ORIGINAL ARTICLE

Growth outcomes in Hirschsprung disease patients following pull-through

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ABSTRACT

Introduction: Despite various definitive methods that are used for treating Hirschsprung's disease (HSCR), there are few studies comparing the effect of different pull-through methods on the growth outcomes of patients. We aimed to compare the effect of three different pull-through methods, namely Duhamel, Soave and transanal endorectal pull-through (TEPT), on HSCR growth outcomes of patients.

Methods: Medical records of all HSCR patients who underwent pull-through at the Dr. Sardjito Hospital, Indonesia between January 2010 and August 2016 were reviewed for their growth outcomes before and after the surgery.

Results: We included 64 HSCR patients, 45 males and 19 females, of which 14, 17, and 33 patients underwent Duhamel, Soave, and TEPT respectively. There were no nutritional status differences in HSCR patients after Duhamel, Soave, and TEPT surgery (p=0.07, 0.17, and 0.79, respectively). Z-score average of weight-for-age did not differ between three surgical methods (p=0.77 and 0.15 for preoperative and postoperative, respectively). In addition, the improvement of nutritional status was achieved in 21.2% HSCR patients after TEPT, 14.3% post Duhamel and 5.9% following Soave procedure, but these differences did not reach a significant level (p=0.34).

Discussion: Our study shows no difference in effect on the growth outcomes in HSCR patients following Duhamel, Soave and TEPT procedure. Further study with a larger sample size is important to give valuable long-term growth outcomes for HSCR patients after pull-through.

KEY WORDS:
Hirschsprung disease; pull-through; Duhamel; Soave; TEPT; growth outcomes

INTRODUCTION

Hirschsprung disease (HSCR) is a congenital disorder characterized by aganglionic colon.[1,2] HSCR occurred on 1 of 5,000 live birth, while the incidence in Indonesia was 1:3,250 of live birth.[3,4]

The definitive therapy for HSCR is removing the aganglionic part and making anastomosis between ganglionic colon with anus (pull-through). This procedure can be performed by transabdominal- or transanal approach, with the most common procedures of Duhamel, Soave and transanal endorectal pull-through (TEPT).[5-10]

Growth pattern can reflect' the general health and nutritional status of patients. Any deviation on its pattern is related with chronic or severe diseases and can cause defect in the development.[11,12] Despite various definitive methods that are used for treating HSCR, there are few studies that compared the effect of different pull-through methods on the growth outcomes of patients.[12-14] Therefore, we aimed to compare the effect of three different pull-through methods, namely Duhamel, Soave and TEPT, on HSCR' growth outcomes of patients.

MATERIALS AND METHODS

Subjects

This was a cohort study of all patients with HSCR who had undergone Soave, Duhamel, or TEPT procedures at the Dr. Sardjito Hospital in Yogyakarta, Indonesia between January 2010 and August 2016. Data was retrieved from' medical records of patients and reviewed including their gender, type of HSCR, age at definitive surgery, gestational age, birth weight, and surgical procedures. We excluded patients with incomplete medical records, deceased, or syndromic HSCR. Data of a total of 64 patient's was included. This study was approved by the Institutional Review Board of the Faculty of Medicine, Public Health and Nursing, Universitas Gadjah Mada/Dr. Sardjito Hospital (KE/FK/1356/EC/2015).

Surgical procedures

All Duhamel and Soave procedures were performed approximately six months after colostomy (multi-staged operation), while TEPT operation was conducted without prior colostomy (one-stage surgery).

Growth outcomes

Growth outcomes of patients were measured before and after surgery and expressed as weight-for-age z scores in relation to growth standards of the age and gender according to the World Health Organization growth chart. These scores were then classified as normal ($z > -2$), underweight ($-3<z<-2$), and severely underweight ($z<-3$). As for growth progression outcome, the most updated weight data were taken which were the shortest measurement time before operation and the

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Table I: HSCR patients' characteristics who underwent pull-through at Dr. Sardjito Hospital, Indonesia

