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Original Research Article

Comparison of the quality of life of children with congenital megacolon (Hirschsprung's disease) post transanal endorectal pull-through with other children's surgical cases in the 5-7-years age group at Moewardi Hospital Surakarta

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ABSTRACT

Background: Congenital megacolon (Hirschsprung's disease) is caused by intestinal innervation abnormalities, due to failure of neuroblast migration from the proximal to the distal intestine so there is no nerve innervation in the distal intestine and causes functional ileus and hypertrophy and excessive intestinal distension. The classic triad of clinical features in neonates is late meconium expulsion, namely more than the first 24 hours, green vomiting, and an enlarged stomach. Management of congenital megacolon is divided into two, namely non-surgical treatment to overcome complications that may occur and to improve the general condition until definitive surgery and surgical treatment namely transabdominal approach (TAB), consisting of Swenson, Duhamel, Rehbein, and Soave, and transanal endorectal pull-through (TEPT). The advantages of TEPT include better cosmetic effects and shorter hospital stays. Quality of life (QoL) is a subjective perception of a person's well-being regarding levels in his life, which includes physical, social, and psychological conditions.
Methods: This research is observational analytical research with a cross-sectional approach conducted at RSUD Dr. Moewardi Surakarta during August - September 2022.
Results: Patients with congenital megacolon (Hirschsprung disease) after TEPT have a worse QoL than patients without congenital megacolon (p value <0.05). All components of the pediatrics quality of life questionnaire (PedsQL™) are influenced by the patient's condition (p value <0.05).
Conclusions: The poor quality of life should increase awareness of the importance of immediate treatment. Difficulty defecating, diarrhea, and abdominal pain can reduce the patient's QoL after TEPT. Family dynamics and support must be prioritized to improve the QoL.

Keywords: Hirschsprung's disease, Congenital megacolon, Transanal endorectal pullthrough, Quality of life

INTRODUCTION

Congenital megacolon (Hirschsprung's disease) is caused by intestinal innervation abnormalities, due to failure of neuroblast migration from the proximal to the distal intestine so that there is no nerve innervation in the distal intestine.[1] Indonesia's population is around 200 million with a birth rate of 35 per million, so it is predicted that 1400 babies will be born with congenital megacolon per

year.[2] Bodian et al stated that aganglionosis in congenital megacolon is not caused by failure to develop extrinsic parasympathetic innervation, but is a primary lesion, so that there is an autonomic imbalance that cannot be corrected by sympathectomy.[3]

Abnormalities in this disease are associated with spasms in the distal colon and internal anal sphincter resulting in obstruction. Therefore, the abnormal part will experience contraction in the distal segment so that the normal part will experience dilation in the proximal part.[4] The pathophysiological basis of congenital megacolon is the absence of propulsive waves and abnormalities or loss of relaxation of the internal anal sphincter caused by aganglionosis, hypoganglionosis, or dysganglionosis in the large intestine.[4]

Based on the location of the aganglionic segment, this disease is divided into a short segment if it reaches the sigmoid, a long segment if it is higher than the sigmoid, total if it affects the entire colon, and universal if it affects the entire intestine up to the pylorus.[2,5]

Diagnosis includes history taking, physical examination, and supporting examinations. The complaints experienced by neonates and children are different. In neonates, meconium comes out late (>24 hours), the stomach is convex and tense, vomiting, and loose stools. Meanwhile, children experience chronic constipation, failure to thrive (failure to grow), and lack of appetite (anorexia).[1]

On physical examination, the stomach is found to be convex or completely bulging, on palpation the stomach is soft to tense, and on auscultation, the bowel sounds are weak or rare. During a digital rectal examination, it is felt that the tip of the finger is pinched by the narrow lumen of the rectum and when the finger is pulled out, the feces will spray out in large quantities and then the bloating in the stomach disappears temporarily.[6]

A plain abdominal radiograph examination was performed on a patient with suspected congenital megacolon and a picture of low-lying intestinal obstruction was obtained. In addition, to confirm the diagnosis, a barium enema is performed with the results of an area of narrowing, in the rectum proximally, the transition area, in the proximal area of narrowing towards the dilatation area, and an area of widening of the lumen, proximal to the transition area. The gold standard for congenital megacolon examination is rectal biopsy.[7]

