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**ADDIS ABABA UNIVERSITY**  
**COLLEGE OF HEALTH SCIENCES**  
**DEPARTMENT OF RADIOLOGY**

**Barium Enema – Pathology correlation in Hirschsprung's  
disease at TASH & Menelik II Hospital, Addis Ababa,  
Ethiopia**

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**Addis Ababa, Ethiopia**

**Oct, 2021 GC**

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Radiology**

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## **ACRONYMS/ABBREVIATIONS**

AAU----- Addis Ababa University

ARM-----Anorectal Manometry

CE-----Contrast Enema

G.C-----Gregorian calendar

HD/HSD-----Hirschsprung's disease

NPV-----Negative Predictive Value

PPV-----Positive predictive Value

RS-----Recto-sigmoid ratio

RSB-----Rectal suction Biopsy

Sn-----Sensitivity

Sp-----Specificity

TASH-----Tikur Anbessa Specialized Hospital

TZ-----Transition zone

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## **ABSTRACT**

**Background:** Hirschsprung's disease is a common cause of pediatric intestinal obstruction. It's a developmental disorder characterized by the absence of ganglion cells in the myenteric and submucosal plexuses of the distal intestine resulting in functional obstruction. Barium enema is a simple and primary study for the evaluation of clinically suspected Hirschsprung's disease.

**Aim:** To review the diagnostic performance of barium enema examinations for the investigation of Hirschsprung's disease.

**Methods:** Institution based prospective cross-sectional study at Tikur Anbessa Specialized - & Menelik II Hospitals was done with clinically HD suspected patients who have a barium enema & biopsy during the study period, September 2020 to August 2021. Convenience sampling technique was used.

**Results:** A total of 28 cases were evaluated during the study period all having a barium enema & biopsy result, of whom 18 were males & 10 were females. The minimum & maximum ages at presentation were 10 days & 10 years respectively with the median age being 11 months. Constipation was the most commonly present clinical symptom followed by abdominal distension & delayed meconium passage which was found in 50% of the participants.

The overall sensitivity, specificity & accuracy of *barium enema* was 95%, 71.4 %, & 88.8 % respectively. Sensitivity, specificity, PPV, and NPV were 90%, 71.4 %, 90% and 71.4 % respectively for *recto-sigmoid index* in barium enema. Sensitivity, specificity, PPV, and NPV were 85 %, 85.7%, 94.4% and 66.7 % respectively for *transitional zone*. Sensitivity, specificity, PPV, and NPV were 75%, 100%, 100% and 58.3 % respectively for *irregular contraction* detected in barium enema. Sensitivity, specificity, PPV, and NPV were 35%, 100%, 100% and 35 % respectively for *mucosal irregularity* detected in barium enema.

**Conclusion:** The accuracy of barium enema for the diagnosis of HSD was 88.8 % & hence barium enema can be used as a reliable primary diagnostic modality for HSD. Irregular contraction & mucosal irregularity were the most specific findings whereas inverted recto sigmoid index & transition zone had a comparably higher sensitivity.

# 1. INTRODUCTION

## 1.1 BACKGROUND

Hirschsprung's disease (HD) is a common cause of functional intestinal obstruction in children caused by a lack of ganglion cells in the distal bowel wall (1,2). The aganglionic segment usually begins at the rectum and extends cranially. Short-segment disease, accounting for about 80%, is confined to the recto-sigmoid region of the colon. Long-segment disease, accounting for 3% to 10%, extends past this region and can affect the entire colon. Rarely the small and large intestines are involved. The controversial ultra-short segment disease extends 3-4 cm cranial to the internal anal sphincter (2, 3, 10).

Although HD may present at different ages and with varied clinical findings, the most common presentation is a neonate with severe constipation or signs of intestinal obstruction (2,3). The disease more frequently presents in infancy, although some patients present with persistent and severe constipation later in life. Early diagnosis is essential to prevent complications such as enterocolitis and colonic rupture.

