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Diagnostic Accuracy of Barium Enema for the Diagnosis of Hirsch-sprung Disease in Children

Zahid Anwar, Naveed Ahmed*, Shafqat Rehman, Tamoor Afzal, Javed-Ur-Rehman, Iftikhar Ahmed

Pak Emirates Military Hospital/National University of Medical Sciences (NUMS) Rawalpindi Pakistan, *Combined Military Hospital, Quetta/National University of Medical Sciences (NUMS) Pakistan

ABSTRACT

Objective: To determine the diagnostic accuracy of Barium contrast enema for diagnosis of Hirsch-sprung disease, taking histopathology as the gold standard.

Study Design: Cross-sectional validation study.

Place and Duration of Study: Department of Paediatric Surgery, Pak Emirates Military Hospital, Rawalpindi Pakistan, from Nov 2019 to May 2021.

Methodology: A total of 251 patients with symptoms of Hirsch-sprung disease were included. All patients were assessed with a Barium enema and full-thickness rectal biopsy. Barium enema was reported for the presence of contrast in the rectum after 24 hours, the presence or absence of transitional zone and a Recto Sigmoid Index less than 1. Full-thickness rectal biopsy was taken 2 cm above the dentate line, and histopathology was performed to see the ganglion cells in Meissner`s and Myenteric plexus.

Results: Histo-pathologically confirmed cases were 120(true positive), where 12(false positive) had no Hirsch-sprung disease on histopathology. In Barium enema negative patients, 113 were true negatives while 06 were false negatives. Overall sensitivity, specificity, positive predictive value, negative predictive value and diagnostic accuracy of Barium contrast enema for diagnosis of Hirsch-sprung disease were 95.24%, 90.40%, 90.91%, 94.96% and 92.83%, respectively.

Conclusion: We concluded that the diagnostic accuracy of Barium contrast enema for diagnosis of Hirsch-sprung disease is quite high, taking histopathology as the gold standard.

Keywords: Barium enema, Hirsch-sprung disease, Rectal biopsy.

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INTRODUCTION

The enteric nervous system is responsible for peristaltic waves in the intestines. In Hirsch-sprung disease (HD), ganglion cells in these plexuses are absent. The function of ganglion cells is the facilitation and coordination of bowel relaxation.[1] Their absence results in a spastic bowel resulting in a dynamic intestinal obstruction with absent or abnormal relaxation of the affected bowel and internal anal sphincter.[2] HD is mostly diagnosed in the newborn period if he/she does not pass meconium within 24-48 hours after birth. Other presentations in infancy can be enterocolitis, vomiting, lack of appetite and abdominal distension.[3,4]

To diagnose HD, the gold standard tool is the histopathological examination of rectal biopsy, revealing the absence of ganglion cells in the submucosal and myenteric plexus.[5] In doubtful cases, anorectal manometry and Barium contrast enema (BCE) may be useful.[6,7] Rectal biopsy is associated with various complications like bleeding, scarring, stricture formation and adverse effects related to anaesthesia. Therefore, initially, some non-invasive modalities like imaging methods are used.[8] Sensitivity and specificity of these methods may differ based on the age and length of the involved segment.[9] Anorectal manometry is not available in most settings, whereas BCE is widely available in almost every setting, even in peripheral hospitals.[10]

The study aimed to establish a sensitivity, specificity, positive predictive value (PPV) and negative predictive value (NPV) of the findings of BCE to diagnose HD so that early management can help better prognosis with less morbidity and mortality. In addition, the objective of the study was to determine the diagnostic accuracy of BCE in diagnosing HD taking histopathology as the gold standard.

METHODOLOGY

The cross-sectional validation study was performed at the Department of Paediatric Surgery, Pak Emirates Military Hospital (PEMH), Rawalpindi Pakistan, from November 2019 to May 2021 after approval from the Hospital Ethical Committee (A/28/EC/219/2020 dated 26 Nov 2020). The sample size calculated considering the prevalence of HD of 14%,

Correspondence: Dr Zahid Anwar, Classified Surgeon, Resident Pediatric Surgeon, Pak Emirates Military Hospital Rawalpindi Pakistan,
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the sensitivity of Barium enema at 90%, and specificity of 80%. Sampling Technique was non-probability, consecutive sampling.

Inclusion Criteria: Children of either gender, aged one day to 13 years, with the history of delayed passage of meconium in neonatal age, symptoms of HD like failure to pass meconium, abdominal distension and constipation and patients with chronic constipation refractory to medical treatment were included in the study.

Exclusion Criteria: Patients with a history of anorectal surgery were excluded

We recruited 251 patients, who were assessed with full-thickness rectal biopsy and Barium enema. Rectal biopsy was the full thickness and was taken 2cm above the dentate line. The specimen was sent for histopathological examination to see the presence of ganglion cells in Meissner's and Myenteric plexus. Based on histopathological findings, patients were divided into two groups, i.e., Group-I with no ganglion cells seen and Group-II with ganglion cells seen on histopathology.

BCE was done under the specialized care of a radiologist competent in paediatric contrast procedures. Barium enema was reported for the presence of contrast in the rectum after 24-hours, presence or absence of transitional zone (TZ), RSI, mucosal irregularities (Jejunization), irregular contractions and cobblestone appearance of the rectum and sigmoid colon. In addition, pre-operative findings of these two diagnostic modalities were noted on a specified patient's proforma.

