

A 3-hour-old girl is in the newborn nursery with choking, coughing, and regurgitation after her first breast milk feeding. She was born at 41 weeks gestation by vaginal delivery to a healthy woman who was compliant with prenatal care. Prenatal laboratory studies were normal. Second- and third-trimester ultrasounds showed polyhydramnios. Clear amniotic fluid was seen after artificial rupture of the membranes 2 hours prior to delivery. Apgar scores were 9 at 1 and 5 minutes of life. The neonate is tachycardic, tachypneic, and hypoxic. Examination shows a crying neonate with copious oral secretions, marked respiratory distress, and abdominal distension. Crackles are heard in the right lower lobe. The anus is patent. Endotracheal and orogastric tubes are inserted. An umbilical venous catheter is placed. X-ray of the chest and abdomen is shown below.







Which of the following is the most likely diagnosis in this patient?

- ☐ A. Bilateral choanal atresia
- ☐ B. Congenital diaphragmatic hernia
- ☐ C. Duodenal atresia



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- ☐ C. Duodenal atresia
- ☐ D. Esophageal atresia with tracheoesophageal fistula
- ☐ E. Isolated esophageal atresia
- ☐ F. Meconium aspiration syndrome
- ☐ G. Vascular ring

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- ☐ A. Bilateral choanal atresia [2%]
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- ☐ C. Duodenal atresia [2%]



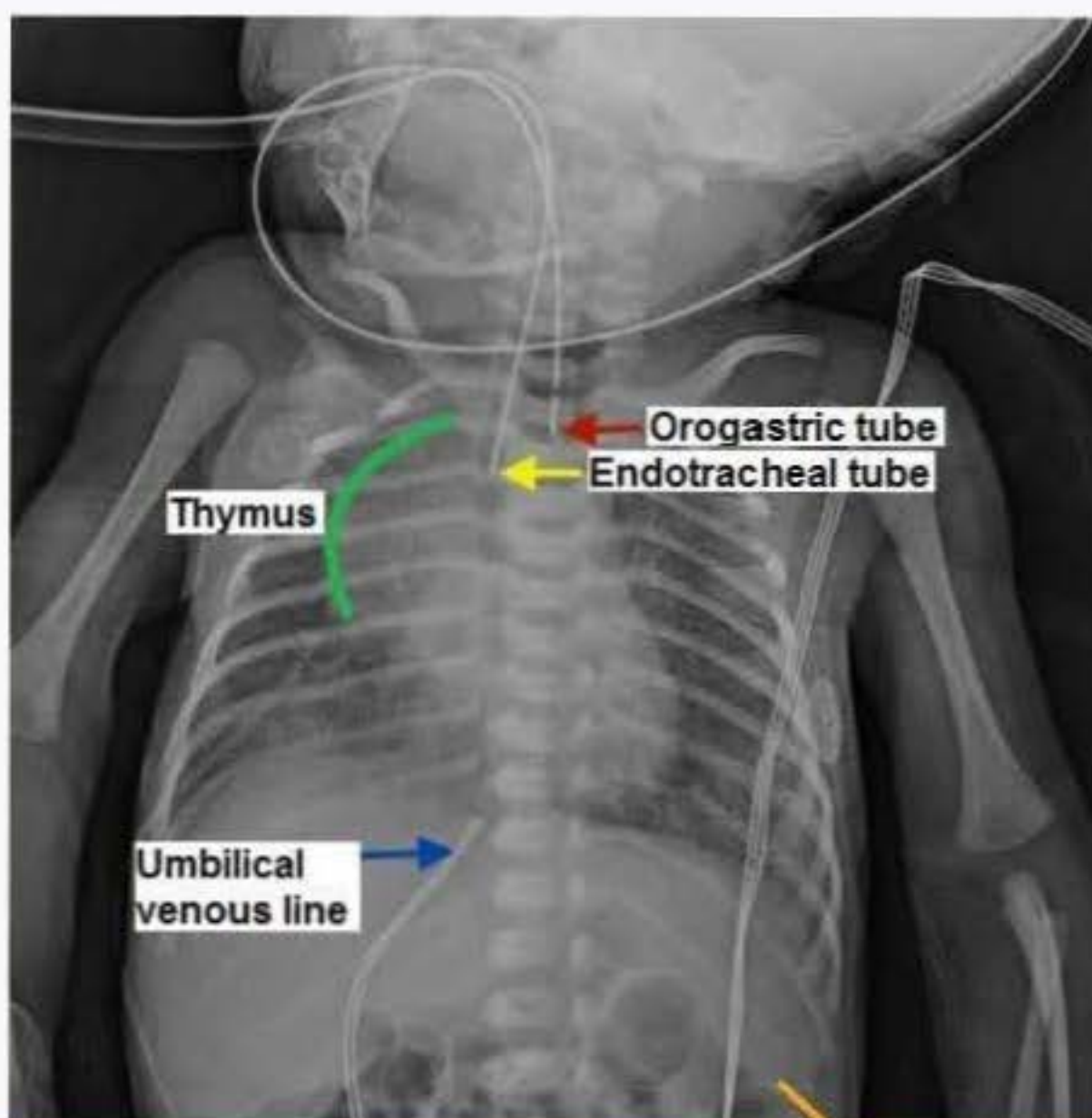
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- ☐ C. Duodenal atresia [2%]
- ☒ D. Esophageal atresia with tracheoesophageal fistula [87%]
- ☐ E. Isolated esophageal atresia [4%]
- ☐ F. Meconium aspiration syndrome [3%]
- ☐ G. Vascular ring [1%]

