Esophageal Atresia Treatment, Pre- and Postoperative Considerations

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Definition

- Esophageal atresia (EA)
  - Congenital defect leading to interrupted esophagus
  - Several types
  - Most infants with EA have another defect called tracheoesophageal fistula (TEF)

- In clinical practice, the term “TEF” often refers to EA with (type C) TEF
Classification

- More than 10 classifications
- Gross classification (1953) most popular in U.S.
Anatomy

A 7.8%  
B 4-5%  
C 85.8%  
D 1.4%  
E 4.2%
Embryology / Etiology

• Unclear
• Disturbance of proximal foregut (mouth to duodenum) separation into
  • respiratory (ventral)
  • and gastrointestinal (dorsal) tubes
• > 50% of EA cases are associated with other malformations
  • VACTERL
• 10% are found in specific chromosomal disorders
  • DiGeorge syndrome
  • Trisomies (13, 21, 18)
  • Opitz syndrome
  • Single gene disorders: Feingold, CHARGE
Embryology

- Foregut
- Neural folds beginning to fuse
- Oropharyngeal membrane
- Heart
- Pericardial coelom
- Hindgut
- Amnion (cut)
- Allantois
- Body stalk

Anterior intestinal portal
Midgut Yolk sac
Posterior intestinal portal

18 days
Embryology

• 4 weeks gestational age
  • Invagination of endodermal tube (gut tube)
  • Development of respiratory diverticulum (ventral to gut)
  • Bifurcation of respiratory diverticulum into two branches (lung buds)
Incidence

- Incidence 1:3000-4500 births
- More common in males
- Male:Female ratio depends on type of EA
  - type E: 2.92
  - type C: 1.44
  - type A: 1.33
- More prevalent in Caucasians (>60%)
- Other risk factors: first pregnancy, advanced maternal age, IVF
Classification
Associated Anomalies

• Presentation dependent on type and associated malformations (overall 50-70%)
• More common in EA without TEF
• Associated malformations
  – Cardiovascular: ~24%
  – Genitourinary: ~21%
  – Gastrointestinal: ~21%
  – Musculoskeletal: ~14%
  – Central nervous system: ~7%
  – Overall incidence: 50-70%
**Associated Anomalies**

- VACTERL (approx. 20% of all EA with TEF)
- 3+ of the following symptoms:
  - Vertebral: 17%
  - Anorectal: 12%
  - Cardiovascular: 20%
  - Tracheal (fistula)
  - Esophageal (atresia)
  - Renal: 16%
  - Limb: 10%
Diagnosis

Prenatal

- Frequently missed: predictive value of US only 20-40%
- Small or absent gastric bubble
- Polyhydramnios >90%

After birth: early symptoms

- Salivation, inability to feed, coughing & choking
- Inability to place OGT/NGT
- TEF: abdominal distention, respiratory compromise
- CXR
  - Upper esophagus/pouch may be outlined by air
  - Contrast XR (air/water soluble)
Classification

GROSS CLASSIFICATION

Type A  Type B  Type C  Type D  Type E
Diagnosis
Diagnosis
Workup

- Rule out associated malformations
- Physical exam
  - Anorectal
  - Cardiac
  - Musculoskeletal
- Xray
  - Air distribution: esophageal pouch, abdominal air?
  - Vertrebral/sacral anomalies
- Ultrasound
  - Renal, sacral, central venous
- Echo
  - Cardiac
  - Anatomy: right or left descending aortic arch
Preoperative considerations

• Involve subspecialists according to diagnosed associated malformations
  • Cardiologist/Cardiac anesthesia
  • Nephrologist
  • Neurosurgeon
  • Infectious disease
Preoperative considerations

• NPO
  • Iv fluids
• Risk for pneumonitis from aspiration of upper pouch
  • sump catheter in upper pouch to low cont suction
  • avoid perforation
• Risk for pneumonitis from gastric reflux
  • broad spectrum antibiotics
• Avoid intubation and positive pressure ventilation
  • Gastrointestinal abdominal inflation via distal TEF
  • Respiratory distress → respiratory failure
• Preparation for surgery
  • Vit K analogue
Anatomy
Operation

