results

Krickenbeck classification results	Frequency (n)	Percent (%)
Verry good	33	82,5
Good	7	17,5
Sufficient	0	0
Poor	0	0
Amount	40	100

This study showed very good results in 33 patients (82.5%), good in 7 patients (17.5%), sufficient and poor in 0%.

Quality of life patient based on the PedsQL questionnaire in Table 5.5.

Table 5: Distribution of Patients According to the Patient's Quality of Life based on the PedsQL questionnaire

PedsQL questionnaire	Frequency (n)	Percent (%)
Physical Function	8	20
Emotional function	12	30
Social function	7	17,5
School functions	0	0

There were 8 patients with physical function disorders (20%), 12 patients with emotional function disorders (30%), 7 patients (17.5%) with social function disorders and 0 patients (0%) with school function disorders.

Distribution of Patients According to the results of the interpretation of the patient's Quality of Life based on the PedsQL questionnaire table 6.

Table 6: Distribution of Patients According to the results of interpreting the Quality of Life of patients based on the PedsQL questionnaire Age 2-4 Years

PedsQL questionnaire	Frequency (n)	Percent (%)
Very good	18	66,7
Good	9	33,3
Sufficient	0	0
Poor	0	0
Amount	27	100

There were 27 patients using the PedsQL questionnaire aged under five (2-4 years). The results of the questionnaire showed that the patient's quality of life was very good in 29 patients

(72.5%), good in 11 patients (27.5%), sufficient (0%) and poor (0%).

Table 7: Distribution of Patients According to the results of interpreting the Quality of Life of patients based on the PedsQL questionnaire Age 5-7 Years

PedsQL questionnaire	Frequency (n)	Percent (%)
Very Good	11	84,6
Good	2	15,4
Sufficient	0	0
Poor	0	0
Amount	13	100

The distribution of patients according to the results of the interpretation of patient quality of life based on the PedsQL questionnaire aged 5-7 years, in 13 patients. Interpretation of quality of life was categorized as very good, good, fair and poor. The results of the questionnaire showed that the patient's quality of life was very good in 11 patients (84.6%), good in 2 patients (15.4%), sufficient (0%) and poor (0%). Statistical analysis in this research to know assosiation between the functional

outcomes in hirschprung patients after Pull through using the Krickenbeck classification and the patient's quality of life using the PedsQL questionnaire. Bivariate analysis using chi square can be seen in the SPSS output.

In Chi Square test requirements, the Krickenbeck classification questionnaire results are categorized into very good and good, in the PedsQL questionnaire they are categorized into very good and good. So the following statistical results are obtained.

	Valid		Missing		Total	
	N	Percent	N	Percent	N	Per cent
kategori skor krikckenbec *	40	100.0%	0	0.0%	40	100
Kategori pedsQL						.0%

Fig 1: Case processing summary

Count		Kategori pedsQL		Total
		sangat baik	baik	
kategori skor krikckenbec	sangat baik	27	6	33
	baik	2	5	7
Total		29	11	40

Fig 2: Kategori skor krikckenbec * Kategori peds QL crolltabulation

	Value	df	Asymptotic Significance (2- sided)	Exact Sig. (2- sided)	Exact Sig. (1-sided)
Pearson Chi-Square	8.212 ^a	1	.004		
Continuity Correction ^b	5.759	1	.016		
Likelihood Ratio	7.385	1	.007		

Fig 3: Chi-Square tests

Fisher's Exact Test				.011	.011
Linear-by-Linear Association	8.007	1	.005		
N of Valid Cases	40				
a. 1 cells (25,0%) have expected count less than 5. The minimum expected count is 1,93.					
b. Computed only for a 2x2 table					

In the statistical test with Chi Square, it was found that Asymptotic Significance (2-sided) < 0.005, with a value of 0.000. This can be interpreted to mean that there is a relationship between functional outcomes and the quality of life of Hirschsprung patients after Pull through.

Discussion

Based on the research results, 135 medical records were obtained from after Pullthrough Hirschsprung patients who were admitted to RSUP M Djamil Padang in 2023 in the inclusion criteria being 40 patients. In this study, there were 22 male patients (55%) more than 18 female patients (45%), although the difference was not that significant. This is in accordance with several previous studies that the incidence of Hirschsprung cases is higher in men than women [2, 22].

