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Neonatal Presentation Of Hirschsprung's Disease Infants At A Tertiary Care Centre, A Prospective Study

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Abstract

Introduction : Hirschsprung's disease is a common cause of neonatal intestinal obstruction. .We conducted a study to evaluate the presentation of Hirschsprung's disease in neonatal and infantile period to aid in early diagnosis and timely management.

Material and Methods: It was a prospective cohort study of newborn and young infants with features of Hirschsprung's disease conducted in our department from Oct. 2014, till Oct. 2018. The diagnosis was based on history, clinical examination, water soluble contrast enema and a definitive rectal biopsy. Transition zone was confirmed by intra-operative frozen section.

Discussion: Total of 25 patients had the disease. Delayed passage of meconium was seen in23 patients (92%). Features of failure to thrive were reported in 40%. Abdominal distension and features of obstruction were seen in 80%. Preoperative rectal biopsy was done for confirmation of diagnosis and intra-operative frozen section for delineation level of aganglionosis. Early diagnosis resulted in primary pull-through in most of the cases.

Conclusion: It is important to rule out Hirschsprung's disease in neonates presenting with delayed passage of meconium and abdominal distension when other obstructive causes stand ruled out.

Keywords: Hirschsprung's disease(HD), Delayed Passage of meconium (DPM)

Introduction

Hirschsprung's disease is the total or partial absence of ganglion cells. As a result the disease presents with features of functional intestinal obstruction in neonates (delayed passage of meconium, constipation, abdominal distension) and associated poor feeding and failure to thrive(1). Among normal full-term infants, 98% pass meconium in the first 24 hours of life and the remainder will pass their first stool by 48 hours [1] It has always been said that over 90% of HD infants fail to pass meconium in the first 24 hours of life [2]. HD can sometimes present with gut prtforation.With a suspicion of HD, irrespective of the site of perforation and the cause of perforation, the neonate has to be surgically managed. The diagnosis can be made with radiographic studies , anorectal monometry and a rectal biopsy [3]. Rectal biopsy is the gold standard for the diagnosis of HD (5).

Material and Methods:

The study was prospective cohort study. It was conducted in the Department of Pediatric Surgery, SKIMS, Soura from Oct. 2014 to Oct. 2018. After proper approval by the institutional ethical committee, all the neonates and infants with suspected Hirschsprung's disease were admitted, registered and evaluated on inpatient basis.

Patients were diagnosed as Hirschsprung's disease based on clinical history, radiological studies and

full-thickness rectal biopsy. Water-soluble contrast helped identify a transition zone between a narrowed aganglionic and dilated, otherwise normally innervated segments. A rectal biopsy was done in patients to confirm HSD . On-table rectal frozen was done to confirm level of aganglionosis.

With a suspicion of HD, patients presenting with perforation , the neonate has to be surgically managed.

Once diagnosed patients were put on a definite rectal wash-out protocol. Parents were taught how to give home based colonic washes with normal saline using a 16 french soft drain till the patient was optimised for a primary pull-through and attained a weight significant enough to withstand a major surgery. Decision regarding timing of surgery in infants diagnosed as Hirschsprung's disease was taken depending on the absence of features of enterocolitis, significant weight gain of around 5kgs and age of baby feasible to withstand significant anesthetic and surgical risks. This decision was tailored to individual cases.

Patients underwent pull-through by different methods depending on patients age, segment involved, feasibility of performing laparoscopy, and clearity of transition zone. In case tasransition was not still seen levelling biopsies were taken to do a pull-through at a later stage.

Results

During this study period 25 children were diagnosed as having Hirschsprung's disease and only 20 patients underwent primary pull-through and were included in the study. Table 1 and 2 show the clinical presentation and results of water soluble contrast study in these patients.

Discussion:

92% patients in our study presented in neonatal period mostly with a history of delayed passage of meconium. In a study by Singh SJ et al[7] reported neonatal presentation was seen in (90%) cases. Similarly Schäppi MG [8] in his study has advocated that > 90% affected children symptoms start in the neonatal period and less than 1% are diagnosed during adult life.

We demonstrated in our study that features of Hirschsprung's and clear transition zone were seen in only 80% patients on contrast studies. So water soluble contrast study has a low sensitivity. This finding is supported by the observation by Schäppi MG [8] et al who has documented a low sensitivity and specificity of contrast enema in comparison with biopsy. However he has advocated that contrast enema can be used as an additional investigation to assess the length of aganglionic segment before surgery. Contrast enema has an important role in the diagnosis of HD. Because of high false positive rate with a strong suspicion of HD the test should always be supplemented by a rectal biopsy.

Conclusion

It is important to rule out Hirschsprung's disease in neonates presenting with delayed passage of meconium and abdominal distension when other obstructive causes stand ruled out. Although rectal contrast study is the initial investigation , rectal biopsy is mandatory for diagnosis.

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Table 1		
Symptoms	Frequency	Percentage
DPM	23	92
Features of failure to thrive	10	40
Abdominal distension and features of obstruction	20	80
Blast sign on DRE	7	28
Hirschsprung's associated enterocolitis	0	0
Constipation	20	80

Table 7: Rectal contrast showing features of HSD in study patients

Features of HSD on rectal contrast study	**Frequency**	**Percentage**
Present	20	80
Absent	5	20
Total	25	100

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