
ORIGINAL ARTICLE

Diagnostic Accuracy of Lateral Abdominal Radiographs among Paediatric Patients in Detecting Hirschsprung Disease

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ABSTRACT

Purpose: The aim of this study was to determine the diagnostic accuracy of lateral abdominal radiographs in the detection of Hirschsprung disease (HSCR) in paediatric patients compared with rectal biopsy as the standard.

Methods: This retrospective study was performed in cases with clinical suspicion of HSCR that had undergone lateral or cross-table prone lateral abdominal radiographs and subsequent rectal biopsies at a tertiary hospital in Metro Manila, Philippines. Reversal of the rectosigmoid index (RSI) on a lateral abdominal radiograph without or with a transition zone was interpreted as positive for HSCR. The radiographic findings were correlated with histopathology.

Results: A total of 72 cases were included. Age ranged from 4 days to 12 years with a mean of 16 months. In all, 62 cases (86%) were positive for HSCR on biopsy, 39 of which had reversed RSI. Ten patients (14%) were negative for HSCR on biopsy, seven of which showed no reversal of the RSI. Overall, the diagnostic accuracy of the lateral abdominal radiograph was 64%. Sensitivity, specificity, positive predictive value, and negative predictive value were 63%, 70%, 93%, and 23%, respectively.

Conclusion: The presence of a reversed RSI on a lateral or cross-table prone lateral abdominal radiograph had a high positive predictive value, indicating a high likelihood of HSCR. Therefore, such views are reliable tools in predicting HSCR in infants and young children. Rectal biopsy should be done for confirmation but patients may no longer need to undergo contrast enema.

Key Words: Biopsy; Constipation; Enema; Hirschsprung disease; Radiology, abdominal

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Data Availability: All data generated or analysed during the present study are available from the corresponding author on reasonable request.

Ethics Approval: This study protocol was approved by the Institutional Scientific and Ethics Review Committees of the Research and Biotechnology Division of the hospital (#CT-18104). The need for informed consent was waived owing to the retrospective nature of the study.

中文摘要

兒科患者腹側位X線片診斷先天性巨結腸的診斷準確性

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目的：與作為標準的直腸活檢相比，本研究確定側位腹部X線片在兒科患者先天性巨結腸的診斷準確性。

方法：這項回顧性研究於菲律賓馬尼拉大都會一家三級醫院進行，對臨床懷疑巨結腸的病例接受側位或橫台俯臥側腹位X線片和隨後的直腸活檢。側位腹部X光片上沒有或有過渡帶的直腸乙狀結腸指數 (rectosigmoid index; RSI) 的反轉解釋為巨結腸陽性。進行放射學檢查結果與組織病理學相關性的分析。

結果：共納入72例。年齡從4天到12歲不等，平均16個月。總共有62例 (86%) 活檢巨結腸陽性，其中39例見RSI反轉。10名患者 (14%) 活檢巨結腸為陰性，其中7名患者的RSI沒有反轉。總體而言，側位腹部X線片的診斷準確率為64%。敏感性、特異性、陽性預測值和陰性預測值分別為63%、70%、93%和23%。

結論：側位或橫台俯臥側腹位X線片顯示RSI反轉具有很高的陽性預測值，表明巨結腸的可能性很高。因此，這些表現是預測嬰幼兒巨結腸的可靠工具。應進行直腸活檢以確認，但患者可能不再需要接受造影劑灌腸。

INTRODUCTION

Hirschsprung disease (HSCR) is the most common cause of paediatric intestinal obstruction.¹ It results from failure of normal bowel innervation due to the arrest of proximal-to-distal migration of vagal neural crest cells, with a variable length of distal bowel lacking parasympathetic Auerbach (intermuscular) and Meissner (submucosal) plexuses.² There is resultant colonic aganglionosis leading to symptoms such as abdominal distension, irregular bowel movements, or chronic constipation.