Diagnosis or exclusion of HD is a frequent exercise in any pediatric practice and poses challenges to pediatric radiologists, pediatricians and pediatric surgeons practicing in resource-limited countries (3). The diagnosis is mainly by radiographic studies, anorectal manometry (ARM) and histological examination of rectal wall biopsies (4).

The radiological diagnosis of HD is possible if films of diagnostic quality can be produced. Contrast enema studies are associated with minimal risk of complications (7). A funnel shaped transition zone on a contrast enema at the junction of aganglionic and ganglionic gut is considered a hallmark for its diagnosis (8).

In developing countries HD poses a diagnostic and therapeutic challenge. In many centers in sub-Saharan Africa, particularly the rural areas, facilities and manpower required for the diagnosis & intervention are not adequately available.

Ignorance and poverty on the part of the parents, late presentation with complications, limited access to trained professionals and limitation of facilities for prompt diagnosis & management are common challenges encountered (2,5,6).

## **1.2 Significance of the study**

Ethiopia, as part of the developing world, resources are limited for establishing an early diagnosis of HD. There are also limited studies with regards to the diagnostic accuracy of barium enema in HD. So this study can serve an input to the existing limited body of local literature in imaging of suspected HD.

## **2.Literature Review**

Hirschsprung's disease, also known as congenital megacolon or congenital colonic aganglionosis, is characterized by aganglionosis (absence of ganglion cells) in the distal bowel extending proximally for variable distances that result in functional intestinal obstruction caused by dysmotility of the diseased segment (1, 2).

HD is a common cause of pediatric intestinal obstruction (8, 9). The worldwide incidence of HD ranges from 1:5000 to 1:10 000 live births and varies among different ethnic groups (Northern European, 1.5:10 000; African American, 2.1:10 000; and Asian, 2.8:10 000). A male-to-female ratio of 3:1 to 4:1 is observed largely in short-segment disease; the sex bias diminishes in long-segment disease with a reported male-to-female ratio of 1:2 to 2:1. The disease is more common in term babies than preterms.(26)The cause of HD is multifactorial, and the disease can be familial or develop spontaneously (2, 10, 11).

The clinical presentation ranges from neonatal intestinal obstruction to chronic progressive constipation in older children. Approximately 80 percent of patients present in the first six weeks of life with difficult bowel movements, poor feeding, and progressive abdominal distention (2, 3, 9). In Africa only 20-40% present at the neonatal age, compared to more than 90% in developed countries (2, 5, 6, 11). A Tanzanian study including a total of 110 HD patients showed a median age of 24 months with six (5.5%) cases in the neonatal period.

A recent retrospective analysis of 10,112 cases of HD over a period of 20 years aimed at reexamining dogma, the age at presentation of common pediatric surgical conditions, however, demonstrated that 6.5% of cases presented within the

neonatal period and 45.9% prior to age of 1 year. The median & mean age of Dx of HD in this study were 1 year & 3 years respectively.(22)

Patients with clinical suspicion for HD should undergo a diagnostic evaluation; which include contrast enema (CE) and ARM. The CE is performed with diluted barium sulfate preferably with normal saline that is administered rectally using 6FR feeding tube.

The reported diagnostic accuracy of CE in the literature is variable. Literatures reported that CE had lower mean sensitivity (70%) and specificity (83%) when compared with ARM and RSB (19). Furthermore, CE in the neonatal period has been shown to be less reliable than in older children. Age less than 30 days was shown to be a predictor of a false-positive result, and CE results in those less than 30 days of age was reported to have a 7-fold higher probability of false-negative results. (3,20,26)

The presence of a radiographical transition zone (TZ) with proximal dilated bowel, retention of contrast on post-evacuation film, irregular colonic contractions, mucosal irregularity, and an abnormal recto-sigmoid ratio are findings that are suggestive of HD (3, 17). The radiographic location of the TZ has been shown to correlate with the length of the aganglionic segment, specifically of the recto sigmoid segment, and may aid in surgical procedure planning (17, 18).

