Esophageal Atresia + Tracheoesophageal Fistula

Jason P. Sulkowski MD
Case

- **Perinatal:**
  - Mother: 29y F G3P2
  - Unremarkable pregnancy
  - Scheduled repeat C-section
  - GA: 38 weeks
  - Weight: 4300 g

- **DOL1:**
  - Noted to have excessive drooling and intolerance of feeds
  - OG passed, resistance at 14 cm
  - X-ray:
• **Physical Exam:**
  - **Vitals:** wnl
  - **HEENT:** wnl
  - **CV:** wnl, no murmurs
  - **Pulm:** bilateral rhonchi
  - **Abd:** wnl
  - **Back:** no vertebral defects, normal anus
  - **Ext:** no skeletal defects
**Imaging:**
- Echo: 2 mm ASD
- Renal Sono: negative
- Spine x-ray: negative

**DOL2:**
- Mild respiratory distress
- Open gastrostomy

**DOL8:**
- TEF/EA repair:
• **Post-Op Course:**
  – Additional chest tube placed due to persistent pneumothorax
  – Swallow study:
• Post-Op Course:
  – Additional chest tube placed due to persistent pneumothorax
  – Swallow study:
  – Early difficulty suckling
  – Now tolerating PO
Questions?
History

• William Durston, 1670
  – “A Narrative of a Monstrous Birth in Plymouth...”

• Thomas Gibson, 1697
  – “We blew a pipe down the gullet, but found no passage for the wind into the stomach.”
  – “...searching what way the wind had passed when we blew from the stomach upwards, we found an oval hole on the fore-side of the gullet opening into the [trachea]...”
• **Timothy Holmes, 1869**
  - *Surgical Management of Children’s Diseases*
  - Suggested possibility of esophageal anastomosis
  - “…the attempt ought not, I think, be made.”

• **Charles Steele, 1888**
  - First operative attempt
  - Unsuccessful
• J. Breenemann, 1913
  – “...the physician who decides to let his little patient die undisturbed can amply justify his course.”

• Mims Gage & Alton Oschner, 1936
  – “…the operation would always be finished as a postmortem procedure.”

• Thomas Lanman, 1940
  – “…it is felt that considerable progress along rational lines is being made...successful treatment is only a matter of time.”
• William Ladd, 1940
Cameron Haight, 1941
Embryology

- Abnormal development of TE septum and lateral esophageal groove
Epidemiology

- 1:3500 live births
- 60-70% white
- 50-60% male
- >80% have associated congenital anomalies
  - More common in pure EA
  - Less common in pure TEF (H-type)
Table I. Baseline demographic and clinical characteristics of neonates with EA/TEF overall and grouped by survival to hospital discharge after the first EA/TEF-related procedure*

<table>
<thead>
<tr>
<th></th>
<th>Total cohort (N = 3,479)</th>
<th>Died (n = 189)</th>
<th>Survived (n = 3,290)</th>
<th>P value</th>
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<tbody>
<tr>
<td></td>
<td>n (% or median (IQR))</td>
<td></td>
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<td></td>
</tr>
<tr>
<td>Male</td>
<td>1,934 (55.6)</td>
<td>101 (53.4)</td>
<td>1,833 (55.7)</td>
<td>.54</td>
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<tr>
<td>Race</td>
<td></td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>White</td>
<td>2,373 (68.2)</td>
<td>106 (56.1)</td>
<td>2,267 (68.9)</td>
<td>.0003</td>
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<tr>
<td>Black</td>
<td>327 (9.4)</td>
<td>30 (15.9)</td>
<td>297 (9.0)</td>
<td></td>
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<tr>
<td>Other/unknown</td>
<td>779 (22.4)</td>
<td>53 (28.0)</td>
<td>726 (22.1)</td>
<td></td>
</tr>
<tr>
<td>Birth weight†</td>
<td>2,580 (2,020, 3,050)</td>
<td>1,868 (1,280, 2,370)</td>
<td>2,604 (2,070, 3,075)</td>
<td>&lt;.0001</td>
</tr>
<tr>
<td>Gestational age in weeks‡</td>
<td>37 (35, 39)</td>
<td>34 (32, 36)</td>
<td>37 (35, 39)</td>
<td>&lt;.0001</td>
</tr>
<tr>
<td>Prematurity</td>
<td>1,286 (37.0)</td>
<td>113 (60.0)</td>
<td>1,173 (35.7)</td>
<td>&lt;.0001</td>
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<tr>
<td>Associated congenital conditions (n, %)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Any other congenital anomaly</td>
<td>2,905 (83.5)</td>
<td>183 (96.8)</td>
<td>2,722 (82.7)</td>
<td>&lt;.0001</td>
</tr>
<tr>
<td>Congenital heart disease</td>
<td>2,420 (69.6)</td>
<td>174 (92.1)</td>
<td>2,246 (68.3)</td>
<td>&lt;.0001</td>
</tr>
<tr>
<td>Gastrointestinal anomaly</td>
<td>707 (20.3)</td>
<td>54 (28.6)</td>
<td>653 (19.9)</td>
<td>.004</td>
</tr>
<tr>
<td>Eye anomaly</td>
<td>109 (3.1)</td>
<td>15 (7.9)</td>
<td>94 (2.9)</td>
<td>&lt;.0001</td>
</tr>
<tr>
<td>Coloboma</td>
<td>39 (1.1)</td>
<td>8 (4.2)</td>
<td>31 (0.9)</td>
<td>&lt;.0001</td>
</tr>
<tr>
<td>Head or neck anomaly</td>
<td>27 (0.8)</td>
<td>4 (2.1)</td>
<td>23 (0.7)</td>
<td>.06</td>
</tr>
<tr>
<td>Respiratory anomaly</td>
<td>564 (16.2)</td>
<td>54 (28.6)</td>
<td>510 (15.5)</td>
<td>&lt;.0001</td>
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<tr>
<td>Palate anomaly</td>
<td>82 (2.4)</td>
<td>11 (5.8)</td>
<td>71 (2.2)</td>
<td>.004</td>
</tr>
<tr>
<td>Musculoskeletal anomaly</td>
<td>919 (26.4)</td>
<td>75 (39.7)</td>
<td>844 (25.7)</td>
<td>&lt;.0001</td>
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<tr>
<td>Genetic anomaly</td>
<td>212 (6.1)</td>
<td>37 (19.6)</td>
<td>175 (5.3)</td>
<td>&lt;.0001</td>
</tr>
</tbody>
</table>

