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Hirschsprung's Disease: A Case Report In Pediatrics

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ABSTRACT

Hirschsprung's disease is a rare motor disorder of the gut which is characterized by the absence of ganglia in distal colon resulting in a functional obstruction. We report a case of a male baby of 15days of life presented with abdominal distension, vomiting, passage of stools in small amounts, greenish gastric aspirate. Physically facial puffiness, rough dry skin and cold peripheries were noticed. On abdominal examination, there were prominent veins, visible intestinal peristalsis and mild hepatospleenomegaly. Ultrasound abdomen shows dilated bowel loops suggesting intestinal obstruction. X-ray abdomen (ERECT) reveals cecal volvulus with intestinal obstruction. Histopathology report shows the impression – features consistent with Hirschsprung's disease. The treatment for Hirschsprung's disease is surgery.

Keywords: Hirschsprung's disease, intestinal obstruction.

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INTRODUCTION

Hirschsprung disease was described by Rusch in the 1691 and popularized by Hirschsprung in 1886, the pathophysiology was not clearly determined until the middle of the 20th century, when whitehouse and Kernohon reported aganglionic of the distal colon as the cause of obstruction in a case series[1]

In 1949, Swenson described the first consistent definitive procedure for Hirschsprung disease, rectosigmodectomy with coloanal anastomosis. Both myenteric and submucosal plexus are absent in patients with Hirschsprung disease. The cause of Hirschsprung disease is most commonly of neuroblasts originality from the neural crest in functional intestinal obstruction.[2,3]

The typical clinical signs of Hirschsprung disease are delayed meconium passage, abdominal distension, vomiting and enterocolitis. More than 80% of all Hirschsprung cases present symptoms in the neonatal period. Only a few of these are having a prenatal diagnosis (mostly performed by intrauterine MRI and ultrasound)[1]. Treatment requires surgical intervention.

We report a case of hirschsprung's disease in 15 days old male child.

CASE REPORT

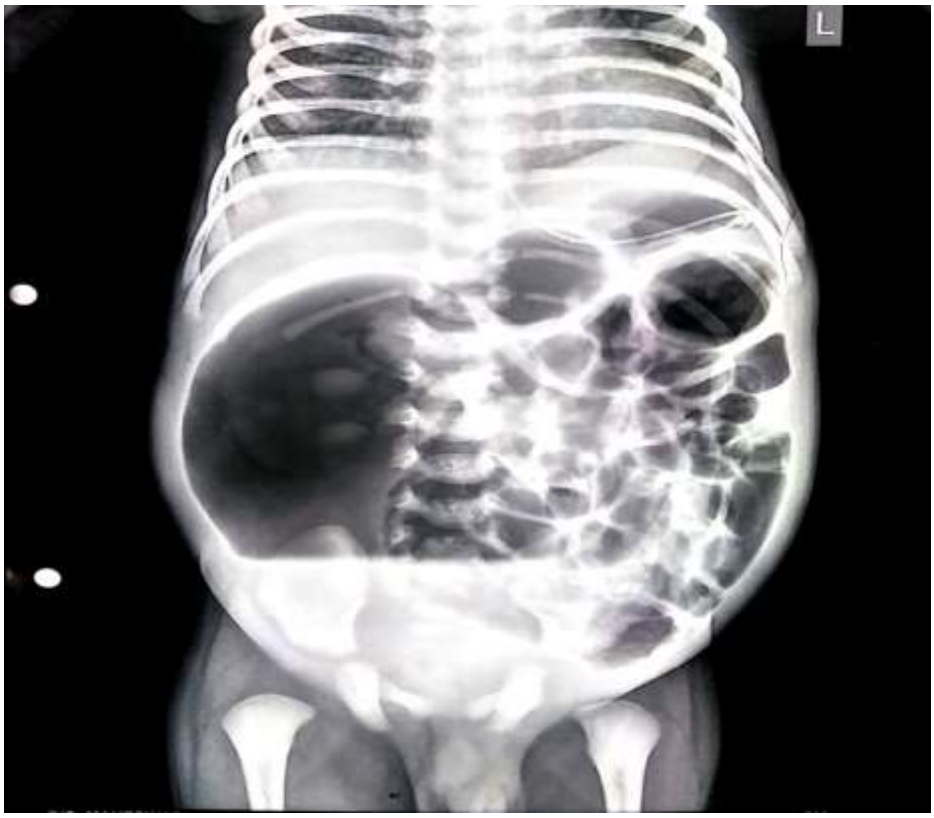
A 30-year-old, third gravida woman, gave birth to a boy at 40 weeks of gestation via normal vaginal delivery and had 3kgs weight. Her marital life was about 6yrs, unconsanguineous marriage in which she had 3 deliveries and 3 children were alive.

