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**The development of frequency of stools over time in children with
Hirschsprung's disease post transanal endorectal one-stage pull-through.**

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Running title: Bowel symptoms in Hirschsprung's disease

Abstract

Background: The transanal endorectal one-stage pull-through (TERPT) procedure in children with Hirschsprung's disease (HD) is frequently used worldwide. In order to give the children's families realistic expectations and to plan the medical care for the period after TERPT, the outcome is of great importance.

Aim: To collect information on the number of stools passed daily after one stage TERPT procedure for HD.

Method: A prospective follow-up study for collecting information on the outcome of planned TERPT from 2005 through 2012 was carried out. A control group consisting of age and gender matched children was used.

Results: The results show an initial high frequency of daily stools, median 12 (3-30), reaching an acceptable situation with median 4 (0-10) stools/day after one year. After four years the number of stools did not differ significantly from healthy controls.

Conclusion: This study shows that it takes four years after TERPT before the number of stools becomes normalized. In order to compare the long term outcome, it would be desirable to have uniform regular reports on the daily frequency of passed stools, incontinence and constipation during the years after TERPT.

Keywords: Transanal endorectal Pull-Through (TERPT); Hirschsprung's Disease (HD); Outcome; Incontinence; Constipation

Background

Hirschsprung's disease (HD) is a rare condition with about 3-4:10000 afflicted newborns. Even though this is a small group of patients, it is a group that requires close monitoring and frequent follow-ups at centers for pediatric surgery.

The transanal endorectal pull-through (TERPT) technique for Hirschsprung's disease, first described in 1998 [1], was intended to be less invasive than prior methods. With minimal intraabdominal dissection the risk of intraabdominal adhesions diminishes. No scars in the perineum mean better cosmetic results and preserve the pelvic structures and innervation.

It is generally known that most patients, initially after the TERPT procedure, will have more frequent stools but that this stabilizes over time [2, 3]. Patients will have to cope with various symptoms both short-term and sometimes lifelong as a result of a shortened intestine, eventual damage to the surrounding structures in the pelvic floor and the possibility of the bowel being left dysfunctional [4].

The few studies that have reported the functional outcome after TERPT [4 – 6] concentrate on the long-term outcome regarding incontinence and constipation. However, knowledge about the short-term outcome during the first year after TERPT and the possible complications is of great importance for the preoperative information and medical planning. A review article [7] finds no published report on the daily frequency of stools. The aim of the study is to report the outcome of TERPT with regard to the daily frequency of stools and its correlation to the length of time after the TERPT procedure.

Material and methods

The study was carried out at a tertiary center for pediatric surgery serving a region with 1.8 million inhabitants and with a nativity of 22 000 newborns per year. In this region all inhabitants have access to health care free of charge at the time when needed and, thus, dropouts due to socioeconomic factors are not expected.

This study included routine prospective follow-up of the patients operated on with HD. The protocol was designed to meet the legislative documentation required in

the country of origin and the regional research ethics committee approved the study (registration number 2010/49). The data are presented in such a way that it is impossible to identify any single patient or control and therefore it was not necessary to obtain approval from the individual patients' guardians.

All children diagnosed with and treated for HD between January 2005 and June 2013 were identified and collected from a prospectively maintained database. A review of their medical records was performed. The data in the prospectively maintained database were collected during the in- and out-patient visits and were asked according to a structured questionnaire.

The diagnosis of HD was made by rectal suction biopsy and a full wall thickness rectal biopsy preoperatively during a prospective study of the two methods [8]. All the patients also underwent preoperative contrast enema to identify the transition zone and if the recto anal relaxation reflex (RAIR) could be disclosed [9]. The patients were then operated on either with one-stage TERPT or primarily with colostomy followed by the TERPT procedure at a later stage. Intraoperative frozen section biopsies were used to confirm ganglionic intestine and were obtained transanally after mobilization of the transition zone. Three experienced colorectal pediatric surgeons carried out all the TERPT procedures, usually at least two together.

After surgery, the patients were monitored at the same tertiary center. The follow-up data were obtained prospectively by the investigators. Initially the follow-up was more frequent and with time, as the symptoms stabilized, with longer intervals according to the children's need. The oldest patient included in the study was 63 months at the last outpatient visit. At each follow-up the guardians were asked about frequency of stools, constipation, and use of medication, perineal retching or other problems they may have experienced.

The patients started with anal calibration/dilations two weeks postoperatively and continued with this usually up to six months post-surgery. The arbitrary scheme used as a guideline when postoperatively calibrating/dilating the anus in children with anorectal malformations (ARM) [10] was used for the patients in this study. If the patient was unable to pass a certain dilator, the patient was defined to have developed a stricture of the anastomosis. The dilatation was then performed under anesthesia.

