

AMSER Case of the Month

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4-year-old male presents with dysphagia

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Patient Presentation

- **HPI:** 4-year-old male with history of esophageal atresia presents with dysphagia and emesis
- **PMHx:** Premature infant of 36 weeks gestation, esophageal atresia, bowel perforation, patent foramen ovale, aspiration pneumonia, chronic difficulty swallowing, G-tube feeds
- **PSHx:** Esophageal atresia repair, gastrostomy tube placement, multiple esophagoscopies
- **FHx:** Mother: allergic rhinitis; Father and paternal grandmother: asthma
- **Allergies:** Chlorhexidine and Tegaderm Ag mesh [silver]
- **Vitals:** BP 96/49, Pulse 89, Temp 98.6 °F, RR 20, SpO2 96%
- **Physical Exam:** Normal physical exam aside from thoracotomy changes

Pertinent Labs

- No pertinent labs

What Imaging Should We Order?

Select the applicable ACR Appropriateness Criteria

Variant 2: **Unexplained oropharyngeal dysphagia. Initial imaging.**

Procedure	Appropriateness Category	Relative Radiation Level
Fluoroscopy biphasic esophagram	Usually Appropriate	☢☢☢
Fluoroscopy barium swallow modified	May Be Appropriate	☢☢☢
Fluoroscopy single contrast esophagram	May Be Appropriate	☢☢☢
Fluoroscopy pharynx dynamic and static imaging	May Be Appropriate (Disagreement)	☢☢☢
Esophageal transit nuclear medicine scan	May Be Appropriate	☢☢☢
CT neck and chest without IV contrast	Usually Not Appropriate	☢☢☢☢☢
CT neck and chest with IV contrast	Usually Not Appropriate	☢☢☢☢☢
CT neck and chest without and with IV contrast	Usually Not Appropriate	☢☢☢☢☢

This imaging modality was ordered by the pediatric surgeon

Findings (unlabeled)



Images obtained were intended to be scout radiographs for the esophagram which was later cancelled as the patient went to surgery instead of completing the esophagram

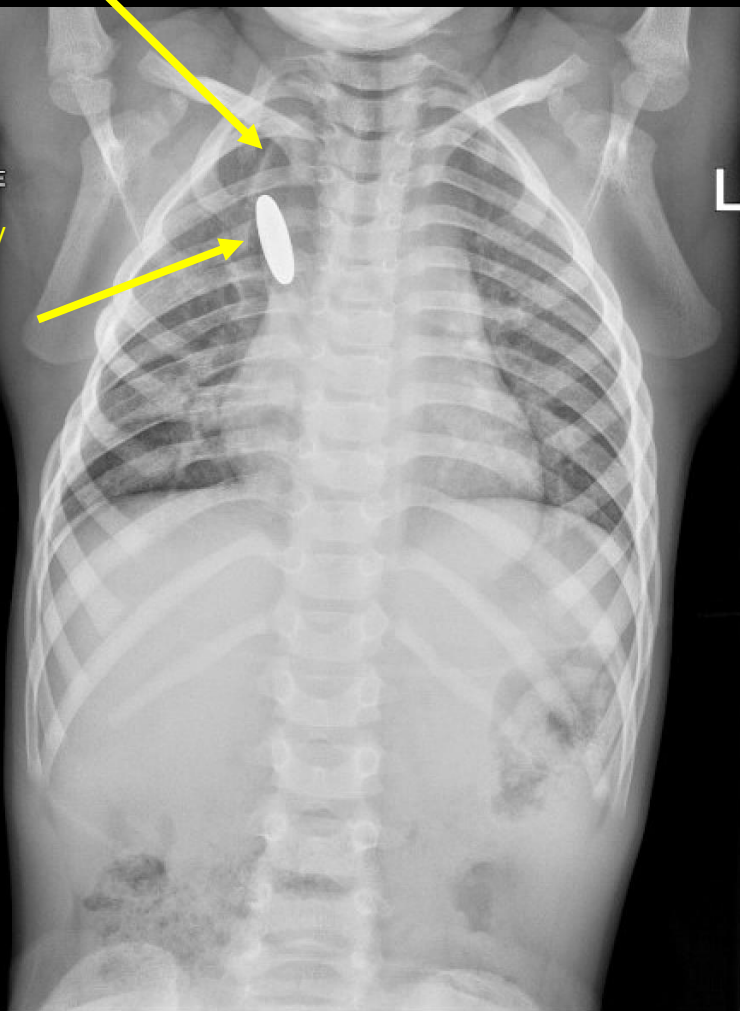
Findings: (labeled)

