

Surgery for Hirschsprung's disease: comparison between the Duhamel method and the transanal endorectal pull-through based on 59 patients and a review of the literature.

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Background: Hirschsprung's disease (HSCR) is a congenital malformation of the enteric nervous system due to the arrest of migration of neural crest cells to form the myenteric and submucosal plexuses. It leads to an aganglionic intestinal segment, which is permanently contracted causing intestinal obstruction. Its incidence is approximately 1/5000 birth, and males are more frequently affected with a male/female ratio of 4/1. The diagnosis is in most cases made within the first year of life. The rectal biopsy of the mucosa and sub-mucosa is the diagnostic *gold standard*.

Purpose: The aim of this study was to compare two surgical approaches for HSCR, the Duhamel technique and the transanal endorectal pull-through (TEPT) in term of indications, duration of surgery, duration of hospital stay, postoperative treatment, complications, frequency of enterocolitis and functional outcomes.

Methods: Fifty-nine patients were treated for HSCR by one of the two methods in our department of pediatric surgery between 1994 and 2010. These patients were separated into two groups (I: Duhamel, II: TEPT), which were compared on the basis of medical records. Statistics were made to compare the two groups (ANOVA test). The first group includes 43 patients and the second 16 patients. It is noteworthy that twenty-four patients (about 41% of all patients) were referred from abroad (Western Africa). Continence was evaluated with the Krickenbeck's score.

Results: Statistically, this study showed that operation duration, hospital stay, postoperative fasting and duration of postoperative antibiotics were significantly shorter (p value < 0.05) in group II (TEPT). But age at operation and length of aganglionic segment showed no significant difference between the two groups. The continence follow-up showed generally good results (Krickenbeck's scores 1; 2.1; 3.1) in both groups with a slight tendency to constipation in group I and soiling in group II.

Conclusion: We found two indications for the Duhamel method that are being referred from a country without careful postoperative surveillance and/or having a previous colostomy. Even if the Duhamel technique tends to be replaced by the TEPT, it remains the best operative approach for some selected patients. TEPT has also proved some advantages but must be followed carefully because, among other points, of the postoperative dilatations. Our postoperative standards, like digital rectal examination and anal dilatations seem to reduce the occurrence of complications like rectal spur and anal/anastomosis stenosis, respectively in the Duhamel method and the TEPT technique.

Since the first surgery for HSCR performed by Swenson in 1948 [1], technical improvements have been made in this specific operative treatment. Nowadays, three surgical methods are known to offer satisfactory postoperative outcomes: the Swenson, the Duhamel and the Soave modified by De La Torre in 1998 [2] into a transanal operation.

This study focused on the Duhamel technique and the Transanal Endorectal Pull-Through (TEPT).

In our department, Duhamel technique was the exclusive technique used until 2004 when TEPT was introduced.

The aim of this study was to find if there were any differences between the two operations after a comparison based on the following criteria:

- The indications and contraindications to the operation: patient's age and weight, patient's origin, length of the affected intestinal segment, previous colostomy
- Duration of surgery
- Postoperative treatments: duration of postoperative fasting, antibiotic therapy, duration of hospital stay, other specific therapy to one or the other technique
- Postoperative complications: ileus, postoperative enterocolitis, adhesive intestinal obstruction, intestinal residual spur, anal/anastomosis stenosis

- Outcomes regarding continence
This helped us to bring out the indications or contraindications to one of these operations.

Materials and methods

For this study, we selected all patients who were diagnosed with HSCR and treated surgically by one of the authors (BJM) at our University Hospital between 1994 and 2010, with one of the two different techniques, which are used in our centre, namely the Duhamel procedure and the TEPT.

Patients were divided into two groups:

Group I (Duhamel), includes forty-three patients (72.9% of all patients) operated between 1994 and 2008.

Group II (TEPT), includes sixteen patients (27.1% of all patients) operated between 2004 and 2010.

For all the patients of this study the Ethics Commission and the Medical Direction of the University Hospital of Lausanne approved the access to their medical records, operative protocols, anesthesia protocols and pathology reports.

The medical records were used to determine the age and weight at operation, the comorbidities, the length of hospital stay, the duration of postoperative antibiotic therapy, the presence of postoperative Hirschsprung associated enterocolitis (HAEC), the existence of a preoperative colostomy and the evolution of the stool continence.

The operative protocols and the pathology reports were used to determine the length of the aganglionic bowel segment. Anesthesia protocols were used to establish the duration of operation.

