CLINICAL VIGNETTE

A Case of Jejunal Atresia

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Case

A 35-week 3-day preterm infant was admitted to the NICU for management of prenatally diagnosed jejunal atresia and respiratory distress. The finding of jejunal atresia and polyhydramnios was initially diagnosed on a 30-week anatomy scan (Figure 1).

The patient's mother was admitted with premature rupture of membranes and underwent induction with oxytocin. The patient then developed prolonged category II fetal heart rate late decelerations at which time oxytocin was turned off, and the NICU team was called for a crash C-section. Following C-section, the patient was found to have a delayed cry and neonatal resuscitation was performed. An enteric tube was placed, which drained 50 mls of clear fluid. Patient was also initially placed on CPAP for poor respiratory effort. APGARs were 6 and 8 at 1 and 5 minutes. Following admission into the NICU, the patient was transitioned to high flow nasal cannula, an enteric tube was placed, and surgery was alerted to patient's arrival. Initial abdominal x-ray demonstrated dilated proximal bowel (Figure 2, Figure 3). Prior to surgery, patient underwent an echo which did not demonstrate any structural congenital cardiac anomalies. Patient was continued on low intermittent suction and went to surgery on the second day of life.

During surgery, the proximal jejunum was found to be dilated with abrupt caliber change and type 1 atresia approximately 6 cm distal to the ligament of Treitz with a large transition zone. A second area of atresia, 2 cm distal to the first area, was also identified. Both segments were resected and sent to pathology. Following resection, the diameter of the two resected bowel ends had greater than 3:1 difference and a tapering enteroplasty was performed with an end-to-end jejunojejunostomy.

Following surgery, the patient returned to the NICU intubated, in stable condition. The patient's preoperative respiratory distress, as well as preference to avoid noninvasive positive pressure ventilation, the patient was kept intubated post-operatively until arrival to the NICU. Shortly after extubatation in the NICU, he developed frequent apneic/bradycardia/desaturation events. He was loaded with caffeine and placed on nasal cannula - intermittent mandatory ventilation with FiO2 40% with improvements in oxygen saturation. Patient was kept NPO with an enteric tube to low intermittent suction. On postop day 5, patient was weaned to room air, and on postop day 6 patient began having green smears per rectum, but no definitive bowel movements. On postop day 8, repeat abdominal xray showed distal bowel gas past the resected point and the enteric tube was discontinued, and trickle feeds started.

Following trickle feeds, the patient had three episodes of emesis and was made NPO again. Feeds were restarted on postop day 10, and a glycerin suppository was given, which resulted in stool output. On postop day 12, he again had an episode of bilious emesis with dilated loops on KUB. Feeds were discontinued, and an enteric tube was placed back to suction. On postop day 16, patient was again started on feeds and tolerated it well. At postop day 22, patient was at goal feeds and was discharged in good health.

Discussion

Jejunal and/or ileal atresia occurs in about 1 in 3,000-5,000 live births and over 30% of affected neonates are born prematurely.[1] There is typically no sex predominance and is rarely associated with chromosomal abnormalities (<1%).[2] Overall, jejunal atresias are more common than ileal atresias and account for roughly 20% of all small bowel atresias. The more distal the atresia, the less frequent the incidence.[3] Unlike duodenal atresias, which are thought to arise from a failure of canalization, jejunal atresias are secondary to an in utero vascular insult leading to ischemic necrosis and resorption of the affected bowel.[4] There have been several different mechanisms reported, which cause decreased fetal gut blood supply, and thus predispose patients to jejunal atresias. These include cystic fibrosis, gastroschisis, midgut volvulus, maternal vasoconstrictive medications, maternal cigarette smoking and cocaine use, and genetic factors.[5-8]

Jejunoileal atresias are classified surgically into four main groups, using the Louw's classification.[9-10] Type I is membranous with serosa continuity and has no mesenteric defect. Type II has serosal discontinuity with a fibrous cord between the proximal and distal ends. Type III has been further subdivided using the Grosfeld criteria with IIIa having mesenteric discontinuity and IIIb known as the "apple peel atresia", present in about 7% of jejunoileal atresias.[10] Apple peel atresia is the result of a large in utero vascular insult with the superior mesenteric artery absent beyond the origin of the middle colic artery. This leads to atresia of a large segment of small bowel and its associated mesentery. This deformity causes a short dilated segment proximally with a larger

decompressed segment distally. The collapsed segment spirals around the distal branches of the ileocolic artery giving the appearance of an apple peel. Type IV has multiple atresias and sometimes is termed "sausages on a string" due to its appearance. Type IIIb and IV atresias are more commonly associated with a significant loss of intestinal length, and thus have a poorer prognosis. Overall, type III atresias are the most common, and type I is the least common form.

