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Original Article

Hirschsprung's disease presenting in the neonatal period in Jos,Nigeria

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Abstract:

***Background*:** While most cases of Hirschsprung's disease are diagnosed during the neonatal period in developed countries, majority of the cases present outside the neonatal period in developing countries. We reviewed our experience with Hirschsprung's disease presenting during the neonatal period to document the presentation and management.
***Patients/Methods*:** A retrospective analysis of the presentation and management of 31 neonates with Hirschsprung's disease over a nine year period in a Tertiary pediatric surgical centre in Nigeria was performed.
Results: From January 1996 – December 2004, 78 children were managed for Hirschsprung's disease in our unit. Thirty-one (39.7%) were aged 30 days or below. The median age at presentation was eight days (range 2-30 days). There were 23 boys and 8 girls. The median weight at presentation was 2.8kg (range 2.3 – 4.5kg). Fifteen weighed <2.5kg. Twenty-two presented with acute intestinal obstruction. Three presented with enterocolitis while in five there was delayed passage of meconium and recurrent constipation. Barium enema was done in 25 and rectal biopsy confirmed diagnosis in 29 babies. In two babies with total colonic aganglionosis (TCA), diagnosis was confirmed by colonic biopsy at laparotomy. Twenty nine had right transverse loop colostomy in the neonatal period; four were under local anesthesia, while the two children with TCA had ileostomy. Four children died before definitive surgery, two with TCA and two with enterocolitis. None had primary pull-through. Two children were lost to follow up after colostomy formation. Twenty two had definitive surgery between the ages of three months and five years. Fifteen had Boley's endorectal pull-through with a single mortality; while seven had Swenson's pull-through, one of whom died.
***Conclusion*:** Few cases of Hirschsprung's disease present during the neonatal period in our environment, many presenting with intestinal obstruction. A high index of suspicion is needed for the early diagnosis of Hirschsprung's disease in environment with limited diagnostic facilities.
Key words: Hirschsprung's disease, neonate, intestinal obstruction.

Introduction

Hirschsprung,s disease is a common cause of bowel obstruction, usually presenting in the neonatal period[1-3]. Diagnosis and treatment is most commonly during the neonatal period in developed countries[2,3] . In Northern Nigeria less than half of the cases of Hirschsprung's disease are diagnosed during the neonatal period [4,5]. This is a review of our experience with Hirschsprung's disease during the neonatal period at a tertiary pediatric surgical centre in North Central Nigeria.

Patients/Methods

Seventy eight children with Hirschsprung's disease were managed at our institution over a 10 year period (January 1996 – December 2004). Thirty-one (39.7%) of them were aged 30 days or below at presentation. The case records of these children were retrospectively reviewed for age at presentation, weight, presenting features, diagnosis, initial treatment, definitive treatment and outcome. **Results** Twenty-three (74.2%) were boys, eight (25.8%) were girls. Their ages at presentation ranged from 2 – 30 days (median 8 days). The weight at presentation ranged from 2.3 – 4.5kg (median 2.8kg). Fifteen (48.4%)weighed less than 2.5kg at presentation. The clinical presentation is summarized in table 1. There was a history of delayed passage of meconium in 27 (87%). In 11 of the children there was delay for 48 hours, in eight the delay was for 72 hours, in three the delay was for four days while the remaining five children passed meconium 7 days after birth. Twenty two presented with features of intestinal obstruction: abdominal distention and bilious vomiting. Five presented with delayed passage of meconium and recurrent constipation; diagnosis was made from

barium enema and rectal biopsy .Three patients resented with enterocolitis - explosive diarrhea and fever.They were receiving treatment for neonatal sepsis before referral to the unit. One child passed meconium within 24 hours of birth, but started having recurrent constipation at the age of 2 weeks. She presented to the hospital at the age of 30 days. Two neonates had total colonic aganglionosis and presented with intestinal obstruction for which laparotomy was done. Three children presented with concurrent inguinal hernia; one being bilateral. Plain abdominal radiographs done in the 22 neonates with intestinal obstruction showed multiple fluid levels in all of them. Barium enema was done in 25 neonates. The barium enema showed a typical change in large bowel caliber between a small or normal distal bowel and an abnormal dilated proximal bowel in 17 (68%) neonates. All the 31 neonates had rectal biopsy . Management of the neonates with acute intestinal obstruction consisted of resuscitation and administration of broad spectrum antibiotics. After stabilization a right transverse loop colostomy was constructed. In five neonates other causes of intestinal obstruction could not be excluded so emergency laparotomy was done. Ileostomy was constructed in two children who had transition zone at the distal ileum. The remaining three children had right transverse loop colostomy. The three neonates that presented with enterocolitis had an initial resuscitation with intravenous fluids and broad spectrum antibiotics and when they were cinically stable, they had rectal biopsy and right transverse loop colostomy was done later. Four of the colostomies were done under local anesthesia using 2% lignocaine; as the patients were too sick to withstand general anesthesia. The site of aganglionosis is shown in table 2. More than 50% of the neonates had transition zones at the sigmoid colon. Colostomy complications included prolapsed colostomy in six children and skin excoriation in 3 children. Two children were lost to follow up after colostomy while one other child returned to hospital 18 months after colostomy formation with enterocolitis. Twenty two children had definitive pull-through between three months and five years of age. Fifteen had endorectal pull-through procedure while seven had Swenson's pull through. Six (27.2%) patients developed post operative complications which are shown in table 3. Death was recorded in seven (22.6%) patients ; four before definitive surgery and three after pull through (one each after endorectal pull through and Swenson's procedure), one other child who was awaiting colostomy closure after endorectal pull-through sustained major burns from hot water at home and died from complications of burns in the hospital.

