

Quality of life and parents' satisfaction with Duhamel's versus transanal endorectal pull-through for the treatment of Hirschsprung's disease in children

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Background The aim of this study is to compare the surgical outcome as well as parents' satisfaction and quality of life for children after the transanal and the Duhamel pull-through operations in a single-center experience.

Patients and methods A retrospective cohort file review was carried out of all cases of Hirschsprung's disease treated surgically in our institution. Patients were classified into group 1 (transanal endorectal pull-through) and group 2 (Duhamel pull-through). Three questionnaires were designed. Demographic data, perioperative data, complications, and the length of follow-up data were analyzed statistically.

Results Six-nine patients were included. Medications were required postoperatively in 27% of the patients in group 1 versus 60.7% of patients in group 2. In group 1, 22.5% of parents were fairly satisfied and 69% of parents were satisfied, whereas in group 2, 31.8% of parents were poorly satisfied, 40.9% were fairly satisfied, and 27.3% were satisfied. Patients were older than 3 years; no patients had poor results in group 1 versus 33.3% in group 2.

Introduction

Hirschsprung's disease (HD) is caused by the failure of ganglion cells to migrate cephalocaudally through the neural crest during weeks 4–12 of gestation [1]. The disease occurs in one of 5000 births [2]. Traditionally, surgical therapy for HD has consisted of a proximal defunctioning colostomy, followed months later by a definitive reconstructive pull-through procedure in which the aganglionic colon is resected and the normally innervated bowel is brought down and sutured to the area just above the anal sphincter [3]. Over the past decade, the surgical management of HD has evolved. The previous gold standard three-staged procedure with a preliminary stoma was replaced by a two-staged procedure; one-stage pull-through is now advocated in many centers worldwide, with results as favorable as multistage procedures [4].

Swenson, Duhamel, and Soave (or endorectal pull-through) procedures were the most commonly performed operations for HD in North America until 1998 [3], when transanal endorectal pull-through was first described [5–7].

Although there are many publications comparing different surgical procedures for the treatment of HD, little is known about parents' satisfaction and the quality of life of children after different operations.

The aim of this study is to compare the surgical outcome as well as parents' satisfaction and quality of life for

Conclusion Our experience with transanal pull-through showed less incidence of postoperative enterocolitis, failure to thrive, redo surgery, and need for anticonstipation medications than that with Duhamel pull-through. Although the anorectal scoring system showed better results in transanal pull-through than Duhamel pull-through in all age groups, it was statistically significant in patients older than 3 years of age. There was a statistically significantly better parent satisfaction and quality of life in the transanal group than the Duhamel pull-through group. *Ann Pediatr Surg* 8:105–110 © 2012 Annals of Pediatric Surgery.

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Keywords: Hirschsprung's disease, parent satisfaction, quality of life and children

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children after the transanal and the Duhamel pull-through operations in a single-center experience.

Patients and methods

The preferred surgical procedure for HD was the two-stage Duhamel pull-through at the pediatric surgical department, Al Wasl Hospital (Dubai) until June 2002. The transanal endorectal pull-through has been performed in Al Wasl Hospital since that date to date.

A retrospective file review was carried out of all cases of HD treated surgically in our institution from June 1998 until June 2010.

Patients were classified into two groups according to the type of surgical procedure they underwent: group 1 (transanal endorectal pull-through) and group 2 (Duhamel pull-through).

Three questionnaires were used: one designed to evaluate bowel function for children 3 years of age or younger (*) and the other for children older than 3 years of age (**), the Wingspread scoring system [8] (3 years is the age at which the majority of children will have achieved day and night anal continence). The third questionnaire (***) measured the quality of life for patients as well as parents'/patients' satisfaction and for the surgical management and outcome. Patients as well as parents were interviewed or contacted by phone to answer the questionnaire by an independent witness (social worker).

