Surgery Today (2022) 52:92–97
https://doi.org/10.1007/s00595-021-02356-9

ORIGINAL ARTICLE

Optimal timing of definitive surgery for Hirschsprung's disease to achieve better long-term bowel function

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Received: 9 December 2020 / Accepted: 12 April 2021 / Published online: 12 August 2021

Abstract

Purpose Few studies have focused on the operative age for Hirschsprung's disease (HD). We evaluated the optimal timing of surgery in HD patients based on their long-term bowel function.

Methods HD was diagnosed in 65 pediatric patients in our institute between 1992 and 2018. Twenty-five patients underwent the Soave–Denda procedure (SD) and 40 underwent transanal endorectal pull-through (TA). We divided these patients into two groups: those who underwent surgery at < 6 months of age (younger group) and those who underwent surgery at 6–12 months of age (older group). We assessed bowel function at 5, 7, and 9 years of age.

Results The bowel function of the patients who underwent the SD did not differ significantly between the groups. Similarly, the total bowel-function scores of the patients who underwent TA did not differ between the groups at any age. However, the soiling score at 7 years of age in the older group of patients who underwent TA was significantly lower than that in the younger group ($p = 0.02$).

Conclusions Our data suggest that to achieve optimal bowel function, TA should be performed at < 6 months of age.

Keywords Hirschsprung's disease · Transanal endorectal pull-through · Soave–Denda procedure · Operative age · Bowel function

Introduction

Hirschsprung's disease (HD) has long been a main target disorder in pediatric patients. Pediatric surgeons developed three standard definitive operations for HD: the Swenson procedure [1], the Duhamel procedure [2], and the Soave procedure [3]. Despite the differences in these procedures, a definitive operation is generally recommended for HD patients when their body weight reaches approximately 5 kg. Many studies have evaluated bowel function after each of these procedures [4–7], but few have focused on the optimal timing of the definitive operation.

Until 1998, we performed the Soave–Denda procedure [8] via an open approach, as a one-step modified Soave procedure (SD) to treat HD in our institution. In 1998, the transabdominal approach was changed to a transanal approach, with or without laparoscopic assistance (TA). Despite the differences in approach, the concept of the operative technique has remained the same in relation to details, such as the mucosectomy level and treatment of the muscle cuff. In a previous study, we evaluated the long-term postoperative bowel function in pediatric patients who underwent definitive operations with the SD or TA procedures [4]. The bowel function of patients who underwent surgery with each of these approaches were similar and improved over time. Furthermore, satisfactory results were observed in both groups 10 years after the operation. We also reported on the long-term outcomes over 18 years in patients who underwent

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definitive operations using the SD procedure [5]. Recently, the TA procedure has become the main procedure for HD and definitive operations are being performed in younger patients [9]. However, these studies did not evaluate the optimal timing for a definitive operation to obtain a favorable bowel function.

The aim of this study is to evaluate the optimal timing for operations to improve the functional outcome of the bowel in HD patients who underwent definitive operations using the SD and the TA procedure.

Patients and methods

The subjects of this retrospective study were HD patients who underwent definitive surgery at less than 1 year of age at Kagoshima University between 1992 and 2018. From among the total 75 patients who underwent definitive surgery, the following 10 patients were excluded from the analysis: 6 who underwent the surgery when they were older than 1 year of age, 1 whose surgery was converted to open from TA, 1 who underwent a reoperation, 1 with trisomy 21, and 1 who died of an unrelated cause). Finally, 25 HD patients underwent the SD procedure, and 40 patients underwent the TA procedure, with or without laparoscopic assistance. We divided the patients who underwent each procedure into two groups: those who underwent surgery at < 6 months of age (younger group) and those underwent surgery at 6–12 months of age (older group).

