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Original Article

Clinical Profile and Outcomes in Neonates with Hirschsprung's Disease in A Tertiary Center: A Retrospective Observational Study

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Abstract

Background: Hirschsprung's disease is a developmental disorder characterized by the absence of ganglia in the distal colon. It is one of the common surgical causes of intestinal obstruction in neonates with incidence of 1 in 1500 live births. This condition requires multidisciplinary care including neonatal and surgical expertise for better outcome. **Aims and objectives:** To describe the clinical profile, complications and outcomes in neonates with Hirschsprung's disease. **Methods:** This was a retrospective observational study of neonates with biopsy proven Hirschsprung's disease admitted in NICU between Jan 2018 to June 2022. The data was collected from hospital records, tabulated, and analyzed using standard statistical analysis. **Results:** A total of 43 Neonates with Hirschsprung's disease were included in the study. Majority were term neonates (86%) with increased male preponderance. Age of presentation being 2nd-5th day. 12% were low birth weight, 20% had sepsis. 16% had shock, 16% had thrombocytopenia, 9% had MODS, 52% required blood transfusion. 18% had renal failure. 14% had coagulopathy. 2% had wound dehiscence. 2% had wound infection. 16% required mechanical ventilation. 9% had CLABSI. 72% had growth failure at the time of discharge. 25% had associated anomalies. Mortality rate was found to be 13%. **Conclusion:** Early identification and transportation of neonates with Hirschsprung diseas may help in decreasing the mortality. Maintenance of infection control is important as there was increase in sepsis in postoperative period. Proper nutrition, early enteral feeding and good post-operative care can improve the outcome.

Key words: Neonatal, Hirschsprung's disease, morbidity and mortality, gastrointestinal anomalies

Introduction

Hirschsprung's disease is a developmental disorder characterized by the absence of ganglia in the distal colon, resulting in a functional obstruction [1]. It is one of the common surgical causes of intestinal obstruction in neonates with reported incidence of 1 in 1500 live births [2]. Common clinical presentation is delayed passage of meconium, bilious vomiting, abdominal distention, poor feeding. It may be associated with other co morbidities like infection, prematurity, cardiac, vertebral, cranial or limb anomalies and failure to gain weight. These neonates are also prone for complications like perforation, sepsis, surgical site infection, wound dehiscence, prolapse of stroma, anastomotic leak etc.. [3]. Untreated ganglionic mega colon in infancy may result in a mortality rate as high as 80%. Hence this study was planned.

Aims and objectives: To describe the clinical profile, complications, and outcomes in neonates with Hirschsprung's disease

Methods

Data of Neonates with biopsy proven Hirschsprung's disease admitted in NICU between Jan 2018 to June 2022 over a period of 5 and half years was collected from hospital records. Neonates with other causes of intestinal obstruction were excluded from the study. Baseline demographic data, clinical details, clinical variables, age of presentation, presence of associated malformations, laboratory data and other variables related to outcome were collected, tabulated and analyzed by standard statistical analysis

Sample size- those cases in the study period were included and those with incomplete or missing data were excluded

Ethics statement: The Ethics Committee of the Indira Gandhi institute of child health waived the need for ethics approval and patient consent for the collection, analysis, and publication of

the retrospectively obtained and anonymized data for this non-interventional study.

Data management and analysis:

The data from the present study was systematically collected and compiled in Microsoft Excel, then statistically analyzed using the Statistical Package for the Social Sciences (SPSS) 26 to draw relevant conclusions. Categorical data was presented as numbers and percentages, while parametric data was presented as mean ± standard deviation (SD). The student's t-test was used to analyse continuous variables. Categorical data was analysed using the Chi-square test or Fisher's exact test, as appropriate. A significance level of $p<0.05$ was considered significant, and $p<0.001$ was considered highly significant.

Results

A total of 43 cases were included in the study. Majority of the neonates (86%) presented in the 1st week of life with majority of neonates being term, (86%) with male predominance (81%).

Most common presentation was abdominal distension followed by bilious vomiting and delayed passage of meconium. Short segment Hirschsprung disease was the most common type and nearly half of the cases were treated with colostomy. Nearly 14 % of the cases had sepsis at admission. Average duration of NICU stay was around 14 days. Post-operative sepsis was seen in nearly 20 % of the cases.

Mortality rate was found to be 13% with septic shock being the leading cause of death in these patients and the Mean duration of NICU stay was 14 days. Twenty five percent of cases had associated other congenital anomalies. Antenatal GDM, resuscitation at birth, condition at admission and post-operative complications were found to be important predictors of mortality.