(*) 3 Years of age or younger bowel function estimation questionnaire

Number	Questions	Answers	Point scores
1	Stool frequency 6 months postoperatively	Constipated (not passing every 3 days spontaneously/very hard stool)	1
		More than six times daily	1
		Normal frequency	0
		Always his anus is stained with stool	2
2	Stool consistency	Soft	0
		Fluid	1
		Hard	1
3	Frequency of perianal excoriation	Always present	3
		Once/2–4 weeks	2
		Less than once a month	1
		None	0
4	Use of oral laxatives	Very frequent (every 1–2 days)	3
		Frequent (every 3–7 days)	2
		Infrequent (every 8th day or more)	1
		None	0
5	Frequency of using enemas	Very frequent (every 1–2 days)	3
		Frequent (every 3–7 days)	2
		Infrequent (every 8th day or more)	1
		None	0
6	Frequency of abdominal distention + diarrhea + fever (enterocolitis) postoperatively	Always present	3
		Once/2–4 weeks	2
		Less than once a month	1
7	Bowel habits of the child	None	0
		Normal	0
		Acceptable	1
		Bad (constipated always or incontinent)	2

(**) The Wingspread scoring system for children older than 3 years of age

Number	Questions	Point scores
1	Totally continent or very occasionally stress-related staining of underclothes without constipation Toilet trained with no medication	Excellent/very good
2	Rarely soils, except during stressful exercise Constipation managed with medication	Good
3	Intermittent soiling, urge incontinence Frequent loose stools or constipation that requires enemas	Fair
4	Constant fecal soiling and smearing Constipation only response to enemas	Poor

(***) Parents' satisfaction/quality of life

Number	Questions	Answers	Point scores
1	Explanation about procedure and complications before surgery	Yes	2
		No	0
		Partially	1
2	Postoperative course of the child in the hospital	Very good	3
		Good	2
		Fair	1
		Bad	0
		Very good	3
3	Postoperative course of the child after discharge	Good	2
		Fair	1
		Bad	0
		Very good	3
		Good	2
4	Satisfaction with the result of the surgery	Fair	1
		Bad	0
		Yes	1
		No	0
5	Recommending this surgery to the child's friend if he had same disease	Very good	3
		Good	2
		Fair	1
		Bad	0
		Very good	3
6	Postoperative bowel habits	Good	2
		Fair	1
		Bad	0
		Very good	3

(continued)

(***) Parents' satisfaction/quality of life

Number	Questions	Answers	Point scores
7	Mental development	Very good	3
		Good	2
		Fair	1
		Bad	0
8	Physical development	Very good	3
		Good	2
		Fair	1
		Bad	0
9	Using bowel wash or laxatives	Regular	2
		Irregular	1
		None	0
		Yes	1
10	Perception that the management was carried out properly	No	0
		None	0
11	Absenteeism from school for HD-related problem	Infrequently	1
		Frequently	2
		None	0
12	Frequency of hospital visits for HD-related complaint	No more follow-up	0
		Once a month	1
		Once/3 month	2
		Once/6 month	3
		Once a year	4
13	Child's relationship with his peers	Very good	3
		Good	2
		Fair	1
		Bad	0

Demographic data, disease presentation, associated congenital anomalies, family history of HD, age at surgery, age at interview, type and stages of surgery, length of aganglionic segment, early postoperative complications (within 30 days), late postoperative complications, total number of enterocolitis attacks, stooling patterns (frequency of bowel motion and need for laxative and/or enemas), presence or absence of failure to thrive (according to growth charts that correlate height, weight, and age), need for redo surgery, length of follow-up were collected, tabulated, and analyzed statistically.

Results

Sixty-nine patients were included in the study, 41 in group 1 and 28 in group 2. Figure 1 shows the sex distribution in both groups.

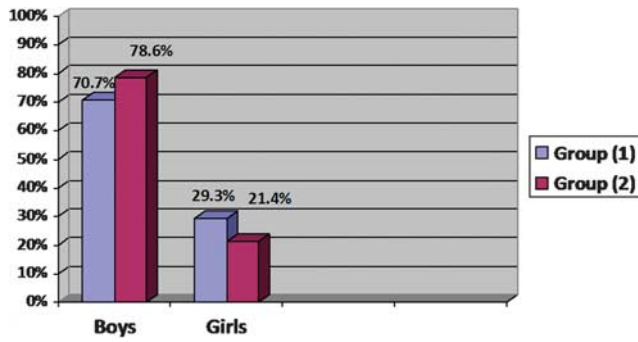
The median age at presentation was 20 days in group 1 and 16 days in group 2. The median age at surgery was 3 months in group 1 and 6 months in group 2. The median age at interview was 30 months in group 1 and 78 months in group 2.

