

Case Report

Prenatal Sonographic Findings in a Fetus with Congenital Isolated Ileal Atresia

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Most cases of isolated nonduodenal bowel atresia are thought to be due to ischemic events from hypotension, vascular accident, volvulus, intussusception and cocaine. We present a case of isolated ileal atresia suggested using results of prenatal sonography which showed multiple markedly dilated fluid-filled intestinal loops with changeable shapes and positions secondary to active peristalsis. Postnatal oral contrast radiography demonstrated obstruction located at the ileal level. Exploratory laparotomy confirmed isolated ileal atresia. Primary end-to-end anastomosis was done. The infant endured the procedure well. Cases with multiple intestinal atresia always have fatal prognosis, even after surgery. Cases of isolated intestinal atresia usually have better prognosis and deserve more attention. Early diagnosis and early intervention are both important for good outcomes.

Key Words

isolated ileal atresia;
prenatal diagnosis;
sonographic findings

Atresias are the most common congenital anomalies of the midgut.¹ The incidence is approximately 2 to 3 in 10,000 births.² Jejunioileal atresia usually occurs more frequently than duodenal atresia. Multiple atresias are found in 7 to 20% of cases.^{1,3} A few cases of intestinal atresia with detailed descriptions of prenatal sonographic findings have been reported.⁴⁻⁹ We present a case of isolated ileal atresia suggested by results of prenatal sonography and confirmed by postnatal radiographs with successful surgery. We also discuss the clinical significance by reviewing recent reports in the literature.

CASE REPORT

A 23-year-old primigravida at 36 4/7 weeks of gestation was referred with suggestion of fetal anomalies. Prenatal sonography (Acuson, Aspen, CA, USA) revealed a relatively enlarged fetal abdomen caused by multiple markedly dilated fluid-filled loops with active peristaltic movements (Fig. 1). The maximal diameter

of the loops was about 5 cm. The anus was found and no obvious signs of dilated rectum or colon were identified. Distal small intestine obstruction was suggested. No other anomalies were found, including polyhydramnios, meconium peritonitis, gastroschisis and defects of other organs (heart, kidney, liver, head and bones). Color Doppler showed normal umbilical artery and ductus venosus flow. After counseling, a 2964-g male infant was delivered by cesarean section due to malpresentation 2 days after sonography. Apgar scores were 7 and 8 at 1 and 5 minutes, respectively. The chromosomal study showed 46, XY. Abdominal distension was noted and greenish bile-like materials were sucked from the stomach. Oral contrast radiographs demonstrated distended bowels, which suggested obstruction at the ileal level (Fig. 2). Exploratory laparotomy on the third day postpartum revealed an isolated ileal atresia, about 45 cm proximal to the ileocecal valve (Fig. 3). After cutting 2 blind ends (about 20 cm in length) and tapering the proximal dilated ileum, end-to-end primary anastomosis was performed. Meconium and contrast medium were passed on the second post-operative day. The in-

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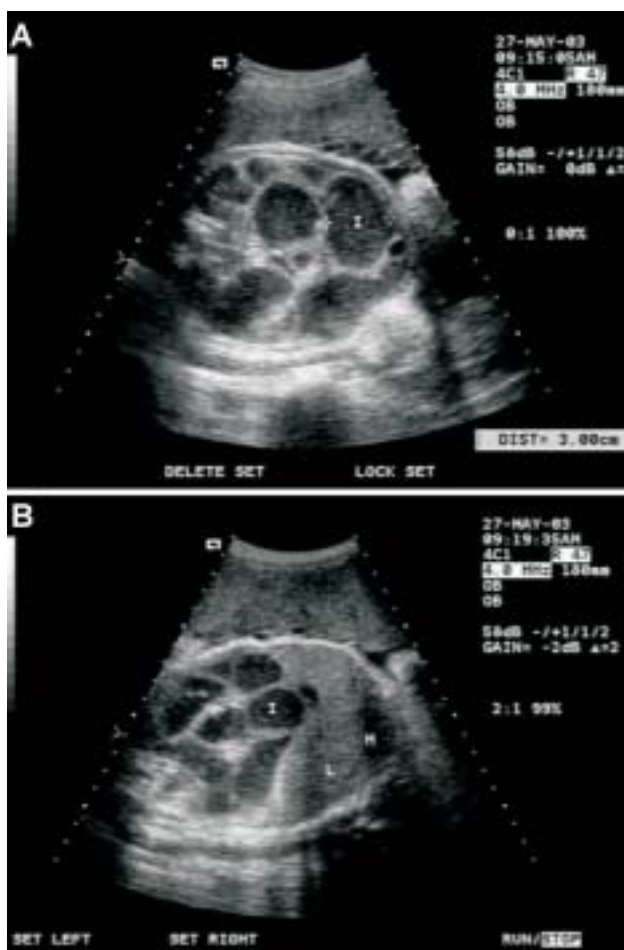


Fig. 1. Multiple dilated fluid-filled intestinal loops (I) presented with changeable shapes and positions secondary to active peristalsis. (A) transverse view, (B) longitudinal view. L = liver, H = heart.