The data abstracted from the patient records included the procedure type, operative details, extent of the aganglionic segment and early complications. We analyzed follow-up data for patients > 3 years of age, retrospectively, and evaluated bowel function according to the evacuation score (ES; defined by the Japan Society of Ano-Rectal Malformation Study Group in 1980) at 5, 7, and 9 years of age [4] (Table 1). We then calculated the total scores and the sum of the frequency of the bowel movement score and the soiling score. A lower score for constipation and incontinence was better, with the optimal score of 8 points indicating excellent bowel function [4]. Incontinence was defined as the inability to control the escape of stool from the rectum despite the patient's consciousness about their bowel movement, whereas soiling was defined as the underwear being soiled with the patient unaware. The main difference related to whether patient was conscious of their bowel movement.

We performed this clinical study according to the ethical guidelines for clinical research from the Japanese Ministry of Health, Labor and Welfare. The Research Ethics Committee of our institution approved this study (registration number: 27–133). All patients provided their informed consent.

Statistical analyses were performed using the unpaired *t* test and Fisher's exact probability. *p* values of < 0.05 were considered significant. Data are expressed as the mean ± standard deviation.

Table 1 Evacuation score defined by the Japan Society of Ano-Rectal Malformation Study Group

Item	Criteria	Score
Frequency of bowel movement	Frequent	2
	Accidental	1
	Absent	0
Constipation	Absent	4
	Accidental	3
	Requirement for enema or suppository everyday	2
	Requirement for colonic irrigation or stool extraction	1
Incontinence	Absent	4
	Incontinence with diarrhea	3
	Accidental	2
	More than twice a week	1
	Everyday	0
Soiling	Absent	2
	Accidental	1
	Everyday	0

Results

SD procedure

Patient characteristics and the extent of aganglionosis

Twenty-five patients with HD underwent the SD procedure, with 14 boys (73.7%) and 5 girls (26.3%) in the younger group and 4 boys (66.7%) and 2 girls (33.3%) in the older group. The extent of aganglionosis was as follows: short segment (ultra-short and recto-sigmoid), $n = 20$ (80.0%); long segment, $n = 5$ (20.0%). Patients with total colon aganglionosis were not included (Table 2).

Table 2 Extent of aganglionosis in the Soave–Denda (SD) procedure

	Younger group $n = 19$	Older group $n = 6$
Recto-sigmoid	15	5
Descending	3	0
Transverse	1	1
Ascending	0	0
Total	0	0

Operative results

The age at operation was significantly lower in the younger group than in the older group (113.11 ± 55.00 days vs. 234.00 ± 19.45 days, respectively; $p < 0.01$). There were no significant differences in weight at the time of operation (6244.0 ± 2127.98 g vs. 7719.17 ± 2127.98 g, respectively; $p = 0.13$), operative time 236.39 ± 46.57 min vs. 270.83 ± 62.81 min, respectively; $p = 0.19$) or blood loss per body weight during surgery (8.91 ± 6.12 ml/g vs. 8.91 ± 6.12 ml/g, respectively; $p = 0.75$). There was also no significant difference in the postoperative hospital stay (25.21 ± 10.09 days vs. 22.17 ± 4.91 days, respectively; $p = 0.50$) (Table 3).

Postoperative bowel function

The mean total ES at 5 years, 7 years, and 9 years in the younger vs. the older group was as follows: 7.00 ± 1.36 ($n = 14$) vs. 6.40 ± 1.24 ($n = 5$), respectively ($p = 0.39$); 7.27 ± 0.83 ($n = 11$) vs. 6.75 ± 0.75 ($n = 4$), respectively ($p = 0.30$); and 7.36 ± 1.22 ($n = 11$) vs. 7.00 ± 0.88 ($n = 4$), respectively ($p = 0.57$). The total ES increased chronologically in both groups, with satisfactory results achieved by at least 9 years after the operation (Fig. 1). There was no significant difference in the total score or in each parameter between the younger and older groups (Fig. 2).

TA procedure

Patient characteristics and the extent of aganglionosis

Forty patients with HD underwent the TA procedure, with 24 boys (82.8%) and 5 girls (17.2%) in the younger group and 10 boys (91.0%) and 1 girl (9.0%) in the older group. The extent of aganglionosis was as follows: short segment (ultra-short and recto-sigmoid), $n = 30$ (75.0%); long segment, $n = 8$ (20.0%); total colon aganglionosis with or without small intestine involvement, $n = 2$ (5.0%) (Table 4).

