Published in final edited form as:

J Pediatr Surg. 2019 June ; 54(6): 1257–1260. doi:10.1016/j.jpedsurg.2019.02.003.

A New Variant of Type III Jejunoileal Atresia

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Abstract

Jejunoileal atresia (JIA) is a congenital defect that can result in significant loss of bowel length. The traditional classification of JIA was first proposed by Grosfeld and includes 4 subtypes. Among these, type IIIB, or apple-peel atresia, is characterized by a proximal atretic jejunum and a distal segment of spiraled bowel that terminates at the cecum. Due to this anatomy, patients with type IIIB JIA are at increased risk for short bowel syndrome and intestinal failure. In this report, we described the case of a neonate with a prenatal diagnosis of JIA. At exploration, she was initially found to have a type IIIB atresia. However, instead of terminating at the cecum, the distal spiraled segment was followed by 75 cm of normal small bowel and mesentery. Surgical correction proceeded with minimal resection and primary anastomosis. She recovered well from this procedure, tolerated full enteral nutrition by mouth, and displayed good weight gain at outpatient follow-up. Due to the unique anatomy of the gastrointestinal tract in this case report, we propose the addition of a new class of JIA, type IIIC, to better reflect its prognostication and surgical management.

Keywords

jejunoileal atresia; apple-peel atresia; prenatal imaging; parenteral nutrition; direct hyperbilirubinemia

Introduction:

Jejunoileal atresia (JIA) occurs with an incidence of 1–3 in 10,000 live births [1]. Studies in canine models have suggested that JIA results from intrauterine vascular accidents, which may explain its association with other ischemic congenital anomalies, such as volvulus and gastroschisis [2]. JIA was first comprehensively discussed by Sutton and Spriggs [3,4]. This was followed by a proposed classification system by Louw in 1967 that included type I-III atresia [5]. Martin and Zerella subsequently introduced the variant of multiple and apple-peel atresia [6], which were combined with the Louw categories by Grosfeld in 1979 [7] and

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Conflict of Interest:

The authors disclose no financial conflict of interest in relation to the work presented in this manuscript.

have since remained the standard classification system for JIA. Type I atresia is characterized by a transluminal septum; type II involves a fibrous cord connecting 2 blind-ending pouches; type IIIa has a V-shaped mesenteric defect; and type IV exhibits multiple atretic segments. A special class of intestinal atresia, type IIIB, which occurs in less than 10% of all JIA [8], consists of proximal jejunal atresia and a short segment of ileum spiraling around a single mesenteric vessel. In this paper, we report a unique variant of type IIIB JIA where a significant amount of normal ileum continued from the “apple peel/Christmas tree” segment up to the ileocecal valve. To our knowledge, this is the first report of this anatomic variant in the literature.

Case Report:

The patient was a newborn 34-week baby girl who carried a prenatal diagnosis of JIA. The mother was a 30-year-old G1P0 woman, with no significant past medical history or family history of JIA. Her initial structural survey at week 18 of pregnancy showed no abnormality except for a low-lying placenta. On follow-up ultrasound (US) at week 28, there was the bowel was notably dilated. She was subsequently referred to our Advanced Fetal Care Center for consultation, presenting at 31 weeks of gestation. There, she underwent repeat US and magnetic resonance imaging (MRI), which confirmed proximal jejunal obstruction and mild polyhydramnios (amniotic fluid index = 25) (Figure 1).

The mother presented at 33 weeks of gestation in active preterm labor with rupture of membranes. She subsequently received betamethasone, penicillin, and azithromycin prophylaxis. The baby was delivered vaginally, and a large amount of amniotic fluid was noted. The infant emerged vigorously with an Apgar score of 8 and 9 at 1 and 5 minutes, respectively. Her birth weight was 2210 g. Physical exam at birth revealed a soft and non-distended abdomen. A nasogastric tube (NGT) was placed for gastric decompression. Postnatal abdominal radiograph was consistent with jejunal atresia (Figure 1). An echocardiogram on day of life (DOL) 1 and showed no structural abnormalities except for an expected patent foramen ovale and small patent ductus arteriosus. A peripherally inserted central catheter (PICC) was placed on DOL 1 for anticipation of prolonged parenteral nutrition (PN) dependence. She required continuous positive airway pressure soon after birth for increased work of breathing but was quickly transitioned to room air by DOL 1.

On DOL 2, she was taken to the operating room for surgical repair of her presumed jejunal atresia. Upon exploration, the bowel was normally rotated. A classic proximal jejunal atresia was encountered a few centimeters distal to the ligament of Trietz and was accompanied by a dilated jejunal bulb (Figure 2A). The distal bowel appeared spiraled around a single vessel and was consistent with a type IIIB or apple-peel JIA and. This portion of the bowel measured 55 cm. Instead of the spiraled bowel terminating at the ileocecal valve, there was an additional 75 cm of distal small bowel with a normal mesentery (Figure 2B). The remainder of the gastrointestinal tract, including the ileocecal valve and the entire colon, was anatomically normal. The proximal 5 cm of the atretic segment was resected due to tenuous vascular perfusion. The bowel lumen was both visually inspected and flushed with saline to confirm the absence of distal obstruction. Bowel continuity was established with an end-to-end primary anastomosis employing the Cheatle technique for size mismatch. In total, the

patient had more than 130 cm of small bowel remaining, and the initially planned gastrostomy tube placement was deemed unnecessary. Pathological examination of the resected small bowel segment showed no obstruction or any other abnormality.