HSCR occurs in approximately 1 per 5000 live births with a male-to-female ratio of 4:1.¹ It is responsible for approximately 15% to 20% of cases of neonatal bowel obstruction, presenting in the newborn period in approximately 80% of cases.²

Initial diagnosis is mainly based on clinical history and examination,¹ but approaches to diagnoses include abdominal radiography, contrast enema, anorectal manometry (which is not always readily available), and rectal biopsy.³

Patients may present with failure to pass meconium within the first 24 to 48 hours of life or may subsequently develop

chronic constipation. Intestinal obstruction may ensue, causing abdominal distention, poor feeding, and poor weight gain. Plain abdominal radiographs are routinely taken as the initial imaging study in the assessment of these patients, oftentimes in anteroposterior upright and supine views with or without a lateral view.

Contrast enema is a valuable tool in the diagnosis of HSCR,³ with an accuracy ranging from 80% to 94%,⁴ sensitivity of 65% to 80% and specificity of 66% to 100%⁵ when compared with anorectal manometry and rectal biopsy. Common findings in contrast enema for HSCR include reversal of the rectosigmoid index or the presence of a transitional zone (RSI: the ratio of the anteroposterior diameter of the rectum at the level of the 2nd sacral vertebral body to the anteroposterior diameter of the sigmoid colon measured at the level of the 1st sacral vertebral body on the lateral view. It is considered normal if the RSI is ≥ 1 [Figure 1], meaning the rectal diameter is wider or at least equal to that of the sigmoid colon. If RSI is < 1 , it is considered abnormal [Figure 2]. A transitional zone is a severe form of reversed RSI showing an abrupt change in colonic calibre with shouldering), delay in barium evacuation > 24 hours after birth, jejunitis of the colon, a cobblestone appearance of the aganglionic segment, and irregular contraction.¹



Figure 1. Lateral abdominal view of a 3-month-old boy showing a normal rectosigmoid index (>1) where the rectal air column (R) is wider than the sigmoid (S) colon.

Although contrast enema is minimally invasive, it causes unnecessary stress and discomfort not only for the patients but also for the parents and/or caretakers.

The gold standard of diagnosis for HSCR remains rectal biopsy, which will demonstrate absence of ganglion cells in the submucosal layer distal to the transitional zone as well as presence of acetylcholinesterase-positive hypertrophic nerve fibres.³

Our primary objective was to assess the diagnostic accuracy of plain lateral abdominal radiography in the diagnosis of HSCR, with the results of the rectal biopsy used as the reference standard. To the best of our knowledge, published studies on plain lateral abdominal radiography in the diagnosis of HSCR as correlated with rectal biopsy are scarce.

METHODS

The study was a retrospective study of paediatric patients, aged ≤ 5 years, suspected of having HSCR, who underwent abdominal radiographs with lateral or cross-table prone lateral views, as a stand-alone study

