www.jmscr.igmpublication.org
Impact Factor 5.84
Index Copernicus Value: 71.58
ISSN (e)-2347-176x ISSN (p) 2455-0450
crossref DOI: https://dx.doi.org/10.18535/jmscr/v5i9.41

IGM Publication
Journal Of Medical Science And Clinical Research
An Official Publication Of IGM Publication

Original Research Article

A Prospective Observational Study to Evaluate the Cases of Suspicious Hirschsprung's Disease in Neonates and Children Using Radiologic Investigation Method

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ABSTRACT

Background: *Hirschsprung's disease (HD) is a congenital intestinal disorder characterised by aganglionosis of the distal bowel. It is a common cause of pediatric intestinal obstruction.*

Aims & Objectives: *A standard contrast enema for HD can sometimes be inconclusive in delineating a transition zone especially in neonates and infants. The aim of this study was to determine the utility and diagnostic accuracy of different radiologic methods to evaluate a checklist of radiologic and clinical signs to determine the probability of HD in suspicious patients.*

Methods: *In a diagnostic accuracy study, 22 children with clinical manifestations of suspected HD attended pediatric OPD in a tertiary care teaching hospital, Indore from June 2015 to June 2017 were assessed. A checklist was used to evaluate the items proposed by contrast enema (CE), based on six subscales, including transitional zone, rectosigmoid index (RSI), irregular contractions in aganglionic region, cobblestone appearance, filling defect due to fecaloid materials and lack of meconium defecation during the first 48 hours after birth. The patients were classified as high score and low score. Sensitivity and specificity were calculated for identifying HD, in comparison with pathologically proved or ruled out HD.*

Results: *Of the 22 patients, 14 (63.6%) cases had HD and 08 (36.4%) cases were without HD. The mean age was 4.68 ± 5.24 months. Abdominal distension, constipation and lack of meconium defecation were the most common clinical symptoms with frequencies of 18 (81.82%), 15 (68.18%) and 13 (59.09%) respectively. In summary, the mean sensitivity of detecting the radiological signs of transition zone, spastic colon, reversed recto-sigmoid index and the overall impression in histological confirmed HD patients are 67.86%, 53.75%, 60.72% and 60.77% respectively. The mean specificity of detecting the absence of the radiological signs of transition zone, spastic colon, reversed recto-sigmoid index and the overall impression*

in histological confirmed non-HD patients are 68.75%, 81.25%, 87.5% and 79.17% respectively. This would in turn give an overall mean specificity rate of 79.17% in successfully excluding HD with the above mentioned radiological signs from the contrast enema.

Conclusion: *In conclusion, our study underscores the importance of combining the information of a transition zone on a plain abdominal radiograph and contrast enema to decide the surgical approach for the correction of Hirschsprung's in developing countries where laparoscopic facilities are not available.*

Keywords: *Hirschsprung's Disease, Neonate, Children, Constipation, Diagnosis, Radiological investigation, Contrast Enema, Sensitivity, Specificity*

INTRODUCTION

Hirschsprung's disease (HD) is a congenital intestinal disorder characterised by aganglionosis of the distal bowel.[1] The clinical symptoms of HD may include delayed passage of meconium beyond 24 hours of life, signs and symptoms of large bowel obstruction such as bilious vomiting, a distended abdomen, feeding intolerance, failure to thrive, severe defaecation problems and enterocolitis.[2]

It is a functional intestinal obstruction that results from the congenital deficiency of the normal myenteric plexus parasympathetic ganglion cells in the distal portion of the large intestine. It remains the most common condition to cause low functional intestinal obstruction in children. It is mostly identifiable at birth, but often presents later, particularly in resource-challenged environments.[3, 4]

