

# Redo pull-through for postoperative complications following pull-through in Hirschsprung disease: a single center experience

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**Purpose:** Although surgical management of Hirschsprung disease (HD) is effective in most patients, some patients experience long-term postoperative complications, and require redo pull-through (PT). The present study evaluated clinical outcomes of redo PT in HD patients at a single center.

**Methods:** Patients with HD who underwent redo PT procedures between 2003 and 2019 were retrospectively reviewed.

**Results:** Thirteen patients were included. Five (38.5%) had undergone initial PT surgery at our center and 8 (61.5%) at other centers. Redo PT procedures were transanal endorectal PT in 12 patients (92.3%) and the posterior sagittal approach in 1 patient (7.7%). Indications for redo PT included pathologic misdiagnosis in 8 patients (61.5%); stricture in 2 (15.4%); and rectal stenosis, obstructing Duhamel pouch and remnant septum in 1 each (7.7%). At a median follow-up of 68 months (range, 3–227 months) after redo PT, 8 patients (61.5%) had normal bowel function, 2 (15.4%) had incontinence, and 1 (7.7%) had constipation.

**Conclusion:** Redo PT procedures could be an effective approach for improving obstructive symptoms in HD patients with anatomic or pathologic reasons following primary PT. Careful selection of patients and discreet indications for redo PT are crucial.

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**Key Words:** Congenital megacolon, Complication, Hirschsprung disease, Pull-through, Redo pull-through

## INTRODUCTION

Hirschsprung disease (HD) is a congenital anomaly of the enteric nervous system causing intractable constipation which requires surgical correction. Following the first successful operation for HD [1], several surgical techniques have been developed. Most patients with HD have favorable outcomes, although some experience long-term postoperative complications, including constipation, fecal incontinence,

recurrent Hirschsprung-associated enterocolitis (HAEC), and failure to thrive (FTT) [2-4]. Obstructive symptoms after a pull-through (PT) occur around 8%–30%, and the causes can be classified as anatomical, pathologic, and functional [4]. Although conservative medical treatment or minor surgical intervention can improve these symptoms, some patients may require redo PT [5]. Indications for redo PT surgery can include pathologic and/or anatomic factors. Anatomical problems can include anastomotic stricture, stenosis, obstructing

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Duhamel pouch, remnant septum, and obstructing Soave cuff, whereas pathologic problems include retained aganglionosis and transition zone pathology [2,5]. Most of the anatomical problems are related to the initial PT procedure, which can be confirmed by rectal examination or contrast enema. In the absence of mechanical obstruction on rectal examination and contrast enema, a rectal biopsy should be performed to determine pathologic reasons [4].

Several studies on redo PT surgery in HD patients have been previously published. However, the outcomes vary by report and long-term outcomes were scarce. Therefore, we aimed to evaluate the clinical outcomes of redo PT surgery in HD patients at a single center.

## METHODS

The medical records of patients who underwent redo PT for HD at a single tertiary center, between June 2003 and October 2019 were retrospectively reviewed. Data regarding clinical characteristics, pathologic results, initial surgical techniques, redo PT techniques, symptoms and indications for redo surgery, complications after redo PT, and follow-up clinical outcomes were collected. The study was approved by the Institutional Review Board of Asan Medical Center (No. 2020-0188). It was performed in accordance with the Declaration of Helsinki and written informed consent was waived due to its retrospective nature.

All redo PT procedures were performed by a single surgeon. Patients were included if they had postoperative obstructive symptoms after initial PT, including refractory constipation and episodes of enterocolitis or FTT. For work-up, contrast

enema (Figs. 1, 2) was done in every patient, except for those confirmed to have pathologic problems by biopsy.

The surgical technique for redo PT was determined by the surgeon according to the patient's state. Posterior sagittal approach was performed in 1 patient in 2003 and transanal endorectal PT (TERPT), which has a similar basic procedure to the Soave procedure, was performed thereafter [6]. TERPT underwent in lithotomy position, and laparoscopy was performed for the mobilization of the colon if necessary.

