MANAGEMENT OF ESOPHAGEAL ATRESIA AND TRACHEO-ESOPHAGEAL FISTULA

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Long Island College Hospital
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CASE PRESENTATION
HPI

- 3 days old baby girl
- 32 weeks gestation
- Emergency C/section due to PROM and fetal distress
- **Birth weight 1670 g**
- **APGAR 5 @ 1mn and 7 @ 5mn**

**IVF with a DONOR EGG**
- DONOR MOTHER- 25 yo healthy
- CARRIER/BIRTH MOTHER- 50 yo, h/o HTN
HPI

- RESPIRATORY DISTRESS
  - Intubated
  - Surfactant
  - Extubated the second day

- IN THE MEANWHILE
  - Passed meconium
  - Neonatal jaundice ⇒ PHOTOTHERAPY
  - Amp/Gent for presumed sepsis
HPI

- **SAME NIGHT**
  - Respiratory distress/labored breathing
  - NCPAP trial $\Rightarrow$ Intubation

- NGT attempted twice w/o success

- Abdominal X-ray
ABDOMINAL XRAY
PHYSICAL EXAMINATION

- Intubated
- Abdomen distended
- Rectal exam - meconium
- No other anomalies detected
- Labs - within normal
ADDITIONAL WORK UP

CARDIAC ECHO

• PDA with left to right shunt
• OSTIAL ASD with left to right shunt

RENAL ULTRASOUND

• NORMAL
DIAGNOSIS

TYPE C ESOPHAGEAL ATRESIA
- with distal tracheo-esophageal fistula
- proximal pouch at T4-T5

ASSOCIATED ANOMALIES
- Vertebral malformations
- PDA
- ASD
Gastrostomy

Broviac catheter placement

Right posterolateral thoracotomy, closure of TE fistula and end-to-end primary esophago-esophagostomy
RIGHT POSTEROLATERAL THORACOTOMY, CLOSURE OF TE FISTULA AND END-TO-END PRIMARY ESOPHAGO-ESOPHAGOSTOMY

- Left lateral decubitus
- Right posterolateral thoracotomy through the 4th intercostal space
- Retropleural dissection underwent
- Ligation/division of the azygos vein
- The gap between the 2 esophageal stumps was about 1.5-2cm
- The fistula was identified, divided and closed with interrupted 5/0 prolene sutures
- Mobilization of proximal pouch
RIGHT POSTEROLATERAL THORACOTOMY, CLOSURE OF TE FISTULA AND END-TO-END PRIMARY ESOPHAGO-ESOPHAGOSTOMY

- Tension-free end-to-end esophago-esophagostomy with interrupted 3/0 silk, posterior row first then anterior after placement of a silastic tube through the anastomosis up to the mouth
- 8 Fr chest tube
- Penrose drain next to the anastomosis
- Closure in layers
POST-OP

- Extubated on POD#4
- TPN post-op

- POD#9 - Gastrograffin swallow showed a small leak
- Tube feeds started and tolerated
- Chest tube removed but Penrose left in place
MANAGEMENT OF ESOPHAGEAL ATRESIA AND TRACHEO-ESOPHAGEAL FISTULAS
# HISTORICAL BACKGROUND

<table>
<thead>
<tr>
<th>Year</th>
<th>Event Description</th>
</tr>
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<tbody>
<tr>
<td>1899</td>
<td>First permanent gastrostomy for a patient with EA by Hoffman.</td>
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<tr>
<td>1941</td>
<td>Cameron Haight (University of Michigan) performed the first successful repair of EA-TEF via a left extrapleural approach w/ fistula ligation and a single layered esophageal anastomosis.</td>
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</table>
HISTORICAL BACKGROUND

- 1943- Haight revised the procedure to a right thoracotomy
- Between 1939 and 1969, he cared for more than 284 infants with EA and reported an overall survival of 52%
Normal embryology - budding of the tracheal primordium from the primitive esophagus and formation of the tracheoesophageal septum which begins caudally and ends cranially.
EMBRYOLOGY/PATHOGENESIS

