A Collaborative Approach to Caring for the Patient with Complex Esophageal Disorders

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Disclosure Information
Speaker: Marilyn Stoops

I have no disclosures.

Objectives

• Discuss the benefits of a multidisciplinary approach to patients with complex esophageal conditions.
• Describe the unique challenges of treating patients with complicated esophageal disorders.
• List the long term issues facing patients with complex esophageal disorders.
TEF/EA

- Incidence
  - 1: 2400-4000 live births
- Associated anomalies occur in up to 50% of patients
- VACTERLY, CHARGE association
- Syndromes: Pierre Robin, DiGeorge

Gross Classification of TEF/EA

- Type A 3-5%
- Type B 2%
- Type C 85%
- Type D 3-5%
- Type E 3-6%

‘Long-Gap’ Esophageal Atresia

- Identify anatomy
- Distance of gap
- Spontaneous growth and hypertrophy of esophageal segments occur at a rate faster than overall somatic growth in the absence of mechanical stretching
- Maximal growth occurs during the 1st 8-12 weeks
Complications Following TEF/EA Repair

• Anastomotic leak: 15-20%
  – Conservative management, rarely reoperation

• Anastomotic stricture: up to 40%
  – Anastomosis under tension
  – History of anastomotic leak
  – Ischemic injury
  – GER

Complications following TEF/EA Repair

• Dysphagia
  – Esophageal peristalsis (manometry) is abnormal in up to 75% of children and adults with history of TEF/EA
  – TEF/EA disrupts normal development of myenteric plexus disordered peristalsis and impaired lower esophageal sphincter

Complications following TEF/EA repair

• GER
  – Incidence is 40-50% and as high as 75%
  – Due to shortening of intra-abdominal esophagus during repair
  – Distortion of Angle of His
  – Abnormal LES due to developmental neuromuscular disturbance
• Contributes to stricture formation
Airway Issues

- Recurrent TEF
- Tracheomalacia (10-20%)
- Vocal cord paralysis (congenital vs. acquired)
- Laryngeal Cleft

Pulmonary Issues

- Pneumonia
  - Recurrent episodes can result in long term bronchiectasis and decreased pulmonary function
  - Can be result of GER, dysmotility, stricture
- Reactive Airway Disease
  - Airway hyperactivity may be due to events in early childhood such as aspiration

Aerodigestive and Esophageal Center

- Mission
  - Provision of interdisciplinary care for patients with complex disorders of the airway and digestive tract
- Goal
  - Improve clinical outcomes for patients by coordinating diagnostic and operative procedures and implementing an integrated treatment plan
Aerodigestive and Esophageal Center

- Pediatric Surgery
- Gastroenterology
- Otolaryngology
- Pulmonary
- Interdisciplinary Feeding Team

Aerodigestive and Esophageal Center

- Additional Disciplines
  - Anesthesia
  - Genetics
  - Cardiology
  - Radiology
  - Speech Pathology
  - OT/PT
  - Social Services
  - Dieticians
  - Other services as needed

Aerodigestive and Esophageal Center

- Referral into the ADSC – issues span the scope of multiple disciplines
- Received by the referral management coordinator
  - Assignment to "lead service" (APN and MD) for management based on pre-determined criteria
- Obtain medical records
- Intake Process
  - Completed by lead service APN
  - Tentative plan for assessment
ADEC Intake

- CCHMC ADEC intake form completed by APN with parent
- Intake is a synopsis of history, evaluates primary problems and concerns, additional factors
  - Identification of care providers locally**
- Tentative plan for assessment is included
- This information is then summarized and presented at Wednesday meeting

ADEC Intake

- Reason for referral
- Current meds & allergies
- Review of past medical history
- Review of systems: HEENT, speech & language, pulmonary, cardiovascular, gastrointestinal, feeding, endocrine, musculoskeletal, neurologic, hematologic
- Review of prior tests & procedures, nursing care, equipment, special needs

“The Wednesday Meeting”

- ADEC Grand Rounds-patients presented by lead service APN
  - Patients going to the OR today
  - New patients
    - Tentative plan for evaluation discussed with input from all disciplines, and adjustments made as necessary
  - Patients seen in last week
    - Findings discussed, plan for next visit
Aerodigestive and Esophageal Center

• Lead service APN
  – Serves as primary contact for family, even if family’s questions deal with another discipline within ADEC
  – Contacts family with initial plan, results from visit week earlier, plans for additional visit
  – Enters all orders
  – Final review of ADEC report along with ADEC primary physician

Aerodigestive and Esophageal Center

• ADEC scheduling
  – Appointments coordinated usually during a 3-5 day span during same week
  – Patient evaluated by multiple divisions, imaging studies as needed
  – Coordination of appointments is key
  – Patient itinerary developed and sent to family

