

**REDO PULL-THROUGH: PATIENT CHARACTERSTICS, INDICATIONS  
AND OUTCOME AT TIKUR ANBESSA HOSPITAL, ETHIOPIA, 2010-  
2019**



**COLLEGE OF HEALTH SCIENCES**

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## **Declaration**

I **Hadush Tesfay** declare that this paper is a result of my independent research work on the topic entitled “**Redo pull-through: patient characteristics, indications & outcome at Tikur Anbessa hospital, Ethiopia, 2010-2019**” in partial fulfillment of the requirements for specialty certificate for General Pediatric Surgery at Addis Ababa University, College of Health Sciences, Department of Surgery, Pediatric Surgery Unit. This work has not been submitted for a degree to any other university. All the references are also acknowledged.

Dr. Hadush Tesfay

Signature: \_\_\_\_\_

Date: \_\_\_\_/\_\_\_\_\_/\_\_\_\_\_

## **Confirmation**

This is to certify that **Hdush Tesfay** has carried out this research work on the topic entitled “**Redo pull-through: patient characteristics, indications & outcome at Tikur Anbessa hospital, Ethiopia, 2010-2019**” under my supervision. This work is original in nature and has not been presented for a degree in any University and it can be submitted for the partial fulfillment of the requirements for the award of the specialty certificate for General Pediatric Surgery.

Dr. Hanna Getachew (Associate Professor of General & Pediatric Surgery)

Signature: \_\_\_\_\_

Date: \_\_\_\_/\_\_\_\_\_/\_\_\_\_\_

## **AKNOWLEDGMENT**

I would like to forward my gratitude to my advisor Dr Hanna Getachew for her continuous mentorship, guidance, assistance and constructive comments throughout the preparation of the research thesis. I also thank my mentors and colleagues in the unit of pediatric surgery for their support.

## ACRONYMES

AAU	Addis Ababa University
CHS	College of Health Sciences
ERPT	Endorectal Pull Through
HAEC	Hirschsprung Disease Associated Enterocolitis
HSD	Hirschsprung Disease
PSARP	Posterior Sagittal Anorectoplasty
PT	Pull Through
TERPT	Transanal Endorectal Pull Through

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## Abstract

**Background:** HSD is a developmental disorder of the intrinsic nervous system of the distal colon. The management is pull through procedure and majority of patients do well after surgery; but 32% of patients continue to have complaints after surgery & up to 8.6% of patients can require repeat pull through.

**Objective:** To describe indications for reoperation, surgical technique adopted, complications, and outcome of patients who underwent a redo pull-through procedure in Tikur Anbessa hospital, Ethiopia from

**Methodology:** Retrospective descriptive study design was used and the sampling procedure was a multi-stage sampling. Descriptive statistics (mean, SD, frequency, percentage, graph and table) was generated by using SPSS version 21.

**Results:** In our study a total of 18(4.87% of all PT) patients underwent redo pull through from 2010-2019. 13 (72.2%) were males & 5 (27.8%) were females with male to female ratio of 2.6:1. Soave pull through was the most frequent type of pull through on first time; done on 16(94.2%) of patients, two being primary TERPT, and Duhamel pull through for one(5.9%) patient. The type of initial PT wasn't known in one. The indication for redo PT was pathologically confirmed persistent/acquired aganglionosis in 8(50%) patients. 5(27.8%) patients had anastomotic site stricture confirmed with examination under anesthesia. Three of them were done for persistent obstructive features with clinical impression of aganglionosis. Twisted colon & anastomotic failure with colonic retraction accounted for one patient each. Swenson procedure was performed on 15 (83.3%)patients & Soave pull through was done on 3(16.7%) patients as the procedure for the redo PT. Long term outcome of patients after redo pull through was measured using Holschneider's FII score. Of the patients FII score was obtained (11 of them), 6 patients achieved good continence,3 has fair continence status & one patient was incontinent & required bowel management program to stay clean. One patient was less than 4 years old & not toilet trained yet. In 7 patients the Holschneider FII score couldn't be obtained.

**Conclusion :** The indication for redo pull through was persistent obstructive features in 77.8% of patients. The causes of persistent obstructive symptoms after an initial pull-through procedure are pull through of aganglionated segment (50%) and anastomotic site stricture (27.8%). 81.8% of those for whom the Holschneider FII score was obtained has fair to good continence.



# **1 Introduction**

## **1.1 Background**

Hirschsprung's disease (HSD) is a developmental disorder of the intrinsic component of the enteric nervous system characterized by the absence of ganglion cells in the myenteric and submucosal plexuses of the distal intestine beginning at the internal anal sphincter and extending proximally for varying distances[1, 2]. The incidence of HSD is estimated to be 1 in 5,000 live births and ranges from 1 in 2,000 to 1 in 12,000 live births[1, 2]. Males are more commonly affected than females with a male to female ratio of 4:1. Most affected patients present with neonatal intestinal obstruction; delayed passage of meconium beyond the first 24 hours is present in approximately 90%. Other manifestations include chronic constipation with failure to thrive, Hirschsprung disease associated enterocolitis, and rarely neonatal cecal/appendiceal perforation [1, 2].

The surgical management for Hirschsprung disease is to remove the aganglionic bowel and reconstruct the intestinal tract by bringing the normally innervated bowel down to the anus while preserving normal sphincter function (pull through procedure). The most commonly performed operations are the Swenson, Duhamel, and Soave procedures[1-3]. Laparoscopy and transanal (endorectal) pull-through (TERPT) techniques were introduced in the late 90s as less invasive procedures and are increasingly becoming surgery of choice[1, 2, 4, 5].

## **1.2 Statement of the problem**

Although surgery is effective in most HSD patients, up to 32% of these patients continue to have symptoms and complaints. These persisting symptoms and complaints include fecal incontinence, constipation and persistent stooling problems, and Hirschsprung disease associated enterocolitis (HAEC). Additional laxative treatment is generally needed and a very small fraction of the patients with obstructive symptoms needs secondary surgery[1, 2, 4, 6]. 3-8.6% of patients who undergo pull through (PT) require redo PT with 43-71% of them showing abnormal histological findings[4, 5, 7, 8]. Indications for redo PT include residual or acquired

aganglionosis, anastomotic stricture, twisting of the pulled through colon, fistulae, constricting cuffs, megarectal pouches, and fecal incontinence or soiling[1, 2, 4, 6].

The average age at primary PT is 5 months-1 year[5-7], mean age at redo PT ranges between 3.5-7.5 years[3, 4, 6, 8], with mean follow up period from primary to redo surgery of 3-5.1 years[3, 5, 6]. 52-75 % of patients who require redo surgery are males[7-10]. Mean follow-up after Redo operation was 3-13.8 years[3, 4, 10, 11].