- Foregut occlusion and failure of recanalisation of the intestinal lumen
- Cephalic NEUROCRISTOPATHY and defective pharyngeal pouch development
- Abnormal growth of the esophagus into the trachea and abnormal lung morphogenesis (it was found that the distal esophagus and fistula tract are of respiratory origin)
EPIDEMIOLOGY

- Incidence – **1/2440 to 1/4500 births** (Europe > USA and Australia)
- Male/female ratio - **1.33 to 2.29** (depending on the type)
- Risk factors
  - First pregnancy
  - Mothers younger than 20
  - Advanced maternal age
OTHER ASSOCIATIONS

- Chromosomal abnormalities (tri 13 and 18)- 6-10%
- Teratogens- OCPs, Thalidomide, estrogen/progesterone
- Hyperthyroid and diabetic mothers
- Genetic syndromes- DiGeorge, Polysplenia sequence, Holt-Oram syndrome, Pierre-Robin syndrome
- Familial- if 1 child affected risk is 0.5-2%; if more than 1 child affected risk is 20%; if one parent affected ⇒ risk 3-4%
## ASSOCIATED ANOMALIES

<table>
<thead>
<tr>
<th>System</th>
<th>Incidence</th>
<th>Description</th>
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</thead>
<tbody>
<tr>
<td>Cardiovascular</td>
<td>35%</td>
<td>Account for most of the death associated with EA; VSD (19%) and ASD (20%) most common; right sided aorta 4%</td>
</tr>
<tr>
<td>Genitourinary</td>
<td>24%</td>
<td>Renal agenesis or Hypoplasia (1%); hypospadias, exstrophy;…</td>
</tr>
<tr>
<td>Gastrointestinal</td>
<td>24%</td>
<td>Anorectal malformations (14%); duodenal atresia (2%); malrotation (4%)</td>
</tr>
<tr>
<td>Neurologic</td>
<td>12%</td>
<td>Neural tube defects (2.3%): hydrocephalus (5.2%); holoprosencephaly (2.3%)</td>
</tr>
<tr>
<td>Musculoskeletal</td>
<td>20%</td>
<td>Limb anomalies (15%); vertebral anomalies (17%)</td>
</tr>
<tr>
<td>VACTERL</td>
<td>20%</td>
<td>Vertebral, Anorectal, Cardiac, Tracheo-Esophageal, Renal, Limb</td>
</tr>
<tr>
<td>Overall Incidence</td>
<td>50-70%</td>
<td>50-70% at least one malformation; most common with EA w/o TEF and least common with H-type TEF</td>
</tr>
</tbody>
</table>
ASSOCIATED ANOMALIES

- **VACTERL**- 20% mainly with EA-TEF
  may have also other midline defects (cleft lip/palate 2%; sacral agenesis 2%)
- **VACTERL-H** with congenital hydrocephalus
- **Unilateral pulmonary agenesis**- rare (37 cases since 1874)
- **Tracheobronchial abnormalities**- 47% (ectopic or absent right upper lobe bronchus, congenital bronchial stenosis, tracheal malformations)
- **Abnormalities in vagal innervation**
### CLASSIFICATION: ANATOMIC

<table>
<thead>
<tr>
<th>TYPE</th>
<th>%</th>
<th>GROSS TYPE</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. EA w/ distal TEF</td>
<td>85.8</td>
<td>C</td>
</tr>
<tr>
<td>2. EA w/o TEF</td>
<td>7.8</td>
<td>A</td>
</tr>
<tr>
<td>3. TEF w/o EA</td>
<td>4.2</td>
<td>E</td>
</tr>
<tr>
<td>4. EA w/ TEF to both pouches</td>
<td>1.4</td>
<td>D</td>
</tr>
<tr>
<td>5. EA w/ proximal TEF</td>
<td>0.8</td>
<td>B</td>
</tr>
</tbody>
</table>
**CLASSIFICATION: PROGNOSTIC**

<table>
<thead>
<tr>
<th>GROUP</th>
<th>SURVIVAL</th>
<th>WATERSTON CLASSIFICATION</th>
</tr>
</thead>
<tbody>
<tr>
<td>A</td>
<td>100%</td>
<td>Birth weight &gt;2500g and otherwise healthy</td>
</tr>
<tr>
<td>B</td>
<td>85%</td>
<td>Birth weight 2000-2500g and well OR higher weight w/ moderate associated anomalies (non cardiac plus PDA, VSD or ASD)</td>
</tr>
<tr>
<td>C</td>
<td>65%</td>
<td>Birth weight &lt;2000g OR higher with severe associated Cardiac anomalies</td>
</tr>
</tbody>
</table>

