The long and short of esophageal atresia

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Outline

• What is esophageal atresia (EA)?
• Diagnosing EA: the role for imaging
• Management of short-gap vs long-gap EA
• The Foker Method
• One patient’s story through her diagnosis, therapy, complications, and cure
EA: Classification

- TEF (8%)
  - Proximal (1%)
  - Both (3%)
  - Distal (84%)

* TEF without EA (H type, 4%)

EA: the basics

- Most common esophageal abnormality (1/2500-1/3500)
- Cause: unclear
- Associated with anomalies >50% of the time
  - Highest risk of VACTERL with isolated EA (no TEF)
- Presentation: at birth with excessive salivation, suffocation, cyanosis, feeding intolerance, pneumonia
- Treatment: repaired surgically
EA: Prenatal diagnosis

- Prenatal: difficult to diagnose
  - US: polyhydramnios + dilated proximal pouch + absence of gastric bubble
  - not specific or sensitive findings, not always present with TEF
  - MR: nonvisualization of intra-thoracic esophagus fundus

Second/third trimester screen: evaluation of fetal presentation, amniotic fluid volume, cardiac activity, placental position, fetal biometry, and fetal number, plus an anatomic survey (gastric bubble)


7. AIUM 2012 Practice Guidelines for the Performance of Obstetric Ultrasound Exams
EA: Post-natal diagnosis

Delivery room:

– Symptoms, impassibility of orogastric catheter past 11-12cm

– AP CXR with air as contrast (avoid aspiration of contrast fluid!): catheter coiled in esophagus

  • Air in stomach, indicates TEF → increased urgency for surgery, high risk of aspiration

  • Absence of air in stomach indicates no TEF
Companion Patients 2, 3: EA with and without TEF on AP Chest X-Ray

Left: note nonprogression of orogastric catheter, lack of gastric air bubble

Right: note nonprogression of catheter, presence of gastric air bubble indicating TEF

8. LearningRadiology.com
9. radiopaedia.org
EA: Treatment

**Pre-op:** full aspiration precautions, VACTERL assessment
- orogastric tube (suction), head of bed 45°, acid suppression, avoid PPV
- renal US, ECHO, etc.

**Primary repair:** thoracoscopic or thoracotic ligation

**Contraindications:**
- Low birth weight, congenital heart disease, long gap length, compromised physiologic status

EA: Recognizing post-operative complications through imaging

- Seven days post-op, **esophogram** is performed
- **Leak** → 15% of cases
- **Strictures** → common, recurrent, tx: serial endoscopic dilation
- **GERD** → common
- **Esophageal dysmotility** → 75%-100% of cases
- **Thoracic wall deformities** → 24% winged scapula, 21% scoliosis

2. Spitz L. Orphanet J Rare Dis 2007; 2: 24
Long-Gap EA: Management

- Primary surgery not possible
- **Gapogram** to assess gap size
  - Treatment is controversial
  - Esophagus grows from swallowing (proximal) and reflux (distal) → repair delay until 1-3 months
  - Other options: replacement vs circular myotony vs surgery under tension vs **Foker method**

**Definition:** gap too big to repair (>3cm or 2 vertebral bodies)


Companion patient 4: Gapogram to assess gap size

- Contrast through enteric tube to assess proximal stump (then removed by enteric tube)
- Contrast (or probe) through gastrostomy to assess distal stump
- Measure gap by vertebral bodies or cm
Long-gap EA: Controversy surrounding repair

OPTIONS

Growth induction

Replacement

Stomach (gastric pull up or gastric tube)

jejunum

colon

Compiled information from:
Long-gap EA: Management by Growth Induction (the Foker process)

- Sutures applied through external proximal and distal stumps, with tag on end
- Sutures are pulled out of patient’s body attached to external traction devices
- With the patient paralyzed, increasing traction is applied over time

Companion patient 5: Serial gapograms show decreasing gap size over time in a patient undergoing the Foker process.

- Over time, traction causes stress leading to natural tissue growth (rather than stretch, which may compromise tissue integrity).
- Serial gapograms are a critical part of the procedure, assessing gap size in preparation for repair.