Since its first description by Harald Hirschsprung, the understanding and the management of the condition has improved greatly. The first successful treatment of the condition was undertaken over six decades ago by Orvar Swenson (12). Since this first definitive operation was described, many other patients have been treated successfully with other operations including the retro-rectal pull-through of Duhamel and its modifications and the endorectal pull-through of Soave procedure and its various modifications (13, 14, 15, 16).

### **3.Objective:**

#### **3.1 General objectives:**

- To assess the diagnostic accuracy of barium enema in Hirschsprungs's disease.

#### **3.2 Specific Objective:**

- Assess specificity & sensitivity of the common radiological findings as compared to the gold standard/ Rectal biopsy/.
- Assess the age & gender distribution of the disease.

## **4. Materials and Method:**

**4.1 Source Population:** children with clinical diagnosis of HD in Black Lion and Menelik II Hospitals who were subsequently evaluated with barium enema study.

**4.2 Study subjects:** All children with clinical diagnosis of HD in Black lion & Menelik II Hospitals who had both barium enema & rectal biopsy.

**4.3 Study Design:** Institution based prospective cross-sectional study at Tikur Anbessa Specialized - & Menelik II Hospitals was done with clinically HSD suspected patients who had a barium enema & rectal biopsy during the study period, September 2020 to August 2021. Convenience sampling technique was used.

A total of 68 barium enema images of clinically HD suspected cases were collected mostly from the PACS of Vision internal medicine specialty clinic & partly received directly from the patients in a form of compact disc (CD). 28 of the 68 had both barium enema & biopsy in the study period, which were then enrolled in the study.

**4.4 Study period:** A prospective study was done from September 2020 to August 2021.

## **4.5 Inclusion & Exclusion Criteria**

### **Inclusion Criteria:**

- Delayed passage of meconium (beyond 48 hours for a full term neonate and beyond 72 hours for a preterm neonate).
- Patients with defecation problems since birth and abdominal distension.

### **Exclusion Criteria:**

- Abdominal radiograph showing multiple air fluid levels.
- The modified Bell staging criteria (21) in which a composite of clinical signs and symptoms (eg, abdominal distention, bloody stools, or hypotension), biochemical parameters (eg, thrombocytopenia or

neutropenia), and radiographic signs (eg, pneumatosis or pneumoperitoneum) was used to exclude NEC.

#### **4.6 Study Variables**

- Age
- Sex
- Clinical presenting symptoms ( Constipation, abdominal distension, delayed meconium passage, vomiting).
- Barium enema imaging findings ( RS, TZ, irregular contraction, mucosal irregularity)
- Final barium Diagnosis
- Biopsy result

#### **4.7. Data collection**

Data was collected by principal investigator and barium enema image interpretation was done by principal investigator and Advisors. A modified structured questionnaire was used to collect information from patient's charts.

##### **4.7.1 Data quality control**

The research advisors have reviewed the modified structured questionnaire before the study is conducted.

#### **4.8. Data Analysis**

After filling the data on to SPSS version 26 and analyzed, tables and figures were drawn using SPSS and Microsoft Excel. Frequency distribution and percentages were obtained for:

- Socio-demographic data (age, sex)
- Cross tabulation was used to describe the frequency and percentage of the barium enema imaging findings.

#### **4.9 Ethical Consideration.**

Data collection was taken after getting permission from ethical committee of the department of radiology and Addis Ababa University. Approval from IRB was obtained and formal letter were written from Radiology Department to the outpatient directorate of TAH & Menelik II Hospitals before commencing the data collection process. Then, for legal purpose data collection of individual record had taken place in the medical records room. After completion of data collection, medical records (charts) were returned back to their original place properly.

#### **4.10 Dissemination of the study findings**

The findings of this study will be submitted to the Department of Radiology, College of Health Sciences, Addis Ababa University as a partial fulfillment of specialty certificate in Radiology. The outcome of this study will be presented to other interested stakeholders such as annual conferences. Finally, the manuscript will be submitted to a reputable scientific journal for possible publication.