Waterston’s 1962 classification separated patients into groups based on birth weight, pneumonia and congenital anomalies:
- Group A were treated with immediate repair
- Group B were treated with delayed repair
- Group C were treated with staged repair
## CLASSIFICATION: PROGNOSTIC

<table>
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<th>GROUP</th>
<th>SURVIVAL</th>
</tr>
</thead>
<tbody>
<tr>
<td>I. Birth weight &gt; 1500g w/o major CHD</td>
<td>97%</td>
</tr>
<tr>
<td>II. Birth weight &lt;1500g or major CHD</td>
<td>59%</td>
</tr>
<tr>
<td>III. Birth weight &lt;1500g and major CHD</td>
<td>22%</td>
</tr>
</tbody>
</table>

SPITZ Classification- most commonly used currently
PRENATAL DIAGNOSIS

- Rarely diagnosed
- Findings-
  - small or absent stomach bubble
  - associated maternal polyhydramnios
- Predictive value only 20-40%
- Fetal MRI can help
CLINICAL FINDINGS

- Earliest sign: Excessive salivation
- First feeding followed by regurgitation, coughing, choking
- Other manifestations:
  - Cyanosis w/ or w/o feeding
  - Respiratory distress
  - Inability to swallow
  - Inability to pass a suction catheter

- If distal fistula ⇒ abdomen distends + chemical pneumonitis ⇒ compression on the chest + worsening respiratory status
DIAGNOSIS

Plain XR showing a Replogle tube in the pouch

0.5-1 ml of barium injected in upper pouch confirm the diagnosis
DIAGNOSIS

- A small upper pouch suggests a proximal TEF
- Air in the stomach confirms a distal TEF
- Absence of air in the abdomen represents EA w/o TEF

- TEF w/o EA (H-type)
  - High index of suspicion needed
  - Barium esophagography in the prone position
  - Confirm with bronchoscopy or esophagoscopy
ADDITIONAL TESTS

- ECHOCARDIOGRAPHY
- RENAL ULTRASOUND
- CHROMOSOMAL ANALYSIS
PREOPERATIVE MANAGEMENT

- **SUMP CATHETER** in upper esophageal pouch
- Position infant to prevent reflux and aspiration
  - Upright sitting position
  - **Head-up prone position**
- Broad spectrum antibiotics
- IVF
- ET intubation if needed but AVOID if possible
Operative Repair: Esophageal Atresia with Distal Tracheoesophageal Fistula

Right posterolateral thoracotomy
A left thoracotomy may be needed in case of right sided arch
Chest entered through the fourth intercostal space

Extrapleural approach is preferred to minimize the risk of empyema in case of leak

Azygos vein is ligated

Fistula, distal pouch and proximal pouch identified
OPERATIVE REPAIR: ESOPHAGEAL ATRESIA WITH DISTAL TRACHEOESOPHAGEAL FISTULA

A. Lower part of esophagus and fistula circumferentially dissected
5/0 or 6/0 prolene sutures are placed and the fistula divided
Check for air leak

B. Distal dissection should be limited
Traction suture proximally to mobilize proximal end
Possible identification of a proximal TEF
Beware of vagal branches

C. May need to do a proximal circular myotomy to further mobilize the proximal pouch
Operative Repair: Esophageal Atresia with Distal Tracheoesophageal Fistula

Single layer anastomosis with 5/0 or 6/0 interrupted suture (silk vs polypropelene or polyglycolic acid)

Corner stitches tied on the outside

Posterior row of interrupted sutures tied on the inside

Complete anastomosis over a tube
THORACOSCOPIC REPAIR OF EA-TEF

Benefits include
- Superior visualization
- Improved cosmesis
- Decreased morbidity of neonatal thoracotomy (winging of scapula, scoliosis, chronic pain, chest wall asymmetry, …)

