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# Diagnosis of Hirschsprung Disease in Neonates Histo-pathological Correlation of Barium Enema X-ray

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**Abstract** **Original Research Article**

***Background:*** A condition that affects the large intestine (colon) and causes problems with passing stool is called Hirschsprung's (HIRSH-sproongz) disease. The condition is present at birth (congenital) as a result of missing nerve cells in the muscles of the baby's colon. ***Objectives:*** The aim of the study was one staged primary transanal endorectal pull through procedure, barium enama X-ray is an important initial diagnostic procedure in the evaluation on the determination of the level of aganglionosis preoperatively for HD in neonates. ***Methods:*** This prospective study was carried out in the Department of Paediatric Surgery and the Department of Radiology & Imaging, Dhaka Shishu Hospital (DSH) & Bangladesh Institute of Child Health (BICH) Dhaka during the period of July 2016 to June 2021. The study was conducted on 45 neonates of either sex admitted in Dhaka Shishu Hospital with Astuspected HD with exclution of patients with pneumoperitoneum on plain X-ray, too sick needed emergency laparotomy and patients who had BE X-ray but did not have rectal biopsy report. ***Results:*** A total of 37 neonates were enrolled in the study. Male Female ratio was 3.1:1. The mean age was 16.5days. The mean weight was 2.87kg. Punch rectal biopsy was done in 31 (83.8%), laparotomy and biopsy in 7(18.9%) cases. Rectosigmoid index was <1 in 31 (83.7%) cases out of which 30 were HD positive and 1 was HD negative as proved by histopathological study. Retention of barium was present in 19(65.5%) out of which 17 were HD positive, 2 HD negative. For retention of barium of barium alone, sensitivity was 65.38%, specificity 33.33%, accuracy 62.1%, positive predictive value 89.5% and negative predictive value 10%. So, retention of barium alone is not reliable for the diagnosis of HD. ***Conclusion:*** Barium enema X-ray could predict the level of aganglionosis in all rectosigmoid HD. It proved that Barium enema x-ray is a sensitive, specific and accurate investigation for the diagnosis of Hirschprung's disease in neonates.

**Keywords:** Hirschsprung Disease, Neonates, Histo-pathology, Barium Enema X-ray.



## INTRODUCTION

Hirschsprung's disease (HD) is congenital aganglionosis of colon. It is reported to be the most common cause of intestinal obstruction in the newborn [1]. Diagnosis of HD has always been a clinical challenge for paediatric surgeons. The mean age, at which the children are diagnosed has progressively decreased [2]. More than 90% cases of HD are diagnosed in the neonatal period these days. Early diagnosis of HD remains a desirable objective since immediate surgicale intervention is of utmost importance to reduce the morbidity and mortality associated with the disease.

Radiological examination is the most important diagnostic modality. Plain x-ray abdomen in erect posture may demonstrate dilated loops of intestines with or without air-fluid levels, paucity of rectal gas, evidence of enterocolitis or free air in case of intestinal perforation [3].

Barium enema x-ray may demonstrate a radiological transition zone (RTZ), rectosigmoid index (RSI) or retention of barium (RB) in 24-hour late film or a microcolon in case of total colonic aganglionosis. RTZ. Is arelatively narrowed aganglionic segment distal to a dilated normal colon. It has been the hallmark for radiographic diagnosis of HD. Failure to visualize a RTZ

in neonates has been previously recognized [4] RSI is of rectum to maximum diameter of sigmoid colon. It is less than I in neonates with HD and equal to or a ratio of maximum um diameter more than I in normal individual and in other pathologic conditions of gastrointestinal tract. lt aids in the diagnosis of neonatal HD in cases with no definite RTZ [5]. RB is considered by many to be important in the diagnosis of HD. Rosenfield study reported that retention of barium is not a specific sign, but it may be the only sign of HD in neonates [6]. Barium enema X-ray is a simple and safe procedure, readily available in most hospital settings. It has been relied on as the initial diagnostic or screening procedure. The diagnostic accuracy of barium enema x-ray reported in different series varies from 76-92% [7]. A 20% rate of false positive and a 43% of false negative results have been reported [8]. Several investigators have reported the presence of radiologic trasition zone on barium enema X-ray in the diagnosis of HD. However, the concordance between the location of radiologic trasition zone and the level of aganglionosis has not been well studied [9]. Some have reported cases in which the radiological transition zone and pathological trasition zone differ. Another study found radiological transion zone to be concordant with aganglionosis [10]. The study found 90% concordace when radiologic transition zone was present [11].

The traditional treatment of HD has been staged surgery that includes creation of colostomy in normally inervated bowel in the neonatal period followed by one of the abdomino-perineal pull through procedures at 6-12 month of age and closure of the colostomy. With the advent of one stage transanal endorectal pull through procedure, which is based in part on the anticipated level of aganglionosis and the extent of dilatation of proximal colon, barium enema x-ray has been increasingly relied on to predict the level of aganglionosis and to observe the extent of proximal colon dilatation [12-14].

Cosidering these facts, the present study was carried out to evaluate the sensitivity, specificity, accuracy and predictivity of barium enema x-ray in the diagnosis of HD in neonates and the concordance of the radiological trasition zone with the level of aganglionosis to implicate surgical approach.

## METERIALS AND METHODS

This cross-sectional, consecutive, prospective study was carried out in the department of Paediatric Surgery and the Department of Radiology & Imaging, Bangladesh Institute of Child Health (BICH) & Dhaka Shishu (Child) Hospital during the period of July 2016 to June 2021. The study was conducted on neonates admitted in Dhaka Shishu Hospital during the period with suspected HD. A total of 45 cases were initially included. Neonates (age ≤28 days) presenting with abdominal distension with or without vomiting and/or history of delayed passage of meconium. The patients whom rectal decompression done prior to contrast enema, were also included after 24 hours. We exclude the patients with huge abdomen distention demonstrating pneumoperitoneum on plain X-ray, patients who were too sick and needed emergency laparotomy and patients who had BE X-ray but did not have rectal biopsy report.

