



Surgical maneuvers for long-segment Hirschsprung pull-through in unique patients

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Abstract

Purpose Patients with Hirschsprung disease affecting the splenic flexure or more proximal segments present a surgical challenge. Mobilizing the transverse colon to the pelvis during a pull-through may obstruct the distal ileum, or the length may be insufficient to reach the lower pelvis. This retrospective study aimed to describe two surgical techniques that facilitate mobilization of the transverse colon and their outcome.

Methods We included patients operated on between April 2017 and April 2024 and analyzed sex, comorbidities, type of pull-through, age at pull-through, history of previous surgeries, cause of the proximal transverse colon pull-through, technique used (Deloyers or Turnbull), complications, postoperative outcome and follow-up. The first technique used was the maneuver described by Turnbull. This operation creates a mesenteric defect and mobilizes the colon into this mesenteric window at the distal ileum level. The second technique was described by Deloyers and involves a 180-degree rotation of the right colon by dissecting the right colon attachment and the hepatocolic ligament. The cecum and the ileocecal valve are placed in the right upper quadrant, and the distal colon is mobilized into the pelvis.

Results We included 13 patients, 12 boys and 1 girl. Eight patients had previous surgeries in another hospital: five had an initial transverse colostomy, and three had an ileostomy. The remaining five had the initial operation in our hospital: two had an ileostomy, two had a colostomy, and one had a primary pull-through. The median age at pull-through was 16 months (4–59 months). We used the Turnbull technique in four patients whose aganglionosis was limited to the middle transverse colon. The Deloyers technique was used in the remaining patients, with ganglion cells in the proximal transverse colon. We left a protective ileostomy in five patients. The median follow-up was 4.5 years (3 months to 10 years). The stoma takedown is pending in one patient.

Conclusion The Turnbull and Deloyers techniques were helpful in patients with aganglionosis affecting the transverse colon.

Keywords Long-segment · Hirschsprung · Aganglionosis · Colonic rotation

Introduction

Mobilizing and pulling through the proximal transverse or right colon may be challenging in unique patients with long-segment Hirschsprung disease. If not performed correctly,

the pull-through can cause tension or compromise the vascular supply. Consequently, anastomosis dehiscence and stenosis are the most frequent complications. Mobilizing the colon in front of the distal ileum also can cause an intestinal obstruction (Fig. 1).

Deloyers in 1958 [1] and Turnbull in 1978 [2] described techniques to mobilize the proximal transverse colon and achieve a colorectal anastomosis proximal to the peritoneal reflection in adult patients. However, in patients with Hirschsprung disease, the colon must be mobilized even more distally than Deloyers and Turnbull referred because the colon must cross the pelvis and reach the vicinity of the anal canal.

Our study aimed to show the experience of patients with Hirschsprung disease who required a pull-through of the

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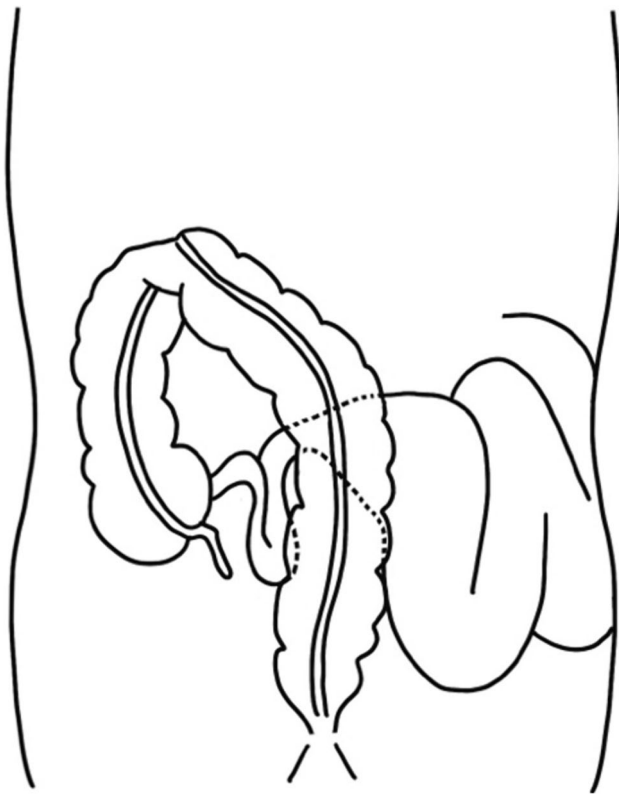


Fig.1 Pull-through with a conventional mobilization of the colon in front of the distal ileum. The pull-through is causing intestinal obstruction

proximal transverse colon in which the Deloyers and Turnbull techniques were necessary.