Limited studies and follow up

Preliminary results
- Stenosis rate 30-50%
- Leak rate 12-15%
- GERD rate 30-50%
- OR time 2-4 hours
Between 1990 and 2006
- 22 neonates underwent TR
- 23 matched controls underwent open repair

<table>
<thead>
<tr>
<th></th>
<th>TR (n=23)</th>
<th>OCR (n=22)</th>
<th>P</th>
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<tbody>
<tr>
<td>OR time (mn)</td>
<td>149.4</td>
<td>179</td>
<td>.18</td>
</tr>
<tr>
<td>Leak</td>
<td>4 (17.4%)</td>
<td>3 (13.6%)</td>
<td>.725</td>
</tr>
<tr>
<td>Strictures</td>
<td>2 (8.7%)</td>
<td>4 (18.8%)</td>
<td>.414</td>
</tr>
<tr>
<td>Early complications</td>
<td>1 (4.35%)</td>
<td>6 (27.2%)</td>
<td>.047</td>
</tr>
<tr>
<td>Mortality</td>
<td>1 (4.35%)</td>
<td>2 (9.1%)</td>
<td>.6</td>
</tr>
</tbody>
</table>
THORACOSCOPIC REPAIR OF TRACHEO-ESOPHAGEAL FISTULAS: A CASE-CONTROL MATCHED STUDY  AL TOKHAIS ET AL.  J PED SURG 2008 43: 805-809

- Time to full enteral feeding
  - TR 10 days
  - COR 16.3 days

- CONCLUSIONS
  - Comparable operative times
  - Comparable leak and stricture rates
  - Comparable rates of mediastinitis, pneumonias or death despite the transpleural approach
  - Lower rate of early complications
  - Earlier feeding
OPERATIVE REPAIR: ESOPHAGEAL ATRESIA with DISTAL TRACHEOESOPHAGEAL FISTULA

- Respiratory distress w/ ET intubation ⇒ gastric distention and more respiratory distress
- Gastric division, banding of gastroesophageal junction or placing ET tube below the level of the fistula
- Bronchoscopic placement of a Fogarty catheter in the fistula
- Emergency gastrostomy- consider underwater seal
OPERATIVE REPAIR: LONG GAP EA

- Staged procedure starting with a gastrostomy
- The gap tends to lessen during the first few months of life due to spontaneous growth

- Non-operative methods to help shrink the gap
  - Upper pouch bougeinage for 6-12 weeks
  - Upper and lower pouch bougeinage
  - Electromagnetic field applied to 2 pieces of metal placed in both pouches
Operative Repair: Long Gap EA

Operative approaches
- Threads with silver olives
- Multistaged extrathoracic elongation technique
- FOKER technique - Traction sutures on both ends of the esophagus that exit through the chest wall and are pulled in opposite directions
Operative Repair: Long Gap EA

Circular Myotomy

- Complications
  - Worsens esophageal motility
  - Esophageal leak
  - Impaction of food particles in the myotomized segment
  - Ballooning of the myotomized segment - pseudodiverticulum

- Variations
  - Spiral myotomy
  - Short horizontal myotomies
Other approaches

- Creation of a full thickness anterior or posterior flap (Gough et al.)
- Extensive dissection of the lower esophagus to or below the diaphragm
- Combined thoracic and abdominal approach- ligation of the left gastric artery, division of the stomach and mobilization of the cardia and fundus into the chest with a partial fundoplication (SchSrli et al.)
OPERATIVE REPAIR: EA WITHOUT TEF

- Almost no esophagus in the chest
- Higher incidence of premature (52%), Down’s syndrome (10-20%) and duodenal atresia (10%)
- On PE- abdomen scaphoid
- AXR- no abdominal air
- Higher incidence of tracheomalacia
OPERATIVE REPAIR: EA WITHOUT TEF

Gastrostomy placed w/i the first 24-48 Hours of life and feeding started

Elongation techniques

Thoracotomy with attempted primary anastomosis

If non-feasible ⇒ Gastric transposition
OPERATIVE REPAIR: ISOLATED (H-TYPE) TRACHEOESOPHAGEAL FISTULA

