# **Case Report**

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# Type IV jejunal atresia in a newborn: a rare birth defect presenting with bilious vomiting

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### **ABSTRACT**

**Background:** Type IV jejunal atresia is a rare birth defect, which occurs due to late intrauterine vascular accidents that results in complete obstruction of intestinal lumen. There is no genetic predilection. It occurs most commonly in proximal jejunum. Clinical presentation includes bilious vomiting, abdominal distension, feeding difficulties, failure to pass stools and/or absence of bowel movements after birth. Here author report a case of premature newborn, who was admitted in NICU at birth in view of prematurity. As the baby had bilious vomiting and bilious NG aspirate on day 3 of life, possibility of intestinal obstruction was kept. It was diagnosed as a case of jejunal atresia by abdominal radiograph displaying dilated stomach, duodenum and proximal jejunum with gasless abdomen. Further jejunal atresia confirmed by the upper GI study exhibiting the level of obstruction. Surgical resection of the atretic segment and primary anastomosis was done. This case emphasizes the need of rapid identification of jejunal atresia and its adequate management to prevent morbidity and mortality associated with fore shortened intestine.

**Keywords:** Abdominal distension, Abdominal radiograph, Bilious vomiting, Intrauterine vascular accidents, Jejunal atresia, Upper GI study

## INTRODUCTION

Jejunal atresia is a rare congenital anomaly, that occurs in only 1 in 12,000 live births. This unusual condition is characterized by complete occlusion of the intestinal lumen. It is one of the major causes of intestinal obstruction. The likely cause of jejunal atresia is late intrauterine mesenteric vascular accidents that causes necrosis and resorption of intestine. Though other factors like in utero intussusception, intestinal perforation, segmental volvulus and thromboembolism have also been shown to cause jejunal atresia. About 10% of newborns with jejunal atresia have cystic fibrosis and meconium ileus. Jejunal atresia generally occur as a single entity (>90%); however multiple atresias occur in 6-20% of patients, mostly in the proximal jejunum.

Newborns with jejunal atresia generally present with bilious vomiting in first 24 to 48 hours of life; however, it may present with abdominal distension, feeding difficulties and non-passage of stool after birth in some cases.

Multiple jejunal atresias are often associated with shorter bowel length, prematurity and high mortality rate; hence necessitating urgent work up and prompt surgical intervention to prevent related complications and undue prolonged NICU hospitalization.

Herein authors report a rare case of multiple jejunal atresia diagnosed on clinical and radiological grounds, followed by surgical intervention and meticulous postoperative care.

#### **CASE REPORT**

A preterm (34 weeks) AGA (1.65kg) male baby was born to primigravida mother by vaginal delivery. Baby cried immediately after birth. Antenatal history was uneventful. Mother had irregular antenatal visits and no antenatal ultrasound was done. On admission, baby was stable, pink and had mild respiratory distress with no abdominal distension. Septic screen was positive.



Figure 1: Preoperative picture of new-born baby.

Baby was kept nil per oral and started on intravenous fluids, intravenous antibiotics and nasal CPAP. On day 2 baby started having yellow color nasogastric (NG)aspirate, which increased in amount and changed into green color in next 24 hours. Baby also had intermittent bilious vomiting despite of continuous NG aspiration. Abdominal radiograph revealed triple bubble sign characterized by gaseous distension of stomach, duodenum and jejunum with gasless abdomen (Figure 2).



Figure 2: Abdominal radiograph showing triple bubble sign with gasless abdomen.

A contrast enhanced upper GI study was done to confirm the level of intestinal obstruction and to exclude midgut volvulus. Upper GI study showed dilated stomach and duodenum with enlarged upper jejunum and lack of passage of contrast agent to distal small intestine (Figure 3).



Figure 3: Upper GI study depicting extent of intestinal obstruction.

Baby was kept NPO and surgery was performed next day. A transverse incision was given at supraumbilical region. The entire intestine was delivered out through the incision to assess the extent of atresia. Proximal jejunum was dilated and there were multiple jejunal atresias extending over approximately 15cm of jejunum. Irrigation of distal bowel with normal saline confirmed the patency of remaining small and large intestine. Resection of affected segment was done followed by primary anastomosis of two ends. Jejunostomy tube was kept in situ.

Postoperatively nothing per oral, nasogastric suction. maintenance of fluid and electrolyte balance was continued. Baby was kept on ventilatory support for one day in NICU. Nasogastric output was monitored and replaced volume for volume. Jejunostomy feeds were started after signs of propulsive peristalsis occurred i.e., clear, low volume nasogastric output, soft flat abdomen and passage of stool. Further it was confirmed by contrast enhanced upper GI study which showed passage of contrast in to the distal bowel. Jejunostomy feeds were started on day 3 postoperatively in small amount and gradually build up to full feeds over next 2 weeks. Baby was discharged home on spoon feeds after 2 weeks.

