Bowel function after transanal endorectal pull-through for Hirschsprung disease – does outcome improve over time?

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ABSTRACT

Background: The reported functional outcome in patients operated with transanal endorectal pull-through (ERPT) for Hirschsprung disease (HD) varies greatly. Some studies suggest better functional outcome in older than in younger HD patients, but there are almost no longitudinal studies that have demonstrated such improvement. Therefore, we aimed to compare functional outcome in a cohort of HD patients over time to assess whether bowel function improves with increasing age.
Methods: Functional outcome in HD patients operated with ERPT from 1998 to 2007 was recorded by standardized interviews by an independent investigator during 2008–2011 and again in 2017/2018. Bowel function was assessed using the Krickenbeck questionnaire. Clinical data were collected prospectively.
Results: 50 of the original 62 patients responded for a second interview. Median age at the two interviews was 8,1 (3,4-16,6) and 15,4 (9,9–25) years respectively. There was no difference in the rate of soiling at first (52%) and second (52%) follow-up. Constipation was reported in 20% of the patients at first, and in 24% at second follow-up. Bowel management was used by 30% and 32% at first and second interview respectively.
Conclusion: Soiling and constipation are common in HD patients several years after surgery, and no improvement of bowel function with increasing age could be demonstrated.
Level of Evidence: Level II.

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Hirschsprung disease (HD) is a congenital disorder characterized by functional intestinal obstruction due to lack of ganglion cells in the distal bowel. The treatment is surgical and entails resection of the affected aganglionic segment and anastomosis of the proximal ganglionic bowel to the distal rectum [1]. Over the past two decades, a minimally invasive, one-stage operation has been established as the preferred surgical strategy [2]. One of the most common procedures is the transanal endorectal pull-through (ERPT), performed with or without laparoscopic or laparotomy-assisted mobilization of the aganglionic colonic segment. ERTP can be carried out in infancy and is considered safe and efficient [4,3]. There is, however, concern about how a low colorectal anastomosis and dilatation of the anal sphincters during the transanal stage of the operation may affect fecal continence [4,5].

A significant number of HD children experience impaired bowel function postoperatively [6,7]. Both soiling and constipation are reported. There is a general assumption among pediatric surgeons that bowel function improves over time in HD patients. This belief is supported by various recent articles, some presenting results from a large number of patients [8,9]. Most of these studies compared bowel function in patients of different age, where older patients as a group proved to have better outcome. However, there are multiple factors that can affect outcome besides age; modification of surgical technique over time, different surgeons, age at time of surgery, and shifting trends regarding follow up procedures. In addition, the majority of the studies have a retrospective design, and conclusions are based on reviews of medical charts. Furthermore, an independent investigator has rarely assessed bowel function, and standardized questionnaires for evaluation of bowel function have seldom been applied. Only a few studies have offered longitudinally collected data allowing direct comparison of outcome over time in HD patients, their findings suggesting that incontinence may remain a problem, whereas constipation should be expected to reduce as the patients grow older [10]. However, to the best of our knowledge, no studies to date have followed the same patients beyond early childhood. We have previously reported functional outcome in HD children undergoing ERPT, and we found reduced fecal control in a significant number of patients [11,12]. As these patients now are reaching adolescence and adulthood, we aimed to assess long-term functional outcome and examine if they had gained a better functional result with increasing age, and thereby test the hypothesis that bowel function improves with age in HD patients.

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1. Method

1.1. Patients

All 62 patients included in a previous study on functional outcome in HD patients were eligible for inclusion in this follow-up study [11,12]. The patients had undergone ERPT at our hospital between 1998 and 2007, and they were prospectively followed according to the department's research protocol. Demographic data were collected at the time of surgery and during follow-up.

1.2. Surgical management

HD was histologically verified by rectal biopsies, and contrast enemas were used to identify the transition zone in all patients prior to surgery. The ERPT was performed as previously described. Shortly, the operation was initially performed in combination with laparotomy before introduction of the completely transanal pull-through in 2001 [11]. In all patients a short, intact muscle cuff was left, and the colorectal anastomosis was fashioned 5–10 mm oral to the dentate line. Frozen sections were taken to confirm presence of ganglion cells in the colon at the anastomotic level.

An one-stage endorectal pull-through was intended in all patients. However, 17 patients had a preoperative diverting colostomy performed before introduction of the one-stage procedure in 1998, or as an emergency procedure for ileus or perforation. Routine anal dilatations started 3 weeks postoperatively, and lasted at least 5 weeks after surgery.

1.3. Interviews

Stoma nurses performed the standardized interviews during outpatient visits between 2008 and 2011 (FU1) to record bowel function. The second interviews were done by telephone by the first author (MVF) during 2017-2018 (FU2). MVF had not been involved in the treatment of the patients, and an identical interview was conducted. In patients less than 12 years of age or with cognitive impairment, the parents were interviewed, or interviews were undertaken in their presence.

1.4. Definitions

Bowel function was classified according to the Krickenbeck score (Table 1). Although not a validated score, it is frequently used to describe bowel function in patients operated for HD and anorectal malformations [13].

