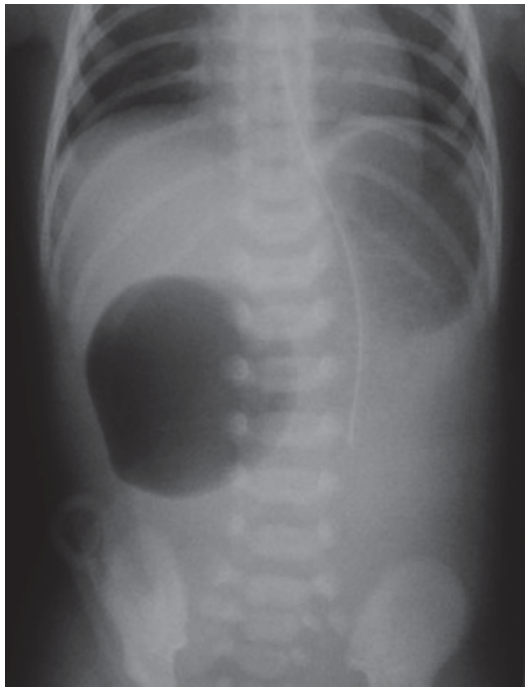
January

ASSESSMENT PROGRESS: Total Questions: **10** Questions Answered: **9** Correct Answers: **4**

Question 9

The newborn nursery staff calls you to evaluate a full-term infant with emesis following each feeding. The nurse reports that the 18-hour-old infant does not have abdominal distention. After reviewing the abdominal radiograph (**Figure 1**) with the resident on call, you discuss historical and clinical findings of the infant's condition.

Figure 1: Duodenal atresia (adapted from http://www.adhb.govt.nz/newborn/teachingresources/radiology/AXR/DuodenalAtresia/AXR-DuodenalAtresiaSupine.jpg).



Of the following, the clinical or historical finding MOST likely associated with this infant's condition is:

- ✓ **A.** additional congenital anomalies
- ○ **B.** hematochezia
- ○ **C.** maternal oligohydramnios
- ○ **D.** nonbilious emesis
- ○ **E.** tender abdomen

Correct

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The infant's abdominal radiograph shows a classic double-bubble sign. Gas or air-fluid levels in the markedly dilated stomach and the proximal duodenal bulb form the two bubbles of the double-bubble sign. The classic double-bubble sign is found in neonates with a congenital duodenal obstruction.

Duodenal atresias and stenosis, which occur in 1 in 7,000 live births, comprise approximately 50% of small intestinal atresias. Congenital duodenal atresia is associated with additional serious anomalies in 50% to 80% of cases, and these anomalies are responsible for much of the morbidity and mortality among patients with duodenal atresia. Congenital heart defects and trisomy 21 are the most common associated conditions. Because of the 30% incidence of congenital heart disease in patients with duodenal atresia, most surgeons will evaluate cardiac anatomy and function before surgically correcting the atresia. Other anomalies that can be seen with duodenal atresias may include intestinal malrotation, esophageal atresia, imperforate anus, heterotaxia, and gallbladder agenesis.

Congenital duodenal obstruction can be complete or partial and may be intrinsic or extrinsic. Extrinsic obstruction to the duodenum may be caused by:

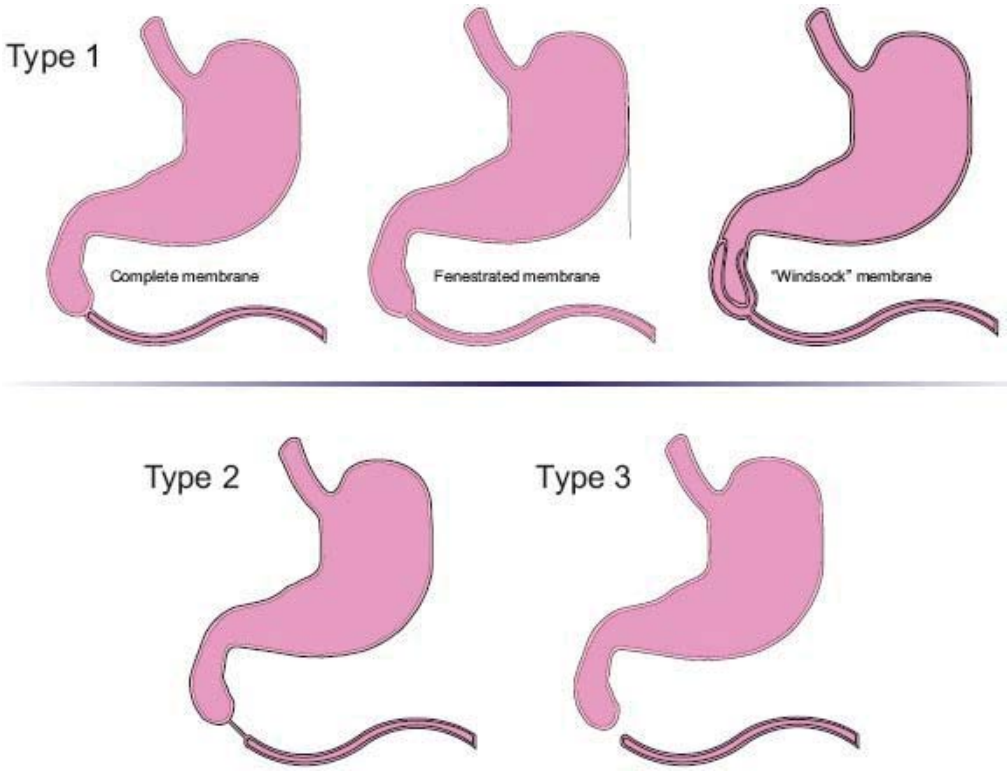
- malrotation with Ladd bands
- preduodenal portal vein
- gastroduodenal duplication
- cysts or pseudocyts of the pancreas and biliary tree
- annular pancreas

