ORIGINAL ARTICLE

Barium Enema in Hirschsprung's Disease : Correlation of Clinical Outcome

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Abstract:

The anticipated level of aganglionosis can influence the surgical planning in Hirschsprung's disease (HD). The aim of this study was to find out the role of barium enema in diagnosis of HD. Barium enema is usually performed in patients referred for investigation for constipation. This prospective study was performed in the department of pediatric surgery Chittagong Medical College & Hospital. During this period a total of 198 patients were clinically diagnosed as HD. All patients were initially evaluated by plain x- ray abdomen in erect posture. Among 198 patients, 43 patients had perforation in plain x- ray and 31 were clinically unstable at presentation. Those 74 patients were excluded from the study. Rest 124 participating patients were divided into three groups: Neonate, Infant and children according to their age. Male to female ratio was 2.44: 1. Barium enema X- ray early film, 24 hours delayed film (Antero-posterior and lateral view) were done for every patient. Length of radiological narrow zone (RNZ), radiological Transitional zone (RTZ) and Recto-sigmoid index (RSI) was measured in cm. Photo of X- ray was taken for every patient. RTZ was demonstrated in 74 (60%) patients. RNZ was demonstrated in 94 (75%) studied population. RSI was measured in 65 (52.42%) subjects. Retention of barium more than 24 hours was found in 95 (75%) patients. Redundant sigmoid colon was found in 54 (43.5%) patients. Right-sided sigmoid colon was present in 46(30.6%) patients. Saw-toothed appearance in spastic colon was seen in 38 (30.6%) patients. Mucosal edema and irregularities were present in 25 (20.2%) patients. Rounded transverse colon was seen in 6 children. Barium mixed with stool was found in 32 (25.8%) patients. Mosaic pattern of colon was present in 10 patients. Visualization of RTZ is a reliable sign of HD. Besides the RTZ, BE in HD has various other radiological features.

Introduction:

Hirschsprung's disease (HD) is a developmental disorder of the enteric nervous system characterized by absence of ganglion cells in the myenteric and missener's plexuses along a variable portion of the distal intestine1. Aganglionosis typically extends upto the recto sigmoid region in approximately 80% cases and the process is nearly always continuous and uninterrupted until the proximal ganglionic segment is reached2. Diagnosis of HD has always been a clinical challenge for pediatric surgeons. It requires consideration of the patient history, clinical findings, radiological evaluation & anorectal manometry. Histopathological examination & histochemical findings are required to confirm the diagnosis.

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Histopathological examination & histochemical findings are required to confirm the diagnosis. Radiological examination is an important diagnostic modality in diagnosis of HD. Radiographic studies begin with anteroposterior or lateral decubitus abdominal films, which commonly show several distended loops of intestine, may show a paucity of air in the rectum, evidence of enterocolitis or free air in case of intestinal perforation3,4. The Barium enema (BE) is a good screening test for HD. It is usually performed in patients referred for investigation for constipation. Swenson et al first described the barium enema findings of relatively narrowed aganglionic segment distal to a dilated normal colon in 19485. The aim of this study is to find out the various findings of barium enema in diagnosing HD.

Materials and method:

This prospective study was performed in the department of pediatric surgery Chittagong Medical College & Hospital. During this period a total of 198 patients were clinically diagnosed as HD. All patients were initially evaluated by plain x- ray abdomen in erect posture. Among 198 patients, 43 patients had perforation in plain x- ray and 31 were clinically unstable at presentation. Those 74 patients were excluded from the study. Rest 124 participating patients were divided into three groups: Neonate, Infant and children according to their age. Neonates were 32(25.8%), Infants were 43(34.7%) and Children were 49 (39.5%). Male was 71% (n=88) and 29% (n= 36) was female. Male to female ratio was 2.44: 1. Barium enema X- ray early film, 24 hours delayed film – A/P and lateral view were done for every patient. Length of radiological narrow zone (RNZ), radiological Transitional zone (RTZ) & Recto-sigmoid index (RSI) was measured in cm. Photo of X- ray was taken for every patient.

Results:

RTZ was fig1) demonstrated in 74 (60%) patients. Out of which neonates were 17, Infants were 23 and children were 34. RNZ (fig2) were demonstrated in 94(75%) studied population. Among them, 26 were neonate, 34 were infants and children were 34.