Data were collected from the patient records including demographics, age at diagnosis, gestational age, birth weight, associated congenital anomalies, need of colostomy prior to pull-through, length of removed aganglionic bowel, number of

stools passed at follow-up and bowel function. Constipation refers to bowel movements that are infrequent or hard to pass [11]. Chronic constipation was defined as the use of daily laxatives to achieve passage of daily stools. Postoperative complications as enterocolitis were also recorded. The authors define enterocolitis as a clinical presentation of diarrhoea abdominal distention and fever [12]. Frequent foul-smelling stools only were not considered to be mild enterocolitis.

The use of medicine to achieve normal bowel function and also if the patient had received an antegrade continence enema (ACE) [13] were noted. The author's registered soiling or faecal incontinence in those older than 3-4 years of age. However, that group is still so small that the data are not relevant.

A control group consisting of a consecutive group of children from the same geographical region as the study group, matched with regard to age and with no prior gastrointestinal problems, was gathered and data were collected concerning demographics and number of daily stools.

Statistical considerations:

In this case-control study we used a continuity corrected chi-squared statistic or Fisher's exact test and the Mann-Whitney U test two-tailed to evaluate the results.

Results

A total of 31 children were diagnosed and operated on using the TERPT technique. The demographic data are listed in Table 1. There were no disclosed intraoperative complications.

Of the 31 patients, a total of seven (23%) received a preoperative colostomy and this group was excluded in the postoperative follow-up. All seven had diverting stomas since the diagnosis HD was not set yet and the dilatation of the intestine was alarming; one of these seven children had total colon aganglionosis and received a preoperative ileostomy. The stool frequency in the seven patients that initially got a stoma did not differ from those operated directly with TERPT. There was no difference in outcome in patients who underwent preoperative colostomy versus no colostomy. Stämmer detta? Kan de då inte argumentera att de ska vara inkluderade?

Three patients were reoperated on due to complications and were also excluded (Figure 1). One patient suffered postoperatively from persistent symptoms of bowel obstruction, e.g. constipation, abdominal distension and abdominal pain and was diagnosed with twisted bowel and underwent reoperation. Two other patients developed postoperative fistualization between the bowel and the urinary tract, one with a recto-vesicoseminal fistula and the other between the rectum and the urinary bladder. They were treated operatively for their severe and unacceptable complications. It is unclear why they developed fistualizations and they were not known to have any perioperative problems.

The cohort of children is summarized in Figure 1, illustrating the drop out and then the included children with HD operated on with one stage TERPT. Two of the included children had Down's syndrome.

The median time for diagnosis was seven days, Table 1. Twenty-four (77 %) of the children were diagnosed within the first month of life. The median time to the TERPT procedure was 34 (range 10-261) days after diagnosis in the cases of one-stage pull-thorough without prior colostomy (Table 2).

The mean length of the intestinal resection and the median observation time are presented in Table 2.

Enterocolitis was noted once in three patients 15 - 29 weeks postoperatively and twice in one of the patients. The number of daily stools for these children at the last follow-up was 3 (1 – 5).

When compared with the control group, Table 3, there were no significant differences between the demographic data.

Functional results:

The number of stools passed daily post-TERPT correlated with the time elapsed since the operation and is demonstrated in Table 4 and Figure 2. There was a significant decrease in the frequency of daily stools over time (Figure 2). The six oldest children (49 – 63 months) passed one stool daily after four years which was then comparable to the control group. All of the six children mentioned had passed more than three stools daily three months earlier.

Eight children (38%) used some form of bowel regulating medication at their last follow up. Of these, three (10%) had an antegrade continence enema (ACE) [13] and two (9.5%) used daily oral medication.

Four (18%) of the children only used enemas if necessary and not daily. One of these four patients had suffered from two episodes of enterocolitis and the parents thus used rectal enemas immediately when necessary.

Discussion

Our findings show an initial high frequency of stools compared with non-afflicted children but that the frequency normalizes during the first four years. This is important for the preoperative counseling with the parents and for the medical planning. The strength of this study is its prospective nature, the fact that all children from a defined region are included and that all the TERPT operations were performed by the same group of pediatric surgeons using the same details of the operative method throughout the study period. The patients are also closely monitored at our center with frequent follow-ups after surgery. Thus our results show the true nature of the immediate postoperative period as well as the long-term results.

