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Sensitivity and Specificity of Barium Enema in the Radiological Diagnosis of Hirschsprung Disease in Children in a Tertiary Pediatric Hospital

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Abstract

Background: *Hirschsprung disease (HSCR) is a congenital disorder characterized by the absence of ganglion cells in the distal bowel, leading to functional obstruction. Although rectal biopsy is the gold standard, barium enema remains a widely used diagnostic tool in pediatric radiology, particularly in resource-constrained settings. This study aimed to assess the sensitivity, specificity, and diagnostic performance of barium enema findings in diagnosing HSCR using histopathology as the reference standard.*

Methods: *This cross-sectional observational study was conducted at the Department of Radiology and Imaging, Bangladesh Shishu Hospital & Institute, Dhaka, Bangladesh, from January 2021 to December 2024. A total of 150 children with suspected HSCR underwent barium enema, followed by confirmatory rectal biopsies. The radiological features, including the transitional zone, rectosigmoid index (RSI) <1, and 24-hour retention films, were analyzed. Statistical analyses were performed using SPSS v25.0.*

Results: *Among the 150 participants, 73.3% were male and 44.7% were less than one month old. Clinically, constipation (80.7%), abdominal distension (78.7%), and delayed meconium passage (76.7%) were the most common symptoms. The 24-hour retention film showed the highest sensitivity (92.5%) and specificity (91.4%), followed by the transitional zone (87.3%, 90.1%), and RSI <1 (75.9%, 88.7%). Diagnostic accuracy was highest in children aged 1–12 months and in patients with short-segment disease.*

Conclusion: *Barium enema, particularly the 24-hour delayed film, demonstrates high diagnostic accuracy and remains a valuable noninvasive tool for evaluating HSCR in pediatric patients.*

Keywords: *Hirschsprung disease, barium enema, pediatric radiology, sensitivity, specificity*

1. INTRODUCTION

Hirschsprung disease (HSCR) is a congenital developmental disorder that is defined by the lack of ganglion cells in the distal bowel, subsequently causing functional intestinal obstruction. It is due to the failure of neural crest cell migration during intestinal development, leading to varying lengths of aganglionosis, most commonly in the rectosigmoid area [1].

The prevalence of HSCR is estimated to be about 1 in 5,000 births, occurring in live births with a male-to-female ratio estimated to be 4:1 [2]. Diagnosis at an early stage is important to prevent complications of enterocolitis, failure to thrive or life-threatening sepsis [3].

HSCR can be classified based on the length of the aganglionic segment into short-segment disease, long-segment disease, and total colonic

aganglionic disease. Short-segment HSCR, which affects the rectosigmoid region, is the most common form and often presents with more distinct radiological features. Long-segment disease, which extends proximal to the sigmoid colon, and total colonic aganglionosis are less common but pose greater diagnostic challenges, especially with barium enema due to diffuse or subtle radiologic findings [4,5]. Therefore, identifying the disease segment is crucial for evaluating radiologic performance and tailoring clinical management.

HSCR clinical manifestation is age and level of aganglionosis dependent. Typical symptoms are a failure to get rid of meconium, slow-moving bowels, persistent constipation, bloating, and throwing up [6]. These symptoms are however not specific and may overlap with other causes of neonatal gangrene of the intestine. Therefore, confirmation of histopathologic examination through rectal biopsy is essential in the conclusive diagnosis, and this is the gold standard [7, 8].

An important role in the initial diagnosis of suspected HSCR belongs to the imaging, contrast (barium) enema. It is non-invasive, widely available and gives structural evidence like a transition zone and a rectosigmoid index <1 and prolonged evacuation on a 24-hour retention film [9]. Those results are suggestive rather than indicative when taken alone. Barium enema diagnostic performance has been variedly reported in different studies as sensitive between 70-95%, depending on technique effectiveness, interpreter experience and age of the patients [10, 11].

Barium enema, despite its limitations, continues to be the main radiological method of screening HSCR in resource-limited settings. This is especially applicable in Bangladesh and other low and middle-income nations, where access to manometry or worthwhile rectal biopsy can be limited. Proper interpretation of radiological signs can help in better clinical diagnoses and minimize unnecessary surgical relays [12, 13].