- 4% of EA
- Choking and cyanotic spells
- Later recurrent pneumonias typically RUL
- Diagnosis: tube video esophagography in the prone position
- Bronchoscopy/esophagoscopy confirms diagnosis
- May place guidewire through the fistula during bronchoscopy
**Operative Repair: Isolated (H-Type) Tracheoesophageal Fistula**

**Cervical approach**
Division of the inferior thyroid artery and middle thyroid vein

*Identify the recurrent laryngeal nerve*
Traction sutures placed on the superior and inferior borders of the fistula

5/0 prolene or silk on the side of the trachea
5/0 silk or vicryl on the side of the esophagus

Muscle tissue interpoistion

**Complications -** tracheal edema and RLN injury
OPERATIVE REPAIR: EA WITH UPPER POUCH FISTULA

- 2 types:
  - Proximal pouch fistula with distal pouch fistula (1.4%)
  - Isolated proximal pouch fistula (0.8%)

- Contrast study with fluoroscopy
- Bronchoscopy and esophagoscopy
- Same repair as distal pouch fistula

- If diagnosed after repair of an EA w/ or w/o TEF ⇒ a cervical approach can be done
OUTCOMES

**SURVIVAL**

85-95% survival

Worse prognosis for isolated EA patients

**FACTORS ASSOCIATED W/ INCREASED RISK OF DEATH AND LONG TERM MORBIDITY**

- Lower birth weight (<1500)/prematurity
- Major CHD
- Severe associated anomalies
- Ventilator dependency
- Long gap
EARLY COMPLICATIONS: ANASTOMOTIC LEAK

- **14-16%** of patients with EA
- **95%** close spontaneously
- **3-5%** are significant and are recognized within 24-48h

- The infant deteriorates from pneumothorax or mediastinitis uncontrolled by drainage
- Poor surgical technique, myotomy, tension, ischemia of the esophageal ends
- Reexploration and attempted repair with a buttress
- Cervical esophagostomy and delayed replacement
EARLY COMPLICATIONS: ESOPHAGEAL STRICTURE

- Stricture requiring dilatation 80%

Treatment
- Bougienage: antegrade/retrograde
- 1-3 dilatations
- R/O GERD- fixe before treating fistula

Associated factors
- Poor technique (tension, 2-layered anastomosis, silk sutures)
- Ischemia
- GERD
- Anastomotic leak
EARLY COMPLICATIONS: RECURRENT TEF

- Occurs in 3-14% of cases
- Due to anastomotic leak and inflammation and erosion through the anastomotic site
- Symptoms ≡ H-type TEF
- Diagnosis
  - Air filled esophagus on plain radiograph
  - Contrast esophagography in the prone position
  - Bronchoscopy
- Treatment
  - Thoracotomy with ligation and a pleural, pericardial or intercostal flap
LATE COMPLICATIONS: GERD

- **30-70% after repair of EA**
- **Symptoms**: vomiting, dysphagia, recurrent strictures, stridor, cyanotic spells, recurrent pneumonia, reactive airway disease

**Diagnosis**
- Upper GI contrast studies
- pH metry
- Manometry is not helpful

**Management**
- Medical management
- 45-75% require surgical treatment
- Nissen, Thal or Nissen-Collis fundoplication
LATE COMPLICATIONS: TRACHEOMALACIA

- 10-25% symptomatic
- Approximately half require surgery

Pathophysiology
- Intrinsic (shorter cartilage and lack of support)
- Extrinsic compression by the aortic arch
- At the level of or just above the site of the TEF generally at the level of the aortic arch

Clinical manifestations
- Barking cough
- Apneic spells 5-10mn after meals ⇒ cyanosis, bradycardia ⇒ cardiorespiratory arrest
LATE COMPLICATIONS: TRACHEOMALACIA

Diagnosis
Bronchoscopy with spontaneous ventilation

Treatment
Mild to moderate symptoms tend to resolve with time
Severe symptoms ⇒ Surgery

Aortopexy - Ascending and arch sutured to the sternum or suspension with a pericardial flap
Tracheal stent/tracheostomy in case of failure
LATE COMPLICATIONS: OTHER LONG TERM ISSUES

- Motility disorders

- Dysphagia
  - 50-90% of adults
  - 30% have choking
  - Early feeding intolerance
  - Failure to thrive
  - May improve with time
REFERENCES


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