Although the exact worldwide incidence is unknown, international studies have reported rates ranging from approximately 1 case per 1500-7000 newborns. This disease occurs more often in males than in females, with a male-to-female ratio of approximately 4:1; however, the ratio in long-segment disease decreases to 2:1.Hirschsprung disease affects all races; however, it is roughly 3 times more common among Asian-Americans.[5, 6]A funnel shaped transition zone on a contrast enema (CETZ) at the junction of aganglionic and ganglionic gut is considered a hallmark for its diagnosis.[7, 8, 9]Knowledge of the extent of aganglionic bowel on contrast enema is important for preoperative planning of trans-anal surgery.[10] HD is accepted as being a sex-linked heterogonous disorder with differing phenotypic expressions and degrees of severity.[11]

Most cases of Hirschsprung disease are diagnosed in the newborn period. Hirschsprung disease should be considered in any newborn that fails to pass meconium within 24-48 hours of birth. It results in delayed meconium defecation, abdominal distension, lack of appetite, vomiting, and enterocolitis.

Several diagnostic tests have been described for patients whom HD is suspected such as demonstration of the absence of rectoanal inhibitory reflex (RAIR) in anorectal manometry and an elevated cholinesterase activity and aganglionosis in rectal suction biopsy. However, these tests can sometimes be expensive and invasive, and specialized equipments may also be required if the test is to be performed on a small size infant or even neonate. As a relatively non-invasive procedure, the use of contrast enema (CE) is sometimes a favorable diagnostic option for suspected HD in infants. A systemic review by De Lorijin et al, had quoted a sensitivity rate of 70% and a specificity rate of 83% in using CE as the initial diagnostic test for the workup of HD. However, the evaluation of the CE can often be subjective and the sensitivity of diagnosis of HD from a CE may sometimes be observer dependent.[12]

Contrast enema is useful in establishing the diagnosis, but full-thickness rectal biopsy remains the criterion standard. Once the diagnosis is confirmed, the definitive treatment is to remove aganglionic bowel and to restore continuity of the healthy bowel with the distal rectum, with or without an initial intestinal diversion.[13,14,15] Currently, approximately 90% of patients with HD are diagnosed in the newborn period.[16] Approximately 1% of patients with Hirschsprung

disease have debilitating incontinence requiring a permanent colostomy.[16]

The length of the aganglionic segment anatomically divides HD into 4 types [17]–

- Short segment HD: ~ 75%, rectal and distal sigmoid colonic involvement only
- Long segment HD: ~ 15%, typically extends to splenic flexure / transverse colon
- Total colonic aganglionosis: 2-13%, also known as Zuezler – Wilson syndrome; occasional extension of aganglionosis into small bowel
- Ultra-short segment HD: 3- 4 cm of internal anal sphincter only, and controversial entity

Although the initial diagnosis is mainly based on clinical history and examination, followed only afterwards by pathological assessment[18], radiographic contrast assessment may be useful in diagnosis.[4,18] A plain radiography may demonstrate a transition zone between a gas-filled colon and a non-dilated proximal colon, as a funnel-shaped region. Contrast enema (CE) with barium may be the first imaging procedure performed in the majority of centers, showing a transition zone, irregular colonic contractions, irregular mucosa suggesting enterocolitis, and an abnormal rectosigmoid index (RSI).[19] These methods may have different sensitivity and specificity, based on age and length of involved region.[20, 21]

The gold standard of diagnosis is the pathological evaluation, revealing the absence of ganglion cells in the submucosal and myentric plexus, resulting in aperistaltism in the affected enteric regions and functional intestinal obstruction. Although the initial diagnosis is mainly based on clinical history and examination, followed only afterwards by pathological assessment[18], radiographic contrast assessment may be useful in diagnosis.[4] The risks associated with rectal biopsy, such as perforation scar, stricture, bleeding and also anesthesia-related adverse effects, results in seeking several non-invasive techniques, such as imaging methods.[22,23]

A plain radiography may demonstrate a transition zone between a gas-filled colon and a non-dilated proximal colon, as a funnel-shaped region [Figure 1]. Contrast enema (CE) with barium may be the first imaging procedure performed in the majority of centers, showing a transition zone, irregular colonic contractions, irregular mucosa suggesting enterocolitis, and an abnormal rectosigmoid index (RSI).[20] These methods may have different sensitivity and specificity, based on age and length of involved region.[22,24,25] Regarding the availability and feasibility of CE in most centers, and considering the role of prompt diagnosis and treatment of disease for the prevention of mis-diagnoses and complications, the use of a non-invasive method, such as CE, would help the physicians to achieve a better management of the disease.