The main goal of operative treatment of congenital megacolon is to remove the aganglionic intestine and reconstruct the intestinal tract by providing a segment of the intestine that has normal innervation to the anus while maintaining normal function of the anal sphincter muscle. There are several operative approaches to congenital megacolon, including the transabdominal approach (TAB) which consists of 4 types of procedures, namely, Swenson, Duhamel, Rehbein, and Soave, as well as transanal endorectal pull-through (TEPT).[8] The transanal endorectal pull through (TEPT) procedure is a procedure where intra-abdominal dissection is not performed because the entire procedure is carried out transanally.[9]

Starting from a circular mucosal incision above the dentate line, a submucosal dissection is performed in a proximal direction. The muscularis is incised circumferentially and the remainder of the dissection is carried outside the rectal wall until the transition zone is identified. After confirmation of ganglion cells in the frozen section, the aganglionic intestine was resected and anastomosis was performed.[9] Single-stage TEPT is felt to be more suitable in cases of infants because it is a minimally invasive procedure that can reduce the risk of complications such as abdominal adhesions and pelvic nerve injury.[9]

A study shows that in a short period, pediatric patients undergoing TEPT can still experience postoperative complications such as enterocolitis, perineal dermatitis, the need for dilatation, and even reoperation as in classic transabdominal pull-through surgery. However, this TEPT procedure results in a shorter period of intestinal paralysis and a shorter hospital stay.[10] In addition, the laparoscopy assisted transanal endorectal pull-through (LATEP) which is preceded by a laparoscopic biopsy can confirm the extent of the area of aganglionosis before Interventional procedures on the colonic mesentery or rectal ablation can be beneficial in neonates whose transition zone level is difficult to predict.[9]

Quality of life is a subjective, multidimensional concept that defines standard levels for emotional, physical, material, and social well-being.[11] A child's quality of life can be assessed from physical, psychological, social relationships, and environmental aspects.[12]

The aim of this study is to determine the quality of life of megacolon patients after TAERPT procedures with other pediatric surgical cases in the 5-7-year age group.

METHODS

The type of research includes observational analytical research with a cross-sectional approach. The data collection process was carried out at the Children's Surgery Polyclinic, RSUD Dr. Moewardi Surakarta, and in the patient's medical records during August - September 2022. The sample calculation in this study used the Lemeshow formula for research sample size, and we obtained a minimum sample of 36 patients . The subjects of this study were all pediatric patients aged 5-7 years with congenital megacolon who received TEPT and without megacolon at Moewardi Regional Hospital.

The inclusion criteria for this study are as follows: patients diagnosed with congenital Megacolon at least more than 6 months after the TEPT procedure with an age range of 5 - 7 years; patients who have complete medical record data, including record number, full name, date of birth, gender,

address, telephone number, age at surgery, aganglion segment length, history, physical examination and other support; patients and patient families who agree to be research subjects; and children without congenital megacolon aged 5-7 years.

The exclusion criteria in this study are as follows: cogenital megacolon patients who have other congenital diseases, the patient has died, and patients undergoing definitive therapy other than TEPT.

Quality of life assessment uses the pediatric quality of life inventory™ (Peds QL™) questionnaire which has 4 domains, namely social, emotional, school, and psychological. Apart from that, this questionnaire has 2 modules, namely a module for children with certain chronic diseases and a module to assess the child's health condition so that it can differentiate the quality of life of healthy children from children suffering from an illness.[13]

The data was then analyzed using the statistical package for social sciences (SPSS) program version 20.0 and a multivariate analysis of the quality of life of congenital megacolon patients after TEPT using the multivariate analysis of variance (MANOVA) test.

This study was approved by the ethics committee in Moewardi Hospital, and have received written informed consent from the patients or the patient's parents.

RESULTS

Demographic data and clinical characteristics in this study can be seen in Table 1.

Table 1: Clinical characteristics of patients.

Characteristics	n	%
Age (years old)		
5	15	41.7
6	14	38.9
7	7	19.4
Total	36	100
Gender		
Man	31	86.1
Woman	5	13.9
Total	36	100
Clinical condition		
With megacolon	15	41.7
Without megacolon	21	58.3
Total	36	100

Respondents in the study were divided into patients with megacolon and without megacolon who were then measured for their quality of life using the PedsQL™ questionnaire which contained 10 questions. The score is interpreted as the higher the score, the higher the quality of life.

Table 2 shows that patients with congenital megacolon (Hirschsprung disease) after transanal endorectal pull-through have a worse quality of life than patients without congenital megacolon (p value <0.05).

Table 2: Influence of the patient's condition on the patient's quality of life based on the results of the PedsQLTM.