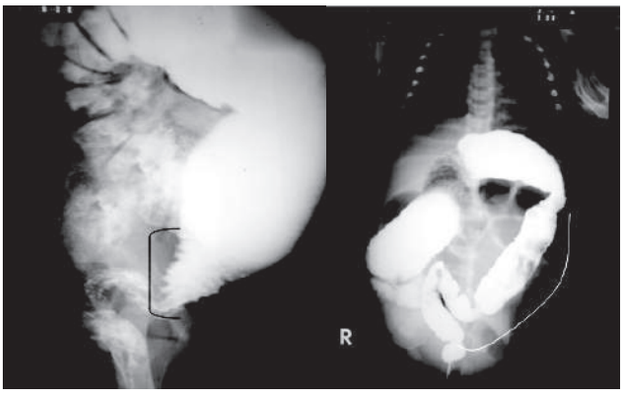

Figure-1: RTZ at recto-sigmoid junction
Figure-2: RNZ extending up to splenic flexure

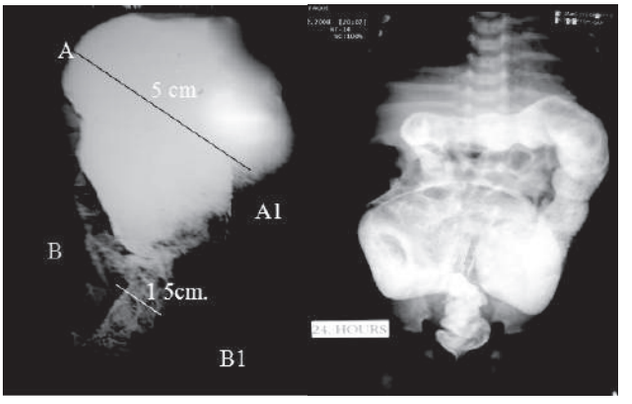

Figure-3: Measuring RSI Figure-4: Retention of Barium after 24 hours with Long redundant sigmoid

RSI (fig3) was measured in 65 (52.42%) subjects. Among them neonates were 14, infants were 25, children were 26. Retention of barium more than 24 hours (fig 4) was found in 95(75%) patient. Among them 30 were Neonates, infant were 30 & children were 35.

Redundant sigmoid colon was found in 54 (43.5%) patients. Among them neonates were 21, infants were 23 and children were 10. Right-sided sigmoid colon was present in 46(30.6%) patients. Out of which neonates were 16,

infants were 18 and children were 20.

Saw-toothed appearance in spastic colon was seen in 38 patients (30.6%). Mucosal edema and irregularities was present in 25 (20.2%) patients, Rounded transverse colon was seen in 6 children. Barium mixed with stool was found in 32 (25.8%) patients. Mosaic pattern of colon was present in 10 patients.

Discussion:

The first report of a patient with HD was made in 1691 by Frederick Ruysch, but it was Danish pediatrician Harold Hirschsprung, who in 1888 published the classic description of congenital megacolon6. Harold Hirschsprung described the clinical and autopsy findings in two infants who had suffered from constipation and died with dilatation and hypertrophy of the colon. Ehrenpreis pointed out that the diagnosis could be made in the newborn[7]. He was the first to appreciate that colon became enlarged and distended because of the obstruction caused by the lack of ganglion cell[2]. Even after 100 years of Harold Hirschsprung's clinical presentation, the condition was considered incurable and uniformly fatal. Over time, major advances have taken place in the diagnosis of HD with histochemistry, anorectal manometry etc. Diagnosis of HD has always been a clinical challenge for pediatric surgeons. It requires considerations of the patient's history, clinical findings, radiological evaluation and anorectal manometry. Rectal biopsy is the gold standard for the diagnosis of aganglionosis .In Bangladesh ,investigative studies are bounded by many limitations . There are very few pathologists experienced in pediatric cases. Most of the histopathological examinations are usually done by general pathologist. Anorectal manometry is not available yet. Kelleher and Blake reported that while rectal manometry remains a reliable screening procedure, radiology still has an important role to play[8].

