## OUTCOMES OF PULL-THROUGH SURGERY FOR HIRSCHSPRUNG'S DISEASE AT MUHIMBILI NATIONAL HOSPITAL, TANZANIA:

### A COMPARATIVE STUDY BETWEEN REHBEIN'S AND CLASSIC SWENSON'S PROCEDURES.

TOM SAMORA EDWARD, MD

MMed (General Surgery) Dissertation Muhimbili University of Health Allied Sciences October 2021

#### MUHIMBILI UNIVERSITY OF HEALTH AND ALLIED SCIENCES

#### SCHOOL OF MEDICINE

#### DEPARTMENT OF SURGERY



#### **DISSERTATION RESEARCH PROPOSAL**

## TITLE: OUTCOMES OF PULL-THROUGH SURGERY FOR HIRSCHSPRUNG'S DISEASE AT MUHIMBILI NATIONAL HOSPITAL, TANZANIA.

# A COMPARATIVE STUDY BETWEEN REHBEIN'S AND SWENSON'S PROCEDURES

#### BY: TOM SAMORA EDWARD, MD REG. NO: HD/MUH/T.259/2018

A Dissertation Submitted in (Partial) Fulfilment of the Requirements for the Degree of Master of Medicine (General Surgery) of the Muhimbili University of Health and Allied Sciences

October 2021

#### CERTIFICATION

The undersigned certify they have read and hereby recommend for examination of a dissertation entitled: "Outcomes of pull-through surgery for Hirschsprung's disease at Muhimbili National Hospital, Tanzania: A comparative study between Rehbein's and Swenson's procedures" in fulfillment of the requirements for the degree of Master of Medicine (General Surgery) of the Muhimbili University of Health and Allied Sciences (MUHAS), Dar es Salaam, Tanzania.

## Dr. Ramadhani H. Khamisi, MD, MMed, CTVS-Fellow (Supervisor)

Date.....

Dr. Mohamed Salim, MD, MMed. (Co-Supervisor)

Date.....

#### **DECLARATION AND COPYRIGHT**

I, **Tom Samora Edward**, declare that this dissertation is my original work and that it has not been submitted to any other university for the award of any degree.

Signature.....

Date.....

This dissertation is copyright material protected under the Berne Convention, the Copyright Act 1999, and other international and national enactments, on that behalf, on intellectual property. It may not be reproduced by any means, in full or in part, except for short extracts in fair dealing, for research or private study, critical scholarly review or discourse with an acknowledgment, without written permission of the Directorate of Postgraduate Studies, on behalf of both the author and the Muhimbili University of Health and Allied Sciences.

#### ACKNOWLEDGEMENT

May Almighty God receive my heartfelt appreciation for his mercy, for granting me strength and health throughout the research period.

My heartfelt gratitude goes to my supervisors, Dr. Ramadhani Khamisi and Dr. Mohamed Salim, for their supervision, support, and unique heartfelt direction during my research effort.

Special thanks go to Prof. Obadia Nyongole, the head of the Department of General Surgery (MUHAS), for his contributions to the study's success.

Miss.Grace Tarimo, in particular, deserves special thanks for her technical assistance.

#### DEDICATION

I dedicate this work to my wonderful Grandmother Rose Ojuku, who has always been supportive of me and has given me courage during the research period.

#### ABSTRACT Background

The pull-through procedure is the definitive surgical management of Hirschsprung's disease. With the emergence of modern procedures, pull-through surgery has undergone significant changes. Despite single-stage transanal-endorectal pull-through (TERPT) indisputable success, there is yet no optimal multistage approach.

#### Objective

This study aimed to compare outcomes following multistage Rehbein's and Swenson's procedures among Hirschsprung's (HD) patients at MNH.

#### Methods

A retrospective cross-sectional study was conducted at Muhimbili National Hospital (MNH), Tanzania. The study involved 49 patients, aged <11years who underwent Swenson's and Rehbein's pull-through from January 2018 to July 2020. Data was retrieved from the patient's medical files and divided into two groups; Rehbeins and Swenson's. The comparison was done in terms of short and long-term complications, operative time, rate of additional operation, length of hospital stay, and rate of redo-pull through. Multivariate logistic regression was performed to determine statistical differences between the two groups.

#### Results

A total of 49 patients (male 36 and female 11) were included, with median ages at diagnosis and pull-through of 35 and 45 months, respectively. Thirty-one (63.3%) had Swenson's surgery, and 18 (36.7%) had Rehbein's procedure. Overall, there were 27/49 (55.1%) patients who had complications. In multivariate logistic regression, Rehbein's surgery had a larger proportion of total complications, 14/18 (77.8%) than Swenson's 13/31 (41.9%); (OR=6.5 (1.4-30.7); p=0.018). The Swenson's group had a significant large number of patients with Voluntary Bowel Movement (22/29 (75.8%) compared to the Rehbein's group's 5/15 (33.3%); p=0.019. Constipation was more common in Rehbein's group 8 (53.3%) than in the Swenson group 5 (17.3%), 6.3 (1.2-33.7); p=0.031. Rehbein's operation had a

greater rate of residual-aganglionic 4/15 (26.7%) than Swenson's 1/29 (3.4%), but was not significant in multivariate analysis p=0.080. Soiling and anastomotic stricture did not differ significantly. In Rehbein's procedure, 9/18 (50%) had more additional operations than Swenson's procedure on 6/31 (19.4%), (OR=4.9 (1.1-21.2); p=0.038). Redo pull-through was significantly higher in Rehbein's group than in Swenson's group (OR=7.1 (1.2-40.9); p=0.028). In terms of operating time, hospital stay length, and readmission rate, there was no difference. The mortality rate was 10.2% and was caused by sepsis and hemorrhage.

#### **Conclusion and Recommendation**

In terms of bowel functional outcome, fewer additional operations, and a reduced rate of redo pull-through, the Swenson procedure outperforms Rehbein's technique. The majority of Swenson's patients have a voluntary bowel movement, which is the primary goal of pull-through.

Sepsis is the major cause of post-pull-through mortality.

Even though these study findings favor Swenson's method over Rehbein's, a bigger prospective multicenter study is needed.

