General Surgery

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Operative Outcomes and Associated Factors of Hirschsprung Disease Patients at Tibebe Ghion Specialized Hospital and Felege Hiwot Comprehensive Specialized HOSPITAL

Adane, Kassa

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BAHIR DAR UNIVERSITY COLLEGE OF MEDICINE AND HEALTH SCIENCES, SCHOOL OF MEDICINE, DEPARTMENTS OF SURGERY

OPERATIVE OUTCOMES AND ASSOCIATED FACTORS OF HIRSCHSPRUNG DISEASE PATIENTS AT TIBEBE GHION SPECIALIZED HOSPITAL AND FELEGE HIWOT COMPREHENSIVE SPECIALIZED HOSPITAL

A THESIS SUBMITTED TO BAHIR DAR UNIVERSITY, COLLEGE OF MEDICINE AND HEALTH SCIENCES, DEPARTMENTS OF SURGERY IN PARTIAL FULFILLMENT OF THE REQUIREMENT FOR SPECIALIZATION IN GENERAL SURGERY

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GENERAL SURGERY RESIDENT, BDU

NOVEMBER, 2015 E.C

BAHIR DAR, ETHIOPIA

OPERATIVE OUTCOMES AND ASSOCIATED FACTORS OF HIRSCHSPRUNG DISEASE PATIENTS AT FELEGE HIWOT COMPREHENCIVE SPECIALIZED HOSPITAL AND TIBEBE GHION SPECIALIZED HOSPITAL BAHIR DAR UNIVERSITY, COLLEGE OF MEDICINE AND HEALTH SCIENCES, SCHOOL OF MEDICINE, DEPARTMENTS OF GENERAL SURGERY

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NOVEMBER, 2015 E.C

Declaration

This is to certify that the thesis entitled "Operative outcomes and associated factors of hirschsprungs disease patients in Felege Hiwot Comprehensive Specialized Hospital from September 2007 to January, 2011 E.C and Tibebe Ghion specialized Hospital, Northwest Ethiopia from January, 2012 to April ,2014 E.C", submitted in partial fulfillment of the requirements for the degree of Master of sciences in Department of surgery, Bahir Dar University, is a record of original work carried out by me and has never been submitted to this or any other institution to get any other degree or certificates. The assistance and help I received during the course of this investigation have been duly acknowledged.

Dr. Adane Kassa

Name of the candidate

Signature

Date

Acknowledgement

First, I would like to take this opportunity to express my profound gratitude and deep regard to Bahir Dar university college of medicine and health science for giving me this opportunity to practice the development of health-related research and also to my advisors Dr. Nebiyu Shitaye and Mr. Taye Abuhaye for their exemplary guidance, valuable feedback and constant encouragement, constructive advice and support since the beginning to the end of proposal.

Secondly, I would like to thank all those whose assistance proved to be a milestone in the accomplishment of my end goals.

Approval of the advisors

I hereby certify that I have supervised, read, and evaluated this thesis entitled "Operative outcomes and associated factors of hirschsprungs disease patients in Tibebe Ghion specialized Hospital from January, 2011 to April ,2014 E.C and Felege Hiwot Comprehensive Specialized Hospital from September 2007 to January 2011 E.C" by Dr. Adane Kassa prepared under my guidance. I recommend the thesis to be submitted for oral defense.

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2. <u>Mr. Taye Abuhaye (Ass't professor of biostatistics)</u> Advisor's name Signature Date

ABSTRACT

Background: Hirschsprungs disease is one of the cause of intestinal obstruction in pediatric patients that requires surgical correction. However, the surgical management sometimes ends with unfavourable outcomes characterized by fatal and nonfatal postoperative complications.

Objectives: To identify operative outcomes and associated factors in Hirschsprungs disease patients at TGSH and FHCSH, Bahir Dar Ethiopia, 2015 E.C.

Method: Institution based cross-sectional study of all biopsy proven Hirschsprungs disease patients in TGSH admitted from January ,2011 E.C to April ,2014 E.C and FHCSH from September 2007 E.C to January 2011 E.C will be included. Patients who fulfill the inclusion criteria will be selected by systematic random sampling from registration logbook. The collected data will check for completeness. The collected data will be coded, entered into EPI data version 3.1 software, and exported to statistical product and service solutions (SPSS) version 25 software packages for further analysis. Descriptive statistics will be used to summarize the data in the form of frequency, mean, standard deviation (SD), and cross-tabulation. Binary logistic regression analyses will be carried out to identify the association between the outcome variable and independent variables. A p-value <0.05 will be considered statistically significant. Adjusted odds ratio (AOR) with 95% confidence intervals (CI) will be used. The results of the analysis will be presented in texts, tables, and figures.

Result: From 218 study participants (HSD patients),156(71.2%) were males and 62(28.8%) females. About 31.2% of HSD patients had unfavorable outcomes. Late presentation after one year of age (p=0.002, AOR=5.002, 95% CI [1.820, 13.750]), the presence of down syndrome (p=0.006, AOR=7.442, 95% CI [1.796, 30.839]), preoperative HAEC (p=0.044, AOR=2.532, 95% CI [1.025, 6.254]) and long segment HSD (p=0.026, AOR= 7.176, 95% CI [1.267, 40.638]) are significantly associated with post-operative unfavorable outcomes of HSD patients.

Conclusion: from this study majority of patients had favorable post-operative outcomes (68.8%). The most frequently observed post-operative complication from this study was constipation which occurs in 18(8.2%) patients after definitive operation.