## RESULTS

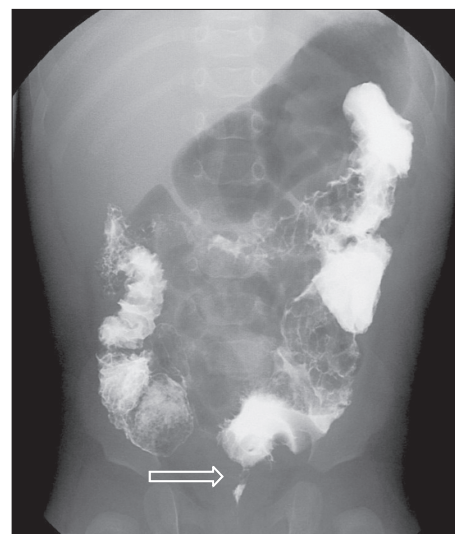
### Demographics

During the study period, 187 patients underwent PT procedure for HD at Asan Medical Center, 13 of which were redo PT. The 13 patients included 9 boys and 4 girls, with median age of 35 months (range, 1–270 months) at the time of redo PT. The median interval between primary and redo PT was 27 months (range, 18 days–168 months). Eight of the patients had primary PT at initial surgery, whereas 5 underwent staged operations. Five patients underwent initial PT at our center and 8 at another center. Initial PT procedures included the Soave procedure in 3 patients, the Duhamel procedure in 3, the Swenson procedure in 2, and TERPT in 5.

At the time of initial PT, aganglionic segments were in the rectum of 3 patients (23.1%), the sigmoid colon in 2 (15.4%), and the descending colon in 1 (7.7%), with 2 patients (15.4%) having total colonic aganglionosis (TCA). The transition zone in 5 patients (38.5%) was not documented (Table 1).



**Fig. 1.** Contrast enema of a patient who had a Duhamel procedure. The enlarged Duhamel pouch was causing obstructive symptoms.



**Fig. 2.** Contrast enema of a patient with retained aganglionosis after transanal endorectal pull-through. The arrow shows the transition zone.

**Table 1.** Demographics and clinical characteristics of patients included in this study

Characteristic	Data
No. of patients	13
Sex, male:female	9:4
Age (mo)	35 (1–270)
Initial PT procedure	
Soave	3
Duhamel	3
Swenson	2
TERPT	5
Aganglionic segment	
Rectum	3
Sigmoid colon	2
Descending colon	1
TCA	2
Unknown	5
Interval between initial and redo PT (mo)	27 (0.7–168)

Values are presented as number only or mean (range). PT, pull-through; TERPT, transanal endorectal pull-through; TCA, total colonic aganglionosis.

**Table 2.** Symptoms and procedures prior to redo pull-through (PT)

Variable	Data
Symptoms before redo PT <sup>a)</sup>	
Constipation with enterocolitis	5 (38.5)
Constipation without enterocolitis	5 (38.5)
Failure to thrive	2 (15.4)
Hematochezia	1 (7.7)
No defecation after initial PT	1 (7.7)
Procedures before redo PT (n = 9, 69.2%) <sup>b)</sup>	
Diverting stoma formation	7 (53.8)
Stoma revision	3 (42.9) <sup>c)</sup>
Anal hegar dilatation	5 (38.5)
Botulinum toxin injection	4 (30.8)
Septoplasty	1 (7.7)

<sup>a)</sup>Numbers do not add to total; some patients presented with more than 1 symptom. One patient who presented constipation with recurrent enterocolitis also showed failure to thrive. <sup>b)</sup>Numbers do not add to total; some patients underwent more than 1 procedure. <sup>c)</sup>42.9% of patients who underwent diverting ostomy formation.

### Assessments and procedures prior to redo pull-through

Ten patients (76.9%) presented with constipation, including 5 with (38.5%) and 5 without (38.5%) episodes of enterocolitis. Other presenting symptoms included FTT in 2 patients (15.4%), hematochezia in 1 (7.7%), and absence of defecation after initial PT in 1 (7.7%). One patient who presented constipation with recurrent enterocolitis also showed FTT (Table 2).

Seven patients (53.8%) underwent diverting ileostomy formation in our center prior to redo PT to alleviate obstructive symptoms during the preoperative evaluation period. All of

**Table 3.** Summary of patients in this study who underwent redo pull-through (PT)

Case No.	Sex	Age <sup>a)</sup> (mo)	Initial PT procedure	Aganglionic segment	Redo indication	Interval <sup>b)</sup> (mo)	Priorostomy <sup>c)</sup>	Redo procedure	Post-redo symptoms
1	Male	16	Soave	Unknown	Anastomotic stricture	8	Yes	PSA	None
2	Male	7	TERPT (L)	Rectum	Residual aganglionosis	6	No	TERPT (L)	Incontinence
3	Female	36	TERPT (L)	Rectum	Rectal stenosis	35	No	TERPT (L)	Incontinence
4	Female	1	TERPT	Rectum	Transition zone	0.7	No	TERPT	None
5	Female	270	Soave	Unknown	Anastomosis stricture	112	Yes	TERPT	None
6	Female	13	Duhamel	Unknown	Remnant septum	10	Yes	TERPT	None
7	Male	185	Duhamel	Unknown	Obstructing Duhamel pouch	168	Yes	TERPT	None
8	Male	52	TERPT (L)	DC	Transition zone	27	No	TERPT	None
9	Male	34	TERPT	Unknown	Transition zone	34	No	TERPT	None
10	Male	35	Duhamel	TI	Transition zone	23	No	TERPT	Diarrhea
11	Male	102	Swenson	TI	Transition zone	102	Yes	TERPT (L)	Diarrhea
12	Male	54	Soave	SC	Transition zone	49	Yes	TERPT (L)	None
13	Male	12	Swenson	SC	Residual aganglionosis	10	Yes	TERPT (L)	Constipation

TERPT, transanal endorectal pull-through; L, laparoscopy-assisted; DC, descending colon; TI, terminal ileum; SC, sigmoid colon; PSA, posterior sagittal approach. <sup>a)</sup>Age at the time of redo PT (months). <sup>b)</sup>Time interval between initial and redo PT (months). <sup>c)</sup>Diverting ostomy formation prior to redo procedure.

them received initial PT at other institutions and were referred to our institution. Three patients who underwent stoma formation had to undergo additional stoma revision due to stoma-related complications, including prolapse in 2 (66.7%) and retraction in 1 (33.3%).

Nine patients (69.2%) underwent other treatment interventions to resolve obstructive symptoms such as constipation, enterocolitis, or FTT before redo PT, including anal dilation, botulinum toxin injection, or septoplasty for remnant septum, but these procedures were not successful (Table 2). For the 4 patients with suspected spasms of the anal sphincters, 5 mg/kg of botulinum toxin was injected between external anal sphincter and internal anal sphincter in 4 divided doses around the anal opening as in the previously published protocol [7]. However, 3 of 4 were found to have transition zone pathology, and an additional patient was found to have anastomotic stricture.

A patient who underwent the Duhamel procedure at another institution showed anastomotic stricture on contrast enema, and during examination under anesthesia, the remaining septum of the anastomosis area was confirmed. Septoplasty was performed via laparotomy sequentially. The anterior wall of original rectum was incised and the remaining septum was removed, but it was ineffective.

Case 10 in Table 3 is 1 of the 2 patients diagnosed with TCA who underwent the Duhamel procedure for initial PT in our institution but subsequently experienced constant constipation and recurrent enterocolitis. Case 11 in Table 3 is the other patient diagnosed with TCA who underwent the Swenson procedure at another institution and was referred to our hospital with uncontrolled diarrhea and intermittent hematochezia. This patient was suspected of sphincter muscle impairment during examination under anesthesia and had severe skin erosion around the anus, thus underwent diverting ileostomy prior to redo PT.

