

# Use Of Barium Enema In Neonates With Suspected Hirschsprung Disease: A Narrative Review

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## Abstract

**Background:** Hirschsprung's disease (HD) is one of the most prevalent causes of intestinal obstruction in children. Although there are several ways to diagnose Hirschsprung's disease, barium enema is the most common first diagnostic approach. During the first month of life, radiographic pictures of the colon obtained with a barium enema frequently appear normal, and in patients with complete colonic involvement, they may remain normal indefinitely.

**Methodology:** To diagnose Hirschsprung's disease in neonatal patients who have received a barium enema, this review looks at recent updates on barium enema in the diagnosis of Hirschsprung's disease in neonates. A search was made on Google Scholar, PubMed, Medline, Hinari, ResearchGate, ScienceDirect, using 'barium enema', 'Hirschsprung's disease' and 'neonates' as search terms from 2005 to 2025. Retrieved data were entered into an

online Google form and transferred to an Excel sheet. A review of relevant full articles was subsequently done. Articles not published in English Language, those published outside the neonatal period and in adults were excluded.

**Results:** The rectosigmoid index is a clear way to aid in the diagnosis of rectosigmoid Hirschsprung disease in newborns, especially when the radiographic images are unclear or hard to read. The presence of recto-sigmoid index and three radiographic signs - rectosigmoid transition zone, barium retention, and stool mixed with barium - strongly indicates the condition.

**Conclusion:** Barium enema is an easy way to diagnose Hirschsprung's disease in neonates particularly in resource poor settings.

**Keywords:** Barium enema, Hirschsprung disease, neonates

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## Introduction

A common aetiology of intestinal obstruction in paediatric patients is Hirschsprung's disease (HD).<sup>1</sup> Hirschsprung's disease is the predominant aetiology of lower intestine obstruction in neonates and babies with large bowel involvement<sup>2</sup>. In full-term infants who are otherwise healthy, it usually manifests as an isolated disease<sup>3</sup>. With a male-to-female ratio of 4:1, the illness is estimated to affect 1 in 5,000 live newborns<sup>4</sup>. According to a Nigerian study, the disease is responsible for roughly 13.85% of juvenile intestinal obstruction cases<sup>5</sup>. Hirschsprung's disease is a condition characterized by the absence of ganglion cells in specific colon areas, causing the enteric nervous system to malfunction due to abnormal development.<sup>6</sup>

Many theories have been suggested to explain how Hirschsprung's disease starts, but the most widely accepted one is that by Okamoto (1967) who proposed a problem with the movement of neuroblasts from the head to the tail of the body during the first 10 weeks of pregnancy<sup>7</sup>. Functional colonic blockage results from the distal colon's varying lengths failing to relax. Consequently, the affected segment(s) fail(s) to relax and hence, obstructs gut motility<sup>8</sup>. Although the entire colon and, rarely, the small intestine may be affected, Hirschsprung's disease primarily affects the rectosigmoid region<sup>9</sup>. Usually beginning at the anus, the aganglionic portion extends proximally<sup>6</sup>. In the typical rectosigmoid type of Hirschsprung's disease, there are four boys for every girl affected, but in the more severe cases, the number of boys compared to girls is less.<sup>10</sup>

Constipation is a prevalent issue among children. Despite being frequently overlooked and underestimated, it can

result in significant discomfort and dysfunction. Organic conditions such as Hirschsprung disease impact about 10% of children who have defecation problems<sup>11</sup>. However, it can be difficult to differentiate between idiopathic constipation (IC) and Hirschsprung disease (HD) in newborns who arrive late<sup>12</sup>. A delayed passage of meconium is a characteristic of the majority of HD cases.<sup>13</sup> According to one report there is a large range in the incidence of this symptom, from 60% to 94%<sup>14</sup>. However, Other reports indicate that this symptom is present in only 65% of HD patients and about 13% of IC patients, suggesting that it may not be a reliable way to distinguish HD from IC<sup>15</sup>. Therefore, finding HD early is important since surgery should be done soon after birth to avoid problems that may arise if the disease is not treated. Furthermore, infants are typically more amenable to surgical procedures from a technical standpoint<sup>16</sup>. In an effort to reduce the need for rectal biopsies—which are occasionally needless and not always risk-free, several studies have reported other non-invasive methods of diagnosing Hirschsprung disease. Efforts have been made to assess the sensitivity and specificity of various clinical data and non-invasive diagnostics. The fact that only 12–17% of biopsies performed on constipated patients revealed a positive result for aganglionosis lends credence to this theory<sup>17</sup>. Additionally, up to 80% of these biopsies may not be necessary<sup>18</sup>. Accordingly, in neonates with low small intestinal obstruction, a preliminary barium enema should be the first non-invasive diagnostic method before thinking about more invasive abdominal exploration. Evidence has also shown that operating on newborns who have never had a barium enema is a significant error when it comes to treating Hirschsprung disease<sup>19,20</sup>. Also,

