ORIGINAL ARTICLE

Quality of life of Hirschsprung disease patients with a stoma

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ABSTRACT

Introduction: Hirschsprung disease (HSCR) is a genetic disorder leading to gastrointestinal obstruction due to the absence of ganglion cells in the submucosal and myenteric plexuses. Treatment typically involves a pull-through surgery, sometimes starting with a colostomy. The COVID-19 pandemic has resulted in restrictions on general patient services, i.e., non-COVID-19 patients, at health facilities, leading to reduced hospital visits, including HSCR patients with stomas. This study aimed to determine whether there were differences in the quality of life (QoL) of HSCR patients with stomas before and after the COVID-19 pandemic.

Materials and Methods: This research was a descriptive study comparing the quality of life (QoL) of HSCR patients with stomas before and after the COVID-19 pandemic. It utilized a cross-sectional study design and assessed QoL using the PedsQL Generic Core Scales 4.0 questionnaire.

Results: There was no significant difference in the QoL of HSCR patients before COVID-19 and after COVID-19, as indicated by parent reports (p=0.88) and child reports (p=0.12). However, there was a statistically significant difference in scores on the social dimension of child reports (p=0.04). Furthermore, there was no statistically significant relationship between parent and child reports (p>0.05).

Conclusion: The QoL of HSCR patients with a stoma before and after the COVID-19 pandemic is similar, except for the social dimension in the child's report. Further studies with more cases are necessary to clarify the findings of this study.

KEYWORDS:
Hirschsprung disease, stoma, quality of life, COVID-19 pandemic; two stages pull-through surgery

INTRODUCTION

Hirschsprung disease (HSCR) is a genetic disorder characterized by a motility issue that causes functional obstruction of the gastrointestinal system.[1] The pathogenesis of HSCR involves the absence of parasympathetic ganglion cells in the submucosal plexus and the myenteric plexus due to disturbances in the migration of neural crest cells to the abdominal wall.[2] The incidence of HSCR in our country is approximately 1 in 3,250 live births.[3] This rate is higher than those of countries in Asia (1 in 3,600), Europe (1 in 6,700), and Africa (1 in 4,800).[4] Treatment for HSCR involves pull-through surgery, which can be performed in one or two stages. In a two-stage pull-through, the procedure starts with creating a stoma through a colostomy.[5] Stoma can enhance the quality of life (QoL) for HSCR patients. However, creating a stoma comes with the risk of potential complications.[6]

Coronavirus disease 2019 (COVID-19) is caused by transmission of the SARS-CoV-2 virus. COVID-19 was declared a pandemic by the World Health Organization (WHO) on March 11, 2020. As a form of control against SARS-CoV-2, the Indonesian government implemented a large-scale social restrictions policy as regulated in Government Regulation Number 21 of 2020 concerning large-scale social restrictions in the context of accelerating the handling of COVID-19.[8] At this time, health service facilities are also affected. Healthcare facilities are reducing services for non-COVID-19 patients and focusing on providing COVID-19 services. This restriction also caused a decrease in patient visits at our institution, including HSCR patients with stomas. Therefore, this research was conducted to determine whether there was a difference in the QoL of HSCR patients with stomas before and after the COVID-19 pandemic.

MATERIALS AND METHODS

Subjects

This research was a cross-sectional study of a population of HSCR patients with a stoma at our hospital in Indonesia. Subjects were taken based on the following criteria: (1) isolated HSCR patients with a stoma at our hospital from January 2018 to May 2022, (2) patients must be at least 2 years old and a maximum of 18 years old, (3) willing to take part in the research and have obtained informed consent from the patient's parents. Subjects with HSCR syndrome, incomplete medical records, and those who could not be contacted/reached were excluded. This study involved 6 HSCR patients with stoma as a group before COVID-19 and 7 HSCR patients with stoma as a group after COVID-19. The ethical research has been agreed upon by our institution's ethical committee.

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Table I: Characteristics of subjects in the study (N=13)

Characteristics	Before COVID-19 n (%)	After COVID-19 n (%)	p-value
Sex			
▪ Male	4 (66)	4 (57)	0.725
▪ Female	2 (33)	3 (42)	
Age at completing the questionnaire/interview (years old)			
▪ 2-4	1 (16)	4 (57)	0.220
▪ 5-7	4 (66)	1 (14)	
▪ 8-12	1 (16)	1 (14)	
▪ 13-<18	0	1 (14)	
Report type			
▪ Parent reports	6	7	0.797
▪ Child reports	3	3	

Table II: Quality of life scores for patients with HSCR, before and after the COVID-19 pandemic (based on parents' reports)

Variable	Before COVID-19 pandemic		After COVID-19 pandemic		p-value
	N	Score (mean ± SD)	N	Score (mean ± SD)	
PedsQL score	6	88.40 ± 13.48	7	87.57 ± 5.35	0.88

Table III: Quality of life scores for patients with HSCR, before and after the COVID-19 pandemic (based on child's reports)

