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Original Research Article

Clinical outcomes evaluation of congenital megacolon (Hirschsprung disease) short segment type after transanal endorectal pull through at Moewardi Hospital Surakarta

Erwin A. Ismail[1]*, Suwardi[2], Ida B. Budhi S. A.[3], Nunik Agustriani[2], Amru Sungkar[3], Setya A. Tusarawardaya[3]

[1]Department of Surgery, Sebelas Maret University, Surakarta, Indonesia
[2]Department of Surgery, Moewardi Hospital, Surakarta, Indonesia
[3]Department of Surgery, Medical Faculty Sebelas Maret University, Moewardi Hospital, Surakarta, Indonesia

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***Correspondence:**
Dr. Erwin A. Ismail,
E-mail: erwinaritama@gmail.com

ABSTRACT

Background: Congenital megacolon occurs in 1 in 5000 live births, with an estimated 1200 cases annually in Indonesia, predominantly affecting males. Mortality and morbidity in these patients can be reduced through improved diagnosis, neonatal care, surgical techniques, and enterocolitis management. The transanal endorectal pull through (TEPT) procedure, a minimally invasive surgery, is the common definitive treatment for congenital megacolon, performed even in neonates. However, TEPT can lead to post-operative anorectal dysfunction, necessitating assessment through fecal continence evaluation. The aims of this study is to understand of the clinical outcome congenital megacolon short segment type after TEPT procedure at Moewardi Hospital, Surakarta.
Methods: The study is a cross sectional descriptive analysis in patients with congenital megacolon short segment type who have received TEPT treatment at Moewardi Hospital Surakarta from 2010 until 2022.
Results: Among the patients, 18.9% exhibited normal anorectal function, 54.1% had good continence, 24.3% had moderate continence, and 2.7% experienced incontinence. Surgery age was mostly between 1 and 12 months (64.9%), with 62.2% achieving good continence. At the 1-5-year follow-up, 48.6% had good continence. Among primarily male patients (86.1%), 43.2% had good continence. Strong correlation (Pearson Chi-square score: 0.001) confirmed the relationship between age at the time of surgery and anorectal function outcomes.
Conclusions: TEPT yielded favorable anorectal function outcomes in short-segment congenital megacolon patients, with surgery age significantly impacting these outcomes. This study supports the hypothesis that surgery age influences anorectal function outcomes after TEPT at Moewardi Hospital, Surakarta.

Keywords: Hirschsprung disease, Transanal endorectal pullthrough, Clinical outcomes

INTRODUCTION

Congenital megacolon, also known as Hirschsprung disease, is a congenital condition characterized by the absence of nerve cells in the intestine, particularly in the region extending from the internal anal sphincter to varying lengths proximally. This condition consistently involves the anus and at least a portion of the rectum, leading to clinical symptoms that manifest as functional impairments in intestinal passage. Early intervention is crucial in the management of congenital megacolon in pediatric patients, aiming to minimize complications and halt the progression of the patient's overall health deterioration. The limited incidence of hospital-treated cases may be attributed to a lack of public awareness of

congenital megacolon, as well as potential delays in initial treatment by paramedics and local medical staff.[1]

The prevalence of congenital megacolon is approximately 1 in 5000 live births. In the context of Indonesia, it is projected that around 1200 infants will be affected by congenital megacolon annually. Furthermore, this condition's gender distribution is considered to be lopsided, with a male-to-female ratio of 4:1. It is imperative to promptly diagnose congenital megacolon in order to mitigate the potentially perilous complications that may arise for the patient, including constipation, enterocolitis, intestinal perforation, and sepsis, which can ultimately lead to fatality.[2]

According to Rochadi, enhancements in the diagnosis of congenital megacolon, intensive care for neonates, surgical procedures, and the management of congenital megacolon accompanied by enterocolitis have the potential to decrease mortality and morbidity rates in affected patients.[3]

The management of congenital megacolon involves the implementation of interim interventions to alleviate intra-abdominal pressure and the implementation of definitive surgical procedures. The transanal endorectal pull trough (TEPT) procedure is widely recognized as a prominent surgical intervention for congenital megacolon. This technique represents a significant advancement in the field of minimally invasive surgery, as it allows for transanal access without the requirement of laparotomy or laparoscopy. Moreover, the TEPT procedure can be conducted from the neonatal stage, as demonstrated by Stensrud et al.[4]

The TEPT technique has demonstrated favorable outcomes, particularly for the rectosigmoid type, as it allows for a reduced treatment duration and early initiation of feeding is administered promptly following surgery.[5] According to Dutta, following the TEPT treatment, there is a reduction in the length of the patient's intestinal segment. Consequently, this alteration may lead to the occurrence of anorectal dysfunction in the post-TEPT period.[6] The most optimal metric for evaluating anorectal outcome and function subsequent to TEPT is the assessment of fecal continence, specifically the patient's capacity to retain fecal matter. Hence, it is imperative to conduct comprehensive monitoring of fecal continence in both the immediate and extended periods following surgery in order to ascertain postoperative results and address any potential problems.[7]

Aim

This study aimed to evaluate the outcomes of patients with congenital megacolon short segment type who underwent TEPT surgery. The assessment of fecal continence index was conducted at Moewardi Hospital Surakarta, starting from the year 2010.

METHODS

This study is a cross sectional descriptive analysis in patients with congenital megacolon short segment type who have received TEPT treatment at Moewardi Hospital Surakarta from 2010 until 2022. The primary data collection method was conducted at the Pediatric Surgery Polyclinic at Moewardi Hospital Surakarta. Meanwhile, the secondary data was collected from the patients' medical records stored in the medical records department of Moewardi Hospital. The study was conducted over the months of August and September in the year 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 study population consisted of patients diagnosed with congenital megacolon short segment type who had TEPT operation at Moewardi Hospital between the years 2010 and 2022. The included patients were under the age of 11 and had a minimum duration of TEPT treatment of six months. The exclusion criteria in this study are patients with congenital megacolon who during the TEPT procedure received a transabdominal assisted procedure, either laparoscopic or laparotomy, patients with other congenital disease, and patients undergoing definitive therapy other than TEPT.

Table 1: Heikkinen scoring system.[8]

Variables	N
Frequency of defecation	
Normal	2
Often (3-5/day)	1
Very often	0
Fecal consistency	
Normal	2
Loose	1
Liquid	0
Soiling	
No	2
Stress/diarrhoea	1
Constant	0
Rectal sensation	
Normal	2
Defective	1
Missing	0
Ability to hold back defecation	
Minutes	2
Seconds	1
Missing	0
Discrimination between formed, loose, or gaseous stools	
Normal	2
Defective	1
Missing	0
Need for therapy (enemas, drugs, napkins)	
No	2
Occasionally	1
Always	0

Data was analyzed using the statistical package for social sciences (SPSS) program version 20.0. The dataset comprised variables such as patient age, age at the time of surgery, gender, age at the time of follow-up, and anorectal function. Fecal continence is measured using a questionnaire based on the Heikkinen scoring system. After a period of 6 months following the surgical procedure.[8]

Heikkinen scoring system is a measurement which is used to evaluate fecal continence (anorectal function) which consists of 7 questions related to defecation problems in the patient (score 14 sufferers have no anorectal disorders at all (normal), score 10-13 good continence, the patient is believed to have no social disturbances, score 5-9 moderate continence, sufferers with limitations in their social life and recurrent obstipation, score 4 or more small = bad/total incontinence).[8]

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

There were 37 patients diagnosed with congenital megacolon short segment type who received TEPT from 2010 – 2022 who were included in the inclusion criteria. The data revealed that anorectal function was normal in 7 patients (18.9%), good continence in 20 patients (54.1%), moderate continence in 9 patients (24.3%), and incontinence in 1 patient (2.7%). At the time of operation, the age distribution for congenital megacolon short segment type patients were largely between 1 and 12 months, with a total of 24 patients (64.9%). The most common outcome of anorectal function in congenital megacolon short segment type patients was good continence at the age of surgery 1-12 months, which occurred in 23 patients (62.2%). The average age of the patients at the time of follow-up in this study was 1-5 years, with 29 patients (78.4%). Good continence at the age of 1-5 years was the most prevalent outcome of anorectal function in congenital megacolon short segment type patients, with as many as 18 patients (48.6%) achieving it. The patients were primarily male, with 31 patients (86.1%), good continence was the most prevalent outcome for anorectal function in this category, with 16 patients (43.2%).

Table 2: Clinical characteristics of patients.

Characteristics	n	%
Age at surgery		
0-1 month	5	13,5
1-12 months	24	64,9
1-5 years	7	18,9
6-11 years	1	2,7
Total	37	100
Age at follow-up		
0-1 month	0	0
1-12 months	2	5,4
1-5 years	29	78,4
6-11 years	6	16,2
Total	37	100
Gender		
Man	32	72,5
Woman	5	13,5
Total	37	100
Anorectal function outcomes		
Normal	7	18,9
Good continence	20	54,1
Moderate continence	9	24,3
Incontinence	1	2,7
Total	37	100

Table 3: Anorectal function outcomes.

