American Pediatric Surgical Association

Standardized Toolbox of Education for Pediatric Surgery

Esophageal Atresia and Tracheo-Esophageal Fistula

APSA Committee of Education
2012-13
2018-19

STEPS
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Esophageal atresia and Tracheo-esophageal fistula

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- Pramod S. Puligandla, MD MSc (2019 edits)
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History

- Newborn child, normal vaginal delivery
- Prenatal ultrasound unremarkable
- Now 3 hours old with difficulty feeding

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History Discussion Slide

- **What other points of the history do you want to know?**

- **Characterization of symptoms:** spitting and coughing during attempted breastfeeding
- **Temporal sequence:** immediate with beginning of feeding
- **Alleviating / Exacerbating factors:** appears fine while not feeding; may have excessive secretions
- **Associated signs/symptoms:** otherwise normal appearing child
- **Pertinent PMH:** vaginal delivery
- **Perinatal:** mild polyhydramnios
- **Meds:** none
- **Relevant Family Hx**: none
- **Relevant Social Hx**: none

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Physical Exam

- **What specifically would you look for?**
  - Vital Signs: HR 135bpm; RR 40/min; O_2Sat 97%sat on RA
  - Appearance: Well appearing
  - Relevant exam findings for a problem focused assessment:
    - oral secretions
    - mild upper abdominal distension

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Studies (Labs, Imaging)

- **What labs are needed?**
  - Standard labs (CBC, X-match)

- **What imaging is needed?**
  - Chest and Abdominal Radiograph, after placement of NG tube

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Gross's Anatomical Classification

- Type A:
  - Esophageal atresia without tracheoesophageal fistula. 8%
- Type B:
  - Esophageal atresia with proximal tracheoesophageal fistula. <1%
- Type C:
  - Esophageal atresia with distal tracheoesophageal fistula. 87%
- Type D:
  - Esophageal atresia with proximal and distal fistula. 1%
- Type E:
  - Tracheoesophageal fistula without atresia. 4%
  - H-type fistula

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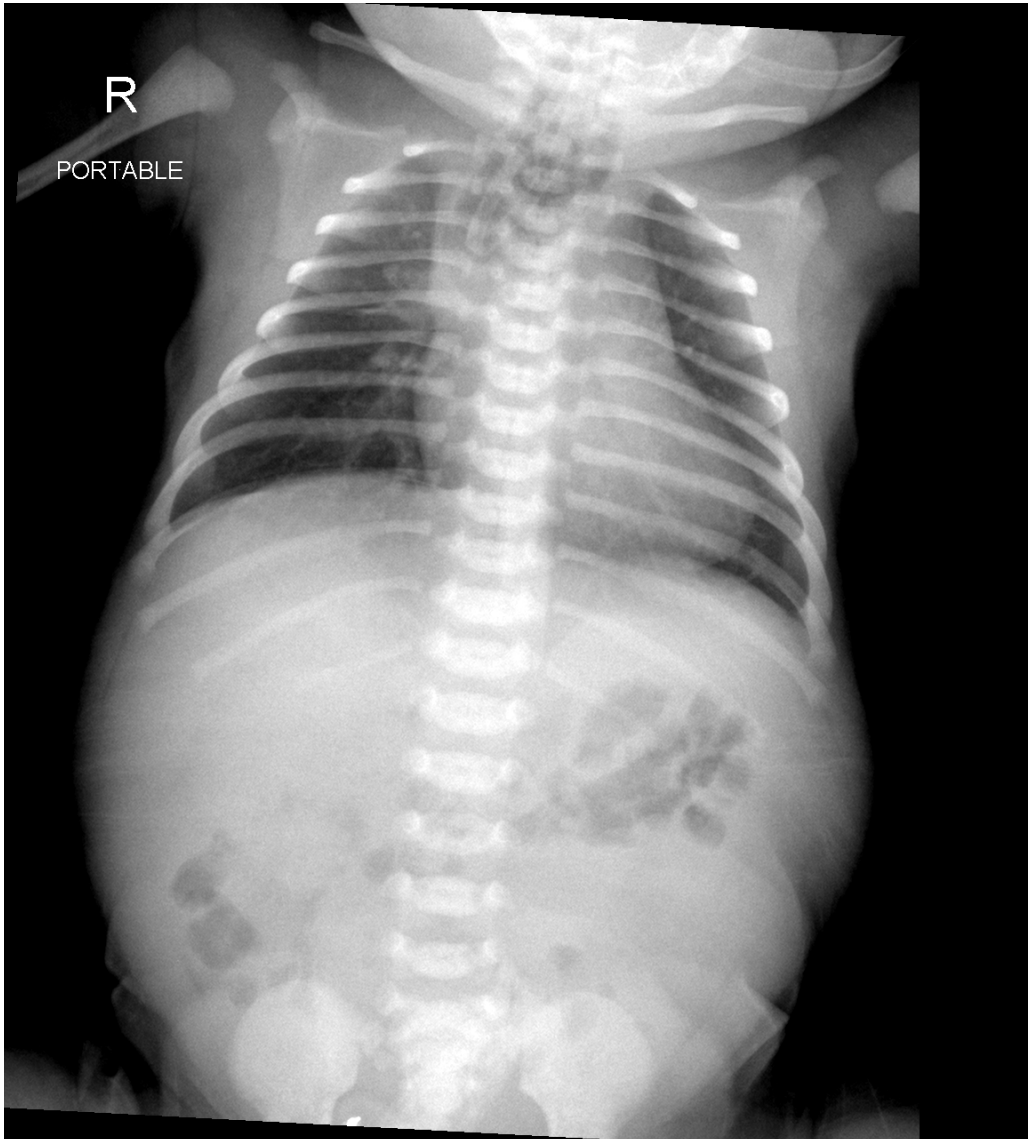
Study Results