Variable	Before COVID-19 pandemic		After COVID-19 pandemic		p-value
	N	Score (mean ± SD)	N	Score (mean ± SD)	
PedsQL score	3	77.65 ± 6.8	3	86.95 ± 1.88	0.12

Table IV: Analysis of the differences in PedsQL scores across dimensions based on parents and children's reports

PedsQL scores	Before COVID-19 pandemic (mean ± SD)	After COVID-19 pandemic (mean ± SD)	p-value
Parent reports			
▪ Physical	83.85 ± 22.20	87.49 ± 5.10	0.68
▪ Emotional	81.66 ± 23.38	83.57 ± 12.14	0.85
▪ Social	96.66 ± 4.08	95.00 ± 6.45	0.59
▪ School	94.16 ± 6.64	84.28 ± 15.39	0.17
Child reports			
▪ Physical	85.41 ± 25.20	86.95 ± 1.88	0.50
▪ Emotional	50.00 ± 17.32	81.66 ± 23.62	0.50
▪ Social	83.33 ± 5.77	98.33 ± 2.88	0.04*
▪ School	86.66 ± 15.27	76.66 ± 20.81	0.50

*, p<0.05 is considered significant.

Table V: Correlation of PedsQL scores between parent's and child's reports

PedsQL scores	Before COVID-19				After COVID-19			
	Parent reports Median (IQR) (N=6)	Child reports Median (IQR) (N=3)	p	r	Parent reports Median (IQR) (N=7)	Child reports Median (IQR) (N=3)	p	r
Physical	92.18 (67.18-100)	100(56.2-100)	0.17	0.96	84.37 (84.36-93.75)	93.75 (81.25-93.75)	0.15	0.97
Emotional	92.5 (53.75-100)	40 (40-75)	0.66	0.50	85 (70-90)	90 (55-90)	0.31	0.88
Social	97.5 (93.75-100)	80 (80)	0.67	0.50	100 (90-100)	100 (95-100)	0.66	0.50
School	95 (88.75-100)	90 (70-90)	0.21	0.94	85 (70-100)	70 (60-70)	0.10	0.98

Data collection
Data were obtained from medical records of HSCR patients with stoma at our hospital from January 2018 to March 2020 for the group before the COVID-19 pandemic and April 2020 to May 2022 for the group after the pandemic. The questionnaire was read to the patient and/or the patient's parents after informed consent was obtained from the doctor in charge.

Statistical Analysis
Data are provided in the form of mean, standard deviation, median, and IQR. A higher outcome score indicates a better QoL. The Shapiro-Wilk test was used to perform a normality test. If the data was normally distributed, the independent t-test method was used.

Ethics approval and consent to participate
Our institution's Medical and Health Research Ethics Committee approved this study. The study was conducted in accordance with the Declaration of Helsinki.

RESULTS

Subject Characteristic
In our study, we observed no significant differences in sex, age at the time of completing the questionnaire, or type of report between the groups before and after the COVID-19 pandemic (p>0.05) (Table I).

Quality of life of patients with HSCR before and after the COVID-19 pandemic
According to parents ' and child's reports, the average QoL score for HSCR patients before and after the COVID-19 pandemic was similar. (p=0.88 and 0.12, respectively) (Tables II and III).

Further analysis described how higher physical and emotional scores were obtained in the group after the COVID-19 pandemic, both from parent reports and child report questionnaires. None exhibited significant differences across the dimensions, except for the social dimension in the child reports (p=0.04) (Table IV).

Next, we determined the correlation of PedsQL scores between parent and child reports. None of the dimensions showed any significant differences (p>0.05) (Table V).

DISCUSSION

Our study revealed that there were more male HSCR patients than female patients. This aligns with prior studies, which indicated that the male-to-female ratio among patients was 4:1.[9-11] Concerning the age characteristics of the subjects when completing the questionnaire, the group of HSCR patients aged 5-7 years before the COVID-19 pandemic surpassed those aged 2-4 years and 8-12 years, with a ratio of 4:1:1. Conversely, among the HSCR patients after the COVID-19 pandemic, those aged 2-4 years showed a higher prevalence compared to patients aged 5-7 years, 8-12 years, and 13-<18 years, with a ratio of 4:1:1:1.

The incidence of HSCR in our province is 1 in 3,250 live births. With a population of 4 million and a birth rate of 36,045, or 9 per thousand, it is estimated that 41 babies with HSCR will be born each year in this province.[12-13] The number of HSCR patients at our hospital is higher yearly, as patients come from our province and the southern part of the neighborhood province.

Previously, neonates diagnosed with HSCR had to undergo a colostomy first and wait until they were 6 to 12 months old to have definitive pull-through surgery. However, this approach has become less common over the last three decades, and single-stage pull-through surgery, particularly transanal endorectal pull-through (TEPT), has gained popularity among pediatric surgeons worldwide[14], including in our hospital.[3] In addition, one-stage surgery offers many benefits, including more uncomplicated pre-surgical care, reduced costs, shorter hospital stays, and sparing the patient and their family from the psychological burden of creating a stoma.[15-16] This may explain why fewer HSCR patients with stomas were included in the research sample taken at our hospital.