Keyword: HSD, HAEC, outcome, down syndrome

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Acronyms and Abbreviations

AOR	Adjusted odds ratio
CI	confidence interval
DS	Down syndrome
ETB	Ethiopian birr
FHCSH	Felege Hiwot comprehensive specialized hospital
HAEC	Hirschsprungs associated enterocolitis
HSD	Hirschsprungs disease
NDS	Non down syndrome
QoL	Quality of life
SD	Standard deviation
SPSS	statistical product and service solutions
TGSH	Tibebe Ghion specialized Hospital
VBM	Voluntary bowel movement

1. INTRODUCTION

1.1 Background

Hirschsprungs disease (HSD) is defined as a functional intestinal obstruction that results from the congenital deficiency of the normal myenteric plexus parasympathetic ganglion cells in the distal portion of the large intestine due to a disruption of normal neural crest cell migration, proliferation, differentiation, survival and/or apoptosis(1).The clinical presentation of Hirschsprungs disease ranges from neonatal intestinal obstruction to chronic progressive constipation in older children. Approximately 80 percent of patients present in the first few months of life with difficult bowel movements, poor feeding, and progressive abdominal distention(2).

The diagnosis is mainly by radiographic studies, anorectal manometry and histological examination of rectal wall biopsies(3). The treatment of hirschsprungs disease is surgical. The goal of surgical management 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. The most commonly performed operations are the Swenson, Duhamel, and Soave procedures(4).

Complications occurring after the surgical repair of Hirschsprungs disease can be temporally categorized into early and late complications. However, there is significant overlap in regard to the time period during which these may occur(5).

Early postoperative complications appear to be consistent, despite varying ages of surgical correction. The are many postoperative complications of Hirschsprungs disease surgery including, anastomotic insufficiency(leak), rectal stenosis, prolonged ileus, intestinal adhesive obstruction, and neorectal retraction, wound or other septic complications may occur with intrapelvic or presacral abscesses related to anastomotic complications. Excessive fluid and electrolyte losses may result in metabolic derangements, particularly in long-segment disease and enterocolitis associated with Hirschsprungs disease. HAEC remains a significant cause of morbidity and mortality(6).

1.2 Statement of the problem

Hirschsprungs disease is a complex surgical problem that appears to have varied health and social outcomes with the age and neurodevelopmental state of patients. In general, long-term outcomes are thought to be good for the majority of patients despite recognized problems with constipation and/or fecal incontinence. However, there are no universally accepted pathways regarding post-operative bowel management programs nor clearly defined follow-up pathways making the current outcome measures difficult to interpret. HSD children have significant reductions in psychosocial QoL and functional outcomes. Psychosocial functioning was affected by increasing age, fecal incontinence, constipation, and dysfunctional elimination. Fecal incontinence also reduced physical functioning QoL in study at a large tertiary pediatric institution between 2004 and 2013 (7, 8).

Compared to the general population, adolescent and adult patients with HSD surgical history tend to have a higher prevalence of fecal incontinence and lower gastrointestinal-related quality of life(9). Enterocolitis and soiling/incontinence constituted the most frequent complications, whether early or late in the postoperative period. Once considered, surgical treatment has reduced the disease mortality to 3% in the developed countries(10). Early detection and definitive diagnosis within the neonatal period can reduce the complication rate, and lead to a better disease outcome. It also allows prompt surgical intervention using the one-stage trans anal endorectal pull-through procedure, thus avoiding the need for multi-stage surgeries and further laparotomies that carry a higher rate of postoperative morbidity. Large studies evaluating complications exclusively in children who underwent neonatal definitive pull-through procedures are scarce. The complications data are mostly extrapolations from studies evaluating definitive repair in infants and older children with HSD(11, 12).

However, we have lack of information about post-operative outcomes and associated factors of HSD patients in Ethiopia even in Africa. The relationship between post-operative short term outcomes and associated factors of HSD patients, like patterns of presentation and management has not been fully quantified and well described. The above facts clearly indicate that the need of studying on operative outcomes and associated factors in TGSH and FHCSH, Bahir Dar, Ethiopia.

1.3 Significance of the study

Despite the fact that Hirschsprungs disease in pediatrics is prevalent in our environment, little work has been done; studies in particular. There is also paucity of local data regarding operative outcomes and associated factors of patients with HSD.

Bahir Dar University will be benefited from this study and we can take it as a benchmark for undertaking preventive measure for operative outcomes of HSD patients. Being the first study in TGSH and FHCSH, the data will be used as a baseline for future studies to be done in this subject matter.

1.4 Objective

1.4.1 General objective

To identify operative outcomes (favorable Vs unfavorable) and associated factors of HSD patients at TGSH and FHCSH Bahir Dar Ethiopia, 2015 E.C.

1.4.2 Specific objectives

- > To describe operative outcomes of HSD patients (favorable or unfavorable)
- > To identify factor associated with operative outcomes of HSD patients

2. Literature review

2.1 Patterns of clinical presentation and operative outcomes of HSD patients

Study in Netherland were done to assess operative outcomes of HSD in terms of age at operation. During the study period 830 patient were reviewed. There was a small increase in the risk of a permanent stoma and a temporary stoma with increasing age at surgery, regardless of the length of the aganglionic segment and operation technique. Age at surgery was not associated with the probability and the severity of constipation and fecal incontinence in long term(13).