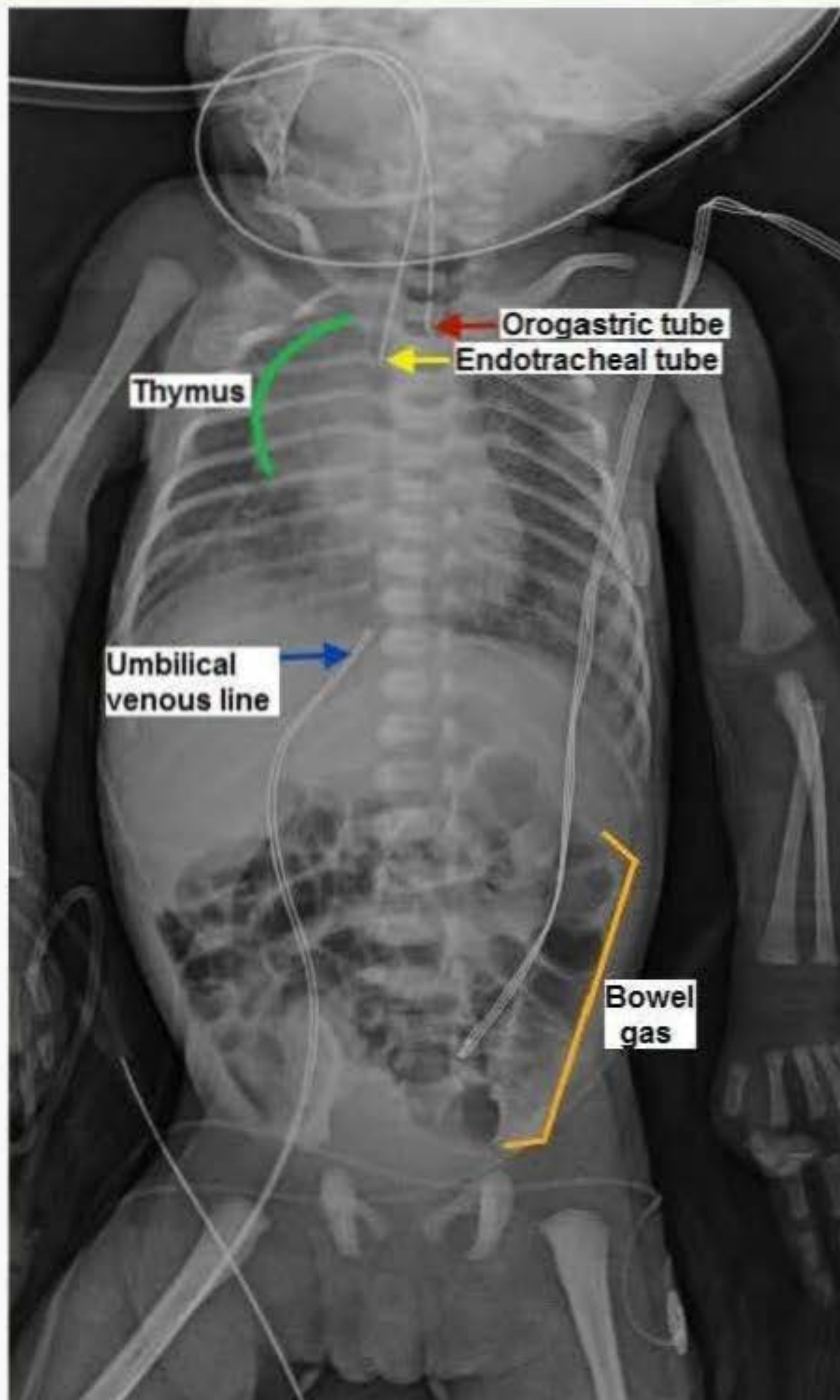
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Explanation:

