Repaired Esophageal Atresia and Tracheoesophageal Fistula and Chronic Dysphagia

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Disclosures

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Case Presentation

• Infant with VACTERL
  • EA/ TEF (Type C)
  • Aspiration
  • Airway abnormalities
    • Type 2 Laryngeal Cleft (LTC)
    • Subglottic Stenosis
    • Tetralogy of Fallot (TOF)
    • Tethered Cord
• Early Interventions:
  • TEF Repair (DOL 1)
  • TOF Repair (4 months)
  • Endoscopic LTC repair (6 months)

12 months: Complicated admission for aspiration following diagnostic rigid bronchoscopy
• Tracheostomy
• G-tube / fundoplication
Subsequent admissions for aspiration and respiratory symptoms:
• 14 months
• 15 months (PICU)
• 18 months

Case Presentation

• Feeding history:
  • NPO since g-tube / fundoplication
  • Recurrent events of coughing-vomiting-aspiration
• Diagnostic questions for GI consult service:
  • Aspiration events despite fundoplication?
  • Does the child need fundoplication revision?
  • Is the family continuing to feed him orally?
  • How to intervene?

Dependent consolidation but no bronchiectasis
Objectives

- EA/TEF: Background
- Key GI/Esophageal Issues:
  - GERD
  - Esophageal motility
  - Dysphagia
- Key Airway/Pulmonary Issues:
  - CHCO Aerodigestive Program experience
  - What’s next for EA/TEF care?

Background: EA/TEF

- Common congenital disorder:
  - 1:2,500
  - 25% of patients: other congenital anomalies

Pathophysiology in EA / TEF

- Esophageal:
  - Anatomic
  - Motility
  - GERD
- Feeding Dysfunction:
  - Anatomic
  - Esophageal
  - Syndromic / Developmental

Outcomes:
- Pulmonary Symptoms
- Feeding Dysfunction
- Esophageal Pathology
- Acute events:
  - Hospitalizations, ALTE
- Interventions
  - Quality of Life

Importance of Multispecialty Care

- Published outcomes are largely retrospective, single center, heterogeneous population
- Authors and editors historically have single specialty focus
- No consensus guidelines on management, surveillance, and follow up

GERD in EA/TEF

- Incidence: Common (27% - 87%)
- Sequelae:
  - Esophageal:
    - Surgical: anastomotic stricture, leak, or recurrent fistula
    - Esophagitis / Peptic Stricture
    - Feeding difficulties / Failure to Thrive
    - Intestinal metaplasia (Barrett’s esophagus): estimated 15% of patients
      - 50 fold higher risk of esophageal carcinoma
      - May be isolated to risk in adults
  - Respiratory:
    - Aspiration events
    - ALTE/apnea
- Treatment:
  - Acid Suppression
  - Surgical

Long Term Esophageal Symptoms and Findings

- Table 3: Esophagitis clinical, radiologic and endoscopic data
- Cartabuke: Gastroenterology Report 2015

Kovesi: Chest 2004
Levesque: JPEN 2011
Cartabuke: Gastroenterology Report 2015
Shawyer: Ped Surg International 2014
Burjonrappa Eur J Ped Surg 2011
Questions: Management and Surveillance of GERD in EA/TEF?

• When to initiate and how long to continue acid suppression?
• When to consider anti-reflux surgery, and how best to perform this?
• When to survey for long term esophageal injury?

Implications of Long Term PPI Use

• In general, PPI use is safe and well tolerated
• Spectrum of long term physiologic alterations:
  • Gut microbiome:
    • Small bowel bacterial overgrowth
    • C. Diff infection
    • Alteration of respiratory microbiome
    • Pneumonia
    • Traveler’s Diarrhea
  • Metabolic consequences:
    • Hypomagnesemia
    • Iron absorption
    • Calcium absorption
    • Bone demineralization
    • B12
  • Cardiovascular / pharmacologic:
    • Methotrexate, Plavix

Anti-reflux surgery in EA/TEF

• Should be considered:
  • Pure EA
  • Long gap disease
  • Recurrent stricture
  • Medically refractory or severe complications of GERD
• Surgical technique:
  • Partial wrap
  • "loose" wrap
• Failure rates: 25-40%

Role of GI Endoscopy in EA/TEF

• Evaluate for and dilate anastomotic stricture
• Evaluate for complications of GERD:
  • Esophagitis
  • Stricture
  • Barrett’s
• Evaluate for non-GERD esophagitis (EoE)
Evaluation of Feeding Difficulties

- Esophagram
- Evaluation of Feeding:
  - Fluoroscopic
  - FEES
- Endoscopy
- Esophageal Motility Study

Case Presentation contd.

- Initial focus on reflux
- Diagnostic questions for ENT/Pulm consult service:
  - How to prevent pneumonia / what is the right sick plan?
  - What daily medical/treatment options will benefit him?
  - What is the path toward trach decannulation?
  - Where are the sites of airway obstruction?

Upper Airway Anomalies in Congenital Tracheoesophageal Fistula and Esophageal Atresia Patients

- 430 patients with EA/TEF at Boston Children’s Hospital
- 139 saw ENT for upper airway evaluation during the study period 2008-2013
Key Airway/Pulmonary Issues

- 20-40% have recurrent pneumonia
- 14-75% bronchitis
- 75% cough
- 75% wheeze
- 10-20% ALTEs/choking/gagging
- 1-22% bronchiectasis
- 70%-90% of adults have low lung function
  - 50:50 restrictive vs. obstructive

Pretty common
Almost universally present
Serious complications in 20%

Questions: Management and Surveillance of Lungs in EA/TEF?