.**Table 1**: **Symptomtomatologys in 31 neonates**

Symptom	**No (%)**
Delayed passage of meconium	27(87)
Abdominal distension	22(71)
Bilious vomiting	22(71)
Fever	3(10)
Diarrhoea	3(10)
Constipation	1 (3)
Total	31(100)

Table II: **Site of aganglionosis**

Site	**No(%**
Rectum	6(19.0)
Sigmoid colon	18(58.0)
Descending colon	3(10.0)
Splenic flexure	2 (6.5)
Ileum	2 (6.5)
Total	31(100)

Table III Post operative complications in 6 children.

S/N	**Procedure**	**Complications**	**Treatment**	**Outcome**
1	**Swenson**	Anastomotic desruption	Colostomy	Survived
2	**Swenson**	Anastomotic desruption	Colostomy	Died
3	**Swenson**	Enterocolitis	Antibiotics,	Survived
4	**Endorectal**	Burst abdomen	Laparotomy	Died
5	Endorectal	Anastomotic stenosis	Sphinterotomy	Survived
6	**Endorectal**	Recurrent constipation	Re-operation	Survived

Discussion

Children with Hirschsprung's disease usually present in the neonatal period in developed countries[1]. In a recent report from Australia, 90% of the cases of Hirschsprung's disease were diagnosed during the neonatal period while only 10% presented outside this period [2] . In Japan neonatal presentation was seen in between 40.1 and 53.4% during three different periods from 1978 – 2002 [6] . In Zaria, most children presented outside the neonatal period [4,5]. In the present study, 31(39.7%) children with Hirschsprung's disease presented during the neonatal period. It is not clear why most of our patients present outside the neonatal period, but a combination of many factors like lack of awareness of the disease, distance to the hospital and financial incapability could have contributed. In our experience some patients presenting to hospital early, are diagnosd late when they first present to the pediatricians who often are preoccupied with treating the neonates for sepsis or gastroenteritis. A high index of suspicion of Hirschsprung's disease by the attending physician would lead to early referral and appropriate investigations.The more common symptoms of Hirschsprung's we encountered in the neonatal period were; bilious vomiting, abdominal distension , delayed passage of meconium and decreased frequency of stools . Momoh JT in a report from Zaria found that 50% of neonates with Hirschsprung's disease had not passed meconium at presentation[6]. In this report, 87% had a history of delayed passage of meconium with more than half having delays of above 72 hours after birth. In Klein's report, 42% of neonates with Hirschsprung's disease had passed meconium within 24 hours and 58% after 48 hours[7] . Delayed passage of meconium especially within 48 hrs may not always mean Hirschsprung's disease, as more premature neonates are surviving today especially in developed countries. Most of our patients presented with intestinal obstruction, which is similar to reports elsewhere in Africa[5,6,8] . There was no intestinal perforation in any of our patients an uncommon presentation which has been reported from Zaria[5] . Three of our patients presented with enterocolitis but the impact of this in our care of these children is the fact that all of them have been mistaken for neonatal sepsis by a pediatrician thereby delaying surgical diagnosis . Delayed presentation is an important cause of increased morbidity and mortality in our environment .The diagnostic accuracy of Barium enema was 68% in this series. Others have reported 94% for barium enema [7] . We do not have the occasion to compare the diagnostic value of barium enema and rectal manometry as we do not have this facility though they are often quoted as having the same value. [9] Most of our patients have their transition zone at the sigmoid colon and this is , similar to most other reports [2,9]. In this study 6.5% had total colonic aganglionosis and the mortality was 100%.All the patients diagnosed during the neonatal period had definitive pull-through in the second stage after the neonatal period in this series. Some patients treated by one stage transanal pull-through as described by De laTorre-Mondragon[10] were managed outside this study period and are not included in this analysis, however a few neonates presenting without complications are suitable and will benefit from this procedure.In our environment as elsewhere in developing countries, few children with Hirschsprung's disease present during the neonatal period. A high index of suspicion especially by the first attending physician and early appropriate investigations are necessary to make early diagnosis and prevent morbidity and mortality.

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