Regarding the availability and feasibility of CE and considering the role of prompt diagnosis and treatment of disease for the prevention of misdiagnoses and complications, the use of a non-invasive method, such as CE, would help the physicians or pediatric surgeons to achieve a better management of the disease. The objective of this study was to review the diagnostic accuracy of different radiological investigations like X-ray and contrast enema in neonates, infants and children with suspected HD and to investigate the potential concordant rate among different radiologists in the interpretation of the CE and erect X-ray radiographs.

MATERIALS AND METHODS

From June 2015 to June 2017, neonates and infants, and children with clinical suspicion of Hirschsprung's disease were enrolled in this prospective observational study that was approved by the institutional ethics committee of a tertiary care teaching hospital, Indore.

Patients' inclusion criteria

1) Delayed passage of meconium (beyond 48 hours for a full term neonate and beyond 72 hours for a preterm neonate)

2) Patients with defecation problems since birth and abdominal distension

Patients' exclusion criteria

1) The modified Bell staging criteria[26] in which a composite of clinical signs and symptoms (e.g., abdominal distention, bloody stools, or hypotension), biochemical parameters (e.g., thrombocytopenia or neutropenia), and radiographic signs (e.g., pneumatosis or pneumoperitoneum) was used to grade the severity of NEC.

2. Abdominal radiograph showing multiple air fluid levels.

Inclusion criteria were having clinical presentation highly suspicious for HD, performing a CE exam and full thickness biopsy. Written and verbal informed consent was taken from the parents who satisfied the inclusion criteria to undergo further investigations. Data on gestational age and first passage of meconium after birth were collected. Prior to per rectal examination, all patients underwent a plain abdominal radiograph and a contrast enema. Patients were divided into three age groups, as follows: < 1 month, 1 - 12 months, and > 12 months.

Plain abdominal radiograph transition zone:

A diagnostic evaluation should begin with abdominal X-rays (diagnostic accuracy: 52%), but normal practice would be to proceed to a contrast enema to evaluate the diagnosis further. A plain abdominal erect radiograph was taken to visualize tapering and abrupt cutoff of left colon gas shadow above the pelvis, which indicated the level of PARTZ [Figure 1]. All plain abdominal radiographs were read by the same radiologist.

Contrast enema

Radiologists performed the contrast enema with the support of a pediatrician in a routine manner using standard CE techniques. Dilute barium-sulfate was administered rectally using a # 6 infant feeding tube placed just within the rectum. No balloon catheters were used. All CE images were read by the same radiologist. The classical finding of a transition zone (CETZ) was considered being a positive result.

Rectal biopsy

The final diagnosis of HD was made by the absence of ganglion cells in a full thickness biopsy (FTB). Biopsy specimens were obtained at 2 cm above the dental line, posteriorly. These specimens were examined for ganglion cells with a hematoxylin-eosin staining and acetylcholine-sterase activity was determined as previously described by Karnovsky and Roots.[27] A biopsy was considered to be positive when the acetylcholinesterase activity was elevated in combination with an absence of ganglion cells.