Only 50% of intestinal atresias are detected by prenatal ultrasound,[11] and the detection rates decrease for more distal lesions. One study found that prenatal ultrasound detected only 52% of duodenal obstructions, 40% of jejunal or ileal obstructtions, and 29% of colonic obstructions.[12] Additionally, ultrasound is limited in detecting the number of obstructions or location as the bowel distal to the obstruction is generally decompressed. Ultrasound is also poor in assessing the viability of the unobstructed distal bowel.[13]

Typical findings on prenatal ultrasound suggesting jejunal atresia are multiple dilated bowel loops (>17 mm in diameter), ascites, echogenic bowel, and polyhydramnios.[2] Of these findings, dilated bowel loops and polyhydramnios after 32 weeks of gestation show the highest sensitivity in detecting jejunal atresia prenatally. However, these findings are nonspecific and can be seen in a variety of anomalies such as meconium ileus, colonic atresia, Hirschsprung's disease, and imperforate anus.[13]

Postnatal presenting symptoms in patients with jejunal atresia are bilious vomiting, abdominal distension and delayed passage of meconium. This presentation can be very similar to that of malrotation and midgut volvulus; therefore, these conditions must be quickly excluded.[2]

Standard imaging for diagnosing jejunal atresia is the abdominal radiograph, which will often show two or three dilated bowel loops in the upper abdomen. This characteristic radiographic feature is called the "triple bubble sign", which corresponds to dilatation of the stomach, duodenum, and proximal jejunum.[14] There should also be absence of air within the lower portion of the abdomen in jejunal atresias. Generally, patients require no further imaging for diagnosis. However, if there are concerns for malrotation, an upper GI can be performed.[2] Contrast enemas are also used both to differentiate jejunal atresia from other pathologies as well as attempt to exclude additional sites of obstruction.[14]

Once the diagnosis of jejunal atresia has been made the initial treatment is nasogastric decompression and fluid resuscitation followed by surgical resection. The surgical approach is variable and depends on the classification of the atresia. Following surgery, short term total parenteral nutrition is required in the majority of cases while normal intestinal function resumes.[2]

The prognosis for jejunal atresia is related to the total length of the remaining intestine and the presence of an intact ileocecal valve. Infants with >100 cm of remaining intestine are likely to require only short term TPN, whereas infants with <25 cm of remaining intestine will require long term TPN as well as intestinal lengthening procedures or even small bowel transplantation.[2] The two commonly used lengthening procedures are the serial transverse enteroplasty procedure (STEP) and longitudinal intestinal lengthening procedure (LILT). Overall survival with jejunal atresias is >90%.[2]

Figures

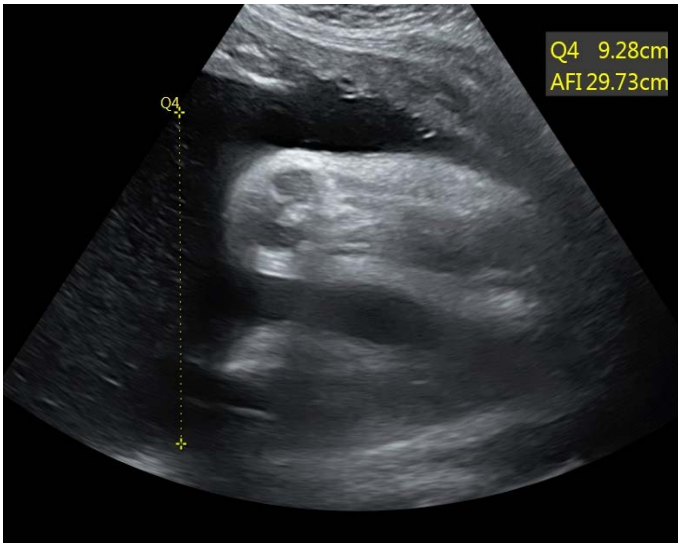

Figure 1: Patient's prenatal 30-week anatomy scan demonstrating polyhydramnios (AFI >24 cm).