Patients using regular enemas, either antegrade or retrograde, or had a stoma due to bowel problems, were automatically given the poorest score. Soiling was defined as involuntary leaking of stool, requiring change of underwear or use of protective pads. Patients who stated general ability to feel urge and to hold the bowel movement until they reached the nearest toilet, or at least for 15 min, were defined as having voluntary bowel movements. Impact on social life was added as an open question at FU2 and was not classified using a scoring system, but graded 0–3 on a scale where 0 denotes severe impact on

social life, 1 restrictions to social life, 2 few adjustments to social life and 3 no impact on social life.

1.5. Ethics

The study has been approved by the institutional review board (2017/4913).

1.6. Statistics

Continuous parameters are presented as median and range, and were analyzed with Mann–Whitney U Test. Categorical variables were compared with a Chi Square test and Fischer's Exact test where applicable. Krickenbeck scores were analyzed as categorical variables, as the Krickenbeck classification is not a linear score. A P value <0.05 was considered statistically significant. Data analyses were performed with IBM SPSS for Windows Version 24.0. Armonk NY, IBM Corp, Released 2016.

2. Results

62 patients were interviewed at FU1. 50 (79%) patients agreed to a second interview (FU2) and were included in this study. 10/50 (20%) were girls. Six patients had an associated syndrome (Downs syndrome 5, Goldberg-Sphrintzen syndrome 1). One boy has since the primary operation been diagnosed with ulcerative colitis, and one girl has been reoperated with colectomy and an ileoanal reservoir due to residual aganglionosis. Age at ERPT was median 89 (11–4035) days. 35/50 (70%) patients were operated completely transanally, whereas 15/50 (30%) patients had a laparotomy-assisted procedure. 41/50 patients had aganglionosis restricted to the rectosigmoid colon. Median age was 8,1 (3,4-16,6) and 15,4 (9,9–25) years at FU1 and FU2, respectively.

Details of continence and constipation at FU1 and FU2 for both syndromic and non-syndromic patients are presented in Table 2. 52% (26/50) of the patients reported soiling at FU1. At FU2, this number had not changed. Daily soiling was reported by 38% (19/50) at FU1 and by 34% (17/50) at FU2. 18% (9/50) of the patients reported worsening of soiling from FU1 to FU2, 20% (10/50) reported improvement, whereas the majority (62%) reported their situation unchanged.

The incidence of soiling was higher at FU1 in patients with an associated syndrome or illness (63%, 5/8) than in otherwise healthy patients (50%, 21/42, p = 0,704), and at FU2, the discrepancy was even larger, with 88% (7/8) of syndromic patients reporting soiling, as opposed to 45% (19/42) in the non-syndromic group (p = 0,056).

Constipation was less common than soiling at both FU1 and FU2. At FU1, 20% (10/50) of the patients reported constipation requiring dietary adjustment, use of laxatives or bowel management, as opposed to 24% (12/50) at FU2. None of the syndromic patients reported constipation at either follow-up.

At FU1 15 patients used some form of bowel management (appendicostomy (ACE) 5, regular rectal enemas 5, colostomy/

Table 1
The Krickenbeck classification system for assessment of bowel function.

Krickenbeck classification	
1. Voluntary bowel movements	Yes/No
Feeling of urge, capacity to verbalize, hold the bowel movement	
2. Soiling	Yes/No
Grade 1: Occasionally (once or twice per week)	
Grade 2: Every day, no social problem	
Grade 3: Constant, social problem	
3. Constipation	Yes/No
Grade 1: Manageable with diet	
Grade 2: Requires laxatives	
Grade 3: Resistant to diet and laxatives	

Table 2
Demographics of patients with Hirschsprung disease operated with transanal endorectal pull-through. Patients were interviewed at two time points; FU1 and FU2.

	No syndrome (n = 42)		Syndrome (n = 8)	
	FU1	FU2	FU1	FU2
Age (years)	7,7	15,3	8,7	17
Soiling				
None	21 (50%)	23 (55%)	3 (38%)	1 (13%)
Grade 1	7 (17%)	7 (17%)	0	2 (25%)
Grades 2–3	14 (33%)	12 (28%)	5 (62%)	5 (62%)
Constipation				
None	32 (76%)	30 (71%)	8 (100%)	8 (100%)
Diet/laxatives	5 (12%)	4 (10%)	0	0
Resistant	5 (12%)	8 (19%)	0	0

Table 3
Characteristics of patients included and not included at the second follow-up.

	Respondents (n = 50)	Non-respondents (n = 12)	P
Age			
Operation (days)	78	398	*0,021*
FU1[1] (years)	7,7	10,7	*0,200*
Syndrome	8 (16%)	1 (8,3%)	*0,675*
Sex			
Girls	10 (20%)	0 (0%)	*0,185*
Boys	40 (80%)	12 (100%)	
Incontinence FU1			
None or grade 1	31 (62%)	9 (75%)	*0,512*
Grades 2–3	19 (38%)	3 (25%)	
Constipation FU1	10 (20%)	2 (17%)	*1,000*

[1] FU1: First follow-up.

ileostomy 5). At FU2 the corresponding number was 16 (ACE 11, regular rectal enemas 4, colostomy/ileostomy 1). 81% (13/16) of the patients using bowel management at FU2 also used bowel management at FU1, but not necessarily in the same form. 41% (7/17) of the patients who reported daily soiling at FU2 did not use any form of bowel management.