All patients with HSD who presented between January 1, 1995, and January 1, 2001, in Children's Hospital of Pittsburgh were retrospectively assessed. Patients were divided into two groups. Group I consisted of infants diagnosed before 30 days of life, and Group II consisted of patients diagnosed afterwards. The symptoms at presentation differed significantly between groups. For younger patients, the most common symptoms were failure to pass meconium within the first 24 h of life and abdominal distention, whereas constipation was the presenting symptom in 100% of older patients. Complications, including postoperative enterocolitis, occurred equally. The delayed diagnosis of HSD patients does not worsen outcomes of older children with HSD patients. This finding implies that these children have a milder form of the disease, perhaps because of adaptation to the aganglionic state(14). Eleven patients with HSD diagnosed after the age of 3 years from 1998 to 2011 in Norway were studied. Age at diagnosis was 3.0 to 9.6 years. Early postoperative complications, especially anastomotic leakage, occurred frequently in children with late-diagnosed HSD. The long-term functional results were comparable with those seen in children operated on as neonates(15). The risk of chronic constipation is greater in children who underwent pull-through surgery at an older age, but surgical method and sociodemographic characteristics were not associated with bowel dysfunction(16). Eighty-two patients were analyzed in United States to compare post-operative outcomes for the neonatal (<31 days) and delayed (≥31 days) pull-through. Forty three were operated upon in the neonatal period and 33 in a delayed fashion. 18 of 49 patients (36.7%) operated in the neonatal period and 16 of 33 (48.5%) operated beyond the neonatal period had at least one postoperative episode

of enterocolitis. 15 of 49 patients (30.6%) in the neonatal period were receiving bowel management for fecal incontinence compared to 5 of 33 (15.2%) operated beyond the neonatal periods(17).

At medical center in Taiwan, 51 cases of neonates with HD between 2002 and 2009 were collected and studied. Patients were divided into two groups based on the time of initial diagnosis: Group I, diagnosis made within 1 week after birth, and Group II after 1 week. There were 25 patients in Group I and 19 in Group II. Patients with preoperative HAEC were more likely to develop adhesive bowel obstruction after operation (33% vs. 3%) and failure to thrive (33% vs.3%). Also, patients with long-segment or total colonic aganglionosis were at risk of developing both postoperative HAEC (85% vs. 29%) and failure to thrive(18).

Between March 1987 and August 2008, in Netherlands' 149 children were operated for Hirschsprung's disease. 20 children of this group were additionally diagnosed with Down syndrome. All children underwent either an open or a laparoscopic Duhamel procedure. Postoperative leak occurred significantly more often in children with Down syndrome (25%) compared to non-down syndrome (NDS) children (0.7%). Postoperative leakage-related abscess formation was higher in the Down syndrome (DS) patients (15%) compared to the non-down patients (0%). Severe constipation was present significantly more often in down children (55%) compared to non-down children (22.3%). There was no difference in incontinence between down and non-down children. Enterocolitis occurred more frequently in down patients after operation (45% vs. 31%)(19). A systematic literature-based search for relevant cohorts was conducted using multiple online databases to compare post-operative outcome of HSD patient with and without associated down syndromes. Sixty-one articles met defined inclusion criteria. Postoperative complications such as recurrent enterocolitis and soiling were significantly more frequent in HSD patients with coexisting Down syndrome. Although not statically significant fecal incontinence and persistent constipation occurred more often after surgical treatment of HSD patients with DS. The mortality was significantly higher in HSD patients associated with down syndrome(20).

A total of 110 HSD patients (M: F ratio= 3.6:1) with a median age of 24 months were studied in northwestern Tanzania. Sixty-four (58.2%) patients had complete intestinal obstruction whereas 38.2% and 3.6% patients had chronic intestinal obstruction and intestinal perforation respectively. No patient had enterocolitis. Constipation (94.5%) was the most common

complaints. About 99.1% patients had colostomy prior to the definitive pull-through. The majority of patients (67.3%) had short segment of aganglionosis. The definitive pull-through was performed in 94 (85.5%) patients (Swenson's pull-through (80.9%), Duhamel's pull-through (12.8%) and Soave's pull-through (4.3%) patients). Postoperative complication rate was 47.3%. During the follow-up period, the results of Swenson's and Duhamel's pull through procedures were generally good in 87.8% and 42.9% of patients respectively. The result of Soave's procedures was generally poor(21).

2.2 Patterns of management and operative outcomes of HSD patients

In Ireland hospital 259 consecutive patients with a confirmed histological diagnosis of HSD during 1975–2003 were examined. Of the study subjects, 77.2% were males and 22.8% females. Intestinal obstruction was the presenting feature in 56.8%, intestinal perforation in 1.9%, and constipation in 29.7%. Down's syndrome occurs in 15.1%. About 80.7% of the patient had recto sigmoid disease, 12% had long segment HSD and 16.6% had preoperative enterocolitis. Various pull through procedures were performed in these patients. Postoperative complications including soiling 10.3% and 21.7% had constipation requiring laxatives or enemas were occurred. There was no difference in bowel function in relation to type of pull through operation. Only 34% of patients with Down's syndrome had normal continence. Patients with HAEC had significantly poorer long-term bowel function as compared to those that did not have HAEC like that of long segment HSD(22).