A checklist was used to evaluate the items proposed by CE as radiologic signs, including six sub-scales: 1) transitional zone (TZ), defined as significant change in intestinal diameter from non-dilated to dilated section during CE; 2) RSI, ratio of largest rectal diameter to largest sigmoid diameter (among proximal, distal, and loop sections), is considered abnormal if it is < 1; 3) bizarre large irregular contractions in aganglionic region, with saw teeth appearance, due to dysrhythmia; 4) cobblestone appearance or mucosal irregularity or proximal colon spasm; 5) filling defect due to fecaloid materials; 6) lack of meconium defecation during the first 48 hours after birth. For subscales 1 and 2, if they were positive, we would consider scoring = 2, and, if they were negative, we would consider scoring = 0. For the other subscales, the positive results had scoring = 1 and negative results had score = 0. Furthermore, the patients were evaluated based on scoring system, as follows: high (5-8), and low (0–4).[28] Data analysis was performed with the Graph Pad Prism 7 software online. Chi-Square and independent t tests were used for analysis and were considered statistically significant at $P < 0.05$.

RESULTS

About 22 pediatric patients (12 neonates, 08 infants and 02 aged for than 1 yr) were diagnosed as suspected HD included in the study, of these, 14 (63.6%) patients had histological confirmed diagnosis of HD and 08 (36.4%) patients had HD

excluded by rectal biopsy. The mean age was 4.68 ± 5.24 months, ranging from 4 days to 1.4 years. Males represented 7 (31.8%) of patients while 15 (68.2%) were female [M: F ratio 0.32:1]. In HD group, 11 subjects showed high score (5-8) and 03 subjects low score (0-4). In the non-HD group, 06 subjects showed low score (0-4) and 02 subjects showed high score (5-8). Abdominal distension, lack of meconium defecation, and constipation were the most common clinical symptoms with frequencies of 15 (78.9%), 11 (57.8%), and 14 (73.68%), respectively. The most common findings in CE were TZ and RSI, respectively (Table 1).

For the 14 patients with histological confirmed HD, the sensitivity of detecting transition zone [Figure 1, 2 and 3] were 10 (71.43%) and 09 (64.29%) by the two independent radiologists respectively, giving a mean sensitivity of 67.86%. For the 08 patients with confirmed non-HD, the specificity of detecting the absence of transition zone were 6 (75.0%) and 5 (62.5%) by the two independently radiologists respectively, giving a mean specificity of 68.75%. The concordant rate for the correct radiological diagnosis was 15/22 (68.31%).

For the 14 patients with histological confirmed HD, the sensitivity of detecting spastic colon were 07 (50%) and 08 (57.14%) by the two independently radiologists respectively, giving a mean sensitivity of 53.57%. For the 08 patients with confirmed non-HD, the specificity of detecting the absence of spastic colon were 6 (75%) and 7 (87.5%) by the two independently radiologists respectively, giving a mean specificity of 81.25%. The concordant rate for the correct radiological diagnosis was 16/22 (67.41%).

For the 14 patients with histological confirmed HD, the sensitivity of detecting reversed recto-sigmoid index were 08 (57.14%) and 09 (64.29%) by the two independent radiologists respectively, giving a mean sensitivity of 60.72%. For 08 patients with confirmed non-HD, the specificity of detecting the absence of reversed recto-sigmoid index were 7 (87.5%) by the two independent radiologists, was giving a mean specificity of 87.5%. The concordant rate for the correct radiological diagnosis was 74.11%.

For the 14 patients with histological confirmed HD [Figure 4], the sensitivity of diagnosing the correct diagnosis of HD from an overall impression were 54.54% and 57.57% by the two independent radiologists respectively, giving a mean sensitivity of 56.06%. For the 08 patients with histological confirmed non-HD, the specificity of diagnosing the correct diagnosis of non-HD from an overall impression were 79.17% and 79.17% by the two independent radiologists respectively, giving a mean specificity of 79.17%. None of the histologically confirmed HD had aganglionic bowel segment extending proximal to the distal two third transverse colon i.e. no long segment HD and no total colonic aganglionosis were included in this study.