Duhamel

The Duhamel procedure (retrorectal transanal pull-through) was described in 1956. It consists in the resection of the aganglionic bowel segment to the peritoneal reflection distally (through a laparotomy or a laparoscopy), according to fresh frozen sections. An incision through the whole posterior rectal wall is made about 1 cm proximal to the dentate line. The healthy bowel is pulled through this incision behind the rectum and anastomosed side to side

with a staple device to the remaining aganglionic segment.

The standard postoperative care in our institution for this procedure includes a 10-day antibiotic therapy (in most cases amoxiciline/clavulanic acid, metronidazole and amikacin), nasogastric tube until bowel movements resume, digital rectal examination at hospital discharge and barium enema one month after surgery.

TEPT

The TEPT method is performed according to De La Torre description in 1998 [2]. TEPT is a transanal modified Soave approach (endorectal pull-through) that aims to avoid laparotomy or laparoscopy. A mucosectomy is performed starting about 1cm proximally to the dentate line. This leaves a muscle cuff until the peritoneal reflection is reached. At this point, the muscle cuff and the peritoneal reflection are incised, and the entire colon is mobilized up to the transition zone. After fresh frozen sections to prove the presence of ganglionic cells, the aganglionic bowel is resected and an anastomosis is made between the ganglionic bowel and the anal canal. The standard postoperative care for this procedure includes anal dilatations with Hegar dilators starting about two weeks after surgery. Dilators sizes are used according to patient's age (see in Table 1) [3] to avoid stenosis of anastomosis and/or smooth muscle sleeve, and antibiotic therapy for 3-5 days with amoxiciline/clavulanic acid, metronidazole and amikacin. No postoperative fasting is needed.

To define the Hirschsprung associated enterocolitis (which has no clear definition) we used the definition proposed by Coran and al. [4] and Elhalaby and al. [5] according to clinical and radiological criteria: the presence of one or more clinical criteria among abdominal distension, explosive diarrhea, vomiting, fever, lethargy, rectal bleeding and colonic perforation, radiological criteria are: colonic dilatation (90% sensitivity but 24% specificity), intestinal cut-off sign (74% sensitivity and 86% specificity).

To summarize, enterocolitis associated with HSCR is defined by abdominal distension and explosive diarrhea with intestinal cut-off sign ("appears when a gaseous intestinal dilatation is

abruptly cut off at the pelvic brim” [6]).
 Krickenbeck’s score (initially developed for the ARMs) was used for the follow-up of stool continence. The score is shown in Table 1.
 Concerning the statistics, for each criterion we used the mean value \pm the standard deviation, and that for each group. Then, to compare the two different groups (I Duhamel, II TEPT), we used the analysis of variance (ANOVA) to determine whether the results were statistically significant ($P < .05$) or not ($P > .05$).

One particularity of this study was that 24 patients (about 41% of all patients) were referred from abroad (Western Africa) leading to difficulties in assessing long-term results of continence.

Table 2: Krickenbeck’s score

1. Voluntary bowel movements	Yes / No
2. Soiling	Yes / No
2.1	Occasionally (one or twice per week)
2.2	Every day, no social problem
2.3	Constant social problem
3. Constipation	Yes / No
3.1	Manageable by changes in diet
3.2	Requires laxative
3.3	Resistant to laxatives and diet

Results

The mean follow-up time in group I was 63.8 ± 44.4 months (mean \pm SD) and in group II: 39.3 ± 20.7 months (mean \pm SD).

Indications

Two indications for the Duhamel method were first the origin of patient and secondly the presence of a colostomy. Patients who were referred from abroad represent 51% of patients in group I and 12% in group II.
 All patients with a colostomy were treated by the

Duhamel method.

With these two observations, we concluded that with patients referred from abroad and/or in the presence of a colostomy, the operation of choice is the Duhamel.

There was no significant difference between the two groups neither in what concerns the age and weight at operation or the length of aganglionic segment, so they can’t be considered as indications for one of both operative techniques.

Comorbidities

Among our 59 patients, two were affected with Down syndrome (3.4%), which is in accordance with the recent literature [7]. Those two patients were treated with the Duhamel method.

We also noted cardiac abnormalities in 3 patients (5.1% of all patients) that were patent foramen ovale and atrial and/or ventricular septal defects. Note that one patient with patent foramen ovale and ventricular septal defect was also affected by Down syndrome.

The cri-du-chat syndrome was reported in one patient.

No other significant comorbidities were present in our other patients.

