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Transanal pull-through for Hirschsprung disease in neonates: A single-center experience

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KEYWORDS

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Hirschsprung disease,
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Outcome

ABSTRACT

Background: Transanal pull-through for the treatment of Hirschsprung disease in the neonatal period remains debatable. This study reported our experience with transanal pull-through in neonates.

Methods: We reviewed medical records of neonates with Hirschsprung disease treated in our department with transanal pull-through between the years 2010 and 2016.

Results: Of the 40 included patients, 31 were male neonates, and 9 were female. The mean age at the time of surgery was 13 days. The mean operative time was 136 minutes. The level of aganglionosis was rectal in 8 cases, rectosigmoid in 19 cases, descending colon in 7 cases, splenic flexure in 3 cases, transverse colon in 2 cases, and ascending colon in one case. A combined abdominal approach was used in 11 patients. There were no intraoperative complications. Major complications were noted in 4 cases: anastomotic leakage in 3 cases of whom one succumbed 3 days postoperatively; and bowel perforation in one case. The most common late postoperative complication was fecal soiling encountered in 25% of cases. Two among 29 patients who reached the age of continence remain incontinent.

Conclusion: Transanal pull-through in neonates has similar outcomes and complications to those of infants and children. Therefore this procedure can be safely employed in neonates however we recommend that the treating team should have an ample learning curve for this procedure in infants.

INTRODUCTION

Hirschsprung disease (HD) is a disorder in the development of the enteric nervous system characterized by the absence of ganglion cells in the myenteric and submucosal plexuses of the distal intestine causing functional intestinal obstruction; one to two cases are reported for every 10000 live births. [1] The surgical treatment consists of the removal of the aganglionic bowel and the bringing of the normally innervated intestine to the anus. The age at surgery and the appropriate approach remain controversial. [2-5]

In our department, during the last decade, we used to treat patients with Hirschsprung disease during the neonatal period by transanal pull-through (TAPT) using the Soave procedure with or without an assisted abdominal approach. The objective of this study was to report our experience with TAPT during the neonatal period and to assess its feasibility and safety compared with a previously published study conducted in our department of patients treated by TAPT after the neonatal period.

METHODS

We conducted a retrospective study on neonates with Hirschsprung disease who underwent TAPT between 2010 and 2016 in the Department of Pediatric Surgery of Fattouma Bourguiba Hospital in Monastir Tunisia, with IRB approval.

Inclusion Criteria:

1. Newborns who received surgical treatment for Hirschsprung disease through the Transanal Pull-through Procedure (TAPT)
2. Patients who underwent surgery before reaching 28 days of age

Exclusion Criteria:

1. Sick or septic neonates, not feasible for primary procedures

2. Cases with insufficient data available in the hospital records
3. Patients with a follow-up period of less than three years

Preoperative management:

The initial management included a nasogastric aspiration and intravenous fluids. Intravenous antibiotherapy was administered in case of enterocolitis or positive C-reactive protein. An early decompression of the colon was obtained using rectal stimulation and irrigation through a rectal tube. A water-soluble contrast enema was performed and the preoperative diagnosis was based on clinical and radiological features.

The approach was decided by the level of the transitional zone on the contrast enema. When the radiologic transitional zone was at the rectosigmoid or the sigmoid colon, the patient was considered eligible for single-stage transanal pull-through while laparoscopic-assisted TAPT was reserved for patients with a transition zone proximal to the sigmoid colon. The final surgery could be done after the newborn was stabilized.

Surgical technique:

The surgery is performed under general anesthesia. When an abdominal approach is indicated, a laparoscopy is performed to identify the transitional zone, perform biopsies, and mobilize the colon using the Georgeson technique for the pull-through procedure. [6]

For the transanal procedure, a caudal block is performed at the beginning to minimize the need for anesthetic agents and to provide postoperative analgesia. Patients are placed in the lithotomy position and a urinary catheter is placed in the urinary bladder. All patients receive a third-generation cephalosporin intravenously. Anus retraction is performed using 8 stay sutures. Submucosal injection of saline solution is used to ease the mucosal dissection. The endorectal mucosectomy starts at 0.5-1 cm above the dentate line and the mucosa is stripped from the underlying muscle distance until we reach the peritoneal reflection. At this level, the muscularis is circumferentially incised to enable bringing down the sigmoid colon through the anus. Frozen section biopsies are performed at the time of the procedure to detect the ganglionosis level. The colon is divided a variable distance above the most distal normal biopsy (minimum 2cm) to prevent any potential transition zone pull-through. The colo-anal anastomosis is done above the dentate line with 4/0 absorbable interrupted sutures.

Postoperative management:

The urinary catheter is removed on the 1st postoperative day. Oral feeding is allowed 24-48 hours postoperatively. The patients are discharged once stooling has started and feeds are well tolerated. Two weeks after surgery, a rectal examination is performed using the little finger to assess the anastomosis. Routine postoperative anal dilatations with Hegar dilators are performed for cases of anastomotic strictures- starting with no. 6 and increasing progressively. Follow-up evaluations are planned weekly during the first month, monthly during the following 3 months, and every 3 months later.

Data collection:

Data were collected on perinatal history, demography, clinical presentation, investigations performed, operative management, postoperative assessment and complications, and outcome variables including stooling habits, length of stay, and occurrence of anastomotic leakage or stricture, incontinence, and enterocolitis

RESULTS

Clinico-demographic characteristics

We included 40 newborns- 36 were term and 4 were preterm newborns; 31 (77,5%) were male and nine were female (22,5%) (M:F 3.4:1); and the average body weight was 3300g (2450-4400g). The average age at presentation was 2 days (0-13 days) and the average age at admission was 7 days (1-27 days). Symptoms were abdominal distension in 100% of cases, emesis in 80% of cases, and delayed meconium passage (more than 48 hours) in 65% of cases; meconium was passed within 48 hours in 20% of the patients. Seven patients had preoperative enterocolitis and required intravenous antibiotics.

Four patients had Down syndrome and 4 other patients had other anomalies (including 2 cardiac anomalies: atrial septal defect in one case and ventricular septal defect in the other case).

A contrast enema was performed in all patients and showed an evident transition zone in 37 cases: 92,5% (rectal transition zone in 7 cases, rectosigmoid in 22 cases, and proximal to sigmoid in 8 cases); for the three other cases, we performed a rectal biopsy to confirm the diagnosis. The average age at the time of surgery was 13 days (4-28 days).

Surgical details and postoperative complications

We performed a primary laparoscopic-assisted pull-through in 9 patients with an extended form of Hirschsprung disease. In these patients, 2 needed conversion to an open approach due to technical difficulty. Primary TAPT was performed in 31 cases

including two patients who needed laparoscopic assistance for colon mobilization. The mean operative time was 136 minutes (70-240 minutes) including the waiting time for frozen biopsy results. The level of aganglionosis was rectal in 8 cases, rectosigmoid in 19 cases, descending colon in 7 cases, splenic flexure in 3 cases, transverse colon in 2 cases, and ascending colon in one case. The intraoperative course remained event-free and none of the patients required a blood transfusion. Bowel starts moving within 48 hours of surgery in 85% of the patients. Oral feeds were initiated within 2 days in 77% of the patients. The average hospital stay was 4.7 days (2-17 days).

Complications

Four patients had major postoperative complications consisting of anastomotic leakage in 3 cases: one patient died 3 days after the surgery due to peritonitis and two patients were re-operated 3 days later and bowel perforation in 1 case re-operated with good evolution (Table 1).

Table 1: Data of patients with major postoperative complications (Please fill the missing information)

Patient	Age/sex/ weight	Level of aganglionosis	Associated anomalies	Complications	Management	outcome
1 (term) operated on 27th DOL	4 days/ male/ 3.2kg	Rectosigmoid	Nil	1. Presented with enterocolitis 2. Lengthy operation (180 min. due to difficult dissection) 3. Anastomotic leakage		Died 3 days postoperatively
2 (term) operated on 4th DOL	2 days/ male/ 3.4kg	Rectal	Down syndrome atrial septal defect	1. Anastomotic leakage	Re-operated: colostomy	Good evolution
3(term) Operated on 15th DOL	17days/male/ 2.7kg	Transverse colon	Nil	1. Anastomotic leakage	Re-operated: ileostomy	Good evolution
4(term) Operated on 6th DOL	4 days/ male/ 2.9kg	Descending colon	Nil	1. Bowel perforation secondary to laparoscopic biopsy	Re-operated: colostomy	Good evolution

Perianal excoriation (22,5%), wound infection (10%), and postoperative enterocolitis (10%) were the most common complications (Table 2)

Table 2: Postoperative complications and its management

Complication	Number (%)	Management
Anastomotic leakage	3(7,5)	Reintervention
Perineal excoriation	9(22,5%)	Local care
Wound infection	4(10%)	Local care and antibiotherapy
Early intestinal obstruction	1(2,5%)	Surgery
Enterocolitis	4(10%)	Antibiotherapy
Anastomotic stricture	2(5%)	Stricturoplasty
Soiling	10	25
Constipation	3	13,3
Anastomotic stenosis	3	7,5
Incontinence	2	5

Fecal soiling between stools was the most common complication reported in 10 cases (25%), it was partially improved by enemas. Among 29 patients who reached the age of continence, two patients remain incontinent, very good improvement was reported for one patient through physiotherapy

DISCUSSION

Hirschsprung disease traditionally dealt with multistage surgery including the formation of proximal diverting colostomy at the time of diagnosis and later pull-through procedure. it was believed that operating on an older child made the pull-through technically easier. [7] Over the past two decades, it has been increasingly recognized that the use of routine colostomy is not always necessary. A large number of pediatric surgeons preferred the single-stage procedure at an early age. [8] The primary TAPT for HD has the advantage of avoiding the colostomy and its related complications including its social stigma. The aim of our study was to evaluate the feasibility, safety, and outcomes of TAPT during the neonatal period.