- How to monitor lung disease?
  - Flexible bronchoscopy
  - Imaging to evaluate for bronchiectasis
  - Lung function testing
- How to prevent chronic lung disease and lung function decline?
  - Airway clearance regimen
  - Inhaled steroids
  - Antibiotics for chronic bacterial bronchitis
  - Surgical treatments for tracheomalacia

Questions: Management of Tracheomalacia?

- Type C EA/TEF is the most common cause of congenital tracheomalacia
- Bronchoscopy: gold standard diagnosis
  - Definition of severity is not agreed upon
- Medical Treatments
  - Cholinergic agonist: bethanechol
  - Muscarinic M2 blockade: ipratropium
- Other Treatments
  - Positive airway pressure
  - Invasive or non-invasive
  - Surgical options
  - Watch and wait

Serious complications in 20%

Surgical Options for TEF

- Tracheopexy and Aortopexy (or Innominate Arteriopexy)
- 3D printed airway stents

TEF/EA Experience Within Our Aerodigestive Program

Multidisciplinary Care of Children With Repaired Esophageal Atresia and Tracheoesophageal Fistula

DeBoer, Emily, MD
Repaired Esophageal Atresia and Tracheoesophageal Fistula and Chronic Dysphagia
The challenge – get the patients into our clinics
These children are symptomatic!

Similar incidence from previous studies

Reinforces need for endoscopy by GI, ENT, and Pulmonary:

Dysphagia in EA/TEF

- Esophageal dysphagia has been clearly established
- Pharyngeal dysphagia reports are conflicting
  - 44 children with EA/TEF
  - 19 (43%) had pharyngeal dysphagia
  - 11 (25%) needed a modified liquid diet
Feeding Characteristics by Age

<table>
<thead>
<tr>
<th>Oral motor problems</th>
<th>Need for modified diet</th>
<th>Complain of food stuck</th>
<th>Behavioral feeding problems</th>
<th>Slow pace of eating</th>
</tr>
</thead>
<tbody>
<tr>
<td>0%</td>
<td>0-6 months 10 (22.7%)</td>
<td>0-6-12 months 5 (11.4%)</td>
<td>0-6-12 months 5 (11.4%)</td>
<td>0-6-12 months 5 (11.4%)</td>
</tr>
<tr>
<td>50%</td>
<td>6-12 months 8 (10.2%)</td>
<td>6-12 months 5 (11.4%)</td>
<td>6-12 months 5 (11.4%)</td>
<td>6-12 months 5 (11.4%)</td>
</tr>
<tr>
<td>100%</td>
<td>12-24 months 8 (10.2%)</td>
<td>12-24 months 5 (11.4%)</td>
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Slide courtesy Jenny Maybee and Jackie Deck

Case Review – Revisited Aerodigestive Program Interventions

- GI:
  - Conversion to post-gastric feeds
  - Serial esophageal dilations
- Feeding:
  - Regular feeding therapy
- Pulmonary:
  - Aggressive airway clearance
  - Antibiotic regimen
- ENT:
  - Tonsillectomy and adenoidectomy
  - Airway reconstruction

Case Review: Outcome

- 2 years later:
  - Trach removed
  - G-tube removed
  - Remains on acid suppression?
  - Pulmonary health improved
  - No hospitalizations or procedures under GA in > 1 year

Future Directions

- Are there “best practice” recommendations that apply to the aerodigestive / multidisciplinary management of this population?
- How do we manage these patients?
- What outcomes do we follow?

Proposed Management Recommendations

1. Early Referral to Aerodigestive Program or Equivalent:
   - Communication and collaboration with Pediatric Surgery and Primary Care
   - First evaluation between 6 – 8 months
   - Subsequent visit: 11-14 months
   - Follow up visits: annually, with interim visits as needed
2. Role and timing of imaging and diagnostic studies:
   - Esophagram / Upper GI series
   - Instrumental Evaluation of Swallowing
   - Chest imaging
   - Pulmonary function
3. Endoscopic / Procedural evaluation and intervention
   - Timing may be influenced by need for esophageal dilation in infants and younger children
   - In older children – recommend screening “triple scopes” if not previously done. Timing dictated by severity of symptoms.
   - Long term endoscopic surveillance for esophageal mucosal injury?
4. Feeding:
   - Standardized protocol for advancement of solids in infants and toddlers
   - Proactive education to families about dysphagia, strictures, and feeding dysfunction
5. GERD Management:
   - Early initiation of acid suppression, and continued follow up and discussion regarding risks / benefits of long term treatment
6. Pulmonary Management:
   - Routine airway clearance
   - Education and initiation of pulmonary sick plan
   - Routine influenza vaccination (? Quality benchmark)
   - Role of palivizumab?
7. Ongoing Multidisciplinary (Aerodigestive) Oversight of child’s medical care
Future Directions: Research and Advocacy

- In the works
  - International GI Guidelines to guide care in EA/TEF
  - International Guidelines to guide respiratory care in EA/TEF
- Multisite coordination of longitudinal data and a research database
  - Demographics
  - Symptoms
  - Diagnostics
  - Medical, Surgical, and Therapeutic Management
  - Developmental Outcomes
- Fellowship training, mentorship, and research in this population

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