Study at Armed Forces Hospital Riyadh between 1991 and 2000 were reviewed. The patients consisted of 33 boys (85%) and six girls (15%). Twenty-five patients (64%) underwent Soave's pull-through, and 13 patients (33%) underwent Duhamel's pull-through. Twenty children (80%) out of the 25 undergoing Soave's pull-through recovered uneventfully, compared with 11 out of the 13 (84%) undergoing Duhamel's pull-through. The complications following Soave's procedure included strictures in two patients (8%), enterocolitis in another two (8%), and

anastomotic leakage in one (4%). The complications following Duhamel's procedure included stricture in one patient (7.6%) and enterocolitis in another (7.6%). The rate of constipation was 16% after the Soave's pull-through compared with 15% after the Duhamel's pull-through(23). Patients who underwent Soave and Duhamel pull-through at Dr. Sardjito Hospital, Indonesia from 2013 to 2016 were studied to assess post-operative outcomes between the two procedures. Fifty-three patients were included in the study (45 males and 8 females. Ninety-three and 88% patients had a VBM following Duhamel and Soave pull-through, respectively. Constipation frequency was significantly higher in Soave than Duhamel groups (24% vs. 4%). whereas soiling rate was similar between Duhamel (21%) and Soave (8%) groups. The risk of constipation was increased ~ 21.7-fold in female patients after Soave procedure and was almost statistically significant. Age at operation were divided into two groups (< 3 years and >3 years). The operative outcome does not affected by the age at operation in both soave (p=0.21) and Duhamel(p=0.6) procedures(24).

HSD patients who underwent the Soave and Duhamel pull-through at Dr. Sardjito Hospital, Indonesia from 2010 to 2015 studied. One hundred patients were involved (Soave: 52 males and 19 females vs. Duhamel: 23 males and 6 females). The HAEC frequency after pull-through was significantly higher in the Duhamel than the Soave group (28% vs. 10%, respectively, p = 0.03). Pre-operative enterocolitis showed a significant association with HAEC following pull-through and the risk of HAEC after pull-through was increased in long-segment aganglionosis compared to short-segment(25).

Comparative study was done in groups of patients with biopsy-proven Hirschsprung's disease at the University of Iowa Medical Center by two different definitive procedures (soave vs Duhamel). All patients in both treatment groups had preliminary colostomies as the first stage of their surgical management. All of the late postoperative complications, like constipation, incontinence, occurred after the modified Duhamel operation, the most common of which was recurrent enterocolitis(26).

2.3 Conceptual framework

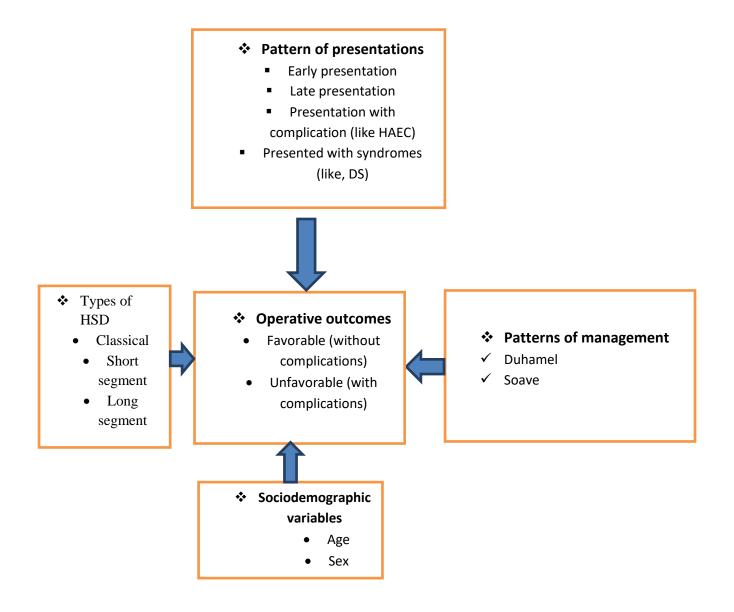


Figure 1: Conceptual framework showing the relationship between operative outcomes and associated factors of HSD patients(14, 18).

3. Methods and Materials

3.1 Study design

An institution-based cross sectional retrospective study will be conducted

3.2 Study period

The study period was from October 17,2015 to October 30,2015 E.C

3.3 Study area

The study was conducted in Tibebe Ghion Specialize Hospital (TGSH), Bahir Dar, Ethiopia. It is located 563 KMs far from Addis Ababa. Tibebe Ghion Specialize Hospital (TGSH) is found about 10km from Bahir Dar city to south direction on the way to Adet District and started to function on January, 2011 E.C. It gives surgical services including for those HSD patients since 2011. TGSH currently runs residency programs in ten specialties and three sub specialties. The hospital provides a tertiary level health care service and it is administered by Bahir Dar University. The hospital has more than 500 beds, all over around 104 beds for surgery in general and 8 beds only given for pediatric surgery.

Felege hiwot Comprehensive Specialize Hospital (FHCSH) is one of the main referral hospital in Amhara region and found in Bahir Dar city kebele 13 along the shore of Lake Tana. The hospital has more than 400 beds and around 103 beds for surgery in general. It has 15 outpatient departments. There are only 5 beds given for pediatric surgery.

3.4 Source of population

All HSD patients at TGSH and FHCSH were source of population

3.5 Study population

All HSD patients admitted at FHCSH from September 2007 to 2011 E.C and TGSH from January ,2011 to April ,2014 E.C.

3.6 Sample population

Biopsy proven HSD patients undergoing definitive surgery at FHCSH from September 2007 to January 2011 E.C and TGSH from January ,2011 to April, 2014 E.C.

3.7 Sample size determination and Sampling procedure

The sample size was determined using confidence interval approach

$$n = \frac{z^2(pq)}{e^2}$$

n= sample size

z=standard normal distribution (typically 1.96)

• p= proportion of HSD patients, it is taken as 50% since there is no similar study published in our country.

q= 1-p

e= acceptable sample or marginal error= 0.05

• There for the sample size is $n = \frac{(1.96)2 (0.5 \times 0.5)}{(0.05)2} = 384$

- According to this the sample size is 384 patients
- The total source population during the study period is estimated 500. The correction formula depicted below was applied.
- nf = n/(1 + n/N) = 384/(1 + 384/500) = 218
- Therefore, systematic random sampling was used and sample size was 218.