In most of the cases (37/40 cases), the water-soluble contrast enema was sufficient to confirm and

characterize the HD. A rectal biopsy was performed for three patients due to diagnostic uncertainty. We operated on HD in the neonatal period without prior rectal biopsy. The final histopathology confirmed the diagnosis in all cases: this allowed us to accelerate the surgical management when the characteristic features of HD were evident on contrast enema.

We also found that contrast enema helped us in identifying the need for laparoscopic assistance in certain patients with an extended type of HD based on the identification of the transition zone: when the transition zone is situated between the rectum and the sigmoid, it is very likely that the pull-through can be performed completely by a transanal approach. In our study, we needed a combined abdominal approach in 11 patients (27.5%) who required a laparoscopic mobilization. This rate seems to be high in comparison to other studies. [9,10] The correlation between the radiographic transition zone and the level of aganglionosis in pathology was 87.5% and was comparable to other studies. [11]

The average operative time in our series was 136 minutes. This is significantly shorter than the average operative time in another study conducted in our department in patients of over 2 years age, who underwent a single-stage Soave endorectal pull-through (240 minutes). [12] There was no significant intraoperative bleeding. This can be explained by the easier mucosal dissection during the neonatal period due to the presence of less adherent mucosa (no repeated episodes of enterocolitis), a less dilated colon above the aganglionic segment, that the mesentery and the colon are less substantial and easier to mobilize and that the blood vessels are easier to control. [4,8,13]

The advances in anesthesia and perioperative care made TAPT for newborns safe and feasible. We had no anesthetic complications in our patients.

The most important post-operative complication in our study was the anastomotic leakage reported in three patients (7.5%). This complication is more frequent for patients treated with TAPT during the neonatal period, according to a meta-analysis conducted by Westfal et al. [14] which can be explained by increased susceptibility of newborns to infections. [15] This complication was neither reported in the study conducted by Bhatia et al. [8] nor by Hassan et al. [4] and reported in only one patient by Ali. [16]

Post-operative enterocolitis is considered one of the main complications of patients with HD before and after definitive treatment. The reported incidence rate in the literature is about 20%. [17,18] The risk factor for post-operative enterocolitis is the history of preoperative Hirschsprung-associated enterocolitis. [17,18] It is believed that routine anorectal dilatations are an effective way to prevent the occurrence of anal stricture and to decrease the rate and severity of post-operative enterocolitis. [4] In our series, we had a significantly lower rate of post-operative enterocolitis (10%) than reported in the literature. Few other studies also reported a lower frequency of this complication. [5,8,9,16]

Although variable complication rates have been reported in the literature, we found many similarities in early complication rates with other studies on TAPT in newborns. [16,19] Teilebelum et al. found a higher rate of early complications in primary pull-throughs than in staged pull-throughs for newborns. [19] Skarsgard et al. found no difference in complication rates between patients who underwent a pull-through during the neonatal period and later. [20]

Stooling patterns have been poorly characterized in most of the series due to the short follow-up times reported in many of these studies. Among our 39 patients who survived, constipation was noted in three patients and improved with dietary management and laxatives. Soiling was present in 10 cases partially improved with enemas. Among 29 patients who completed three years of follow-up, 27 cases were continent. In comparison with the study conducted in our department for TAPT in newborn patients and patients above 2 years old, we found no statistically significant difference in the occurrence of complications between the two groups (Table 3).

Table 3: comparison of the occurrence of complications between patients who underwent TAPT in the neonatal period and those above 2 years in our department

Complication	TAPT in newborns	TAPT >2 years	p
Death	1/40	0/20	1
Anastomotic leakage	3/40	1/20	1
Perianal excoriation	9/40	5/20	1
Enterocolitis	4/40	1/20	0,65
Anastomotic stenosis	3/40	4/20	0,2
Soiling	10/40	4/20	0,75
Constipation	3/40	2/20	1
Incontinence	2/29	0/20	0,5

Our results are similar to those reported in the literature on TAPT for newborns (4,19) and better than other reports which find a high rate of defecation disorders (22% to 35%). (21)

A few studies have been conducted to assess the outcomes of TAPT for newborns; Most of them were limited to early outcomes due to the short follow-up period (8,16) or limited to a small cohort. Our study has several strengths; it is one of the very few studies in the literature with an acceptable cohort with a follow-up period of more than three years. There are

some limitations to our study: it is a retrospective study and a control group is not included.

CONCLUSION

TAPT during the neonatal period is a feasible procedure that offers the benefits of easier surgery than for older children. There were no statistically significant differences concerning post-operative outcomes but a randomized prospective study is suggested for getting a better level of evidence.

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