According to an analysis of the final PedsQL scores reported by parents, the HSCR patient group exhibited a higher mean score following the COVID-19 pandemic. However, the difference between the two groups was not statistically significant. This aligns with a previous study suggesting that children with congenital diseases may already have disorders from the beginning, indicating that the COVID-19 pandemic might not affect children's psychosocial conditions.[17]

In analyzing the physical dimension scores, both groups received moderate classifications. Additionally, the group demonstrated better physical scores in reports from children and parents after the COVID-19 pandemic. However, no significant differences were found. This finding aligns with previous research18, which indicated that there were no significant differences in physical dimension PedsQL scores in children before and after the COVID-19 pandemic.

Next, we examined the emotional and social dimensions. We observed contrasting findings: The social dimension indicated a good QoL with a score > 90, while the emotional dimension showed a poor QoL19, ranking lowest among all PedsQL dimensions. This aligns with a previous study suggesting that children with long-term physical illnesses not involving cerebral function are more vulnerable to emotional disorders than those exhibiting antisocial behavior.[20]

The results regarding the emotional dimension also indicate higher scores in both child and parent reports after the COVID-19 pandemic. However, this difference was not statistically significant. The emotional dimensions are often related to fear and sadness regarding their different conditions compared to normal children. Many patients also experience concerns about stoma leakage during outdoor activities.[21]

In the social dimension, a few problems were identified. The most common complaint was "Unable to do things that his friends can do." Many individuals point out that activities like swimming are not accessible to HSCR patients with a

stoma. Analyzing the difference in scores between the two groups revealed that the score was higher in child reports after the COVID-19 pandemic. The large-scale social restrictions implemented during the pandemic resulted in children being isolated from their friends, leading to a lack of relationships among peers essential for developing children's social skills.[22-23] Other factors impact children's social lives, especially during a pandemic, including parental support and environmental conditions.[24] However, this study did not evaluate other factors that could impact patients' quality of life in the social dimension. One reason the score was higher after the COVID-19 pandemic compared to before may be that no issues were identified in the social dimension post-pandemic, as individuals had fewer opportunities to socialize due to extensive social restrictions.

Two groups showed similar scores on the school dimension. Children struggling with stool control often lack self-confidence and dislike attending school.[25] Interestingly, a change in school regulations was likely underlying the scores that tended to be lower in the group after the COVID-19 pandemic. These changes impact children's mental health.[26-27]

PedsQL scores are categorized as good (>90), moderate (90-75), and poor (<75) [19]. Our study indicated that the QoL for patients with HSCR was categorized as moderate in both the pre and post-pandemic COVID-19 groups. Additionally, the QoL for HSCR patients with stomas is not significantly different from that of normal children.[28-30]

Several factors contribute to a good QoL for HSCR patients with stomas. One of these factors is the parents' knowledge about stoma care. A strong knowledge base also contributes to maintaining a high QoL.[31] This also aligns with a prior study suggesting that education can significantly influence patient quality of life.[32] This education can enhance patient knowledge to prevent future complications.[33-34]

The family's role is also essential in achieving a good QoL for HSCR patients with stomas. These patients require support from family and medical professionals to adjust to social and psychological challenges.[35] Family support can also enhance the patient's self-confidence and psychological well-being.[35-37]

Patients with stomas in our HSCR study reported a good QoL. They were similar to healthy children in terms of QoL, perhaps influenced by several factors, such as the adequate education of the parents, which enabled them to understand how to care for the stoma and achieve good physical function in HSCR patients' stoma. In our study, strong family support for HSCR patients with stomas positively influenced their self-confidence and psychological well-being, ensuring that their social, emotional, and educational functions remained intact. However, our research did not evaluate other factors, such as education and family support, that could affect the quality of life for HSCR patients with a stoma. Every individual's experience is unique. Some children may face more significant challenges, while others may adapt effectively. The small sample size in this study could be seen as a limitation, as it may not adequately represent the broader population.

Our study found no significant correlation between parent and child reports. The consistency between child and parent reports on the PedsQL is low, with intra-class coefficients ranging from 0.02 to 0.23.[38] This occurs because various factors can affect the level of agreement between parent and child reports, including the child's age, the domain being assessed, and the parent's QoL.[39]

CONCLUSION

The QoL for HSCR patients with a stoma before and after the COVID-19 pandemic is similar, except for the social dimension in children's reports. There was no significant correlation between child and parent reports. Further studies with more cases are necessary to clarify our findings.

List of Abbreviations

COVID-19: Coronavirus disease 2019; HSCR: Hirschsprung disease; QoL: quality of life; TEPT: transanal endorectal pull-through; WHO: World Health Organization.

CONFLICT OF INTEREST

The authors declared no potential conflicts of interest regarding this article's research, authorship, and/or publication.

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