CERTIFICATION	i
ACKNOWLEDGEMENTii	i
DEDICATIONiv	/
ABSTRACT	/
TABLE OF CONTENTS	i
List of Tablesix	۲
List of Figuresix	۲
LIST OF ABBREVIATIONS	۲
DEFINITION OF TERMSx	i
CHAPTER ONE 1	1
1.0 Introduction1	1
1.2 Literature review	3
1.3 Problem statement6	3
1.4 Rationale of the study6	3
1.5 Research Hypothesis6	3
1.6 Objectives	7
1.6.1 Broad Objective	7
1.6.2 Specific Objectives	7
CHAPTER TWO	3
2.0 Methodology	3
2.1 Study design	3
2.2 Study population	3
2.3 Study area	3
2.4 Sample size estimation and selection	3
2.5 Data collection methods	9
2.6 Surgical Techniques:	9
2.7 Variables 10	)
2.8 Investigation tools validity and reliability10	)
2.9 Surgical Techniques:10	)
2.9 Data analysis11	

#### TABLE OF CONTENTS

2.10 Study limitations and mitigation	11
2.11 Ethical considerations	11
CHAPTER THREE	12
4.0 RESULTS	12
CHAPTER FOUR	18
4.0 DISCUSSION	18
CHAPTER FIVE	21
5.0 Conclusions and Recommendations	21
5.1 Conclusions	21
5.2 Recommendations	21
REFERENCES	22
APPENDICIES	26
Appendix 1: INVESTIGATION TOOLS	26

#### List of Tables

Table 1 Patient's characteristics distribution (N=49)	12
Table 2 Comparison of Short-term (<30 days) complications after Rehbein and	
Swenson's procedure	14
Table 3 Comparison in operative time, length of hospital stay, and rate of	15
Table 4 Functional outcome of Rehbein and Swenson's procedure	16
Table 5 The causes of post pull-through mortality	17

## List of Figures

#### LIST OF ABBREVIATIONS

HAEC	Hirschsprung associated enterocolitis
HD/HSCR	Hirschsprung's disese
L-HSCR	Long aganglionic segment
LOS	Length of hospital stay
MNH	Muhimbili National Hospital
S-HSCR	Short aganglionic segment
ТАВ	Transabdominal approach
TCA	Total colonic aganglionosis
TERPT	Transanal-endorectal approach
U-HSCR	Ultra short segment
VBM	Volunatary Bowel Movement

#### **DEFINITION OF TERMS**

- 1. Fecal incontinence (FI); the recurrent uncontrolled passage of fecal material at a person of at least 4 years old.
- 2. **Constipation;** the recurrent difficulty, infrequent or incomplete passing of stool for at least 3 months.
- 3. **Post-pull-through mortality;** the deaths associated with complications of pull-through surgery.

#### CHAPTER ONE

#### 1.0 Introduction

Hirschsprung's disease is a congenital colon motor dysfunction caused by neural crest cells failing to fully migrate to the distal intestine.(1,2,3) Hirschsprung's disease, often known as congenital megacolon, is characterized by an aganglionic colon that causes functional intestinal obstruction. (1,3) One out of every 5000 live newborns is affected by this disease. (1,3) At a 4:1 ratio, males are more affected than females.(1, 2, 3, 4)

The definitive treatment for HD is pull-through surgery. (1,3) Pull-through surgery aims to resect the aganglionic colon and anastomose a healthy ganglionic colon to the anus while maintaining anal sphincter function. (1,3) This can be done as a primary (one-stage) or multi-stage operation that takes 3 to 9 months after the colostomy has been leveled. (3,4,5,6) There are several multi-stage pull-through procedures. Swenson, Rehbein, Soave, and Duhamel are some of the most regularly used trans-abdominal multi-stage pull-through. (3,4,5) Classic Swenson's procedure involves the resection of the whole aganglionic colon followed by end-to-end colo-anal anastomosis. (7) The Rehbein's procedure is similar to low anterior resection, involving resection of the aganglionic colon, but anastomosis is done between the colon and the distal rectum. (8,9)

The primary (single-stage) Transanal-endorectal pull-through (TERPT) is done only on patients diagnosed early during the neonatal period and who have no complications. (3, 5, 10) Staged pull-through is performed on late-diagnosed patients with a substantially dilated proximal bowel where a primary anastomosis can not be done, as well as patients with enterocolitis, intestinal perforation, or malnutrition (3,5,10). Multistage Pull-through is still the most common approach in Sub-Saharan Africa due to late patient presentation. (5,11,12,13,14).

The patient's age, sex, comorbidities, concomitant additional congenital malformations, length of aganglionic bowel, and surgical technique influence the prognosis after pull-through surgery. (3,4,5)

The Transanal-endorectal pull-through (TERPT) provides the best outcomes compared to various multistage techniques.(6,10,15,16) However, no clear evidence that a particular multistage technique (Swenson's, Rehbein's, Duhamel's, or Soave's) is superior to another (11,12,13) As a result, the surgeon's decision to choose a particular technique is solely based on the surgeon's training, experience, and preference. (11,17)

This study compared the outcomes of Swenson and Rehbein's procedures in children with Hirschsprung's disease at Muhimbili National Hospital, Tanzania.

#### **1.2 Literature review**

#### Pull-through outcomes

The various pull-through procedures have been used with inconsistent results.(18,19) Apart from other factors that influence outcomes, the type of pull-through technique performed is a critical component in surgery success. (4,6,17) There is no optimal multistage technique.(17, 19) As a result, the choice of a multistage technique is largely determined by the surgeon's training, preferences, experience, and available resources.(11,17) The Swenson's and Rehbein's procedures are among the techniques done in staged pull-through. Swenson's procedure is one of the most commonly staged procedures in many centers, while Rehbein's has become infamous despite being the simplest procedure. (3,5,20).

