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Clinical History: A 22 year old G1P0 female was referred from an outside clinic with report of an abnormal abdominal mass seen on a second trimester fetal ultrasound. Serial second and third trimester fetal ultrasounds were performed at our institution. The baby was delivered near term via induced vaginal delivery. An abdominal ultrasound was performed on the baby's first day of life and the neonate underwent exploratory laparotomy with resection and temporary colostomy on the second day of life. The patient subsequently underwent right hemicolectomy with creation of an ileocolostomy at age 2 months.

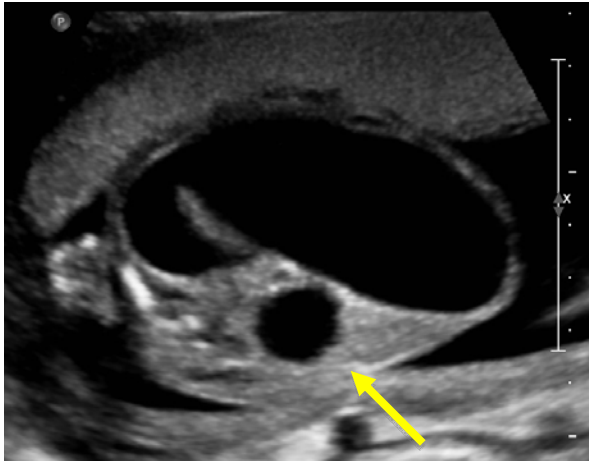
Figures:

FIGURE 1. Ultrasound images of the fetus at 20 weeks 5 days estimated gestational age.
A: Sagittal midline US image demonstrates a large cystic abdominal mass.

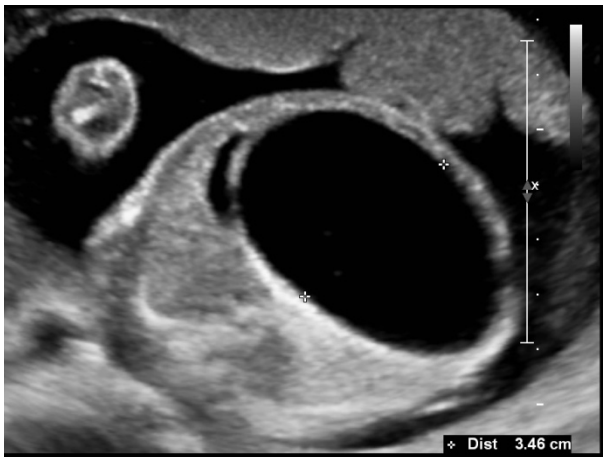


B, C: (B) The cystic mass is noted to be serpiginous on this transverse oblique US image of the fetal abdomen. The mass is separate from the stomach (yellow arrow) and follows the expected course of the transverse colon. (C) The mass maximally measures 3.5 cm in the mid abdomen.

B.



C.



D: On this transverse ultrasound image at the level of the sacrum, the serpiginous cystic mass resembles a blind ending loop of bowel.



E: Transverse US image of the fetal abdomen shows that the distended cystic mass contains some echogenic debris.