### Redo pull-through: indications and surgical techniques

Indications for redo PT in the 13 patients included pathologic problems in 8 (61.5%), stricture in 2 (15.4%), and rectal stenosis, obstructing Duhamel pouch, and remnant septum in 1 each (7.7%). Of the 8 patients with pathologic problems, 2 had retained aganglionosis and 6 had transition zone pathology. Of the 13 redo PT operations, 12 (92.3%) were TERPT, including 5 with laparoscopy assisted, and 1 (7.7%) was performed via the posterior sagittal approach (Table 3).

### Postoperative clinical outcomes

Two patients (15.4%) experienced early complications including postoperative ileus and a ureter injury during redo PT procedure due to severe adhesion. Five of the 7 patients who

had a diverting stoma prior to redo PT performed stoma closure 4 to 6 months after redo PT, and the other 2 had stoma closure concurrently. None of the patients experienced anastomotic complications including leakage, abscess, or perforation.

After a median follow-up period of 68 months (range, 3–227 months), 8 patients (61.5%) had voluntary bowel movements, 1 (7.7%) needed laxatives for constipation, and 2 (15.4%) were incontinent. One patient with fecal incontinence showed spontaneous improvement of symptoms at the age of 7 years, which was 69 months after surgery. The patient with TCA who underwent the Duhamel procedure for primary PT and underwent redo TERPT was monitored by a pediatric gastroenterologist in the outpatient clinic due to recurrent ileitis with constant diarrhea. The other TCA patient with suspected sphincter muscle impairment underwent laparoscopic-assisted redo TERPT due to transition zone pathology. However, since the diarrhea was not alleviated, this patient underwent permanent ileostomy formation 20 months after the redo PT procedure (Table 3).

## DISCUSSION

Although surgery is usually successful in patients with HD, some patients experience postoperative morbidities, including constipation, fecal incontinence, and enterocolitis [2–4]. Improvements are often provided by medical treatment or minor surgical interventions, but some patients may require redo PT [5]. The present study evaluated long-term clinical outcomes in patients who underwent redo PT for various indications, with all procedures performed by a single surgeon.

Of the 13 cases requiring redo PT during the study period, 8 (61.5%) did so for pathologic issues, as confirmed by examination of the removed tissue specimens. Pathologic problem is usually caused by a pathologist's incorrect assessment of presence or absence of ganglion cells on frozen section of specimen, or by a surgeon's technical failure not going far enough above the pathological transition zone [4]. Three patients underwent redo PT based on biopsy results without contrast enema. Case 4 in Table 3 had regular ganglion cells in the frozen biopsy specimen during initial PT. However, the patient had no stool passage after the surgery, and the final pathology report indicates that, with a few ganglion cells present, this patient underwent redo PT 18 days after the initial PT. Case 8 in Table 3, who suffered from refractory constipation, was found to have a decrease in ganglion cell density only upon reassessment of the pathology slides 26 months after initial PT; this patient underwent redo PT without re-biopsy during the work-up period. Two sequential rectal biopsies of case 2 in Table 3 with recurrent enterocolitis and FTT after initial PT confirmed that the bowel was normally ganglionated. Contrast enema of this patient showed functional obstruction,



for which this patient underwent redo PT, and examination of the final specimen showed transition zone pathology. These cases indicate the importance of the evaluation of adequate and sufficient specimens by experienced and reliable pathologists [8].