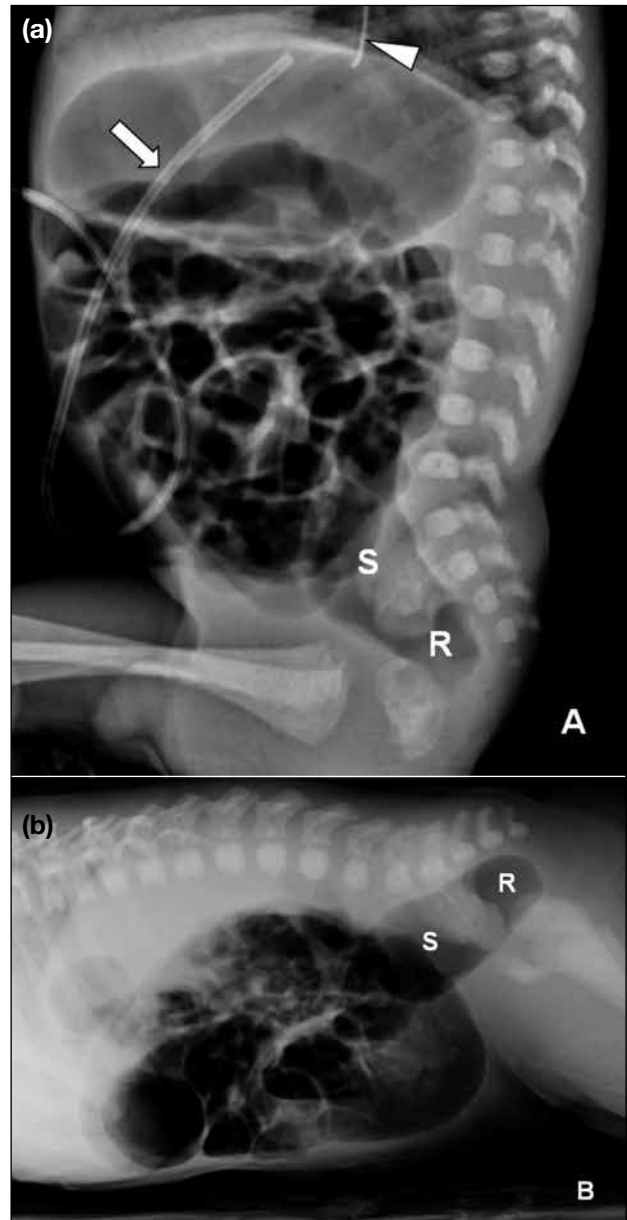


Figure 2. (a) Lateral abdominal radiograph of a 4-day old neonate, and (b) cross-table prone lateral view of a 14-day old infant; both show a reversed rectosigmoid index (<1) where the rectum (R) is narrower than the sigmoid (S) colon. Also seen in (a) are an umbilical venous catheter (arrow) and the tip of an orogastric tube (arrowhead).

or as scout views prior to a barium or water-soluble contrast enema, and subsequent rectal biopsy at St. Luke's Medical Center–Quezon City from January 2011 to December 2020 and at St. Luke's Medical Center–Global City from January 2010 to December 2020.

Clinical data and histopathological results were retrieved from the hospital's HealthCare System. Demographic

information such as age and gender were recorded. Clinical suspicion of HSCR was due to the presence of abdominal distention, delayed passage of meconium, and/or constipation.

The digital radiographs were reviewed via the Picture Archiving and Communication System. The lateral views or cross-table prone lateral views were used for evaluation. The presence of an abnormal RSI (<1) with or without a transitional zone was considered a positive reading. Evaluation of the digital images was done by a single paediatric radiologist of more than 12 years' experience, who was blinded on case information and rectal biopsy findings.

Patients who had undergone prior gastrointestinal surgery for any reason and those who had a cross-table supine lateral view were excluded from this study.

The rectal biopsy results were interpreted as positive if the histopathological report mentioned any of the following findings: absence of ganglion cells/aganglionosis, immature ganglion cells, or hypoganglionosis. Absence of these findings or an inconclusive report was considered negative.

The findings from the lateral radiographs were then correlated with the rectal biopsy results using the four-fold table technique (Table) to compute for the accuracy, sensitivity, specificity, positive predictive value, and negative predictive value. To determine the relationship between patient age and gender with radiographic findings and biopsy results, independent sample *t* tests and Fisher's exact test (with a Chi-square test for small samples) at a 5% level of significance, were utilised.

RESULTS

A total of 72 paediatric cases clinically suspected of having HSCR were included in the study. In all, 46 were male and 26 were female, with a male-to-female ratio of 1.8:1, with age ranging from 4 days to 12 years old (mean age 16.09 ± 32.16 mo). Those with positive lateral radiographs had a mean age of 12.17 ± 32.05 months; negative lateral radiographs being 21.59 ± 32.02 months; positive rectal biopsy being 11.20 ± 23.68 months; and negative rectal biopsy being 32.88 ± 48.47 months. The differences in age and gender in relation to the radiographic findings and biopsy results were not statistically significant.