## 5. RESULT

A total of 28 cases were evaluated during the study period all having a barium enema & biopsy result, of whom 18 were males & 10 were females. The minimum & maximum ages at presentation were 10 days & 10 years respectively with the median age being 11 months. Only 5 patients were in the neonatal period (17.9%).

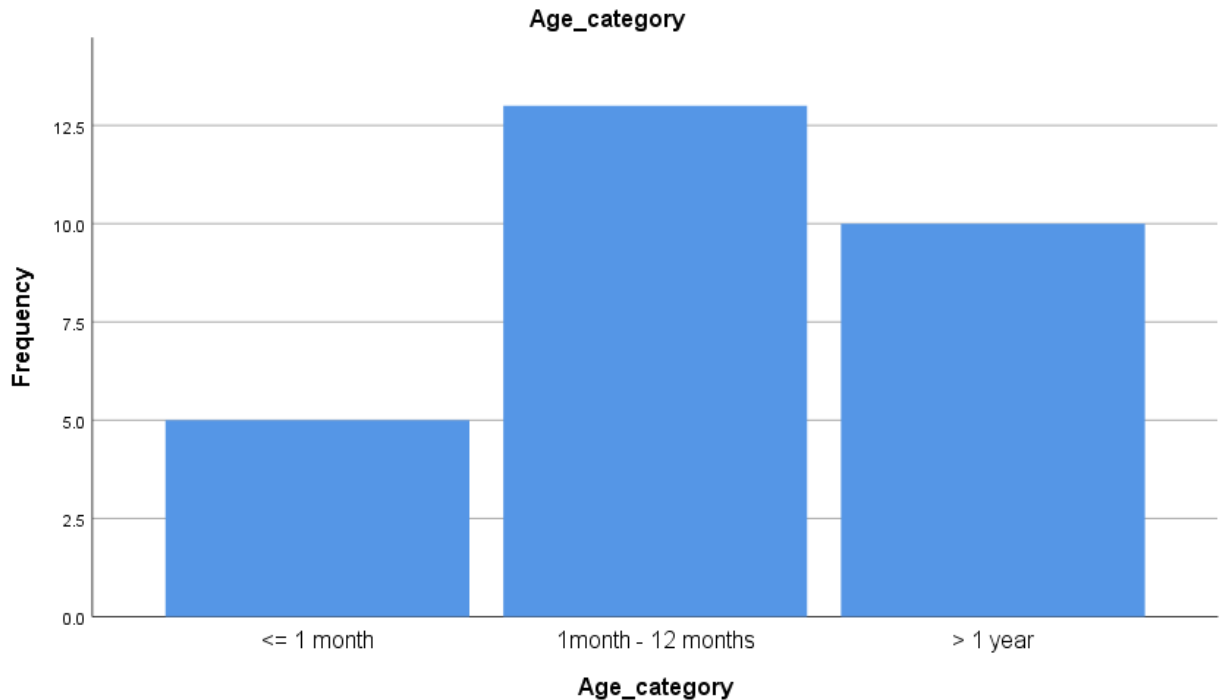


Figure 1: Age category frequency (All participants with barium enema & rectal biopsy)

Constipation was present in all of the cases, abdominal distension in 27 cases (96.4%) & history of delayed meconium passage was present in 14 cases (50%). Vomiting was present in 3 cases (10.7%).

Barium enema study was suggestive of HSD in 22 of the cases of whom 19 were aganglionated, 2 ganglionated & 1 inconclusive upon rectal biopsy. The remaining 6 barium studies were not suggestive of HSD & upon histopathologic evaluation ganglion cells were demonstrated in 5 & not demonstrated in the remaining 1.

The median age of diagnosis of HSD in these 20 biopsy confirmed cases was 1 year & M: F ratio was 3:1. Only 3 of the confirmed cases were in the neonatal age group.