Now that we know about different types of EA, how to diagnose each by imaging, and how imaging is used with some of the management options, let’s meet our patient!
First, let’s go through our patient’s diagnostic studies
Our Patient: AP Chest X-Ray at delivery

Pause to evaluate and continue for labeled findings.
Did you remember to assess for EA AND for TEF? This is a key distinction that changes the management of patients with EA.

Our Patient: Impassibility of NG tube on AP Chest X-Ray at delivery

NG tube impassible proximally

Paucity of gastric gas, indicating isolated EA (without TEF)
Our patient: Presentation and Diagnosis

- GT is a 50-day-old ex-32-weeker who was diagnosed prenatally with isolated EA (no TEF).
- Postnatally, a diagnosis of EA without TEF was confirmed, as you saw on chest X-Ray done at delivery.
- She lives in Tennessee, and was transferred to Boston Children’s for definitive care.
Our Patient: Pre-operative evaluation

Two major considerations:

• Aspiration precautions

• VACTERL assessment
  – She showed no signs of any other anomalies (normal ECHO, no other GI/anal anomaly, no limb anomalies, no vertebral anomalies, normal kidneys)
Our Patient: Gapogram showing 34 mm gap size

Water-soluble barium gapogram with contrast injected into the enteric tube proximally and contrast injected into a catheter passed through the gastric tube distally. A 34 mm gap between distal and proximal esophageal stumps was visualized. Note the ruler behind the patient, to calibrate findings.
Our Patient: Imaging safety considerations

Remember:
- Use water-soluble contrast in case of leak.
- Remove contrast from upper esophageal pouch immediately after! These patients have a blind-ending esophagus and it is important to be prepared with suction after imaging is performed to avoid aspiration.
Our Patient: Management

- As seen on gapogram, her gap size is 34 mm (dx: long-gap EA)
- She came to Children’s for the Foker process
- Her Foker 1 process (thoracotomy with placement of sutures and traction devices) was uncomplicated, and the ensuing slides follow the serial gapograms that assessed her stump approximation for esophageal anastomosis
Our patient, day 1 status post Foker 1 process: **External traction devices** and esophageal stump tags on portable supine chest x-ray

POD1 after Foker 1 process. Note the external traction devices attached to sutures at each esophageal stump. The **proximal** and **distal** stumps are tagged with markers. Barium gapogram is needed for definitive gap size assessment.
Our patient day 5: External traction devices with spacers and esophageal stump tags on portable supine chest x-ray

Note the external traction devices, now with spacers to increase the tension on the stumps. The proximal and distal stumps are tagged with markers, and the distance between is measured at 11.63mm (need gapogram for accurate assessment).
Our patient day 5: Injection of contrast proximally and distally on barium gapogram

Water soluble barium injected through enteric tube proximally and through catheter in the gastric tube distally. At this point in time, the proximal esophagus and distally the stomach are visualized.
Our patient day 5: High attenuation contrast extravasation on barium gapogram

In this next image from the same study, the upper esophageal pouch appears intact, but it is evident that high-attenuation contrast has extravasated from the distal gastric catheter into the pleural space, indicating leak.
After this, a repeat thoracotomy was performed and the ruptured distal esophageal stump was repaired.
Our patient day 30: Close approximation of proximal and distal esophageal stumps on barium gapogram.

Water soluble barium study highlighting approximation of proximal and distal stumps, no extravasation of contrast.

With the stumps well-approximated, thoracotomy with esophageal anastomosis was next performed.
Our patient post-operative day 1: Passage of NG tube to stomach on supine chest x-ray
Our patient post-operative day 1: Evidence of stricture formation on barium esophagogram
Companion patients 6, 7:

Strictures on barium esophagrams

10-year-old female with chicken stuck in esophagus due to stricture.

30-day-old male with evidence of stricture.
Summary

We have seen:

• Key imaging signs to diagnose EA +/- TEF, prenatally and postnatally
• Imaging as it is used to evaluate gap size in EA (gapograms)
• How the Foker process uses serial gapograms to evaluate progress
• Imaging of complications in EA, including leak and strictures

We have discussed:

• Key considerations in diagnosis and management of long and short gap EA, including safety precautions and pre-operative planning
• Different treatment options exist for long-gap EA
• Complications are common, but rarely severe
Thank you!

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References

References


