History

- 8 y.o. boy admitted on 2/17/06 with a diagnosis of severe gastroesophageal reflux disease. He was scheduled to undergo an elective revision of his colonic interposition graft performed 7 years earlier for esophageal atresia. He had multiple prior admissions for aspiration pneumonia and received tube feedings for failure to thrive.
- PMH: Asthma, GERD
- Allergies: latex
- Birth Hx: 32 ½ weeks, polyhydramnios
- PSH: esophagostomy and gastrostomy '97, colonic interposition ‘98
- Medications: Advair, Singular, Prevacid, Albuterol
Physical

- Wt 64lbs 75%
- Cardiac: no murmurs
- Chest: CTA b/l
- Abd: well healed upper midline scar, RUQ feeding tube, NT/ND, no masses
- Genitalia: normal, testes descended b/l
- Rectal: normal tone
- Ext: no c/c/e
Laboratory Values

\[
\begin{align*}
\frac{135}{4.0} & \quad \frac{98}{25} & \quad \frac{10}{0.6} & \quad 129 \\
6.6 & \quad \frac{13.8}{39} & \quad 293 \\
\frac{13.3}{28.1} & \quad 1.1
\end{align*}
\]
Intraop Events

- Procedure: thoracoabdominal exploration (median sternotomy), tapering coloplasty, partial resection colon interposition, Nissen fundoplication, Stamm gastrostomy
- EBL: 50cc
- Fluids: 1700cc
- OR time: 5 hours
Postop Course

- POD#1 - Extubated in PICU
- POD#6 – Esophogram and return of bowel function
- POD#7 - Tube feeding started
- POD#8 - Full liquids started
- POD#15 – Discharged home tolerating soft diet
Management of Esophageal Atresia
Historical Perspective

- 1670 – Durston - First reported case in conjoined twins
- 1888 – Steele - First attempted surgical correction of pure EA by gastrostomy
- 1939 – Levin and Ladd - First survivors of staged repair of EA-TEF
Historical Perspective

- 1941 – First successful primary repair - Haight
  - Left extrapleural approach
  - Primary single layer anastomosis
  - Fistula ligation

- “In recent years there has been no more dramatic advance in surgery than that which has taken place in the treatment of congenital atresia of the esophagus.” Gross R.

Dr. Cameron Haight
Historical Perspective

- Gradual increase in survival rates from 1941 to present 95-99%
- Advances in neonatal intensive care, nutritional support, surgical management, antibiotic therapy
Epidemiology

- Incidence 1 in 3,570 to 4,000
- Slightly higher frequency in males
- 85% cases with tracheoesophageal fistula
- Higher rate low birth weight and preterm birth
Epidemiology

- 57.3% with atresia only, 36.4% with other major malformations, 6.3% chromosomal anomalies*
- 1 in 10,000 white births 0.55 in 10,000 non white births
- Higher rate in monozygotic twins *

Embryology

- Derived from the primitive foregut
- 4<sup>th</sup> week of gestation trachesophageal diverticulum forms from the laryngotracheal groove
- Tracheoesophageal septum develops during the 4<sup>th</sup> to 5<sup>th</sup> weeks
- Elongates with descent of heart and lungs
- 7<sup>th</sup> week reaches final relative length
Clark DC. *American Family Physician*, 59(4), 1999
Etiology

- Aberration at 26-32 days gestation
  - Differential growth rate
  - Cellular differentiation
  - Apoptosis

- Notochord abnormalities – sonic hedgehog-Gli signaling pathway

- Neural crest abnormalities

Presentation/Diagnosis

- Prenatal ultrasound
  - Polyhydramnios (1 in 12)
  - Small or absent stomach
  - Distended blind esophageal pouch

- Prenatal MRI
  - Blind esophageal pouch
Presentation/Diagnosis

- Postnatally
  - Excessive drooling
  - Fine frothy bubbles in mouth and nose
  - Choking, coughing, cyanotic episodes, and regurgitation with feeding
  - Inability to pass 10Fr OGT
Imaging