In total, 62 patients (86%) were confirmed positive for

Table. Relationships between radiographic findings (test) and biopsy results (gold standard).*

| Radiographic finding | Biopsy results | | Total |
|----------------------|----------------|----------|-----------|
| | Positive | Negative | |
| Positive | 39 (54%) | 3 (4%) | 42 (58%) |
| Negative | 23 (32%) | 7 (10%) | 30 (42%) |
| Total | 62 (86%) | 10 (14%) | 72 (100%) |

* Data are shown as No. (%).

HSCR on rectal biopsy, 39 of which were also positive on the lateral abdominal radiographs. Ten patients (14%) were negative for HSCR on biopsy, seven of which were likewise negative on the radiographs (Table).

In using lateral abdominal radiography to detect HSCR, its sensitivity and specificity were computed to be 63% and 70%, respectively. The positive predictive value was high at 93%, while the negative predictive value was only 23%. Overall accuracy was 64%.

DISCUSSION

A plain (non-contrast) abdominal radiograph is the initial study for patients manifesting with abdominal distention, failure to pass meconium in 24 to 48 hours after birth, and/or constipation. This is routinely taken in anteroposterior supine and upright views, along with a lateral view. Additional cross-table prone or supine lateral views may also be included in patients with possible intestinal obstruction and to better visualise the rectosigmoid air column.⁶⁻⁹ These are easily done, taking less than 5 minutes, with minimal radiation exposure, and are also relatively cheap and ubiquitous.

A contrast enema using either barium solution or water-soluble iodinated contrast material has 80% to 94% accuracy⁴ in detecting HSCR, more accurate than a lateral plain abdominal study (56%). Most common findings appreciated were presence of a transitional zone and a reversed RSI.^{1,4,5,10,11} However, this procedure is more invasive because it necessitates insertion of a rectal tube.¹² It is likewise more expensive, takes more time, and involves some ionising radiation exposure.

A study by Pratap et al in 2007⁴ compared the plain abdominal radiograph transitional zone (PARTZ) with the contrast enema transitional zone (CETZ) in diagnosing HSCR. Their study concluded that PARTZ and CETZ matched with the level of the transitional zone in 22 (92%) and 13 (72%) patients, respectively.⁴

PARTZ was said to be better than CETZ in predicting the level of the transitional zone in cases of inconclusive contrast enema.⁴

This study obtained an overall diagnostic accuracy of 64% with sensitivity of 63%, specificity of 70% and negative predictive value of 23% in predicting HSCR using the lateral abdominal radiograph, which is comparably low relative to contrast enema and rectal biopsy. This is actually not unusual because decreased or non-visualisation of the rectosigmoid air column is common in patients without bowel preparation, due to presence of stool and/or fluid within the distal colonic segments. It may even be empty and thus physiologically collapsed. The positive predictive value, however, was very high at 93%, indicating that detection of a reversed RSI (<1) without or with a transitional zone on a plain lateral or cross-table prone lateral radiograph correlates with high likelihood that the patient has HSCR. Age and gender were non-contributing factors.

The authors recognise some limitations in the methodology. This was a retrospective study and thus the radiological technique was not fully controlled resulting in non-uniform image quality. Moreover, both lateral and cross-table prone lateral views were taken into consideration, and the latter view is considered more superior in the visualisation of the rectosigmoid colon. It is therefore the recommendation of the authors that a prospective study be done using standardised imaging parameters in acquiring only the cross-table prone lateral view. Lastly, the number of true negatives is limited because it is rare for surgeons to perform a biopsy on patients not suspected of having HSCR.

In conclusion, the diagnostic accuracy of the lateral abdominal radiograph in the detection of HSCR among paediatric patients ≤ 12 years prior to rectal biopsy for suspected HSCR is only 64%. However, there was a high positive predictive value of 93% which means that if a reversed RSI (<1) is appreciated on the lateral abdominal radiograph, there is also a high likelihood that the patient has HSCR disease. Therefore, a lateral or cross-table

prone lateral abdominal radiograph that demonstrates a reversed RSI is a reliable view in the detection of HSCR in infants and young children. Rectal biopsy may be done for confirmation but patients may no longer need to undergo contrast enema.

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