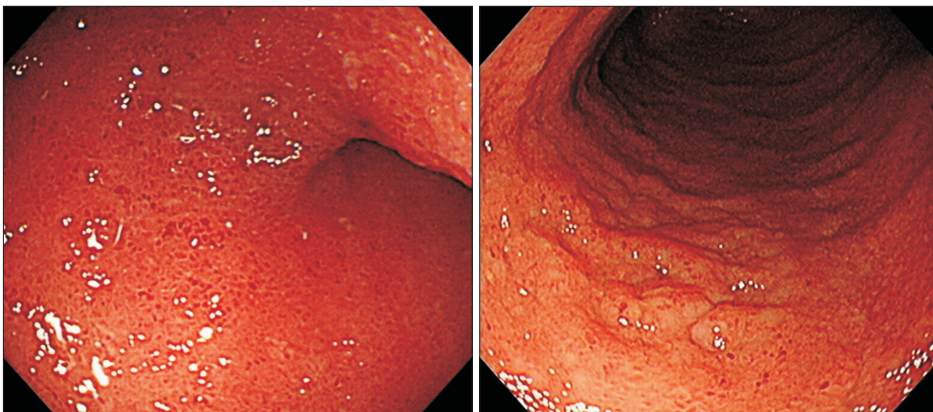
Two patients (15.4%) experienced early complications within 30 days after redo PT. One patient experienced postoperative ileus, which improved after conservative management, whereas the other experienced a ureter injury during redo PT due to severe adhesion, with this patient later requiring an additional ureteroureterostomy. None of the patients experienced an anastomotic leak, abscess, or perforation. This incidence was lower than in previous studies [9,10], which regarded preoperative stoma formation as advantageous. All 7 patients who underwent diverting ostomy formation prior to redo PT were referred from other institutions after initial PT. Since obstructive symptoms such as refractory constipation or HAEC were severe in patients who were referred from other institutions, diverting ostomy was performed to prevent obstructive symptoms during preoperative evaluation period. Five of the 6 patients who did not undergo stoma formation prior to redo PT had undergone initial PT at our institution, with initial PT in the sixth performed by a pediatric surgeon trained in our center. The absence of detailed medical records made it difficult to perform redo PT shortly after initial PT. Diverting ostomy formation prior to redo PT may be a safe option, enabling preoperative work-up with sufficient time to prevent the recurrence of obstructive symptoms. In addition, early complications after redo PT can be reduced by delayed stoma closure [11]. However, 42.9% of the patients with diverting ostomy in this study and 28% of pediatric patients with colostomy in a previous study had stoma-related complications [12], suggesting the need for careful patient selection.

Three patients (23.1%) experienced incontinence after redo surgery, with 1 showing symptom improvement during follow-up, an incidence lower than in previous studies [2,10]. It is important to preserve the anal canal during PT procedures [4,13],

as the low incidence of incontinence in this study appears to be due to careful preservation of the dentate line throughout the procedure.

Two patients (15.4%) diagnosed with TCA reported recurrent diarrhea after redo PT. One of these patients (case 10 in Table 3), who underwent redo PT for transition zone pathology, was diagnosed by a pediatric gastroenterologist with chronic active ileitis and possibility of Crohn disease (Fig. 3). This patient is currently on medication for Crohn disease and has been showing improvement in symptoms since. A meta-analysis reported that, of HD patients who developed inflammatory bowel disease (IBD) after PT operation, 41% had TCA and 45% showed aganglionic involvement of the long segment [14]. The possibility of IBD should be considered in patients with extensive colonic aganglioneosis if uncontrolled diarrhea persists after successful PT operation. The other TCA patient (case 11 in Table 3) with suspected sphincter muscle impairment eventually required a permanent ileostomy due to repeated diarrhea after redo PT. This patient was also diagnosed with chronic ileitis by the same gastroenterologist and received medical treatment, but symptoms did not improve. Findings in this patient once again indicate the importance of preserving the anal canal during initial PT [4,13].

The strengths of this study were its relatively long-term follow-up period after redo PT and that all operations were performed by a single expert surgeon, which makes all surgeries comparable by similar surgical skill. However, the present study had several limitations. Firstly, its retrospective design can lead to selection bias and 5 patients referred from other centers might not include sufficient data. Secondly, it has wide variability and heterogeneity of the patient population including initial length of aganglioneosis, type of primary PT, highly variable patient age, highly variable interval between initial and redo surgery, and the presenting symptoms leading to redo PT. Thirdly, included number of patients is too small to reach any conclusions, though we attempted to show long-term follow-up results of redo PT.