The intention when operating on a patient with Hirschsprung's disease is to ensure regular and spontaneous bowel motions without incontinence. Several studies have focused on the long-term follow-up [14, 15] after the pull-through operation, usually comparing different operating techniques. The focus has been on constipation and incontinence using different scoring systems [6, 14, 15]. The most common criticism of these studies is the lack of clear criteria for defining incontinence and constipation, that the studies rely on interviews with parents and patients or documentation of symptoms in patients' records with no clear-cut criteria of how the severity of constipation or soiling is graded or if these problems are chronic or not.

Zhang et al [3] concluded that the stooling function was fine in most of the patients after one stage pull-through operation for HD. They showed a very low rate of constipation and no cases of incontinence. The difference from the present study is that they only had three follow-up points, one and three months after surgery as last follow-up.

There have been no studies showing any statistically significant advantages of early versus late completion of surgery in respect to long-term stooling habits [6, 15, 16, 17] although it is generally thought that the earlier the child has the operation the better their future stool control will be; early sensation of passage of feces through the

anal canal should allow early re-establishment of some anorectal reflex. One study [3] shows that the younger the patients are at the time of surgery, the faster the stooling patterns recover. However, another study [17] found that neonates had a significantly longer stabilization period than that of the non-neonates. Most of our patients were operated on as neonates and, thus, our results do not reflect the outcome of an early versus later approach when operating on the patients with HD.

It is well known that patients operated on with TERPT may suffer in the immediate postoperative period from a very high frequency of stools, up to 30-40 a day. This causes much problem with painful perianal excoriations and the guardians have to change diapers frequently. The few studies that have focused on this issue have limited follow-ups [3, 7, 18, 19].

The results of this study show a high frequency of the number of daily stools passed during the first postoperative months. The number of daily stools has not reached an acceptable situation until four years after the TERPT operation. This information is of importance in patient counseling and when informing the children's guardians on the children's situation during the postoperative period. Kim et al [17] reports a postoperative stabilization period of 9.4 weeks to reach normal stooling patterns and normal findings in abdominal radiography. This is a significantly shorter recovery period than our findings. The difference may be attributed to the definition of normal stooling patterns and in Kim et al stools passed was not compared with a healthy control group. Van Leeuwen et al [19] report results similar to those from our study concerning lapse of time to normalization of stooling patterns with a trend towards a decreasing number of stools per day over the first 12 to 18 months.

The mean length of removed bowel in our study population was 17 cm and only one child had more than 25 cm removed during the one-stage pull through procedure. It is generally known that the longer the length of the removed aganglionic segment the more severe the stool disorder will be. Zhang et al [3] found that in patients with frequent stools and soiling there was a loss of the natural sigmoid loops on contrast enema and the pulled through colon had a straight course; this was particularly seen in aganglionic segments over 30 cm. Since the resection length in the present study did not vary much, we have not correlated it to the number of stools.

Constipation occurred in eight (38%) patients in the group but only five (22%) of these used daily bowel regulation medications. This number is lower than the number presented by other authors [16], which showed that the rate of

constipation was 28% after primary pull-through operation. On the other hand, it is higher than that of Zhang et al [3], which reported a constipation rate of 9%. Van Leeuwen et al [19], report a constipation rate of 22%, which is close to ours. As long as there is no clear consensus on how constipation is reported, the true number is hard to predict.

Enemas or oral laxatives were used by 22% of the children operated on with TERPT. The bowel management was considered to be a factor influencing the results. However, when excluding the children on bowel management with regular enemas, the results did not significantly change the conclusion reported here.

Loperamid, Imodium®, is sometimes used to reduce the number of stools in children with very frequent passage of stools [12]. None of the children in this study used or had been using this drug.

The children who had suffered from enterocolitis did not differ significantly in the number of daily stools at the last follow-up compared with the median number of daily stools for the other children with HD operated on with TERPT. Thus, it is not possible to explain their enterocolitis by obstipation or infrequent passage of stools. This is in line with a recent review [20] where the authors could not correlate the frequency of daily stools to the frequency of enterocolitis. The number of children with enterocolitis in our study is similar with that reported in a literature review [12, 20].

The weakness of our study is the small number of children included as well as the short duration of the observation time, which does not enable the discovery of all long-term problems. However, the duration of the observation was long enough to answer the question posed here. The number of children excluded from the study is high, Figure 1, mainly due to an initial treatment using a diversion through a stoma as the first operative intervention before the ano-rectal reconstruction. Secondly, the number of reoperations is higher than that reported in a recent literature report [21]. Thirdly, the study does not compare with other methods used for the operative reconstruction of children with HD. More studies on the outcome of the operative treatment comparing different methods are needed [22].