Multidisciplinary evaluation in the OR

• Patient taken to OR with all involved disciplines present (ENT, pulm, GI, ped surg)
  – Esophagoscopy
  – Flexible bronchoscopy
  – MLB (rigid bronchoscopy)
Multidisciplinary evaluation in OR

• After 'triple scope', all disciplines meet together with family in consultation room
• Initial findings and impressions are discussed and treatment plan developed
• Final discussion of findings and plan done at next Wednesday meeting, after all results have been reported

Aerodigestive and Esophageal Center

• Clearly defined roles and expectations of all members of team
• Clearly defined lines of communication
• Clearly defined leadership
• Shared vision among all involved
• Institutional support
  – Significant amount of administrative support for complex program

Aerodigestive and Esophageal Center

• Institutional support
  – Sufficient administrative support is key
    • Scheduling- all diagnostic procedures, clinic appointments, operating room procedures
    • Insurance/pre-certing
    • Documents (itinerary, organization for Wednesday meeting, ADRC reports)
    • Communication with families regarding scheduling/Insurance
  – 2 operating rooms dedicated ADEC each Wednesday
Our Experience

June 2011-December 2013
41 patients evaluated

<table>
<thead>
<tr>
<th>Referral Diagnosis</th>
<th>Number of patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Prior TEF/EA Repair</td>
<td>20</td>
</tr>
<tr>
<td>Acquired esophageal conditions</td>
<td>9</td>
</tr>
<tr>
<td>Primary repair of TEF/EA</td>
<td>8</td>
</tr>
<tr>
<td>Other (infectious, duplication, congenital stricture)</td>
<td>4</td>
</tr>
</tbody>
</table>

Prior TEF/EA repair

<table>
<thead>
<tr>
<th>Referral Diagnosis</th>
<th>Number of patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Stricture/pneumonia</td>
<td>5</td>
</tr>
<tr>
<td>Recurrent pneumonia</td>
<td>5</td>
</tr>
<tr>
<td>Dysphagia/pneumonia</td>
<td>4</td>
</tr>
<tr>
<td>Esophagus not in continuity</td>
<td>2</td>
</tr>
<tr>
<td>GER</td>
<td>1</td>
</tr>
<tr>
<td>GER with esoph diverticulum</td>
<td>1</td>
</tr>
<tr>
<td>Esophageal stricture</td>
<td>2 total=20</td>
</tr>
</tbody>
</table>

Stricture/Pneumonia

- 3 out of 5 patients had prior esophageal perforation following dilation
- Significant bronchiectasis from aspiration
## Recurrent Pneumonia

- 4 out of 5 patients had airway anomalies
  - 2 deep intra-arytenoid notches (laryngeal cleft)
  - 1 previously unknown unilateral vocal cord paralysis
  - 1 recurrent TEF

## Acquired Esophageal Conditions

- Caustic Ingestion: 6 patients
- Damage from foreign body ingestion: 3 patients
- Normal esophageal mucosa

## Caustic Ingestion

<table>
<thead>
<tr>
<th>2 days post injury</th>
<th>4 mos post injury</th>
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</thead>
<tbody>
<tr>
<td><img src="image1" alt="Image" /></td>
<td><img src="image2" alt="Image" /></td>
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</tbody>
</table>
Esophageal Replacement

“Every effort should be made to conserve the native esophagus, as no other conduit can replace its function in transporting food from the oral cavity to the stomach satisfactorily.”
Meyers, 1974

Esophageal Replacement

• Decision based after interdisciplinary evaluation, every other treatment has been tried and failed
  – Repeated dilations, steroid injections, stents
• Significant pulmonary disease often drives decision
  – Unresponsive to CPAP (if trached), airway clearance, vest therapy

Esophageal replacement
What to use?

• Colon interposition
  – Long-term, more GI issues
• Gastric pull-up
  – Long-term, more respiratory issues
• Jejunal interposition
  – Very few studies with small number of patients
Esophageal Replacement

**What to use?**

- Prior surgical history of patient
- Other concerns (pulmonary disease)
- Preferred technique of surgeon

- Nothing works as well as native esophagus

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Esophageal Replacement

- Colon interpositions as replacements

- Preoperative evaluation
  - Barium enema
  - FEES (fiber optic endoscopic evaluation of swallowing) to look at vocal cords

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Esophageal Replacement

- Ascending or descending colon
  - Decision in OR based on evaluation of blood supply to segment

- Pyloroplasty
- Avoidance of vasopressor drugs post op
- UGI POD #7
- Enteral feeds post op (G tube vs J tube)
Esophageal Replacement: Coordination in OR with ENT