It is noteworthy that two patients in our study had a positive family history (3.4%) illustrating the genetic predisposition of HSCR [8].

Complications

Complications after surgery are summarized in table 3.

Postoperative enterocolitis was found in 20.9% of patients in group I and 12.5% of patients in group II. This showed no statistically significant difference between the two groups.

We reported no postoperative adhesive intestinal obstructions in group I. One patient in group II (6.3%) had three episodes of postoperative adhesive intestinal obstructions: respectively 2 and 3 years postoperatively.

Rectal spur was present in two patients in group I (4.6%) who had to be re-operated.

One patient with anal stricture in group II (6.3%) required long-term dilatation. The standard postoperative dilatation hadn’t been performed

carefully in this patient.

Colostomy

Colostomy was present in 23 patients (53.5%) in group I (respectively 0% in group II). These colostomies had been performed first for bowel obstruction and secondly for HAEC. Thirteen patients (56.5%) had colostomy for bowel obstruction and two after ceecal perforation (8.7%). Indications for colostomy were not known for all patients. Fifteen patients (65.2%) with colostomy were referred from abroad.

Four patients of group I died postoperatively, what represents 6.8% of all patients and 9.3% patients of group I. These four patients had been referred from abroad. Three of them died probably because of enterocolitis, and the cause is unknown for the last one. We report no death in group II.

Statistical analysis

The age at operation was 32.2 ± 41.2 months in group I, and 15.8 ± 29.2 months in group II ($p > .05$).

The weight at operation was 11.7 ± 7.6 kg in group I and 8.2 ± 6.4 kg in group II ($p > .05$).

The length of aganglionic bowel segment, according to the pathological exam, was 16.4 ± 13.0 cm in group I and 19.4 ± 20.5 cm in group II ($p > .05$).

These first three points of comparison between group I and II showed no statistically significant difference.

The operating times were, in minutes, 329.7 ± 71.8 in group I and 238 ± 139.8 in group II ($p < .05$).

Postoperative antibiotic therapy was 11.9 ± 6.4 days in group I and 7.4 ± 3.9 days in group II ($p < .05$).

Fasting times in days were, 3.3 ± 1.3 and 0.6 ± 0.9 respectively in group I and II ($p < .05$).

Hospital stay was 13.2 ± 2.1 days for group I and 9.6 ± 4.7 days for group II ($p < .05$).

Those four next comparison points showed statistically significant differences between the two groups ($p < .05$).

This shows that operation duration, postoperative antibiotics use, postoperative fasting time and hospital stay is significantly

shorter in the TEPT group.

All statistics are resumed in table 4.

Continence outcomes

The continence follow-up with the Krickenbeck's score showed generally good results in both groups (scores 1; 2.1; 3.1), with a tendency to constipation (score 3.2 and 3.3) in group I and slightly more soiling (2.2 and 2.3) in group II. Figure 1 and 2 (respectively Duhamel and TEPT) summarize the overall continence follow-up according to time after surgery.

To compare the two groups more precisely, we assessed the continence 36 months after surgery (or the first time after 36 months).

Twenty-eight patients in group I and nine patients in group II were still followed at 36 months or after. To simplify the comparison, we added the scores 2.1 (soiling occasionally) and 3.1 (constipation manageable by changes in diet) to the normal continence (score 1) admitting that those two score were tolerable socially and manageable conservatively.

In group I, 60.7% of patients had a normal continence, 3.6% had soiling and 35.7% were constipated. The continence was assessed at 55 ± 20.1 months for these 28 patients.

In group II, 55.5% of patients had a normal continence, 22.2% had soiling and 22.2% were constipated with a continence assessed at 41.3 ± 7.9 months.

Both groups could be compared because there was no significant difference in patients' ages between them at continence assessment.

Discussion

Indications

The first indication for the Duhamel procedure was the origin of the patients. This operation involves no heavy postoperative care. That is why, together with the fact that most of the patients referred from abroad had a colostomy (65.2%), 92% of them had a Duhamel operation. Especially anal dilatations, which are important for good postoperative outcomes after TEPT to reduce the rate of anal stricture.

Our second indication for the Duhamel method was the presence of a preoperative colostomy. Indeed, if a colostomy had already been performed, the idea of a minimal invasive

surgery (TEPT) had less sense than another open surgery (Duhamel).