- Prenatal ultrasound
  - Good screening test
  - 42% sensitive
  - Polyhydramnios in combination with small or absent stomach
  - No significant role postnatally
Imaging

- Prenatal MRI
  - Following positive US findings
  - 100% sensitivity and 80% specific
  - Positive if esophagus absent in mid chest
  - No significant role postnatally
Imaging

- Plain Radiography
  - Confirmatory test
  - OGT in esophageal pouch
  - Presence or absence of gas in the abdomen
  - Assess gap length
- Anomalies
  - Pneumonitis
  - Atelectasis
  - Cardiac
  - Vertebral
  - Aortic arch
Imaging

Coiled OGT

Gasless Abdomen
Imaging

- Contrast studies
  - Rarely needed
  - Identification of proximal fistula
  - Measuring gap length
  - Should use water soluble contrast
  - Perform under fluoroscopy
Imaging/Diagnostics

- Bronchoscopy
- Esophagoscopy
- CT
- MRI
Classification

Clark DC. *American Family Physician*, 59(4), 1999
Associated Anomalies

- VACTERL – vertebral, anorectal, tracheoesophageal, radial, renal
- Trisomy 18 and 21 (7%)
- Congenital heart disease (25%)
- Urinary tract (22%)
- Orthopedic – vertebral and radial (15%)
- GI – i.e. duodenal atresia, imperforate anus (22%)
Management

- Minimal handling to minimize gastric distention and regurgitation
- NPO!!
- Avoid bag-mask ventilation
- Maintain in partial upright position 45°
- Repeated upper esophageal pouch suctioning minimum q10min or low continuous
- Transfer to tertiary pediatric institution for management and definitive care
Management

- IV Tx - 10% dextrose with hypotonic NaCl solution
- Vitamin K analog preoperatively
- Broad spectrum antibiotics
- TPN if surgery delayed for several days
Preoperative Investigations

- Plain radiograph of torso ± contrast
- Echocardiography
- Renal ultrasonography
- Bronchoscopy – selectively
- Esophagoscopy - selectively
Management of Anomalies

- Congenital heart disease
  - Non-duct-dependent conditions – early EA repair
  - Duct dependent disease – PGE1 infusion – EA repair when stable followed by cardiac surgery
  - Right aortic arch – EA through left thoracotomy
Management of Anomalies

- Urinary Tract
  - Reflux associated nephropathy
  - R/O bilateral renal agenesis or severely multicystic dysplastic kidneys
  - Renal scan if absent on US
Management of Anomalies

- Gastrointestinal
  - Careful anorectal exam – EA repair followed by anorectal anomaly
  - Duodenal atresia – combined EA repair duodenoduodenostomy
  - High imperforate anus – combined EA repair colostomy
Management of Anomalies

- Chromosomal
  - EA repair contraindicated in trisomy 18
  - 70% mortality with EA + major chromosomal anomaly
- Orthopedic – no contraindication to EA repair
  - Vertebral i.e. Hemivertebrae – progressive scoliosis
  - Radial club hand
  - Absent thumb
  - Hip dislocation
  - Talipes equinovarus
Surgical Therapy

- Determinants
  - Type of esophageal anomaly
  - Condition of infant
  - Other congenital anomalies present
Surgical Therapy

- Staged Approach
  - Pure EA and select EA and TEF
  - Stamm gastrostomy
  - Delayed fistula division and esophageal reconstruction
Surgical Therapy

- EA and TEF
  - Fistula division with primary esophageal anastomosis
  - Right thoracotomy via 4th ICS
  - Fistula divided close to trachea with air-tight ligation
  - Mobilization of proximal segment with circular myotomy if extra length required
  - Single layer closure with absorbable suture knots internal
  - Feeding tube placed across anastomosis
Surgical Therapy

- Pure esophageal atresia
  - Delayed repair gastrostomy then primary anastomosis
  - Esophageal lengthening
    - Bougienage
    - Circumferential or spiral myotomy
    - Proximal and distal esophageal mobilization
    - Multistaged extrathoracic lengthening
  - Cervical esophagostomy
    - Failed repair
    - Primary anastomosis impossible
Surgical Therapy