3.8 Inclusion criteria and Exclusion criteria

3.8.1 Inclusion criteria

Preoperative biopsy proven HSD patients under the age of 18 years and underwent definitive operation

3.8.2 Exclusion criteria

- Patients with inconclusive preoperative biopsy results
- Patients who lost from regular follow up
- Patients with incomplete medical record
- Total colonic HSD

3.9 Variables

3.9.1 Independent variable

***** Sociodemographic related variables

- o Sex
- o Age

* Pattern of presentations related variables

- ✓ Early presentation
- ✓ Late presentation
- \checkmark Presentation with complications
 - HAEC
- ✓ Presentation with syndromes
 - Dawn syndrome

* Types of HSD

- Classic HSD (recto sigmoid HSD)
- Short segment HSD
- Long segment HSD

✤ Pattern of management related variables

- o Duhamel
- o Soave

3.9.2 Dependent variable

- Operative outcomes of HSD patients (favorable or unfavorable)
- Favorable outcomes (patients without post-operative complications)
- Unfavorable outcomes (patients with complications like, HAEC, stricture, wound infection......death)

3.10 Operational and term definitions

Pull-through Procedure is a technique 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.

Soave procedure is a procedure by doing a sub mucosal endorectal dissection and placing the pull-through bowel within a "cuff" consisting of aganglionic muscle.

Duhamel procedure involves bringing the normal colon down through the bloodless plane between the rectum and the sacrum and joining the two walls to create a new lumen, which was aganglionic anteriorly and normally innervated posteriorly.

Incomplete data means patients without clear written evaluation of their progress during inpatient and outpatient follow-up period.

Short segment HSD means absence of ganglion cells mostly in rectum

Long segment HSD means transition zone proximal to mid transverse colon

Total colonic HSD means all the colon even distal ileum is agangleonosis

Classic HSD (recto sigmoid) means transition zone is in recto sigmoid area

3.11 Data collection instruments

Pretested checklist or formats prepared in English, adapted from similar pieces of literature was used as a data collection instrument to extract the data from the charts(7, 14, 19, 24). Data was collected by retrospective chart review by trained two data collectors. During data collection, one medical record officer and one nurse diploma holders working at Adissalem Hospital were involved. Using the medical record number (MRN) of patients, data collectors were traced and collect data from the charts using a checklist.

Supervision of the data collection process was made by the principal investigator. The supervisor was cross check for completeness and consistency of collected data daily

3.12 Data processing and analysis

The collected data was coded, entered into EPI data version 3.1 software, and exported to SPSS version 25 software packages for further analysis. Descriptive statistics was used to summarize the data in the form of frequency, mean, standard deviation (SD), and cross-tabulation. Binary logistic regression analyses were carried out to identify the association between the outcome variable and independent variables. Variables with P-value <0.25 in bi-variable logistic regression was used for multivariable logistic regression. A p-value <0.05 was considered statistically significant. Adjusted odds ratio (AOR) with 95% confidence intervals (CI) was used. Model fitness tests will be checked to assess whether the necessary assumptions are fulfilled. The results of the analysis were presented in texts, tables, and figures.

3.13 Quality assurance

Before the actual data collection, data collectors and supervisors was trained on the contents of the checklist, data collection methods, and ethical concerns. Therefore, data collectors become familiar with the checklist.

Five percent of the sample was pre-tested and the checklist was modified and edited based on the findings. During data collection, both supervisors and the principal investigator were checked the data for its completeness and missing information at each point. Furthermore, data was checked during entry and compilation before analysis.

3.14 Ethical considerations

For this study to proceed, letter of permission or ethical clearance was obtained from research ethics committee of BDU collage of medicine and health science, TGSH and FHCSH prior to study conduction. Though it was not possible to obtain participants informed consent, Names and other personal information which can violate the confidentiality of the study participants was not exposed to third party for any other reason. Any information was kept confidential and only used for research purpose.

3.14 Dissemination of the study

After the data is analyzed, the results of the research will be presented to Bahir Dar University College of medicine and health sciences, department of surgery. The findings will be published in a relevant scientific journal and disseminated online so that they can be of use for other academic researchers and clinical practitioners. It will also be presented on different conferences, and professional society meetings like Ethiopian Society of surgery. The data can also serve as a base line for future studies.

4. RESULT

4.1 Clinical presentation of HSD patients

During the eight-year study period, there were total of 500 HSD patients' admitted in TGSH and FHCSH surgical ward. From the study subject of 218 patients,167(76.3%) patients had repeated vomiting,213(97.3%) had abdominal distension, 180(82.2%) had failure to pass meconium within 24 to 48 hours (after delivery),10(4,6%) patients had down syndrome,25(11.4%) had preoperative HAEC at presentation (see table 1).

Table 1: Clinical presentation of HSD patients from September,2007 to January, 2011E.C atFHCSH and from January ,2011 to April 2014 E.C at TGSH, Bahir Dar, Ethiopia, (N=218).

Clinical presentation	Frequency	Percent(%)
Vomiting	167	76.3
Abdominal distension	213	97.3
Failure to pass meconium	180	82.2
Down syndrome	10	4.6
Preoperative HAEC	25	11.4

4.2 Socio-demographic characteristics of study participants

From 218 study participants (HSD patients),156(71.2%) were males and 62(28.8%) females. Age at presentations was varies among patients ,133(60.7%) patients were less than one month,62(28.8%) were age from one month to twelve months and 23(10.5%) of patients were age greater than one year.