Dilated proximal esophagus projecting to the right of midline from chronic distention in the setting of prior esophageal atresia post repair

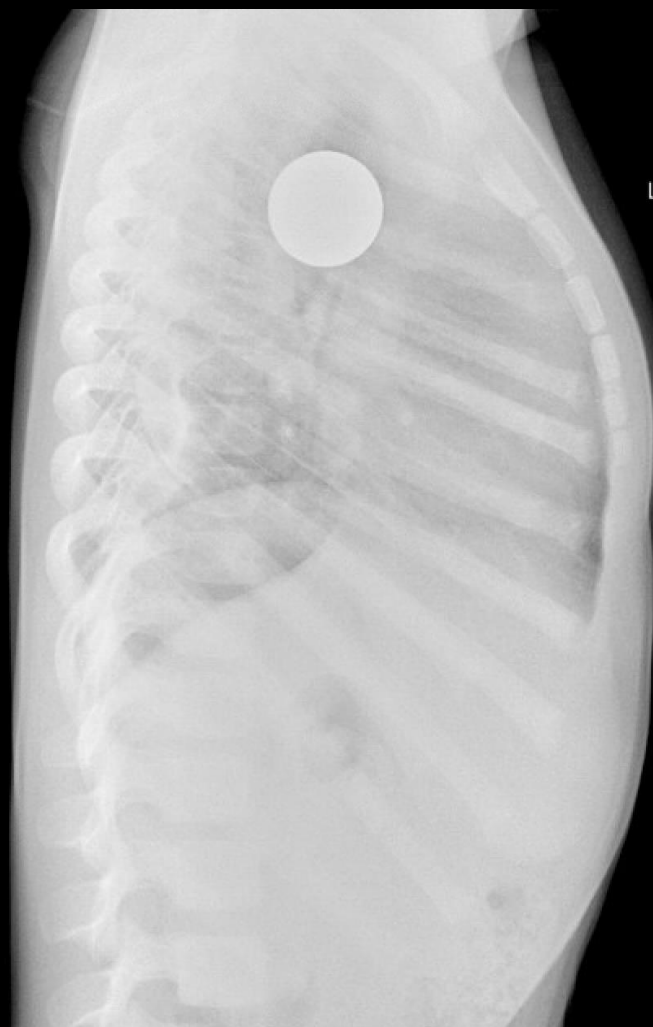
Rounded radiopacity within the right upper chest representing a coin within the patient's proximal esophagus

SUPINE

L



L
LATERAL



Additional findings:

Lungs clear

Cardiomediastinal silhouette is normal in size

No pleural effusion or pneumothorax

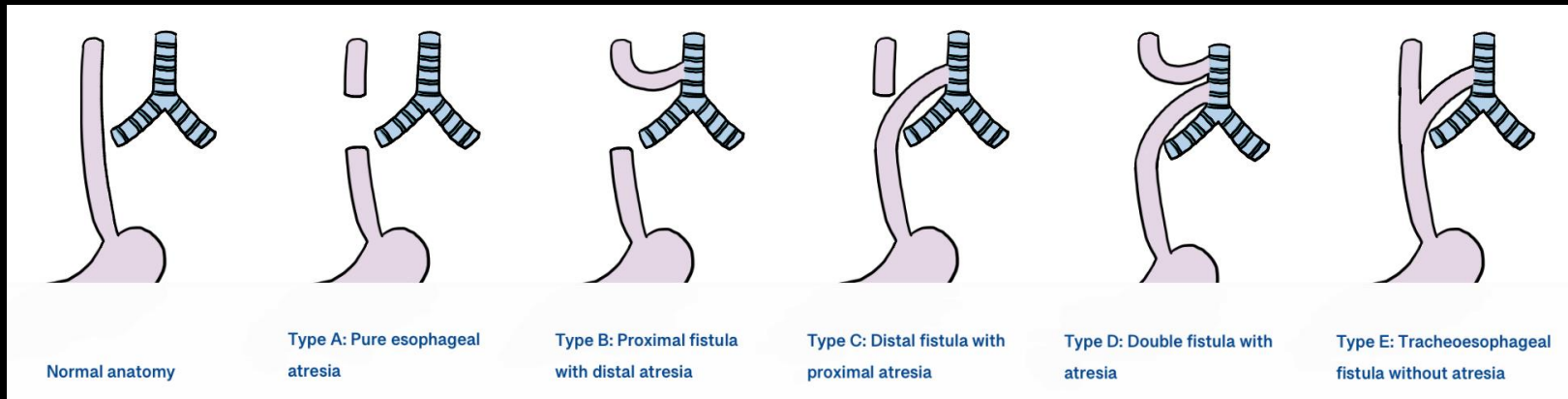
Final Dx:

Type A esophageal atresia status post repair with stricture, now post stricture repair with a retained swallowed coin

Case Discussion

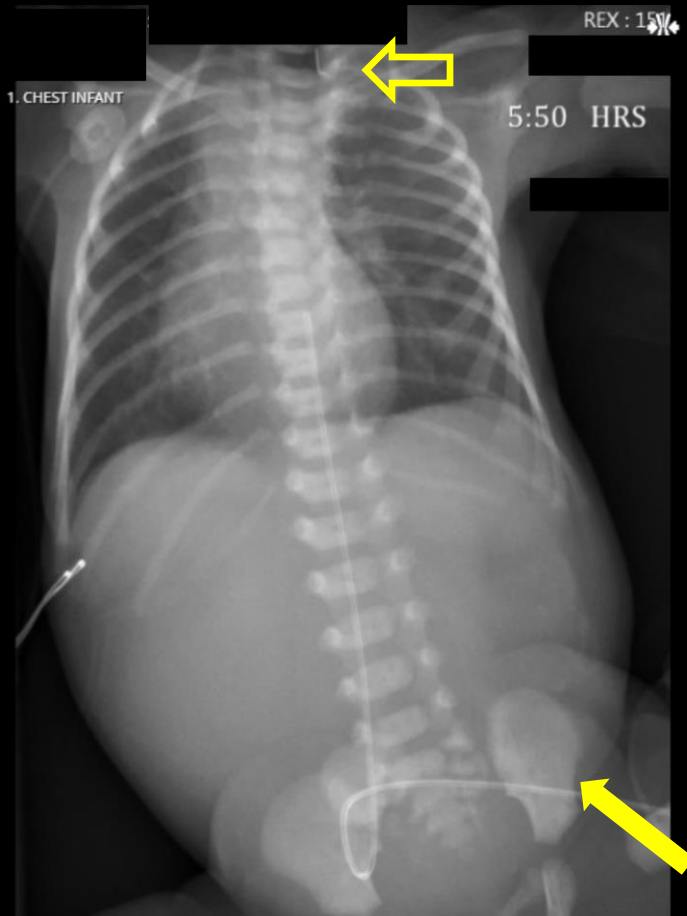
Background

- Esophageal atresia (EA) is a congenital anomaly in which the esophageal lumen is discontinuous, with a separation typically ≥ 3 vertebral bodies¹. It can occur with or without a tracheoesophageal fistula(s) (TEF)²
- Classification of EA (Types A-E) is based on location of the atresia and presence of a tracheoesophageal fistula(s), as depicted in the image below³
- In the United States, EA has a prevalence of about 2.3 per 10,000 live births while it is 1 in 2,500 to 1 in 4,500 worldwide⁴
 - Type C has a prevalence of 84% - *most common*³
 - Type A has a prevalence of 8%³ – *the patient had Type A esophageal atresia at initial surgery*
- Though diagnosis is often made after birth, polyhydramnios on prenatal ultrasound can raise suspicion of EA²
 - Postnatal symptoms that raise clinical suspicion for EA include the inability to handle secretions and respiratory distress, with further confirmation from the inability to insert an orogastric tube into the stomach¹



Adapted from Pinheiro et al.