In summary, the mean sensitivity of detecting the radiological signs of transition zone [Figure 2, 3], spastic colon, reversed recto-sigmoid index and the overall impression in histological confirmed HD patients are 67.86%, 53.57%, 60.72% and 56.06% respectively. The mean specificity of detecting the absence of the radiological signs of transition zone, spastic colon, reversed recto-sigmoid index and the overall impression in histological confirmed non-HD patients are 68.75%, 81.25%, 87.5% and 79.17% respectively. This would in turn give an overall mean specificity rate of 79.17% in successfully excluding HD with the above mentioned radiological signs from the contrast enema.

Table 1. Contrast enema findings in those with and without Hirschsprung's disease [Data are presented as No. (%)]

Contrast enema [CE] findings	With Hirschsprung [n=14]	Without Hirschsprung [n=08]	*P* Value
Transitional zone	67.86%	31.25%	P = 0.1051
Recto-sigmoid index	60.72%.	12.5%	P = 0.0319
Irregular contraction	53.75%	18.75%	P = 0.1167
Mucosal irregularity	28.57%	12.5%	P = 0.3979
Cobblestone appearance	21.42%	-	-
Delay in passing meconium within 48 hours after birth	57.14%	37.5%	P = 0.3865

A plain abdominal radiograph showed tapering of left colon gas with an abrupt cutoff indicative of a transition zone (PARTZ) in 19/22 (86.36%) patients. The PARTZ was located at rectosigmoid in 18 (81.81%), midsigmoid in 3 (13.64%) and at descending colon in 1(4.5%). Of the 22 patients with a PARTZ, 14 (63.63%) had a matching level of aganglionosis [p = 0.0853, 95% CI (-5.907 to 48.091)]

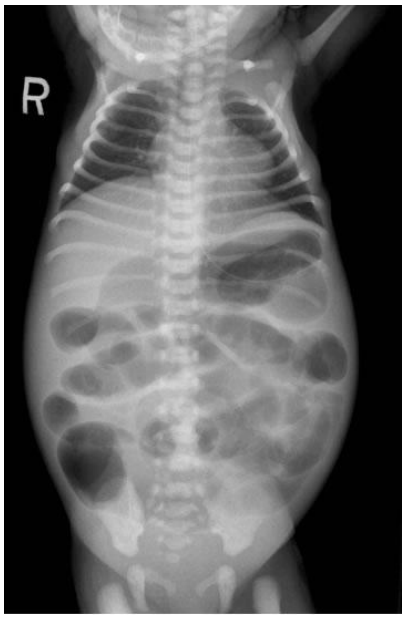

Figure 1: A plain AP view erect abdominal X-Ray shows multiple loops of dilated bowel, absence of rectal gas shadow, loaded colon, and multiple air-fluid levels (with intestinal obstruction) and gas under diaphragm in perforative peritonitis.