The neonates of either sex with clinical features of intestinal obstruction were evaluated, admitted and managed initially according to the protocol. Barium enema was preformed on the neonates matching selection criteria. Further investigations and management followed the hospital protocol (fig. 1). The aims and objectives of the study, the procedure, its assistance in making rational approach of the case management and potential complications were explained to the patient's parents/legal guardians in easily understandable local language and then informed consent was taken.

**Barium enema (BE) examination**

A thin dilute barium in normal saline is traditionally used. A total of 4 exposures were taken; 1 each for plain x-ray in erect posture, BE anterior-posterior (AP) view, BE lateral view and 24-hour late film. We used a volume of barium milk prepared with normal saline calculated from body weight (5-15 ml/kg) to instillate optimum amount of barium and to prevent water intoxication. The calculated volume of the contrast was neither too less to visualize the radiologic transition zone or rectosigmoid index nor too much to overload the sigmoid. The following materials were required for barium enema: Barium sulphate powder, Normal saline, and 10Fr plain rubber catheter, 50 cc disposable syringe, 2% lidocaine HCI lubricant jelly, a Bowl and stirrer. A thin dilute barium milk was prepared by mixing barium sulphate powder with normal saline of a volume of 5-15 ml/kg body weight until the suspension was of milk like consistency (Barium sulphate powder 5gm/10 ml Normal saline).

**Procedure of barium enema**

1. At first a plain x-ray abdomen including domes of diaphragm in erect posture was taken, the wet film was viewed to rulc out presence of pneumoperitonium and multiple gas-fluid levels.
2. Patient was kept in suspended lithotomy or lateral position on the table or on mother's lap.
3. A 10 Fr plain rubber catheter (lubricated with 2% lidocaine jelly) was inserted into the anorectum upto 2-3cm from anal verge. Open end of catheter connected with a funnel of 50 cc syringe by a saline tube.
4. Barium milk (5-15ml/kg body weight) was allowed to run slowly by gravity at a height less than three feet into the anal canal so that it would not distend the aganglionic segment. Usually 15-45ml was sufficient depending upon the degree of dilatation of distal colon.

5. The tube was withdrawn and Antero-posterior and lateral view radiographs were taken immediately and a late film (24 hours) was taken to see hold-up of barium.