Individual radiologic features have been tried to justify the same by several studies. Having a transitional zone has exhibited high specificity but erratic sensitivity [14, 15]. Even a rectosigmoid index <1 has been regarded as a good predictor in the past, but more recent studies have cast doubt on its usage in neonates [16]. The delayed film after 24 hours has shown itself to be a rather promising signifier, and in some cases, is more medically accurate in diagnosis [17].

A large body of research, however, reveals a wide range of variability in barium enema diagnostic yield across populations. This inconsistency is attributable to a number of factors such as inter-observer variability, disease segment length and the patient age among others [18, 19]. Not much has been studied when it comes to the sensitivity and specificity of barium enema in children, especially in the Bangladeshi setting. The current study attempts to fill this gap by determining the accuracy of specific barium enema findings, including transitional zone, rectosigmoid index <1, and 24-hour retention, against the actual histopathological diagnosis of HSCR. This study aimed to provide evidence-based knowledge that can be used to understand the utility of barium enema in local practice by evaluating such characteristics in a large pediatric population in a tertiary hospital in Bangladesh.

2. METHODOLOGY & MATERIALS

This was a cross-sectional observational study conducted at the Department of Radiology and Imaging, Bangladesh Shishu Hospital & Institute, Dhaka, Bangladesh, over four years from January 2021 to December 2024. The study included 150 children with clinically suspected Hirschsprung disease (HSCR) referred for barium enema evaluation, followed by confirmatory histopathology.

2.1. Sample Selection

Inclusion Criteria

- Children aged <18 years with clinical features suggestive of HSCR.
- Patients referred for radiological evaluation by barium enema.
- Histopathological confirmation is available through rectal biopsy.

Exclusion Criteria

- Unsatisfactory biopsy specimen.
- Patients with incomplete or missing imaging or biopsy data.
- Children with known anorectal malformations or previous bowel surgery.
- Parents or guardians who did not give informed consent.

2.2. Data Collection Procedure

Data were collected using standardized forms capturing demographic, clinical, and radiological data. Barium enema was performed according to

institutional protocols, and the images were interpreted by experienced radiologists. The clinical features were recorded from the medical records. Histopathological confirmation of HSCR was obtained through a rectal biopsy.

Data collection was ensured for completeness and accuracy through double-entry and review. Informed consent was obtained from the parents or guardians of all participants, and confidentiality was strictly maintained throughout the study.

2.3. Statistical Analysis

Data were analyzed using SPSS version 25.0. Descriptive statistics were used to present the frequencies and percentages. Diagnostic accuracy was assessed by calculating the sensitivity, specificity, positive predictive value (PPV), negative predictive value (NPV), Youden Index, and Kappa agreement. Diagnostic metrics were stratified by age group and disease segment type to evaluate subgroup variations in radiologic performance.

3. RESULTS

Table 1. *Baseline characteristics of the study population (n=150)*

Characteristics		Frequency (n)	Percentage (%)
Age Group	<1 month	67	44.67
	1–12 months	51	34.00
	>12 months	32	21.33
Sex	Male	110	73.33
	Female	40	26.67
Birth weight <2.5 kg		39	26.00
Term delivery (≥37 weeks)		124	82.67
NICU admission post-birth		18	12.00
Family history of HSCR		5	3.33

Table 1 shows the baseline characteristics of the 150 children included in the study. The majority were aged <1 month (44.7%), followed by 1–12 months (34.0%) and >12 months (21.3%). Most patients were male (73.33%). Low birth weight (<2.5 kg) was observed in 26% of cases, 82.67% were born at term, 12% required NICU admission after birth, and 3.33% had a family history of HSCR.

