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CASE SERIES ON HIRSCHSPRUNG'S DISEASE

Dr. N. C. K. Reddy[1] M.S,M.Ch(AIIMS)
Paediatric surgeon,
NICE Hospital, Shanthi Nagar,
Hyderabad, Telangana
dr.nckreddy@gmail.com
Phone no. 9440057888

Corresponding author: **Dr. Geetha Shavali[2*]** MBBS, DRTD, MD
Associate professor, Dept. of Physiology,
Osmania Medical College
Hyderabad, Telangana
geethsmbbs@gmail.com
Phone no. 7989452276

ABSTRACT
Food in the intestine always moves from oral to the aboral end with proximal constriction and receptive relaxation, this is the law of the gut. One congenital condition violating this law is Hirschsprung's disease. The defect is failure of the caudal migration of neural crest cells, which are the precursors of ganglion cells in the Meissner's (submucosal) and Auerbach's (myenteric) plexuses resulting in loss of myenteric reflexes and causing megacolon in neonates. Surgical intervention is the only option for this condition. Such neonatal cases admitted at a paediatric tertiary care centre, with final diagnosis confirmed by histopathological examination have been presented below. The follow up examination up to two years of all the three neonates showed normal weight gain and normal developmental milestones.

KEYWORDS
Hirschsprung's disease; megacolon; law of gut; Meissner's plexus; Auerbach's plexus; abdominal perianal pull through

CASE 1
Case one was a single, male, appropriate for gestational age (AGA) baby born to a primi, diabetic mother. Lower segment caesarean section done at an obstetric centre, baby cried immediately after birth. No active resuscitation was required. APGAR score 8-10. On day of life (DOL)-2, baby had episodes of bilious vomiting and abdominal distension, shifted to neonatal intensive care unit (NICU), kept NPO (nil per oral) and was started on antibiotics. X-ray abdomen revealed grossly distended large bowel with minimal rectal gas. Baby was shifted to the pediatric tertiary care centre. Paediatric surgeon advised for Cologram which showed features of intestinal obstruction due to Hirschsprung's Disease. On DOL-7, baby was operated. An abdominal perianal pull through was performed after resection of 5cm of aganglionic descending colon segment. Resected tissue was sent for histopathological examination which showed proximal resected end with ganglionic cells in the submucosal and myenteric plexus, mid zone and distal resected ends showing no ganglion cells, suggestive of Hirschsprung's disease. Postoperatively, Leukocyte-reduced red blood cells and plasma

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protein fraction transfusions were given. Minimal feeds started on postoperative day-4, baby tolerated well and passed stools. Reached full feeds on day of life-12 and discharged. One year eight months past surgery baby is thriving well with normal bowel habits and stable vitals.

CASE 2

Case 2 was a singleton, male, AGA (appropriate to gestational age) baby born to a primi, Rh-negative mother by lower segment caesarean section. Baby cried immediately after birth; no active resuscitation required. On day of life-2, baby developed abdominal distension with history of non-passage of meconium since birth. Abdominal X-ray showed multiple air fluid levels. On day of life-3, baby passed small quantities of meconium but abdominal distension continued. From DOL-6 to DOL-16, soap water enemas were given on alternate days due to which baby passed minimal amounts of stools. On DOL-17, baby was brought to the emergency room of the paediatric tertiary care centre with complaints of bilious vomiting. On examination, baby was hemodynamically stable but with massive abdominal distension. There were signs of failure to thrive. Face was emaciated with dry lips. All four limbs looked disproportionate to the torso. Drastic decrease in weight from birth weight of 3kg to 2.2kg at admission. Baby's blood group was Rh positive. Baby was started on intravenous fluids. Cologram was performed which showed features of obstruction due to Hirschsprung's disease. Baby was operated on DOL-19. An abdominal perianal pull through was done. Postoperatively, axillary PICC (Peripherally inserted central catheter) line malfunctioned. A femoral line with ultrasound guidance was instituted. Packed RBC and fresh frozen plasma were administered. On postoperative day-6, minimal feeds were started. Enteral feeds with EBM and RCM (formulated food products) were administered. Reached full feeds on postoperative day 8. Remained euglycemic throughout the period of stay.

CASE 3

Case 3 was a late, preterm, AGA female with TTNB (transient tachypnoea of the new born) with intestinal perforation (sigmoid colon perforation).

Baby was admitted at three hours of life in NICU with history of respiratory distress since birth. At admission DOWNE's score was 2. Baby was started on HFNC (high flow nasal cannula) support with flow rate of 5L/min and FiO2 25%. Chest X ray was suggestive of TTNB. Respiratory distress improved with oxygen support and baby was weaned to room air on day 3 of life. Kept NPO, on I/V fluids for the first 48 hours. On day 3 of life, tube feeds were started but there was severe intolerance and recurrent vomiting. There was abdominal distension but the OG (orogastric) aspirate was clear. X-ray abdomen showed distended large bowel loops. A lower GI contrast was done which was suggestive of Hirschsprung's disease. OG aspirate turned bilious on day of life 5. Baby was taken up for open laparotomy on day 7 of life. Intraoperative findings revealed sigmoid colon perforation. Resection and reanastomosis was done with peritoneal wash. Postoperatively peristalsis improved. Feeds started on postoperative day 9, and reached full feeds by postoperative day 16. Baby observed for 10 days and discharged.

ABBREVIATIONS

AGA: appropriate for gestational age
DOL: Day of life
NICU: neonatal intensive care unit
NPO: nil per oral
PICC: peripherally inserted central catheter
EBM and RCM: formulated food products
TTNB: transient tachypnoea of the new born
OG: orogastric

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CONFLICT OF INTEREST Nil

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