Original Research Article

Incidence and outcome of neonatal Hirschprung's disease in a government tertiary care setup

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Abstract

Objective: The main objective of this study was to know the Incidence and outcome of neonatal Hirschprung's Disease in a government tertiary care setup. **Methods:** It was a cross sectional retrospective study, neonates admitted with history of abdominal distension, delayed passage of meconium at birth and constipation into NICU of government tertiary care center, Mysore were included. The information was obtained over a period of 6 months between January 2017 to June 2017 by collecting the case details from medical records. **Results:** There were total admission of 1531 in the NICU between January 2017 to June 2017.Out of which 45 neonates presented with different complaints , 26(57.7%) neonates presented with abdominal distension,17(47.7%) Presented with delayed passage of meconium, 2(4%) neonates presented with constipation Out of 45 cases presenting with above complaints, only 16 cases were diagnosed to be Hirschprung's disease. Out of 16 cases of Hirschprung disease, 14 cases were operated with primary procedure being the reversible colostomy. Two cases could not be operated, cases of sepsis with severe hemodynamic instability and both babies succumbed to death before surgery. Out of 14 cases which underwent primary surgery, 13 cases colostomy functioned well, babies were started on feeds by 48 hours, improved and discharged, one case died of sepsis following primary surgery **Conclusion:** Hirscprung's disease is found to be one of the common surgical condition in our NICU with incidence being 35.5% and therefore high clinical suspicion, appropriate investigations and early surgery has better outcome

Key Word: Hirscprung's disease, Constipation, Neonates

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INTRODUCTION

Hirschprung's disease is congenital disorder of colon involving both myenteric and submucosalplexures, where primary pathogenic entity is absence of enteric ganglionic cells. The first report of Hirschprung's disease dates to 1961 described by dutch anatomist Fredrick Ruysch.[1] Hirschprung's disease affects all races; however, it is roughly 3 times more common among Asian-Americans. The typical clinical signs in the neonatal period are of abdominal distension, delayed passage of meconium at birth and constipation. More than 80% of all Hirschsprung's cases present symptoms in the neonatal period.[2] The Gold standard investigation is histopathological examination of a rectal biopsy specimen, showing the absence of ganglia structures. Acetyl-cholinesterase staining reveals hypertrophied nerve trunks throughout the lamina propria and muscularispropria layers of the bowel wall.[2]Early diagnosis and prompt treatment of Hirschsprung's disease will result in a significantly improved quality of life for the patient, and may alleviate potentially life-threatening complications.

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PATIENTS AND METHODS

It was a cross sectional retrospective study ,neonates admitted with history of abdominal distension, delayed passage of meconium at birth and constipation into NICU of government teritary care center, Mysore were included. The information was obtained over a period of 6 months between January 2017 to June 2017 by collecting the case details from medical records.

Statistical Analysis: The data was analysed by frequency, percentage, and mean.

RESULTS

There were total admission of 1531 in the NICU between January 2017 to June 2017.Out of which 45 neonates presented with varying complaints of abdominal distension, delayed passage of meconium and constipation , 26(57.7%) neonates presented with abdominal distension,17(47.7%) Presented with delayed passage of meconium, 2(4%) neonates presented with constipation(table 1).

Table 1: symptoms wise distribution of cases

Symptoms	Number of cases (%)
Abdominal distension	26(57.7%)
Delayed passage of meconium	17(47.7%)
Constipation	2(4%)

Out of 45 cases presenting with above complaints, only 16 cases were diagnosed to be Hirschprung's disease and the remaining cases were as follows (fig 2).

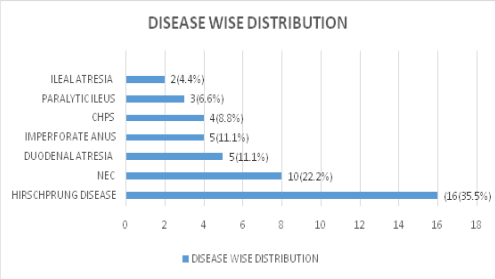

Figure 1: Disease wise distribution of cases

Out of 16 neonates suspected with Hirschprung's Disease, 13(81%) were males and 3(19%) were females. Biopsy was done for all the cases out of which 12 (75%) were biopsy positive for Hirschprung disease. Out of 16 cases of Hirschprungdisease, 14 cases were operated with primary procedure being the reversible colostomy. Two cases could not be operated, cases of sepsis with severe hemodynamic instability and both babies succumbed to death before surgery. Out of 14 cases which underwent primary surgery, 13 cases colostomy functioned well, babies were started on feeds by 48 hours, improved and discharged, one case died of sepsis following primary surgery.

DISCUSSION

Hirschsprung Disease is a disease that should be diagnosed in the newborn period. Hirschsprung Disease should be considered in any newborn that fails to pass meconium within 24-48 hours of birth.[4]Currently, approximately 90% of patients with Hirschsprung disease are diagnosed in the newborn period.[6]Untreated aganglionicmegacolon in infancy may result in a mortality rate as high as 80%. Operative mortality rates for any of the interventional procedures are very low. Even in cases of treated Hirschsprungdisease, the mortality rate may approach 30% as a result of severe enterocolitis[5] If Hirschsprung disease is suspected, neonates and children should be assigned to a center where pediatric specialists areavailable to make the diagnosis and to provide definitive care.[7] Consult with pediatric surgeons and pediatric gastroenterologists. Genetic consultation may be indicated (if a heritable or chromosomal anomaly is suspected).It is important that conversations between patients and/or caregivers and clinicians take place regarding transitions in carefrom pediatric providers to adult providers, and that plans for transitional care be implemented early.[8] Hirschsprung disease cannot be prevented; however, perceptive clinical acumen may prevent delays in the diagnosis

Limitations of our study

- Small sample size
- Shorter duration of study

CONCLUSION

Hirschsprung's disease is an uncommon cause of a common condition. Early detection of the disease will lead to a much improved quality of life and may avert potentially lethal complications. In an infant or young child with intractable constipation from birth, which is resistant to conservative management, Hirschsprung's disease is a diagnosis which needs to be considered and excluded. Hirscprung's disease is found to be one of the common surgical condition in our NICU with incidence being 35.5% and therefore high clinical suspicion ,appropriate investigations and early surgery has better outcome.

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