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Case Report

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# Congenital Cecal Atresia: An Uncommon Entity

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## Abstract

Though bowel atresia-associated intestinal obstruction (IO) is a frequently reported pediatric surgical emergency, IO resulting from cecal atresia (CA) has been seldom reported. Herein, we report a rare case of a 7-day-old female neonate, who presented with gross abdominal distention, bilious vomiting, and failure to pass meconium since birth. Physical examination revealed tense and distended abdomen, and abdominal X-ray disclosed dilated loops of the bowel, and absence of the bowel gas pattern in the lower abdomen, thereby suggesting small bowel obstruction. Emergency laparotomy revealed markedly dilated terminal ileum with atretic cecum and normal appearing appendix. The affected segments were excised and side-to-side ileo-ascending colon anastomosis was achieved. Histopathological examination of the excised section revealed Type I CA. Postoperatively, the patient recovered well, and examination at follow-up visits suggested satisfactory development with above 25th percentile on the growth chart.

**Keywords:** Cecal Atresia; Intestinal Obstruction; Pediatric Emergency

## Introduction

In the neonatal period, intestinal obstruction (IO) is a frequent emergency condition, requiring early diagnosis and subsequent surgical management [1]. Jejunum and ileum are the predominantly affected intestinal segments, while colon is seldom involved [2]. Due to its rarity, colonic atresia has an incidence of 1:20000 to 1:66000 live births [3]. In a retrospective analysis from India, only 5.7% neonates had colonic atresia [1]. In the colon, the commonly affected segments, in decreasing order, include sigmoid, splenic flexure, hepatic flexure, and ascending colon [4]. However, to the best of our knowledge, a case of isolated cecal atresia (CA) has not been reported yet from India. Herein, we report a rare case of CA in a newborn female.

## Case Report

A 7-day-old female twin neonate presented with gross abdominal distention, bilious vomiting, and failure to pass meconium since birth. She was born at 32 weeks' gestation by lower segment caesarian section to a multiparous 25-year-old woman and weighed 2.02 kg at birth. The prenatal ultrasound scan did not reveal any gross fetal anomaly, and there was no evidence of polyhydramnios. At presentation, the patient had tachycardia, while other vital parameters were normal. Physical examination revealed tense and distended abdomen. An X-ray of the abdomen disclosed dilated loops of the bowel, and absence of the bowel gas pattern in the lower abdomen (Figure 1). Based on these findings, ileal atresia was suspected.

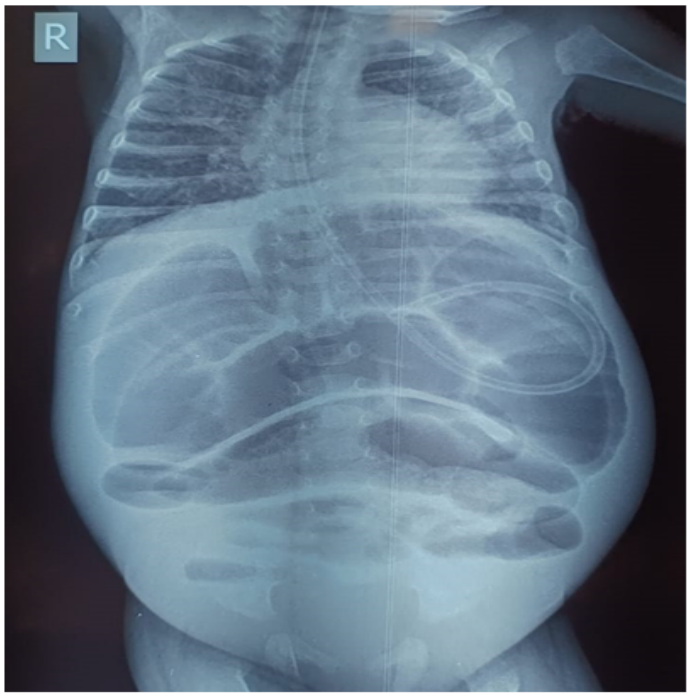


**Figure 1:** Abdominal X-ray illustrating dilated bowel loops, and absence of the bowel gas pattern in the lower abdomen

Laparotomy was performed via right infraumbilical transverse incision. On exploration of the bowel, the terminal ileum was found to be markedly dilated, and the cecum was atretic with normal appearing appendix (Figure 2). The grossly distended terminal ileum, atretic cecum, and appendix were excised with 1 cm margin (2×1.5×0.3 cm, Figure 3). The absence of any distal obstruction was confirmed by irrigating the lumen of ascending colon with normal saline, and side--to-side ileo-ascending colon anastomosis was achieved. The excised section was sent for histopathological examination which demonstrated cecal atresia.