Effect	Value	Sig.
Pillai's trace	0.918	0.000
Wilks' lambda	0.082	0.000
Hotelling's trace	11.202	0.000
Roy's largest root	11.202	0.000

Table 3 shows that all components of the PedsQL™ are influenced by the patient's condition (p value <0.05).

We also use the same method to compare the patient's age with the patient's quality of life. However, the results we obtained were not significant.

Table 3: Effect of patient condition with each component of the PedsQLTM.

Dependent variable	df	F	Sig.
Abdominal pain	1	37.438	0.000
Discomfort when eating	1	38.636	0.000
Limitation when eating and drinking	1	43.029	0.000
Swallowing problems	1	38.824	0.000
Nausea and chest burns	1	51.262	0.000
Weakness and vomiting	1	38.636	0.000
Bloating	1	7.556	0.010
Difficult to defecate	1	186.184	0.000
Bloody feces	1	31.086	0.000
Diarrhea	1	82.642	0.000

DISCUSSION

TEPT is a definitive operative procedure for congenital megacolon which is carried out in one stage. In this surgical method, a transanal dissection is performed outside the intestinal lumen wall to separate the aganglionic segment.[14]

QoL in patients with congenital Megacolon increases in the physical aspect but decreases in the psychosocial aspect, this continues with increasing age. The most common anomalies in congenital megacolon patients are cardiac (32%), genitourinary (18%), and vertebral/spinal (10%). Adaptation is needed from each patient mentally and emotionally and support from parents to have a higher QoL.[12]

Most children with congenital megacolon have a fairly good quality of life, although not as good as their healthy peers. The majority of children can carry out routine physical activities, academic activities, and good social

relationships with their friends. Factors that may influence the patient's quality of life include age at the time of surgery, gender, type of surgery performed, degree of aganglionosis, initial stoma diversion, episodes of enterocolitis, and bowel dysfunction.[15]

Bowel function and quality of life can be inconsistent, this may be due to symptoms and psychological factors, such as anxiety and unhappiness which can have a negative influence. In addition, parental stress and self-efficacy are associated with child health outcomes and may play a mediating role between children's health-related behaviors and quality of life.[16]

The quality of life of patients with congenital megacolon after surgery was found to be quite good with increasing age, which is in line with the results of several other studies, although improvement in intestinal dysfunction did not increase significantly. In addition, there was no difference between the responses of pediatric patients and parents regarding the child's post-operative quality of life, this could be due to parents overestimating the improvement in post-operative quality of life.[17]

In childhood, behavioral problems towards the environment become more prominent because patients may experience rejection from peers, depression, or anxiety about their differences and difficulties at school. Therefore, family dynamics and support are important for a patient's quality of life.[18]

Several studies conducted in England and Ireland did not prove that there was a difference in race, gender, or various lengths of aganglionosis on the quality of life of patients with post-operative congenital Megacolon. Therefore, regardless of the surgical technique used, the pull-through procedure is seen only as part of the management of children with congenital megacolon and requires appropriate post-operative management, including a management program in bowel preservation and patient psychological factors.[19]

Quality of life indicators in the group of healthy children showed a higher quality of life compared to children who suffered from congenital megacolon before surgery. Analysis of the quality of life of children with congenital megacolon after surgery showed a significant improvement within 12 months after surgery compared to the group of healthy children with an average level of 76.7-84.7%. Patients with post-operative congenital megacolon allow better results in the recovery of key parameters in the early postoperative period such as duration of analgesia, recovery of peristalsis, and time to start enteral feeding.[20]

The quality of life of children with congenital megacolon assessed by kidscreen was significantly higher, from the parent's perspective, when compared with the reference population for children of the same sex and age, and tended to be higher from the children's perspective. The quality of life of school-age children with congenital megacolon is higher when compared with children of the same sex and age in the reference population. In addition, children with congenital megacolon can also maintain normal intelligence and do not have dysexecutive disorders.[21]

Limitation in this study is that a larger sample size is required, due to a number of patients who could no longer be contacted and moved their residence during the time of data was collected.

CONCLUSION

The poor quality of life of patients with congenital megacolon after TEPT compared with children without congenital megacolon makes us increasingly aware of the importance of immediate treatment in this case. The most disturbing condition of patients with congenital megacolon after the TEPT procedure is difficulty defecating, diarrhea, and abdominal pain, this can reduce the patient's quality of life. Family dynamics and support must be prioritized to improve the quality of life of children with congenital megacolon so that they do not experience problems when they grow up.

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