The overall sensitivity, specificity & accuracy of barium study is 95%, 71.4 %, & 88.8 % respectively.

**Table 1. Barium enema & biopsy result cross tabulation.**

<b>BIOPSY RESULT</b>	<b>BARIUM ENEMA RESULT</b>		
	<b>Suggestive of HD</b>	<b>Non-suggestive</b>	<b>Total</b>
<b>No ganglion cell</b>	<b>19</b>	<b>1</b>	<b>20</b>
<b>Ganglion cell seen</b>	<b>2</b>	<b>5</b>	<b>7</b>
<b>Non diagnostic</b>	<b>1</b>	<b>0</b>	<b>1</b>
<b>Total</b>	<b>22</b>	<b>6</b>	<b>28</b>

Frequency, Sensitivity, specificity, PPV, NPV of the barium enema findings in HSD (recto-sigmoid ratio <1, transition zone, irregular contraction & mucosal irregularity in those with HSD & w/o HSD) are demonstrated in the following 4 consecutive tables:

**Table 2.** Frequency of rectosigmoid index in barium enema study in subjects with & without HSD

	<b>RS &lt; 1</b>	<b>RS &gt; 1</b>
<b>HSD ( +Ve)</b>	<b>18</b>	<b>2</b>
<b>HSD ( -Ve)</b>	<b>2</b>	<b>5</b>

**Table 3.** Frequency of transition zone (TZ) in barium enema study in subjects with & without HSD

	TZ- Present	TZ- Absent
HSD ( +Ve)	17	3
HSD ( -Ve)	1	6

**Table 4.** Frequency of irregular contraction in barium enema study in subjects with & without HD.

	Irregular contraction- Present	Irregular contraction- Absent
HSD ( +Ve)	15	5
HSD ( -Ve)	0	7

**Table 5.** Frequency of Mucosal irregularity in barium enema study in subjects with & without HD.

	Mucosal Irregularity- Present	Mucosal irregularity- Absent
HSD ( +Ve)	7	13
HSD ( -Ve)	0	7

**Table 6. Summary specificity, sensitivity, PPV & NPV of Barium enema findings in HD**

BARIUM ENEMA FINDING	Sp ( %)	Sn (%)	PPV(%)	NPV(%)
Recto-sigmoid index	71.4	90	90	71.4
Transition zone	85.7	85	94.4	66.67
Irregular contraction	100	75	100	58.33
Mucosal Irregularity	100	35	100	35

## **6. DISCUSSION**

In this study, the median age of presentation for HD confirmed cases was 1 year which is a younger age as compared to 2 years in a Tanzanian study which enrolled 110 cases(2). And it's same as the finding in a retrospective study of the age at presentation of common pediatric surgical conditions: Reexamining dogma , which included a total of 10,112 HD cases.(22 )

The gender distribution of biopsy confirmed HD cases which was 3:1 in our study is comparable with other studies. In the previously mentioned Tanzanian study the M:F ratio was 3.6:1. (2)

The sensitivity, specificity & accuracy of barium enema in the Dx of HD in this study was 95%, 71.4% & 88.8% respectively. According to another study which enrolled 111 patients (25), the sensitivity & specificity of barium enema was found to be 76% & 97% respectively. In a systematic review of the literature, de Lorijn et al. reported that CE had lower mean sensitivity and specificity of 70% and 83%. As compared to these studies, our study suggested that barium enema is more sensitive than it's specific.

In this study irregular contraction was found in 15 out of 20 cases of HSD & was not detected in those without HSD, making the specificity & PPV of this finding 100%, a result which is the same as with other studies done by Alehossein (23) & Mehran Peyvateh (24) which enrolled 60 & 55 patients respectively, in which no irregular contraction was detected in all cases without HD.

Mucosal irregularity was found in 7 out of 20 cases of HD & was not detected in those without. Likewise, no mucosal irregularity was detected in those without HD in a study done by Alehossein (23) & Mehran Peyvateh(24).