On postoperative day (POD) 3, the patient passed the meconium, and breast feeding was re-initiated on POD 5, 2 ml per 2

hr initially and then gradually to full breast feed. The patient was discharged on POD 10 in stable condition. She was followed-up on POD 14, and at monthly intervals for 3 months, and examination showed satisfactory development with above 25th percentile on the growth chart.

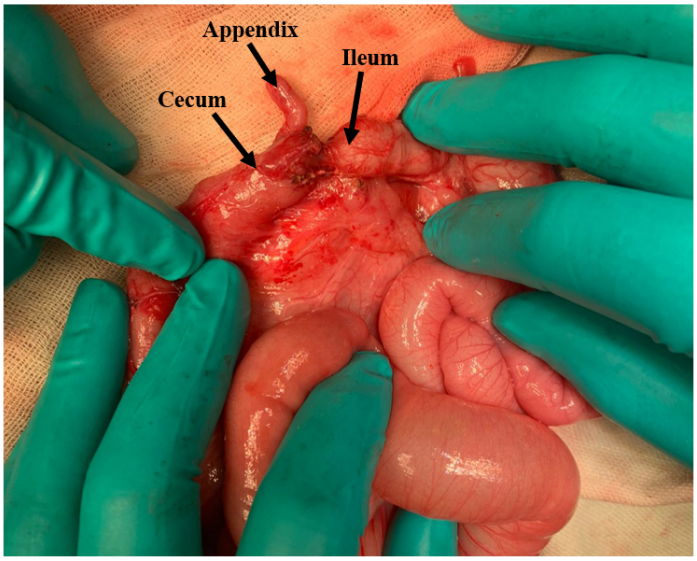


**Figure 2:** Intraoperative pictograph depicting markedly dilated terminal ileum, atretic cecum, and normal appearing appendix

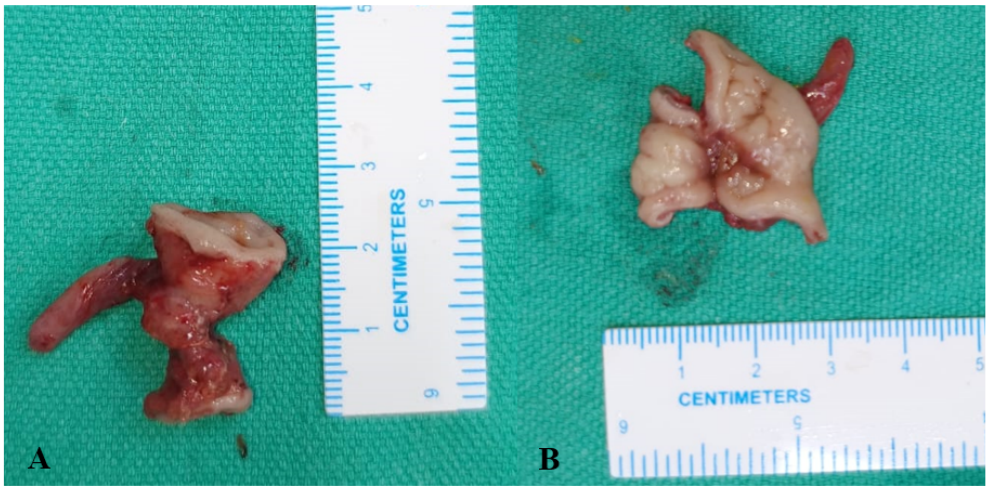


**Figure 3:** Excised bowel segment depicting terminal ileum, atretic cecum, and appendix were excised with 1 cm margin (2×1.5×0.3 cm)

## Discussion

IO can be classified as high and low IO. While, the former includes duodenal atresia, jejunal atresia, and malrotation, the latter includes ileal atresia, colonic atresia, Meckel's diverticulum with bands, and Hirschsprung's disease (HD) [1]. Colonic atresia, a rare cause of neonatal IO, accounts for 1.8 – 15% of all gastrointestinal atresia. In 1954, Robert A. De Bord documented the first patient with cecal atresia [5]. In this report, we document the first case of CA in our country to the best of our knowledge, and demonstrate satisfactory growth following side-to-side ileo-ascending colon anastomosis.