how long the barium stays in the body is important to check before surgery, as a 24-hour delay in barium retention can also indicate Hirschsprung disease. De Lorijn stated that the details of interpreting a barium enema are just as important as anorectal manometry and rectal biopsies<sup>21</sup>. A barium enema's accessibility is another favorable advantage to its usage<sup>19</sup>. A barium enema is a type of x-ray procedure that specifically involves the use of a contrast agent. A contrast enema uses a special liquid that has barium or iodine to highlight the large intestine, colon, and rectum; this x-ray test of the lower digestive system is called a barium enema. Barium sulphate is administered into the colon to render the large intestine, including the rectum, visible in X-ray images. Clearer imaging is made possible by barium's enhancement of specific bodily parts<sup>22</sup>. The barium enema, which shows the area where the normal wide intestine meets the narrow, non-functioning gut, is the first choice for diagnosing Hirschsprung disease. About 70 to 90 percent of cases show this transition zone<sup>23</sup>. The intestinal segment above the problematic location usually exhibits obstruction-related symptoms, such as thickening and distention. When dilatation takes place, the proximal colon gets dilated, but the afflicted section of the colon may appear normal. Besides finding a transitional zone, a low rectosigmoid index (which is the size of the rectum compared to the size of the sigmoid colon), a delay in getting rid of leftover barium after 24 hours, and having leftover barium all indicate that there may be a disease<sup>24</sup>. However, as the transition zone between normal and aberrant ganglion cells may not be fully established in neonates, its usefulness may be restricted. It is crucial to note that the colon will not enlarge and expand in suspected

neonatal cases of Hirschsprung disease until the intestines have been operating normally for at least three months<sup>25</sup>. Barium enema investigations may show no transition zone in the large intestine in these cases<sup>26</sup>. Additionally, a transition zone will not exist in newborns with total colon aganglionosis, which is a sign that a rectal biopsy is necessary<sup>27</sup>. The diagnosis should usually be made in the first few days or years of a child's life. Up to 90% of patients with Hirschsprung disease can be diagnosed by the time they are one month old, and the majority are recognized by the time they are one year old if they are not discovered during the neonatal stage<sup>13</sup>. Radiographic contrast tests could help with the first diagnosis. There are reports indicating that patients suspected of having Hirschsprung disease should be first evaluated using a barium enema due to its non-invasiveness, availability and accuracy.

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This review aims to examine the literature on the value of radiologic findings for diagnosing Hirschsprung disease in neonatal patients who have received a barium enema. Since the results of a contrast enema can appear normal in both total colonic Hirschsprung disease and Hirschsprung disease in the first three months of life, the goal is to assess how useful this procedure is for newborns who are suspected of having Hirschsprung disease.

### Methodology

A search of the following databases including Google Scholar, PubMed, Medline, ResearchGate, ScienceDirect between 2005 and 2025 was done using the following barium enema, neonates, newborn, and Hirschsprung disease as search terms. The data were stored in a google form.

## Results

A total of 30 articles were retrieved but only 15 met the inclusion criteria.

Zhou et al, study enrolled 58 patients and found that the 24-hour delayed film of barium enema was crucial for predicting neuropathological segments in patients with HSCR. The barium retention level also contributed to the decision-making of surgical physicians. The Darmajaya et. al. study found that barium enema can be an early diagnostic tool for infants suspected of Hirschsprung's disease. In Darmajaya study -all the patients reviewed, sensitivity, specificity, positive and negative predictive value of barium enema for diagnostic of Hirschsprung's disease was 95.5%, 87.5%, 97.7%, and 77.8%. This was similar results to the above studies. More than 75 percent of the articles concluded that a barium enema may serve as a first diagnostic instrument for infants suspected of having Hirschsprung's disease.<sup>28-36</sup>