Case Discussion

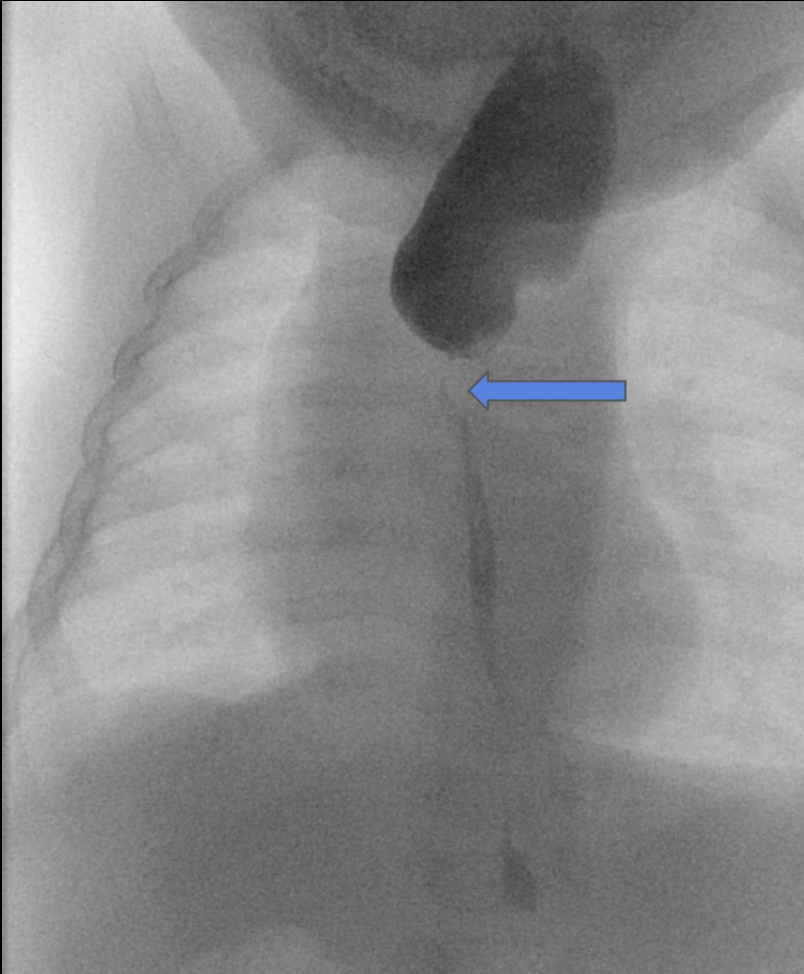


First radiograph done < 24 hours after birth

- The patient's initial chest and abdomen radiograph does not demonstrate bowel gas due to the absence of a distal tracheoesophageal fistula⁴
 - If this were EA type C or E, air from the trachea would enter the stomach through the distal fistula and esophagus
 - TEF increase the risk for aspiration pneumonia due to reflux of gastric contents into the trachea/lungs⁴
- Associated congenital anomalies may be present and are collectively referred to as part of the VACTERL association (Vertebral, Anorectal, Cardiac, Tracheoesophageal, Renal, Limb)¹
 - VACTERL is diagnosed if 2 or more anomalies are present alongside EA²
 - Isolated EA is associated with VACTERL in about 11.5% of patients¹
 - Congenital heart disease is the VACTERL diagnosis that is most frequently associated with EA/TEF⁴

Gasless abdomen compatible with esophageal atresia without a distal tracheoesophageal fistula. Enteric tube tip in proximal esophageal pouch also seen (open arrow)