- Especially helpful in caustic ingestion or in patients with prior procedures
- Will use information from FEES to decide operative approach
- Decrease in OR time

Esophageal Replacement
June 2011-December 2013

- 13 colon interpositions
- 12/13 patients NPO at time of initial evaluation
- 5/13 patients underwent complex airway reconstruction due to congenital or acquired condition of esophagus/airway

Colon interposition

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>Number of patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Caustic ingestion</td>
<td>5</td>
</tr>
<tr>
<td>Stricture resulting in pulmonary complications</td>
<td>3</td>
</tr>
<tr>
<td>Esophagus not in continuity (prior repair)</td>
<td>2</td>
</tr>
<tr>
<td>Long-gap esophageal atresia (primary repair)</td>
<td>2</td>
</tr>
<tr>
<td>Injury from foreign body</td>
<td>1</td>
</tr>
</tbody>
</table>
Caustic Ingestion

- Esophagectomy due to increased risk of esophageal carcinoma
- Occurs in 2-30% of patients
- Develops 1 to 3 decades after ingestion
- Bypassing esophagus does not prevent the development of cancer

Esophageal Replacement Outcomes

<table>
<thead>
<tr>
<th>Complications</th>
<th>Number of patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Anastomotic leak (resolved with conservative management)</td>
<td>2/13</td>
</tr>
<tr>
<td>Stricture</td>
<td>8/13</td>
</tr>
<tr>
<td>Loss of graft</td>
<td>1/13</td>
</tr>
<tr>
<td>Re-operation (resection of stricture)</td>
<td>1/13</td>
</tr>
</tbody>
</table>

Esophageal Replacement Strictures

- Dilations
  - < 10: 4 patients
  - > 10: 4 patients

- Of the 4 patients requiring >10 dilations, 3 had history of caustic ingestion as indication for colon interposition
Outcomes

<table>
<thead>
<tr>
<th>Progress towards PO</th>
<th>Number of patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>All PO</td>
<td>2</td>
</tr>
<tr>
<td>PO with supplemental enteral feeds</td>
<td>6</td>
</tr>
<tr>
<td>Limited PO</td>
<td>2</td>
</tr>
<tr>
<td>Nothing by mouth (graft loss, nerve damage from caustic ingestion, severe cleft lip/palate)</td>
<td>3</td>
</tr>
</tbody>
</table>

Lessons Learned

• Evaluation of vocal cord function prior to esophageal replacement

• Difficult strictures after interposition in patients with history of caustic ingestion
  – Anastomosis very proximal in neck
  – Proximal esophagus that looks ‘normal’ may be damaged on cellular level—affects healing

• Pulmonary status important in determination of whether native esophagus can be salvaged in patient with history of prior TEF/EA repair

• In patients who have had TEF/EA repair and present with recurrent pneumonia without obvious etiology, look for airway anomalies
Moving Forward

- Long-term follow-up
  - Potential for redundany of colon
  - GER and possible aspiration
  - Monitor and minimize respiratory complications
  - Continue to work to advance patients to PO feeds
  - Growth and development
  - Quality of life
- Continue to enhance collaborative care

Case Study

- 2 ½ year old male s/p foreign body ingestion
- TEF, esophageal stricture and stent
- Multiple attempts at repair, multiple thoracotomies and neck explorations over 18 month period
- Esophageal stricture, pooling of secretions and aspiration resulting in recurrent pneumonia
- Trached, vent dependent at night, G-tube fed with gagging/retching, increases saliva, drooling and need for suction, vocal cord paralysis

Initial evaluation

High resolution chest CT
UGI
Clinic appointments
MLB
Flex bronch
EGD (GJ tube placement)
FEES
Findings

- Aspiration and bronchiectasis. Also upper airway issues
- Esophagus- very proximal pinhole stricture, remainder of esophagus viewed retrograde was friable and narrow with diverticular out pouching just distal to stricture.
- GJ tube for jejunal feeds to help decrease gagging/retching and amount of oral secretions
- Confirmed vocal cord paralysis with impaired secretion management

Recommendations

Due to significant pulmonary disease, esophageal stricture unresponsive to conservative management and status of native esophagus:
- Esophageal replacement
- Weaning of ventilator after esophageal status optimized
- Airway reconstruction (glottic airway)
- Decannulation

Current status

Esophageal stricture (<10 dilations)
Passed sleep study, now off ventilator
Flex bronch- airways clean, no evidence of inflammation or aspiration
Repeat FEES—safe for PO
Tolerating PO (all types of food), night feeds
Airway reconstruction scheduled
Questions

References