#### Comparison of early complications of Rehbein's and Swenson's procedures

The common early pull-through complications include anastomotic leak, haemorrhage, sepsis, stricture, neorectal retraction, prolonged ileus, intestinal adhesive obstruction, surgical site infection and pelvic abscess.(3,5). Most of the studies found no significant difference in short term complications in various pull-through techniques. (20) Visser R. et al found common early complications after Rehbein to be anastomotic leakage, wound infection and sepsis. (20) Zganjer M. et al. reported the Rehbein procedure to have an excellent outcome in terms of fewer early complications. (9) A study done in Bangladesh found the classic Swenson procedure to have a reasonable good outcome with lower early complication rates. (21). Sowande A. and his collegue reported Swenson's procedure to be effective as definitive surgical management for Hirschsprung's disease.(14) In their study the commonest early complications were intestinal obstruction, anastomotic leak, and pelvic abscess, but only fewer patients had those complications. (14)

#### The difference in operative time, hospital length of stay and rate of Redo pullthrough in Rehbein's versus Swenson's procedure

Mahmud et al reported mean operating time and hospital stay time of 114.4 minutes and 6.18 days respectively in transabdominal Swenson's procedure (22). Those findings were better than in Rehbein's techniques documented by Visser R and his colleagues, where the mean operating time was 155 minutes and the length of hospital stay was 8 days. (20.)The rate of Redo-pullthrough after Rehbein's procedure is 8% (20)

#### Fuctional outcomes following Rehbein's and Swenson's procedure

The functional outcome is a reliable measure of the pull-through of long-term complications. (18, 23) The success of the definitive surgical management of HD is determined by whether the patient has a voluntary bowel movement or still has persistent bowel obstructive symptoms or fecal incontinence. (18,23). Long-term or functional complications are fecal incontinence and persistent constipation. (3,4,24)

The incidence of post-surgery constipation has been considerably higher in Rehbein's group than other pull-through types (5,20,25). In a multicentre study involving 200 patients from Germany, Switzerland, and Austria, they found a higher incidence of persistent obstructive symptoms in Rehbein's group than in other techniques, including Swenson. But the rate of soiling was higher in Swenson than in other techniques. (26) Visser R and his colleagues found that 80% of patients who underwent Rehbein's procedure had constipation. (20) The persistent obstructive symptoms in Rehbein's procedure have been associated with a residual aganglionic colon as the bowel is resected higher up in the rectum. (20,25). In a study involving 124 patients after Rebein's procedure, they found a constipation rate of 12.9%. (9) A study done in Nigeria to evaluate the outcomes of Swenson's procedure found a post-operative constipation rate of 3%. (14)

Anastomotic stricture is lower in the Rehbein procedure by 4%. (20) Zganjer M et al. found an anastomotic stricture rate of 2.4% following Rehbein's pull-through. (9) Sowande A. and colleagues discovered an anastomotic stricture in 3% of patients

who underwent Swenson procedure. (14) R. Rassouli et al. revealed an anastomotic stricture rate of 9.9% following Swenson's procedure. (26)

The rate of fecal incontinence is less in Rehbein's procedure compared to other techniques. (20) In one study of 124 cases post-Rehbein's procedure follow-up, the rate of soiling was 5.7% (9) Wester T et al reported a higher rate of 27% soiling after Rehbein's surgery. (25) A study done in Bangladesh revealed soiling in 6% of patients who underwent the Classic Swenson procedure. (22) Another study on Swenson procedure outcomes found a soiling rate of 9.1% (14)

#### The causes of post pull-through mortality

In developed countries, post-pull-through mortality is less than 2%. (1,27) In Sub-Saharan Africa, post-pull-through mortality is very high, reported to be 11.8% in one study and rising to 21.8% when colostomy-associated mortality is included. (11) The high mortality is seen more in patients presented late. (11,12) In Kenya, Ongeti et al. reported a 12.9% postoperative mortality rate, with all deaths occurring within 1 to 2 days following the pull-through. (12) Hirschsprung's associated enterocolitis remains the major cause of both preoperative and postoperative deaths. (3, 28, 29) Enterocolitis, sepsis, and metabolic abnormalities were documented as causes of postoperative mortality by Tander et al. (30) Pini and his colleagues found enterocolitis and congestive heart failure to be the causes of deaths post-pullthrough. (31)

#### 1.3 Problem statement

Most of pull-through surgeries performed in Sub-Saharan Africa are multi-stage procedures. (4,11-14)This is due to the late presentation of HD patients in this area, necessitating routine multi-stage pull-through while restricting the use of one-stage, transanal-endorectal pull-through as the optimal approach. (4,11-4). Aside from the late presentation's role in the poor outcome in Sub-Saharan Africa, the type of multistage procedure performed has a significant impact on the outcome. (11,12,14) However, the differences in outcomes across different multistage approaches are not well established. This necessitates a study of comparison of the outcomes, strengths, and failures of common multistage pull-through surgeries used in this locality.

#### 1.4 Rationale of the study

Type of pull-through technique is a modifiable predictor factor for surgery outcomes. This study will provide the strengths and failures of a specific multistage pull-through technique which form a basis for improvement of care in patient with Hirschsprung's disesase.

#### **1.5 Research Hypothesis**

#### The null hypothesis:

There is no difference in outcomes between HD patients undergoing Rehbein's and those undergoing Swenson's procedure at MNH.

#### 1.6 Objectives

#### 1.6.1 Broad Objective

To compare the outcomes of Rehbein's and Swenson's pull-through procedures in Hirschsprung's disease (HD) patients at MNH.

#### **1.6.2 Specific Objectives**

- To compare and contrast the early complications of Rehbein's and Swenson's procedures at MNH.
- 2. To compare and contrast the rate of redo-pull-through and the length of hospital stay after the Rehbein versus Swenson procedures at MNH.
- 3. To compare the bowel functional outcomes after Swenson and Rehbein procedures at MNH.
- 4. To detremine the causes of post pull-through mortality in Hirschsprung's disease (HD) patients at MNH.

#### CHAPTER TWO

#### 2.0 Methodology

#### 2.1 Study design

This study was hospital-based, retrospective cross-sectional.

#### 2.2 Study population

All HD patients attended at MNH from January 2018 to July 2020

#### 2.2.1Inclusion Criteria

- All histologically confirmed HD patients who underwent Rehbein's and Swenson's pull-through procedures from January 2018 to July 2020

#### 2.2.1 Exclusion criteria

- -HD patients not histologically confirmed
- -Patient's with missing required information from the medical files.

#### 2.3 Study area

This study was conducted at Muhimbili National Hospital (MNH), which is a National Referral and University teaching hospital located at Dare es salaam, Tanzania. MNH has a 1500 bed capacity and attending 2000 outpatients per day. The study was specifically done in the Pediatric Surgery unit.

The Pediatric surgery unit provides surgical services to children under the age of eleven years. But this excludes orthopedic conditions, neurosurgical, urological, and plastic conditions which are attended in their respective specialties. Hernias, anorectal malformations, Hirschsprung's disease, and pediatric tumors are the most common conditions managed in this unit.

#### 2.4 Sample size estimation and selection

The consecutive sampling technique was used. All histologically diagnosed HD patients who underwent pull-through at MNH from Jan 2018 to July 2020 informations retrieved from medical records. They were divided into two groups, Rehbein's and Swenson's group.

#### 2.5 Data collection methods

The structured checklist was used to extract data from medical records of all HD patients diagnosed histologically and who underwent pull-through surgery. The following variables were recorded: patient's demographic data (age and sex), histological results for rectal biopsy, age at diagnosis, age at pull-through, length of aganglionosis bowel, HD associated syndromes, HD associates enterocolitis, pull-through technique, Principle Srgeon, operation duration, documented post-operation complications, post-surgery hospital stay time, post-operation readmission, Cause of readmission, Re-do procedure and its indication, death and its cause, stooling pattern, Nutritional status, Immunosupression (HIV/AIDs).

The minimum follow-up was 6 months and maximum was 2 years and 6 months.

#### 2.6 Surgical Techniques:

All patients had colostomy before pull-through. Through the stoma, bowel irrigation with normal saline done for 3 to 5 days before surgery.

There were no criteria to opt for either Rehbein or Swenson. The pull-through was done mainly by three Pediatrics surgeons.

The Classic Swenson's technique was completed through abdominoperineal incisions, involved the resection of the aganglionic colon followed by coloanal anastomosis 1cm above the dentate line.(19) In Rehbein's procedure the only abdominal incision was made, the aganglionic colon was resected and colorectal anastomosis was done to the rectal stump.

No intraoperative frozen section biopsy was done, therefore the completeness of aganglionic segment resection based on intraoperative identification of the transition zone supplemented with preoperation radiological findings.

In 42 patients pull-through completed in two-stages and 7 patients underwent threestaged pull-through.

#### 2.7 Variables Dependent Variable:

- 1. **Shor-term complications;** postoperative enterocolitis, sepsis, surgical site infection, paralytic ileus, burst abdomen, perineal skin excoriation, wound dehiscence, metabolic abnormalities, hemorrhage, surgical site infection, anastomotic leak, early intestinal obstruction and mortality.
- 2. **Bowel functional outcome;** voluntary bowel movement, post-operation constipation and soiling.
- 3. **Other surgery outcome indicators;** operative time, duration of hospital stay, readmission rate, additional surgery and rate of redo-pull-through.

#### Independent Variables:

The independent variables were Rehbein's and Swenson's procedure.

#### 2.8 Investigation tools validity and reliability

The checklist for data collection was tested before starting data collection.

#### 2.9 Surgical Techniques:

All patients had colostomy before pull-through. Through the stoma, bowel irrigation with normal saline done for 3 to 5 days before surgery.

There were no criteria to opt for either Rehbein or Swenson. The pull-through was done mainly by three Pediatrics surgeons.

The Classic Swenson's technique was completed through abdominoperineal incisions, involved the resection of the aganglionic colon followed by coloanal anastomosis 1cm above the dentate line.(19) In Rehbein's procedure the only abdominal incision was made, the aganglionic colon was resected and colorectal anastomosis was done to the rectal stump.

No intraoperative frozen section biopsy was done, therefore the completeness of aganglionic segment resection based on intraoperative identification of the transition zone supplemented with preoperation radiological findings.

In 42 patients pull-through completed in two-stages and 7 patients underwent threestaged pull-through.

#### 2.9 Data analysis

The Categorical data were presented in frequencies or percentages while continuous variables were displayed as means or median. The Chi-square and Fisher's exact test were used for comparison of groups in categorical data and the t-test was used for comparison in continuous variables. The variables with a p value less than 0.20 in univariate analysis were subjected to multivariate logistic regression model to control confounders. Odds ratios with 95% confidence interval were reported. The p-value<0.05 was considered statistically significant. Data were analyzed using SPSS version 25.

#### 2.10 Study limitations and mitigation

 Incomplete documentation in the patients' case file, resulted in missing relevant of information.

#### **Mitigations**

- Patients with Incomplete case files documentation were excluded in the analysis of the particular missing data.

#### 2.11 Ethical considerations

The proposal was submitted to the Directorate of Research and Publication of MUHAS for ethical clearance approval.

Also, permission to research MNH was sought from the before the study was carried out.

The patient's information obtained was handled in great secrecy and care. The data were entered into a computer database by code to mask participant identity. Only investigators and authorized personnel got access to this information, to maintain confidentiality during the study.

The patient's particulars and medical information were not used for other purposes apart from this study. Confidentiality observed.

#### CHAPTER THREE

#### 4. 0 RESULTS

#### Table 1 Patient's characteristics distribution (N=49)

Characteristics		Rehbein n(%)	Swenson n(%)	p- value <sup>a</sup>
Sex	Male		22 (71)	0.743
	Female	4 (22)	9 (29)	
Mean Age at Diagnosis	s (months)	38.4	41.5	0.687
Mean Age at pull-through (months)		47.9	50.0	0.623
Aganglionosis length	Short segment Long segment Total colonic	10 (55.6) 6 (33.2) 1 (5.6)	19 (61.3) 9 (29) 1(3.2)	0.960
	aganglionosis Data Missing	1 (5.6)	2(6.5)	
Associated	Down's Syndrome	0	2	
congenital anomaly	Waardenburg Syndrome	0	1	
	Congenital heart disease	1	0	0.004
	Cryptochidism	1	0	0.221
	Posterior urethral valve	0	1	
Prepull-through HAEC		1	1	
Mean follow-up time (n	nonths)	18.1	21.2	0.555

<sup>a</sup>, t-test; HAEC, Hirschsprung Associated Enterocolitis

A total of 55 HD patients underwent Swenson's and Rehbein's surgery from January 2018 to July 2020. Forty-nine out of 55 met the inclusion criteria and were included in the study. Males were 36 (73.5%) and females were 13 (26.5%), with a male to female ratio of 2.8:1. Of the six excluded from the study, four had incomplete medical records, one was over the age of ten, and one still had a colostomy after three staged pull-throughs. The median age at diagnosis and at the time of the pull was 35 months and 45 months respectively. Swenson's procedure was performed on 31 (63.3%) while Rehbein's procedure was performed on 18 (36.7%). There were

no significant differences in the distribution of preoperative sociodemographic and clinicopathological characteristics between the two groups. There was no malnourished or immunocompromised patient at the time of definitive surgery. The distribution of patient characteristics across Swenson's and Rehbein's groups is shown in Table 1.

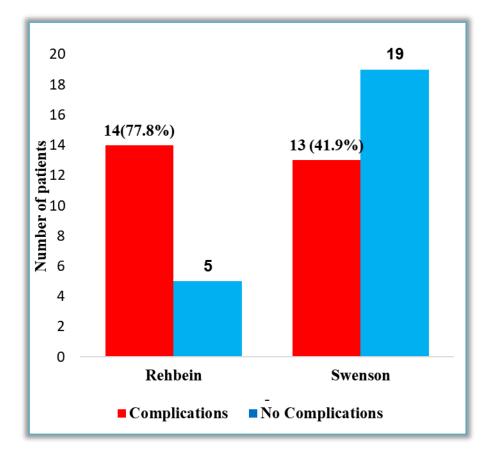


Figure 1 The overall pull-through complications

Patients with post-pull-through complications accounted for 27/49 (55.1%) of the total. In Multivariate logistic regression, Rehbein's method 14/18 (77.8%) had a considerably higher proportion of total complications than Swenson's group 13/31 (41.9%); (OR=6.5 (1.4-30.7) p=0.018) as illustrated in figure 1.

Complications	Rehbein N=18	Swenson N=31	Total (% of 49 cases)	p-value <sup>a</sup>
Postoperative enterocolitis	0	1(3.3)	1	1.000
Paralytic ileus	1(5.6)	0	1 (2)	0.367
Burst abdomen	1 (5.6)	1 (3.2)	2(4.2)	1.000
Perineal skin excoriation	0	2 (6.5)	2(4.2)	0.526
Wound dehiscence	2 (11.1)	0	2(4.2)	0.130
Metabolic abnormalities	2(11.1)	0	2 (4.2)	0.130
Haemorrhage	3(16.7)	2(6.5)	5(10.2)	0.342
Mortality	3 (16.7)	2( 6.5)	5 (10.2)	0.342
Surgical site infection	0	4(12.9)	4(8.2)	0.282
Sepsis	4(22.2)	2 (6.5)	6	0.175
Anastomotic leak	4(22.2)	3 (9.7)	7(14.7)	0.398
Early Intestinal Obstruction	1(5.6)	2 (6.5)	3(6.1)	1.000

Table 2 Comparison of Short-term (<30 days) complications after Rehbein and</th>Swenson's procedure

<sup>a</sup>,Fisher's exact test

The most common short-term complications were an anastomotic leak (14.7%), sepsis (12.2%), hemorrhage (10.2%), surgical site infection (8.2%), and mortality (10.2%). The short-term complications were not statistically different between Swenson's and Rehbein's groups (Table 2). The mortality rate was higher in

Rehbein's 3 (16.7%) than in Swenson's group 2 (6.5%), but was not statistically significant. The causes of the deaths were sepsis and hemorrhage.

# Table 3 Comparison in operative time, length of hospital stay, and rate ofredo pull-through after Rehbein's and Swenson's procedures

		Univariate analysis		Multivariate an	alysis
	Mean ± SD /N (%)	MD/COR (95% CI)	p-value	AOR (95% CI )	p-value
Operative time (mins)					
Swenson	190.6±42.8	1 (Reference)			
Rehbein	181.7± 41.8	9(-16.3 - 34.3)	0.479 <sup>*</sup>		
Length of Hospital stay (days)					
Swenson	8.9 ± 6.7	1 (Reference)			
Rehbein	10.1 ± 5.6	-1.2(-5.0 – 2.6)	0.927 <sup>*</sup>		
Additional operation					
Swenson	6/31(19.4)	1 (Reference)			
Rehbein	9/18(50)	4.2(1.2-14.0)	0.025	4.9(1.1-21.2)	0.038
Readmission rate					
Swenson	6/31(19.4)	1 (Reference)			
Rehbein	6/18(33.3)	0.4(0.1- 1.7)	0.223		
Redo pull-through					
Swenson	3/31(9.7)	1 (Reference)			
Rehbein	7/18(38.9)	5.9(1.3- 27.2)	0.025	7.1(1.2-40.9)	0.028

\*, t-test; COR,Crude Odds Ratio; AOR, adjusted Odds Ratio; MD, Mean Difference; SD, Standard Deviation.

In multivariate analysis, it was more likely to have additional operations following Rehbein's procedure on 9/18 (50%) than after Swenson's procedure on 6/31 (19.4%), a statistically significant (OR=4.9 (1.1-21.2), p=0.038). In Rehbein's group, Redo pull-through was significantly higher than in Swenson's group (OR=7.1 (1.2-40.9) p=0.028). The operation duration and postoperative length of hospital stay didn't differ significantly between the two groups.

		Univariate analysis		Multivariate analysis	
	N (%)	COR (95% CI)	p-value	AOR (95% CI )	p-value
Voluntary Bowel Movement					
Swenson Rehbein	22/29(75.8%) 5/15(33.3%)	1 (Reference) 0.2(0.04-0.6)	0.006	0.1(0.02-0.7)	0.019
<b>Constipation</b> Swenson Rehbein	5/29 (17.2) 8/15(53.3)	1(Reference) 5.5(1.4-22.2)	0.013	6.3(1.2-33.7)	0.031
<b>Soiling**</b> Swenson Rehbein	2/28(7.1) 2/15(13.3)	1(Reference) 0.4(0.1-3.6)	0.583		
Anastomotic stricture Swenson Rehbein	3/29(10.3) 2/15(13.3)	1(Reference) 0.9(0.1-5.7)	1.00		
<b>Residual aganglionosis</b> Swenson Rehbein	1/29(3.4) 4/15(26.7)	1(Reference) 10.2(1-101.5)	0.039	9.2(0.7-114.7)	0.085

#### Table 4 Functional outcome of Rehbein and Swenson's procedure

COR, Crude Odds Ratio; AOR, Adjusted Odds Ratio; VBM, Voluntary Bowel Movement; \*\*, assessed only to those aged ≥3years

Functional outcome was assessed in 44 patients, excluding deaths as they occurred within two weeks after definitive surgery. In Multivariate logistic regression, the Swenson's group had a significant large number of patients with Voluntary Bowel Movement (22/29(75.8%)) compared to Rehbein's group 5/15(33.3%); p=0.019. The Rehbein group had a higher rate of postoperative obstructive symptoms 8(53.3%) than Swenson's procedure 5(17.2%), a statistically significant (p=6.3 (1.2-33.7); p=0.031). The rate of residual-aganglionic was observed more in Rehbein's procedure 4/15 (26.7%) compared to Swenson's group 1/29 (3.4%), but this difference was statistically insignificant in multivariate analysis. There were no

differences in the rates of fecal incontinence and anastomotic stricture between the two groups (Table 4).

		Univariate analysis Multivariate analysis		Univariate analysis Multiva		alysis
Deaths Causes	Mortality N (%)	COR (95% CI)	p-value	AOR (95% CI )	p-value	
Sepsis	3(6.1%)	20(2.4-174.1)	0.010	13(0.6-270.4)	0.094	
Haemorrhage	2(4.1%)	9.1(1.1-77.4)	0.075			
Total Mortality	5/49(10.2%)					

#### Table 5 The causes of post pull-through mortality

COR, Crude Odds Ratio; AOR, Adjusted Odds Ratio

The post-pull-through deaths were caused by Sepsis 3 (6.1%) and intra-abdominal hemorrhage (4.1%) The causal relationship between sepsis and death was significant in univariate analysis (p=0.010) but not in multivariate analysis (p=0.094).

#### CHAPTER FOUR

#### 4.0 DISCUSSION

Swenson's technique is the most often employed for staged pull-through surgery in many centers, while Rehbein's has largely been abandoned. (3,5,11,25) Despite being the simplest technique, Rehbein's procedure is less done due to claims of a significant large aganglionic segment left in situ. (20,25,26) Our center is one of the few facilities that still does Rehbein's surgery. This study compared the outcomes of Rehbein's and Swenson's techniques.

This retrospective analysis has shown that Swenson's procedure produces better overall outcomes than Rehbein's technique. When compared to Swenson's group, Rehbein's group had 6 times higher odds of suffering postoperative complications. The significant complication rate in Rehbein's may be attributable to the aganglionic rectal stump left behind. These findings are comparable to the previous studies suggesting; increased post-surgery complications following Rehbein's than in other techniques (20,25).

Short-term complications in Rehbein's and Swenson's groups were not significantly different. This conclusion is not unique to Rehbein versus Swenson, but it's consistent with findings from other studies looking at early complications in various pull-through procedures. (20) The early pull-through complications are not specific to the type of technique. (1) This explains the similarity observed between these two techniques. As a result, perioperative factors such as postoperative care, rather than the pull-through method used, could play a role in the development of short-term problems.

After Rehbein's procedure, the risk of additional one or more procedures was 4.9 times higher than after Swenson's procedure. This greater rate of subsequent surgery following Rehbein's implies a high proportion of complications that necessitate surgical intervention.(20,25)

Rehbein's group had a larger number of patients with intra-abdominal hemorrhage than Swenson's procedure (16.7% versus 6.5%), but the difference was not

statistically significant. This discovery is disputed because of the level of pelvic dissection, which is lower in Rehbein's, implying a lesser likelihood of pelvic neurovascular injury.(9) The deviation from proper pelvic plane dissection of classic Rehbein's technique may have contributed to this finding.(1)

Rehbein's technique resulted in a significantly higher rate of Redo-pull through (38.9%), compared to 19.4% after Swenson's surgery. This observation is similar to the report of Visser R. and his colleagues on the comparison of Rehbein and transanal endorectal pull-through. (20) It is comparable to other studies suggesting very low redo pull-through after Swenson's procedure(19) Residual aganglionic colon, which is the absolute indication for Redo-pull-through, may have contributed to the pronounced Redo-pull through after Rehbein's. (25) Between Rehbein's and Swenson's procedures, there were no significant variations in mean operating time, length of hospital stay, or readmission rate.

Postoperative constipation was more common in Rehbein's procedures than in Swenson's (46.7% versus 14.3%), which was statistically significant. The probability of having postoperative persistent obstructive symptoms was 6.3 times higher in Rehbein's group than in Swenson's. Several earlier research has come to similar conclusions. (9,20,25,26) The amount of aganglionic colon that remains in place after Rehbein's operation could be a predictor of obstructive symptoms and other consequences as a squealer of blockage. (9,25)

Rehbein's group had a higher rate of soiling than Swenson's (14.4% versus 6.9%), but it wasn't statistically significant. These findings differ from those of a previous study by Visser R and colleagues, who found that the rate of soiling was lower in Rehbein. (20) Rassouli R et al discovered that following Rehbein, fecal incontinence was lower than with Swenson and other techniques. (26) The increased proportion of soiling after Rehbein's in this study could be due to encopresis. (3,32)

There was significant higher rate of Residual's agaglionic colon in Rehbein's group (22.2%) compared to Swenson's group (3.2%). The finding is comparable to other

studies (9,25,26) This higher rate of residual aganglionosis is the result of technique which is not radical hence leave behind large part of aganglionic colon. (25)

The postoperative deaths were caused by sepsis and hemorrhage. This association between post-pull-through sepsis and mortality is consistent with prior studies. (29, 30) In contrast to the previous reports, in this study there was no documentation of enterocolitis as the cause of death.(3, 28, 29)There might be under-diagnosis of Hirschsprung's associated enterocolitis (HAEC).An anastomotic leak was the source of sepsis in all three septic shock deaths. Hemorrhage was found to be the second leading cause of death after surgery in this study.No other study has mentioned bleeding as a cause of death after a pull-through.(28,29,30) One of the two patients who died of an intra-abdominal hemorrhage had entire colonic aganglionosis, which is associated with a high risk of death.The second patient who died of hemorrhagic shock was admitted directly from the operating room to the General ward.Both of the bleeding patients died within 24 hours after the surgery. Although it was beyond the scope of this study to assess postoperative care, based on those two hemorrhage cases, there is a sense of the quality of postoperative care to the contribution of immediate mortality after the pull-through.

#### CHAPTER FIVE

#### **5.0 Conclusions and Recommendations**

#### 5.1 Conclusions

There is no difference in short-term complications between Rehbein's and Swenson's procedures.

The Swenson procedure outperforms the Rehbein technique in terms of less additional operation and fewer redo pull-through.

Swenson's technique has better functional outcomes than Rehbein's procedure. The majority of Swenson's patients have a voluntary bowel movement, which is the primary goal of pull-through.

Sepsis is the major cause of post-pull-through mortality.

#### 5.2 Recommendations.

Even though this study favors Swenson's method over Rehbein's, a bigger prospective multicenter study is needed.

All post-pull-through patients should be admitted to ICU at least for the first 48 to 72 hours to ensure close monitoring and early detection of any immediate complications.

#### REFERENCES

- Puri P., Nakamura H. Epidemiology and Clinical Characteristics of Hirschsprung's Disease. In: Puri P. (eds) Hirschsprung's Disease and Allied Disorders. Springer, Cham. 2019.
- Miyahara K, Kato Y, Koga H, Dizon R, Lane GJ, Suzuki R, et al. Visualization of enteric neural crest cell migration in SOX10 transgenic mouse gut using time-lapse fluorescence imaging. J Pediatr Surg. 2011 Dec;46(12):2305–8.
- 3. Holcomb G, Murphy M, Ostlie D. Ashcraft's Pediatric Surgery 6th Edition.2014: 474–488 p.
- Moore SW. Hirschsprung disease: current perspectives. Open Access Surg. 2016;(May 2016):39.
- Emmanuel A. Ameh, Stephen W. Bickler, Kokila Lakhoo, Benedict C. Nwomeh DP. Paeds Surgery Part 1. Paediatr Surg A Compr Text- Vol 2. 2011;385–8.
- Sulkowski, Jason & Cooper, Jennifer & Congeni, Anthony & Pearson, Erik & Nwomeh, Benedict & Doolin, Edward & Blakely, Martin & Minneci, Peter & Deans, Katherine. Single-Stage Versus Multi-Stage Pull-Through for Hirschsprung's Disease: Practice Trends and Outcomes in Infants. Journal of Pediatric Surgery. 49. 10.1016/j.jpedsurg.2014.06.002.
- Nishijima E. Swenson's Procedure. In: Taguchi T., Matsufuji H., leiri S. (eds) Hirschsprung's Disease and the Allied Disorders. Springer, Singapore 2019.
- Holschneider A., Rassouli R. Rehbein's Procedure (Deep Anterior Resection).
  In: Holschneider A., Puri P. (eds) Hirschsprung's Disease and Allied Disorders. Springer, Berlin, Heidelberg(2008).

- Zganjer M, Cigit I, Car A, Visnjić S, Butković D. Hirschsprung's disease and Rehbein's procedure--our results in the last 30 years. Coll Antropol. 2006 Dec;30(4):905-7. PMID: 17243568.
- 10. Teitelbaum DH, Cilley RE, Sherman NJ, Bliss D, Uitvlugt ND, Renaud EJ, Kirstioglu I, Bengston T, Coran AG. A decade of experience with the primary pull-through for hirschsprung disease in the newborn period: a multicenter analysis of outcomes. Ann Surg. 2000 Sep;232(3):372-80.
- 11. Mabula JB, Kayange NM, Manyama M, Chandika AB, Rambau PF, Chalya PL. Hirschsprung's disease in children: A five year experience at a University teaching hospital in northwestern Tanzania. BMC Res Notes. 2014;7(1):1–9.
- 12. Ongeti K, Saidi H, Ogeng'O J, Tharao M. Experience with hirschsprung's disease at a tertiary hospital in Kenya. Ann African Surg. 2009;4(July):8–12.
- 13. Nasir AA, Adeniran JO and Abdur-Rahman L. Hirschsprung 's disease: 8 years experience in a Nigerian teaching hospital. Journal of Indian Association of Pediatric Surgeons 2007(ISSN:0971-9261) Vol 12.
- 14. Sowande OA, Adejuyigbe O. Ten-year experience with the Swenson procedure in Nigerian children with Hirschsprung's disease. Afr J Paediatr Surg 2011;8:44-8.
- 15. Yan BL, Bi LW, Yang QY, Wu XS, Cui HL, Amornyotin S. Transanal endorectal pull-through procedure versus transabdominal surgery for Hirschsprung disease: A systematic review and meta-analysis. Med (United States). 2019;98(32).
- 16. Somme S, Langer JC. Primary versus staged pull-through for the treatment of Hirschsprung disease. Semin Pediatr Surg. 2004 Nov;13(4):249-55.