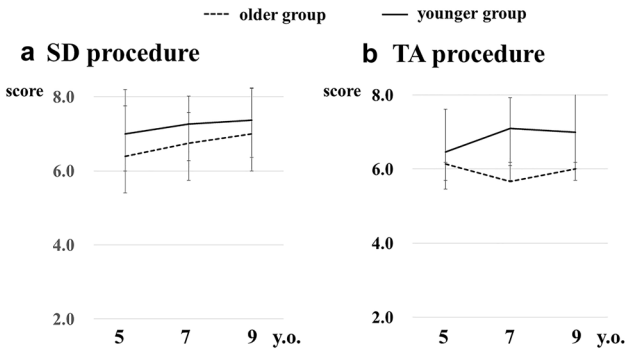

Fig. 1 Total evacuation scores. **a** Soave–Denda (SD) procedure. **b** Transanal endorectal pull-through (TA) procedure

Operative results

The age at operation was significantly lower in the younger group than in the older group (89.9 ± 45.89 days vs. 221.27 ± 33.39 days, respectively; $p < 0.01$) and the weight at operation was also significantly lower in the younger group (5241.45 ± 1370.59 g vs. 7239.27 ± 1454.86 g, respectively; $p < 0.01$). There were no significant differences between the younger and older groups in blood loss per body weight during the operation (4.96 ± 4.46 ml/g vs. 4.95 ± 3.93 ml/g, respectively; $p = 0.99$), operative time (293.17 ± 111.45 min vs. 289.73 ± 96.4 min, respectively; $p = 0.93$) or postoperative hospital stay (20.0 ± 10.4 days vs. 20.27 ± 10.62 days, respectively; $p = 0.94$) (Table 5).

Postoperative bowel function

The mean total ES at 5 years, 7 years, and 9 years in the younger vs. older groups was as follows: 6.47 ± 1.15 ($n = 6$) vs. 6.14 ± 1.46 ($n = 6$) ($p = 0.58$); 7.10 ± 0.83 ($n = 6$) vs. 5.67 ± 1.89 ($n = 6$) ($p = 0.07$); and 7.00 ± 1.41 ($n = 6$) vs. 6.00 ± 1.00 ($n = 2$) ($p = 0.44$). The total ES increased chronologically in both groups, with satisfactory results achieved by at least 9 years after the operation (Fig. 1). There was no significant difference in the total score between the younger and older groups.

Table 3 Operative results of the Soave–Denda (SD) procedure

	Younger group $n = 19$	Older group $n = 6$	p
Age at pull-through (days)	113.11 ± 55	234 ± 19.45	< 0.01
Weight at pull-through (g)	6244 ± 2127.98	7719.17 ± 924.37	0.13
Operation time (min)	236.39 ± 46.57	270.83 ± 62.81	0.19
Blood loss/BW (ml/g)	8.91 ± 6.12	8.07 ± 2.28	0.75
Postoperative hospital stay (days)	25.21 ± 10.09	22.17 ± 4.91	0.50