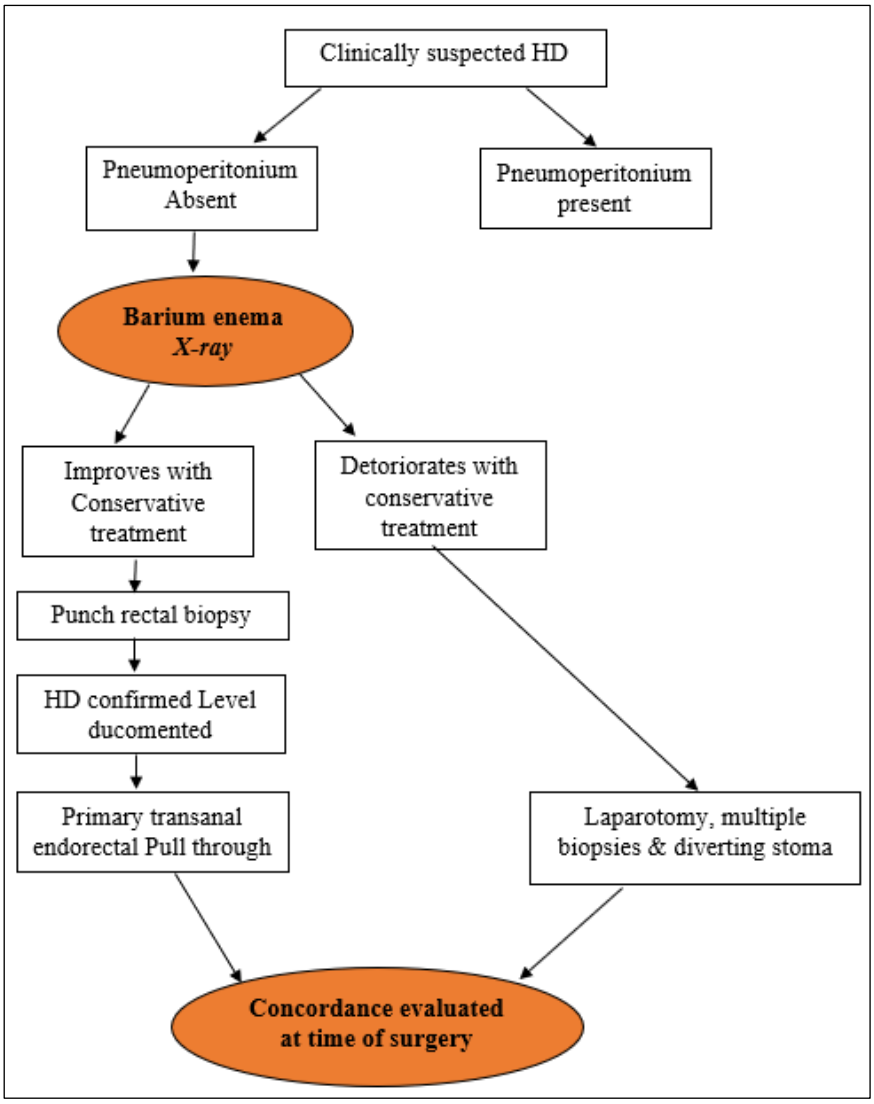


**Fig. 1: Flow chart of the study design**

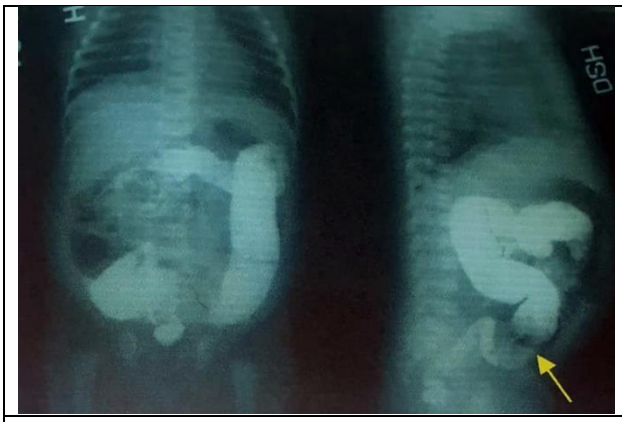

**Fig. 1: Barium enema x-ray showing radiological transition zone**

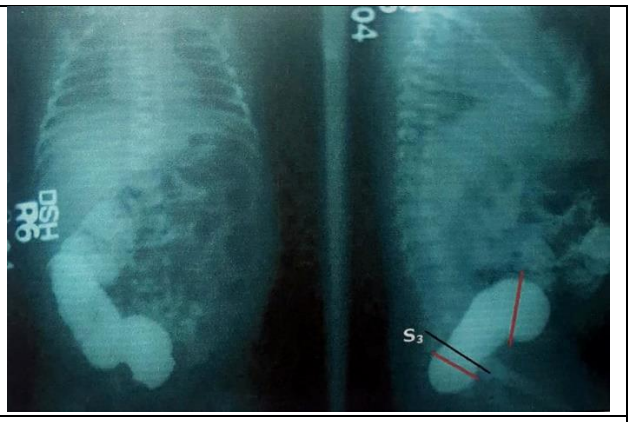

**Fig. 2: Barium enema X-ray showing rectosigmold index**

**Fig. 3: 24-hour late film showing retention of barium**

**Fig. 4: 24-hour late film showing no retention of barium**

**Interpretation of barium enema findigus**

The radiologist interpreted the findings in the barium enema X-ray, who was kept blind regarding the histopathological report. The radiological transition zone is subjective finding on barium enema. The dilated proximal colon was obvious on barium enema in both A/P and lateral (usually sigmoid colon) with narrow distal segment views. It was absent or questionable in some cases. The racto-sigmoid index in an objective measurement with a numerical value. It was determined in the lateral BE film by dividing the maximum diameter of rectum at any level below 3d sacral segment by maximum diameter of sigmoid above the 3d sacral vertebra. The measurements were obtained along a transverse axis at right angle to the longitudinal axis of the colon at that point. The antero-posterior barium enema film helped to determine the course of sigmoid colon particularly when it overlaid the rectum. The microcolon is narrow usually <Icm in diameter. Retention of barium in 24-hour late film was evaluated.

The following outcome variables were studied: Demographic and clinical variables:

Age, Sex, Weight, Family history, Clinical presentation, Associated anomalis.

Radiologic variables: Dilated loops of intestines with/without air-fluid levels, Paucity of rectal gas, Colon cut off sign, Radiological transition zone (RTZ), Rectosigmoid index (RSI), Retention of barium in late film, Microcolon

**Histopathologic correlation**

Panch rectal biopsy was done to confirm the diagnosis. Those patients who underwent laparotomy had multiple rectal and colonic biopsies taken to evaluate the level of aganglionosis.

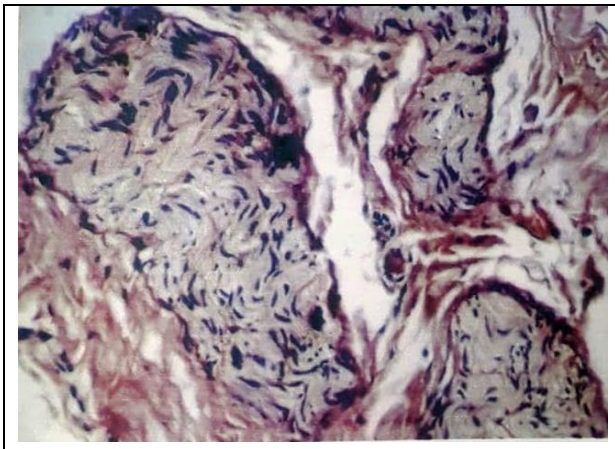

**Fig. 5: Photomicrograph showing hypertrophied nerve bundles in submucous layer in HD (H&E stain)**

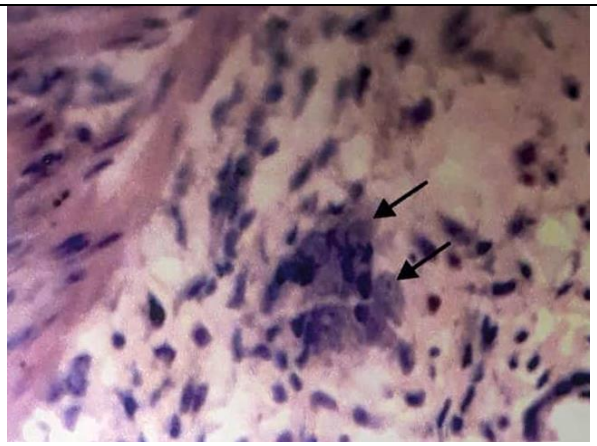

**Fig. 6: Photomicrograph showing ganglion cells in submucous layer in normal colon**

**Data Collection and Analysis**

Data were collected in predesigned data sheet by taking history, meticulous clinical examination, relevant investigations and operative findings. All the relevant collected data were compiled on a master chart. The outcome variables were sub-grouped. Percentages were calculated to find out the proportion. For the validity of the study outcome, sensitivity, specificity, accuracy, positive and negative predictive values of barium enema were calculated. For significance of differences, Pearson's X test was applied. P value <0.05 was taken as statistically significant. The results were presented in suitable tables and diagrams.