- 17. Arts E, Botden SM, Lacher M, Sloots P, Stanton MP, Sugarman I, Wester T, de Blaauw I. Duhamel versus transanal endorectal pull through (TERPT) for the surgical treatment of Hirschsprung's disease. Tech Coloproctol. 2016 Oct;20(10):677-82.
- 18. Widyasari A, Pravitasari W, Dwihantoro A and Gunadi .Functional outcomes in Hirschsprung disease patients after transabdominal Soave and Duhamel procedures. Widyasari et al. BMC Gastroenterology (2018) 18:56.
- Mohajerzadeh L, Tabari A, Rouzrokh M, Mirshemirani A, Sadeghian N et al. Comparison Between Swenson and Soave Pull-Through in Hirschprung Disease. Ann Colorectal Res. 2015 December; 3(4): e32700.
- 20. Visser R, Van De Ven TJ, Van Rooij IALM, Wijnen RMH, De Blaauw I. Is the Rehbein procedure obsolete in the treatment of Hirschsprung's disease? Pediatr Surg Int. 2010;26(11):1117–20.
- 21. Mahmud K, Khan A, Yasmeen BN, Islam M, Karim S. Classic Swenson abdomino perineal pull through in the treatment of Hirschsprung's Disease - 3 years experience in a tertiary care Paediatric Hospital in Bangladesh. Northern Int. Med. Coll. J. [Internet]. 2017 Apr. 30;8(1):185-8.
- 22. Mahmud, Khalid, MK Islam, M Rashedul Alam, and Sabbir Karim. "A Comparative Study Between Swenson Abdominoperineal Pull-through and Transanal, Full Thickness, Swenson Like Approach for Hirschsprung Disease". Bangladesh Critical Care Journal 5, no. 1 (May 11, 2017): 23–27.
- 23. Meinds RJ, van der Steeg AFW, Sloots CEJ, Witvliet MJ, de Blaauw I, van Gemert WG, et al. Long-term functional outcomes and quality of life in patients with Hirschsprung's disease. Br J Surg. 2019 Mar;106(4):499–507.
- 24. Langer JC, Rollins MD, Levitt M, Gosain A, Torre L de la, Kapur RP, et al. Guidelines for the management of postoperative obstructive symptoms in children with Hirschsprung disease. Pediatr Surg Int. 2017 May;33(5):523–6.

- 25.T Wester , L Zetterlind, K Fredin, L Olsen. Postoperative obstructive symptoms are common after Rehbein's procedure for Hirschsprung's disease. Eur J Pediatr Surg. 2006 Apr;16(2):100-3.
- 26. R Rassouli , A M Holschneider, M Bolkenius, G Menardi, M R Becker et al. Long-term results of Rehbein's procedure: a retrospective study in Germanspeaking countries. Eur J Pediatr Surg. 2003 Jun;13(3):187-94.
- 27. Friedmacher F, Puri P. Hirschsprung's disease associated with down syndrome: a meta-analysis of incidence, functional outcomes and mortality. Pediatr Surg Int. 2013;29(9):937–46.
- 28. Gosain A, Frykman PK, Cowles RA, Horton J, Levitt M, Rothstein DH, et al. Guidelines for the diagnosis and management of Hirschsprung-associated enterocolitis. Pediatr Surg Int. 2017;33(5):517–21.
- 29. Wang, JS., Lee, HC., Huang, FY. et al. Unexpected mortality in pediatric patients with postoperative Hirschsprung's disease. Ped Surgery Int 20, 525–528 (2004).
- 30. Tander B, Rizalar R, Cihan AO, Ayyildiz SH, Ariturk E, Bernay F. Is there a hidden mortality after one-stage transanal endorectal pull-through for patients with Hirschsprung's disease? Pediatr Surg Int. 2007;23(1):81–6.
- 31. Pini Prato, A., Rossi, V., Avanzini, S. et al. Hirschsprung's disease: what about mortality?. Pediatr Surg Int 27, 473–478 (2011).
- 32. P. Saadai · A. F. Trappey1,2 · A. M. Goldstein3 · R. A. Cowles4 · L. De La Torre et al. Guidelines for the management of postoperative soiling in children with Hirschsprung disease. Pediatric Surgery International (2019) 35:829– 834.

#### **APPENDICIES**

#### Appendix 1: INVESTIGATION TOOLS Data Collection check list:

Check list serial number.....

#### PART A: Patient particulars

Patient's name: .....

Hospital reg. number: ..... Phone number.....

Sex.... (a) Male (b) Female

Age.....

#### PART B: Diagnosis

- 1. Age at Diagosis.....
- 2. Age at the pull-through procedure.....
- 3. Histology results

a) Hisrchsprung's disease diagnose (HP.....)

b)No HD

- 5. Classification based on any of Radiology intra-operative findings and/or frozen section results:
  - a) Short aganglionic segment (S-HSCR)
  - b) Long aganglionic segment (L-HSCR)
  - c) Total colonic aganglionosis (TCA)
  - d) Ultra short segment (U-HSCR)

5. Was the patient diagnosed to have Hirschsprung's associated enterocolitis before surgery

a)Yes

b)No

6. Is the patient has another associated congenital anomaly or syndrome

a)Yes

b)No

If yes, mention that congenital anomaly.....

#### PART C: Treatment

- 1. Pull-through surgery
  - (a) Date...../...../.....
  - (b) Pull-through stages used...... (i) One stage/ (ii) Multistage
  - (c) Technique.....
  - (d) Operating time (duration).....
- 2. The postoperative patient admitted at
  - a) ICU
  - b) General pediatric surgery ward
- 3.Early postoperative complications ... Yes/No.... if yes which complication:
  - a) Bleeding
  - b) anastomotic leak
  - c) surgical site infection
  - d) Sepsis
  - e) Wound dehiscence
  - f) Prolonged ileus
  - g) Electrolyte imbalance
  - h) Enterocolitis
  - i) Perineal excoriation
  - j) Others.....

- 4. Length of hospital stay post pull-through......days
- 5. Late complication
  - a)Obstructive symptoms (Persistent Constipation, abdominal distension, vomiting)
  - b)Fecal incontinence (soiling)
  - c) Enterocolitis
- 6. The cause of persistence obstructive symptoms post pull-through
  - a) Stricture
  - b) Residual aganglionosis colon,
  - c) motility disordereds in the residual colon or small bowel,
  - d) twist in the pulled through bowel
  - e) Internal sphincter achalasia
  - f) Not applicable
- 7. Post Pull-through Readmissions......yes/no

If yes, how many admissions ......Reason.....

8. Additional operation.....yes/no

If yes, indication.....

9. Redo pull-through.....yes/no

If yes, indication.....

10. Postoperative Death (operative mortality)

- a) Yes
- b) No

11. If yes, occured at which time after surgery......days

- 12. Cause of death
  - a)Enterocolitis
  - b) Sepsis
  - c) Shock (Haemorrhagic/neurogenic)
  - d) Metabolic abnormalities
  - e)Others.....