Esophageal Atresia and Treacheoesophageal fistula

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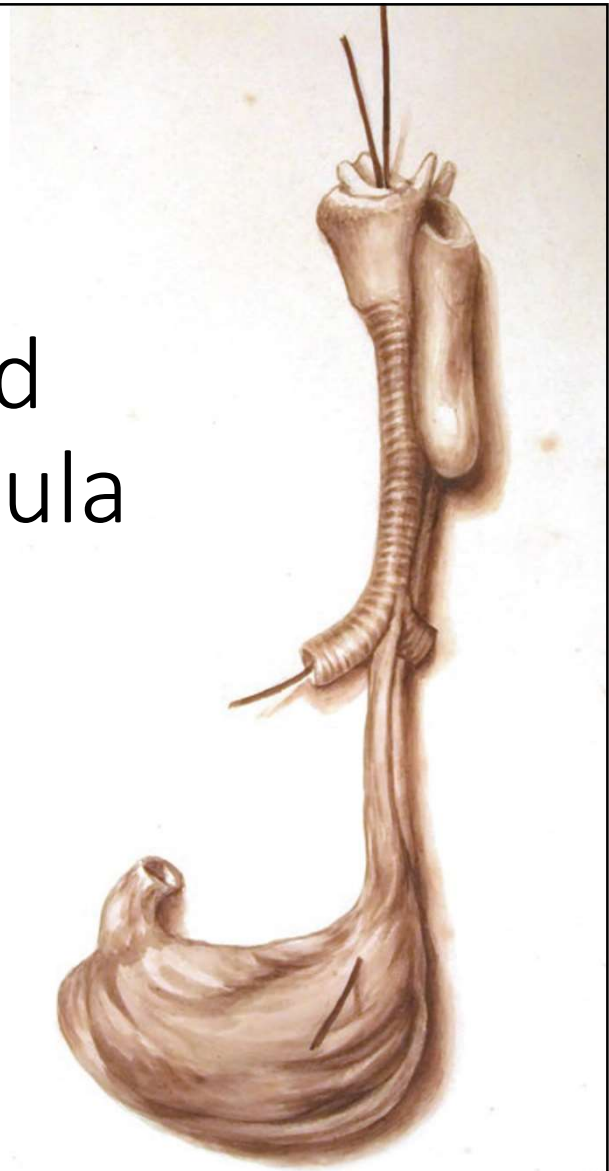

Introduction

- Repair of EA/TEF is one of **greatest landmarks** in neonatal surgery
- Advances in aesthesia and NICU have led to > 95% **survival**
- Current interest is focusing on **morbidity** and **quality of life** of survivors.
- **Debate** continues with regard best management of
  - Pure [long-gap] atresia
  - RAA
  - GERD
  - Stricture
  - Tracheomalacia

Outline

1. Embryology
2. Epidemiology
3. Diagnosis & Treatment
4. Operative details
5. Pure TEF (H-type)
6. Pure EA (long gap)
7. Outcome

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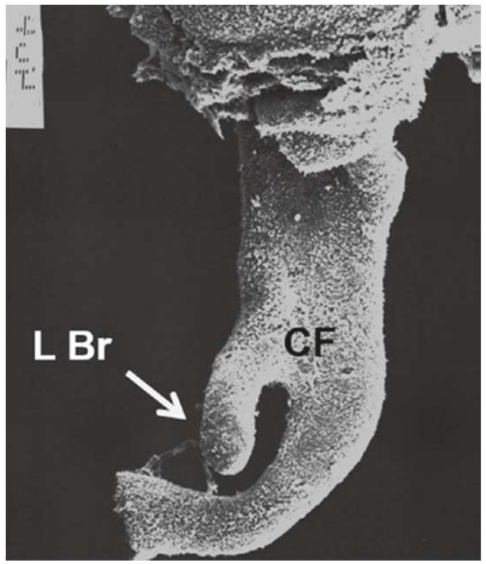
1. Embryology

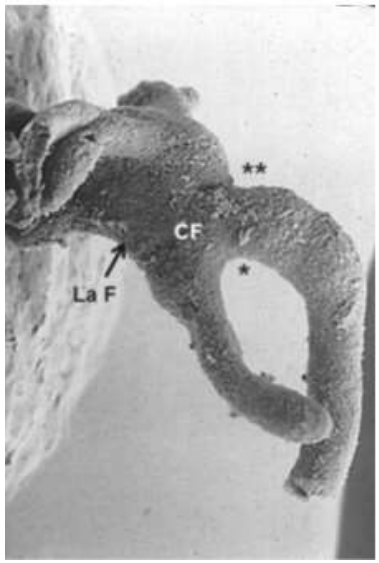
The embryology of the foregut is still subject to controversy. What is known is that during the 4th week foregut starts to differentiate into respiratory and esophageal part.