## RESULTS

A total of 3920 patients were admitted in the Department of Surgery, Dhaka Shishu (Children) Hospital during July 2003 to June 2005. Out of them, 204 were Hirschsprung's disease (HD), which was 5.2% of total surgical admission. A total of 86 (42.1%) neonates were diagnosed to have HD. A total 45 neonates who met the selection criteria underwent the Barium enema (BE) examination. All patients had some form of rectal decompression like glyceine suppository, rectal enema, tube or foreign body stimulation of anorectum before admission. The BE examination was performed after restricting rectal decompression for at least 24 hours. One neonate having congenital cardiac anomaly expired before rectal biopsy could be taken and was excluded. Parents of one neonate refused rectal biopsy and subsequent treatment. Biopsy reports were unavailable in 3 cases. These 4 cases were also excluded from the study. BE could diagnose 3 cases were not included in the study. So finally, the study population comprised of n-37 cases and statistical analysis was done on this population. Nine patients presented with complications, 7 with septicaemia and 2 with enterocolitis. These patients required aggressive resuscitation with intravenous fluid and broad-spectrum antibiotics. They underwent barium enema X-ray after stabilization. The age range of study population was 2-28 days. The mean ± SD age at presentation was 16.499.41 days. There were 3 preterm babies (8.1%). Out of 37 neonates, 28 were males and 9 were females giving rise to male: female ratio of 3.1.1 (fig. 2).

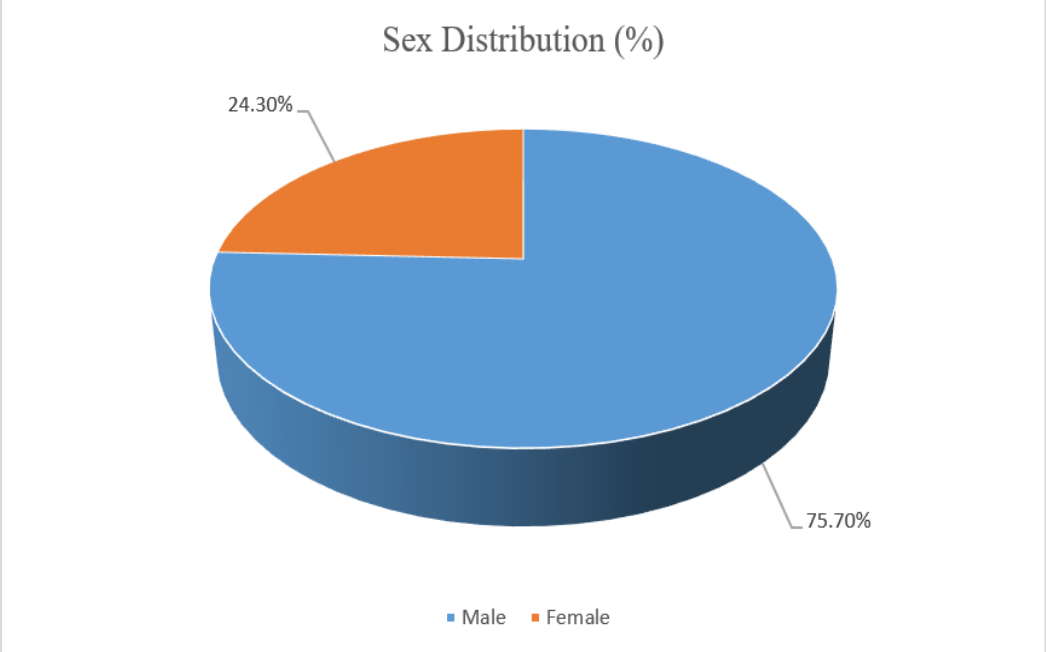


**Fig.2: Sex distribution of study population**

Body weight of the study population ranged from 1.6-4.5 Kg. The mean ± SD weight was 2.87 ±0.63 kg. There was no family history of HD in any of the cases. The parents of 8 cases (21.6%) had consanguineous marriage.

**Clinical Presentation**

All the patients (100%) presented with abdominal distention. Twenty-nine patients (78.4%) had constipation and 20 (54.1%) had vomiting. There were 13 patients (35.1%) who failed to pass meconium by 48 hours. One patient (2.7%) had assisted passage of meconium, while 16 (43.2%) had no history of delayed or assisted passage of meconium Nine patients (24.3%) presunted with complications, 7 (18.9%) with septicemia and 2 (5.4%) with enterocolitis (table 1). Associated anomalies One patient (2.7%) had Down's syndrome (table 1).

**Table I: Clinical presentation of study population (n=37)**

| Sex | N=37 | % |
|---|---|---|
| Male | 28 | 75.7 |
| Female | 9 | 24.3 |
| Associated anomalies | | |
| Down's syndrome | 1 | 2.7 |
| Preterm | 3 | 8.1 |
| Presentation | | |
| Abdominal distention | 37 | 100 |
| Constipation | 29 | 78.4 |
| Vomiting | 20 | 54.1 |
| H/O passage of meconium | | |
| Beyond 48 hours | 13 | 35.1 |
| Assisted passage of meconium | 1 | 2.7 |
| No H/O delayed or assisted passage of meconium | 16 | 43.2 |
| Complications | **9** | **24.3** |
| Septicaemia | 7 | 18.9 |
| Enterocolitis | 2 | 5.4 |

**Plain X-ray abdomen in erect posture**

Dilated loops of intestine and paucity of rectal gas were present in all cases (100%). Dilated loops of intestine with air fluid levels were present in 14 patients (38%). Colon cut off sign was present in 2 (5.4%). There was no free gas in any of the cases.

**Table II: Plain X-ray abdomen findings in study population (n=37)**

| | N=37 | % |
|---|---|---|
| Dilated loops of intestines | 37 | 100 |
| Paucity of rectal gas | 37 | 100 |
| Dilated loops of intestines with air-fluid levels | 14 | 38 |
| Colon cut off sign | 2 | 5.4 |

**Histopathology**

Punch rectal biopsy was preferred method of rectal biopsy in our institute. Thirty-one neonates (83.7%) had punch rectal biopsy out of which 27 (87.1%) proved to be HD positive and 4 (12.9%) to be HD negative.

Laparotomy and multiple colonic biopsies and diverting stoma were needed in 7 neonates (18.9%). In one case both procedures were done. Extramucosal (seromuscular) biopsies were taken from narrow segment, dilated segment and another one 3-5 cm proximal to it. Five neonates (71.4%) were proved to have rectosigmoid HD and 2 (28.6%) were TCA histopathologically.

The specimen was stained with haematoxylin and eosin. No histochemistry using acetylcholine esterase staining was done in any case. Demonstration of absence of ganglion cells and presence of hypertrophied nerve fibers confirmed the diagnosis of HD.