Fig. 2. Oral contrast radiograph showed the markedly dilated small intestine (arrows) at the ileal level.

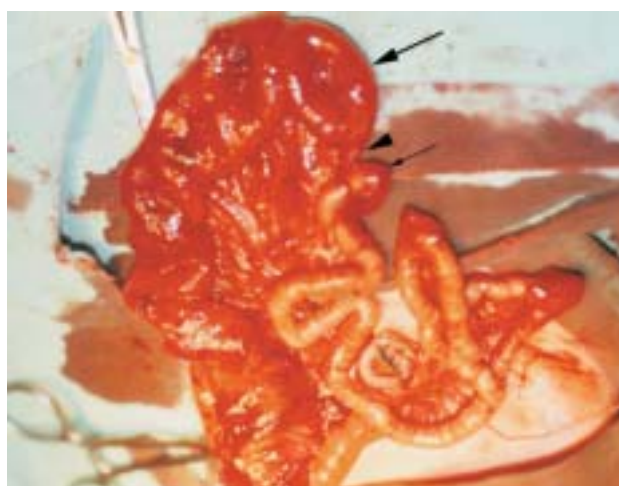


Fig. 3. Intraoperative photograph showed isolated type III atresia with complete defect (arrow head) between the dilated proximal ileum (large arrow) and the small distal ileum (small arrow).

fant endured the procedure well and was discharged 2 weeks after surgery.

DISCUSSION

The embryonic origins are different for duodenal atresia and jejunoileal atresia. Duodenal atresia results from failure of recanalization of lumen before the 10th gestational week.^{1,10} Some genetically determined factors may act to induce the transition from solid stage to hollow stage. Autosomal recessive gene and trisomy 21 are partially responsible for the duodenal atresia.^{2,10} Most cases of isolated nonduodenal bowel atresia are thought to be due to ischemic events from hypotension (due to “brain-sparing” effect in the fetus), vascular accident, volvulus, intussusception and cocaine.² The events resulting in ischemic necrosis of a segment of the intestine may develop after the onset of fetal swallowing, so that meconium may be present in the distal bowel beyond the atresia.¹ Hereditary multiple intestinal atresia has been associated with autosomal recessive genetic syndromes including cystic fibrosis.^{2,4-6,11} Intestinal atresia can be classified into 4 types.¹² Type I atresia is caused by a luminal diaphragm with continuing outer muscular layer. In type II, the 2 blind ends of the bowel are attached by a fibrous cord. Type III

atresia is complete separation of the bowel ends. Type IIIa atresia is associated with a V-shaped mesenteric defect. Type IIIb atresia is referred to apple-peel atresia. Multiple atresia is classified as type IV. Our case was type IIIa.

Differential diagnosis of fetal abdominal cysts should include origins from the ovary, urinary organ, hepatic-biliary organ and intestine. Multiple cysts with obvious peristalsis inclined the consideration of intestinal lesions. More loops dilated usually means more distal obstruction and less severity of the polyhydramnios. Absent signs of dilated rectum or colon in our case excluded the possibility of imperforate anus. Intestinal volvulus often presents as a whorl-like cystic mass and fixed dilated loops arranged in 2 to 3 parallel segments.¹³ Isolated ileal atresia in our case showed multiple markedly dilated fluid-filled loops with active peristaltic movements. The loops were shown communicating after careful tracing of the course of the dilated loops using real-time ultrasound. Previous report has mentioned the possibility that torsion of dilated bowel may occur.⁷ Congenital isolated ileal atresia has been reported as a single cyst-like lesion due to the incomplete torsion of the dilated proximal small bowel.⁸ Late detection may lead to intestinal necrosis, resulting in meconium peritonitis. However, the findings in our case were different.

Patients with multiple intestinal atresia always have poor prognoses, even after surgery.^{6,11} Cases with prenatally closed gastroschisis always suffer from extended intestinal atresia from the jejunum to the proximal transverse colon and lead to death.¹⁴ Those with isolated intestinal atresia usually have better prognoses, especially when located at more distal portion, diagnosed prenatally and managed prior to the development of severe distention or perforations of bowels. Accordingly, isolated intestinal atresia deserves more attention. The favorable prognosis in our case resulted from the absence of meconium peritonitis, meconium ileus, malrotation, polyhydramnios, abnormal chromosome and other congenital anomalies. Early obstetric intervention, successful pediatric surgery and meticulously intensive care of the newborn also helped achieve the

positive outcome.

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