70-94% of patients who have undergone redo PT and who don't have neurological problem and are above the age of toilet training have voluntary stooling without the requirement of medications and are continent[3, 8, 10, 11]. The papers that have been published on redo PT recommend that the procedure has a satisfactory outcome when done on carefully selected patients; appropriately selected children undergoing a Redo PT can achieve good results, with comparable continence rates to those undergoing a primary PT.

### **1.3 Rationale of the study**

Up to 32% of patients who undergo pull through procedure continue to have symptoms and complaints after the surgery; majority of these patients respond well to non-surgical management. 3-8.6% of patients who undergo pull through (PT) require redo PT, and should be managed with a center based management guidelines to have satisfactory outcome. To our knowledge few published studies are available regarding redo pull through in Africa and no studies in our center. So, the results of this study can provide :

- Baseline data about the rate of redo PT, the contributing factors and management outcome in our hospital
- Used to compare our level of care with other centers
- Help in the development of management guideline for patients who are not doing well after pull through
- Help improve our practice of care

## 2. Literature review

### 2.1 Patient profile and indications for redo PT

The average age at primary PT is 5 months-1 year[5-7], mean age at redo PT ranges between 3.5-7.5 years[3, 4, 6, 8], with mean follow up period from primary to redo surgery of 3-5.1 years[3, 5, 6]. 52-75 % of patients who require redo surgery are males[7-10].

Jiang et.al reported that 72 patients (8.6% of all for whom PT was done) had redo operation. The patients' age at the primary operation ranged from 1 month to 6 years, with an average of 5 months. The interval between initial and redo operation varied from 12 months to 5 years (mean, 3 years)[5].

Lawal et.al showed in their study that 16 children (median age, 7.5 [2-17] years) presented at 6 to 66 months after initial pull-through (performed at median, 5.5 months [5 days-2 years]). Fourteen were boys, and 2 were girls[6].

In Dingemans' et al. study sixteen patients were included with a median age at study of 6 years and 9 months (range 1-25 years) and age at the time of redo surgery being 4 years and 7 months (range 2 months to 21 years). Ten of them were males, 6 females.[4]

Van Leeuwen et al. performed redo PT in 19 patients. Thirteen of the 19 patients were boys and 6 were girls. One patient had a family history positive for Hirschsprung's disease, and one patient had Trisomy 21[10].

Pini-Prato et al performed 70 redo PT in their study. Twenty-two patients (61%) complained of obstructive symptoms, 11 (30%) recurrent or severe acute HAEC, and 10 (28%) severe chronic constipation. Seven patients (19%) experienced both obstruction and enterocolitis. Twenty-four patients (67%) required a stoma to relieve symptoms before reoperation (ileostomy in 10, colostomy in 18). Six had it fashioned elsewhere before admission to our institution. The enterostomy was performed to decompress the bowel in case of HAEC or severe bowel distension to avoid contamination and further inflammation in case of leakage or pelvic fibrosis and to cope with unresponsive nursing in all patients with TCSA. Of 24 patients who required a

stoma, 9 (37.5%) experienced complication(s) (prolapse in 7, retraction and/or stenosis in 2, and skin excoriation in 3), which required some sort of surgical reintervention in 2 (8%)[8].

Of the 16 patients for whom redo PT was done by Dingemans et al. 4 patients(25%) were fecally incontinent & 2 patients(13%) reported staining. In 14 patients (88%) other treatment interventions (anal dilation or botulinum toxin injection treatment) were performed before the redo surgery, but were not successful[4].

The primary pull-through was Soave procedure (or its modifications) in 50- 83.3% patients, Duhamel (or its modifications) in 13.8-26.3%, Rehbein in 5.2-18.7%, Swenson in 12.5-15.7% and a posterior sagittal approach (PSARP) in 2.7%[4, 8, 10].

Of the 19 redo PT Van Leeuwen et al. performed, the primary pull-through procedures included 10 endorectal pull-throughs (ERPT), 5 Duhamel procedures, 3 Swenson procedures, and 1 Rehbein procedure[10]. Pini-Prato et al performed 70 redo PT in their study. 36 patients had complete follow up. Of the 36, the failed pull-through was a Soave procedure (or its modifications) in 30 patients, a Duhamel (or its modifications) in 5, and a posterior sagittal approach (PSARP) in 1 patient[8].

Indications to reoperation were abnormal histological findings in 43-71% of them [4, 5, 7, 8, 12]. Anastomotic stricture, anastomotic leak with subsequent stenosis , cuff stricture , cuff abscess with subsequent rectal fibrosis ,persistent or recurrent enterocolitis, and twisted pulled through bowel contributed for the remaining patients.

Hassan et al. performed 33 redo PT procedures over 13 years. The indications were; obstructive symptoms due to retained or quired aganglionic segment in 15 patients, severe anastomotic stricture in 7 patients & enterocolitis with or without constipation in 11 patients[12].

Pini-Prato et al performed 70 redo PT in their study. Indications to reoperation were residual aganglionosis or hypoganglionosis in pulled-through colon in 41 patients, retained aganglionic rectum (high anastomosis) in 10, cuff stricture in 5, anastomotic stricture in 3, persistent or recurrent enterocolitis in 2, leakage with subsequent perirectal abscess and fibrosis in 2, and

twisted pulled-through bowel in 1. Indications could not be retrieved in 6 patients because of improper notes compilation[8].

In Dingemans et al. 16 redo PT the indication for redo TERPT was a suspected mechanical obstruction in or surrounding the distal colon and anal canal in all patients; in 7 patients pathological origin of the colon (transition zone or residual aganglionosis) was proven preoperatively[4].

## 2.2 Redo Surgery

Mean age at redo PT ranges between 3.5-7.5 years[3, 4, 6, 8]. The choice of a specific technique for redo pull-through is quite variable in various reported series[12].

Lawal et al. analyzed the results of 16 patients for whom redo PT was done for histopathologic indications only. The reoperation consisted of a transanal pull-through (n = 15) or a redo via a posterior sagittal approach (n = 1), preferred because of excessive pelvic scar tissue. Of the 15 who had a transanal pull-through, 10 had laparoscopy or laparotomy to complete the colonic mobilization. In 5 patients, the redo was done transanally alone. The median length of bowel resected at the reoperation was 15 (6-31) cm[6].

Pini-Prato et al performed 70 redo PT in their study. Of the 36 patients who had complete follow up twenty-two patients underwent an endorectal pull-through (Soave and/or its modifications), 9 a Duhamel procedure, and 5 a Swenson. Mainly endorectal pull-through was used because of its extreme versatility. However, in some instances, other procedures were preferred because of impaired rectal blood supply, severe perirectal inflammation and fibrosis, and surgeon's judgement[8].

In Dingemans et al. 16 redo PT, the type of surgical technique for the redo procedure consisted of a Swenson-like TERPT in most cases (94%)[4].

### **2.3 Pathology of resected specimen**

Dingemans et al. performed sixteen redo endorectal pull-through surgery in their center between 2007 and 2015. In all patients, pathology was obtained by biopsy during surgery and from the final specimen. Final pathology indicated that 25% patients had residual aganglionosis, in 25% the presence of transition zone was observed in the resected bowel. In another 19% residual cuff tissue was removed. A torsion of the distal bowel was reported in 13%; in 13% patients fibrotic and/or ischemic tissue was seen. In all cases the proximal segment of the redo-resected bowel had normal ganglion cells and no abnormal nerves [4].

Lawal et al. analyzed the results of 16 patients for whom redo PT was done for histopathologic indications only. All of the 16 children had hypertrophic nerves. Ten had hypertrophic nerves with normal ganglion cells. In the remaining 6, our biopsy showed both absent ganglion cells and hypertrophic nerves[6].

### **2.4 Early post redo operation complications**

Pini-Prato et al performed 70 redo PT in their study. 36 patients had complete follow up. Of the 36, twelve patients (33%) experienced postoperative complications, namely, adhesions and obstruction in 7, incisional hernia in 2, anastomotic stricture in 2, and enterocolitis in 1. Eight of these patients were managed conservatively, whereas 4 required some sort of surgical intervention (adhesiolysis, abdominal wall plasty, anal dilatation, and botulinum toxin injection). Those complications were experienced regardless of surgical technique adopted, length of aganglionosis , and associated anomalies.

Dingemns et al. performed 16 redo PTs. Following redo surgery, 5 patients (31%) reported 7 complications within 30 days after redo surgery: anastomotic dehiscence (3×), infection leading to abscess (2×), a rectovaginal fistula (1×) and an enterocutaneous fistula (1×). All these patients needed additional surgery because of these complications & eventually recovered completely from these complications[4].

## 2.5 Post redo operation outcome

Mean follow-up after Redo operation was 3-13.8 years[3, 4, 10, 11].

Pini-Prato et al performed 70 redo PT in their study. 36 patients had complete follow up. Post redo follow-up was  $6.6 \pm 4.3$  years. Two patients younger than 4 years were excluded from fecal continence assessment. Excellent to good continence (wingspread  $\geq 2$ ) was described by 24 (70%) of 34 patients, fair continence by 6 (18%), and poor by 4 (12%). Average daily bowel movements at follow-up were  $2.4 \pm 2$ . 17% of patients required dietary limitation to contain symptoms. Frequent laxatives administration and/or repetitive cycles of decontamination were required in 11%. Chronic constipation in the long term was experienced by 14% of patients. 30% experienced recurrent episodes of enterocolitis and/or severe chronic perineal excoriations in the postoperative period; all except one patient settled with conservative treatment and one patient was still experiencing recurrent enterocolitis and excoriations 5 years after surgery. Significant continence improvement was experienced by 10 patients in the long term with mean age at symptoms resolution of  $8.5 \pm 3$  years [8].

In the Lawal et al study of 16 patients who had redo PT for retained or acquired aganglionosis, median follow-up period was 16 months (3 months-10 years). Eleven (85%) of the 13 patients who are above the age of toilet training have voluntary bowel movements, with normal stooling while on little or no medication (laxatives). One child with Down's syndrome had the initial pull-through bowel anastomosed to the anocutaneous junction and still has a diverting stoma. Another patient has soiling, despite having an intact anal canal and daily enemas. One patient was lost to follow-up. The remaining 2 patients are less than 3 years of age and have not been assessed for fecal continence. No patient had recurrent enterocolitis requiring irrigations after the reoperation, and all thrived with good weight gain[6].

Hassan H et al reported in their study that 33 patients underwent a redo surgery for Hirschsprung's disease. The outcome in 28 patients, who were older than 3 years at time of last follow up visit are; normal or near normal bowel habits in 16 (57.1 %), recurrent constipation and obstructive symptoms in 5 (17.9%), and rank incontinence or repeated soiling in 7 (25%)[12].

In Schweizer et al study seventeen patients with Hirschsprung's disease aged 2 to 9 years (median, 4.6 years) underwent a repeated pull-through procedure because of unresponsive symptoms after an initial operation. Surgical revision was indicated by incomplete resection of the transition zone in 16 patients, anastomotic strictures in 9 patients, and fistulas in 2 patients. All 17 patients underwent Redo Duhamel pull-through procedure. Median follow-up after Redo operation was 9 years (range, 1-23 years). In 14 of 17 patients, the obstructive symptoms disappeared completely immediately after Redo Duhamel procedure and did not recur. These patients characterize their stooling pattern as completely normal. The outcome of two patients is characterized by grade 2 or 1 constipation, in whom the surgeon records the result as unsatisfactory, but the patients characterize their stooling pattern as satisfactory. In one patient, the outcome couldn't be judged as yet because the pull-through procedure was only 1 year back and colostomy has not been removed[11].



### **3. Objectives**

#### **3.1 General objective**

To describe indications for reoperation, surgical technique adopted, complications, and outcome of patients who underwent a redo pull-through procedure at TASH from January 2010-December 2019

#### **3.2 Specific objectives**

1. Determine the indications for redo pull through in patients who underwent redo pull through at TASH from 2010-2019
2. Determine the causes of persistent obstructive symptoms after an initial pull-through procedure in patients who underwent redo pull through at TASH from 2010-2020
3. Describe the preferred surgical technique used for redo pull through in patients who underwent redo pull through at TASH from 2010-2019
4. Describe the early postoperative complications in patients who underwent redo pull through at TASH from 2010-2019
5. Determine the early & long term outcome of patients after redo pull through in TASH from 2010-2019

## **4 Materials & methods**

### **4.1 Study setting**

This is a hospital based study in Tikur anbesa specialized hospital (TASH), Addis Ababa, Ethiopia. TASH is a tertiary & teaching hospital with the highest pediatric surgical patient load in the country.

The unit of pediatric surgery in Tikur anbesa specialized hospital currently has seven pediatric surgeons, two fellows and seventeen residents. It has an inpatient ward with twenty beds for elective admissions, five to ten beds for emergency admissions with their nursing staff, and six elective OR tables per week. It is a center where over 1000 elective pediatric surgical procedures are performed annually & an average of 4-6 pull through procedure are done monthly (taken from monthly audit reports).

### **4.2 Study design**

This study was conducted with a retrospective study design. Charts of patients who had undergone redo PT from January 2000- December 2019 were reviewed & telephone interview of parents/ guardians were conducted as a source of data, and the data was analyzed.

### **4.3 Source population**

The source population was all patients with HSD who underwent pull through procedure from January 2010- December 2019.

### **4.4 Study population**

The study population was all patients who underwent PT procedure 2 or more times from January 2010- December 2019.

#### **Inclusion criteria**

- All patients who underwent a redo procedure after a previous ineffective pull-through for HSD from January 2010- December 2019 in TASH

- Patients who required both a redo pull-through and a sphincterotomy-myectomy either preoperatively or postoperatively
- Minimum follow-up of 12 months

#### **Exclusion criteria**

- Dilatation under anesthesia

### **4.5 Sample size**

Patients who satisfy the inclusion criteria were included.

### **4.6 Sampling procedure**

The sampling procedure was multistage sampling where all patients with HSD who underwent PT from January 2010- December 2019 were identified first, and then those who had undergone PT two or more times were selected and used as a study population.

### **4.7 Data collection procedure**

#### **4.7.1 Instrument**

Structurally developed and pretested questioner was used as a data collection instrument. The questioner was developed by the researcher in English language and was an examiner-administered questioner filled from patient chart records & telephone interviews after being pre-tested for standardization.

#### **4.7.2 Study variables**

#### **Dependent variables**

- Type of redo pull through procedure
- Fecal continence status
- Presence & degree of constipation
- Occurrence of post redo enterocolitis
- Number of daily bowel movements

## **Independent variables**

- Age & sex of the patient
- Presence/absence of associated anomalies
- Clinical findings of the patient before redo surgery
- Investigation results
- Type of first pull through procedure
- Post pull through complications
- Pre redo patient management

### **4.7.3 Enumerators/supervisors**

Data collection was done by the researcher and research assistants(colleagues) who are familiar with the management of patients with HSD. Data completeness and consistency was supervised & checked by the researcher daily.

### **4.7.4 Procedure of data collection**

The questioners were filled, checked for completeness & consistency and entered to SPSS version 25.

## **4.8 Quality assurance**

Quality was controlled by pre-testing the questioners for standardization, checking for completeness & consistency after each questioner was filled, and data cleaning after entry.

## **4.9 Analysis**

Descriptive statistics were reported as absolute frequencies and percentages for qualitative data. Means and standard deviations were used to describe quantitative variables.

#### 4.10 Operational definitions

- Redo pull-through: revision of a previously performed pull through for hirschsprung disease
- Continence: clean between bowel movements.
- Hirschsprung disease associated enterocolitis :explosive diarrhea or abdominal distension requiring hospitalization for hydration and antibiotics
- Staining: stripes in underwear with no need for a change of underwear or use of additional aids.
- Soiling: staining with the need of additional aids.
- Fecal incontinence: fecal loss of larger amounts of stool with a social impact

## 5 Results

### 5.1 Initial presentation

A total of 369 patients underwent pull through for HSD from 2010- 2019 in Tikur Anbessa hospital, Ethiopia. Of those, 18(4.87%) patients underwent redo pull through, of which two had two unsuccessful PT procedures. 13 (72.2%) were males & 5 (27.8%) were females, with male to female ratio of 2.6:1.

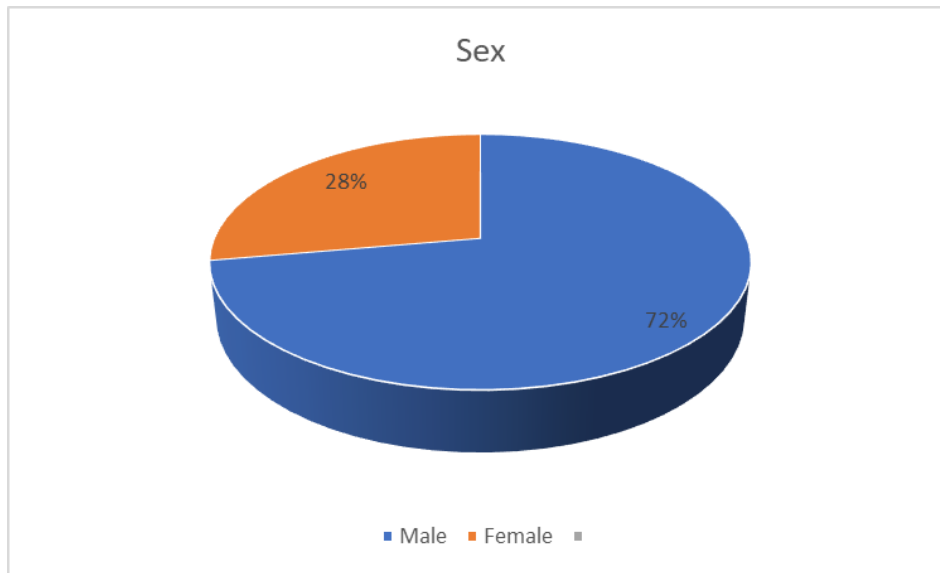


Figure 1. Patients' sex

The median age at the time of first pull through was 24 months with inter-quartile range of 21.25-33 months. The youngest child was aged 3 months and the eldest was 125 months of

Table 1. Age at first PT

Age at first pull through (in months)		
N	Valid	18
	Missing	0
Mean		30.33
Median		24.00
Mode		24
Std. Deviation		25.941
Variance		672.941
Range		122
Minimum		3
Maximum		125
Percentiles	25	21.25
	50	24.00
	75	33.00

Soave pull through with trans-abdominal mobilization of the colon was the most frequent type of pull through on first time; done on 14(82.4%) of patients.. Primary trans-anal endorectal Soave pull through was done for 2 (11.8%)patients, and Duhamel pull through for one(5.9%) patient. Data about the first pull through was not retrieved for one patient.

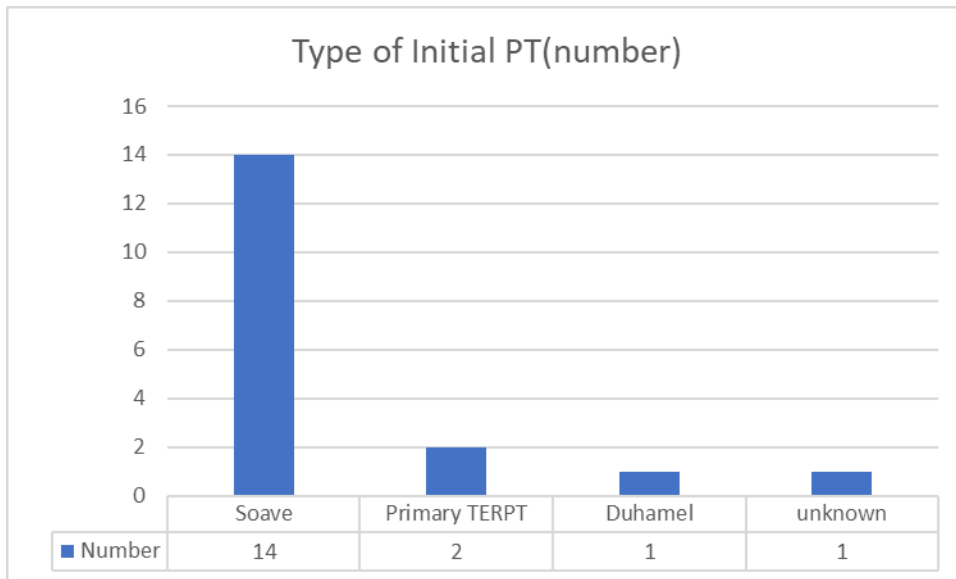


Figure 2. Type of initial PT

The pathologic evaluation result of the resected segment was obtained in 12 patients only; in six of the patients data on pathologic result wasn't found in the charts. Pathologic evaluation of the proximal end of the resected out colon showed ganglionic submucosa & musculari propria in 5(41.7% of biopsies) patients, aganglionic submucosa & muscularis propria in 6 (50% of biopsies) patients, and it was hypoganglionic in one patient. Normal sized nerve fibers were reported in the submucosa & muscularis propria of two of the ganglionated biopsies, and two of the aganglionated biopsies showed hypertrophic nerve fibers in Meissner's & Aurbach's plexuses.

The postoperative course in the first month after surgery was smooth in 11(61.1%) patients. Three patients(16.7%) developed intestinal obstruction, one patient had rectal retraction with cuff abscess, & intrabdominal collection, abdominal wound dehiscence & ECF occurred in one patient each.

Table 2. Early postoperative course after first PT

<b>Early postoperative course after 1st PT</b>	<b>Frequency</b>	<b>Percent</b>
Smooth course	11	61.1
Rectal retraction with cuff abscess	1	5.6
Intraabdominal collection	1	5.6
Intestinal obstruction	3	16.7
Wound dehiscence	1	5.6
Intraadominal collection, then ECF	1	5.6
<b>Total</b>	<b>18</b>	<b>100.0</b>

## 5.2 Redo Pull Through

The most frequent mode of presentation for redo PT was persistent obstructive features in 10(55.6%)patients. Four(22.2%) patients presented with persistent obstructive features and HAEC. One patient had anastomotic stricture, but he wasn't obstructed as he had a protective stoma. Early retraction of PT segment with cuff abscess occurred in 1(5.6%) patient, another one presented with fecal soiling, and for one patient redo PT was done for aganglionic PT segment though he wasn't symptomatic.



Clinical evaluation, examination under anesthesia, barium enema & rectal biopsy are the diagnostic modalities used before redo PT.

The indication for redo PT was pathologically confirmed persistent/acquired aganglionosis in 8(50%) patients. 5(27.8%) patients had anastomotic site stricture confirmed with examination under anesthesia. In three of the patients the redo surgery was done for persistent obstructive features with the clinical impression of persistent/acquired aganglionosis, of which barium enema was suggestive of HSD in one. One patient had twisted colon & one patient had anastomotic failure with colonic retraction.

Figure 3: pie-chart of indications for redo PT

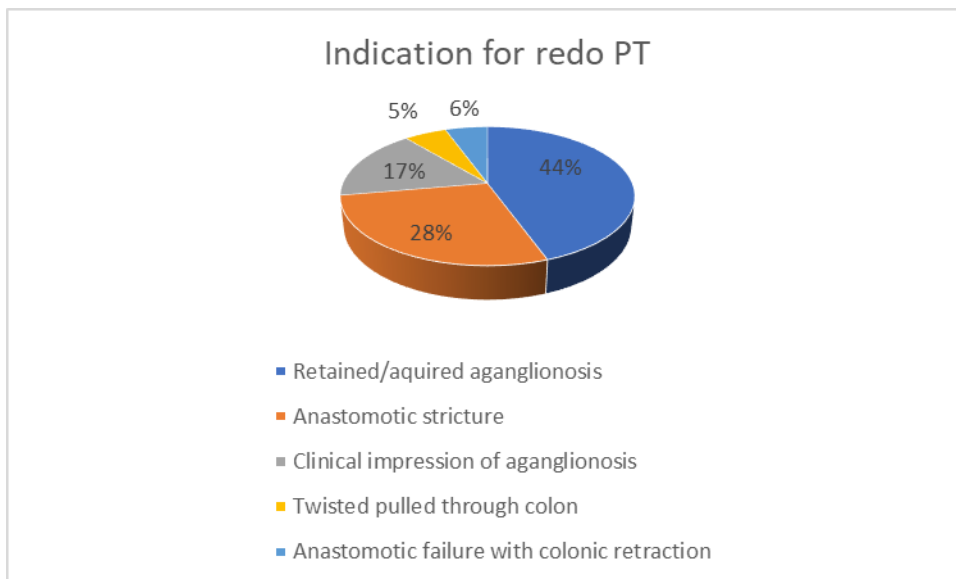


Figure 3. Indications for redo PT

The median age at the time of the second pull through was 48 months with inter-quartile range of 33.5-73.75 months. The youngest age was 10 months & the eldest was 168 months. The median inter-PT interval was 14.5 months, with a range of 1 month to 11.5 years.

Table 3. Age at redo PT

Age at redo PT (months)

N	Valid	18
	Missing	0
Mean		57.7222
Median		48.0000
Mode		36.00 <sup>a</sup>
Std. Deviation		41.95068
Variance		1759.859
Range		158.00
Minimum		10.00
Maximum		168.00
Percentiles	25	33.5000
	50	48.0000
	75	73.7500

Swenson procedure was performed on 15 (83.3%) patients; fourteen of them were done with abdominal mobilization of the colon & One was done transanally. Soave pull through was done on 3 (16.7%) patients; two of them transanally & one with abdominal mobilization.

Table 4. Type of redo PT

Type of redo PT		Frequency	Percent
Valid	Transanal endorectal soave pull through	2	11.1
	Transanal endorectal Swenson pull through	1	5.6
	Swenson with abdominal mobilization	14	77.8
	Soave with abdominal mobilization	1	5.6
	Total	18	100.0

The resected segment was subjected to pathologic evaluation in 14 patients. Thirteen (92.9%) of the biopsies showed ganglionic submucosal & myenteric plexuses, seven (50%) of them also showed normal looking nerve fibers in the Meissner's & Auerbach's plexuses. One biopsy (7.1%) was hypoganglionic.

The postoperative course in the first month after surgery was smooth in 15 patients ( 83.3%).one patient had rectal retraction & reanastomosis was done. One patient developed perianal fistula which had whitish discharge at the last follow up and one patient had electrolyte imbalance which was corrected before discharge.

Table 5. Post redo PT early complications

<b>Post redo PT early postoperative complications</b>		Frequency	Percent
Valid	Smooth course	15	83.3
	Rectal retraction	1	5.6
	Fistula	1	5.6
	Others	1	5.6
	Total	18	100.0

Five patients (27.8%) had a protective stoma done during the second PT procedure. The stoma was closed 8 months later in one patient & 12 months later in another patient. For the three patients, the time of closure wasn't documented.

### 5.3 Outcome

The median age at last follow up was 86.7 months with inter-quartile range of 70.5-108 months. Two patients were less than the age of 4 years (42 months each), of which one is toilet trained but one isn't trained yet.

Table 6. Age at last follow up

Age at last follow up( in months)		
N	Valid	18
	Missing	0
Mean		90.22
Median		86.50
Mode		108
Std. Deviation		37.151
Variance		1380.183
Range		138
Minimum		42
Maximum		180
Percentiles	25	70.50
	50	86.50
	75	108.00

Holshneider FII was obtained for 11 patients. Six patients have score of 10-14, which corresponds with good continence. Three patients have a score of 7, 8 & 9 with a fair continence, one patient has a score of 3, but he is 42 months old with recent introduction of solid meals & started toilet training. One has a score of 4 and has recently been started on bowel management program. On the remaining 7 patients FII couldn't be obtained, but documentation of their continence status was obtained in 6 patients. One patient is continent, 2 patients have night time soiling, 2 patients have night time & intermittent daytime soiling & the last one is incontinent & on daily enema.

## 6 Discussion

Hirschsprung's disease (HSD) is a developmental disorder of the intrinsic component of the enteric nervous system characterized by the absence of ganglion cells in the myenteric and submucosal plexuses of the distal intestine beginning at the internal anal sphincter and extending proximally for varying distances[1, 2]. The incidence of HSD is estimated to be 1 in 5,000 live births and ranges from 1 in 2,000 to 1 in 12,000 live births[1, 2]. Males are more commonly affected than females with a male to female ratio of 4:1. The surgical management for Hirschsprung disease is pull through procedure.

Although surgery is effective in most HSD patients, up to 32% of these patients continue to have symptoms and complaints. These persisting symptoms and complaints include fecal incontinence, constipation and persistent stooling problems, and Hirschsprung disease associated enterocolitis (HAEC). Additional laxative treatment is generally needed and a very small fraction of the patients with obstructive symptoms needs secondary surgery[1, 2, 4, 6].

### 6.1 patient profile before redo pull through

In our study a total of 18(4.87% of all PT) patients underwent redo pull through from 2010- 2019 in Tikur Anbessa hospital, Ethiopia. This was in the range of the rate reported by Ralls et al. & Jiang et.al where 3% & 8.6% had redo operation respectively.13 (72.2%) were males & 5 (27.8%) were females with male to female ratio of 2.6:1. The Male-to-Female ratio was similar to Pini-prato et al. study from Genoa, Italy where it was 2.47:1[8].The median age at the time of first pull through was 24 months with inter-quartile range of 21.25-33 months. The youngest child was aged 3 months and the eldest was 10.4 years old. Our patients had the first pull through at a later age compared to those from Wuhan, China reported by Jiang et.al who had the primary operation at an age range of 1 month to 6 years, with an average of 5 months[5]. Lawal et.al from USA also reported that median age at first PT was 5.5 months with a range of 5 days-2 years[6]. The age difference could be explained by the late presentation of our patients as the median age at initial presentation was 7 months with inter-quartile range of 10 days-24.5 months.

In our study, Soave pull through was the most frequent type of pull through on first time; done on 16(94.2%) of patients, with trans-abdominal mobilization of the colon required in 14 patients. In two patients primary trans-anal endorectal Soave pull through was done and Duhamel pull

through for one(5.9%) patient. Of the 19 redo PT Van Leeuwen et al. performed, the primary pull-through procedures included 10 endorectal pull-throughs (ERPT), 5 Duhamel procedures, 3 Swenson procedures, and 1 Rehbein procedure[10]. Pini-Prato et al performed 70 redo PT in their study. 36 patients had complete follow up. Of the 36, the failed pull-through was a Soave procedure (or its modifications) in 30 patients, a Duhamel (or its modifications) in 5, and a posterior sagittal approach (PSARP) in 1 patient[8]. Compared to these, our centre looks to have a tendency to do Soave PT more frequently.

In our study, the most frequent mode of presentation for redo PT was persistent obstructive features in 10(55.6%) patients. Four(22.2%) patients presented with persistent obstructive features and HAEC. One patient had anastomotic stricture, but he wasn't obstructed as he had a protective stoma. Early retraction of PT segment with cuff abscess occurred in 1(5.6%) patient, another one presented with fecal soiling, and for one patient redo PT was done for aganglionic PT segment though he wasn't symptomatic. This was similar to a study by Pini-Prato et al who analyzed results of 36 patients. Twenty-two patients (61%) complained of obstructive symptoms, 11 (30%) recurrent or severe acute HAEC, and 10 (28%) severe chronic constipation. Seven patients (19%) experienced both obstruction and enterocolitis [8]. Of the 16 patients for whom redo PT was done by Dingemans et al. 4 patients(25%) were fecally incontinent & 2 patients(13%) reported staining[4].

On physical exam evaluation at presentation 13 patients(72.2%) had abdominal distention, 6 (33.3%) had anastomotic site narrowing and in 5 (27.8%) patients there was positive blast sign on digital rectal exam. Anastomotic stricture dilatation under anesthesia was done for 6(33.3%) patients, two of whom had a protective stoma done at the time of dilatation & another one was already on colostomy; 11 (61.1%) of the patients eventually required diversion ileostomy/colostomy. Of the 16 patients for whom redo PT was done by Dingemans et al., in 14 patients (88%) other treatment interventions (anal dilation or botulinum toxin injection treatment) were performed before the redo surgery, but were not successful[4].

The indication for redo PT was pathologically confirmed persistent/acquired aganglionosis in 8(50%) patients. 5(27.8%) patients had anastomotic site stricture confirmed with examination under anesthesia. In three of the patients the redo surgery was done for persistent obstructive features & HAEC with the clinical impression of persistent/acquired aganglionosis, of which

barium enema was suggestive of HSD in one. One patient had twisted colon & anastomotic stricture & the last one was done for anastomotic failure with retraction of PT colon. Hassan et al. performed 33 redo PT procedures over 13 years. The indications were; obstructive symptoms due to retained or acquired aganglionic segment in 15 patients, severe anastomotic stricture in 7 patients & enterocolitis with or without constipation in 11 patients[12]. Pini-Prato et al performed 70 redo PT in their study. Indications to reoperation were residual aganglionosis or hypoganglionosis in pulled-through colon in 41 patients, retained aganglionic rectum (high anastomosis) in 10, cuff stricture in 5, anastomotic stricture in 3, persistent or recurrent enterocolitis in 2, leakage with subsequent perirectal abscess and fibrosis in 2, and twisted pulled-through bowel in 1. Indications could not be retrieved in 6 patients because of improper notes compilation[8]. In Dingemans et al. 16 redo PT the indication for redo TERPT was a suspected mechanical obstruction in or surrounding the distal colon and anal canal in all patients; in 7 patients pathological origin of the colon (transition zone or residual aganglionosis) was proven preoperatively[4].

In our study the median interval between the first & redo PT was 14.5 months with inter-quartile range of 6 months to 2.5 years. This was comparable to Lawal et.al study that showed interval between initial and redo operation from 12 months to 5 years (mean, 3 years)[5] & Lawal et.al study where 16 children presented at 6 to 66 months after initial pull-through.