User Id: [REDACTED]







Anomalies of the esophagus and trachea are common and affect 1 in 3500 births. Improper formation during the 4th and 5th gestational weeks can result in various versions of **esophageal atresia and tracheoesophageal fistula**. The most common defective arrangement is proximal esophageal atresia with a fistula between the trachea and distal esophagus.

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Anomalies of the esophagus and trachea are common and affect 1 in 3500 births. Improper formation during the 4th and 5th gestational weeks can result in various versions of **esophageal atresia and tracheoesophageal fistula**. The most common defective arrangement is proximal esophageal atresia with a fistula between the trachea and distal esophagus.

Prenatal ultrasound shows **polyhydramnios**, as the affected fetus cannot swallow amniotic fluid. Excessive drooling as well as **choking, coughing, and regurgitation with initial feeding attempts** are seen immediately after birth. The presence of the **enteric tube in the proximal esophagus on x-ray** and inability to pass a feeding tube into the stomach is extremely suggestive of esophageal atresia. The tracheoesophageal fistula permits air entry into the gastrointestinal tract, and the stomach and intestines can become quite distended with each breath, especially in the ventilated patient. In addition, gastric fluid can reflux into the distal esophagus through the fistula and into the trachea and lungs, causing **aspiration pneumonia**. This patient's crackles and x-ray infiltrates in her right lower lung are consistent with pneumonia.

As many as half of patients with tracheal and esophageal defects have additional anomalies. Workup for VACTERL (vertebral, anal atresia, cardiac, tracheoesophageal fistula, renal, limb) association should be considered.

**(Choice A)** Bilateral choanal atresia typically presents with cyanosis and respiratory distress during feeding that improve when the infant cries.

**(Choice B)** **Congenital diaphragmatic hernia** presents as cyanosis and respiratory distress immediately after birth. Polyhydramnios can occur as a result of esophageal compression. However, the deviation of abdominal viscera into the thorax results in a scaphoid-appearing abdomen. In addition, x-ray shows a displaced cardiac silhouette, bowel in the thorax, and a gasless abdomen.

**(Choice C)** **Duodenal atresia** can cause polyhydramnios and vomiting with initial feeds but not respiratory distress. The abdomen would not be distended and intestinal gas would be absent on x-ray as air cannot pass the duodenum. In addition, gastric tubes can be inserted without resistance and x-ray would show air in the stomach and proximal duodenum ("double-bubble sign").

**(Choice E)** Esophageal atresia without fistulous connection to the trachea is rare and would not cause sudden respiratory distress with feeding.

**(Choice F)** Neonates born after their due date are at increased risk of passing meconium in utero. Meconium aspiration syndrome can cause respiratory distress and infiltrates on x-ray if neonates inhale the meconium-stained amniotic fluid during birth.



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**(Choice F)** Neonates born after their due date are at increased risk of passing meconium in utero. Meconium aspiration syndrome can cause respiratory distress and infiltrates on x-ray if neonates inhale the meconium-stained amniotic fluid during birth. This etiology is unlikely given the clear amniotic fluid and presence of enteric tube in the proximal esophagus.

**(Choice G)** **Anomalous vascular branches of the aortic arch** can cause stridor and dysphagia due to compression of the trachea and esophagus. However, naso- or orogastric tubes can be advanced into the stomach.