Variables	Anorectal function outcomes									
	Normal		Good continence		Moderate continence		Incontinence		Total	
	N	%	N	%	N	%	N	%	N	%
Age at surgery										
0-1 month	0	0	3	8,1	3	8,1	0	0	6	16.2
1-12 months	4	10.8	13	35.1	6	16.2	0	0	23	62,2
1-5 years old	3	8,1	4	10.8	0	0	0	0	7	18.9
6-11 years old	0	0	0	0	0	0	1	2.7	1	2.7
Age at follow-up										
1-12 months	0	0	2	5.4	0	0	0	0	2	5.4
1-5 years old	6	16.2	18	48.6	7	18.9	0	0	31	83.8
6-11 years old	1	2.7	2	5,5	0	0	1	2.7	4	10.8
Gender										
Man	5	13.5	16	43.2	7	18.9	1	2.7	29	78.4
Woman	2	5.4	4	10.8	2	5.4	0	0	8	21.6

The Pearson Chi-square score is 0.001 (0.05), indicating that age at the time of operation is strongly related to anorectal function results. Therefore, the hypothesis in this study, which claims that there is a relationship between age at the time of surgery and the outcome of anorectal function in congenital megacolon short segment type patients after the TEPT treatment at Moewardi Hospital in Surakarta, has been supported by the results of empirical research.

DISCUSSION

Congenital megacolon is a complex genetic condition that is distinguished by the lack of ganglion cells in the myenteric plexus and intestinal submucosa, leading to functional blockage. The classification of congenital megacolon can be delineated into three distinct categories, namely short segment, long segment, and entire colonic aganglionosis, based on the extent of aganglionosis.[9] This study focused on a sample population consisting of patients diagnosed with short segment congenital megacolon who were eligible candidates for the transanal endorectal pullthrough surgery, which is considered the gold standard treatment for this condition.

Total transanal pull-through has several advantages in comparison to other surgical procedures. Firstly, it involves minimum resection of the normal ganglion, hence preserving the integrity of the ganglion cells. This is beneficial as it helps maintain normal bowel function post-surgery. Additionally, this procedure has been associated with a decreased length of hospital stay, resulting in reduced healthcare expenses. Moreover, the lower risk of intestinal obstruction further contributes to the overall positive outcomes of total transanal pull-through. In the context of megacolon, Hirschsprung's enterocolitis is a potential complication that may manifest both before to and following the completion of definitive surgical intervention.[10] The findings of this research brief indicate that patients with megacolon who underwent a transanal endorectal pullthrough treatment demonstrated favorable outcomes in terms of anorectal function, with low occurrence of anorectal function abnormalities.[11]

This study revealed that patients with congenital megacolon short segment type who underwent TEPT at Moewardi Hospital Surakarta achieved favorable outcomes in terms of anorectal function. Specifically, normal results were observed in 7 patients (18.9%), good continence in 20 patients (54.1%), moderate continence in 9 patients (24.3%), and 1 patient (2.7%) experienced incontinence. The findings of this study demonstrate that the incidence of problems related to anorectal function abnormalities following TEPT surgery is minimal. Consequently, TEPT may be considered as the optimal surgical approach for patients presenting with short segment megacolon.

Certain surgeons may opt to delay the performance of the TEPT surgery till the infant reaches a few months of age. Children are permitted to get instruction from their parents regarding the proper techniques for doing personal hygiene tasks such as washing out or engaging in rectal irrigation in the comfort of their own homes. There exist multiple rationales for the implementation of this waiting strategy, including the potential facilitation of the surgical procedure through enhanced visualization, the expectation that the enlarged proximal colon will naturally reduce in size, and, in certain instances, the logistical consideration of operating room scheduling constraints. One potential risk associated with this technique is the potential occurrence of enterocolitis in the infant during the waiting time.[12]

The present study revealed a median age of 3 months for surgical intervention, aligning with the viewpoint expressed by Tore and Langer, who advocate for deferring surgery till later infancy rather than doing it on early neonates. According to Gandhi et al, the technique known as TEPT has favorable outcomes when conducted on children across various age groups, thereby mitigating the need for abdominal exploration surgery. The median age for surgical intervention ranges from 6 months to 3 years, with a mean age of 9 months.[13]

This study revealed a statistically significant association between the age at surgery and the results of anorectal function, as indicated by a p value of 0.001. According to the study conducted by Onishi, it is typically recommended to proceed with definitive surgery once the patient's weight reaches approximately 5 kg. However, there are instances where surgery may be postponed if prioritizing treatment for other related anomalies, such as congenital heart disease or chromosomal anomalies, is deemed more beneficial in enhancing the patient's overall condition prior to the surgical intervention. The majority of pediatric surgeons at alternative institutions undertake decisive surgical interventions for patients with megacolon once their weight reaches approximately 5 kg. The recommended time for the TA operation in infants, namely from birth to 15 months of age. The findings suggest that it is advisable to execute the TEPT before the infant reaches 6 months of age.[14]

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 patients that diagnosed with congenital megacolon short segment type who underwent a TEPT procedure exhibited favorable outcomes in terms of anorectal function, with low occurrence of anorectal function problems. A significant correlation has been observed between the age at when surgery was performed with congenital short segment megacolon and the outcome of anorectal function following the TEPT procedure.

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