Like age at presentation, age at the time of operation varies among the study subjects, 198(90.8%) were age less than three years and 20(9.2%) were age greater than three years (see table 2).

Table 2: Socio-demographic characteristics of HSD patients from September,2007 to January, 2011 E.C at FHCSH and from January ,2011 to April 2014 E.C at TGSH, Bahir Dar, Ethiopia, (N=218).

Variables	Category	Frequency	Percent(%)
Sex	Male	156	71.2
	Female	62	28.8
Age at presentation	Less than one month	133	60.7
	One month to twelve months	62	28.8
	Greater than one year	23	10.5
Age at operation	Less than three years	198	90.8
	Greater than three years	20	9.2

4.3 Management of HSD patients

From all 218 cases of HSD patients,189(86.7%) were underwent soave pull through,29(13.3) were underwent Duhamel pull through and colostomy was done for all patients (100%) before the two procedures. There was no any patient underwent primary pull through from this study. Intraoperatively,170(78%) patients were having classical (recto sigmoid) HSD,41(18.8%) short segment HSD and 7(3.2%) long segment HSD (see table 3).

Table 3:Type of management and intraoperative finding of HSD patients from September,2007 to January, 2011 E.C at FHCSH and from January ,2011 to April 2014 E.C at TGSH, Bahir Dar, Ethiopia, (N=218).

Variables	Category	Frequency	Percent(%)
Types of operation	Soave pull through	189	86.7
	Duhamel pull through	29	13.3
	Colostomy	218	100
Types of HSD segment	Classical(recto	170	78

sigmoid)		
Short segment	41	18.8
Long segment	7	3.2

4.4 Management outcomes

Among 218 HSD patients,150(68.8%) were having favorable outcome and 68(31.2%) unfavorable outcome.

The most frequently observed post-operative general complication was constipation which occurs in 18(8.2%) patients after definitive operation. The second most common complication was incontinence which occurs in 14(6.4%) patients. Other general complications like, wound infection 12(5.5%), anastomosis leak 2(0.9%), incisional hernia 6(2.7%), anal stenosis 6(2.7%) occurs after pull through procedure.

The most common colostomy related complication was colostomy necrosis which occurs in 6(2.7%) patients. The second common complication was colostomy retraction which occurs in 4(1.8%) patients. Colostomy prolapse occurs in 3(1.4%) and colostomy stenosis in 1(0.5%) of patients.

Table 4: Operative outcomes of HSD patients from September,2007 to January, 2011 E.C atFHCSH and from January ,2011 to April 2014 E.C at TGSH, Bahir Dar, Ethiopia, (N=218).

Operative outcomes	Frequency	Percent (%)
Favorable	150	68.8
Unfavorable	68	31.2

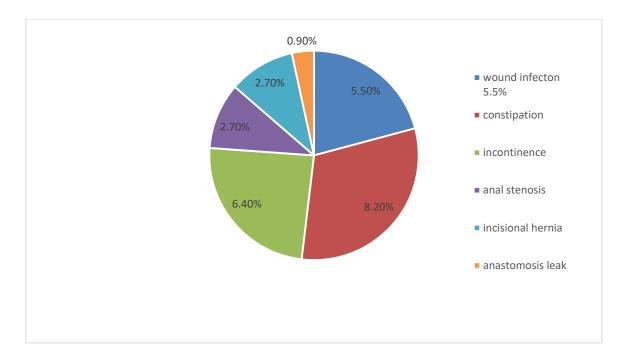


Figure 2: Pie chart of general post-operative complication of HSD patients from September,2007 to January, 2011 E.C at FHCSH and from January ,2011 to April 2014 E.C at TGSH, Bahir Dar, Ethiopia, (N=218).

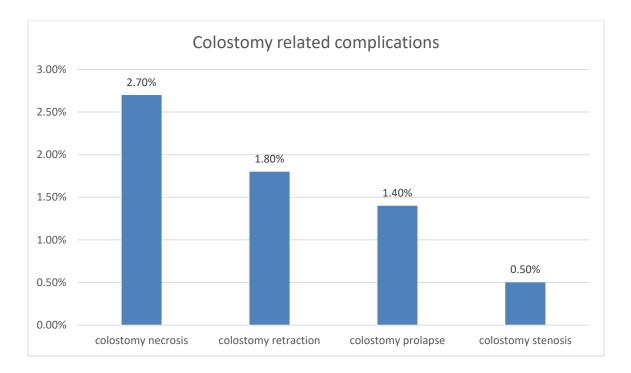


Figure 3: Colostomy related complication of HSD patients from September,2007 to January, 2011 E.C at FHCSH and from January ,2011 to April 2014 E.C at TGSH, Bahir Dar, Ethiopia, (N=218).

4.5 Factors associated with operative outcome of HSD

Bivariate and multivariable logistic regression analysis was used to calculate odds ratios and corresponding 95% confidence intervals for associated factors of operative outcomes of HSD patients in TGSH and FHCSH. From the independent variables, age at presentation, preoperative HAEC, associated down syndrome and long segment HSD are significantly associated with operative outcomes of HSD patient. After checking for the presence of association (p<0.25) in binary logistic regression, these variables were re-entered into multivariate logistic regression to assess the strength of association between these variables and operative outcomes of HSD by controlling the confounding effect of other variables. Late presentation after one year was significantly associated with post-operative complications (unfavorable outcome) (p=0.002) and adjusted odds ratio(AOR) of 5.002 and 95% CI [1.820, 13.750]. The presence of down syndrome also significantly associated with unfavorable outcome(p=0.006), AOR of 7.442 and 95% CI [1.796, 30.839]. Preoperative HAEC also significantly associated with unfavorable outcome (p=0.044), AOR of 2.532 and 95% CI [1.025, 6.254]. long segment HSD also significantly

associated with unfavorable outcome of HSD (p=0.026), AOR of 7.176 and 95% CI [1.267, 40.638] (see table 5)

Table 5: factor affecting operative outcomes of HSD patients from September,2007 to January,2011 E.C at FHCSH and from January ,2011 to April 2014 E.C at TGSH, Bahir Dar, Ethiopia,(N=218).