#### Educational objective:

Esophageal atresia with tracheoesophageal fistula should be suspected when a newborn chokes and coughs during the first feeding. Attempts at naso- or orogastric tube insertion will fail and x-ray will show the tube in the proximal esophageal pouch.

#### References:

1. **Current knowledge on esophageal atresia.**
2. **Current status of prenatal diagnosis, operative management and outcome of esophageal atresia/tracheo-esophageal fistula.**



Media Exhibit

esophageal atresia & tracheoesophageal fistula

### Various types of esophageal atresia & tracheoesophageal fistula



Esophageal atresia with distal fistula (~85%)



Isolated esophageal atresia (~8%)



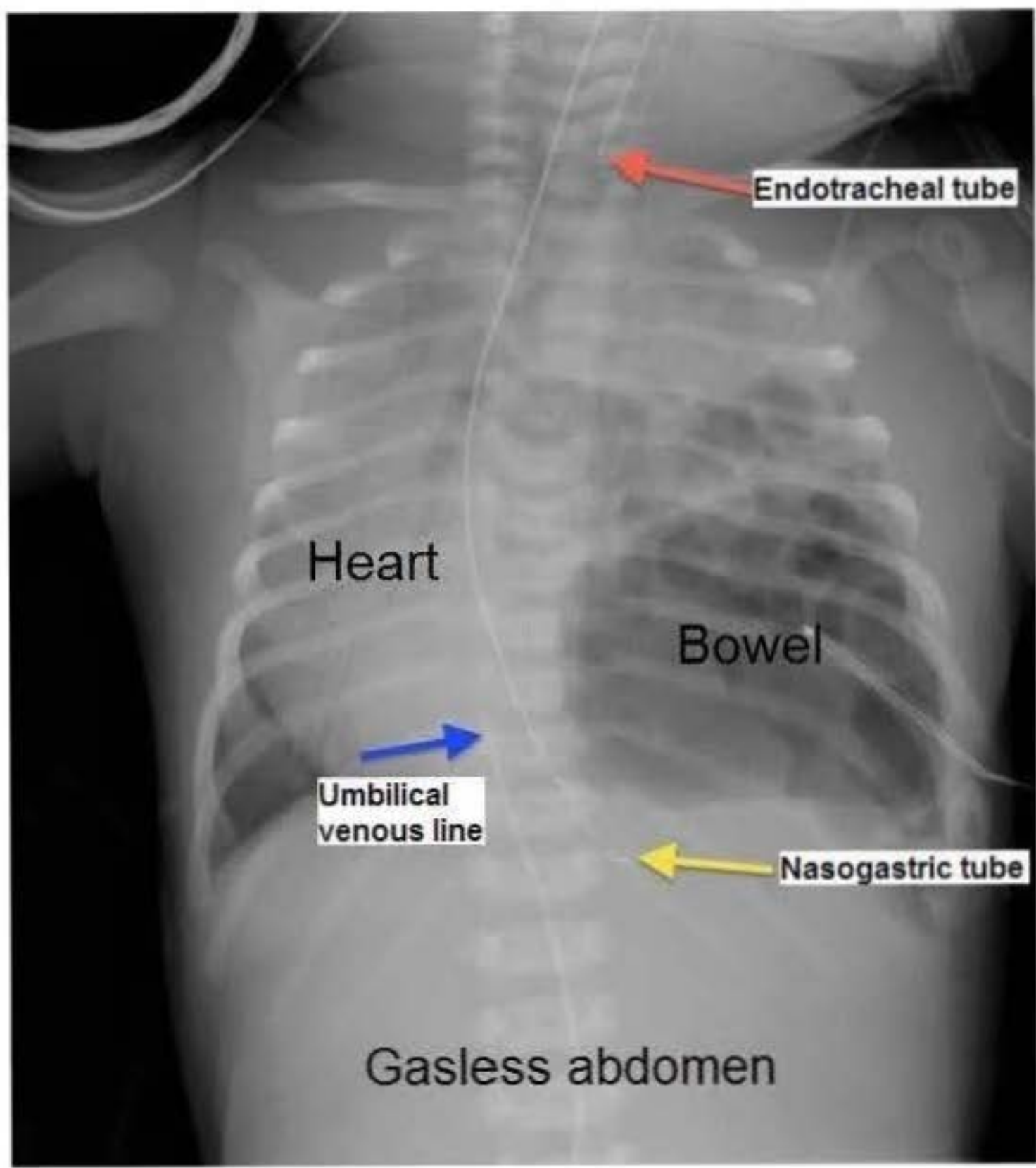
H-type tracheoesophageal fistula (~4%)





Media Exhibit

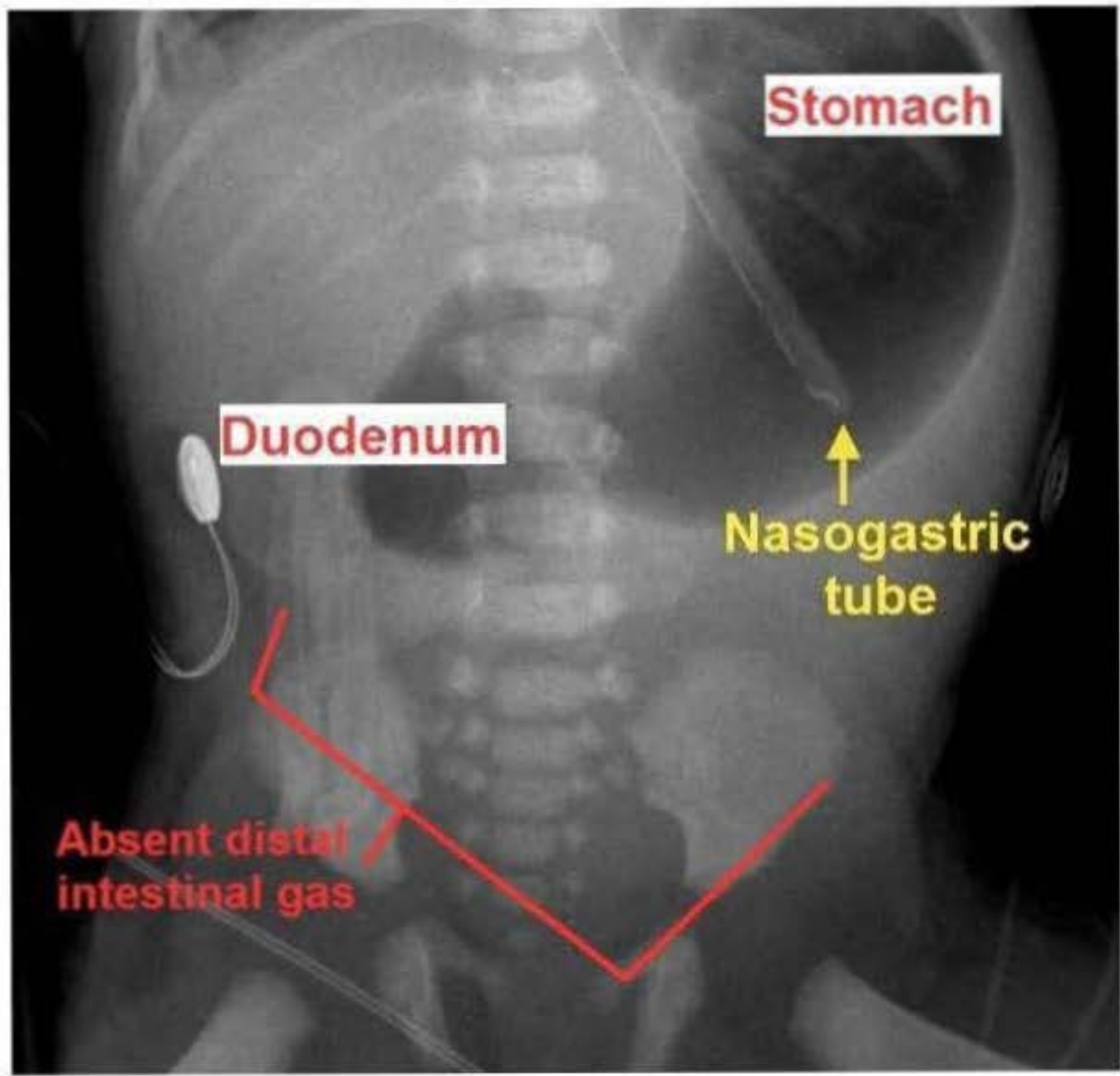
tal diaphragmatic hernia





Media Exhibit

al atresia

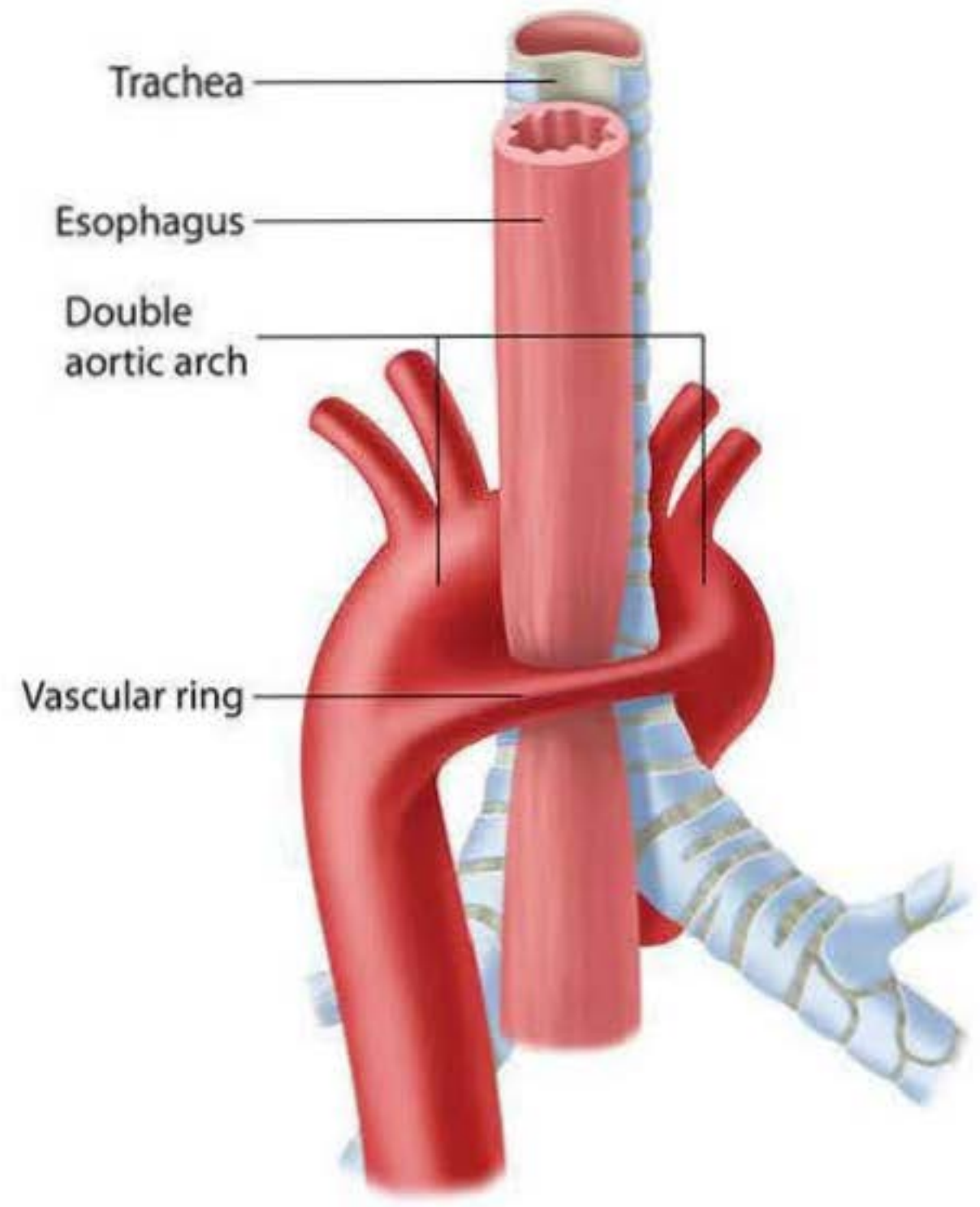




Media Exhibit

ring, posterior view

Vascular ring, posterior view



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