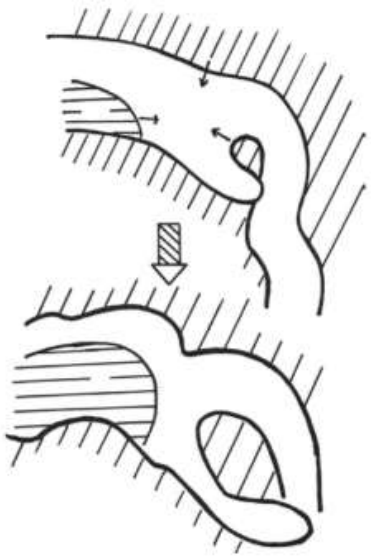
Separation of trachea and esophagus

Foregut is separated by 6th – 7th wk

- Traditional theory = lateral folds fuse to form septum
- SEM (chick embryos) = 3 folds approach each other to reduce foregut (don't fuse)
  - Dorsal fold = between esophagus and pharynx
  - Cranial fold = larynx fold
  - Caudal folds = tracheoesophageal fold







Separation of common foregut: seen in scanning electron microscope of normal chick embryos. Schematic drawing demonstrates the 3 folds approaching each other.

Embryologic theories of EA/TEF

No embryological theory successfully explains all the anatomical variants

- **Failure of fusion of TE septum**
- **Relative deficiency of tissue** (preferential incorporation of tissue into the trachea)
  - increased number of tracheal rings, longer trachea in the Adriamycin rodent model
  - association of **13 pairs of ribs** with long-gap EA

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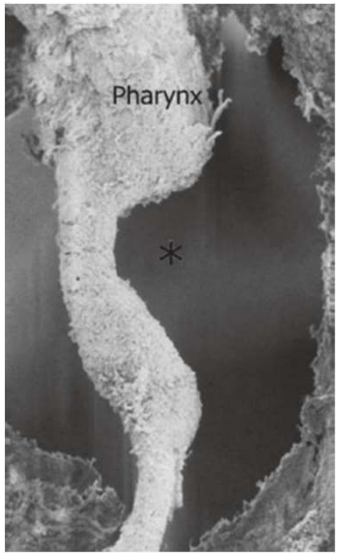

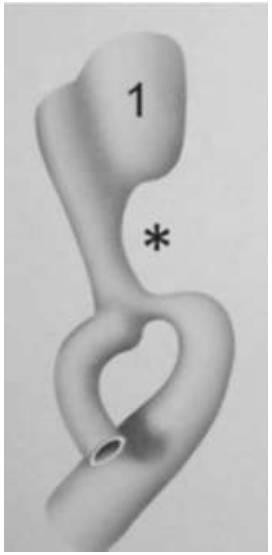

Embryologic theory of EA with TEF (chick model) : dorsal fold between pharynx and larynx grows too deep into the common foregut space.

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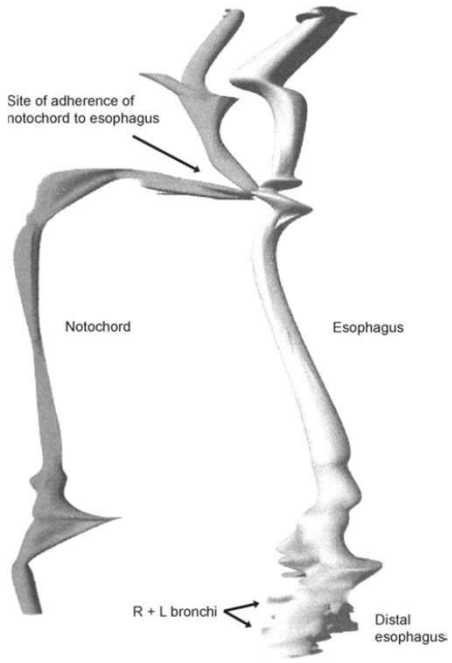