The follow-up period ranged from 6 months to 4 years in group 1, mean 1.7 ± 1.1 years, and from 1 to 8 years, mean 3.6 ± 2.2 years, in group 2.

In group 1, 16.8% of patients had congenital anomalies in the form of neurological impairment (2.4%), cardiovascular, combined Down syndrome and heart anomalies, and Down syndrome (4.8% each).

In group 2, 17.9% of patients had congenital anomalies (10.7% of them had neurological impairment) (Fig. 2).

Fig. 1



Sex distribution in both groups.

In group 1, 9.8% of patients had a positive family history of HD, whereas in group 2, only 7.1% of patients had a family history of HD (Fig. 3).

The most common presentation in group 1 was delayed passage of meconium and chronic constipation (61 and 22%, respectively). In group 2, the most common presentation was delayed passage of meconium and enterocolitis with distension (53.6 and 21.4%, respectively) (Fig. 4).

The most common segment for HD was the rectosigmoid segment in groups 1 and 2 (95.1 and 89.3%, respectively) (Fig. 5).

Early postoperative complications

There was a significant difference between the two groups in the occurrence of early postoperative complications. In group 1, 34% of patients developed early postoperative complications in the form of excoriations and soiling (29.2 and 4.8%, respectively), whereas in group 2, only 10.7% of patients developed early complications in the form of excoriation ($\chi^2 = 4.92, P < 0.05$).

Late postoperative complications

There was no significant difference between the groups in the late postoperative complications ($\chi^2 = 0.337, P > 0.05$). In group 1, 53.7% of patients developed late complications, most commonly excoriations and stricture (27 and 9.8%, respectively), whereas in group 2, 60.7% of patients developed late complications, most commonly constipation and excoriation (32.1 and 14.3%, respectively) (Table 1).

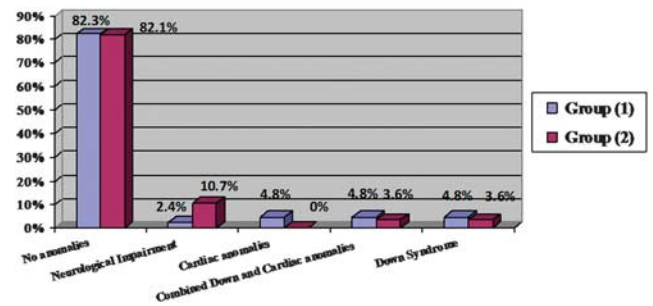
Preoperative enterocolitis

There was no significant difference between the groups in the occurrence of preoperative enterocolitis. Table 2 shows that 14.6 versus 14.3% of patients in groups 1 and 2, respectively, had preoperative enterocolitis.

Postoperative enterocolitis

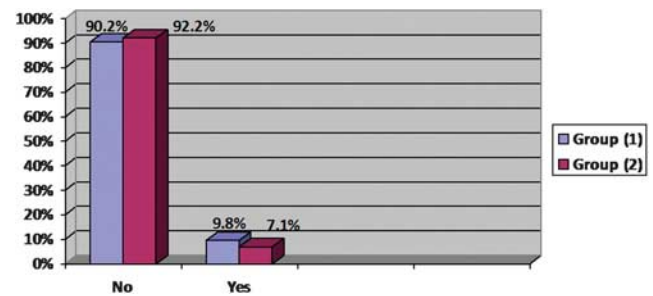
No patients in group 1 developed postoperative enterocolitis versus 32.1% of patients in group 2; there was a significant difference ($\chi^2 = 8.3, P < 0.05$) (Table 3).

Fig. 2



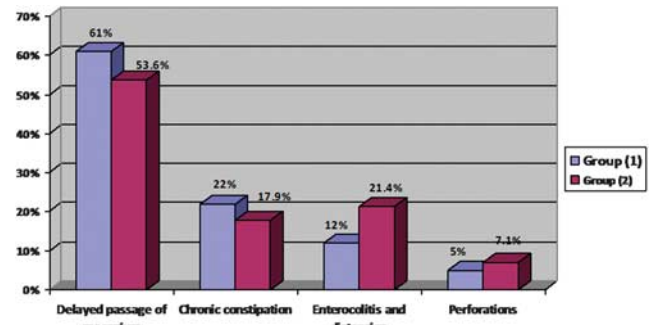
Associated congenital anomalies in both groups.