Statistical Package for Social Sciences (SPSS) version 20.0 was used for the data analysis. Quantitative variables were expressed as mean±SD and qualitative variables were expressed as frequency and percentages. For rectal biopsy and Barium enema findings, a two-by-two table was used to determine Sensitivity, Specificity, PPV and NPV.

RESULTS

In this study, the age range was from 1 day to 13 years, with a mean age of 5.84±2.21 years. Of 251 patients, 148(68.96%) were females, and 103(41.04%) were males. All the patients were subjected to BCE, which supported the diagnosis of HD in 132 patients. Histopathology confirmed HD in 120(true positive) cases, whereas 12(False Positive) had no HD on histopathology. In BCE-negative patients, 113 were true-negative, while 6 were false-negative (Table-I).

Table-I: Diagnostic Accuracy of Barium Contrast Enema for the Diagnosis of Hirsch-sprung Disease taking Histopathology as Gold Standard (n=251)

	Positive Result on Histopathology	Negative Result on Histopathology
Positive result on Barium Contrast Enema	120 (True Positive)	12 (False Positive)
Negative result on Barium Contrast Enema	06 (False Negative)	113 (True Negative)

The parameters like sensitivity, specificity, PPV, NPV and diagnostic accuracy of BCE for the diagnosis of HD taking histopathology as the gold standard is 95.24%, 90.40%, 90.91%, 94.96% and 92.83%, respectively, as summarized in Table-II.

Table-II: Summary of Diagnostic Parameters of Barium Contrast Enema (n=251)

Diagnostic Parameters	Values
Sensitivity= True Positive/(True Positive +False Negative)	95.24%
Specificity= True Negative /(True Negative +False Positive)	90.40%
Positive Predictive Value= True Positive/(True Positive+False Positive)	90.91%
Negative Predictive Value= True Negative/(True Negative+False Negative)	94.96%
Diagnostic Accuracy=(True Positive+True Negative)/All Patients	92.83%

DISCUSSION

Hirsch-sprung disease is a cause of constipation due to colonic aganglionosis. This rare disorder is a type of functional bowel obstruction.[11] The involved length of the bowel can vary. In suspected cases, clinicians are always keen to either promptly diagnose or rule out HD because if not identified early and treated promptly, it can lead to significant morbidity. It is generally a combination of different diagnostic modalities to confidently diagnose or rule out HD.[12] However, the gold standard for diagnosis remains the full-thickness rectal biopsy that confirms the absence of ganglion cells.[13] But a rectal biopsy is not free of complications like bleeding, perforation, sepsis and anaesthesia-related risks.[14]

A systematic review conducted in May 2006 has shown that the sensitivity of BCE in excluding HD can be as high as 80% – 88% and reported FN rates are up to 30%.[11] In our study, BCE supported the diagnosis of HD in 132 patients. Histopathology confirmed HD in 120(true positive) cases, whereas 12(False-Positive) had no HD on histopathology. In BCE-negative patients,

113 were true-negative, while 06 were false-negative. Overall sensitivity, specificity, PPV, NPV and diagnostic accuracy of BCE for the diagnosis of HD taking histopathology as the gold standard is 95.24%, 90.40%, 90.91%, 94.96% and 92.83%, respectively.

A study was conducted by Reid *et al.* in which BCE was compared with anorectal manometry and rectal biopsy to exclude HD after the neonatal period. All the features of BCE, like TZ, RSI, irregular contractions and mucosal irregularity in terms of sensitivity, specificity, PPV, and NPV, were analysed. This study had values comparable to ours in terms of TZ, and the values in the rest of the three factors were inferior to ours. The logical explanation for this can be that we did not individually analyse the different features of BCE but analysed it as a whole.[15] A study by Richard Skaba demonstrated that out of the four features of BCE mentioned above, TZ and RSI are the most common signs. In older children, some other factors may also come into play to predict the sensitivity and specificity of BCE to exclude HD. Those factors are age, gender and duration of disease.[16]

Shankar *et al.* conducted a study in two centres and found that the sensitivity and specificity of BCE to diagnose HD has a wide range, between 60% to 100%.[17] These findings are in contrast to ours. The reasons can be much fold like the techniques used, selection of patients and interpreting skills of the radiologist.

Proctor *et al.* demonstrated the correlation between TZ on BCE and the level of aganglionosis in HD, both for short and long-segment disease. The sensitivity and specificity of BCE for short-segment HD were 92% and 75%, respectively, and for long-segment values, 63% and 100% for long-segment disease, respectively.[18] A study conducted in Nigeria analysed different features of BCE suggestive of HD, and it was concluded that out of four features, TZ and RSI are more common in HD. Out of these two, the TZ was considered pathognomonic for HD.[19] In our study, different features of BCE suggestive of HD were not separately assessed.

However, in our study, we did not stratify the different features of BCE as our main aim was the comparison of BCE with rectal biopsy to prove its usefulness in diagnosing HD early.

CONCLUSION

This study concluded that the diagnostic accuracy of BCE for the diagnosis of HD is quite high. This modality can enable us to diagnose or rule out HD even in remote settings, leading to an early referral and prompt intervention resulting in decreased morbidity and mortality. Therefore, we recommend that BCE be used routinely as a prime method for diagnosing HD in order to reduce the rate of negative rectal biopsies.

Conflict of Interest: None.

Author's Contribution

Following authors have made substantial contributions to the manuscript as under:

ZA & NA: Study design, drafting the manuscript, data interpretation, critical review, approval of the final version to be published.

SR & TA: Critical review, drafting the manuscript, approval of the final version to be published.

JR & IA: Data acquisition, Conception, data analysis, approval of the final version to be published.

Authors agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

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