Teitelbaum et al described that classic radiographic finding of HD is a narrow, spastic distal intestinal segment with a dilated proximal segment[2]. In our studied population (n=124) BE of 60% (74) patients showed a funnel or cone like transition zone between the dilated proximal bowel and the narrowed distal segment suggestive of HD. In 20 % of these children, the funnel or cone was not typical. We assume this finding might be due to longer duration of irrigation before doing enema or due to the use of the stalk of betel leaf for passage of meconium. Naria also mentioned that manipulation before doing enema may interfere with demonstration of RTZ[9]. He identified RTZ in 80% of his patients. Roshenfield et al opined that digital examination and irrigation prior to contrast examination does not affect visualization of RTZ10. We did not find RTZ in 52% of our Neonates. Thirty five percent of the neonates of Rosenfield et al and 15% of Klein did not show any RTZ[10,11]. RTZ in 48% of our neonates with subsequent confirmation by peroperative & histopathological examination showed that, if BE in the neonatal period demonstrate a transition zone, no other investigations are needed to diagnose HD. Berman and Hope mentioned that RTZ is more often visualized in the older age group. Which is similar to our findings also, with 53% infant and 63% children showing RTZ in BE[12,13].

Ehrenpresis first described the use of delayed radiograph obtained 24 and 48 hours after BE to document poor emptying of the colon and this sign was considered by many to be important in the diagnosis of HD[7,12, 14-16]. We have also found the same finding in 76.6% of our studied population. In case of normal colon if BE is done for intussusception or chronic constipation, after 24 to 48 hours small amount of barium is collected as bolus, but in case of HD the remained barium is often seen to be uniformly mixed throughout the colon and becomes apparent as the formed feces collect behind the aganglionic segment .We have 50 (40%) patients who had no RTZ but there were retention of barium for more than 24 hours. Puri et al also thought that TZ may be accentuated and is more easily identified in delayed film[7]. Marfan & Griffith found unusually long and folded sigmoid colon in HD[6]. 59% of our studied population had also

redundant sigmoid colon. We assume that long sigmoid colon is a positive barium enema finding for HD. we have seen that the children who had redundant colon in BE usually complain of constipation more than those who did not have redundant sigmoid colon. Brummer et al found redundant colon in patients with constipation but who did not have HD in rectal biopsy[17]. These patients might have some sort of IND or motility disorders. More research is needed to look into this finding. Right sided sigmoid colon was present in 47% of our patients. In these patients, some times RTZ was not evident but pathological extension of narrow & transitional zone was greater than radiological length. Swenson and Hope described irregular contraction in the aganglionic segment as a reliable sign of HD[5,13] . The irregular contraction is thought to be due to denervation hypersensitivity of the smooth muscle or may be due to colitis[10]. 30.6% of our studied population had 'Saw – Toothed' contraction. Typical RTZ may not be apparent in early period. Normally transverse rectal diameter is wider than that of sigmoid colon. In HD, widest rectal diameter is usually smaller than widest diameter of sigmoid colon. RSI has been used for the diagnosis of HD in neonates[18]. RSI was <0.6 in 55% of our patients. Because of the folding of colon, we failed to measure RSI in more than half of our patients. We have seen mucosal edema and thickened bowel wall in 20 % (25) of our patients. As most (14) of these patients were more than one year old, we think that prolong stasis of fecal matter leads to inflammation of the bowel wall and causes edema & thickening. Puri also mentioned this feature as a diagnostic tool[19].

Spastic colon was found in 7.3% of our patients. This group of patients had no RTZ and whole of the colon showed irregular bizarre appearance. Hope described irregular contraction in the spastic segment as a reliable sign of HD and it is thought to be due to denervation hypersensitivity[13]. We also found rounded hepatic and splenic flexures in half of our patients with spastic colon. Hawakawa introduced this as a diagnostic feature for total colonic aganglionosis[20].

Antero-posterior radiograph will often hide a ganglionic segment, may be due to excessive insertion of enema tube or repeated irrigation before enema and also due to differences among the skills of the operators. It is usually best seen in the lateral radiograph. This emphasizes the need to have experienced radiologists understanding pediatric problems, better co-operation and flexibility between the surgeons and the radiologists.

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