Materials and methods

This was a retrospective and descriptive study in which we reviewed our databases and included patients with Hirschsprung disease who were operated on from April 2017 to April 2024 and required the Deloyers or Turnbull technique.

The following variables were recorded and analyzed: sex, comorbidities, type of pull-through, age at pull-through, history of previous surgeries, cause of proximal transverse colon pull-through, technique used (Deloyers or Turnbull), complications, postoperative outcomes, and follow-up.

Results

Thirteen patients, 12 males and 1 female, with Hirschsprung disease underwent the Deloyers or Turnbull technique. One patient had trisomy 21. In all patients, the pull-through was performed using the Swenson technique. The median age

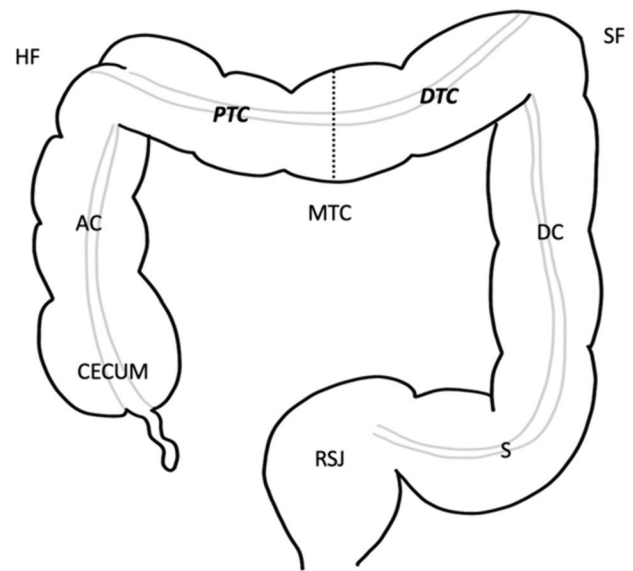


Fig.2 Segments of the large bowel. AC ascending colon, HF hepatic flexure, PTC proximal transverse colon, MTC middle transverse colon, DTC distal transverse colon, SF splenic flexure, DC descending colon, S sigmoid, R rectum

of the pull-through was 16 months (4–59 months). Eight patients had prior surgeries before being referred. Five had a transverse colostomy, and three had an ileostomy. Apart from ileostomy, two patients had a history of a complicated pull-through with a significant long stenosis of the colon.

Five patients were referred for suspected Hirschsprung disease and had no history of previous surgeries. Hirschsprung disease was confirmed by rectal biopsy. Initially, they were treated with rectal irrigations and showed partial improvement. The contrast enema showed a transition zone proximal to the splenic flexure. A primary pull-through was performed in one patient, a colostomy was created in two, and an ileostomy was performed in the other two.

The cause of the pull-through of the proximal transverse colon in five patients was the section of the marginal artery or Drummond's artery. This section occurred when a transverse colostomy was created. Extensive ischemic stenosis was noticed in two patients with a history of primary pull-through and aganglionosis proximal to the middle transverse colon in six patients.

The Turnbull technique was used in four patients. In these patients, the middle transverse colon was used for the pull-through. The Deloyers technique was used in nine patients where the proximal transverse colon, a shorter segment, was used for the pull-through (Fig. 2). After the pull-through in these patients, an ileostomy was created in five patients. Ileostomy closure was performed months later in four patients. One patient is awaiting ileostomy closure.

One patient had an intra-abdominal abscess resolved with medical management. This patient had a long stenosis after

the pull-through and was referred for redo surgery. The redo pull-through involved a complicated pelvic dissection.

The current median age of patients is 5 years (2–12 years). The median follow-up time was 4.5 years (3 months–10 years). Eleven patients had fecal control. The patient with Down syndrome remained clean through the use of a daily enema. One patient is still awaiting an ileostomy closure. Table 1 summarizes the results.

Discussion

Conventionally, mobilizing the transverse colon in front of the small bowel [3] to perform the pull-through is possible in most patients with Hirschsprung disease. However, in particular circumstances, it is necessary to use the proximal transverse colon or even more proximal large bowel. In these cases, the surgeon must use conventional mobilization of an intestinal segment with a length that does not comfortably reach the vicinity of the anal canal. In this way, the pull-through is at risk of tension or compression of the vascular pedicle, which can cause ischemia or necrosis of the colon. In our experience, this alternative method also obstructs the distal ileum (Fig. 1). Sileri et al. [4] also reported this complication.