Characteristics	Duhamel (n=14) (n, %; mean ± SD)	Soave (n=17) (n, %; mean ± SD)	TEPT (n=33) (n, %; mean ± SD)	p-value
Gender				
• Male	10 (71.4)	14 (82.4)	21 (63.6)	0.39
• Female	4 (28.6)	3 (17.6)	12 (36.4)	
HSCR type				
• Short-segment	12	17	33	0.07
• Long-segment	2	0	0	
• Total colon aganglionosis	0	0	0	
Age at definitive surgery (years)	3.22 ± 2.22	3.25 ± 2.76	0.81 ± 1.83	0.0002*
Gestational age (weeks)	38.33 ± 1.87	37.55 ± 2.07	37.31 ± 2.66	0.40
Birth weight (gram)	3085 ± 632.48	3205 ± 387.62	3049 ± 397.82	0.52

*, significant (p<0.05); HSCR, Hirschsprung disease; TEPT, transanal endorectal pull-through

Table II: Comparison of nutritional status pre- and after pull-through in HSCR patients

Nutritional status	Duhamel (n=14)			Soave (n=17)			TEPT (n=33)		
	Preoperative (n, %)	Postoperative (n, %)	p	Preoperative (n, %)	Postoperative (n, %)	p	Preoperative (n, %)	Postoperative (n, %)	p
Severely underweight	0	4 (28.6)	0.07	4 (23.5)	6 (35.3)	0.17	2 (6.1)	2 (6.1)	0.79
Underweight	3 (21.4)	2 (14.3)		0	2 (11.8)		6 (18.2)	7 (21.2)	
Normal	11 (78.6)	8 (57.1)		13 (76.5)	9 (52.9)		25 (75.7)	24 (72.7)	

HSCR, Hirschsprung disease; TEPT, transanal endorectal pull-through

Table III: Z-score average of weight-for-age for HSCR patients pre- and after definitive surgery

Nutritional status	Nutritional status			p	Postoperative			p
	Duhamel (n=14)	Soave (n=17)	TEPT (n=33)		Duhamel (n=14)	Soave (n=17)	TEPT (n=33)	
Severely underweight	N/A	-4.09	-5.17	0.77	-4.89	-4.28	-3,14	0.15
Underweight	-1.95	N/A	-2.53		-2.61	-2.43	-2.56	
Normal	-0.98	-0.83	-0.75		-0.97	-0.90	-0.67	
Total average	-1.19	-1.60	-1.34		-2.33	-2.27	-1.22	

N/A, not applicable; HSCR, Hirschsprung disease; TEPT, transanal endorectal pull-through

Table IV: Nutritional status changes in HSCR patients after pull-through

Nutritional status changes	Duhamel (n=14) (n, %)	Soave (n=17) (n, %)	TEPT (n=33) (n, %)	p
Worsened	4 (28.6)	5 (29.4)	6 (18.2)	0.34
Steady	8 (57.1)	11 (64.7)	20 (60.6)	
Improved	2 (14.3)	1 (5.9)	7 (21.2)	

longest measurement time after operation and then classified as worsened (i.e. decrease nutritional status after surgery), steady (i.e., no change in nutritional status after surgery), or improved (i.e. increase nutritional status after surgery).

All patients received the same post-operative nutritional support. The measurement of postoperative nutritional status was performed by the same staff at our hospital to ensure the uniformity.

Statistical analysis

The data were presented as mean±Standard Deviation (SD) or frequencies (percentages). We conducted Wilcoxon's Signed Rank test to compare pre- and postoperative growth outcomes for each surgical procedure. Variables in baseline characteristics, postoperative growth outcomes between surgical procedures, and the weight measurement range of each procedure were compared with Kruskal Wallis test. p-value<0.05 was considered significant.

RESULTS

HSCR patients' characteristics

We analysed 64 HSCR patients who underwent pull-through, consisting of 14 Duhamel, 17 Soave, and 33 TEPT. There were 45 male and 19 female patients, with 97% infants having a short-segment aganglionosis. There was no difference in the gestational age and birth weight among three surgical methods (p=0.40 and 0.52, respectively) (Table I).