Variable	Category	Outcomes		COR(95%CI	AOD(95%CI	P-
		Unfavor	favorable))	value
		able				
Sex	Male	52(33%)	104(67%)	1.437(0.744,2	1.150(0.562,2.	0.702
				.779)	355)	
	Female	16(25.8	46(74.2%	1	1	
		%))			
Age at	< 1 month	37(27.8	96(72.2%	1	1	
presentatio		%))			
n	1 to 12 months	18(29%)	44(71%)	1.061(0.545,2	1.374(0.678,2.	0.378
				.068)	786)	
	>12 months	13(56.5	10(43.5%	3.373(1.361,8	5.002(1.820,1	0.002
		%))	.358)	3.750)	
Age at	< 3 years	59(29.8	139(70.2	1	1	
operation		%)	%)			
	\geq 3 years	9(45%)	11(55%)	1.928(0.759,4	0.901(0.267,3.	0.866
				.896)	035)	
Preoperativ	Yes	12(48%)	13(52%)	2.258(0.971,5	2.602(1.034,6.	0.042
e HAEC				.252)	546)	
	No	56(29%)	137(71%)	1	1	
Down	Yes	7(70.%)	3(30%)	5.623(1.407,2	7.441(1.796,3	0.006
syndrome				2.464)	0.839)	
	No	61(29.3	147(70.7	1	1	
		%)	%)			

Types of	f Soave	57(30.2	132(69.8	1	1	
pull		%)	%)			
through	Duhamel	11(37.9)	18(62.1%	1.415(0.628,3	1.907(0.768,4.	0.164
)	.187)	738)	
Types of	Classical (recto	50(29.4	120(70.6	1	1	
HSD	sigmoid)	%)	%)			
	Short segment	13(31,7	28(68.3%	1.114(0.534,2	0.815(0.2351,	0.636
		%))	.326)	1.896)	
	Long segment	5(71.4%)	2(28.6%)	6.00(1.126,31 .959)	7.176(1.267,4 0.638)	0.026

5. DISCUSSION AND IMPLICATION

Intestinal obstruction was the most common presentation of HSD patients with complaints of abdominal distension (97.3%), repeated vomiting of ingested matter (76.3%) and constipation or failure to pass meconium within 24-48 hours (82.2%) during initial presentation similar to other studies like, study done in northwest Tanzania(21, 22). Down syndrome occurs in 4.6% of the cases in this study which is slightly lower than study done in Ireland (13.4%) and HAEC occurs in 11.4% in this study almost similar to other studies (16.6%)(22).

In this study, majority of the patients were males which accounts 71.2% and females 28.8% with ratio of 2.5:1. But being male or female didn't affect operative outcomes of HSD patients(p=0.702). During the initial presentation, 60.7% of the cases presented within the neonatal age (less than 30 day), 28.8% were between one month and twelve months and 10.5% of the cases after the age of one year. Majority of patients at operation were below the age of three years. In similar study at Armed Forces Hospital Riyadh and other studies, most of the patients were males and also most of the them were present in neonatal age(23-25).

From this study 68.8% patients were having favorable outcome and 31.2% unfavorable outcome. Late presentation after one year of age was significantly associated with post-operative complications (p=0.002) than early presentation with adjusted odds ratio(AOR) of 5 times more common than patients presented at younger age. From the above statement it is clear that patients presented after the age of one year are 5 times more likely to develop post-operative complications (unfavorable outcome) than patients presented at younger age. About 56.5% patients above one year of age at presentation develops post-operative complications than younger age at presentation (29%). This is similar to study in Norway in contradict to study in children's hospital of Pittsburgh which implies that these children have a milder form of the disease, perhaps because of adaptation to the aganglionic state (14, 15). But age at operation didn't affect operative outcome didn't affected by the age at operation in both soave (p=0.21) and Duhamel(p=0.6) procedures(17, 24).

The most frequently observed post-operative complication from this study was constipation which occurs in 18(8.2%) patients after definitive operation and the second most common complication was incontinence which occurs in 14(6.4%) patients. Other complications like, wound infection occurs in (5.5%), anastomosis leak (0.9%), incisional hernia (2.7%), anal stenosis (2.7%) occurs after pull through procedure. The most common colostomy related complication in this study was colostomy necrosis which occurs in 2.7% of patients. The second common complication is colostomy retraction which occurs in 1.8% of patients. Colostomy prolapse occurs in 1.4% and colostomy stenosis in 0.5% of patients. Similar study at Armed Forces Hospital Riyadh, the complications following Soave's procedure were strictures in two patients (8%), enterocolitis in another two (8%), and anastomotic leakage in one (4%). The rate of constipation was 16% after the Soave's pull-through compared with 15% after the Duhamel's pull-through higher than in this study but not statistically significant(p=0.134)(23).