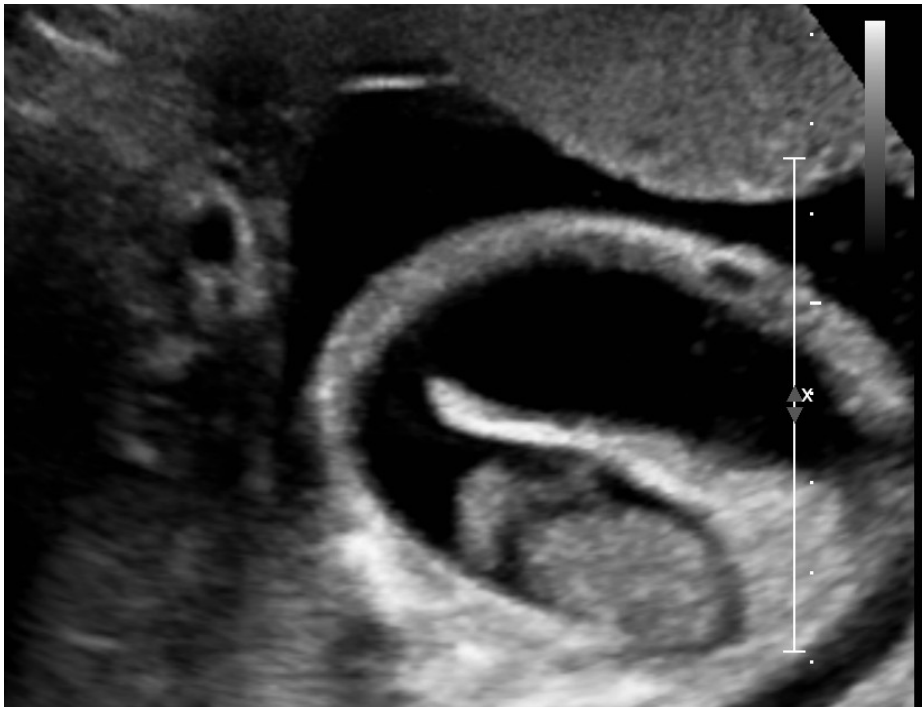


FIGURE 2: Follow up ultrasound at 21 weeks, 3 days
Transverse image of the fetal abdomen demonstrates decompression of the cystic mass (blue arrow) and interval development of ascites (yellow arrow). These new findings suggest interval perforation. The mass remains tubular in shape and mucosal hyperechogenicity is suggestive of the echogenic bowel wall mucosa/bowel signature.

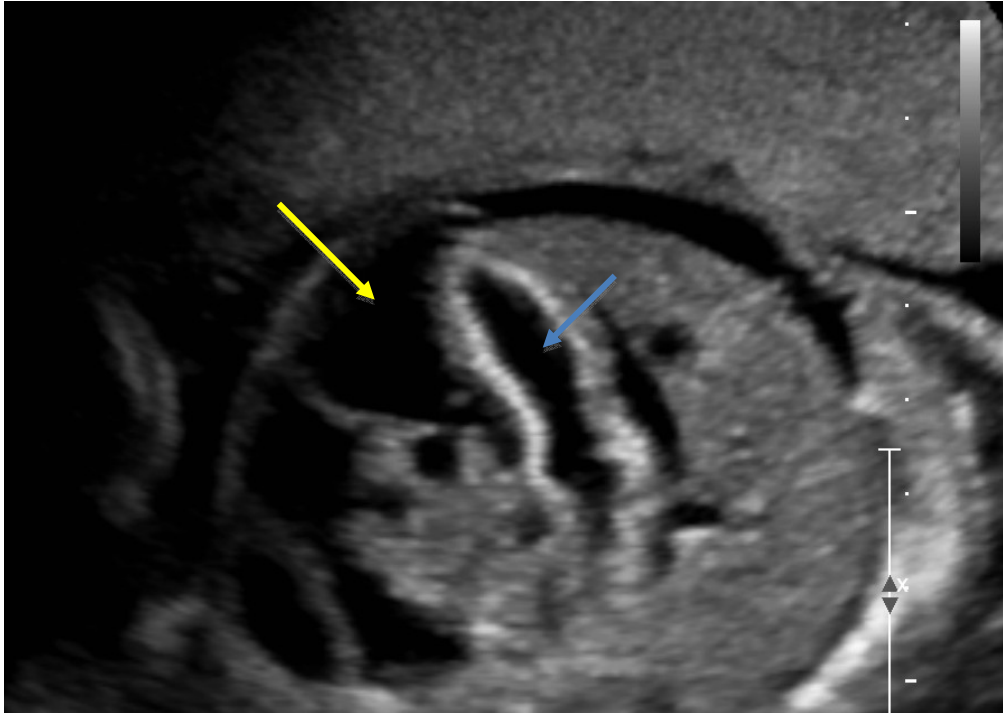


FIGURE 3: Neonatal abdominal ultrasound, day 1
Note large debris filled intra-abdominal collection. The surrounding rim has an echogenic inner layer, hypoechoic middle layer and echogenic outer layer, suggestive of the classic US bowel wall signature.