- Emergency operation rarely necessary
  - Exception: emergent ligation of large distal TEF leading to respiratory failure due to GI inflation
    - Gastrostomy, ligation of fistula
- Surgical repair within 24-48 hours AFTER thorough workup
  - Preoperative bronchoscopy +/- blockage of TEF
  - Open approach
  - Thoracoscopic approach
Operation

- Preoperative bronchoscopy
Operation

• Blockage of fistula with Fogarty balloon catheter
Operation

- Blockage of fistula with Fogarty balloon catheter
- Fluoroscopy
Operation

• Steps of open repair of type C EA with TEF via thoracotomy
  • Patient in lateral position
  • Curved thoracotomy from anterior axilla to vertebral muscles (right side; left side if aortic arch on the right)
  • Division or retraction of chest wall musculature
  • Division of intercostal musculature of 4th intercostal space
  • Extra-pleural dissection to postero-medial cavity
  • Identification and division of azygos vein
  • Identification, ligation and division of TEF
  • Identification, dissection of proximal pouch
  • End-end anastomosis of distal and proximal esophagus
  • NG/OG tube
  • Chest tube
Anatomy
Operation

Patient position and curved thoracotomy
Operation

Extra-pleural dissection
Operation

- Ligation and division of distal TEF
- Mobilisation of proximal and distal esphagus
Operation

Anastomosis of proximal pouch to distal esophagus
Operation

- Thoracoscopic repair of **type C EA with TEF**
  - Preoperative bronchoscopy
    - Identification of TEF
    - Blockage with Fogarty balloon
  - Similar intrathoracic operative steps
  - Lateral position, exposing right chest
  - Placement of three trocars 3-5 mm
Operation
Video: Thoracoscopic repair

Thoracoscopic TEF Repair

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Pediatric Surgery
Anesthesia

• Challenges
  • Neonate
  • Associated anomalies
  • Modified lateral position
    • Unilateral lung collapse on operated side
    • Reduced tidal volumes due to compression of contralateral lung
  • Respiratory compromise from air drainage into GI tract via TEF
  • Dislodgement of TEF block into trachea → airway occlusion
Postoperative course

- NICU, iv/pr analgesia, iv fluids
- Antacid treatment: Histamine-2 blockers or proton pump inhibitors
- Controversial feeding regimen
  - Early OGT feeding if OGT/NGT placed during surgery
  - NPO for 4-7 days, followed by esophagram (swallow study or retrograde via gastric tube)
  - Start po feeds after leak has been ruled out
  - D/C chest tube (CT) after feeds tolerated
- Broad spectrum Abx until CT removed
- No need for extended intubation in most cases
- D/C home when full feeds tolerated po
- Follow up 1/6/12 months postop
Outcomes

• Challenges
  • Neonate
  • Associated anomalies
  • Modified lateral position
    • Unilateral lung collapse on operated side
    • Reduced tidal volumes due to compression of contralateral lung
  • Respiratory compromise from air drainage into GI tract via TEF
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Early Complications

- Sepsis
- Anastomotic leak
  - Common, ~15-20% of all patients
  - Contributing factors: de-vascularized esophagus, high tension anastomosis, poor technique
  - >90% spontaneous resolution if adequately drained, iv Abx
  - 3-5% of all leaks are caused by early anastomotic disruption
    - in the first 48 hours
Late Complications

• Esophageal stricture
• Dysphagia
  • 50-90% of adults after TEF repair report occasional dysphagia
  • 30% report problems with choking
• Recurrent TEF
  • 3-14%
  • Usually occur in early postop period, it can be missed
  • Watch for coughing/choking, recurrent pneumonias
  • Diagnosis: contrast esophagram or bronchoscopy
  • Re-surgery indicated (newer techniques: cauterization, coagulation)
• Gastroesophageal Reflux Disease
• Tracheomalacia
Esophageal Stricture