Associated Syndromes

• **VACTERL**
  - Vertebral
  - Anorectal
  - Cardiac
  - TEF
  - Esophageal
  - Renal
  - Limbs

• **CHARGE**
  - Coloboma
  - Heart
  - Atresia choanae
  - Retardation
  - Genital hypoplasia
  - Ear deformities

• Down syndrome
• DiGeorge sequence
• Pierre Robin sequence
• Feingold syndrome
• Fanconi Anemia
• Polysplenia sequence
• Holt-Oram syndrome
• Townes-Brock syndrome
• Bartsocas-Papas syndrome
• McKusick-Kaufman syndrome
• Schisis association
Diagnosis

• Prenatal diagnosis rare

• Infants usually symptomatic within hours of birth
  – Excessive drooling
  – Intolerance of feeds
  – Cyanosis (with or without feeds)
  – Respiratory distress
    • Can be clinically significant with distal TEF due to gastric distension

• Inability to pass an OG/NG tube
  – Inject air or dilute barium to visualize the proximal pouch

• H-type diagnosis difficult
  – Often presents with coughing during feeding and frequent pneumonia
  – Bronchoscopy & esophagoscopy to diagnose
Initial Management

- Prevention of aspiration pneumonitis
  - Place Replogle tube to suction in upper pouch
  - Upright positioning

- Antibiotics

- Fluids

- Avoidance of endotracheal intubation
Respiratory Distress

• Occurs in infants with large distal TEF
  – Increasing gastric distension presses cranially on diaphragm
  – Gastric rupture possible

• Endotracheal intubation is often necessary
  – Exacerbates the problem

• Management options:
  – Banding at GE junction
  – Pass ETT beyond fistula
  – Fogarty balloon to occlude fistula
  – Gastrostomy tube
    • May need to pass retrograde Fogarty

  – Best option: emergency thoracotomy and division of TEF
Pre-Operative Bronchoscopy

• Proximal TEF rare (6%)

• Some surgeons routinely perform bronchoscopy before starting EA repair
  – Early identification of proximal TEF may prevent need for re-operation later

• Retrospective review of >100 patients over 20 years at single center
  – 54% of EA without distal TEF had a proximal TEF
  – Some attempt to identify proximal TEF is indicated

Thoracoscopic Approach

- 104 patients from multiple centers
- Description of outcomes
- Results similar to open procedures
  - Compared to previously published studies of open TEF/EA repairs
- Major advantage is prevention of possible chest deformities from thoracotomy
  - Winged scapula
  - Asymmetry of thoracic wall
  - Scoliosis
  - Mammary maldevelopment

Open vs Thoracoscopic

• Meta-analysis of 4 comparative studies

• Results were statistically similar for:
  – Any intraoperative complications
  – Any postoperative complications
  – Anastomotic leaks
  – Anastomotic strictures

• Formal RCT is needed

Long-gap EA

- Distance between two pouches precludes safe anastomosis
  - No specific definition of “long-gap”

- More common in Type A (pure EA)

- Place gastrostomy and wait
  - As infant grows the gap will narrow
Other Approaches