Though there is no sex predilection, intestinal atresia is associated with prematurity and low birth weight [6]. Additionally, twin pregnancy is a risk factor for intestinal atresia [7]. Likewise, our patients was delivered prematurely, had low birth weight, and born out of twin pregnancy, thus had a higher risk of developing CA. The patients with high IO, mainly present with vomiting, while those with low IO predominantly present with abdominal distension associated with delayed or non-passage of meconium [1]. Likewise, our patient presented with gross abdominal swelling, bilious vomiting, and inability to pass meconium.

The pathogenesis of colonic atresia is still controversial. Various disorders, including intestinal hernia, intestinal stricture, or intestinal obstruction, believed to act as direct triggers, rarely lead to colonic atresia. It is hypothesized that these disorders trigger a vascular event and induce atresia. Additionally, several congenital anomalies, including gastroschisis, choledochal cyst, imperforate anus, omphalocele, and vesico-intestinal fistula, are reported to be associated with colonic atresia [3].

HD, a developmental defect, occurs simultaneously in a minimum of 2% population with an incidence of 1:5000 live births. It results in functional obstruction due to lack of ganglionic cells in the distal colon. As per a school of thought, in each patient with colonic atresia, HD should be ruled-out prior to surgical intestinal anastomosis.(1) A newborn with absence of meconium passage within 48 hours of birth coupled with abdominal distention, feeding difficulty, emesis (bilious or non-bilious), explosive diarrhea, and seldom pneumoperitoneum or signs of peritonitis raises a suspicion of HD. On contrast enema, HD illustrates characteristic finding of the rectosigmoid ratio reversal (<1), presence of a transition zone, and a spiculated or sawtooth appearance of the rectal mucosa [8]. Neonates with HD can be managed with elective surgery; however, those with CA require emergency laparotomy. In our patient, HD was not ruled-out prior to primary anastomosis; however, the biopsy of the resected specimen demonstrated presence of ganglionic cells. Additionally, IO resulting from midgut malrotation in neonates is a common condition with an incidence of 1:500 live births and the symptomatic incidence of 1:6000 live births. Thus, malrotation should be suspected in a neonate presenting with sudden onset of bilious vomiting [1]. While, Meckel's diverticulum, the commonest gastrointestinal congenital malformation, occurs in 2 – 4% live births, and the neonates characteristically present with bowel obstruction and intestinal bleeding [9]. In our patient, both malrotation and Meckel's diverticulum were ruled out intraoperatively.

Delay in symptom recognition leads to higher chances of complications, including perforation and sepsis [1]. A routine abdominal X-ray demonstrates dilated bowel loops, a contrast enema shows microcolon [10]. Abdominal ultrasonography and magnetic resonance imaging have good accuracy in the diagnosis for congenital bowel anomalies [11,12]. In our patient, the abdominal X-ray showed the dilated bowel loops, and absence of the bowel gas pattern in the lower abdomen, thus supporting the diagnosis of IO. However, the diagnosis of CA was confirmed intraoperatively.

In 1979, Grosfeld et al. proposed an intestinal atresia classification, and divided them into five main types: Type I (mucosal atresia), Type II (atretic ends of the intestine attached by a fibrous tissue band), Type IIIa (atretic ends disconnected by a V-shaped mesenteric defect), Type IIIb (apple skin deformity with proximal colonic atresia and a single retrograde blood supply to the distal end), and Type IV (multiple atresias).(13) Of these types, Type IIIa is frequently observed [3]. However, our patient had Type I CA with mucosal involvement.

The choice of surgery is primarily dictated by the intraoperative findings [10]. In our patients, intraoperative exploration revealed markedly dilated terminal ileum with CA and normal appendix. These parts were excised, and side-to-side ileo-ascending colon anastomosis was achieved. Several surgeons have used this approach, and have reported satisfactory results. As a practice, the occurrence of postoperative ileus can be prevented by dissecting a part of the colon proximal to the anastomosed bowel [10]. Likewise, we excised the affected bowel segments with 1 cm margin.

Intestinal and colonic atresia are associated with good long-term outcomes [14,15]. However, the presence of associated anomalies, described above, increases the risk further [3]. In

this report, we evaluated the short-term outcome in our patient, and the growth was found to be satisfactory.

## Conclusion

To conclude, CA is an extremely rare anomaly with very few cases reported in the literature. The affected neonate frequently presents signs and symptoms of IO. Clinical examination and abdominal X-ray help in reaching the diagnosis. However, intraoperative bowel exploration leads to definitive diagnosis. Excision of the atretic colon segment together with 1 cm healthy segment margin followed by ileocolic anastomosis leads to satisfactory outcome and prevents recurrence.

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