Embryologic theory of EA /TEF (Adriamycin rat model): Abnormal nothocord (Ventral misplacement and prolonged adherence to foregut) results in abnormal development of the mesenchyme

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Etiology

- **Genetic**
  - Three separate genes have been associated with EA/TEF: *MYCN, CHD7, SOX 2*
  - 10% incidence of chromosomal abnormalities (**trisomy 18** and **Down's syndrome)**
  - Risk for a 2nd child 0.5–2%, if >1 child is affected 20%, Vertical transmission is 3–4%.
- **Enviromental**
  - thalidomide exposure
  - methimazole in early pregnancy, prolonged use of contraceptive pills
  - maternal diabetes

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2. Epidemiology

EA/TEF presents in many forms and it should be thought of as a spectrum of anomalies. There are a large number of rare variants. It is not necessary to list them all but surgeon should be aware that bizarre variants do occur.

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Incidence

- 1 in 2500–3000 live births
- Slight male preponderance of 1.26 : 1
- More common in twin pregnancy
- Higher risk of **prematurity**

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Associated anomalies

50% is syndromic *(VACTREL, CHARGE, SCHISIS).* "H" type has least associated anomalies & pure EA highest

- Cardiac (~30%)
- Urogenital (~20%)
- Vertebral (6–21%)- thoracic vertebra
- Limb (~ 10 %)
- Anorectal (~10 %)
- **GI** (~10%)– duodenal atresia, IHPS
- **Tracheobronchial** variant (~50%)- trachoemalacia, laryngeal cleft, esophageal/tracheal stenosis, tracheal bronchus

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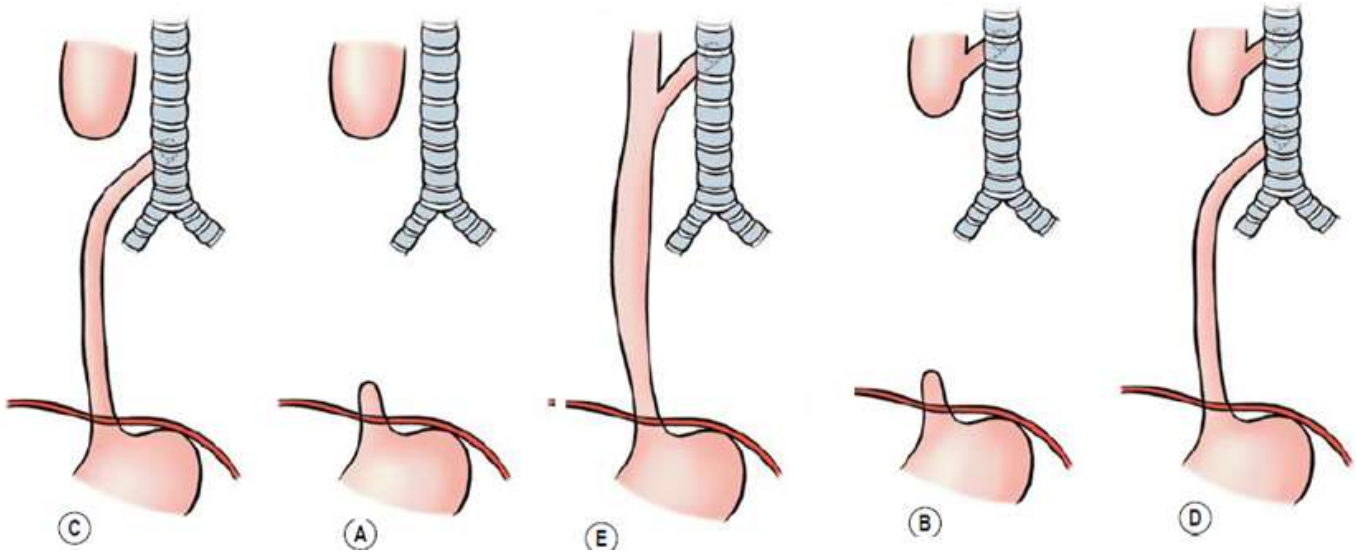

Gross classification (widely used): 85% Type C, 10% type A, 3% Type E (H- type), 2% Type B, <1% Type D