Fig. 3



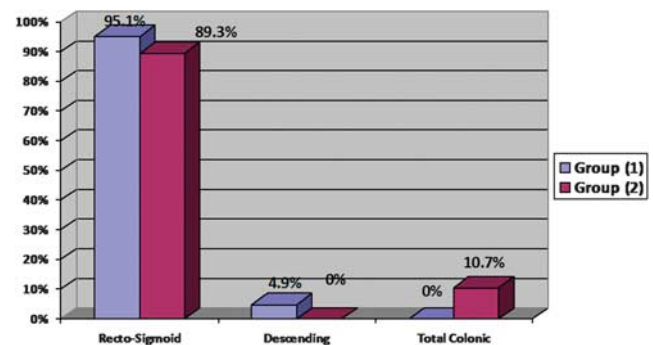
Family history of Hirschsprung's disease in both groups.

Fig. 4



Clinical presentations in both groups.

Fig. 5



Affected segment of Hirschsprung's disease in both groups.

Table 1 Late postoperative complications for both groups

Groups	Late complications							Total
	No	Excoriations	Constipation	Stricture	Soiling	Overflow incontinence	Adhesive obstructions	
Group 1	19 (46.3%)	11 (27%)	3 (7.3%)	4 (9.8%)	2 (4.8%)	2 (4.8%)	0	41 (100%)
Group 2	11 (39.3%)	4 (14.3%)	9 (32.1%)	0	1 (3.6%)	1 (3.6%)	2 (7.1%)	28 (100%)

Table 2 Frequency of preoperative enterocolitis in both groups

Groups	Preoperative enterocolitis			χ^2 P-value
	No	Yes	Total	
Group 1	35 (85.4%)	6 (14.6%)	41 (100%)	0.002 $P > 0.05$
Group 2	24 (85.7%)	4 (14.3%)	28 (100%)	

Table 3 Frequency of postoperative enterocolitis in both groups

Groups	Postoperative enterocolitis		χ^2 P-value
	No	Yes	
Group 1	41 (100%)	0	15.2 $P < 0.05$
Group 2	19 (67.9%)	9 (32.1%)	

Colostomy

All the patients in group 2 required colostomy, whereas only 14.6% of patients in group 1 required colostomy ($\chi^2 = 48.5$, $P < 0.05$).

Frequency of bowel motion

Analysis of the frequency of bowel motion at the time of the questionnaire interview showed no significance difference between the groups. In group 1, 53.8% of patients had bowel movements two to three times/day compared with 53.6% of patients in group 2 (Table 4).

Failure to thrive

4.8% of patients versus 28.6% of patients in groups 1 and 2, respectively, showed postoperative failure to thrive that was statistically significant ($\chi^2 = 7.5$, $P < 0.05$).

Redo operation

Only one patient in group 1 (2.4%) required a redo operation compared with five patients (17.9%) in group 2, with no statistically significant difference.

Anticonstipation medications

Table 5 shows that there was a significant difference in the use of anticonstipation medications between both the groups ($\chi^2 = 7.9$, $P < 0.05$).

Results of questionnaire interview

Thirty-six patients (88%) in group 1 versus 22 patients (78.5%) in group 2 could be reached for the questionnaire interview.

The anorectal function was rated using the Wingspread scoring system for patients older than 3 years of age.

In patients older than 3 years of age, there was a significant difference between the scores of both groups. In group 1, 20% of patients had an excellent function compared with 4.8% of patients in group 2 ($t = 2.8$, $P < 0.05$) (Table 6).

In patients 3 years of age or younger, in group 1, 65% of patients had good bowel function (score 0–6), 19% of patients had fair bowel function (score 7–12), and 16% of patients had poor bowel function (score 13–17).

In group 2, only one patient had fair function.

In terms of parents' satisfaction, there was a significant difference between the two groups. In group 1, 69% of the parents were satisfied compared with 27.3% in group 2 ($\chi^2 = 8.4$, $P < 0.05$) (Table 7).

Discussion

Postoperative results in the surgical management of HD appear to be satisfactory. Despite this good overall outcome reported, many studies have shown a higher than anticipated incidence of problems after surgery for HD [9].