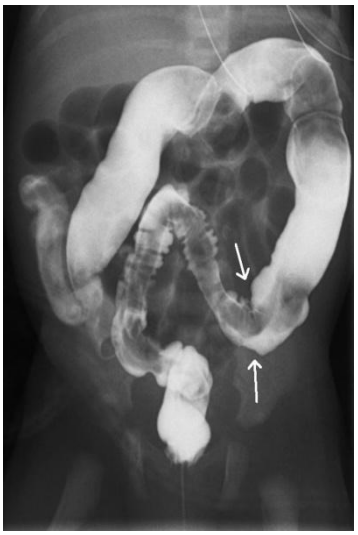

Figure 2: Post evacuation film demonstrates the transition point more obviously

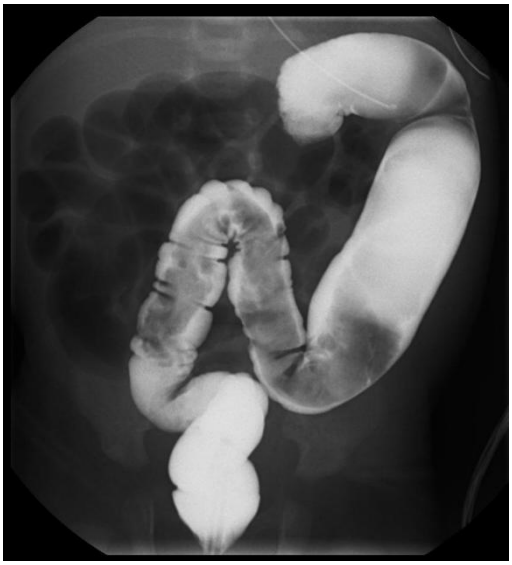

Figure 3: Barium enema demonstrates a reduced calibre rectum and sigmoid (the rectum is smaller than the descending colon) with a saw-tooth appearance to the wall. A transition point is seen at the junction between sigmoid and descending colon.

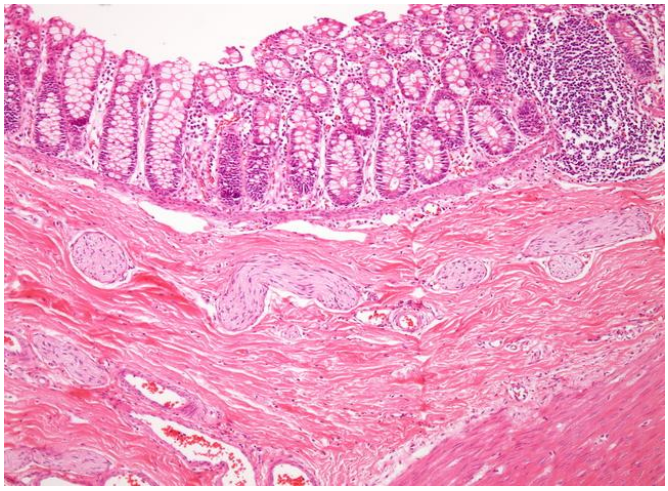

Figure 4: Hirschsprung disease is characterized by the absence of parasympathetic ganglion cells in both submucosal (Meissner) as well as myenteric (Auerbach) plexuses in the distal gastrointestinal tract. This image is from a suction biopsy of the rectum showing absence of ganglion cells in the submucosal plexus as well as the presence of hypertrophic submucosal nerve fibers.

DISCUSSION

The presenting features of HD vary depending on the length of the aganglionic segment and the severity. The gestational age at delivery also influences the clinical features, as HD rarely presents in preterm babies. The presentation may be of the following:

Delayed passage of meconium: In healthy term neonates, delay of more than 48 hours is indicative of HD though this feature occurs in only about 50% of neonates with HD. In preterm babies, delayed passage of meconium is normally seen hence this feature cannot suggest HD.

Neonatal bowel obstruction: Distal bowel obstruction is a presenting feature in about 25% of newborns who have HD and in 15-20% of all intestinal obstructions in the neonates [Figure 2].[29] The classical features are distended abdomen, bilious vomiting, fever, dehydration, lethargy, not passed meconium, and occasionally dilated peristaltic loops visible on the per abdominal examination in the neonate with a normal anus are almost diagnostic of HD. Insertion of simple rubber catheter and baby passing large quantity of meconium and flatus are characteristic of HD. Intestinal obstruction presents with bile-stained vomit, and abdominal distention is often present by day 2. In developing countries, presentation may be significantly late, and the main symptoms include abdominal distention, constipation, or diarrhea, with failure to thrive and developmental delay. These children often have a history of early onset of mild constipation followed by acute low intestinal obstruction. The early onset of chronic constipation (often since birth) is an indication to exclude HSCR. Stools when passed are irregular and passed with great difficulty.[30]

Neonatal bowel perforation: 5 percent of children with Hirschsprung's disease have bowel perforation, colonic[30] or ileal and accounts for about 10 percent of all newborn bowel perforations.