Table 2. *Clinical Characteristics of Children with Suspected Hirschsprung Disease (n=150)*

Clinical Feature	Frequency (n)	Percentage (%)
Delayed meconium passage (>48h)	115	76.67
Constipation	121	80.67
Abdominal distension	118	78.67
Vomiting	76	50.67
Failure to thrive	64	42.67
Explosive stool after DRE	38	25.33
Enterocolitis at presentation	13	8.67

Table 2 presents the clinical characteristics of children with suspected HSCR. Constipation (80.67%), abdominal distension (78.67%), and delayed passage of meconium beyond 48 hours (76.67%) were the most common symptoms. Vomiting occurred in 50.67%, failure to thrive in 42.67%, explosive stools after digital rectal examination in 25.33%, and enterocolitis at presentation in 8.67% of patients.

Table 3. *Cross-tabulation Between Barium Enema Findings and Histopathological Diagnosis*

Radiologic Feature	True Positive	False Positive	False Negative	True Negative
Transitional Zone	69	7	10	64
Rectosigmoid Index < 1	60	8	19	63
24-hour Retention Film	74	6	6	64

Table 3 compares the radiological findings from barium enema with histopathological diagnosis. The 24-hour retention film showed the highest number of true positives (74) and the lowest false negatives (6), while the transitional zone and rectosigmoid index <1 showed higher false negatives (10 and 19, respectively).

Table 4. *Diagnostic Accuracy of Barium Enema in Detecting Hirschsprung Disease*

Feature	Sensitivity % (95% CI)	Specificity % (95% CI)	PPV (%)	NPV (%)	Youden Index	Kappa (95% CI)
Transitional Zone	87.3 (78.5–93.2)	90.1 (81.2–95.4)	90.8	86.5	0.77	0.78 (0.72–0.84)
Rectosigmoid Index <1	75.9 (65.8–83.8)	88.7 (79.3–94.4)	88.2	76.8	0.64	0.63 (0.55–0.70)
24-hr RetentionFilm	92.5 (84.8–96.9)	91.4 (82.8–96.4)	92.5	91.4	0.84	0.81 (0.75–0.87)

Table 4 describes the diagnostic accuracy of the three main radiologic features. The 24-hour retention film demonstrated the highest sensitivity (92.5%) and specificity (91.4%), with a Youden Index of 0.84 and a Kappa value of 0.81. The transitional zone had a sensitivity of 87.3% and a specificity of 90.1%. The rectosigmoid index <1 had lower sensitivity (75.9%) but retained high specificity (88.7%).

Table 5. *Sensitivity and Specificity of Barium Enema by Age Group and Disease Segment Type*

Subgroup		Sensitivity (%)	Specificity (%)
Age Group	<1 month	90.0	91.7
	1–12 months	92.9	92.3
	>12 months	83.3	87.5
Disease Segment	Short segment	90.5	91.4
	Long segment	85.0	87.5
	Total colonic aganglionosis	70.0	80.0

Table 5 presents subgroup analyses of diagnostic accuracy by age group and disease segment. Sensitivity and specificity were highest among children aged 1–12 months (92.9% and 92.3%, respectively), followed by neonates (<1 month) with values of 90% and 91.7%. Children older than 12 months had the lowest diagnostic performance, with a sensitivity of 83.3% and specificity of 87.5%. Diagnostic performance was strongest in short-segment disease (sensitivity 90.5%, specificity 91.4%), while total colonic aganglionosis showed reduced sensitivity (70.0%) and specificity (80.0%).

4. Discussion

This study aimed to assess the diagnostic accuracy of certain findings on a specific barium enema in diagnosing Hirschsprung disease (HSCR) in pediatric patients using histopathological examination as the gold standard. The key analyzers included the transitional zone, rectosigmoid index (RSI) <1, and 24-hour retention film. Although the transitional zone and RSI <1 had higher values, our results were similar to those of existing studies, showing that the 24-hour retention film was the most accurate in terms of diagnostics, followed by the transitional zone and RSI <1.

The results obtained with 24-hour retention film showed a sensitivity of 92.5 percent and specificity of 91.4 percent, and there was an excellent agreement (Kappa = 0.81), suggesting the strong diagnostic reliability of the method. This supports the results of Wong et al., who also recorded a high sensitivity and specificity of delayed evacuation in diagnosing HSCR [17]. In another study conducted by Msomi et al., it was noted that the 24-hour retention film was more reliable than those captured immediately upon evacuation, particularly for long-segment disease detection [20]. These findings support the contention that delayed imaging provides time for the retained contrast to better demarcate aganglionic areas.