We thought that one of the indications for TEPT would be the length of aganglionic bowel segment, which we expected to be significantly shorter than in group I. Elhalaby and al. described that an extension of aganglionic segment further than the sigmoid colon is an essential limitation for TEPT [9]. Our results showed that it was not the case and that the aganglionic bowel segment was even longer in group II. Even excluding one patient with total aganglionic colon segment plus 17 cm of ileum (a total of 78 cm of bowel resected) that, after discussion, should have been operated by the Duhamel method, the results remain statistically non-significant ($p > .05$).

The advantages of TEPT according to De La Torre and al. are, the absence of laparotomy or laparoscopy, shorter surgery, less abdominal contamination and adhesions and a better aesthetic outcome [10]. But some disadvantages were also brought out, such as possible injury to the anal muscular sphincters [4] and slightly inferior long-term continence results [11].

Two of our clear contraindications for the Duhamel and the TEPT methods in our centre, are a patient younger than 3 months and/or under 4.5 kg. The aim is to avoid injury to muscular sphincters.

Statistical analysis

The follow-up time was significantly different between our two groups. This is explained by the fact that Duhamel method was used in our centre long before TEPT was introduced.

In spite that Duhamel was introduced long before TEPT, the difference of the follow-up times was not important due to the fact that some patients referred from abroad and thus operated according to Duhamel were lost to follow-up.

TEPT showed clear advantages over the Duhamel method, especially concerning the hospital stay, which was significantly shorter. This can be explained by a shorter postoperative fasting period together with a shorter postoperative antibiotic therapy.

Our results are in accordance with those of

Langer and al. who found that with TEPT the duration of hospital stay was significantly shorter [12] and the results of Hadidi [13], which say that TEPT was shorter to perform and had shorter hospital stay and earlier recovery than other approaches.

Complications

A study of Minford and al. showed an enterocolitis rate of 3% in the Duhamel group and 35% in the Transanal Coloanal Anastomosis (TECA) group [14]. Compared with our results (respectively 20.9% and 12.5%), we have higher rate of enterocolitis in the Duhamel group and lower in the TEPT group. An other comparison can be made with a study of Elhalaby and al. that showed a rate of 11% postoperative enterocolitis in the Duhamel group [5] and a study of Kim and al. [15] who reported 35% of postoperative enterocolitis in the TERP (Transanal Endorectal Pull-Through) group. The different definitions of enterocolitis can be at the origin of these differences.

Rectal spur was found in two patients (4.6%) in group I, which is lower than what Minford and al. reported (18% of rectal spur) in their Duhamel group [14]. This may be due to the control digital rectal examination we performed at the hospital discharge in order to diagnose and treat the spur as soon as possible.

The anastomosis stenosis is also an important postoperative complication. Stricture was present in only one of our patients in group II (6.3%). We had a lower stricture rate in our patients than Mindford and al. did (19% of stricture, in his "TECA group") [14] and Kim and al. (9% of anastomosis stenosis in TERP group) [15]. This might be explained by the standard postoperative anal dilatation performed in all patients with TEPT.

The death of four of our patients (all from abroad) might be related to the lack of careful postoperative surveillance in their home country.

The TEPT method tends to replace the Duhamel technique with the time because of its minimal invasive approach as shown in figure 3. But there is a lack of evidence particularly concerning the long-term continence outcomes.

Our study shows a slight tendency of soiling in TEPT patients. But the comparison between the two groups remains difficult on this point because of the unequal follow-up time.

In order to improve the evaluation of continence follow-up, a specific score for HSCR could be used, for example the continence score used by N. L. Yanchar and al. [16], and a standardized way for the follow-up could also be used to assess the continence, for example every 3 months.

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Acknowledgements

Roulet G.

Surgery for Hirschsprung's disease:

Table 1 : Postoperative dilatations: Hegar dilator size

Patient's age	Hegar dilators size to be reached
1-4 months	12
4-8 months	13
8-12 months	14
1-3 years	15
3-12 years	16
>12 years	17

Table 2: Krickenbeck's score (already in article)

Table 3: Complications

Complications	Duhamel	TEPT	P Value
Postoperative enterocolitis	9 (20.9%)	2 (12.5%)	0.469587
Postoperative adhesive intestinal obstructions	0 (0%)	1 (6.3%)	0.101535
Rectal spur	2 (4.6%)	0 (0%)	0.390107
Anastomosis stenosis	0 (0%)	1 (6.3%)	0.101535