At last follow up 7% (3/42) of non-syndromic patients reported their social life to be restricted, and 24% (10/42) made some adjustments to their social life as a direct consequence of impaired bowel function. In syndromic patients none found themselves restricted, but 25% (2/8) reported to make adjustments due to bowel problems.

2.1. Drop-out analysis

A drop-out analysis revealed that there were no significant differences in the rate of soiling or constipation between respondents and non-respondents at FU1 (Table 3).

3. Discussion

The main finding of this longitudinal cohort study in HD patients operated with ERPT, was that no improvement of bowel function from early school-age to mid-teens could be demonstrated. This finding contrasts the general assumption that bowel function in HD patients improves with age since several studies have shown better bowel function in older than in younger HD patients [8,9,14]. There are several plausible explanations for the differing results. In contrast to previous studies, which are mostly cross-sectional and comparing outcome of younger with that of older patients, this is a prospective follow-up study in which we have assessed bowel function over time in the same patient cohort. Additionally, we applied standardized, objective questionnaires, and an independent investigator communicated with the patients. This differentiates from several studies where bowel function was either registered from hospital records or by the surgeon.

The dominating long term complaint in this study was soiling. Patients with soiling did not have constipation. Therefore we believe the soiling was caused by true fecal incontinence and not overflow incontinence. Important contributors to fecal continence are the anal sphincters, colonic motility, anorectal sensation, and the sub-epithelium of the anal canal [5,12,15–17]. All these factors can be affected during transanal surgery, and thereby cause soiling. Especially, focus has been drawn to anal sphincter damage and the distance from the dentate line to the level of the anastomosis. If there is no intact anal canal and/or damage to the anal sphincters, one cannot expect that bowel function will significantly improve even if the patient gets older. Thus, to preserve the anal canal and avoid sphincter damage are of vital importance during the ERPT.

Contributing to the reported high rate of soiling at FU2, may be that the patients' expectations of normal bowel function change as they grow older. A child may be more inclined to consider minor soiling as normal, whereas an adolescent or adult may not tolerate even slight staining of underwear. Consequently, our definition of fecal soiling as

the need for change of underwear and/or use of pads, may not precisely describe the actual grade of soiling, and minor improvements in bowel function may not have been picked up. Furthermore, we have given all patients requiring bowel management a poor score and chosen not to look at them separately, as we regard the need for such measures to be indicative of a poor functional result after primary surgery.

The rate of patients needing bowel management varies in comparable studies [8,9,14], and is relatively high in this patient population. The indication for starting bowel management may be different in different institutions. Based on previous studies on the detrimental effect of soiling on psychosocial health [18,19], our team is extremely focused on achieving perfect continence, enabling the patients to live their lives feeling completely sure that they will not experience any fecal accidents. Therefore, patients experiencing only occasional soiling may be offered bowel management. Also, in our experience many patients are reluctant to try to wean off bowel management because it enables them to control their bowel movements in a predictable manner.

The actual percentage of patients using bowel management did not change during the follow-up period. However, the type of bowel management changed as particularly ACE became more common among the older patients. This practice reflects our department's tradition of avoiding rectal procedures in all patients that have the slightest reluctance for rectal enemas because several studies have shown psychological negative effects of rectal procedures [20].

Constipation was less common than incontinence at both follow ups, and the rates are similar to those of comparable studies. However, we found little improvement with increasing age. As for soiling, it is possible that the anastomotic level plays an equally important role when it comes to residual obstructive symptoms. Intestinal dysmotility and internal sphincter achalasia may also contribute to difficulties emptying the bowel.

The main strengths of this study are the longitudinal design, that standardized questionnaires were used, and that an independent researcher assessed bowel function. The relatively small number of patients and the heterogeneity of the cohort are the main limitations, challenging the power of statistical analyses. Also, even though the interviews were intended to be similar, two different persons performed the interviews, and this may have influenced the results. We did not reach all patients for a second interview. However, the dropout analysis showed that there were no major differences between the patients who were included and those who were not. Therefore, we believe the participating patients are representative of our patient population. A more extensive follow-up span could also have been beneficial as the age difference among the patients is fairly large. It is possible that the youngest patients would have experienced improvement if observed longer, and, more importantly, that the oldest patients at FU1 may already have reached their potential for improvement. Lastly, we have not performed a systematic assessment of the integrity of the patients' anal canal with inspection, manometry and anal endosonography in this study. This would undoubtedly have provided valuable information in interpreting outcome and identifying predictive factors for functional result.

4. Conclusion

To conclude, this longitudinal study in HD patients has not shown any improvement in bowel function as patients reach adolescence and early adulthood. Thus, when pediatric surgeons counsel HD patients and their parents, it is important to give a realistic view on what to expect in regards to future bowel function and potential for long term improvement.

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