The presence of down syndrome also significantly associated with unfavorable outcome (p=0.006) in this study then those without down syndrome with AOR of 7.442 times more common than none down patients. From the above statement it is clear that patients with down syndrome are 7.442 times more likely to develop post-operative complications than patients without down syndrome. Similar study in Netherlands, compared to none down children,

children with DS have a higher rate of postoperative complications. Severe constipation was present significantly more often in DS children compared to NDS children (p < 0.01). Enterocolitis occurred more frequently in DS patients after operation (p = 0.038)(19, 20).

From this study 86.7% of HSD patients were underwent soave pull through and 13.3% of patients were underwent Duhamel pull through procedure and colostomy was done for all patients (100%) before definitive operations. There was no any patient underwent primary pull through from this study. It might be due to the absence of parenteral nutrition and late presentation of patients. Patients underwent Duhamel procedure have more unfavorable outcomes (37.9%) than soave procedure (30.2%) but not statistically significant (p=0.164) from this study. This is similar to study in Ireland but contradict to study in Indonesia which is HAEC frequently occurs after pull-through was significantly higher in the Duhamel than the Soave group (28% vs. 10%, respectively, p = 0.03)(22, 25).

The presence of preoperative HAEC also significantly associated with unfavorable outcome (p=0.044) than those without HAEC with AOR of 2.532 times more common than those without preoperative HAEC. From the above statement it is clear that patients with preoperative HAEC are 2.532 times more likely to develop post-operative complications than patients without HAEC. From this study 16% of patients with preoperative HAEC is developing wound infection and 12% develop incontinence. Similar study in Taiwan (p=0.013) and other study showed that patients with preoperative HAEC were more likely to develop post-operative complications than those that patients that preoperative HAEC were more likely to develop post-operative complications than those that patients with preoperative HAEC were more likely to develop post-operative complications than those not having preoperative HAEC(18).

From this study, 78% of cases were having classical (recto sigmoid) HSD,18.8% of patients were having short segment HSD and 3.2% long segment HSD. This is similar to study in Taiwan, Netherlands and Ireland (13, 18, 22), but contradict to study in northwestern Tanzania, the majority of patients (67.3%) had short segment aganglionosis(21). Patients having long segment HSD also significantly associated with unfavorable outcome of HSD patients (p=0.026), with AOR of 7.176 times more common than classical (recto sigmoid) and short segment HSD. From the above statement it is clear that patients with long segment HSD are 2.532 times more likely to develop post-operative complications than patients with recto sigmoid and short segment HSD. Similar studies in Taiwan and Ireland showed that post-operative complications like

constipation, incontinence and HAEC more commonly occurred in patients with long segment HSD than classical (recto sigmoid) HSD(18, 22).

6. CONCLUSION AND RECOMMENDATION

6.1. Conclusion

Intestinal obstruction was the most common presentation of HSD patients. Majority of HSD patients had favorable post-operative outcomes (68.8%). Majority of the patients in this study were males which accounts 71.2% and females account 28.8% of the cases. Late presentation after one year of age was significantly associated with post-operative complications (p=0.002).

The most frequently observed post-operative complication from this study was constipation which occurs in 8.2% of patients after definitive operation and the second most common complication was incontinence which occurs in 6.4% of the case. The most common colostomy related complication in this study was colostomy necrosis which occurs in 2.7% of patients. Patients having long segment HSD also significantly associated with unfavorable outcome of HSD patients (p=0.026). The presence of preoperative HAEC and down syndrome also significantly associated with unfavorable outcome (p=0.044, p=0.006 respectively), but types of operation, sex and age at definitive operation were not significantly affect operative outcomes of HSD patients.

6.2. Limitation of the study

- > This study is not representative to the general population since it is institutional based.
- Since this study is retrospective it may miss some important variables.

6.3. Recommendations

- Public awareness should be increased on clinical manifestation of HSD to bring their child early.
- Based on our findings we suggest that health professionals in the hospital should increase public awareness on HSD by providing appropriate health information.
- Attention should be given to patients presented with preoperative HAEC and down syndrome in HSD.
- Further research using prospective study design is warranted as a way to overcome the limitations of secondary data in the current retrospective research that preclude generalization to the whole population.

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8. APPENDIX

8.1 Checklists

Part I: Sociodemographic factors

1. Age.....

2.Sex

- 0 M
- 0 F

Part II: Presenting complaints

- 1. Vomiting
 - o Yes
 - o No
- 2. Abdominal distension
 - o Yes
 - o No
- 3. Failure to pass meconium(constipation)
 - o Yes
 - o No
- 4. Others
- 5. Age at onset of symptoms.....
- 6. Age at definitive operation.....
- 7. Associated syndromes
 - o Yes

- o No
- If yes, what is the syndrome?.....
- 8. Preoperative HAEC
 - o Yes
 - o No

Part III: Workup

- 1. Barium enema done
 - o Yes
 - o No
 - If yes where is the location of transition zone.....
- 2. Rectal biopsy done
 - o Yes
 - o No

Part IV: Types of operation (types of pull through)

- 1. Primary pull through
 - o Duhamel
 - o Soave
- 2. Staged pull through
 - o Duhamel
 - o Soave

Part V: Types of HSD segment

- Short segment HSD
- Long segment HSD
- Classical (recto sigmoid) HSD

Part VI: Post-operative complications

1. General complications

- Wound infection
- Anastomosis leak or disruption
- Enterocolitis
- Adhesive small bowel obstruction
- Constipation
- Incontinence
- Anal stenosis which needs dilatation
- Others.....
- No complication
- 2. Colostomy related complications
 - o Prolapse
 - \circ Retraction
 - o Stenosis
 - o Necrosis
 - Others
 - No complication

Part VII: General outcomes

- Unfavorable
- Favorable