**Analysis of barium enema and histopathological findings**

The histopathological reports proved 31 HD positive and 4 HD negative. The remaining 2 cases were total colonic aganglionosis (TCA). Barium enema predicted 30 true positive cases, 3 true negative cases and I case of false positive and false negative each. The remaining 2 cases were TCA. Sensitivity of barium enema X-ray to diagnose HD was 96.77%, specificity was 75%, accuracy was 94.3%, positive predictivity value was 96.77% and negative predictivity value was 75%. Out of 35 cases, radiological transition zone was present in 15 cases and these were true positive. In remaining 20, radiological transition zone was absent in 16 cases that were HD positive, 4 that were HD negative. So, for radiological transition zone alone, 15 were true positive, 16 were false negative, 4 were true negative and no false positive. The sensitivity for radiological transition zone was 84.4%, specificity 100%, accuracy 88.5%, positive value 100% and negative predictive value only 20% (Table III).

**Table III: Comparing the predictive power of barium enema findings in study population (n=35*)**

| | RIZ (n=35) | RSI | RSI without RTZ (n=20) | RB (n=29) |
|---|---|---|---|---|
| TP | 15 | 30 | 17 | 17 |
| TN | 4 | 3 | 3 | 1 |
| FP | 0 | 1 | 1 | 2 |
| FN | 16 | 1 | 1 | 9 |
| Sensitivity | 84.40% | 96.77 | 93.70% | 65.38% |
| Specificity | 100% | 75% | 75% | 33.33% |
| Accuracy | 88.50% | 94.30% | 90% | 60.10% |
| PPV | 100% | 96.77% | 93.76% | 89.50% |
| NPV | 20% | 75% | 75% | 10% |

*Two TCA cases were excluded

**Table IV**

| | | | |
|---|---|---|---|
| RTZ | Radiological transition zone | FP | False positive |
| RSI | Rectosigmoid index | FN | False negative |
| RB | Retention of barium | PPV | Positive predictive value |
| TP | True positive | NPV | Negative predictive value |
| TN | True negative | | |

Rectosigmoid index is <1 in HD and 21 in normal subjects. In 31 subjects rectosigmoid index was <1 out of which 30 were HD positive and 1 was HD negative. In 4 subjects rectosigmoid index was 21 out of which 3 were HD negative and 1 was HD positive. So, for rectosigmoid in index 30 were true positive, I false positive, 3 true negative and 1 false negative. The sensitivity for rectosigmoid index was 96.77%, Specificity was 75%, accuracy was 94.3%, positive predictive value was 96.77% and negative predictive value was 75% (table IV).

When rectosigmoid index for 20 cases where radiological transition zone was absent, was considered, the rectosigmoid index was <1 in 16 cases, of which 15 were true positive and I was false positive as proved histopathologically. Rectosigmoid index was 21 in 4 cases, of which 3 were true negative and I was false negative. The sensitivity for rectosigmoid index where radiological transition zone was absent was 93.7%, specificity 75%, accuracy 90%, positive predictive value 93.76% and negative predictive value 75% (table IV).

Late film was available for 29 patients. In 19 cases barium retention was present, out of which 17 were true positive and 2 were false positive as proved histopathologically. In 10 cases barium retention was absent, out of which I was true negative and 9 were false negative. The sensitivity for barium holds up was 65.38%, specificity was 33.33%, accuracy was 62.1%, positive predictive value was 89.5% and negative predictive value was 10% (table IV).

**Concordance of Aganglionosis**

In order to compare the level of aganglionosis predicted by barium enema X-ray with the pathological aganglionosis, the large intestine was divided into ten regions as rectum, distal sigmoid, prosimal sigmoid, descending colon, splenic flexure, left transverse colon, right transverse colon, hepatic flexure, ascending colon and caecum. Concordance was defined as the presence of histological aganglionosis in the same region as predicted by barium enema and discordance as presence of pathological aganglionosis in one or more segment proximal or distal to the one predicted by barium enema. In the present study, there was no case of long segment HD, the concordance/discordance estimation was limited to rectum, distal sigmoid and proximal sigmoid only.

Ten neonates who had short segment HD demonstrated by barium enema X-ray and confirmed by punch rectal biopsy underwent primary transanal endorectal pull through with frozen section facility. All were short segment HD concordant with barium enema finding (table V).

Seven neonates underwent laparotomy. Out of the 7 cases, 2 had total colonic aganglionosis. Barium enema could help the diagnosis in one case but it was not conclusive in the other. There was obvious transition zone at rectosigmoid region at laparotomy in 3 cases with definite radiological transition zone and the agaglionosis, which was later, confirmed by multiple biopsies. In the remaining 2 cases with rectosigmoid index <1 but no obvious radiological transition zone, at laparotomy there was no obvious.

Transitional zone but sigmoid colon was dilated up to the peritoneal reflection. The biopry at distal sigmoid showed ganglion cells while, rectal biopsy demonstrated absence of ganglion colls and hypertrophied nerve fibers. Thus, barum enema could predict the level of aganglionosis in all rectosigmoid disease (table V).

**Table V: Level of agangliononia by barium enema X-ray compared to operative and histologic findings**

| Level of aganglionosis | | | | | |
|---|---|---|---|---|---|
| BE | Operative/histologic finding | Concordance | Discordance | *$X^2$/P | Remark |
| Distal sigmoid | Distal sigmoid | 15 | 0 | 4.35/<0.05 | S |
| TCA | TCA | 0 | 0 | | |
| Inconclusive | TCA | 0 | 1 | | |

| | | | |
|---|---|---|---|
| *X | test with Yates correction | BE | Barium enema |
| S | Sigmificant | TCA | Total colonic aganglionosis |

The $X^2$ test with Yates correction was applied to see the level of sigmificance of concordance of barium enema. It was significant (X/p=4.35/<0.05).

## DISCUSSION

Hirschsprung's disease (HD), congenital aganglionosis of colon, is reported to be a common cause of intestinal obstruction in neonates [15]. The incidence of HD in two largest series was I in 5257 and 1 in 4697 and the estimated incidence accepted in general is 1 in 5000 live births [16].