In this study 85% & 90 % of HD cases have transition zone & RS <1 respectively, making these signs the most sensitive barium enema findings in HD, a finding consistent with other studies as well. In a study done by Mehran Peyvateh (24) 90% & 76.67% had TZ & inverted RS index respectively. In Alehossein et al. paper(23) TZ & inverted RS were present in 94% & 86% of the cases respectively.

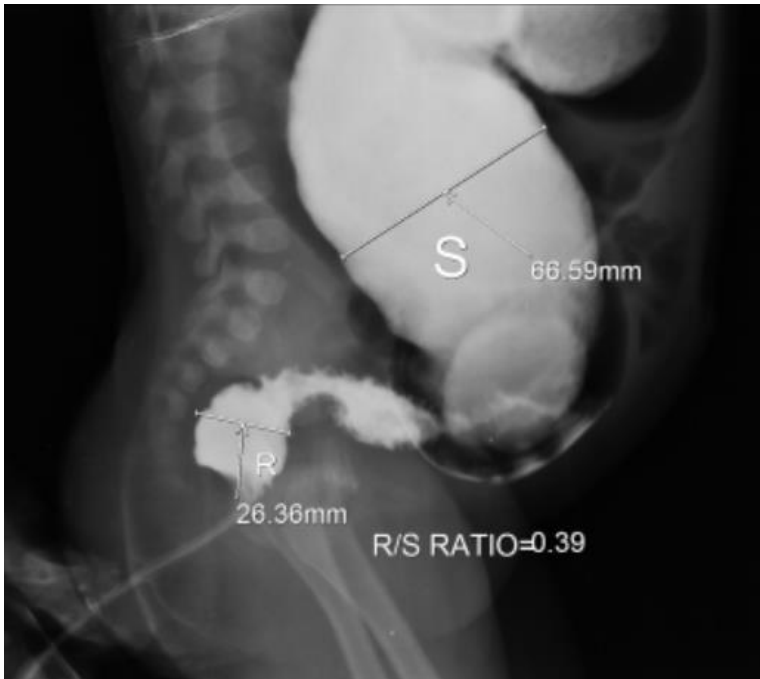
Unlike these studies, however, RS<1 was more sensitive than transition zone in our study.

**Table 7.** Comparison b/n ours, Mehran's & Alehossein study findings.

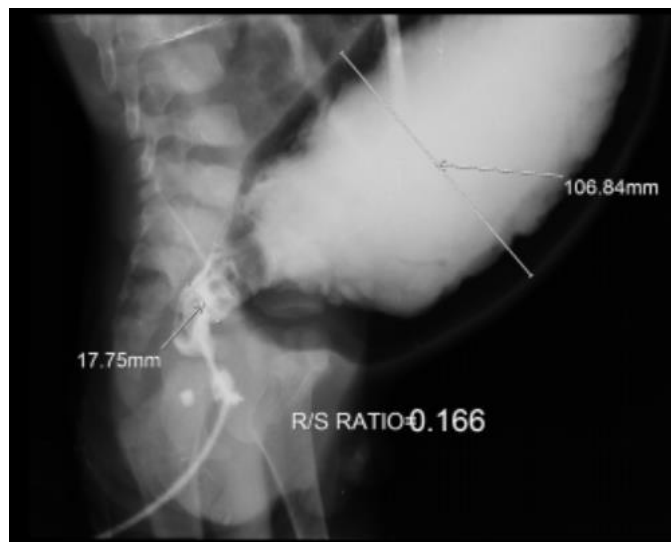
<b>BARIUM ENEMA FINDINGS</b>	<b>Sp (%)</b>			<b>Sn (%)</b>			<b>PPV (%)</b>			<b>NPV (%)</b>		
	<b>Our study</b>	<b>Mehran (24)</b>	<b>Alhousen (25)</b>	<b>Our study</b>	<b>Mehran (24)</b>	<b>Alhousen (25)</b>	<b>Our study</b>	<b>Mehran (24)</b>	<b>Alhousen (25)</b>	<b>Our study</b>	<b>Mehran (24)</b>	<b>Alhousen (25)</b>
RS < 1	71.4	83.3	68.4	<b>90</b>	76.6	86	90	78.1	83.7	<b>71.4</b>	82.1	72.2
TZ	85.7	80	68.4	85	<b>90</b>	<b>94.4</b>	94.4	81.8	85	66.6	<b>88.8</b>	<b>86.7</b>
Irregular contraction	<b>100</b>	<b>100</b>	<b>100</b>	75	46.6	41.6	<b>100</b>	<b>100</b>	<b>100</b>	58.3	65.2	47.5
Mucosal Irregularity	<b>100</b>	<b>100</b>	94.7	35	23.3	18.3	<b>100</b>	<b>100</b>	76.5	35	56.6	36.5