The patient was kept *nil per os* after surgery and required PN for caloric support. During this time, her direct bilirubin (DB) gradually increased and peaked at 3.0 mg/dL, presumably from PN-induced cholestasis. An abdominal ultrasound was performed and showed gallbladder sludge but a normal biliary tree. She started having bowel movements on post-operative day (POD) 12, and breast milk was subsequently initiated via a nasal feeding tube. Her feeds were gradually advanced, and she achieved full enteral intake on POD 20. She was transitioned to full oral intake on POD 24 and subsequently discharged on POD 27. At discharge, her weight was 2490 g and DB was 2.3 mg/dL.

She continued to be followed in the outpatient setting for monitoring of both DB and weight gain. At one-month follow-up after discharge, her DB had normalized. She had gained 1100 g since discharge and continued to tolerate breastfeeding with good appetite. She was scheduled to return for the next follow-up visit at one year of age.

Discussion:

JIA is being increasingly diagnosed by US in the prenatal period [9], although the detection rate is highly variable [10]. Despite being poor diagnostic criteria by themselves, the presence of polyhydramnios and small bowel dilation in the third trimester aid in the diagnosis of JIA [11]. As demonstrated in this case report, JIA is accompanied with a high incidence of premature delivery [12], and frequent prenatal evaluation is warranted. At specialized fetal centers, the addition of MRI imaging has provided important adjunctive information for the prenatal diagnosis of gastrointestinal abnormalities [13]. The use of multiple imaging modalities therefore allows for more informative prenatal counseling and the establishment of postnatal treatment strategy.

Patients with jejunoileal atresia usually have prolonged ileus after surgical correction. Full enteral intake is not typically established until 3 weeks after surgery [8,14,15]. This prolonged dependence on PN can put them at risk for IFALD [16,17], a prevalent and severe complication of PN dependence, which manifests as hepatic steatosis, cholestasis, fibrosis, and even progression to cirrhosis and end-stage liver disease [18]. In addition to the diagnosis of JIA, the patient in this case report was also born prematurely, another well-known risk factor for IFALD [19]. Despite demonstrating biochemical evidence of PN-induced cholestasis, this patient was expected to wean from PN within a reasonable amount of time. For that reason, no additional treatment was necessary.

Over the years, operative mortality for JIA has significantly decreased [20]. However, associated comorbidities, especially short bowel syndrome (SBS), continue to pose clinical challenges [8]. Although there is no clear difference in mortality among the classes of jejunoileal atresia, type IIIB and IV are more commonly associated with SBS, which can occur in up to 1/3 of these cases [8,21]. The lack of bowel length in type IIIB atresia sometimes necessitates creative bowel-preserving procedures in order to minimize the risk

of developing SBS [22]. In this unusual case of apple-peel atresia, the abundance of bowel length negated the need for such strategies. Given that the patient had an additional 75 cm of healthy distal ileum, there was a discussion in the operating room as to how the apple peel deformity should be handled – preservation and anastomosis to the apple peel deformity versus resection and anastomosis to distal small bowel with normal mesentery. We elected a conservative surgical approach and only resected the most proximal 5 cm of the apple peel deformity. Given that long-term outcome in patients with type IIIB atresia proves satisfactory even in the face of SBS [23], we expect excellent prognosis in this patient, who possesses a nearly full complement of bowel.

This case report presented a unique variant of JIA, where the gastrointestinal tract anatomy did not fit with any of the Grosfeld classes. Therefore, we propose the addition of a new class – type IIIC – to describe this variant of JIA (Figure 3). The establishment of an updated classification system can facilitate better report of these cases in the literature, enhance our understanding of the disease, and improve its management and prognostication.

Acknowledgements:

The authors acknowledge Mrs. Kristin Johnson (Vascular Biology Program, Boston Children's Hospital) for her work in preparation of the figures. Research funding for this study was provided by the National Institutes of Health Grant 5T32HL007734 (DTD).

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Figure 1:
A coronal steady-state coherent magnetic resonance image (A) shows the prominent fetal stomach, dilated duodenal sweep, and short segment of dilated jejunum (curved arrow). The appearance is remarkably similar to the abdominal radiograph (B) obtained shortly after delivery.

Figure 2:
Surgical exploration demonstrates a dilated jejunal bulb (J), apple-peel atretic segment (AS), and normal ileum (I).

Figure 3:
Proposal of a new class of jejunoileal atresia – type IIIC.