Neonatal enterocoliis: Sudden onset of diarrhea with or without blood in the stools in neonates should raise the suspicion of HD. About 30% of neonates with HD develop enterocolitis. The stool very foul smells, explosive and is associated with severe abdominal distension, fever, lethargy, dehydration and occasionally generalized sepsis. Hirschsprung's enterocolitis is a life threatening condition if toxic megacolon develops and identification and treatment must be prompt. **5. Constipation:** Constipation is the neonates could be associated with poor feeding, abdominal distension, straining to defecate, failure to thrive, jaundice, occasional non-bilious vomiting and an empty rectum on per rectal examination followed by explosive passage of feces and flatus after removal of the finger. Neonates could be passing normal stools before presenting with constipation. Appropriate investigations should be done if these features are present so as not to misdiagnose the condition.

The hallmark radiological feature of HD is the presence of a transition zone on a contrast enema (CETZ) [Figure 2, 3].[31]Although CETZ remains the most accurate diagnostic sign for Hirschsprung's disease, it is not specific enough to delineate the transition zone in neonates and

infants.[32, 33] The clinical presentation depends not only on the aganglionosis length [Figure 4] but also the age of presentation. More than 90% of the cases can be diagnosed in the neonatal period but are frequently overlooked in poorly resourced health situations, with late presentation is therefore common. A delay in passage of meconium is the most pertinent observation in the neonate suspected of having HD (80%).[31] In the present study we had abdominal distension, constipation and lack of meconium defecation were the most common clinical symptoms with frequencies of 18 (81.82%), 15 (68.18%) and 13 (59.09%) respectively. Whereas normal babies pass meconium within 24 hours, and even up to 48 hours, any baby who passes no or little meconium even after 24 hours should be investigated for HD.[3]

Abdominal distention occurs in almost 100% of the cases and may be marked. But in the present study we observed 18 (81.82%) of cases. Alehossein M et al[16] also observed abdominal distension, lack of meconium defecation, and constipation were the most common clinical symptoms with frequencies of 77.7%, 72.2%, and 53%, respectively. Sahoo R et[34] al also observed abdominal distension, lack of meconium defecation, and constipation were the most common clinical symptoms with frequencies of 15 (78.9%), 11 (57.8%), and 14 (73.68%) respectively. Our findings also matching with the findings reported by Alehossein M et al[16] and Sahoo R et al[34]. A family history of HSCR or severe constipation is not infrequent. Other associated anomalies may be present in approximately 16% of HSCR cases.[3]

Features can often be demonstrated radiologically but varies [Figure 1 & 2], and it is not sensitive enough to exclude HD. However, contrast studies may give an indication of the transition zone level [Figure 3]. Diagnosis is then confirmed by other tests (e.g., histology of rectal biopsies [Figure 4].[3] Previous studies have shown the transition zone and reversed recto-sigmoid index as the most common radiological signs in CE.[14, 35, 36] The sensitivity and specificity of CE in the diagnosis of HD have shown a wide range, between 60%-100%. This could be due to the difference in patient's selection: patients of different age and extent of disease; and difference in the skill levels of the radiologists.[14, 36]

We chose the following radiological signs: transition zone, spastic colon, reversed recto-sigmoid index, mucosal irregularity, cobblestone appearance and the overall impression of the CE films because these were the most commonly described features that had been reported in the contrast enema films in our centre. The challenging issues of intestinal biopsy are higher cost, more invasiveness, longer hospital stay and need to follow-up.[37] Therefore, in this study; we compared our diagnostic scoring system with full thickness intestinal biopsy [Figure 4], as the gold standard diagnostic method.

In a similar study, Donovan et al. introduced a scoring system with eight items in CE, with scores 0 and 1 for negative and positive results, respectively and the patients were divided into three groups: high (scores 6-8), moderate (scores 4-5), and low (scores 1-3) probability. However, this method of scoring was not very effective in our study and we decided to consider two points for positive results in RSI and TZ and one point for positive results related to lack of meconium defecation, cobblestone appearance, irregular contraction, and level of filling defect.[9] Previous studies have shown the TZ and RSI as the most common signs in CE.[14, 38, 39] The sensitivity and specificity of CE in the diagnosis of HD has shown a wide range, between 60% to 100%.[14, 38-40] This may be due to different techniques and also types of patient selections and different skill levels of the radiologists.