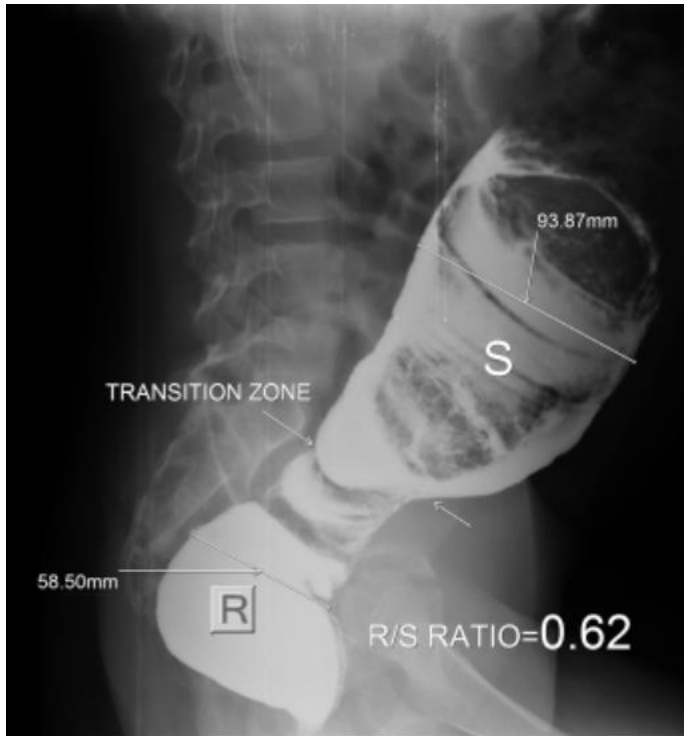
A) Biopsy confirmed HSD, 10 days old neonate, F (*Inverted rectosigmoid index & transition zone at rectosigmoid junction are demonstrated*).



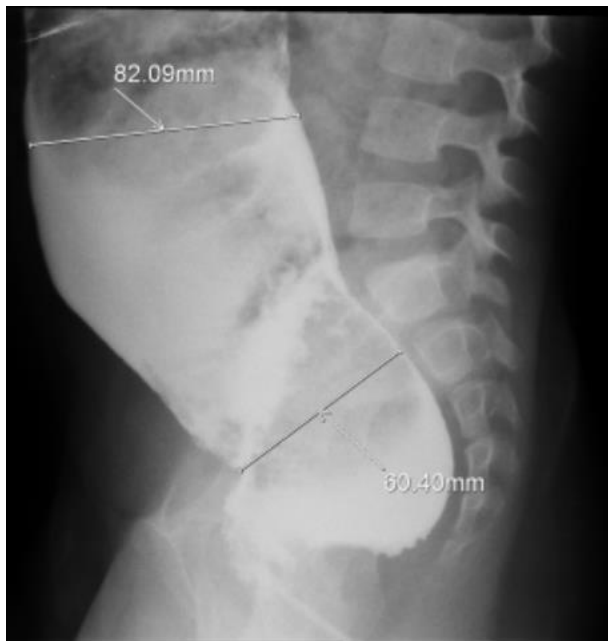
B) Biopsy confirmed HSD, 03 months, F (*Inverted RS index, transition zone at rectosigmoid junction & bizarre, irregular contractions are demonstrated*).