Table 4: Statistical analyses

	Duhamel	TEPT	P value
Number of patients	43 (72.88%)	16 (27.12%)	
Male	33 (76.75%)	8 (50%)	
Female	10 (23.25%)	8 (50%)	
Origin:			
Switzerland	20 (48.8%)	14 (87.5%)	0.004056
TdH	23 (51.2%)	2 (12.5%)	0.004056
Follow-up time (months) (mean ± SD)	63.8 ± 44.4	39.3 ± 20.7	0.045388
Age at operation (month) (mean ± SD)	32.15 ± 41.23	15.81 ± 29.18	0.120106

Operative weight (kg) (mean \pm SD)	11.7 \pm 7.6	8.2 \pm 6.4	0.088770
Length of aganglionic segment (cm) (mean \pm SD)	16.35 \pm 13	19.4 \pm 20.5	0.409891
Preoperative colostomy	23 (53.5%)	0 (0%)	<.0001
Operation duration (minutes) (mean \pm SD)	329.7 \pm 71.8	238 \pm 139.8	0.009196
Postoperative fasting (days) (mean \pm SD)	3.3 \pm 1.3	0.6 \pm 0.9	<.0001
Postoperative antibiotics (days) (mean \pm SD)	11.9 \pm 6.4	7.4 \pm 3.9	0.020324
Hospital stay (days) (mean \pm SD)	13.2 \pm 2.1	9.6 \pm 4.7	0.000125

Figure 1: Duhamel continence follow-up according to months after surgery

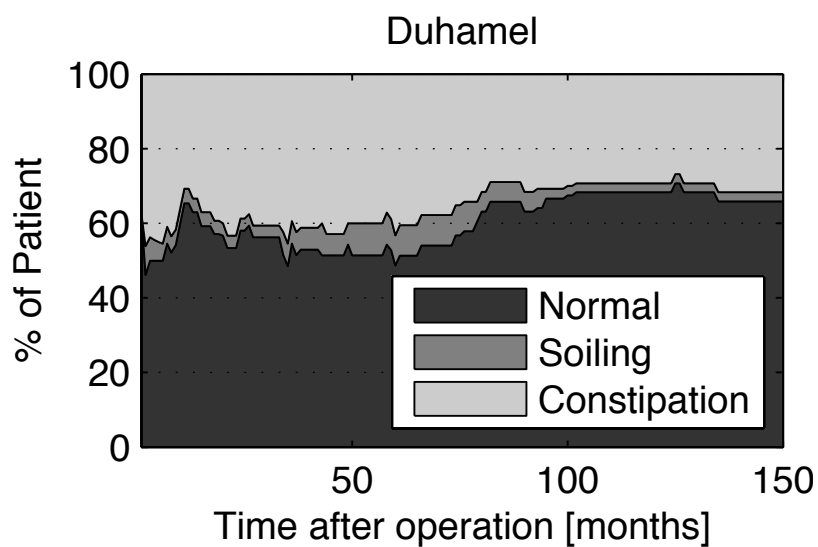


Figure 2: TEPT continence follow-up according to months after surgery

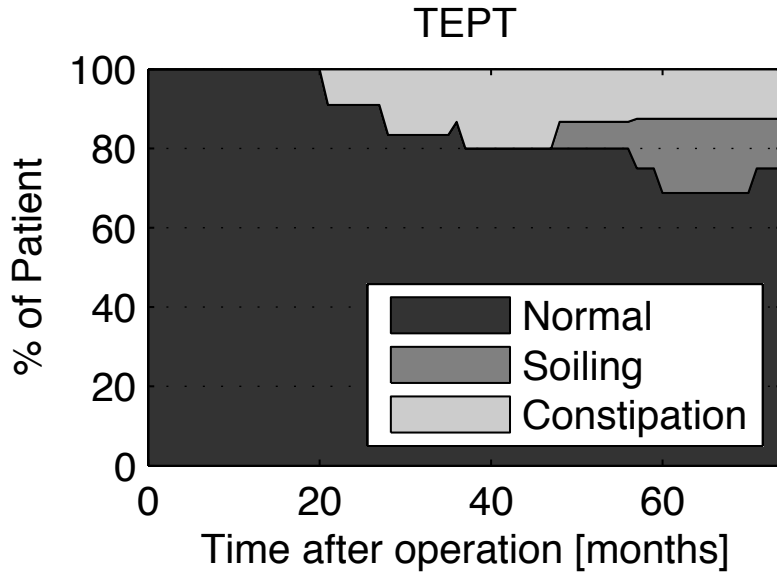


Figure 3: Changes in the use of Duhamel and TEPT over time