p values of < 0.05 were considered to indicate significance

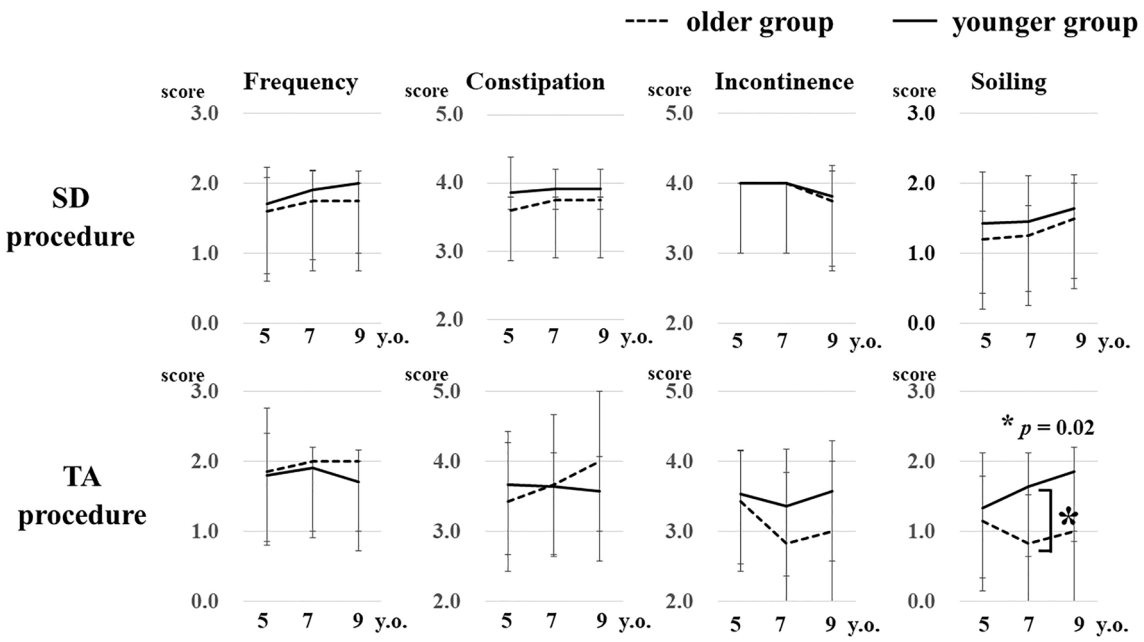

Fig. 2 Evacuation score according to the type of procedure. Upper panel: Soave–Denda (SD) procedure. Lower panel: transanal endorectal pull-through (TA) procedure. p values of <0.05 were considered to indicate significance

Table 4 Extent of aganglionosis in the transanal endorectal pull-through (TA) procedure

	Younger group $n=29$	Older group $n=11$
Recto-sigmoid	21	9
Descending	2	0
Transverse	3	1
Ascending	2	0
Total	1	1

The soiling score at 7 years of age was significantly higher in the younger group than in the older group (1.64 ± 0.48 vs. 0.83 ± 0.69, respectively; $p = 0.02$) (Fig. 2).

Discussion

This study evaluated the bowel function of HD patients who underwent definitive operations using the SD procedure or the TA procedure, to elucidate the optimal timing of surgery to achieve best functional outcomes in terms of bowel function. We divided the patients into those who underwent surgery at < 6 months of age (younger group) and those who underwent surgery at 6 to 12 months of age (older group).

The major findings of this study were as follows: There was no significant difference in the clinical backgrounds of the younger and older groups; the ES improved chronologically after both procedures, with satisfactory results achieved for at least 10 years after surgery; the total ES were comparable at any age; and the soiling scores of the older TA group patients were significantly lower than those of the younger TA group patients at 7 years of age.

Table 5 Operative results of the transanal endorectal pull-through (TA) procedure

	Younger group $n=29$	Older group $n=11$	p
Age at pull-through (days)	89.9 ± 45.89	221.27 ± 33.39	<0.01
Weight at pull-through (g)	5241.45 ± 1370.59	7239.27 ± 1454.86	<0.01
Operation time (min)	293.17 ± 111.45	289.73 ± 96.4	0.93
Blood loss/BW (ml/g)	4.96 ± 4.46	4.95 ± 3.93	0.99
Postoperative hospital stay (days)	20.0 ± 10.4	20.27 ± 10.62	0.94

p values of <0.05 were considered to indicate significance

Pediatric surgeons have developed various definitive operations for HD, including open and laparoscopic procedures [10]. Since De la Torre et al. [11] introduced the TA procedure in 1998, it has been adopted throughout the world because of its simplicity and less invasive nature, allowing pediatric surgeons to treat neonates and small infants. Many surgeons have reported the correlation between the operative procedure and the bowel function of patients with HD; however, these studies have not focused on the timing of the operation. The present study is the first that evaluates the optimal timing of definitive surgery for HD.