- Esophageal replacement
  - Colon –
    - Right, left, or transverse
    - Retrosternal or posterior mediastinal
    - Passive conduit
Colon Interposition

- Pros
  - Acts as a conduit antiperistaltically or isoperistaltically
  - Good vascular supply via marginal artery
  - Can be placed in esophageal bed of posterior mediastinum
  - Has mucous shield, which protects against reflux
  - Responds to acid with a peristaltic rush for clearance
  - Minimizes/eliminates tension on the upper and lower esophageal segments

Colon Interposition

Cons

- Requires 3 anastomoses
- Empties more slowly than the esophagus
- Requires preoperative bowel preparation
- Long surgical procedure with extensive mobilization
- Dilates and becomes redundant over time
- Slows food transit

Gastric Tube

- Reversed (antiperistaltic) tube
  - More common
  - Supplied by left gastroepiploic artery
- Nonreversed (isoperistaltic) tube
  - Supplied by the right gastroepiploic artery

Gastric Tube

- Pros
  - Does not become dilated, tortuous, or redundant
  - Less risk of ischemia because of robust blood supply
  - Simplified construction through stapler use
  - Has a favorable anatomic location in the upper abdomen
  - Requires fewer anastomoses
  - Has comparable diameter and occupies less space in the thorax and neck
  - Has only one suture line
  - Requires no bowel preparation and is a faster procedure

Gastric Tube

- Cons
  - Extensive gastroesophageal reflux
  - Leaves a small gastric reservoir
  - Creates a long suture line
  - May result in gastric outlet obstruction
  - May be unable to reach high in neck
  - Difficult to place in posterior mediastinum

Gastric Transposition

**Pros**
- Readily available and easily mobilized stomach
- Involves single anastomosis
- Adequate length available
- Has excellent blood supply
- Involves a technically easy procedure
- Has low incidence of leaks and strictures

Gastric Transposition

Cons

- Large bulky possibly causing space problems intrathoracically
- Reflux
- Possible stricture or aspiration due to lack of gastroesophageal valve
- Poor gastric emptying
- May affect pulmonary function
- Results in depleted iron stores causing anemia
- May not reach as high in neck as other methods because of blood supply

Jejunum

- Pros
  - Jejunal caliber similar to that of normal esophagus
  - Functions as reliable food transporter
  - Results in low incidence of leaks and strictures
  - Functions as an effective gastroesophageal barrier
  - Does not require a bowel preparation

Jejunum

- Cons
  - Length of conduit limited by blood supply
  - Infarction commonly resulting from passage through chest
  - Procedure more technically difficult
  - Requires 3 anastomoses
  - Has high peptic ulcer susceptibility
  - Blood supply lacking marginal artery
  - Has high failure rate

Complications

- Anastomotic leakage 14-21%
- Fistula recurrence 3-14%
- Esophageal strictures 40%
- Gastroesophageal reflux 40-70%
- Tracheomalacia 10-20%
- Esophageal dysmotility

Thoracoscopic Repair of Esophageal Atresia and Tracheoesophageal Fistula
A Multi-Institutional Analysis

George W. Holcomb III, MD, MBA,* Steven S. Rothenberg, MD,§ Klaas M. A. Bax, MD,†
Marcelo Martínez-Ferro, MD,§ Craig T. Albanese, MD,∥ Daniel J. Ostlie, MD,*
David C. van Der Zee, MD,† and C. K. Yeung, MD‡

- 104 patients with EA and distal TEF only
- Mean operative time 129.9 min, mean days of hospitalization 18.1
- 5 operations converted to open (4.8%)
- 11.5% developed early leak
- 31.7% required esophageal dilatation
- 24% required fundoplication later
- 3 deaths

Annals of Surgery 2005;242:422-430
Summary

- Once a death sentence EA and TEF has a survival rate approaching 100% with advances in neonatal care and pediatric surgery
- Primary esophageal repair is the accepted approach if possible
- The selection of esophageal conduits should be made on an individual basis