Six of our patients had aganglionosis proximal to the middle transverse colon, which is viewed as an “unpreventable cause” that requires performing a pull-through with a short colon via the Deloyers procedure. In contrast, ligating the marginal artery while creating a transverse colostomy interrupts the communication between the superior and inferior mesenteric arteries. Consequently, mobilization of the distal transverse colon is not feasible. This is a “preventable cause”. This situation affected five patients in this cohort, so the proximal transverse colon had to be used for the pull-through, and a significant length of large bowel with ganglion cells was lost.

An ischemic pull-through caused a long stenosis in two patients. This other “preventable cause” led to use of a short proximal transverse colon and special maneuvers to achieve the pull-through. Ensuring a pull-through with no tension and adequate blood supply is essential to avoid a long stenosis.

The Deloyers or Turnbull procedure was performed in a specific segment of the colon. The Turnbull procedure was suitable when the middle transverse colon was used for the pull-through. This technique did not allow the pull-through of a more proximal colon. In patients with a proximal transverse colon segment (Fig. 2), we recommend the Deloyers technique.

When the colon proximal to the hepatic flexure (for example, the cecum and a few centimeters of the ascending colon) is pulled through, patients could have multiple evacuations

Table 1 Demographics, surgical characteristics and outcome in 13 patients with unique long-segment Hirschsprung

Patient	1	2	3	4	5	6	7	8	9	10	11	12	13
Sex	M	M	M	F	M	M	M	M	M	M	M	M	M
Age at pull-through (months)	34	28	23	14	23	59	6	8	4	16	6	42	13
Colon used for the pull-through	PTC	MTC	MTC	MTC	PTC	PTC	PTC	PTC	PTC	PTC	MTC	PTC	PTC
Type of mobilization	Deloyers	Turnbull	Turnbull	Turnbull	Deloyers	Deloyers	Deloyers	Deloyers	Deloyers	Deloyers	Turnbull	Deloyers	Deloyers
Bowel control	yes	no*	yes	yes	ileostomy	yes	yes	yes	yes	yes	yes	yes	yes
Follow-up (months)	54	82	31	55	3	5	6	102	61	122	106	7	52

M male, F female, PTC proximal transverse colon, MTC middle transverse colon. *Patient with trisomy 21

or functional postoperative obstructive symptoms. This short right segment, in our experience, has insufficient motility to produce regular bowel movements. In these cases, our preference is to perform a pull-through with the ileum.

Deloyers and Turnbull described their techniques for colorectal problems in adults. Therefore, these techniques could not be familiar for pediatric surgeons in Hirschsprung disease to resolve the pull-through in unique patients. Three books dedicated to pediatric surgery [5–7] mention colon rotation without giving more details about how to perform it or bibliographic references. In addition, this colon rotation is managed by use of techniques to correct total colon aganglionosis, such as left colon patches, which can confuse readers of those books, and in Hirschsprung disease, confuse readers of publications about use of long-segment techniques [8].

Turnbull procedure

Dr. Rupert Turnbull created a mesenteric window in the distal ileum and mobilized the colon through this window. This maneuver enables resolving the pull-through of the transverse colon (Fig. 3). This maneuver was described in 1978 in adult patients [2]. However, there are previous publications by Toupet [9] in 1961 and Hays in 1976 [10].

One problem that could arise before this maneuver is the creation of an internal hernia through the mesenteric defect. None of the revised articles report this complication [2, 4,

11–13]. Fixation of the transverse colon to the mesentery to avoid this has been described [2, 4, 14]. Other series do not mention this fixation [11, 12]. We have not read any literature reports of the use of this maneuver in the pediatric population.

Deloyers procedure

Dr. Lucien Deloyers described the rotation of the ascending colon preserving the ileocolic artery as an axis, which allowed the anastomosis without tension from the ascending colon to the proximal rectum above the peritoneal reflection (Figs. 4a and b) [1]. In children with Hirschsprung disease, the colon must pass through the pelvis and an anastomosis proximal to the anal canal must be performed (Fig. 4c). Therefore, the colon runs longer, passing through the entire pelvic length. Unlike the procedure in adults, the procedure becomes more complex than the original technique in patients with Hirschsprung disease. Other types of rotations, not applicable to Hirschsprung disease, have been described by different authors [15, 16].