Case Discussion

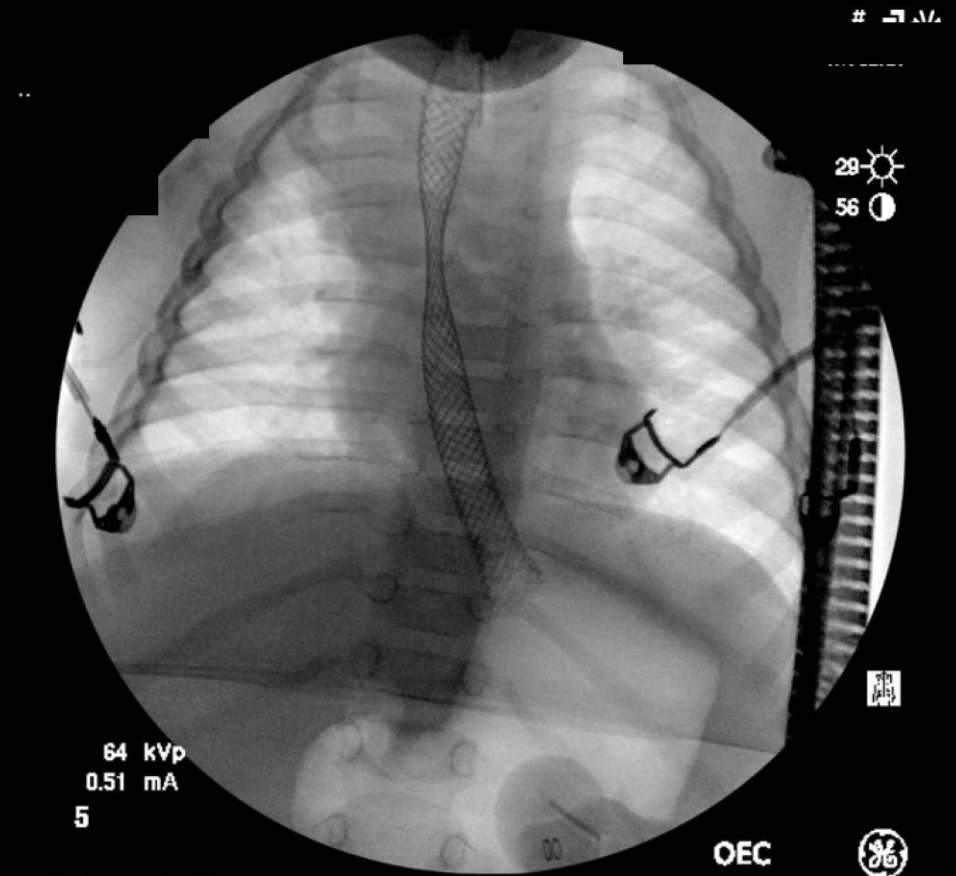


Arrow pointing to the patient's anastomotic stricture

- A post-operative stricture was identified on an esophagram after esophageal atresia repair in 2020 (prior to the new onset dysphagia).
- Stricture at the site of anastomosis can occur after EA +/- TEF repair with symptoms including feeding/swallowing difficulty, drooling, poor weight gain, coughing and aspiration, and is considered the most common complication following surgical repair⁵
- Strictures, either anastomotic, congenital or gastroesophageal reflux (GER) related can lead to dysphagia. Many EA +/- TEF patients also have GER. Anastomotic strictures are most common and are often amenable to balloon dilatation, as compared to congenital strictures related to complete cartilaginous rings (usually more distal).
- Upon the patient's presentation of new onset dysphagia and prior medical history, an esophageal stricture was initially suspected and an esophagram was ordered. However, the study was cancelled as a coin was detected and there was concern for a persistent/recurrent esophageal stricture (no significant stricture seen at the time of coin removal with the patient now post esophagoplasty/stricturoplasty). The retention of the coin was likely related to dysmotility, another common finding in patient's post EA/TEF and repair.

Case Discussion

- Esophageal dilations are the first-line treatment for anastomotic strictures post surgical repair and are primarily used for symptom relief⁵
- The patient's past surgical history involved over 10 dilations performed over a 5-month period, but because treatment was refractory, a decision was made to perform stent placement across the anastomotic stricture (see image)
- The rationale of using esophageal stents after failure of medical and endoscopic treatment is to maintain patency of the esophageal lumen and expand the stricture⁵
- That same year, the patient underwent stent removal with esophagoplasty due to persistent dysphagia despite stent placement (prior to the swallowed coin removal)



References:

1. McGowan NA, Grosel J. An overview of esophageal atresia and tracheoesophageal fistula. *JAAPA*. 2022;35(6):34-37. doi:[10.1097/01.JAA.0000830180.79745.b9](https://doi.org/10.1097/01.JAA.0000830180.79745.b9)
2. Pinheiro PFM. Current knowledge on esophageal atresia. *WJG*. 2012;18(28):3662. doi:[10.3748/wjg.v18.i28.3662](https://doi.org/10.3748/wjg.v18.i28.3662)
3. Singh A, Middlesworth W, Khlevner J. Surveillance in Patients With Esophageal Atresia/Tracheoesophageal Fistula. *Curr Gastroenterol Rep*. 2017;19(1):4. doi:[10.1007/s11894-017-0541-5](https://doi.org/10.1007/s11894-017-0541-5)
4. Baldwin D, Yadav D. Esophageal Atresia - StatPearls - NCBI Bookshelf. NIH. <https://www.ncbi.nlm.nih.gov/books/NBK560848/>
5. Tambucci R, Angelino G, De Angelis P, et al. Anastomotic Strictures after Esophageal Atresia Repair: Incidence, Investigations, and Management, Including Treatment of Refractory and Recurrent Strictures. *Front Pediatr*. 2017;5:120. doi:[10.3389/fped.2017.00120](https://doi.org/10.3389/fped.2017.00120)