- **Kimura technique**¹
  - Esophagostomy advancement

- **Foker technique**²
  - External traction

Outcomes and Risk Stratification

• 85-95% survival

• Risk stratification
  – Waterston (London, UK)
  – Poenaru (Montreal, Canada)
  – Spitz (London, UK)
  – Teich (Columbus, OH)
  – Okamoto (Kobe, Japan)
# Waterston Classification

<table>
<thead>
<tr>
<th>Class</th>
<th>Description</th>
<th>Survival</th>
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<tbody>
<tr>
<td>A</td>
<td>&gt;2500 g and healthy</td>
<td>100%</td>
</tr>
<tr>
<td>B</td>
<td>2000-2500 g and healthy</td>
<td>85%</td>
</tr>
<tr>
<td></td>
<td>&gt;2500 g with congenital anomalies</td>
<td></td>
</tr>
<tr>
<td>C</td>
<td>&lt;2000 g</td>
<td>65%</td>
</tr>
<tr>
<td></td>
<td>&gt;2000 g with severe cardiac anomalies</td>
<td></td>
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</tbody>
</table>

<table>
<thead>
<tr>
<th>Class</th>
<th>Description</th>
<th>Survival</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>&gt;2000 g</td>
<td>100%</td>
</tr>
<tr>
<td></td>
<td>No major cardiac anomaly</td>
<td></td>
</tr>
<tr>
<td>II</td>
<td>&lt;2000 g</td>
<td>81%</td>
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<tr>
<td></td>
<td>No major cardiac anomaly</td>
<td></td>
</tr>
<tr>
<td>III</td>
<td>&gt;2000 g</td>
<td>72%</td>
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<tr>
<td></td>
<td>Major cardiac anomaly</td>
<td></td>
</tr>
<tr>
<td>IV</td>
<td>&lt;2000 g</td>
<td>27%</td>
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<tr>
<td></td>
<td>Major cardiac anomaly</td>
<td></td>
</tr>
</tbody>
</table>

Predictors of Mortality

• Cohort study, PHIS database
  – 3479 patients at 43 hospitals

• In-hospital mortality: 5.4%

• Predictors:
  – Low birth weight
  – Congenital heart disease
  – Other congenital anomalies
  – Pre-operative mechanical ventilation

• Hospital volume not associated with worse mortality

Complications

• Early
  – Anastomotic leak
  – Stricture
  – Recurrent TEF

• Late
  – GERD
  – Tracheomalacia
Esophageal Leak

- 13-16%
- Managed with drainage & nutrition
- Advantage of extrapleural approach
  - 95% will close spontaneously
- 3-5% will require re-operation
  - Repair
  - Buttress with pericardium, pleura, intercostal muscle
  - Cervical esophagostomy if needed
Stricture

- Up to 80% have stricture requiring dilation

- Frequently present with esophageal foreign body
  - Food bolus

- Most respond to 1-3 episodes of bougienage

- Identify concurrent GERD and treat

- Rarely anastomotic revision is required
Recurrent TEF

- 3-14%
- Usually related to anastomotic leak
- Frequently presents in delayed fashion
  - Presentation mirrors that of Type E
- Open ligation
- Endoscopic options
GERD

- **30-70%**
  - 57% (Sulkowski JP et al.)

- **Management:**
  - Thickening of feeds
  - Prone / upright position
  - Acid reducing agents

- **Leads to worse overall outcomes**
  - Failure to thrive
  - Recurrent pulmonary infections
  - Intractable strictures

- **45-75% undergo antireflux surgery**
Tracheomalacia

- 10-25%

- Weakness causing collapse of airway during expiration
  - “Barking” cough
  - Recurrent infections
  - Apneic spells

- Most improve over time

- Aortopexy for severe symptoms and life-threatening apnea
Esophageal Replacement

• **Indications:**
  - Failure to anastomose long-gap
  - Leak / stricture not amenable to revision of anastomosis

• **Options:**
  - Colon
    • Right
    • Transverse
    • Left
  - Stomach
    • Pull-up
    • Reversed gastric tube
  - Small bowel
    • Interposition
    • Free
Excessive drooling and mild respiratory distress are seen 8 hours after birth. An abdominal x-ray shows complete lack of air in the GI tract. What is the most likely diagnosis?

- A. Hirshsprung’s disease
- B. Tracheoesophageal fistula, H-type
- C. Pyloric atresia
- D. Esophageal atresia without tracheoesophageal fistula
- E. Esophageal atresia with tracheoesophageal fistula
Question 2

• Repair of esophageal atresia and tracheoesophageal fistula can be complicated by which of the following problems?

  – A. Esophageal stricture
  – B. Anastomotic leakage
  – C. GERD
  – D. Recurrent fistula
  – E. All of the above
• Thank you
  – Francesca Velcek MD
  – Yuko Shimotake MD
  – Christopher Turner MD

• Main source: