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ORIGINAL ARTICLE

Are all patients with short segment Hirschsprung's disease equal? A retrospective multicenter study

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Abstract

Background/purpose Short segment Hirschsprung's disease (HSCR) carries a better prognosis than long segment disease, but the definition of short is controversial. The objective of this study is to determine anatomically the extent of disease involvement that would be associated with a better functional outcome.

Methods This is a retrospective multicenter ($n=3$) study with patients (≥ 3 years) who had transanal pullthrough operation done for aganglionosis limited to the recto-sigmoid colon were reviewed. The extent of disease involvement and bowel resection was retrieved by reviewing the operative records as well as histopathological reports of the resected specimens. Clinical assessment was performed according to the criteria of a seven-itemed bowel function score (BFS) (maximum score = 20). Manometric assessment was performed with anorectal manometry.

Results The study period started from 2003 to 45 patients were studied with median age at assessment = 52.0 months and operation = 3.0 months. The disease involvement was categorized into upper sigmoid-descending colon (DC) ($n=8$), sigmoid colon (SC) ($n=12$), upper rectum (UR) ($n=14$) and lower rectum (LR) ($n=11$) according to the level of normal biopsy result. There was no significant difference in the age of assessment between the four groups. The median BFSs in the DC, SC, UR and LR were 13, 15, 17 and 17, respectively ($p=0.01$). Nine patients from the DC and SC groups reported soiling for more than twice per week. Sub-group analysis comparing patients with and without the entire sigmoid colon resected revealed worse functional outcomes in terms of the incidence of soiling (40.7 vs 22.2%, $p=0.05$) and the BFS (14 vs 18, $p=0.04$) in the former group. Anorectal manometry did not reveal any significant difference between the four groups, but a higher proportion of patients in the UR and LR groups appeared to have a normal sphincter resting pressure (DC vs SC vs UR vs LR = 62.5 vs 75.0 vs 85.7 vs 80.0%, $p=0.10$).

Conclusion Patients with short segment HSCR are not equal at all. HSCR patients with aganglionosis limited to the rectum without the need of removing the entire sigmoid colon have a better bowel control and overall functional score. Less bowel loss and colonic dissection maybe the underlying reasons. Although future studies with a larger sample size and a longer follow-up period are required to validate the results of this study, it has provided a new insight to the current understanding of short segment disease in HSCR.

Keywords Hirschsprung's disease · Aganglionosis · Anorectal manometry · Transanal

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Introduction

Hirschsprung's disease (HSCR) is characterized by the absence of ganglion cells (aganglionosis) in the gut resulting

in functional intestinal obstruction. The incidence is around 1 in 5000 newborns and is slightly more common in Asians [1]. Since the migration of the vagal neural crest cells follows the cranio-caudal direction during the embryonic development, aganglionosis always starts at the most distal part of the rectum with a variable proximal extension. Depending on the disease extension, HSCR is classified into short segment (80%), long segment (15%) and total colonic aganglionosis (5%) [2]. For many years, short segment disease has been broadly defined as aganglionosis limited to the recto-sigmoid colon [3]. It is generally accepted that short segment disease carries a better prognosis. Under this classification, patients with aganglionosis requiring the resection of a small length of the rectum or the entire recto-sigmoid colon would be grouped under the same category as short segment disease in most of the clinical studies on HSCR. However, it is well known that the sigmoid colon plays an important role in maintaining a normal bowel function and therefore, the functional outcome should be different when it is resected or left in-situ [4]. Moreover, the function of the residual cells maybe different in patients with a different degree of aganglionosis. Therefore, grouping all the patients with rectal or recto-sigmoid disease under the same category maybe oversimplified and cannot appropriately reflect the true prognosis of different patients. The objective of this study is to evaluate whether patients with different length of 'short segment' HSCR indeed carry the same prognosis and to determine anatomically the extent of disease involvement that would be associated with the best functional outcomes based on clinical and manometric assessments.

Materials and methods

Subjects

This is a retrospective multicenter study conducted in the three tertiary pediatric surgical centers in our locality. The study period started from 2003 when the first transanal endorectal pullthrough (TEPT) procedure was performed. The medical records of HSCR patients currently older than 3 years with previous TEPT operation were reviewed. Only those with the aganglionosis limited to the recto-sigmoid colon were selected for further evaluation. The participants who have given their consents were invited to undergo clinical and anorectal manometric studies to assess their anorectal functions. Patients with severe learning disability; inability to co-operate during the study; concomitant anorectal/neurological anomaly and history of re-do operation due to failure of previous procedure were excluded. The original pathology reports (including the intra-operative frozen section record as well as the full report of the resected colon) were reviewed. The histological features being noticed were mainly the first site (the most distal part of colon) where there were an adequate number of normal-looking ganglion cells. In addition, the presence of transition pullthrough was checked by reviewing the transection margin. The patients were divided into four groups [upper sigmoid-descending colon (DC), sigmoid colon (SC), upper rectum (UR) and lower rectum (LR)] according to the most distal level of normal ganglionic innervation. The results of clinical and manometric assessments in different groups were compared. This study has been approved by the hospital ethic committee and was done in accordance with the principles outlined in the Declaration of Helsinki.

Clinical assessment

Clinical assessment was conducted with the seven-itemed bowel function score (BFS) proposed by Rintala [5] (Table 1). This multivariate scoring system assesses several issues such as the ability to hold and report defecation, frequency and severity of constipation and soiling as well as the social impact. These questions required simple answers which could be responded by the patients or their caretakers. The maximum total score is 20 with the median and mean scores for healthy control being 20 (range 14–20) and 19.1 ± 1.3 according to previous studies [6, 7]. A value of ≥ 18 (more than 90% of controls) was taken as the lower limit of normality in this study.

Anorectal manometry

Anorectal manometry is a non-invasive test to assess the anorectal physiology. In this study, the participants underwent manometric assessment in their respective centers where the primary operation was performed. The manometry systems used in different centers were products of the same company [medical measurement systems (MMS)] and the parameters concerned in this study (sphincteric resting/squeezing pressure and anorectal sensation) were measured in the same way among the three centers. Combining the data from two previous studies regarding the manometric findings in pediatric population as well as the values from six age-matched healthy children recruited for another study, the reference values for normal sphincteric resting pressure was 30–60 mmHg and squeezing pressure was 50–120 mmHg [8, 9]. Anorectal sensation was recorded by distending the balloon with various volume of air. The procedures were performed without sedation and all the participants were discharged on the same day.

Data collection and processing

The data were analyzed with standard statistical package [Windows version 21.0; SPSS Inc, Armonk (NY), US].

Table 1 The seven-itemed bowel function scoring system proposed by Rintala was used as the clinical assessment tool

Ability to hold back defaecation	
Always	3
< 1X/week	2
Weekly	1
No voluntary control	0
Feels/reports the urge to defaecate	
Always	3
Most of time	2
Uncertain	1
Absent	0
Frequency of defaecation	
Every day to twice a day	2
More often	1
Less often	0
Soiling	
Never	3
Stain < 1X/week	2
Frequent staining/soiling	1
Daily soiling	0
Accidents	
Never	3
< 1X/week	2
Weekly	1
Daily	0
Constipation	
Never	3
Manageable with diet	2
Manageable with laxatives	1
Manageable with enemas	0
Social problems	
Never	3
Sometimes	2
Restriction in social life	1
Severe social problems	0
Maximum score	20

The maximum score is 20 and 18 was considered as the lower limit of normality in this study

Continuous variables were expressed as medians (ranges) and compared using the Kruskal–Wallis test. Categorical variables were compared using the chi-square test. A p value of less than 0.05 was considered to be statistically significant.

Results

Since 2003, a total of 86 patients from the three centers have suffered from HSCR and 64 patients were classified of having short segment disease with TEPT procedure performed. Eleven patients met the exclusion criteria and another 8 patients did not consent for the manometric procedure. As a result, 45 patients were included in this study (Table2). There were more male patients (male:female = 34:11) and the overall median age was 52.0 months (range 36–172 months). The median ages at diagnosis and the time of pullthrough operation were 1.5 months (range 0.5–56 months) and 3.5 months (range 0.5–60 months) respectively. Eight patients (17.8%) required a defunctioning stoma before TEPT procedure because of intestinal obstruction or enterocolitis. The operations were performed in three centers with eight surgeons involved. The all followed the standard principles of TEPT procedure although minor technical variations existed between different centers. The overall incidence for constipation (defined according to the Rome III criteria) and soiling (more than twice per week) were 17.8 and 33.3%, respectively. The overall median BFS was 16 (range 7–20). Five patients had developed recurrent (more than once) postoperative enterocolitis. According to the classification, the patients were categorized into DC ($n = 8$, 17.8%), SC ($n = 12$, 26.7%), UR ($n = 14$, 31.1%) and LR ($n = 11$, 24.4%). In all the cases, the transection site was found to have an adequate number of normal-looking ganglion cells and the possibility of transition zone pullthrough was excluded.

Comparing the four different groups, there was no significant difference in the median age at operation (DC vs SC vs UR vs LR = 4.0 vs 2.5 vs 3.5 vs 4.5 months, $p = 0.82$) or assessment (DC vs SC vs UR vs LR = 60.0 vs 46.5 vs 60.5 vs 55.0 months, $p = 0.69$). The incidence of constipation (defined according to the Rome III criteria) was highest in the SC group, but this was not statistically significant (DC vs SC vs UR vs LR = 12.5 vs 25.0 vs 14.3 vs 18.2.0%, $p = 0.12$). On the other hand, the incidence of soiling (more than twice per week) was significantly highest in the SC group followed by the DC group (DC vs SC vs UR vs LR = 37.5% vs 50.0 vs 28.5 vs 18.2%, $p = 0.04$). The median BFSs in the DC, SC, UR and LR were 13, 15, 17 and 17, respectively ($p = 0.01$). There was no significant difference in the incidence of recurrent post-pullthrough enterocolitis (DC vs SC vs UR vs LR = 12.5 vs 16.6 vs 14.3 vs 0%, $p = 0.45$). The results of clinical assessment of the four groups were summarized in Table 3.

The clinical outcomes between patients with different levels of colonic resection were also compared (Table 4). Among the 45 patients, 18 patients from the LR and UR groups had colonic transection at the level or distal to the distal sigmoid colon (i.e., recto-sigmoid junction or upper rectum) while the rest of the patients ($n = 27$) all had colonic transection proximal to the distal sigmoid colon that was regarded as having a recto-sigmoidectomy. There was no significant difference in the incidence of constipation between the two groups but patients with the entire sigmoid colon resected were found to have a higher incidence of soiling

Table 2 Demographic data of the 45 patients recruited in the current study

Variables	$n=45$ % (n) or median (range)
Sex	
Male	75.6% ($n=34$)
Female	24.4% ($n=11$)
Current age (months)	52 (36–172)
Age at diagnosis (months)	1.5 (0.5–56)
Age at pullthrough operation (months)	3.5 (0.5–60)
Number of patient required pre-op stoma	8 (17.8%)
Incidence of constipation*	17.8% ($n=8$)
Incidence of soiling (> 2 times per week)	33.3% ($n=15$)
Bowel function score	16 (7–20)
Incidence of recurrent post-pullthrough enterocolitis	11.1% ($n=5$)
Level of aganglionosis	
Upper sigmoid-descending colon (DC)	17.8% ($n=8$)
Sigmoid colon (SC)	26.7% ($n=12$)
Upper rectum (UR)	31.1% ($n=14$)
Lower rectum (LR)	24.4% ($n=11$)
Level of bowel transection	
Proximal to distal sigmoid colon (recto-sigmoidectomy)	60% ($n=27$)
At the level or distal to distal sigmoid colon	40% ($n=18$)

*Constipation is defined according to the Rome III criteria

Table 3 Comparison of clinical assessment results between different levels of aganglionosis

	DC ($n=8$)	SC ($n=12$)	UR ($n=14$)	LR ($n=11$)	p value
Current age (months)[#]	60.0 (50–126)	46.5 (40–132)	60.5 (38–172)	55.0 (36–150)	0.69
Age at pullthrough operation (months)[#]	4.0 (2–32)	2.5 (1–36)	3.5 (1–60)	4.5 (0.5–20)	0.82
Incidence of constipation*	12.5% ($n=1$)	25.0% ($n=3$)	14.3% ($n=2$)	18.2% ($n=2$)	0.12
Incidence of soiling (> 2 times per week)	37.5% ($n=3$)	50.0% ($n=6$)	28.5% ($n=4$)	18.2% ($n=2$)	0.04
BFS	13 (7–16)	15 (8–17)	17 (14–20)	17 (12–20)	0.01
Incidence of recurrent post-pullthrough enterocolitis	12.5% ($n=1$)	16.6% ($n=2$)	14.3% ($n=2$)	0% ($n=0$)	0.45

*Constipation is defined according to the Rome III criteria

[#]Values are expressed as median (range)

Table 4 Comparison of clinical assessment results between different levels of colonic resection

	Proximal to distal sigmoid colon (recto-sigmoidectomy) ($n=27$)	At the level or distal to distal sigmoid colon ($n=18$)	p value
Current age (months)[#]	78.0 (48–144)	66.0 (36–172)	0.32
Age at pullthrough operation (months)[#]	5.0 (0.5–60)	4.0 (0.5–48)	0.57
Incidence of constipation*	18.5% ($n=5$)	16.7% ($n=3$)	0.13
Incidence of soiling (> 2 times per week)	40.7% ($n=11$)	22.2% ($n=4$)	0.05
BFS	14 (7–16)	18 (13–20)	0.04
Incidence of recurrent post-pullthrough enterocolitis	14.8% ($n=4$)	5.5% ($n=1$)	0.19