The patients studied were aged ≥ 3 years because the children are considered capable of toilet training and their functional outcomes can be assessed completely using the Krickenbeck classification [8, 15]. In this study, of the 40 patients studied, 20 patients were aged 3 years old, 7 patients aged 4 years, 3 patients aged 5 years and 3 patients aged 6 years and 7 patients aged 7 years, with a mean age of 4.2 years.

The results of the functional outcomes of patients after pullthrough, there were 35 (87.5%) with voluntary bowel movements and a small percentage without voluntary bowel movements 5 (12.5%). This finding is due to a good post-operative wound healing process and minimal anastomotic strictures that occur in the rectal muscle cuff after the pullthrough procedure [23]. In this study, a low incidence of soiling was found with never soiling 33 (82.5%), grade I was 6 (15%) and grade II was 1 (2.5%). This is in accordance with the literature which explains that after pullthrough Hirschsprung patients can experience overflow incontinence due to constipation because the pullthrough procedure allows damage to the anal canal. In previous research it was also found that soiling events could be caused by enterocolitis [15, 24-26].

Constipation after the pullthrough procedure in this study was 8 patients (20%) lower than (25%) in the previous study. This constipation may occur because the aganglionosis in the proximal colon is not completely removed or the external anal sphincter has not been properly relaxed during the pullthrough

procedure. Female patients also have a higher risk of experiencing constipation after pull through than men. This was also found in this study in 5 (62.5%) women and 3 (37.5%) men. In the general population without other gastrointestinal disorders, it has been proven that women have a higher rate of constipation due to hormonal factors [22]. In women who enter puberty, symptoms of constipation become difficult to assess because constipation is influenced by hormonal factors. However, we do not have data regarding the rate of constipation in after pull-through Hirschsprung children who are or have gone through puberty. Therefore, it would be interesting to conduct a cohort study to compare the frequency of constipation between female and male adolescent Hirschsprung patients in future studies.

Statistical analysis in this study, it was found a relationship between functional outcomes and the quality of life after pullthrough Hirschsprung patients. This is in accordance with the literature which states that after pullthrough patients have better physical function, symptoms and signs, emotional function, social function and cognitive function. This literature also states that parenting style factors can influence the function and quality of life of patients after pullthrough [25, 26].

The results of this study show that the majority after Pulthorough patients with excellent Krickenbeck classification results also have a very good quality of life. This is consistent with previous literature that parents report better social functioning post-surgery, such as good social relationships with their child's friends [28, 29]. The potential explanation for this finding is that children with chronic illnesses experience a 'response shift', that is, they respond to the new reality they live with the disease and adapt to functional norms. In other words, they are able to shift emotionally and mentally to living with their chronic illness. This theory explains why parents observe better social functioning in their children, based on the parenting style of the children. However, parental responses alone are not the only reliable method for assessing quality of life social functioning in children with chronic illnesses. This is because differences in reported quality of life persist in their children, and a combination of both is recommended [24, 26, 30, 31].

Some, that children with lower Krickenbeck classification results also had a lower quality of life. This may occur possibly

because patients who have not lived long enough with Hirschsprung's disease may not be able to manage their functional symptoms effectively which impacts their quality of life [32].

The results of functional outcomes and quality of life in other studies also show similar things even though their implementation uses clinical definitions for constipation, post-operative obstructive symptoms, and incontinence which vary greatly between studies and are obtained differently based on the data collection methods used [28]. In addition, The pediatric population is heterogeneous as they may have neurological disorders from associated diseases that correlate with increased constipation and incontinence scores [33, 34].

Conclusion

In this study, Patients taken in this study were aged ≥ 3 years, with more males than females.

Some patients feeling absence of voluntary bowel movements, soiling and after pullthrough constipation which affect physical function, emotional function and social function as obtained from the PedsQL questionnaire. However, the PedsQL score results are still in the good and very good categories.

Statistically, it was found that there a relationship between the functional outcomes of after pullthrough patients based on the Krickenbeck classification and the patient's quality of life using the PedsQL questionnaire.

However, perceptual evaluation is also needed in assessing functions that influence children's quality of life with more informants (Children, parents, teachers) in clinical research, to support patients and their families after surgery.

Prospective, multicenter, and longitudinal research are needed with consistent monitoring of functional outcomes and quality of life for children after Pullthrough into adulthood.

Further research on patient quality of life with a larger sample based on surgical technique and time of post-operative quality of life evaluation. And research on the relationship between constipation in Hirschsprung patients with a colon after Pullthrough that is still aganglionic.

Conflict of Interest

Not available

Financial Support

Not available

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