Intrinsic atresia and stenosis likely stem from failure of the duodenum to recanalize after obliteration of the lumen by epithelial proliferation during the first trimester. Atresias of the duodenum have three morphologic appearances **(Figure 2)**.

- Type I atresias have a luminal membrane, which occasionally takes on a windsock appearance**.**
- Type II atresias have a dilated proximal and narrowed distal segment separated by a fibrous cord.
- Type III lesions are characterized by a discontinuity between the segments.

Figure 2: Variants of congenital duodenal atresia.

Feedback



The fluid-filled structures of the double bubble can be detected with prenatal ultrasonography at 22 to 23 weeks' gestation in 15% to 20% of cases of duodenal atresia. In one series, maternal polyhydramnios was noted during prenatal screening in 75% of cases.

The clinical presentation of an infant with congenital duodenal obstruction depends on the presence or absence of a membrane aperture and on its size and location with respect to the ampulla of Vater. The distal intestinal tract may be gasless, as it was in the infant in the vignette, or it may have a small amount of air from a small opening or microperforation. Because the site of the duodenal obstruction is distal to the ampulla in more than 80% of cases, the classic presentation includes bilious vomiting within 24 hours of birth.

Neonates with a midgut volvulus can also present with bilious emesis. A midgut volvulus, which is present at surgery in approximately 50% of neonates with malrotation, should be considered in the differential diagnosis of any neonate presenting with an upper gastrointestinal obstruction. The appearance of the duodenum on a plain radiograph can be used to help differentiate a midgut volvulus from duodenal atresia. In the classic double-bubble sign, as seen in the neonate in the vignette, the duodenum will appear distended and rounded from the chronic intrauterine obstruction. If the stomach is distended and the duodenum is of normal caliber, malrotation with duodenal obstruction secondary to Ladd's band or a volvulus should be considered. Occult or gross blood from intestinal ischemia and mucosal injury can be seen in neonates with a midgut volvulus but is uncommon in neonates with duodenal atresia. Initially the abdomen may not be distended in neonates with a volvulus. Persistent intestinal obstruction and vascular insufficiency will result in abdominal distention and tenderness, findings that are unusual in neonates with congenital duodenal atresia.

An abdominal radiograph with a double-bubble sign and without intestinal air beyond the duodenum in an infant without abdominal distention or tenderness suggests complete congenital duodenal obstruction. In such a circumstance no additional abdominal radiographic imaging is necessary before surgery. Unstable infants may require echocardiography and an upper gastrointestinal contrast study to help distinquish between hemodynamic compromise caused by a midgut volvulus or a cardiac anomaly that may be associated with the duodenal atresia.

References:

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American Board of Pediatrics Content Specification(s):

11_Gastroenterology: Know the pathogenesis of atresias, stenosis, diverticulae, and duplications of the small intestine including those associated with annular pancreas

11_Gastroenterology: Know the clinical manifestations of atresias, stenosis, diverticulae, and duplications of the small intestine including those associated with annular pancreas

11_Gastroenterology: Know the approach to diagnosis and management of atresias, stenosis, diverticulae, and duplication of the small intestine including those associated with annular pancreas

11_Gastroenterology: Know the approach to diagnosis and management of infants with malrotation and/or volvulus of the small intestine

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