Similar to previous studies, the most common signs in CE were TZ and RSI [Figure 3]. Among these, the TZ is pathognomonic for HD. CE would have both diagnostic and therapeutic efficacy in neonates. However, in absence of TZ, the HD may not be ruled out. In older children, the first differential diagnosis of HD is functional

megacolon. In several review studies, the frequency of HD in those with TZ was reported at 72%, while in the presence of other signs, it reduced to half.[41]

A diagnostic evaluation should begin with abdominal X-rays [Figure 1] (diagnostic accuracy: 52%), but normal practice would be to proceed to a contrast enema to evaluate the diagnosis further. Essentially, it is necessary to look for signs of low intestinal obstruction and distended bowel loops of different calibers on the abdominal radiographs. Erect abdominal views can demonstrate air–fluid levels, indicating intestinal obstruction. There would be absence of rectal air, and the lateral view may occasionally demonstrate a narrow rectum. If small bowel obstruction is prominent, a longer aganglionic segment must be considered.[4]

A reduced size of rectum or rectosigmoid, with transition to a dilated, proximal colon on contrast enema, is typical of short-segment (rectosigmoid) aganglionosis. The radiologic study should show variation in lumen size (the so-called transition zone). In addition, an irregular or "sawtooth" appearance may be present (Figure 3).[3] These findings may vary, particularly in neonates, as the transition zone may not have developed sufficiently, or in patients with extended aganglionic segments (TCA).[3]

The essential diagnostic feature on contrast enema is demonstration of the narrow aganglionic segment with dilatation of the proximal bowel segment, a reversed rectosigmoid ratio, and a demonstrable transitional segment (Figure 3). The aganglionic segment may be irregular, demonstrating a saw-tooth mucosal appearance, probably as a result of mucosal edema and muscular fasciculations.

A further delay in the clearing of contrast (barium sulfate) within 24 hours is also a reliable sign, and a follow-up X-ray should be performed the following day.[3] In rectosigmoid aganglionosis, the rectosigmoid ratio (ratio of the diameter of the rectum to the sigmoid colon; normal ratio, 1:1) may prove a useful guide, but considerable variation may exist at different ages and aganglionic lengths. In one study,the rectosigmoid index and radiological transitional zone supported the histopathologic diagnosis in 79%–87% of the cases.[29]

The histopathologic diagnosis of HSCR essentially rests on observing the absence of ganglion cells in the intermyenteric plexuses, as well as observation of the presence of enlarged peripheral nerve trunks in the submucosa and the observed increased proliferation of neurofibrils in the lamina propria and the muscularis mucosa on special stains (absent in normally innervated intestinal wall). These findings may vary, particularly in neonates and in long-segment aganglionosis.[3]

CONCLUSION

Hirschsprung is an uncommon disease seen in all races and may result in decreased quality of life due to chronic constipation and periodic obstructions. It may also be fatal in severe cases with enterocolitis, functional obstruction, and intestinal perforation. The diagnosis of Hirschsprung's disease should be suspected in patients with a history of abdominal distension, constipation and appropriate radiographic findings. Although the intestinal biopsy is the diagnostic gold standard, the first diagnostic method is CE. The hallmark radiological feature of HD is the presence of a transition zone on a contrast enema (CETZ). Among these, the TZ is pathognomonic for HD. This may be due to different techniques and also types of patient selections and different skill levels of the radiologists. CE would have both diagnostic and therapeutic efficacy in neonates. In conclusion, our study underscores the importance of combining the information of a transition zone on a plain abdominal radiograph and contrast enema to decide the surgical approach for the correction of Hirschsprung's in developing countries where laparoscopic facilities are not available. Further multicentric studies, with larger sample sizes, are suggested for comparison of all diagnostic methods.

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