Tracheoesophageal fistula

Liz Sim
Case Presentation

Pt is a 38+4 week twin gestation
- C section 2/2 breech with intrauterine growth retardation
- Apgar 9/9
- 2410 g
- Maternal Hx 35, G1P1, IVF, prenatal care uncomplicated

Transferred to NIUC when excessive and copious frothy oral secretions were noted
- NG tube unable to be passed farther than 10cm
CXR
Case Presentation

VACTERL was ruled out
- CXR, renal ultrasound and echo

On exam
- Anal stenosis but passed meconium
- Dimple over coccygeal area
Operative Course

Ex lap, stamm gastrostomy, right posteriolateral retropleural thoracotomy, resection of TEF with end to end esophago-esophagostomy

- Retropleural dissection at 5th intercostal space
- TEF was transected and tied off
- Proximal esophagus was at 1st rib
- 3-0 silk sutures placed on either side of distal esophagus and the proximal blind end of esophagus. Sutures were then placed along the posterior wall of the distal esophagus and to the blind end.
- Anterior wall of the esophagus was incised and the lumen was entered placing a 6.6 F silastic catheter as a stent
- Anastomosis was completed and covered with Tisseel.
- 10F chest tube was placed and a penrose in the retropleural space
Post Op Course

POD 3 started on glucose via PEG

POD 4 persistent pneumothorax, increase work of breathing
- needing 50% oxygen, eventual reintubation
- CXR showed small pneumothorax
- Gastrograffin study showed leak of esophageal anastomosis
Post Op Course

Right posterolateral thoracotomy, thoracic lavage, drainage of anastomotic leak, and debridement of TEF repair

- Fibrin sealant was intact and removed
- Minimal salivary secretions were aspirated
- Irrigated and tisseel application
- Chest tube and penrose placed in retropleural space
Post Op Course

Feeds advanced to goal via gastrostomy
POD 7/10 extubated
POD 10/13 febrile negative cultures, abx started
POD 12/15 penrose fell out chest tube inserted
POD 15/18 gastrostomy feeding stopped due to reflux, one of two chest tubes removed
POD 32/35 esophogram negative, oral feeds initiated
POD 34/37 final chest tube removed, on full oral feeding.
Questions
epidemiology

1:27 to 4.55 per 10,000 live births

Most cases are sporadic
- Slightly higher in twins

Survival
- Without congenital heart disease 95% vs with 67%
embryology
Embryology/theories

Tracheoesophageal septum: Fusion of invaginating lateral longitudinal ridges, creating a septum dividing foregut and respiratory system

Cephalic neurocristopathy: a clear association of neural crest-implicated cardiovascular anomalies (aortic arch, conotruncal and membranous ventricular septal defects), thymic, thyroid, parathyroid, and facial malformations suggests that the pathogenesis of EA may be related to defective pharyngeal arch development
Voight classification
<table>
<thead>
<tr>
<th>Group</th>
<th>Features</th>
<th>Survival (%)</th>
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</thead>
<tbody>
<tr>
<td>I</td>
<td>&gt;1500 grams, no major cardiac anomaly</td>
<td>98.5</td>
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<tr>
<td>II</td>
<td>&lt; 1500 grams or major cardiac anomaly</td>
<td>82</td>
</tr>
<tr>
<td>III</td>
<td>&lt; 1500 grams and major cardiac anomaly</td>
<td>50</td>
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<tr>
<td>Class</td>
<td>Birth weight</td>
<td>Anomaly</td>
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<tr>
<td>-------</td>
<td>--------------</td>
<td>---------</td>
</tr>
<tr>
<td>I</td>
<td>$\geq 2$ Kg</td>
<td>Nil</td>
</tr>
<tr>
<td>II</td>
<td>$&lt; 2$ Kg</td>
<td>Nil</td>
</tr>
<tr>
<td>III</td>
<td>$\geq 2$ Kg</td>
<td>Major cardiac anomaly</td>
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</table>
Diagnosis and clinical findings

Poor detection rate prenatal

Most become symptomatic within hours of life
- Excessive salivation
- Feeding followed by regurgitation, choking or coughing
- Cyanosis and respiratory distress
- Inability to pass NGT

Babygram
- Shows coiling of tube
- Can pass air
- If needed can give contrast agent
Pre-op

Prevention of pneumonitis
- replogle esophageal pouch
- Upright position
- Broad spectrum antibiotic coverage
- Pulmonary physiotherapy

Rule out VACTERL or other associated diseases
- Down syndrome
- Digeorge
Surgery

Goal: Division of fistula with primary anastomosis of esophagus

Right posterior lateral thoracotomy at 4th intercostal space
- Muscle sparing to prevent post operative shoulder morbidity
- Extra pleural to prevent empyema in case of leak, but rather get esophagocutaneous fistula which closes in 1-2 weeks

Azygos vein is identified – can be ligated

Main structures: upper pouch, distal TEF trachea, vagus nerve

Circumferentially dissect distal trachea and close fistula with absorbable suture with a cuff of esophagus to minimize stricture
Surgery

Proximal pouch
- Identified by pushing NGT
- Traction suture
- Mobilization of upper pouch and incised.
- End to end anastomosis with interrupted silk sutures over a small feeding tube
  - End to side not encouraged due to risk of recurrent TEF

Chest tube for drainage

Multi layer closure
Laparoscopic repair

Superior visualization, improved cosmesis

Decreased morbidity
  - Scoliosis, winged scapula, chronic pain, shoulder weakness, chest wall asymmetry, maldevelopment

Technically demanding
Long Gap Esophageal atresia

Delayed primary repair with gastrostomy
Upper pouch bougienage BID for 6-12 weeks
Electromagnetic bullets in both ends of esophagus
Extra thoracic elongation technique
  ◦ Upper esophagus mobilized and brought out as an end cervical esophagostomy
  ◦ Q2-3 weeks esophagus mobilized and translocated down anterior chest wall
Folker technique: Traction sutures on proximal and distal esophageal pouches
  ◦ Brought out through chest wall and serially pulled for 10-14 days
Circular myotomy of upper esophagus
  ◦ Complications: leak, impacted food, pseudodiverticulum, stricture
Kimura technique: circular myotomy
Long Gap Esophageal Atresia

If no closure by 3 months of age begin to think of esophageal replacement
  ◦ This requires a spit fistula in preparation for gastric transposition or colon conduit
  ◦ Allows for sham feeds.

Conduit depends on several factors
  ◦ Colon replacement
  ◦ Reversed gastric tube(tubularized greater curvature)
  ◦ Jejunum
Complications

Early
- Anastomotic leak (13-16%)
- Anastomotic stricture (80%)
- Recurrent TEF (3-14%)

Late
- GERD (30-70%)
- Tracheomalacia (clinically 10-25%)
- Respiratory disease
- Disordered esophageal peristalsis
Summary

There are five types
Survival depends on birth weight and malformations
Babies heal well, leaks may spontaneously close
There are multiple techniques for lengthening a long gap atresia
References


Li, Jiaang et all. A scoring system to predict mortality in infants with esophageal atresia: A case control study. Medicine 2017 Augus 96(32)e7755