## **6.2 Redo surgery**

In our study the median age at the time of the second pull through was 4 years with inter-quartile range of 2.8-6.1 years. The youngest was 10 months & the eldest was 14 years. This was comparable with Dingemans' et al. study from the Netherlands where age at the time of redo surgery was 4 years and 7 months (range 2 months to 21 years)[4] & Pini-prato et al. Study from Genoa, Italy with a mean age at redo of  $4 \pm 3.1$  years[8]. However, Wilcox et.al from London, England found that the mean age at repeat pull-through was 6 years (range, 1 to 13)[3].

The choice of a specific technique for redo pull-through is quite variable in various reported series[12]. In our study Swenson procedure was performed on 15 (83.3%) patients & Soave pull through was done on 3(16.7%) patients; 15 of them were done with abdominal mobilization of the colon & three were done transanally. This was comparable to a study by Lawal et al. From USA who analyzed the results of 16 patients for whom redo PT was done for histopathologic

indications only. The reoperation consisted of a transanal pull-through (n = 15) or a redo via a posterior sagittal approach (n = 1), preferred because of excessive pelvic scar tissue. Of the 15 who had a transanal pull-through, 10 had laparoscopy or laparotomy to complete the colonic mobilization. In 5 patients, the redo was done transanally alone [6]. Dingemans et al. From Netherlands also reported similar technique. Of 16 redo PT, the type of surgical technique for the redo procedure consisted of a Swenson-like TERPT in most cases (94%) [4]

Pini-Prato et al of Italy performed 70 redo PT in their study. Of the 36 patients who had complete follow up twenty-two patients underwent an endorectal pull-through (Soave and/or its modifications), 9 a Duhamel procedure, and 5 a Swenson. Mainly endorectal pull-through was used because of its extreme versatility. However, in some instances, other procedures were preferred because of impaired rectal blood supply, severe perirectal inflammation and fibrosis, and surgeon's judgement [8].

### **6.3 Early post redo operation course**

In our study, the postoperative course in the first month after surgery was smooth in 15 patients (83.3%). One patient had rectal retraction & reanastomosis was done. One patient developed perianal fistula which had whitish discharge at the last follow up and one patient had electrolyte imbalance which was corrected before discharge. Pini-Prato et al performed 70 redo PT in their study. 36 patients had complete follow up. Of the 36, twelve patients (33%) experienced postoperative complications, namely, adhesions and obstruction in 7, incisional hernia in 2, anastomotic stricture in 2, and enterocolitis in 1. Eight of these patients were managed conservatively, whereas 4 required some sort of surgical intervention [8]. Dingemans et al. performed 16 redo PTs. Following redo surgery, 5 patients (31%) reported 7 complications within 30 days after redo surgery: anastomotic dehiscence (3×), infection leading to abscess (2×), a rectovaginal fistula (1×) and an enterocutaneous fistula (1×). All these patients needed additional surgery because of these complications & eventually recovered completely from these complications [4].

### **6.4 Post redo operation outcome**

In our study, the median age at last follow up was 86.7 months with inter-quartile range of 70.5-108 months with median duration of follow up of 26.5 months & inter-quartile range of 16.5-50.25 months. Two patients were less than the age of 4 years (42 months each), of which one is



toilet trained but one isn't trained yet. Holshneider FII was obtained for 11 patients. Six patients have score of 10-14, which corresponds with good continence. Three patients have a score of 7, 8 & 9 with a fair continence, one patient has a score of 3, but he is 42 months old with recent introduction of solid meals & started toilet training. One has a score of 4 and has recently been started on bowel management program. On the remaining 7 patients FII couldn't be obtained, but documentation of their continence status was obtained in 6 patients. One patient is continent, 2 patients have night time soiling, 2 patients have night time & intermittent daytime soiling & the last one is incontinent & on daily enema.

In the Pini-Prato et al. study, 36 patients had complete follow up. Post redo follow-up was  $6.6 \pm 4.3$  years. Two patients younger than 4 years were excluded from fecal continence assessment. Excellent to good continence (wingspread  $\geq 2$ ) was described by 24 (70%) of 34 patients, fair continence by 6 (18%), and poor by 4 (12%). Average daily bowel movements at follow-up were  $2.4 \pm 2$ . 17% of patients required dietary limitation to contain symptoms. Frequent laxatives administration and/or repetitive cycles of decontamination were required in 11%. Chronic constipation in the long term was experienced by 14% of patients. 30% experienced recurrent episodes of enterocolitis and/or severe chronic perineal excoriations in the postoperative period; all except one patient settled with conservative treatment and one patient was still experiencing recurrent enterocolitis and excoriations 5 years after surgery. Significant continence improvement was experienced by 10 patients in the long term with mean age at symptoms resolution of  $8.5 \pm 3$  years [8]. In Schweizer et al study, Median follow-up after Redo operation was 9 years (range, 1-23 years). In 14 of 17 patients, the obstructive symptoms disappeared completely immediately after Redo Duhamel procedure and did not recur. These patients characterize their stooling pattern as completely normal. The outcome of two patients is characterized by grade 2 or 1 constipation, in whom the surgeon records the result as unsatisfactory, but the patients characterize their stooling pattern as satisfactory. In one patient, the outcome couldn't be judged as yet because the pull-through procedure was only 1 year back and colostomy has not been removed[11]. The outcome in 28 patients, from Hassan H et al of Egypt who were older than 3 years at time of last follow up visit are; normal or near normal bowel habits in 16 (57.1 %), recurrent constipation and obstructive symptoms in 5 (17.9%), and rank incontinence or repeated soiling in 7 (25%)[12].

## **7. Conclusion**

The indications for redo pull through was persistent obstructive features in 77.8% of patients. The causes of persistent obstructive symptoms after an initial pull-through procedure are pull through of aganglionated segment (50%) and anastomotic site stricture (27.8%). The preferred surgical technique used for redo pull through in patients who underwent redo pull through is Swenson pull through, which was done for 83.3% of patients. Long term outcome of patients after redo pull through was measured using Holschneider's FII score. Of the patients FII score was obtained (61.1%), good continence was achieved in 54.5%, fair continence in 27.2% & one patient (9.1% ) was incontinent & required bowel management program to stay clean. One patient was less than 4 years old & not toilet trained yet.

## **8 Recommendations**

We need to take full thickness rectal biopsy, & pathology report should include comments on the nerve trunks.

Reversed rectosigmoid ratio on barium enema can not be seen after pull through. We need to improve our radiologic understanding & reporting.

Patients for whom the proximal end pathology reported as aganglionated after PT were subjected to possibly avoidable procedures before the redo surgery. They should have the redo PT on the early postoperative period.

standardized fecal continence status measuring tool should be used in the follow up.

## **Ethical issue**

Ethical clearance was obtained from the research and publication committee of the department of surgery, Addis ababa university - school of medicine. Oral informed consent was taken from Parents/care givers of participants over telephone interviews.