**Fig. 3.** Endoscopic findings of a patient with total colonic aganglioneosis after redo pull-through showing hyperemic mucosa with aphthous ulcers. Endoscopic biopsy reported chronic active ileitis with dense lymphoplasmacytic infiltration in lamina propria and cryptitis.

In conclusion, redo PT procedures could be effective approach for improving obstructive symptoms in HD patients with anatomic or pathologic reasons following primary PT. Careful selection of patients and discreet indications for redo PT are crucial.

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### Conflict of Interest

No potential conflict of interest relevant to this article was reported.

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Writing – Review & Editing: All authors

## REFERENCES

- Swenson O, Bill AH Jr. Resection of rectum and rectosigmoid with preservation of the sphincter for benign spastic lesions producing megacolon; an experimental study. *Surgery* 1948;24:212-20.
- Peña A, Elicevik M, Levitt MA. Reoperations in Hirschsprung disease. *J Pediatr Surg* 2007;42:1008-14.
- Levitt MA, Dickie B, Peña A. Evaluation and treatment of the patient with Hirschsprung disease who is not doing well after a pull-through procedure. *Semin Pediatr Surg* 2010;19:146-53.
- Ahmad H, Yacob D, Halleran DR, Gasior AC, Lorenzo CD, Wood RJ, et al. Evaluation and treatment of the post pull-through Hirschsprung patient who is not doing well; update for 2022. *Semin Pediatr Surg* 2022;31:151164.
- Ralls MW, Coran AG, Teitelbaum DH. Redo pullthrough for Hirschsprung disease. *Pediatr Surg Int* 2017;33:455-60.
- De la Torre-Mondragón L, Ortega-Salgado JA. Transanal endorectal pull-through for Hirschsprung's disease. *J Pediatr Surg* 1998;33:1283-6.
- Ralls MW, Coran AG, Teitelbaum DH. Reoperative surgery for Hirschsprung disease. *Semin Pediatr Surg* 2012;21:354-63.
- Lawal TA, Chatoorgoon K, Collins MH, Coe A, Peña A, Levitt MA. Redo pull-through in Hirschsprung's [corrected] disease for obstructive symptoms due to residual aganglionosis and transition zone bowel. *J Pediatr Surg* 2011;46:342-7.
- Ralls MW, Freeman JJ, Rabah R, Coran AG, Ehrlich PF, Hirschl RB, et al. Redo pullthrough for Hirschsprung disease: a single surgical group's experience. *J Pediatr Surg* 2014;49:1394-9.
- Dingemans A, van der Steeg H, Rassouli-Kirchmeier R, Linssen MW, van Rooij I, de Blaauw I. Redo pull-through surgery in Hirschsprung disease: short-term clinical outcome. *J Pediatr Surg* 2017;52:1446-50.
- Li Q, Zhang Z, Xiao P, Ma Y, Yan Y, Jiang Q, et al. Surgical approach and functional outcome of redo pull-through for post-operative complications in Hirschsprung's disease. *Pediatr Surg Int* 2021;37:1401-7.
- Nour S, Beck J, Stringer MD. Colostomy complications in infants and children. *Ann R Coll Surg Engl* 1996;78:526-30.
- Bischoff A, Frischer J, Knod JL, Dickie B, Levitt MA, Holder M, et al. Damaged anal canal as a cause of fecal incontinence after surgical repair for Hirschsprung disease: a preventable and under-reported complication. *J Pediatr Surg* 2017;52:549-53.
- Nakamura H, Lim T, Puri P. Inflammatory bowel disease in patients with Hirschsprung's disease: a systematic review and meta-analysis. *Pediatr Surg Int* 2018; 34:149-54.