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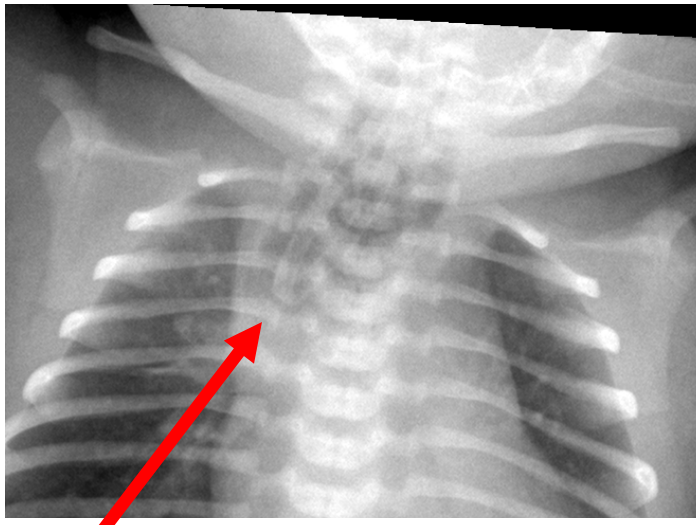
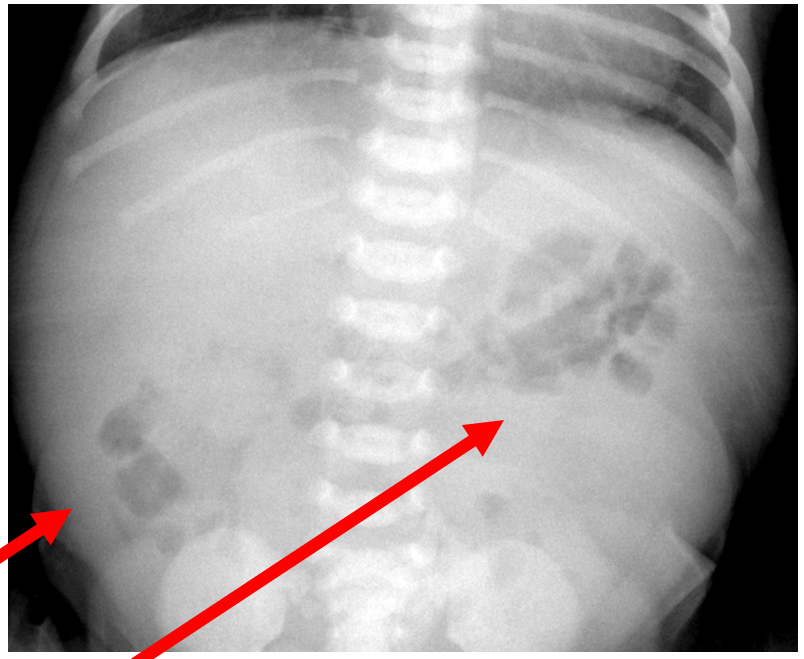


