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Barium enema with reference to rectal biopsy for the diagnosis and exclusion of Hirschsprung disease

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Background: Hirschsprung disease is congenital disease caused by a lack of ganglion cells in the distal bowel wall which results in functional obstruction of the aganglionic segment due to failure of relaxation during peristalsis. Barium enema is the best imaging modality to diagnose Hirschsprung disease but the gold standard to confirm the diagnosis is biopsy.
Methods: A retrospective study to assess barium enema accuracy relative to rectal biopsy in diagnosing Hirschsprung disease was done at Tikur Anbessa Specialized Hospital and data were collected from August 1st to 14th 2010. Seventy one patients who had undergone both rectal biopsy and barium enema examination between January 2002 and December 2009 were studied. In this study patients' record were reviewed and radiologic findings were compared with histopathology results. The results were analyzed using SPSS16.
Results: In our study the mean age at diagnosis was 19 months which is a late diagnosis compared to other studies. Hirschsprung disease was more common in males and recto sigmoid was the most common area of transition zone. Large number of patients had emergency operation and diagnosis was late. Total percent agreement of barium enema and biopsy to diagnose HD is 79.1% and Kappa agreement of 0.34. In children greater than 1 year old accuracy of barium enema was very high (91.1%).
Conclusion and recommendation: Sensitivity of barium enema is less in neonates. If barium enema has positive result intervention can be done safely, especially in children above 1year old. Biopsy is not always necessary for diagnosing Hirschsprung disease.

Introduction

Hirschsprung disease(HD) is frequently encountered problem in our country. The disease can present in neonatal period with acute intestinal obstruction. Associated complications can be seen especially if it is diagnosed late.
The disease more frequently presents in infancy, although some patients present with persistent and severe constipation later in life. Clinical presentation in infants includes vomiting, poor feeding, poor weight gain, and progressive abdominal distension.
Early diagnosis is essential to prevent complications such as enterocolitis and colonic rupture. A rectal suction biopsy can detect hypertrophic nerve trunks and the absence of ganglion cells in the colonic submucosa, confirming the diagnosis.
The radiological diagnosis of Hirschsprung disease is possible if films of diagnostic quality can be produced. Contrast enema studies are affordable and at the same time associated with minimal risk of complications. The choice of the best diagnostic modality or developing criteria-based diagnoses of Hirschsprung

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disease from a combination of the available diagnostic modalities, may avoid or reduce the complication rates associated with late diagnosis.

Patients and Methods

This is a cross-sectional retrospective study conducted in Tuikur Anbessa Specialized Hospital, Addis Ababa. The data were extracted from pediatric surgical operating room registered file of patients with HD from January 2002 to December 2009. The study subjects were all Ethiopian patients in pediatric age group. All study participants had both barium enema examination result and rectal biopsy result. thus 71 participants were recruited. Data were collected using a structured data collection format by principal investigator. Patient card number was retrieved from paediatric operating room registry and cards were retrieved from registration department of the hospital. Individual patient records were reviewed.

Data including demographics and presenting symptoms were extracted. It also included barium enema final radiological diagnosis and site of transition zone of Hirschsprung disease. The result of pathology specimen was also filled from patient chart. Radiological diagnosis of barium enema was reported by final year radiology resident which were always done after consultation of senior radiologists. Biopsy results of patients were also reported by senior pathologists in Tikur Anbessa Specialized Hospital pathology department.
Biopsy specimens were taken by senior paediatric surgeons.

The collected data were checked for completeness and cleaned. Statistical Package for Social Sciences for window (SPSS) version 16 was used to enter and analyze the data. The total agreement between barium enema and rectal biopsy in the diagnosis of HD was calculated. Kappa agreement between two investigation modalities was also measured. Permission to conduct the research was given from the department of radiology and IRB of College of Health Sciences. Code number rather than patients name was used for data collection. The data collected were also confidential and only analyzed by the principal investigator.

Results

From total 71 patients who were diagnosed to have Hirschsprung disease and have both biopsy result and barium enema study 58 (81.7%) were males and 13 (18.3%) were females. The male to female sex ratio was 4.5 to 1.Their age range distribution was 7(9.9%) less than 30 days, 29(40.8%) between 1 month and 1 year and 35 (49.3%) were greater than 1 year (Table 1). The mean age at diagnosis of Hirschsprung disease was 19months.

Table 1. Age Distribution of Patients underwent both Rectal Biopsy and Barium Enema.

Age in Months	Frequency	Percentage
<30 days	7	9.9
1month to 1year	29	40.8
> 1year	35	49.3
Total	71	100

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Table 2. Distribution of Barium Enema findings.

Barium Enema Findings	Frequency	Percentage
Suggested HD	53	74.6
Normal	13	18.3
Other diagnosis	5	7.0
Total	71	100.0

Table 3. Barium enema results with pathology in patients underwent both rectal biopsy and barium enema at Tikur Anbessa Specialized Hospital from January 2002 to December 2009.

Pathology results	Barium enema result			Total
	Suggested HD	Normal	Other diagnosis	
No ganglion cell	**49**	**8**	**5**	**62**
Ganglion cells seen	**1**	**4**	**0**	**5**
Non conclusive	**3**	**1**	**0**	**4**
Total	**53**	**13**	**5**	**71**

Most patients 43 (60.6%) presented with constipation from birth, 23 (32.4%) had both constipation and abdominal distension and the remaining five presented with acute obstructive symptoms. Barium enema studies in 53(74.6 %) suggested HD, 13 (18.3%) had a normal study and 5 (7%) were reported as showing other diagnosis such as micro colon in 2 and meconium plug syndrome in 2 (Table 2). From the 53 (74.6%) patients whose barium enema study result suggested HD; 25 showed inversion of rectosigmoid ratio as evidence and in 31 patients' site of transition correctly identified at rectosigmoid area. Transitional zone was reported at proximal colon in one patient and at descending colon in 2 cases.

Rectal biopsy was conclusive and identified HD in 62(88.7%) patients with no ganglion cells visible, 5 (7%) cases were reported as normal with visible ganglion cells and 4(5.6%) cases as non-conclusive. The reason for being inconclusive biopsy result was not including the sub mucosal layer and inadequate specimens (Table 3). Total percent agreement of barium enema and biopsy to diagnose HD is 79.1% and barium enema and rectal biopsy have Kappa agreement of 0.34. In children greater than 1 year old accuracy of barium enema is high (91.1%).

Discussion

In this study mean age at diagnosis of HD is 19 months which is a delayed diagnosis compared with other studies. A report from India found the mean age at diagnosis to be only 18 days[1,2]. our study excluded neonates and infants who presented with acute intestinal obstruction[3] and failure to pass meconium since rectal biopsy was taken after emergency surgical intervention without barium enema study. This study also showed HD is common in males 81.7% and in majority of cases recto sigmoid disease is seen. This is comparable with other studies[4,5,6,7].

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In this study a strong relationship was noted between a positive result of barium enema and age. Only in 2 of 7 neonates was the barium enema consistent with biopsy.. Barium enema diagnosis of neonates and infants is difficult and some authors recommended that neonates who do not pass meconium in the first 48 hours of life should undergo rectal suction biopsy to establish the diagnosis of congenital megacolon[8,9]. Total percentage agreement of barium enema and biopsy to diagnose HD is 79.1% and Kappa agreement of 0.34. Landis and Koch suggested kappa agreement less than 0.4 represents poor agreement[10]. This can be explained by poor agreement seen between barium enema and rectal biopsy in neonates. In children greater than 1 year old accuracy of barium enema is high (91.1%). One study indicated diagnosis of HD with barium enema findings in children over 2 years could reach a diagnostic accuracy of up to 100%[2].

Most authors took radiological demonstration of a transition zone as the most important diagnostic sign on barium enema but actual detection rate is low[3,11]. In our study transition zone is only reported in 46% of patients diagnosed to have HD. Other important signs like retention of barium 24 to 48hours and stool mixed with barium correlated better with the presence or absence of HD than did any of these features alone[10].

In a country with limited resources barium enema can be used as first line imaging and combined with anorectal manometry or biopsy[12,13,14].

Conclusion and Recommendation

Sensitivity of barium enema is less in neonates. If barium enema is diagnostic intervention can be done safely, especially in children above 1year old. Biopsy is not always necessary for diagnosing Hirschsprung disease. Mean age at diagnosis is late in our study and further study may be needed to know the reason.

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