In our institution, we usually perform definitive operations when the patient's body weight reaches about 5 kg, although the operation is sometimes delayed if treatment for associated anomalies such as congenital heart disease or chromosomal anomalies takes priority. Most pediatric surgeons in other institutions perform a definitive operation for HD when the patient's body weight reaches about 5 kg. However, individual surgeons and institutional criteria have decided on the timing of surgery without any evidence base. The timing of the TA procedure ranging from newborns to patients of 15 months of age has been reported [12–14]. However, no reports have evaluated the optimal timing for definitive operations. The results of this study suggest that the TA procedure should be performed by at least 6 months of age. From the viewpoint of operative damage after the establishment of the sphincter muscle function, the optimal timing of the TA procedure would be within 4 months of age.

The incision line of mucosectomy on the anal canal may be correlated with postoperative anorectal function. If mucosectomy is commenced too high, then there is a tendency for constipation, whereas if it is commenced too low, then there is a tendency for staining. Denost et al. [15] reported that the risk of fecal incontinence after inter-sphincteric resection for rectal colon carcinoma was directly related to the tumor level and the height of the anastomosis. They reported that a "good" continence result required a distance of > 1 cm between the tumor and the anorectal ring, and anastomosis higher than 2 cm from the anal verge. All of the patients in our series underwent a definitive operation with the same mucosectomy incision line, located 5 mm above the dentate line. Thus, the mucosal incision level of our procedure may have prevented any impairment of anorectal function.

The most serious long-term problem after a definitive operation for HD was fecal soiling rather than constipation. Frequent diarrhea in addition to incontinence and soiling would impair the patient's quality of life. Several surgeons have reported that bowel function improved significantly with age [4]. However, among the patients who underwent the TA procedure in the present study, some suffered these complications until they were 9 years of age. The technical problems of the TA procedure might have contributed to the result. The pediatric surgeons manipulated the internal and external anal sphincter muscle directly and the superficial external anal sphincter muscle was stretched excessively and damaged during anal canal dilatation for transanal manipulation. These muscles were damaged more easily in the TA procedure than in the SD procedure. Moreover, the tissue around the anus in the older patients had matured and the muscle might have been easily damaged, because it was more difficult to stretch than in the younger patients.

Another possible reason is maturation and establishment of the function of the internal and external anal sphincter muscles. In human adults, type I muscle fibers account for approximately 75% of the external anal sphincter muscle [16]. Kuroda et al. evaluated the histological difference of the external anal sphincter muscle in patients with anorectal malformation [17]. They reported that type IIc fibers, defined as primitive and multipotent tissue, were observed in the external anal sphincter muscle of newborns. In contrast, the percentage of type I muscle fibers in patients older than 6 months of age was the same as that in adults. The function of the external anal sphincter muscle is important for preventing fecal soiling [18]. Operative damage after the establishment of sphincter muscle function may impair the bowel function of HD patients, as we observed in our older patients who underwent the TA procedure.

Moore et al. reported that HD is part of a complex spectrum that involves both the neurological condition and the smooth muscle [19]. Thus, a definitive operation before maturation of the external anal sphincter muscle fibers may affect bowel function. Further studies are needed to evaluate the correlation between maturation of the external anal sphincter muscle at the time of the definitive operation and the long-term bowel function of HD patients. The bowel function and evacuation score improved significantly with age in our previous study [4] and those of others [6, 7]. Most patients assessed at 11 years of age had satisfactory bowel function. In the present study, we evaluated bowel function up until 9 years of age. Further studies should be performed to clarify the longer term outcomes and optimal timing of the operation to improve bowel function.

Acknowledgements This study was supported by Grants-in-Aid for Scientific Research from the Japan Society for the Promotion of Science (JSPS: 20K08934, 20K17558, 20K10403, 20K08933, 19K10485, 19K09150, 19K09078, 19K03084, 19K18061, 19K17304, 19K18032, 18K08578, 18K16262 17K10555, 17K11514, 17K10183, 17K11515, 16K10466, 16K10094, 16K10095, 16K10434, 16H07090), research grant from The Mother and Child Health Foundation, research grant from the Kawano Masanori Memorial Public Interest Incorporated Foundation for Promotion of Pediatrics.

Funding No funding was received for this article.

Declarations

Conflict of interest We have no conflicts of interest to declare in association with the present study.

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