*Constipation is defined according to the Rome III criteria

[#]Values are expressed as median (range)

(constipation = 18.5 vs 16.7%, $p = 0.13$ and soiling = 40.7 vs 22.2%, $p = 0.05$). The median BFS was also significantly lower in patients with complete loss of the sigmoid colon (14 vs 18, $p = 0.04$). The incidence of recurrent post-pullthrough enterocolitis was higher in patients with recto-sigmoidectomy but the results was not statistically significant (14.8 vs 5.5%, $p = 0.19$).

Regarding manometric assessment, 75.6% patients ($n = 34$) were found to have a normal sphincteric resting pressure. The results of sub-group analysis showed that a higher proportion of patients in the UR and LR groups had a normal sphincteric resting pressure, but this was not statistically significant (DC vs SC vs UR vs LR = 62.5 vs 75.0 vs 85.7 vs 72.3%, $p = 0.10$). The median values of sphincteric resting pressure in the DC, SC, UR and LR groups were 25.0 vs 43.0 vs 38.0 vs 29.0 mmHg, respectively ($p = 0.37$). The median values of squeezing pressure between the four groups were comparable without statistical significance (DC vs SC vs UR vs LR = 55.0 vs 68.0 vs 45.0 vs 62.0 mmHg, $p = 0.57$). Regarding anorectal sensation, the median values for the volume of air to elicit the first anal sensation were 30, 40, 20 and 20 ml in the DC, SC, UR and LR groups respectively ($p = 0.88$). The results of manometric assessment were summarized in Table 5.

Discussion

Unlike the other anorectal disorders with more precise anatomical description, HSCR is traditionally divided into three board categories based on the extent of aganglionosis. Short segment HSCR, which is widely accepted as aganglionosis limited to the recto-sigmoid colon, is the most common form of this disease. The operative principle for HSCR surgery includes the resection of the aganglionic bowel while preserving as much normal bowel as possible. As a result, the actual segment of bowel resected in each individual patient is actually different despite they are grouped under the same category of 'short segment disease'. The underlying physiology responsible for a normal bowel function is highly complex. The length of residual bowel in particular the presence or absence of the sigmoid colon as well as the function in the residual cells are an important determinant of the subsequent anorectal function [10]. Therefore, the functional outcome of aganglionosis involving a small part of the rectum is expected to be different from those with more extensive involvement.

The current study focused on the patients after transanal endorectal pullthrough procedure which is one of the most popular operations for short segment HSCR in recent years after multiple studies showing its favorable results compared to other techniques [11–14]. Only one operation was chosen to minimize the difference related to the surgical procedure. Although the operations were carried out in different centers by multiple surgeons, the operative principle was similar and followed the original description by De la Torre and Ortega [15]. In all the operations, bowel transection was performed at 5–10 cm proximal to the normal biopsy site according to the individual surgeon's judgement. Therefore, the patients with aganglionosis at the sigmoid-descending junction with bowel transection at the descending colon were also included in this study. A short length of aganglionic muscular cuff was left behind in the distal rectum and colo-anal anastomosis was performed transanally above the dentate line using absorbable sutures. To achieve a definitive judgement about the bowel function and an accurate manometric assessment, only patients older than 3 years with toilet training were invited to participate in this study and thus eliminating other factors which may affect the assessment.

The overall incidences of post-pullthrough constipation and soiling in the current study were less when compared with those reported in international publications [16–18]. This maybe attributed to the differences in the definition of constipation and frequency of soiling used in this study. Patients with a shorter segment of aganglionosis (UR and LR) were found to have higher BFS and hence better bowel function than those with aganglionosis extending into the sigmoid colon (DC and SC). We postulate that the loss of the entire sigmoid colon maybe the underlying reason. In patients with a shorter length of aganglionosis (UR and LR), less colonic dissection is required and the sigmoid colon is at least partially or completely preserved. This postulation is supported by the other sub-group analysis which revealed a better functional outcome in patients without the entire sigmoid colon resected. The curve-shaped sigmoid colon is normally responsible for the temporary storage

Table 5 Comparison of anorectal manometry assessment results between different levels of aganglionosis