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## Annexes

### Annex I: Questioner

#### I. Identification:

Name\_\_\_\_\_ MRN\_\_\_\_\_ Age  
\_\_\_\_\_ Sex\_\_\_\_\_ phone No\_\_\_\_\_

#### II. Initial presentation

1. Age at initial presentation\_\_\_\_\_
2. Symptoms/signs at initial presentation to our hospital
  - a. Neonatal lower intestinal obstruction
  - b. Chronic constipation
  - c. Hirschsprung disease associated enterocolitis
  - d. Referred after colostomy was done
3. Any associated anomaly identified
  - a. Congenital heart disease
  - b. Urinary tract anomaly
  - c. CNS anomaly
  - d. Down's syndrome
  - e. Other syndromic association
4. Diagnostic work-up at initial presentation
  - A. Barium enema
    - a. Clear transition zone
    - b. Reversed recto-sigmoid ratio
    - c. Delayed barium hold up
    - d. Saw tooth appearance of colonic mucosa
    - e. Distended colon
    - f. No features suggestive of HD
  - B. Rectal biopsy
    - a. Absent ganglion cells in submucosa &/or muscularis propria
    - b. Hypertrophic nerve fibers in submucosa &/or muscularis propria

- c. Few ganglion cells in submucosa &/or muscularis propria
- d. Normal appearing ganglion cells & nerve fibers in submucosa &/or muscularis propria
- e. Inconclusive finding or inadequate biopsy

C. Leveling colostomy & biopsy

- a. Transition zone identified during exploration
- b. Absent ganglion cells in submucosa &/or muscularis propria
- c. Hypertrophic nerve fibers in submucosa &/or muscularis propria
- d. Few ganglion cells in submucosa &/or muscularis propria
- e. Normal appearing ganglion cells & nerve fibers in submucosa &/or muscularis propria
- f. Inconclusive finding or inadequate biopsy

5. Management offered at initial presentation

- a. Home based rectal stimulation and irrigation
- b. Diversion colostomy
- c. Primary pull through

III. Initial pull-through

1. Age at first pull through\_\_\_\_\_
2. Type of pull through procedure
  - a. Primary transanal endorectal pull through
  - b. Soave pull through with abdominal mobilization
  - c. Swenson pull through with abdominal mobilization
  - d. Duhamel pull through with abdominal mobilization
3. Intraoperative findings
  - a. Distance of endorectal dissection from dentate line\_\_\_\_\_
  - b. Smooth endorectal dissection & mobilization
  - c. Difficult endorectal dissection with adhesion
  - d. Length of cuff\_\_\_\_\_
  - e. Transition zone & reverse transition identified
  - f. Length of the resected colon\_\_\_\_\_

- g. Segment of pulled through colon: 1) Rectosigmoid junction/distal sigmoid 2) proximal sigmoid/descending colon 3) splenic flexure 4) transverse colon 5) hepatic flexure/ascending colon
- h. Adequate bowel preparation
- i. Inadequate bowel preparation with contamination
- 4. Early postoperative complications
  - a) Smooth postoperative course
  - b) Rectal retraction
  - c) Cuff abscess
  - d) Intra-abdominal collection
  - e) HAEC
  - f) Fistula
  - g) Others\_\_\_\_\_

#### IV. Presentation for Redo pull through

- 1. Duration since first pull through\_\_\_\_\_
- 2. Main complaint
  - a. Persistent obstructive symptoms
  - b. Fecal incontinence
  - c. Recurrent HAEC
  - d. Failure to thrive
- 3. Physical examination finding
  - a. Distended abdomen
  - b. Visible peristalsis
  - c. Palpable fecaloma
  - d. Abdominal tenderness
  - e. Rectal anastomotic narrowing
  - f. Tight cuff
  - g. Rectum loaded with stool
  - h. Dilated and capacious rectum
  - i. Positive blast sign
  - j. Good anal sphincter tone

- k. Poor anal sphincter tone
    - l. Other
- 4. Diagnostic work-up/procedure
  - a. Examination under anesthesia
  - b. Barium enema
  - c. Rectal biopsy
  - d. Others
- 5. Management before redo pull through
  - a. Dietary modification
  - b. Laxatives
  - c. Antibiotics
  - d. Rectal/colonic irrigation
  - e. Bowel management program
  - f. Anal dilatation
  - g. Botulinum toxin injection
  - h. Sphincterotomy/ myomectomy
  - i. Other\_\_\_\_\_
- 6. Surgical approach for redo
  - a. Transanal endorectal soave pull through
  - b. Transanal endorectal Swenson pull through
  - c. Abdominoperineal approach
  - d. Other
- 7. Pathology of resected segment (proximal end)
  - a. Absent ganglion cells in submucosa &/or muscularis propria
  - b. Hypertrophic nerve fibers in submucosa &/or muscularis propria
  - c. Few ganglion cells in submucosa &/or muscularis propria
  - d. Normal appearing ganglion cells & nerve fibers in submucosa &/or muscularis propria
  - e. Inconclusive finding or inadequate biopsy

8. Was protective stoma done
  - a. Yes
  - b. No
9. Early postoperative complications
  - a) Smooth postoperative course
  - b) Rectal retraction
  - c) Cuff abscess
  - d) Intra-abdominal collection
  - e) HAEC
  - f) Fistula
  - g) Others\_\_\_\_\_

V. Follow up & Postredo outcome

1. Age at last follow up\_\_\_\_\_
2. Fecal continence status
  - a. Continent
  - b. Staining
  - c. Soiling
  - d. Incontinent
3. Degree of constipation
  - a. No constipation
  - b. Constipation treatable with dietary modification
  - c. Constipation requiring laxatives
  - d. Constipation requiring enema
4. Bowel movements per day\_\_\_\_\_
5. Post redo HAEC
  - a. Yes, if so how many attacks\_\_\_\_\_
  - b. No



## Annex II: Participant Consent Form

This consent form is to be filled by parents or legal guardians of the patients.

This patient information collection sheet is intended to assess the indications for reoperation, surgical technique adopted, complications, and outcome of patients who underwent a redo pull-through procedure at Tikur Anbessa Specialized Hospital (TASH), Addis Ababa, Ethiopia. The study will be conducted through reviewing secondary data and patient evaluation. The study will give some evidence and information for governmental and non-governmental organizations which work in the area pediatric colorectal surgical care at all levels (national, regional and district) level by providing basic information on the indications, surgical technique, complications, and outcome of patients. Information which is necessary for the study will be taken from patients chart, log books and patient evaluation. The individual patients will not be subjected to any harm, but can receive medical intervention of any sort intended only for patient care only; but these interventions may be used as an input for the study. The information extracted from secondary or primary source will be kept and remain confidential. Moreover, no personal identifiers will be used during result publication and dissemination.

Do you agree to participate in the study?

A. Yes, If so Name of parent/legal guardian.....Signature.....

B. No