Coiled NG tube
= esophageal atresia



Air in GI tract =
tracheoesophageal fistula

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Case Discussion

- **Diagnosis**
  - Esophageal atresia with tracheo-esophageal fistula
- **Plans**
  - VACTERL work-up (including physical exam)
  - Essential investigations pre-op:
    - Cardiac Echo to r/o major congenital heart disease
    - Ensure infant has urinated (or get US to rule out renal issue)
  - Consent: Rigid bronchoscopy and right thoracotomy, EA/TEF repair
  - Operative: 1: bronchoscopy
    2: ligation and division of TE Fistula
    3: esophageal anastomosis

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VACTERL Complex of Associated Anomalies

- **V**ertebral (tethered spinal cord, bony anomalies)
  - Sacral X-ray, spine ultrasound
- **A**norectal (imperforate anus)
  - Physical exam
- **C**ardiac (ASD, VSD, other structural anomalies)
  - echo
- **T**racheo **E**sophageal Fistula
- **R**enal (hydronephrosis, duplicate/absent kidneys)
  - Renal ultrasound
- **L**imb Anomalies (radial anomalies)
  - Physical exam

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Interval steps before / instead of surgery

- **Head-up position to minimize aspiration with NE tube to suction in upper pouch**
- **Timing of surgery can be quite emergent, as every inspiration may be diverted into the stomach**
  - **Can lead to severe abdominal distension and respiratory compromise** (decreased ventilation and reduced diaphragmatic excursion)
  - **Keep infant breathing spontaneously to reduce ventilation of fistula**

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Operation

- Confirmation of diagnosis via bronchoscopy, opportunity to localize fistula and possibly occlude with balloon catheter
- Right thoracotomy to close/divide fistula
- Mobilization of proximal/distal esophagus and create tension free anastomosis
- Same can be done thoracoscopically

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Complications

- **Intra-operative:**
  - Long gap atresia, (more than 2 vertebral bodies)
    - Can try to mobilize upper and lower pouches to reduce tension on anastomosis
    - If above does not work, then could tack upper and lower pouches to prevertebral fascia and come back to repair in 8-12 weeks
    - Alternative operations: "Foker" technique (not in premature infant)

Complications

- **Peri-operative:**
  - Air leak at tracheal repair site
  - Anastomotic leak of esophagus
  - Anastomotic stricture of esophagus
- **Long Term**
  - Gastroesophageal reflux w/wo stricture formation
  - Tracheomalacia

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Post-operative Management

- **Routine milestones for post op care**
  - Assessment of patency of esophagus with esophagram (post-op day 5-7)
  - Oral feeds
  - Gastroesophageal reflux prophylaxis

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Questions

- **Newborn with flat abdomen, and gasless appearance on abdominal X-ray. Which type of atresia?**
  - A
  - B
  - C
  - D
  - E

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Questions

- **Newborn with flat abdomen, and gasless appearance on abdominal X-ray. Which type of atresia?**
  - A
  - B
  - C
  - D
  - E

Answer: A or B

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Questions

Postoperative day 30 at home, patient after type C EA/TEF repair starts to take very long time to finish her bottle. Why?

A delayed presentation of mediastinitis

B undiagnosed cardiac anomaly

C anastomotic stricture of esophagus

D recurrent tracheo-esophageal fistula

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Questions

Which type of esophageal atresia or tracheo-esophageal fistula is most difficult to diagnose/presents the latest??

- A
- B
- C
- D
- E

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Questions

Which type of esophageal atresia or tracheo-esophageal fistula is most difficult to diagnose/presents the latest??

- A
- B
- C
- D
- **E (H-type fistula without atresia)**

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Final Discussion/Review

- 1:3-4000 births, no risk factors
- distal TE fistula (Type C) is most common
  - 87% proximal atresia with distal fistula
- Respiratory distress can create emergency
- Often associated with tracheomalacia
  - Seal like barking cough
- VACTERL associated anomalies

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Acknowledgement Slide

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