	DC ($n = 8$)	SC ($n = 12$)	UR ($n = 14$)	LR ($n = 11$)	p value
Sphincteric resting pressure (mmHg)[#]	25.0 (10.0–53.5)	43.0 (26.5–58.0)	38.0 (15.5–46.5)	29.0 (11.0–41.5)	0.37
% of patients with normal sphincteric resting pressure	62.5% ($n = 5$)	75.0% ($n = 9$)	85.7% ($n = 12$)	72.3% ($n = 8$)	0.10
Sphincteric squeeze pressure (mmHg)[#]	55.0 (18.5–80.0)	68.0 (23.0–92.5)	45.0 (25.5–76.5)	62.0 (20.0–94.5)	0.57
Volume of air to elicit the first anal sensation (ml)[#]	30 (10–50)	40 (10–60)	20 (10–60)	20 (10–40)	0.88

[#]Values are expressed as median (range)

of feces and losing it will result in the disturbance of the braking system [19]. This may explain the observation that the patients with complete recto-sigmoidectomy performed had a higher incidence of soiling, which is close to statistical significance, than those who had the sigmoid colon at least partially preserved. The overall number of recurrent post-pullthrough enterocolitis is too small ($n=5$) for a statistically significant analysis but it seems that removing the entire colon maybe associated with a higher incidence of this complication. Another postulation to the observed differences between patients in the four different groups is related to the degree of the underlying cellular dysfunction in the remaining colon. However this postulation cannot be proved in the current clinical study and should be addressed in future laboratory studies.

Anorectal manometry was also performed to provide an objective assessment of the sphincteric function. Its application as a follow-up study for HSCR has been reported previously [20]. The majority of patients could still have a normal sphincteric pressure suggested that the injury due to stretching during the transanal procedure may not be a severe one as reported in the other study [21]. Although a higher proportion of patients with aganglionosis limited to the rectum (UR and LR) had a normal sphincteric pressure, this finding was not statistically significant. The actual sphincteric resting and squeezing pressures were also similar between the four groups without statistical significance. The lack of significant differences in the comparisons maybe related to the small sample size. However, it may also reflect that the injury of the anal sphincter which accounts for more than 85% of the resting pressure was similar during the operations [22]. Since the degree of anal stretching during the operations was similar regardless of the extent of disease involvement, the severity of injury to the anal sphincter should be the same. Some patients could still retain a normal sphincteric resting pressure suggested that leaving a short muscle cuff does not seem to create the problem of increasing the anal pressure. In addition, a comparable result in the assessment of anorectal sensation is another piece of evidence to suggest that the operative damage to the anorectal innervation was similar between the four groups.

According to the manometry findings, there was no significant differences observed in all the measurements between the four groups and we postulated that the most likely reason would be the small sample size in this study. On the other hand, patients with disease limited to the rectum had a significantly better functional outcome in term of soiling and the BFS. Besides the small sample size, we believe the lack of correlation between manometric findings and the clinical outcomes may also be related to the underlying physiology of bowel control. The control of defecation also involves other mechanisms (such as large bowel motility, personal habit etc) which were not assessed by anorectal manometry. Furthermore, some of the items in the BFS such as the severity of constipation (diet/laxative/enema dependent) and social impact are relatively subjective and hence the findings of clinical assessment maybe different from those of manometric assessment.

This study is limited by its retrospective nature and is associated with different forms of bias. In addition, this is a multicenter study and multiple surgeons from different centers were involved in the operations. To minimize the variation in surgical procedure, we therefore included only patients receiving TEPT. According to our understanding, the operating surgeons all followed the principle of TEPT, but variations in operative techniques and the length of the muscle cuff did exist. The analysis was therefore subjected to these confounding factors. There was also a lack of consensus about the optimal level of bowel transection with respect to the normal biopsy site. This variation in practice might have resulted in patients with the same extent of aganglionosis undergoing different levels of colonic transection. Future prospective study with a standardized protocol is recommended to address these limitations.

Conclusion

In conclusion, HSCR patients who are under the traditional classification of ‘short segment disease’ actually have a different functional outcome depending on the actual extent of the aganglionosis and bowel resection. Patients with aganglionosis limited to the rectum without the need of removing the entire sigmoid colon appeared to have a better post-operative bowel function and control than those with more proximal extension. Combining clinical and manometric assessments, the differences in the functional outcomes are believed to be related to less colonic resection and the preservation of the sigmoid colon instead of the effect of the operation. Future clinical and laboratory studies with a larger sample size and longer follow-up period are required to explain the results of this study. Nonetheless, this study has provided a new insight to the current definition and understanding of ‘short segment disease’ in HSCR. Patients labeled as having short segment HSCR but requiring the resection of a major part of the sigmoid colon may represent a different disease entity. Special considerations should be given to this group of patient during peri-operative counseling as well as post-operative bowel management program. Patients with short segment disease are not equal at all.

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Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest with respect to this manuscript.

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