Quality of life remains a difficult concept to assess and is influenced by the physical, psychological, spiritual, functional, and social well-being of an individual. The discrepancies in the quality of life reported following surgical correction of HD may be attributed to the study design, the details of the investigations carried out, or the lack of an objective independent observer [9].

Recently, transendorectal pullthrough (TERPT) has become the most popular procedure for the treatment of HD, but overstretching of the internal anal sphincter remains a critical issue, which may impact the long-term continence outcome. Because TERPT is a relatively new procedure, there is only one report, to our knowledge, that compares the long-term outcome of TERPT with the conventional transabdominal pull-through [10].

Our current study is the first (to our knowledge) to compare TERPT with Duhamel pull-through in terms of the rates of complication as well as quality of life and parents' satisfaction.

In our study, the most common length of aganglionic segments in both groups was rectosigmoid (95.1% of patients vs. 89.3% of patients in groups 1 and 2, respectively).

Although there were three cases of total colonic aganglionosis in group 2, early postoperative excoriation

Table 4 Frequency of bowel motion in both groups at the time of the last follow-up visit before the study

Groups	Bowel movement				Total
	4 times or more/day	2–3/day	Once/3 days	Once/4 days or more	
Group 1	15 (36.6%)	22 (53.8%)	2 (4.8%)	2 (4.8%)	41 (100%)
Group 2	3 (10.7%)	15 (53.6%)	3 (10.7%)	7 (25%)	28 (100%)

Table 5 The need for laxatives and/or enema in both groups

Groups	Medications		χ^2 P-value
	No	Yes	
Group 1	30 (73%)	11 (27%)	7.9 P<0.05
Group 2	11 (39.3%)	17 (60.7%)	

was higher in group 1 (29.2% of patients vs. 10.7% of patients in groups 1 and 2, respectively); overall, two cases had persistent soiling (a 2-year-old boy with down syndrome and a 5-year-old neurologically normal boy) in group 1. Langer *et al.* [3] have reported an incidence of 11% of early postoperative excoriation in his largest multicenter series for transanal pull-through.

Excoriation was still the most common late postoperative complications in group 1 (27%) followed by stricture (9.8%) and constipation (7.3%). The Langer *et al.* [3] series reported a 4% incidence of postoperative stricture. The four cases of strictures developed in our early experience with transanal pull-through, but after we adopted a protocol of postoperative dilatation for 3 months, Moreno cases of strictures were found.

In group 2, constipation was the most common late postoperative complication (32.1%), followed by excoriation (14.3%). El-Sawaf *et al.*'s [10] study also reported a higher incidence of constipation in the abdominal than the transanal pull-through group (38.1 vs. 20%, respectively). Moore *et al.* [9] reported constipation in 26% of patients versus 9% of patients who underwent Duhamel versus transabdominal Soave procedures, respectively.

There were two cases of adhesive bowel obstruction in group 2 that required surgical release. The incidences of incontinence and overflow incontinence were almost similar in both the groups.

Although there was almost the same percentage of patients who had preoperative enterocolitis in both the groups, there was a statistically significant difference in the incidence of postoperative enterocolitis (nil in group 1 vs. 32.1% in group 2). El-Sawaf *et al.* [10] reported an incidence of postoperative enterocolitis of 45 versus 61.9% in the transanal versus abdominal pull-through groups. Both Langer *et al.* [3] and Moore *et al.* [9] studies reported a 6% incidence of postoperative enterocolitis for transanal pull-through and abdominal pullthrough respectively.

Although we prescribed oral metronidazole for all patients in group 1 postoperatively for 1 month, it is still not clear whether this led to a decrease in the incidence of postoperative enterocolitis in the patients in group 1. Another possible explanation may be under-reporting of cases of enterocolitis in our hospital as parents seek medical advice in other hospitals.

In terms of bowel movement, 53.8 of patients in group 1 versus 53.6% of patients and in group 2, respectively, had two to three normal bowel movements per day at the time of the questionnaire interview. Langer *et al.* [3] reported that 80.5% of children had normal bowel function after undergoing transanal pull-through as reported by their parents or care givers. EL Sawaf *et al.* [10] study revealed similar continence outcomes in transanal and abdominal pullthrough although the postoperative period was longer in transanal group.