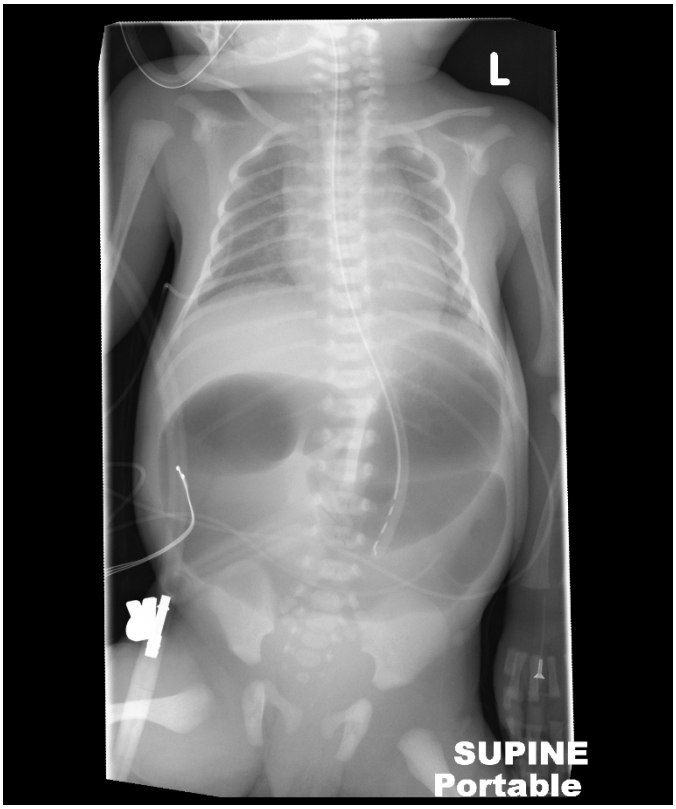

Figure 2: Initial KUB showing severely dilated loops of small bowel.

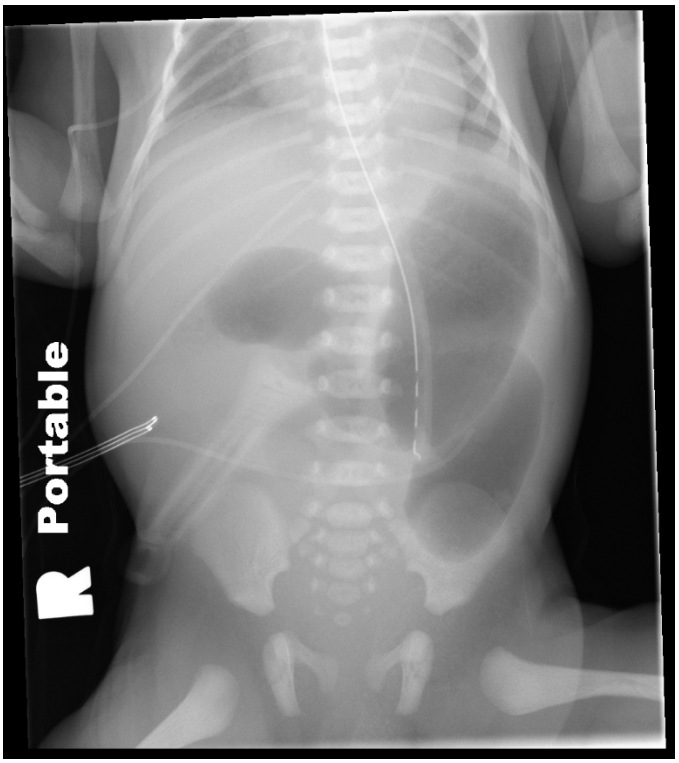

Figure 3: Four hour follow-up KUB showing decreased proximal duodenum distension.

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