The anatomy behind this maneuver leaves the cecum and ileocecal valve under the liver, and the colon has a single vascular pedicle from the ileocolic artery. The torsion or compression of the vascular pedicle in the pelvis can compromise the pull-through. The right colon was fixed to the right gutter to reduce this risk in patients. This fixation also

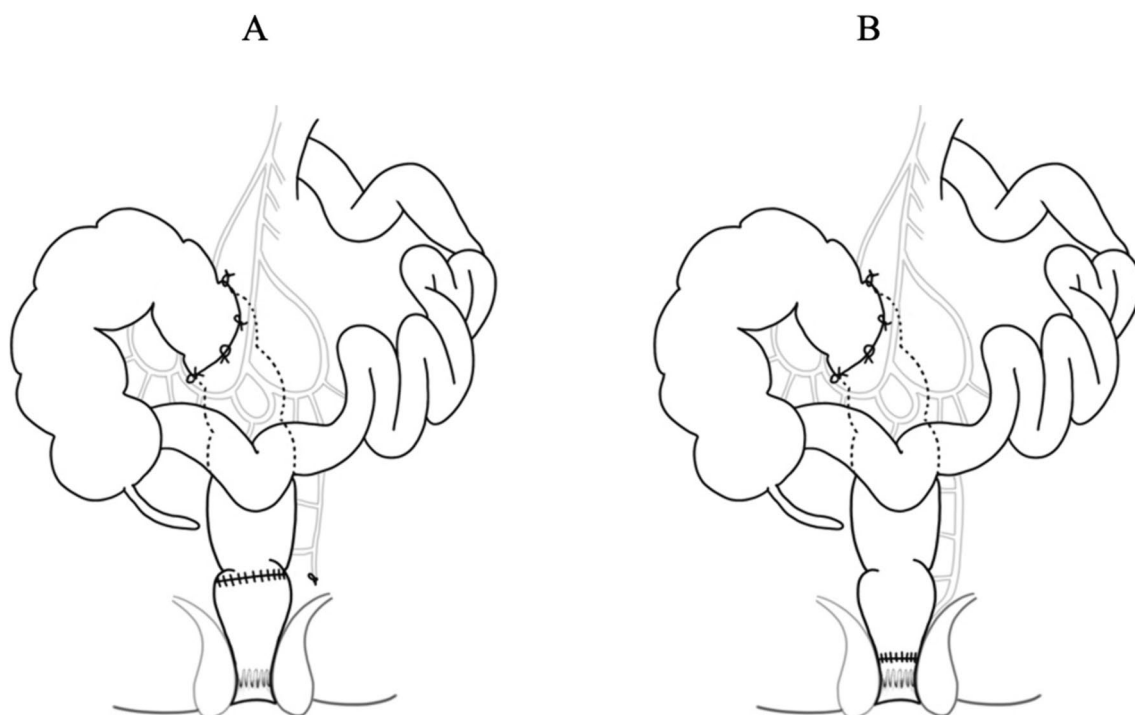


Fig.3 Turnbull procedure. **A** The large bowel passes through the mesenteric defect and the anastomosis is above of the peritoneal reflection in adults. **B** same maneuver in patients with Hirschsprung with a colorectal anastomosis above the pectinate line