In our study, there was a higher incidence of constipation in group 2 (25%) than in group 1 (4.8%); however, 36.6% of patients in group 1 versus 10.7% of patients in group 2 had bowel movements four times or more per day at the time of the questionnaire interview.

There were statistically significant differences in the incidence of failure to thrive between groups 1 and 2, respectively (4.8 vs. 28.6%) during the follow-up period in our study. Thirty-nine percent of the patients in Moore *et al.*'s [9] study were over 50th percentile in terms of weight for age (WA); most of the patients whose WA was below the 3rd percentile were in the younger age group. Moore *et al.* [9] concluded that normal WA regained with time after surgical correction of HD.

One patient (2.4%), a 6-year-old female in group 1, required redo surgery for persistent anastomotic stricture; in group 2, five patients (17.9%) required redo surgery in the form of division of persistent pouch spur. This difference between both groups was statistically significant. A total of six patients (14.6%) required redo surgery in the El-Sawaf *et al.* [10] study (five for abdominal pull-through and one for transanal pull-through); their results indicated significantly poorer long-term outcome in the redo group. Follow-up of our redo cases indicated that only two patients in group 2 still had persistent constipation, although they had no residual spur and no residual aganglionic segment in the pull-through segment.

In terms of the need for laxatives and/or enemas, 27% of patients in group 1 versus 60.7% of patients in group 2 still require medications for the management of their constipation; the results were statistically significant.

Table 6 Bowel functions in children older than 3 years of age in both groups using the Wingspread scoring system

Groups	Bowel function >3 years				Number of patients >3 years of age interviewed	<i>t</i> <i>P</i> -value
	Poor	Fair	Good	Very good/excellent		
Group 1	0	2 (40%)	2 (40%)	1 (20%)	5	2.8 <i>P</i> <0.05
Group 2	9 (42.9%)	10 (47.6%)	1 (4.8%)	1 (4.8%)	21	

Table 7 Parents' satisfaction and quality of life in both groups

Groups	Parents' satisfaction			χ^2 <i>P</i> -value
	Unsatisfied	Fairly satisfied	Satisfied	
Group 1	3 (8.5%)	8 (22.5%)	25 (69%)	8.4 <i>P</i> <0.05
Group 2	7 (31.8%)	9 (40.9%)	8 (27.3%)	

The Langer *et al.* [3] study reported that 9.3% of patients still require medications to manage their constipation. The El-Sawaf *et al.* [10] study showed that 19.5% of patients still require medications to manage their constipation; Moore *et al.* [9] reported that 13% of patients of the study groups still require medications to manage their constipation.

Quality of life and patient/parent satisfaction discussion

In patients older than 3 years of age, there was a statistically significant better bowel function in group 1 than in group 2 using the Wingspread scoring system. Although the same results were obtained in patients 3 years of age or younger, it was not statistically significant.

The total stooling score in the El-Sawaf *et al.* [10] study was higher in the transanal pull-through group (12.75 ± 8.07) in comparison with that of the abdominal pull-through group (11.28 ± 7.75), although the results were not statistically significant.

Analysis of the degree of parent's satisfaction/quality of life in our study indicated that there was statistically significant more parent satisfaction in group 1 versus group 2. The Bai *et al.* [11] study of quality of life after the Swenson procedure showed that 40% of patients had good quality of life, 46.7% had fair quality of life, and 13.3% had poor quality of life. In the Bai *et al.* [11] study, there was a strong association between fecal continence and the quality of life in patients.

Heij *et al.* [12] used a questionnaire on anorectal function and quality of life after Duhamel pull-through surgery; they concluded that the majority of patients have impaired anorectal function after Duhamel's operation and that there was no indication that this impairment improved with time. In the Moore *et al.* [9] study, the

majority of patients (94%) appeared to be well-adjusted members of the society.

Conclusion

Our experience with transanal pull-through showed lower incidence of postoperative enterocolitis, failure to thrive, redo surgery, and need for anticonstipation medications than that with Duhamel pull-through.

Although the anorectal scoring system showed better results in transanal pull-through than Duhamel pull-through in all age groups, it was statistically significant in patients older than 3 years of age.

There was a statistically significant better parent satisfaction and quality of life in the transanal group than the Duhamel pull-through group.

Acknowledgements

Conflicts of interest

There are no conflicts of interest.

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