The baby with 15 days old was presented with abdominal distension, vomiting and passage of stool which is liquid in consistency in small amounts for the previous 13 days. On the day of admission he had yielded 15 ml of greenish gastric aspirate He had a history of excessive cry and feed was poor. There was no history of convulsion or any other neonatal illness. There was no history of any significant maternal illness during pregnancy or in the past. Physical examination revealed facial puffiness, rough dry skin and cold peripheries. There was no neck swelling or umbilical hernia. On abdominal examination, there were prominent veins, visible intestinal peristalsis and mild hepatosplenomegaly. The rest of the systemic examination was normal.

Complete blood count showed increase in haemoglobin content with 17.1g/dl (normal value - 11.5-16.5 g/dl) and all other counts are normal. CRP was elevated to 9 mg/dl (normal range 0-6 mg/dl). Random blood sugar was found to be normal. Serum electrolytes such as sodium, potassium and chlorides are found to be with in the range. Total bilirubin content was increased to 5.6mg/dl (normal – 0-1mg/dl), direct bilirubin increased to 5.2mg/dl (normal – 0.2-0.8mg/dl) and direct bilirubin was found to be normal. Blood urea and serum creatinine were found to be normal.

Ultrasound abdomen shows dilated bowel loops noted in the right iliac fossa with distended small bowel loops – likely intestinal obstruction and advised X-ray abdomen (ERECT). X-ray abdomen ERECT shows cecal volvulus with intestinal obstruction. Histopathology report (moderate biopsy) reveals features consistent with hirschsprung's disease.

X–ray abdomen (ERECT)



The treatment of this condition is by surgical management. However, initial medical management is needed in order to stabilise the patient before any surgical treatment is undertaken. Medical management includes the correction of any fluid or electrolyte imbalances, antibiotic therapy if enterocolitis is found and rectal decompression using rectal irrigations and tubes till the surgery is undertaken.

The basic aim for the definitive surgical treatment of this disease is the resection of any aganglionic segment, which is followed by a pull-through of any ganglionic bowel down to the anus.

Pre-operative treatment:

Drugs prescribed	Dosage	Frequency
Inj Meropenem	20mg/kg	BD
Inj Vancomycin	15mg/kg	BD
IVF Iso-P	10ml/hr	

Surgical therapy:

Pre-operative: Abdomen was found to be distended but the anus was patent. Operation: Mapping laparotomy and colostomy.

Post operative:

Wound developed to heal after a few days and the patient was returned to the operational theatre to repair the stoma and clean the bowel. Individual stitches were used to close the wound layers Blood group of the child was B +ve and was kept in reserve.

Post-operative treatment:

50 ml of fresh whole blood was transfused.

Managed nil by mouth and medications are as follows

Drugs prescribed	Dosage	Frequency
Inj Ceftriaxone	50mg/kg	BD
Inj Amikacin	20mg/kg	BD
Inj Metronidazole	15mg/kg	BD
IVF Iso-P	100ml	8th hourly

Follow up:

Following the operation, all his vitals and other parameters were observed to be in improving state and the wound started healing.

After a week the patient condition was significantly better and allowed oral feeding and continued antibiotic therapy.

DISCUSSION

Hirschsprung's disease is a congenital disorder that is characterised by the complete absence of ganglion cells in myenteric and submucosal plexus for a variable length of colon.[4] Ganglion cells act as points within the enteric nervous system to help coordinate and facilitate bowel relaxation. With their absence, the aganglionic areas of the bowel become spastic, thus causing distal intestinal obstruction. [5]

Hirschsprung's disease is a disease that should be diagnosed early in the new born period. Hirschsprung's disease should be considered in any newborn that fails to pass meconium with in 24-48 hours of birth. Untreated aganglionic megacolon in infancy may result in a mortality rate as high as 80%. Symptoms include bowel obstruction with bilious vomiting, abdominal distension, poor feeding and fail to thrive. About 10% of children present with diarrhea caused by enterocolitis, this may progress to colonic perforation causing life threatening sepsis.

Diagnosis of disease made from physical examination, abdominal X-ray, contrast enema, rectal biopsy, anorectal manometry. In all cases of hirschsprung's disease, surgery is the definitve treatment.[6]

CONCLUSION

Our case was presented with abdominal distension, vomiting and passage of stool which is liquid in consistency in small amounts. Neonatologists should be careful in evaluation in any infant who does not pass meconium with the first 24 hours of life after birth or who had abdominal distension with small amount of passage of stools. Pediatrician should be suspected in newborn intestinal obstruction, Hirschsprung's disease and consider appropriate treatment or surgical intervention without any delay. We the clinical pharmacists should play a role in assessing the condition of the neonates and counselling their parents about the condition, monitoring points and precautions to be taken after discharge from the hospital.

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