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Case Report

Paediatrics Section

Congenital Absence of Ileum, Caecum and Appendix in a Preterm Neonate: A Rare Case Report

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ABSTRACT

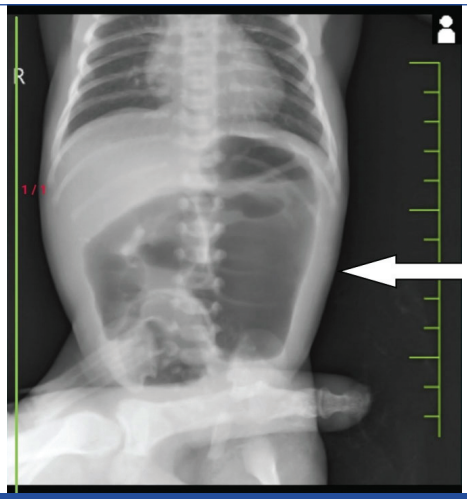
There are numerous birth defects that can affect the gastrointestinal tract, starting from the oesophagus or stomach to the small and large intestines. Intestinal atresias are one of the most common causes of intestinal obstruction in newborns. The most common location is the jejunum, followed by the duodenum and the colon. Congenital absence of the ileum, caecum, and appendix is an extremely rare condition. Here, an exceptional instance of congenital absence involving the distal jejunum, ileum, caecum, and appendix in a male child is presented, who presented with chief complaints of multiple episodes of bilious vomiting after initiating the first feed and non passage of meconium. Upon initial examination, the patient was suspected to have an intestinal obstruction and subsequently underwent an exploratory laparotomy. However, intraoperative findings revealed a grossly dilated proximal jejunal loop, and the next segment of the intestine was the narrow ascending colon. In due course, the baby recovered well and was discharged. Currently, there is no documented occurrence of a comparable case involving a viable neonate in the existing body of scholarly literature. Surgeons need to understand positional variations and congenital anomalies, as well as the procedures needed to identify these abnormalities during surgery.

Keywords: Ascending colon, Congenital atresia, Intestinal obstruction, Jejunum

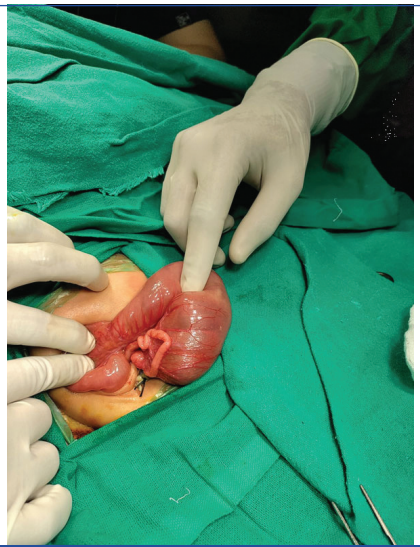
CASE REPORT

A male child, weighing 2058 grams, born to a primigravida mother at the gestational age of 35 weeks by vaginal delivery, was brought to the neonatology department at 12 hours of life with chief complaints of multiple episodes of bilious vomiting after initiating the first feed and non passage of meconium. The antenatal history was not significant. As the patient was from a rural background, no earlier ultrasound was done. On admission, the baby appeared lethargic, had a weak cry, and was vitally stable with a heart rate of 134 beats/min, a respiratory rate of 44 cycles/min, blood pressure of 64/40 mmHg, and SpO_2 of 95%. On systemic examination, the abdomen was shiny, grossly distended. Palpation revealed a soft, non tender abdomen with no organomegaly, and bowel sounds were absent on auscultation. The infant was kept nil by mouth, initiated on total parenteral nutrition, started on empirical antibiotics (Aampicillin 50 mg/kg 12 hourly and gentamicin 5 mg/kg/day for five days), and had a nasogastric tube inserted into the gastric cavity. The blood tests yielded normal results, while the X-ray of the erect abdomen [Table/Fig-1] indicated notable bowel dilatation accompanied by a scarcity of gas in the pelvic region.

The clinical presentation and X-ray revealed intestinal atresia, and the baby was taken for emergency exploratory laparotomy. Preanaesthetic evaluation revealed normal vitals and blood parameters. A right transverse supraumbilical incision was made, revealing a grossly dilated proximal Jejunal loop which terminated blindly 20 cm from the duodeno-Jejunal flexure. There was an absent distal jejunum and complete absence of the ileum, appendix, and caecum. A narrow ascending colon, transverse colon, and descending colon were noted [Table/Fig-2]. The patency of the colon was confirmed by milking saline from the ascending colon down to the rectum, which excluded distal intestinal obstruction. Excisional tapering of the dilated distal jejunum was done, followed by end-to-end anastomosis of the tapered jejunum with the ascending colon. The baby had a good postoperative recovery. On the seventh surgical day, the nasogastric tube was withdrawn, and nursing commenced. The patient exhibited no instances of bilious vomiting upon consuming breast milk and was discharged



[Table/Fig-1]: Significant dilatation of bowel with paucity of gas in the pelvis.