C,D. Biopsy confirmed HSD 2 years old male, control film & Barium enema resectively. (*Inverted rectosigmoid index with marked proximal dilatation & transition zone at rectosigmoid junction are noted*).



E) Biopsy confirmed HSD, **10 years old, F** (*Inverted rectosigmoid index & funnel- shaped transition zone at rectosigmoid junction are visualized*).



F) 2 years & 6 months old, M, **Biopsy= Ganglionated**. (*Inverted rectosigmoid index is seen*).

## **7. CONCLUSION**

The accuracy of barium enema for the diagnosis of HSD was 88.8 % & hence barium enema can be used as a reliable, primary diagnostic modality for HD. Irregular contraction & mucosal irregularity were the most specific findings whereas inverted recto sigmoid index & transition zone had comparably the highest sensitivity.

## **8. LIMITATIONS**

Comparing the Sensitivity & Specificity of the barium enema findings in different age groups was not done due to the limited number of HD cases especially in the neonatal period.

Post evacuation film for delayed meconium passage was not mostly performed, so it was not included as a study variable.

Getting a complete data including the barium enema images, biopsy reports & charts of the patients was the major challenge which required a lot of time & energy.

## **9.RECOMMENDATION**

A larger scale longitudinal study of this kind is recommended to further look & compare the sensitivity & specificity of barium enema for the diagnosis of HSD in different age groups. The availability of a fluoroscopy machine at Black lion Hospital, Radiology department will be of high significance to achieve this in addition to benefiting patients a lot and residents as well.

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**11.ANNEX**

**Questionnaire**

**Pediatric Barium Enema Diagnostic Accuracy with Radiology-Pathology  
Correlation**

**Data Collection Tool**

**I. CLINICAL DATA:**

**A. Patient**

Name : \_\_\_\_\_ MRN: : \_\_\_\_\_ Age: : \_\_\_\_\_

Sex: :M      F

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

Singleton:

Twin:

Presenting compliant

i. Failure/Delay to pass meconium

ii. Abdominal distention

iii. Constipation [Refractory]

iv. Anal opening

v. Vomiting

vi. External genitalia [anomalies]

**B. Mother**

Maternal age during pregnancy 1. <20       2. 20-35       3. >35

ANC follow up 1. Yes       2. No

**IF YES Prenatal Ultrasound Documentation**

Known maternal chronic illness 1. Yes       2. No

IF YES 1. DM       2. Hypertension       3. Seizure Disorder       4.

**Other**

C. Sibling with similar problem 1. Yes       2. No

IF YES [specify\*] \_\_\_\_\_

D. Number of family members \_\_\_\_\_

**E. Average monthly income of the family [Total]**

1. <1500 ETB       2. 1500-3000ETB       3. 3000-5000ETB       4. >5000ETB

**II. IMAGING FINDING:**

- a. Rectosigmoid Index 1. <1  2. > 1
- b. Transition Zone 1. Present: level  2. Absent
- c. Irregularity of the Mucosa 1. Present  2. Absent
- d. Bowel Caliber 1. Normal  2. Distended  3. Small   
[Microcolon]
- e. Delay in Barium Evacuation after 24 hours 1. Present   2. Absent   
3. Not present
- e. Irregular contraction 1. Present  2. Absent
- f. Fistula 1. Present [specify]  \_\_\_\_\_ 2. Absent
- g. Location of the Rectal Pouch [specify] \_\_\_\_\_
- h. Other Congenital Anomaly 1. Yes [specify] \_\_\_\_\_  2. No

**III. SURGICAL [INTRA-OP] FINDING:**

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**IV. BIOPSY RESULT**

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