All the studies concluded that barium enema X-ray is a trustworthy and accurate test for Hirschsprung disease (HD) in newborns, helping to show how much of the intestine is affected, especially in cases of rectosigmoid HD.<sup>37</sup>

Seeing the radiological transition zone in a barium enema for newborns is a good sign of Hirschsprung's disease, but not seeing it does not mean the disease is ruled out.<sup>38</sup>

The rectosigmoid index is a clear way to help diagnose rectosigmoid Hirschsprung disease in newborns, especially when the radiological transition zone is unclear or hard to see. The retention of barium at 24 hours following the late film is inadequate for diagnosing HD.<sup>39</sup>

The 24-hour evacuation of barium does not conclusively rule out the possibility of HD. The combination of three radiographic

signs (rectosigmoid transition zone, barium retention, and stool mixed with barium) was better at showing the presence of HD than any one sign by itself.<sup>32</sup>

## Discussion

The results of this study using barium enema validated the subsequent facts: The barium enema X-ray is a reliable and precise diagnostic tool for Hirschsprung disease (HD) in neonates, effectively illustrating the extent of intestinal involvement, particularly in instances with rectosigmoid HD.

The presence of the radiological transition zone in a barium enema for neonates is indicative of Hirschsprung's disease; nevertheless, its absence does not exclude the diagnosis.<sup>35</sup>

The rectosigmoid index is an effective tool for diagnosing rectosigmoid Hirschsprung disease in neonates, particularly when the radiological transition zone is ambiguous or difficult to discern. The retention of barium at 24 hours post-late film is insufficient for diagnosing HD<sup>36</sup>. The findings of this research indicate that diagnosing gastrointestinal issues in neonates is complex due to the potential for misinterpretation caused by residual barium.

The 24-hour barium evacuation does not definitively exclude the potential for HD<sup>32</sup>. The amalgamation of three X-ray indicators (rectosigmoid transition zone, barium retention, and stool mixed with barium) demonstrated superior efficacy in indicating the presence of HD compared to any individual sign alone. A comparison of radiographs obtained 24- and 48-hours post-evacuation may aid in differentiating Hirschsprung's disease from meconium plug syndrome. Distinguishing between

these two disorders is crucial for appropriate therapy, as Hirschsprung's disease typically necessitates surgical intervention, although meconium plug syndrome can frequently be managed non-surgically<sup>26</sup>. Mahboubi et al. also examined rectal manometry and barium enema tests in relation to Hirschsprung disease. Their research determined that if both the barium enema and manometry tests confirm Hirschsprung's illness, a rectal biopsy is unnecessary prior to surgery, as the tests are likely to accurately diagnose the condition<sup>40</sup>. A prompt and precise diagnosis can profoundly influence patient outcomes and diminish the likelihood of complications. Comprehending the magnitude of the impacted area is essential for devising suitable surgical intervention. Timely and precise diagnosis using techniques like the barium enema can greatly enhance prognoses for infants afflicted with Hirschsprung disease. Techniques such as barium enemas enhance diagnostic accuracy, facilitating prompt interventions that improve the quality of life for affected newborns and promote healthier futures. It is important to note that in the context of Hirschsprung-associated enterocolitis, the application of barium enema was found to have restricted clinical value and presented hazards, especially in the case of perforation in neonates<sup>41</sup>.

All of the aforementioned investigations demonstrate that anorectal manometry and barium enema have comparable sensitivity and specificity. To investigate suspected Hirschsprung disease, a contrast enema proved helpful; nevertheless, a negative study does not rule out the problem. They all concluded that the rectal suction biopsy was the most accurate test for diagnosing HD, and those with indeterminate findings would benefit from

a lower threshold to perform follow-up rectal biopsy in clinically suspect newborns for HD. A rectal biopsy, however, might not be required prior to surgery if both studies (barium enema and manometry) show the condition because there is a strong likelihood that both will be positive. But due to the unavailability of anorectal manometry in many underdeveloped countries and the availability of barium enemas, this exam should be considered the first-line imaging in a neonate suspected to have Hirschsprung disease. Also, the rectosigmoid index should be routinely used in neonates if the transition zone is not obvious.

### Conclusion

Barium enema is an easy way to diagnose Hirschsprung's disease in neonates particularly in resource poor settings.

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