The transitional zone was observed in 76 patients, with a sensitivity of 87.3% and specificity of 90.1%. This feature is widely used in diagnosing HSCR but is prone to inter-observer variability. Abbas et al. reported similar accuracy and emphasized the need for experienced radiologists to minimize misinterpretation [21]. Although reliable, transitional zones may be difficult to identify in neonates because of incomplete bowel calibration. Anwar et al. also highlighted the role of radiologist expertise in reducing diagnostic errors [22]. Taxman et al. noted the challenge of identifying the zone in early infancy, when anatomical differences are subtle [23].

The rectosigmoid index (RSI) <1 exhibited the lowest diagnostic accuracy overall, with sensitivity and specificity of 75.9% and 88.7%, respectively. Although it is a commonly used radiologic marker, its utility in neonates is

limited because of the similar diameters of the aganglionic and ganglionic bowel segments, as noted by Reid et al. [18]. In contrast, the RSI showed improved performance in older children, particularly those aged 1–12 months, likely due to better bowel distension and anatomical definition. In our study, the highest diagnostic accuracy was observed in children aged 1–12 months (sensitivity, 92.9 %; specificity, 92.3%), followed by those older than 12 months. Neonates (<1 month) demonstrated lower accuracy, possibly because of less distinct radiologic features in early life. These findings are consistent with those of Noviello et al., who also reported improved radiologic clarity in children beyond the neonatal period [24].

Furthermore, the diagnostic accuracy varied with the length of the aganglionic segment. Barium enema demonstrated the highest accuracy in short-segment disease, where radiologic markers, such as the transitional zone and rectosigmoid index <1, are more readily visible. In long-segment disease and total colonic aganglionosis, these features may be less distinct or absent, leading to reduced sensitivity and specificity of the diagnosis. This observation is consistent with the findings of Rosenfield et al. and Hailemariam et al., who reported similar difficulties in the radiologic diagnosis of extensive aganglionosis using barium enema [4, 5]. These findings highlight the importance of considering segment length in the interpretation of imaging and in choosing complementary diagnostic modalities.

The effects of the technical quality and patient preparation on imaging outcomes are another major factor to consider. As De Arruda Lourencao et al. claim, insufficient bowel cleansing or hypervolume intraluminal air gas can overlie important features like the transitional zone or rectosigmoidal diameter [25]. We have achieved standardization of technique, but there can well be variation in image quality between patients, which is a realistic limitation.

Our findings reiterate that although barium enema is not foolproof, it is a useful, noninvasive diagnostic tool, especially in resource-limited areas. It is most accurate in diagnosis when several features are evaluated simultaneously and analyzed by qualified individuals who understand the age-related anatomy of the bowel.

Although full-thickness rectal biopsy is the gold standard for diagnosis, our results, as well as those of Peyvasteh et al., suggest that barium enema is an effective first-time test for diagnosis [14]. Radiologic diagnosis can be used to prioritize the occurrence of biopsy assists, avoid futile biopsies, and speed up administration in high-risk cases. Further training, standardization of protocols, and the use of auxiliary technologies may increase the reliability and practical value of this tool in clinical practice.

5. Conclusion

This study demonstrates that barium enema remains a valuable diagnostic tool for Hirschsprung's disease in children, especially in settings where access to rectal biopsies is limited. Among the evaluated radiologic features, the 24-hour retention film showed the highest sensitivity, specificity, and diagnostic agreement, followed by the transitional zone imaging. The rectosigmoid index (<1) demonstrated better performance in the 1-12 months age group. While the diagnostic accuracy was reduced in neonates, it was enhanced in older children. Accuracy also remained the highest in short-segment disease. These findings highlight the continued relevance of barium enema as a first-line, noninvasive investigation in the radiologic evaluation of HSCR in children

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Conflicts of Interest

There are no conflicts of interest

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