[Table/Fig-2]: Intraoperative picture showing grossly dilated jejunal loops with narrow colon.

on the 19th day following the surgical procedure. The parents were explained regarding the nutritional morbidity associated with this condition and were advised to have regular follow-up. However, the baby succumbed at home after one week.

DISCUSSION

Atresia is a prevalent aetiological factor contributing to neonatal intestinal obstruction. Current thinking suggests that a vascular injury within the uterus is to blame [1]. Four forms of intestinal atresia are now recognised in the medical community's categorisation scheme [2]. The reported incidence of intestinal atresia ranges from 1.3 to 3.5 per 10,000 live births, of which approximately 20% are associated with a chromosomal anomaly [3]. This particular instance represents a rare occurrence of intestinal atresia characterised by the complete absence of the ileum, caecum, and appendix. To the best of our understanding, this particular instance represents the initial occurrence in which the infant underwent a successful operation and subsequently received discharge while being nourished through breastfeeding.

Short bowel syndrome may be avoided with surgical techniques such as intestinal plication and proximal tapering enteroplasty. Bianchi's recommended longitudinal intestinal lengthening and tailoring operation, as well as Javid PJ et al., supported serial transverse enteroplasty treatment, are both surgical therapies intended for individuals with a minimum amount of functional small intestine [4]. In this particular instance, a tapering enteroplasty procedure was conducted on the distal jejunum, which was subsequently followed by an end-to-end jejuno-colic anastomosis. Functional obstruction at the site of anastomosis is a frequently observed postoperative complication in cases of intestinal atresia. Literature reports a few cases of congenital absence and abnormalities of the intestine [5-8].

Tripathy PK et al., reported a case of a 16-day-old female newborn who underwent exploratory laparotomy and was found to have her jejunum and ileum absent from birth [5]. Blind termination of the dilated duodenum was observed, and the following section of the intestine consisted of a microcolon and a peanut-sized caecum. The appendix and caecum were removed. To verify colonic patency, saline was pushed into the colon and milked in a microcolon all the way to the rectum. The duodenum was opened at the distal dependent portion, and a 5-0 polyglactin suture was used to perform a single-layer end-to-back anastomosis with the ascending colon. The baby's postoperative recovery went smoothly.

Salim A et al., reported the first-ever occurrence of congenital absence of the appendix in association with malrotation. A Ladd's procedure was performed in the standard fashion, with evisceration of bowel loops, division of Ladd's bands, broadening of the mesentery, and replacement of bowel loops within the abdominal cavity in a non rotated position [7]. Sham M and Singh D conducted a study on a neonate who presented with near-total jejunoileal atresia [8]. Despite the severity of the condition, the neonate managed to survive for a period of three months.

In 1884, Thomas WJTL documented a postmortem discovery concerning a premature infant born at seven months gestation, which revealed a total absence of the jejunum, ileum, and a significant portion of the colon [9]. There are only two documented cases of congenital absence of the small bowel. In both instances, the affected individuals exhibited intact duodenums and colons. Infants commonly exhibit symptoms such as bilious emesis, abdominal distention, and failure to pass meconium. The clinical manifestation exhibits variability depending on the specific site of the atretic obstruction. Proximal atresia is characterised by the occurrence of notable bilious emesis, whereas distal atresia is commonly associated with abdominal distention accompanied by the presence of multiple dilated bowel loops [5,10]. Louw JH and Barnard CN were the first to provide evidence supporting the hypothesis that vascular injury during pregnancy is the primary cause of jejunoileal atresia [11]. Intestinal atresia is classified according to research performed by Louw JH and Barnard CN in 1955. There are four types of intestinal atresia. Type 1 is characterised by a mucosal web or diaphragm. Type 2 involves an atretic cord located between two blind ends of bowel, with an intact mesentery. Complete mesenteric separation at the blind ends of the gut (type 3a) results in a V-shaped mesenteric gap. Finally, type 3b has a big mesenteric gap and looks like an apple peel or Christmas tree. Type 4 presents with the presence of multiple atresias, exhibiting a characteristic string-like morphology reminiscent of a sausage. Nevertheless, it is worth noting that complete jejunoileal atresia has not been included in any of the existing classification systems [11]. The present case cannot be classified as it is a case of complete absence of the ileum, caecum, and appendix. While newborns with total low obstruction should have a contrast material enema examination, those with full high obstruction often do not need further radiologic screening after radiography. Since each incidence of partial intestinal blockage requires a particular course of treatment, all patients with this condition must have an upper gastrointestinal series. Ultrasonography (US) can distinguish between colonic and small bowel blockage in cases of low intestinal obstruction. Furthermore, US is helpful in the accurate detection of intestinal duplication cysts as well as meconium ileus and meconium peritonitis [12].

CONCLUSION(S)

The management of long gap intestinal atresia continues to pose challenges. In the present case, the patient was suspected to have an intestinal obstruction upon initial examination and subsequently underwent an exploratory laparotomy. However, intraoperative findings revealed a grossly dilated proximal jejunal loop, and the next segment of the intestine was a narrow ascending colon. Surgeons need to understand positional variations and congenital anomalies, as well as the procedures needed to identify these abnormalities during surgery.

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