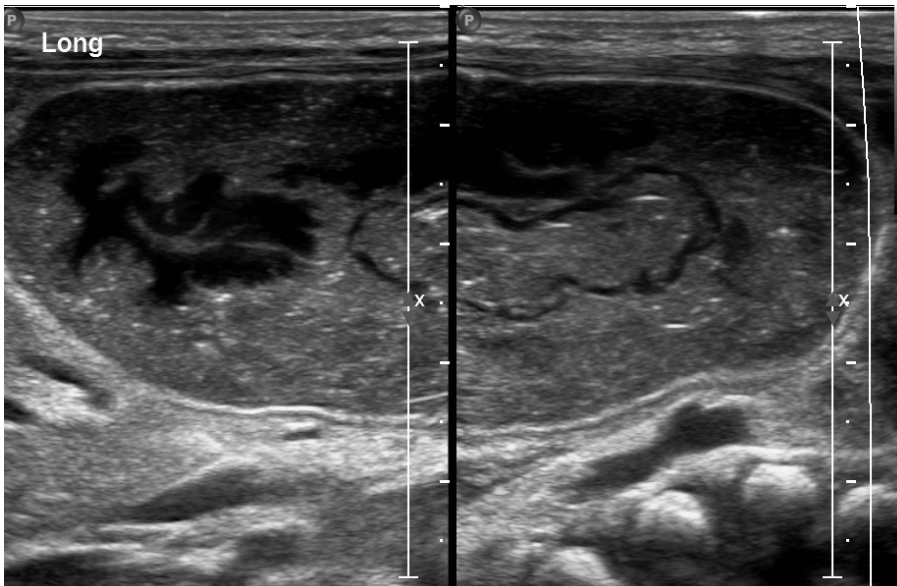


FIGURE 4: Water soluble contrast enema performed at 2 months of age. Note: small caliber distal colon (microcolon) with normal rectosigmoid ratio and no evidence for strictures.



Diagnosis: Colonic atresia (CA).

Discussion:

CA is an extremely rare cause of fetal intestinal obstruction and constitutes approximately 1.8% to 15% of all intestinal atresias (1). The incidence has been estimated at about 1 in 66,000 births (2). The etiology of most cases of CA is widely accepted to be a vascular insult to the fetal intestine from volvulus, internal hernia, intussusception, or strangulation in tight gastroschisis (3). Although cases with genetic and familial occurrence of CA have been reported, most reports of CA are sporadic (4). A 4:3 male to female ratio reveals a slight male predominance for CA and most newborns with CA are reported to be full term (3).

CA is a type of low intestinal obstruction, involving the distal ileum or colon, as opposed to high intestinal obstructions that involve bowel proximal to the midileum. CA is often indistinguishable from obstruction of the distal ileum on imaging, especially when the atresia is located in the ascending colon (5). As seen in our case, the colon proximal to the point of atresia is often massively dilated. If the ileocecal valve is intact, CA can become the functional equivalent of a closed loop obstruction and has a high risk of perforation (3). Barium enema examination usually reveals a distal microcolon with obstruction to the retrograde flow of barium at the site of atresia (5). The ultrasound features of CA include dilation of the distal small bowel and proximal colon with increased echogenicity of the involved bowel secondary to retained

meconium (5). Ultrasound differentiation between small bowel obstruction and colonic obstruction in young children is possible only if the distal portions of the colon are visualized and appear collapsed.

In the fetus, CA becomes even more difficult to differentiate from other congenital gastrointestinal abnormalities that may present as an abdominal cystic mass. The differential diagnosis on obstetric ultrasound is therefore broad and includes enteric duplication cyst, peritoneal pseudocyst, bowel volvulus, or other causes for low intestinal obstruction such as anal atresia (6).

With regard to prognosis, CA is the most favorable type of congenital bowel atresia with respect to morbidity and mortality, since in the absence of small bowel atresia, the normal length of the small intestine should allow for normal bowel function (3). The incidence of associated congenital anomalies is also much lower relative to other intestinal atresias (1). In fact, CA has been reported to be an isolated anomaly in about two thirds of cases (3).

Treatment of CA is primarily operative with primary anastomosis for atresias proximal to the splenic flexure and colostomy but delayed anastomosis for those occurring distal to the splenic flexure being the classic choice for most surgeons (3). Statistically significant high mortality in cases of CA operated on after the first 72 hours of life highlight the importance of early diagnosis and treatment of this entity (3).

References:

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