In his first discussion of this disease, Haroid Hirschprung described the occurrence of the disease in two neonates. Later studies treated this as a disease of children, again pointed out that the diagnosis could be made in the newborn [17]. The major reason for emphasizing the diagnosis of HD in neonatal period or in early infancy has been the high mortality in this age group due to sepsis from underlying enterocolitis. Another large survey in Japan in 1983, which reviewed 1628 cases of HD, found 48.7% cases diagnosed in neonatal period [16]. Klein *et al.*, in 1984 reported 70% of HD diagnosed in neonatal period. In our institute 42.1% cases were diagnosed in the first month of life. Early diagnosis of HD is favoured by high index of suspicion of this disease in neonates presenting with intestinal obstruction.

Our study supported several of previously reported demographic parameters. We found the overall male female ratio 3.1:1, which approximated to 3:1 and 3.8:1 [16, 18]. The prematurity rate on out study was 8.1%, which was Similar to the rate reported by Klein and Philippart in 1993 [19].

In the American Academy of Pediadrics survey, Kleinhaus reported 74% short segment, 15% long segment and 8% total colonic aganglionosis (TCA) and Ikeda and Goto (1983) in Jnapanese survey found 80%, 20% and 8.5% respectively [20]. In the present study, 94.6% was short segment HD and 5.4% was TCA but no case of long segment HD. This may be due to small study population in comparison to them.

We did not have any familial cases. There are reports of familial involvement in 3% case [16]. The most common anomaly associated with 2.9% and Goto's study (1984) it was 4.35%. In our study it was 2.7%.

Majority of patents presented in autumn (37.9%) and summer (29.7%). There were 18.9% cases in spring and 13.5% in winter. This may reflect the agricultural workload, planning of deliveries, etc. Klein found higher number of patients in autumn, winter, and lower number in summer and spring [21]. They also showed that the incidence did not vary significantly from year to year.

Most of the patients belonged to poor socioeconomic class (64.86%). Only 35.14% were from the non-poor class. This may be because most of the affluent people attend private hospitals for healthcare services for their convenience.

All neonates presented with abdominal distention (100%), 29 (78.4%) with constipation and 20 (54.1%) with vomiting. This clinical picture of intestinal obstruction was also noted in other series (35.1%) cases. In Jung's study it was 53%; in Klein, Coran and Wesley, it was 58% [37]. In our study one patient (2.7%) had history assisted Passage of meconium. There was no history of passage of meconium in first 48 hours of life could not rule out the possibility of HD nor failure to pass meconium supported the diagnosis of HD. Enterocolitis was present at time of diagnosis in 2 (5.4%) patients [22].

Recently one-stage primary transanal endorectal pull through procedure is gaining popularity. As this surgical procedure depends on the level of aganglionosis, it is mandatory to determine the level of aganglionosis preoperatively. Barium enema has been increasingly relied on to predict the level of aganglionosis [23].

In the present study, initially plain X-ray abdormen in erect posture was done in all cases. Dilated loops of intestine and paucity of rectal gas were present in all cases (100%). Dilated loops of intestine with air fluid levels were present in 14 (38%) and colon cut off sign in 2 (5.4%) cases. There was no free gas in any of the cases.

In all cases (n=37) barium enema X-ray was performed. Microcolon was evident in a case of total colonic aganglionosis but it was inconclusive in another one.

The inaccuracy of barium enema X-ray in the diagnosis of HD was emphasized. Yet the diagnostic accuracy of barium enema reported in different series varies from 76-92% [24]. In our study the diagnostic accuracy of barium enema was 94.3%. This was partly because we took meticulous precaution in performing the barium enema X-ray and partly because we included rectosigmoid index as a diagnostic variable.

Punch rectal biopsy was performed in 31 (83.8%) neonates out of whom 27 (87.1%) were proved to be HD positive and 4 (12.9%) to be HD negative. Laparotomy, multiple colonic biopsies and diverting stoma were needed in 7 (18.9%) neonates as in one case both procedures were done. Extramucosal biopsies were taken from narrow segment, dilated segment and another one 3.5 cm proximal to it. Two (28.6%) neonates who had total colonic aganglionosis at laparotomy were proved subsequently by hisopathology. Remaining 5 (71.4%) were diagnosed to be HD positive and 4 to be HD negative histopathologically.

The barium enema finding of radiological transition zone, a relatively narrowed aganglionic segment distal to a dilated normal colon, was first described by Swenson visualization of radiological transition zone has been the hallmark for radiographic diagnosis of HD [25].

However, this finding is less often evident in neonates, probably the duration and/or severity of intestinal obstruction in newborn is not adequate enough to produce the changes to be evident radiologically. This has been recognized previously [27]. Therefore, failure to visualize radiological transition zone does not rule out HD in neonates.

In our study, only 15 neonates (42.9) with HD showed radiological transition zone in barium whereas proctor reported visualization of radiological transition zone 92% cases [26]. Rosenfield reported visualization radiological transition zone in 65% neonates and Klein *et al.,* (1984) in 85% neonates. In the present study radiological transition zone was not visualized in 20 (57.1%) neonates out of them 16 (45.7%) were true positive, 4 (11.4%) were true negative as confirmed by histopathology. So, for radiological transition zone alone 15 were true positive, 16 were false negative, 4 were true negative and no false positive. Thus, statistical analysis showed that sensitivity of radiological transition zone alone was 48.4%, specificity 100%, accuracy 88.5%, positive predictive value 100% and negative predictive value 20% and FN 45.7% [27]. Rosenfield in their study found the sensitivity of radiological transition zone to be 54%, specificity 100%, positive predictive value 100%, negative predictive value 54%, no false positive and 46% false negative. In both the studies the analysis was comparable.

A typical radiological transition zone may not be apparent during the first days of life. Sometimes it may be equivocal or not readily demonstrable. In such condition rectosigmoid index may be of value in the diagnosis of HD. In normal newborn and in cases of meconium plug syndrome and other pathological state of gastrointestinal tract, the rectum was commonly found to be wider than sigmoid. Io HD the widest rectal diameter is usually smaller than the widest diameter of the sigmoid even in absence of radiological transition zone. Rectosigmoid index is obviously superfluous when radiological transition zone is in the form of marked disproportion between the rec rectum and present widest dimeasurement, a numerical value obtained by dividing the widest diameter of rectum by the sigmoid colon. Rectosigmoid index is a widest diameter of sigmoid. Rectosigmoid index is <1 in HD and 21 in normal subjects and other gastrointestinal tract phthology [28].

Rectosigmoid index was not intended to be used in the rare instances of aganglionosis involving the long segment or total colon or for HD. terminal ileum. The In the present study 31 (83.7%) cases had rectosigmoid index <1 out of which 30 were HD rectosigmoid region remains the most common site positive and I was HD negative as confirmed by histopathology. In the remaining 4 subjects with rectosigmoid index 30 were HD negative and I was HD positive. So, for rectosigmoid index 30 were true positive, I was false positive, 3 true negative and I false negative. Statistical analysis showed the sensitivity for rectosigmoid index alone to be 96.77%, specificity 75%, accuracy 94.3%, positive predictive value 96.77% and negative predictive value 75% In other studies rectosigmoid index was not analysed and did not use rectosigmoid index as they found that radiological transition zone was clearly visible in all cases in which rectosigmoid index was positive [27]. But this was not observed in the present study.

When rectosigmoid index alone was considered where definite radiological transition zone was not visualized in 20 cases, the rectosigmoid index was <1 in 16 cases, of which 15 were true positive and 1 was false positive as proved histopathologically. The rectosigmoid index was 21 in 4 cases, of which 3 were true negative and I was false negative. The sensitivity for rectosigmoid index where radiological transition zone was absent was 93.7%, specificity 75%, accuracy 90%, positive predictive value 93.76% and negative predictive value 75% [27]. Pochaczevsky and Leonidas in their study found that the rectosigmoid index was less than I in all cases of histopathologically proved HD in neonates where radiological transition zone was not evident [28].

Recotosigmoid index can be assumed to be the subtle change in the Caliber of rectosigmoid produced by intestinal obstruction due to HD in early life. It is obviously superfluous when radiological transition zone is evident. Conversely, radiological transition zone con be assumed to be exaggerated rectosigmoid index. It can

be particularly applied in situation where radiological transition zone is not readily apparent in neonates.

Delayed evacuation of barium of barium is commonly present in neonates and infants with constipation and is not a reliable indicator of HD [29]. They reported high false positive rate (85%) in addition to having lowest sensitivity (57%) and specificity (27%). Late film was available for 29 cases (78.4%) in the present study. In 19 (65.5%) cases retention of barium was present, out of which 17 were HD positive and 2 were HD negative as confirmed by histopathological study. In 10 cases retention of barium was absent out of which I was HD negative and 9 were HD positive. So, for retention of barium 17 were true positive, 2 false positive, 1 true negation and 9 false negative. The sensitivity for retention of barium alone was 65.38%, specificity 33.33%, accuracy 62.1%, positive predictive value 89.5% and negative predictive value was 10%. Rosenfield found the senditivity of retention of barium to be 86%, specificity 38%, positive predictive value 69%, negative predictive value 62.5%, false positive rate 31% and false negative rate 37.5% [27].

The correlation between location of radiological transition zone and pathologic level of aganglionosis has not been well studied. Rosenfield found radiological transition zone (location not specified) to match the level of aganglionosis identified at the time of surgery in all cases compared. Procter reported that radiological transition zone was concordant in 79% of radiological transition zone matched the level of aganglionosis in all cases of rectosigmoid disease at the time of surgery [26].

Ten neonates who had short segment HDD demonstrated by barium enema and confirmed by histopathology underwent primary transanal endorectal pull through with frozen section facility. All were short segment HD concordant with barium enema finding. Seven neonates underwent laparotomy, of them 2 had total colonic aganglionosis. Barium enema could help the diagnosis in 1 case but was not rectosigmoid region in the other. There was obvious transition zone at rectosigmoid region at laparotomy in three cases as predicted by radiological transition zone in barium enema. The level of aganglionosis as predicted by barium enema was subsequently confirmed by histopathology. In 2 cases with no definite radiological transition zone but rectosigmoid index <1, there was no obvious transition zone but sigmoid colon was dilated up to the peritoneal reflection. The aganglionosis a distal sigmoid was confirmed by histopatholoty. Thus, barium enema could predict the level of aganglionosis in all rectosigmoid HD. Statistical analysis showed that the occurrence of concordance was significant ($X^2$/p-4.35/<0.05).

**Limitation of the Study**

1. The study was conducted on a small population.
2. There were no cases of long segment HD in the study.
3. The patients were followed up for a short period only.

## CONCLUSION

The result of the present study supported the following facts regarding barium enema X-ray: Visualization of radiological transition zone in barium enema is highly reliable sign of Hirschsprung's disease, but failure to visualize it does not rule out the disease. The rectosigmoid index is an objective measurement, which is accurate in the diagnosis of rectosigmoid HD in the newborn, particularly in those cases where radiological transition zone is not readily apparent or its presence is questionable. Retention of barium at 24 hours late film cannot be used alone to diagnose HD. Evacuation of barium at 24 hours does not always rule out HD. Barium enema X-ray is a sensitive, specific and accurate investigation for diagnosing HD in neonates can correctly predict the level of aganglionosis, particularly in case of rectosigmoid HD-

## RECOMMENDATION

Barium enema should be done as initial investigation in all suspected cases of Hirschsprung's disease in neonatese din are population is required to reproduce the similar results. 2 Further multicentric study larger

## REFERENCES

1. Swenson, O., Sherman, J. O., & Fisher, J. H. (1973). Diagnosis of congenital megacolon: an analysis of 501 patients. *Journal of Pediatric Surgery*, *8*(5), 587-594.
2. POLLEY JR, T. Z., CORAN, A. G., & WESLEY, J. R. (1985). A ten-year experience with ninety-two cases of Hirschsprung's disease including sixty-seven consecutive endorectal pull-through procedures. *Annals of surgery*, *202*(3), 349-355.
3. Boley S.J., Dinari G. & Cohen M.1. (1978), 'Hirschsprung's disease in the newborn', *Perinatol*, vol 5, pp. 45- 60.
4. Ricketts, R.R. (2003). "Hirschsprung's disease', in Surgical Directives: Pediatric Surgery, ed. P. Mattei, Lippincott, *Williams and Wilkins, Philadelphia*, pp. 371-376.
5. POCHACZEVSKY, R., & LEONIDAS, J. C. (1975). THE" RECTO-SIGMOID INDEX" A MEASUREMENT FOR THE EARLY DIAGNOSIS OF HIRSCHSPRUNG'S DISEASE. *American Journal of Roentgenology*, *123*(4), 770-777.
6. KE, G. (1995). Primary laparoscopic pull-through for Hirschsprung's disease in infants and children. *J Pediatr Surg*, *30*(7), 1-7.

7. Hirose, R., Hirata, Y., Yamada, T., Kawana, T., Taguchi, T., & Suita, S. (1993). The simple technique of rectal mucosal biopsy for the diagnosis of Hirschsprung's disease. *Journal of pediatric surgery*, *28*(7), 942-944.
8. Albanese, C. T., Jennings, R. W., Smith, B., Bratton, B., & Harrison, M. R. (1999). Perineal one-stage pull-through for Hirschsprung's disease. *Journal of pediatric surgery*, *34*(3), 377-380.
9. DH, T. (1998). Hirschsprung's disease and related neuromuscular disorders of the intestine. *Pediatric surgery*, *2*, 1381-1418.
10. Kleinhaus, S., Boley, S. J., Sheran, M., & Sieber, W. K. (1979). Hirschsprung's disease a survey of the members of the surgical section of the American academy of pediatrics. *Journal of pediatric surgery*, *14*(5), 588-597.
11. Cass, D. T. (1990). Neonatal one-stage repair of Hirschsprung's disease. *Pediatric surgery international*, *5*, 341-346.
12. De la Torre, L., & Ortega, A. (2000). Transanal versus open endorectal pull-through for Hirschsprung's disease. *Journal of pediatric surgery*, *35*(11), 1630-1632.
13. Teeraratkul, S. (2003). Transanal one-stage endorectal pull-through for Hirschsprung's disease in infants and children. *Journal of pediatric surgery*, *38*(2), 184-187.
14. Elhalaby, E. A., Hashish, A., Elbarbary, M. M., Soliman, H. A., Wishahy, M. K., Elkholy, A., ... & Hamza, A. F. (2004). Transanal one-stage endorectal pull-through for Hirschsprung's disease: a multicenter study. *Journal of pediatric surgery*, *39*(3), 345-351.
15. Jung, P. M. (1995). Hirschsprung's disease: one surgeon's experience in one institution. *Journal of pediatric surgery*, *30*(5), 646-651.
16. Ikeda, K. & Goto, S. (1983) 'Diagnosis of Hirschsprung's disease in Japan. An analysis 1628 patients', *Ann. Surg*, vol. 199, pp. 400-405.
17. DH, T. (1998). Hirschsprung's disease and related neuromuscular disorders of the intestine. *Pediatric surgery*, *2*, 1381-1418.
18. Russell, M. B., Russell, C. A., & Niebuhr, E. (1994). An epidemiological study of Hirschsprung's disease and additional anomalies. *Acta Paediatrica*, *83*(1), 68-71.
19. Nakayama, D. K. (1997). Critical care of the surgical newborn. *(No Title)*.
20. Kleinhaus, S., Boley, S. J., Sheran, M., & Sieber, W. K. (1979). Hirschsprung's disease a survey of the members of the surgical section of the American academy of pediatrics. *Journal of pediatric surgery*, *14*(5), 588-597.
21. Klein, M. D., Coran, A. G., Wesley, J. R., & Drongowski, R. A. (1984). Hirschsprung's disease in the newborn. *Journal of Pediatric Surgery*, *19*(4), 370-374.
22. Jung, P. M. (1995). Hirschsprung's disease: one surgeon's experience in one institution. *Journal of pediatric surgery*, *30*(5), 646-651.
23. Elhalaby, E. A., Hashish, A., Elbarbary, M. M., Soliman, H. A., Wishahy, M. K., Elkholy, A., ... & Hamza, A. F. (2004). Transanal one-stage endorectal pull-through for Hirschsprung's disease: a multicenter study. *Journal of pediatric surgery*, *39*(3), 345-351.
24. Yadav, L. (2017). *Biology of Hirschsprung Disease: Pathomorphological, Histochemical, Immunohistochemical and Genetic (RET Gene) Study of the Enteric Nervous System* (Doctoral dissertation, Rajiv Gandhi University of Health Sciences (India)).
25. Swenson, O., Sherman, J. O., & Fisher, J. H. (1973). Diagnosis of congenital megacolon: an analysis of 501 patients. *Journal of Pediatric Surgery*, *8*(5), 587-594.
26. Proctor, M. L., Traubici, J., Langer, J. C., Gibbs, D. L., Ein, S. H., Daneman, A., & Kim, P. C. W. (2003). Correlation between radiographic transition zone and level of aganglionosis in Hirschsprung's disease: Implications for surgical approach. *Journal of pediatric surgery*, *38*(5), 775-778.
27. Rosenfield, N. S., Ablow, R. C., Markowitz, R. I., DiPietro, M., Seashore, J. H., Touloukian, R. J., & Cicchetti, D. V. (1984). Hirschsprung disease: accuracy of the barium enema examination. *Radiology*, *150*(2), 393-400.
28. POCHACZEVSKY, R., & LEONIDAS, J. C. (1975). THE" RECTO-SIGMOID INDEX" A MEASUREMENT FOR THE EARLY DIAGNOSIS OF HIRSCHSPRUNG'S DISEASE. *American Journal of Roentgenology*, *123*(4), 770-777.
29. Taxman, T. L., Yulish, B. S., & Rothstein, F. C. (1986). How useful is the barium enema in the diagnosis of infantile Hirschsprung's disease?. *American journal of diseases of children*, *140*(9), 881-884.