In conclusion, the results of this study show that the frequency of daily stools passed will be high for a long time during the postoperative phase after the TERPT procedure. The results are of importance for preoperative counseling and when

planning for the postoperative care of children with HD undergoing the TERPT procedure.

Legends to the figures

Figure 1

A figure illustrating the included children with Hirschsprung's disease (HD) operated on with Transanal endorectal one-stage pull-through (TERPT) and the reasons for drop-out. N = number.

Figure 2

The figure shows the number of stools passed daily post transanal endorectal one-stage pull-through (TERPT) for Hirschsprung's disease (HD) in 21 children. The findings are correlated with the duration of time in months/years from the TERPT operation as well as to a control group of healthy children. After 48 months the number of stools passed daily by the children with HD is no longer significantly different from that of the healthy control group of children. $P = 0.1674$. Statistical method: Mann-Whitney U test two-tailed. Diagram created by Excel® Microsoft Corporation, Redmond, WA, USA.

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Table 1

The demographic data of the children in the total study group (N=31)

Male	22 (71%)
Female	9 (29%)
Birth weight median (mean 3662) range	3745 g, 2420 - 4800
Age at diagnosis median	7 (1-1039 days)
Preoperative colostomy	7 (24%)

Table 2

Data regarding included patients only (N=21).

	Median (range)
Time from birth to TERPT, days	41 (15-1235)
Time from histopathological diagnosis to TERPT, days	34 (10-261)
Duration of observation, months	23 (<1-63)
Length of bowel removed, cm	17 (10 – 30)

Table 3

The demographic data of the children in the study group compared with those in the control group.

	Study group	Control group	P value
Number	21	54	
Median age, months (range) at the study	48 (2-72)	50 (14-92)	0,5515*
Female/male	9 (29%)/12 (71%)	15/39	1**

*Mann-Whitney U test two-tailed

**Fishers Exact probability Test, two tailed

Table 4

The number of stools passed every day in the study group compared with the control group.

Age group	Number of daily stools: Median (range)		P value*
	Study group	Control group	
total	5 (1 - 35)	1 (1 - 2)	<0,001
> 2 years	3 (1 - 20)	1 (1- 2)	<0.01
>3 years	3 (1- 20)	1 (1 - 2)	<0.01
>4 years	1 (1 – 6)	1 (1 - 2)	0,1674

***Mann-Whitney U test two-tailed**

Figure 1

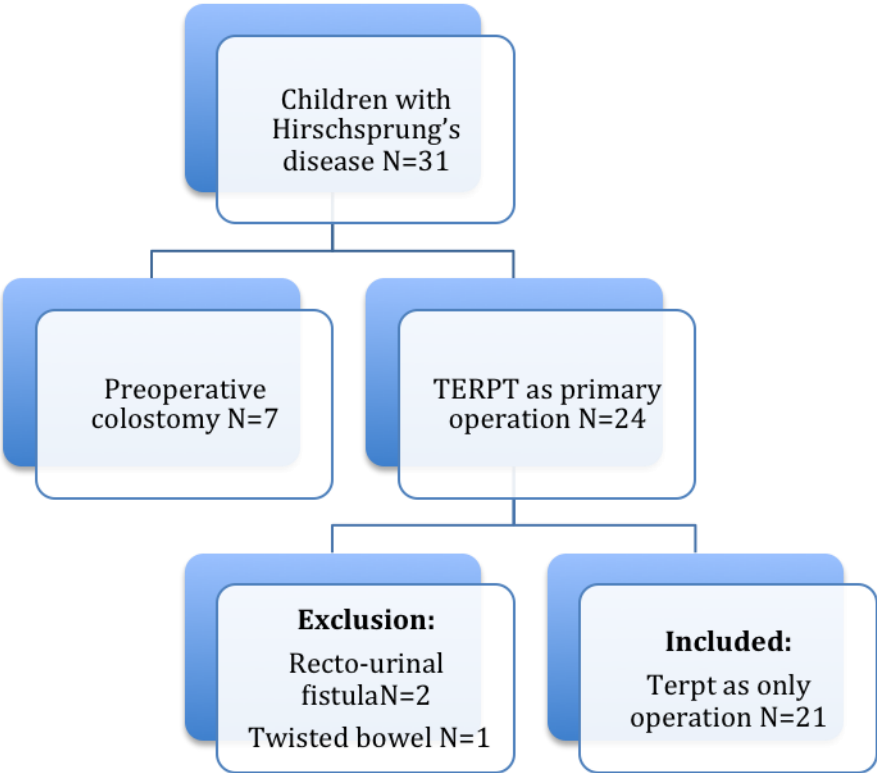


Figure 2