- Common postoperative finding: reported -80% of patients!
- Defined as narrowing of the esophagus causing symptoms like dysphagia, recurrent aspiration pneumonias
- Similar rates open vs thoracoscopic repair
- Treatment
  - Aggressive GER treatment. Acid “bathing” of the anastomosis causes scarring and calcification
  - Antegrade dilation with bougies or balloon dilators
    - 50% of all strictures present in the first few months after surgery and need 1-3 dilations
  - Injection of triamcinolone or mitomycin c (antineoplastic drug)
  - Resection of recalcitrant stricures with primary anastomosis or replacement with bowel
GERD

- Very common (~70%) after surgical EA/TEF repair
- Lifelong tendency for GERD
  - Increased risk for late complications like Barrett esophagus
- Esophageal motor dysfunction (primary or postop)
  - Poor peristalsis
- Shortening of intra-abdominal esophagus → impairment of lower esophageal sphincter function
- All patients should be discharged on antacid meds
  - Duration: controversial: at least one year
- “pathologic GERD”: vomiting, dysphagia, recurrent esophageal stenosis, respiratory symptoms (Asthma, recurrent pneumonias)
  - Extensive GERD treatment & GI workup: pH/impedance probe
  - 40-70% require antireflux operation: Nissen wrap
Tracheomalacia

- “Floppy” trachea, tendency to collapse during expiration
- 75% of patients with EA/TEF, less frequently in patients with pure EA
- Level of tracheal collapse in the region of or above previous TEF site (behind aorta)
- Symptoms can mimic anastomotic stricture, recurrent TEF or GERD
- Diagnosis established by bronchoscopy
- Treatment
  - Expectant in mild or moderate cases
  - Aortopexy in severe cases
Tracheomalacia

Anatomy aorta/trachea

Aortopexy
Long Gap Esophageal Atresia

- No clear definition for “long” gap
- Primary repair not possible due to gap between proximal and distal esophagus
  - Common in pure EA
- No rush for initial surgical repair if no TEF present
- Delayed repair after initial G tube placement
  - Spontaneous growth of upper pouch
  - +/- bougienage has been applied
- Primary (thoracoscopic) repair
- Lengthening procedures (Foker) with secondary repair
Long term outcome

- Life-long impaired esophageal motility +/- GERD +/- stricture
- Dysphagia
  - 50-90% of adults
- Occasional choking
  - 30% of adults
- Recurrent respiratory infections due to aspiration (-60%)
- Thoracic deformities: rare after muscle sparing approach
  - Winging of scapula 5%
  - Scoliosis 1-2%
  - Ipsilateral shoulder elevation -5%
Long term outcome
Bringing it all together: Case

- 29 weeker, 810g with prenatal diagnosis of tetralogy of Fallot
- Respiratory distress at birth, immediate intubation
- Failure of Replogle placement
- CXR
Case

- Diagnosis if EA with type C TEF
- Increasing abdominal distension on day 3 of life
  - Progressive ventilator support
- Respiratory decompensation on day 4 of life

- Surgical options?
Remember: Anatomy

A  7.8%
B  4-5%
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Case

- Surgical/medical options? (instable patient, 780g, cardiac anomaly, abdominal distension, metabolic/respiratory acidosis)
  - Advance ET tube passed fistula?
  - Block TEF with balloon?
    - Bronchoscopy?
    - Open?
  - Definitive repair?
    - Open or thoracoscopic?
  - Ligation of the fistula, open or thoracoscopic?
  - Gastric decompression and placement of G tube?
Case

- Patient too unstable for thoracoscopy, long surgery
- 2.5-cm transverse mini-laparotomy
- Stamm gastrostomy for GT placement
- Via gastrostomy site, a 6-Fr Fogarty catheter was advanced under fluoroscopic guidance into the TEF to block it
Case

- Feeding via G tube, ventilation now safe after blockage of TEF
- → patient stabilized, gaining weight
- Patient underwent TEF ligation after 3 weeks
- Definite thoracoscopic EA repair with anastomosis after another 6 weeks
- Cardiac surgery at age of 5 months
Case

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• Cardiac surgery at age of 5 months
To the north of Kalispell is Glacier national park. It is located in Montana and Canada (Alberta/British Columbia).