Table 1: Clinical details, course, and complications

Parameters	**Values**
Age of presentation#(days)	5(2-5)
Presenting complaints *	
Abdominal distention	31(72%)
Bilious vomiting	5(11.6%)
Delayed/ non passage of meconium	6(13%)
Feed intolerance	1(2%)
Condition at admission *	
Shock	4(9%)
Respiratory distress	9(20%)
Hypoglycemia	4(9%)
Sepsis at admission*	6 (14%)
gram positive	1 (16%)
gram negative	3(50%)
fungal	2 (33%)
Type *	
Short segment	30 (69%)
Long segment	13(31%)
Procedure done*	
Colostomy	19 (45%)
Pull through procedure	1 (2%)
Congenital malformation*	
External	4 (9%)
Cardiac	7 (16%)
Vertebral	1(2%)
Mechanical ventilation *	7(16%)
Growth failure @	27(72%)
Duration of NICU stay (days) $	14±11 days
Duration of TPN(days)$	7±6 days

* numbers (percentage), #Mean (interquartile distance) @ only neonates who were discharged are considered (37), $ mean± SD

Table 2: Antenatal, At birth and demographic details

Parameters	**Values**
Mother's age$	23±3
PIH*	3(6.9%)
Antenatal drugs*	3 (6.9%)
GDM*	1(2.3%)
Amniotic fluid volume*	
Polyhydramnios	3 (6.9%)
Oligohydramnios	2 (4.6%)
Type of delivery*	
NVD	27 (62%)
LSCS	16 (37.2%)
Gestational age*- Term	37 (86%)
Sex* -Male	35 (81%)
Birth weight$	2763±405

$ mean± SD * numbers (percentage)

Table 3: Post-operative complications

Parameter	**Value**
Sepsis 72 hrs. after surgery*	9 (21%)
Shock*	7 (16%)
Thrombocytopenia *	7 (16%)
MODS*	4(9%)
Renal failure *	8(18%)
Coagulopathy *	6(14%)
Wound dehiscence*	1(2%)
Prolapse of stroma*	1(2%)
Need for Re exploration*	1(2%)
Wound infection *	1(2%)
CLABSI*	4 (9%)
PNALD*	1(2%)

* numbers (percentage)

Table 4: Comparison of various significant factors among the survivors and expired cases

Variables	Discharge (37) #	Death (6)#	P value
Resuscitation	5.4%	33.2%	0.029 *
Condition at admission			
Lethargic	18.9%	66.4%	0.013*
Respiratory failure	0%	66.4%	0.003*
Shock	2.7%	49.8%	0.002*
Hypoglycemia	2.7%	49.8%	0.002*
Post-operative complications			
Shock	1%	99%	0.001*
Thrombocytopenia	1%	99%	0.001*
MODS	0%	66%	0.000*
Renal failure	5%	49%	0.033*
Coagulopathy	0%	99%	0.000*
Wound dehiscence	1%	16.6%	0.012*
Prolapse of stroma	0%	33.1%	0.012*
PNALD	0%	16.6%	0.012*

percentage in each group who had the particular variable, * significant predictors of mortality

Discussion

Hirschsprung's disease is a developmental disorder characterized by the absence of ganglia in the distal colon, resulting in a functional obstruction. It is one of the common surgical causes of intestinal obstruction in the neonates. The incidence is reported as 1 in 1500 live births. A total of 43 cases were which were admitted in our NICU over the period of last 5 years were included. Polyhydramnios is found to be associated with Hirschsprung's according to study done by Vermesh M [4]et al and in our study 3 cases had Polyhydramnios, but is nonspecific to help in antenatal diagnosis, and approximately 92% of mothers with HSCR babies had a normal antenatal history in a study done by Moore SW et al [5], in line with our study. Majority of the neonates (86%) presented in the 1st week of life with mean age of presentation being 5th day, while previous studies had the mean age of presentation was 18th day [7]. This might be due to early diagnosis and referral due to increased awareness about the disease. Hirschsprung's disease has

previously been recognized as being rare or absent In premature babies, although it is not excluded in this group. In study done by Sharp NE et al [6], 12.2% of babies weighed under 2.5 kg at birth, illustrating its low prevalence in this group. More recent reports [6] indicate a rising prevalence in premature babies. In our study only 14% were preterm which is in line with study done by Ali et al [7]and many other studies. There was a male predominance in our study with male: female ratio being 4.5:1 which was similar to the international reported ratio of 2.9:1 to 4.5:1. [8]. Condition at admission was found to be statistically significant predictor for mortality, emphasizing on the significance on early diagnosis, prompt referral and transport of the neonate. In our study most common presentation was abdominal distension followed by non-passage or delayed passage of meconium and bilious vomiting which was similar to study by Pini Prato A et al[9]. A majority cases had short segment Hirschprungs which was consistent with Ali et al whose study showed most of the patients treated surgically had a short segment disease (70.5%). [7].

Nearly 20% cases had postoperative sepsis, which could be due to prolonged TPN administration, decreased intestinal motility and bacterial translocation in these infants. Growth failure at the time of discharge was present in 72% emphasizing the importance of nutrition in post-operative period. Associated anomalies was present in 25% of which 16% neonates had cardiac anomalies. In study done by Klein MD et al [10] 35% had associated congenital anomalies, implying that screening the neonates for cardiac anomalies in Hirschprungs disease help in early diagnosis. Mortality rate was found to be 13% with septic shock being the leading cause of death in these patients whereas Infection and malnutrition were identified as two of the main causes of a high post-operative mortality of 35% in study done by Momoh JT [12]

Conclusions

Hirschsprung's disease is one of the most common surgical cause of intestinal obstruction in neonates. Early identification and appropriate postoperative care will yield better outcomes.

Data Availability: The datasets used and/or analyzed during the current study available from the corresponding author on reasonable request.

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Author's contributions

Sangeeta CS.; Prathik BH. Shared in collection of data, carried out the study, designing and coordinated the implementation of the study.

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None

Conflict of interest

None

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