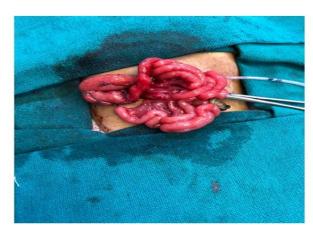


Figure 4: Multiple jejunal atresias on laparotomy.

#### **DISCUSSION**

Jejunal atresia is one of the major causes of intestinal obstruction in newborn, which occurs due to late intrauterine vascular insults. It is characterized by complete absence of mesentery, which causes jejunum to twist around an artery that supplies blood to intestine. Eventually leading to necrosis and resorption of intestine. Favara et al also suggested antenatal vascular accident as causative factor for intestinal atresia.5 There is no male to female variation. Till the year 2007, there were approximately 57 cases of jejunal atresia reported in medical literature. 6 In many cases, Jejunal Atresia is seen to arise sporadically. Though in some cases, it is inherited, and therefore, a positive family history of the condition is the biggest risk factor for a child being diagnosed with the condition. Maria et al reported a case of multiple intestinal atresia in two consecutive siblings in 2018.8

There are four types of atresia. Type I atresia constitute luminal webs or membranes with mural continuity. Type II atresia is characterized by blind ends joined by fibrous cord. Type IIIa atresia has two disconnected ends without a fibrous cord and a mesenteric gap. Type IIIb atresia has two separated ends along with large mesenteric defect. Type IV atresia is characterized by multiple segments of atresia. Our patient had a Type IV jejunal atresia comprising of multiple atretic segments of jejunum.

Jejunal atresia is often diagnosed antenatally by prenatal ultrasound which shows dilated bowel proximal to blockage and polyhydramnios. Though in present case, there were no records of antenatal ultrasound and there was no history of polyhydramnios. After birth, proximal jejunal atresia present with bilious vomiting, abdominal distension and absent bowel sounds, while distal atresia present with jaundice and delayed clinical onset of signs. In present case, baby had bilious vomiting along with sluggish bowel sounds suggestive of proximal jejunal atresia.

Jejunal atresia is confirmed by abdominal radiograph characterized by triple bubble sign and contrast enhanced upper GI study displaying the extent of atresia. <sup>10</sup> Timely surgical intervention is the main stay of therapy. Excision of atretic segment and primary anastomosis is done followed by comprehensive postoperative care. <sup>11</sup>

## CONCLUSION

Jejunal atresia is one of the major causes of intestinal obstruction. Proximal jejunum is more commonly affected than distal jejunum. Predominantly present with bilious vomiting. Prompt work up, early diagnosis, definitive surgical management and comprehensive postoperative care reduce the chances of mortality and morbidity.

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#### REFERENCES

- 1. Louw JH, Barnard CN. Congenital intestinal atresia: observations on its origin. Lancet 1955;269:490-1.
- 2. Rasquin A, Di Lorenzo C, Forbes D, Guiraldes E, Hyams JS, Staiano A, et al Childhood functional gastrointestinal disorders: child/ adolescent. Gastroenterol. 2006;130(5):1527-37.
- 3. Roberts HE, Cragan JD, Cono J, Khoury MJ, Weatherly MR, Moore CA. Increased frequency of cystic fibrosis among infants with jejunoileal atresia. Am J Med Genet. 1998;78(5):446-9.
- 4. Puri P, Fujimoto T. New observations on the pathogenesis of multiple intestinal atresias. J Pediatr Surg.1988;23(3):221-5.
- 5. Favara BE, Franciosi RA, Akers DR. Enteric duplications: thirty-seven cases: A vascular theory of pathogenesis. Am J Dis Children. 1971;122(6):501-6.
- 6. Jejunal Atresia. NORD.2007. Available at: http://rare diseases.org/ rare.diseases/ jejunal atresia.
- 7. Mishalany HG, Najjar FB. Familial jejunal atresia: three cases in one family. J Pediatr. 1968;73(5):753-5.
- 8. De Nardo MC, Douch C, Yee L, Patel D. Consecutive siblings, each with two separate small bowel atresias. BMJ Case Rep. 2018;2018.
- 9. Martin LW, Zerella JT. Jejunoileal atresia: a proposed classification. J Pediatr Surg. 1976;11(3):399-403.
- 10. Mandell J. Core radiology. Cambridge University Press; 2013.
- 11. Shahjahan M, Ferdous KM, Mitul MA, Islam MK. Management of jejunoileal atresia: our 5-year experience. Chattagram Maa-O-Shishu Hospital Med Coll J. 2013;12(3):52-5.

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