Comparison of nutritional status in HSCR patients following different pull-through

We determined whether the pull-through effect on the nutritional status of HSCR patients. No nutritional status differences were noted in HSCR patients after Duhamel, Soave, and TEPT surgery (p=0.07, 0.17, and 0.79, respectively) (Table II).

We analysed the differences of Z-score average of weight-for-age for HSCR patients among groups. No significant

differences were seen among the three surgical methods (p=0.77 and 0.15 for preoperative and postoperative, respectively) (Table III).

Next, we compared the changes in the nutritional status after pull-through. The improvement of nutritional status was achieved in 21.2% HSCR patients after TEPT, 14.3% post Duhamel and 5.9% following Soave procedure. However, these differences were not significant (p=0.34) (Table IV).

DISCUSSION

Our study shows that the effect of three pull-through methods on the growth outcomes of HSCR patients is not significantly different. It is shown by z-score average of weight-for-age for HSCR patients pre- and post-pull-through (Table III). Our findings were similar with More et al.12. However, we had two novelties: 1) we analysed the effect of three pull-through procedures on the growth outcomes of HSCR patients; and 2) we compared the growth outcomes of HSCR patients among those three methods. More et al.[12] determined the growth outcomes of HSCR patients after laparoscopic assisted endorectal pull-through and Duhamel, but did not compare the effect of those two methods. They reported that HSCR the birth weight of patients and one-year after pull-through weight were in the same percentile range.[12]

Interestingly, the nutritional status preoperative and postoperative Duhamel procedure almost reached a significant level (p=0.07; Table II). It might be associated with the evidences that some Duhamel patients had complication postoperatively, such as Hirschsprung-associated enterocolitis (2) and rectovaginal fistula (1). More multicentre and prospective cohort studies with a larger number of patients are important to clarify our findings. In addition, the surgical procedures were chosen according to the paediatric surgeon preference. These facts should be considered during the interpretation of our findings.

Our study also gives information that the total average of weight-for-age of our HSCR patients is less than the normal population mean (Table III). These findings were compatible with Neuvonen et al.,[13] where the height-adjusted relative weight (-3.0% [range, -38%, +35%]) in non-syndromic HSCR infants who underwent TEPT procedure was less than the normal population mean. More et al.,[12] also found that the length-for-age curve of their HSCR patients, although still normal, was in the 50th percentile. Moreover, Veras et al.,[14] reported that their infants with HSCR had a higher risk for growth impairment.

Differences in the growth of each child may be affected by gender. We found that ~70% of our patients were males. Different gender can cause differences in growth pattern, onset of growth spurt, body size, and bone maturity in children. The growth rate in girls at birth will be a little slower than boys, then it will be the same at the age of seven months, and faster until the age of four years.[15] Ghaemmaghami et al.,[16] found that there were significant differences in weight gain between boys and girls aged 0-2 years. They described that boys have a higher body weight compared to girls. This could have influenced by the presence of testosterone production in boys.[15,16] These findings should be considered during the interpretation of our study.

Most (51.6%) our patients underwent TEPT (Table I). This procedure had gained a popularity among paediatric surgeons as it can be performed safely even in the neonatal phase, supported by advances in the field of neonatal anaesthesia, monitoring during surgery, and parenteral nutrition. As a single-stage repair, TEPT doesn't need a stoma before definitive surgery.[13,17]

Furthermore, Gutbrod et al.[18] found that a term infant who had normal birth weight would have better growth outcomes compared to preterm infants. Infants with lower gestational age and accompanied by very low birth weight would result in poor weight gain and slower head growth.[18-20] Our study showed that there was no difference in gestational age and birth weight between HSCR patients who underwent Duhamel, Soave and TEPT procedure.

In this study, we determined the growth outcomes following pull-through and focused on the weight and height of patients. Furthermore, our report was a retrospective design and a follow-up of patients only according to the medical records. Therefore, some data were missing, becoming a weakness of our study. Moreover, growth outcomes assessments were not measured with a uniform timeline in all patients showing only shows a short-term growth outcome.

CONCLUSIONS

Our study shows no difference effect on growth outcomes in HSCR patients following Duhamel, Soave and TEPT procedures. Further study with larger sample size would give valuable long-term growth outcome for children with HSCR patients.

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