



UNIVERSITY of  
**ROCHESTER**  
MEDICAL CENTER

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**DEPARTMENT OF IMAGING SCIENCES**

**Imaging Sciences Interesting Cases**

**CASE 14**

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**CLINICAL PRESENTATION:** Newborn with trisomy 21 and bilious emesis.

**IMAGING FINDINGS:** Plain film of the abdomen after air insufflation of the GI tract via indwelling enteric tube shows the classic “double bubble” sign of duodenal atresia - a gas filled stomach and duodenal bulb with no gas distally.



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**Figure 1**

## **DIAGNOSIS: Duodenal Atresia**

**DISCUSSION:** Duodenal atresia represents complete obliteration of the duodenal lumen. A duodenal diaphragm (or duodenal web) is thought to represent a milder form of the same process, with incomplete obliteration of the lumen. The etiology of duodenal atresia and stenosis is unknown. Failure of recanalization of the duodenal lumen remains the favored theory, as opposed to intrauterine vascular ischemia. In most cases, duodenal atresia occurs below the ampulla of Vater. Duodenal atresia rarely occurs proximal to the ampulla. Bilious emesis in neonates aged 24 hours or younger is the typical presentation of atresia or severe stenosis. Minimal duodenal obstruction in mild stenosis or duodenal membrane may have few symptoms and present later in life.

The incidence of duodenal atresia in the US is 1 per 6000 births. Intrinsic congenital duodenal obstruction constitutes two thirds of all congenital duodenal obstructions, with extrinsic compression from an annular pancreas comprising the other third. If duodenal atresia or significant duodenal stenosis is left untreated, the condition rapidly becomes fatal as a result of electrolyte and fluid imbalance.

**ASSOCIATIONS:** About 30% of patients with duodenal obstruction have trisomy 21. Other problems associated with trisomy 21 include cardiac defects (most commonly ventricular septal defects and endocardial cushion defects), as well as Hirschsprung disease. Duodenal anomalies can be associated with other GI and biliary tract abnormalities (malrotation, esophageal atresia, imperforate anus, annular pancreas, gallbladder or biliary atresia, vertebral anomalies). In addition, duodenal atresia can be associated with congenital abnormalities in other systems, such as the VACTERL (vertebral, anal, cardiac, tracheal, esophageal, renal, limb) syndrome. One half of neonates with duodenal atresia or stenosis are born prematurely.

**IMAGING:** Plain radiographs demonstrating the classic "double-bubble" appearance with no distal gas are characteristic of duodenal atresia. Distal bowel gas indicates stenosis, incomplete membrane, a hepatopancreatic ductal anomaly, or other causes of proximal obstruction such as malrotation with midgut volvulus.

**TREATMENT:** Duodenoduodenostomy is curative.

## **REFERENCES:**

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2. Forrester MB, Merz RD. Population-based study of small intestinal atresia and stenosis, Hawaii, 1986-2000. Public Health. 2004 Sep;118(6):434-8. [PubMed]