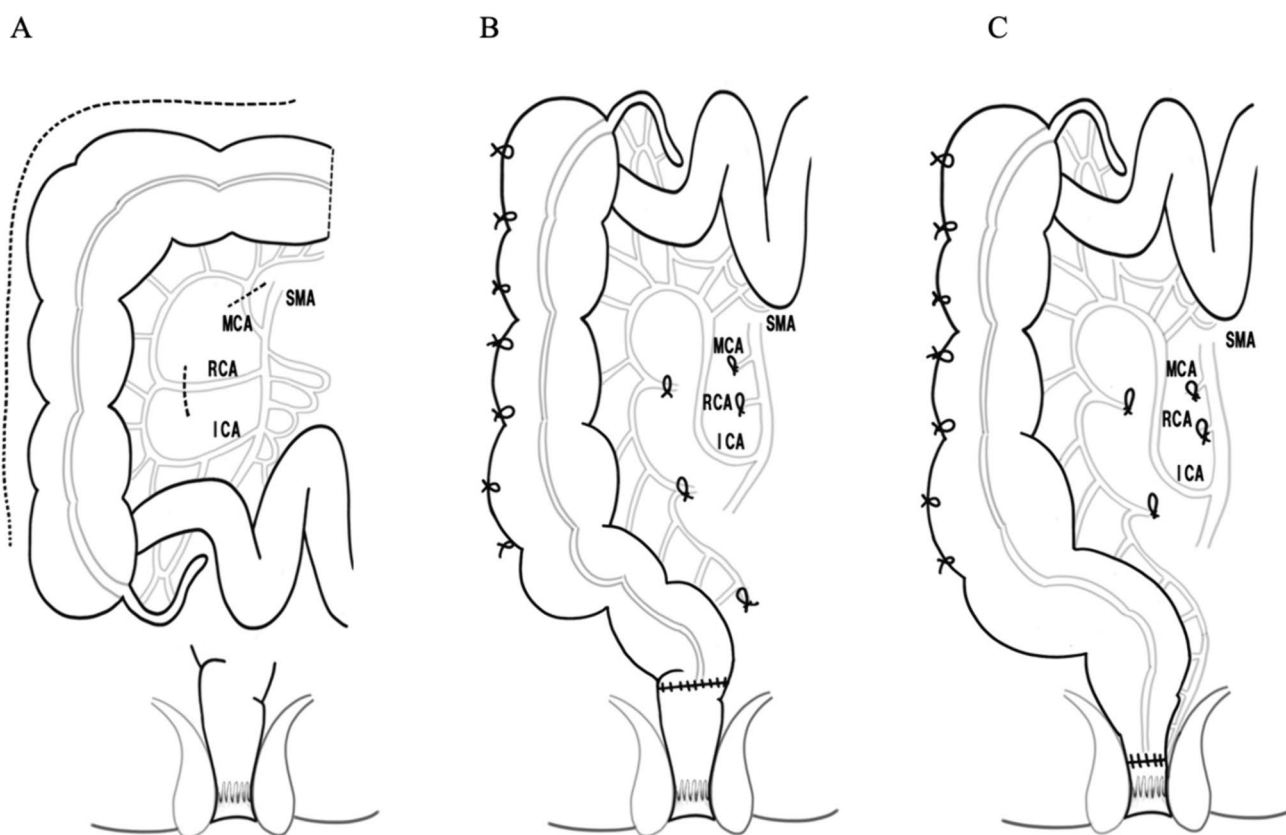


Fig. 4 Deloyers procedure. **A** The position of the right colon before the section of the MCA and the RCA. The dotted line indicates the area to be dissected to mobilize the colon. **B** The position of the colon after the rotation. In adult patients, the anastomosis is proximal to the peritoneal reflection. **C** Procedure used in patients with

Hirschsprung's disease. The colon is mobilized inside the pelvis, and the anastomosis is above the pectinate line. *SMA* superior mesenteric artery, *MCA* middle colic artery, *RCA* right colic artery, *ICA* ileocolic artery

has been described by Kontovounisios [17]. We did not find any report of colon necrosis that used this maneuver [1, 18–27]. Only Mulsow [28] reported 80 adult patients, of whom two had postoperative cecum necrosis, presumably due to irrigation of the pedicle.

The results of studies in adults suggest performing a prophylactic appendectomy during the surgical procedure to avoid the complexity of the diagnosis of acute appendicitis. However, in pediatric colorectal surgery, the appendix must be preserved because at a later age, it could be helpful in the creation of a conduit for administering antegrade enemas.

Conclusion

The Deloyers and Turnbull techniques proved very useful in resolving the pull-through in unique patients with Hirschsprung disease in whom the colon segment to be pulled-through was proximal to the middle transverse colon.

Pediatric surgeons need to become more familiar with these techniques to treat patients with Hirschsprung

disease. Particularly, these techniques are useful when the surgeon faces the need to perform a pull-through of the proximal transverse colon or even more proximal segments and the conventional mobilization does not allow reaching the anal canal or could cause an ischemic or high-tension pull-through. Preserving the marginal artery is essential when creating a colostomy in patients with Hirschsprung disease. The Turnbull technique is more reproducible, but it was insufficient in all cases. In our cohort, it was enough to mobilize the middle transverse colon, but not enough for more proximal segments. The Deloyers technique appears to be more complex but allows the proximal transverse colon to be mobilized to reach the anal canal.

A limitation of this study was its retrospective nature and the small number of patients.

Author contributions Study conception and design: MZ and AR Data acquisition: MZ, LM and AR Analysis and data interpretation: MZ, LM and AR Drafting of the manuscript: MZ, LM and LDT Critical revision: MZ, LM, AR and LDT

Data Availability No datasets were generated or analysed during the current study.

Declarations

Competing interests The authors declare no competing interests.

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