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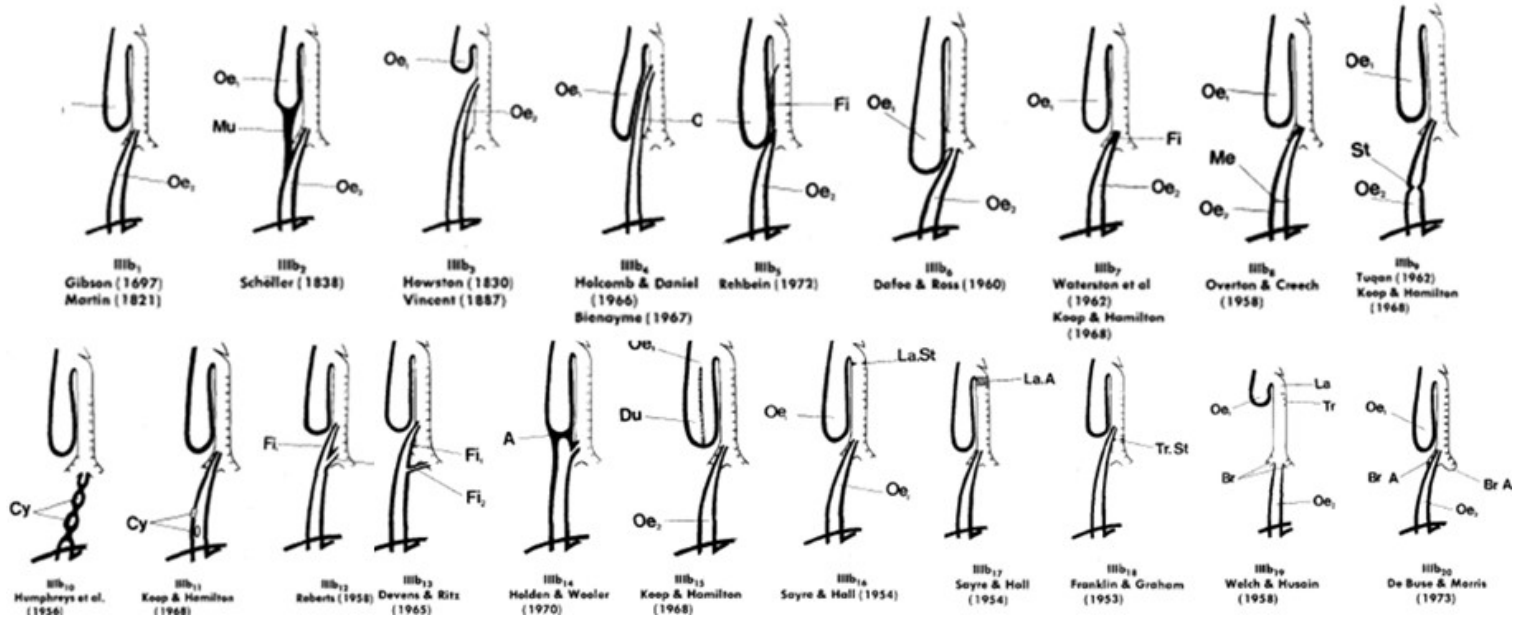

Kluth classification (most detailed): classifies EA into 10 types and ~100 subtypes.

20 subtypes of Kluth type IIIB/Gross type C (EA with distal TEF) : *short gap, log gap, high inserting fistula with short upper pouch, high inserting fistula with overlap, fistula is long atretic strand, long proximal pouch with overlap, fistula closed at connection to trachea, additional membranous atresia of distal segment, additional stenosis of distal segment, multiple cysts connected by fibrous strand replace distal esophagus, multiple cysts on distal esophagus, two fistulas to trachea, one fistula to trachea and one to bronchus, esophageal continuity maintained externally, duplicated proximal esophagus, stenosis of larynx, tracheal stenosis below entrance of fistula, high atresia, atresia of bronchi*

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3. Diagnosis and Treatment

Diagnosis is confirmed when 10 F tube can't be introduced beyond 10 cm from the gums. Confirmation with plain X-ray is not essential for the diagnosis.

Antenatal Diagnosis

- **Detection rates vary** widely in fetal medicine centers (9–24%).
  - Rarely diagnosed antenatally unless pure atresia
  - high rate of false positive scans (50% later proven not to have EA after birth)
- **Signs**
  - polyhydramnios (50%)
  - absent or small stomach bubble
  - dilated cervical esophagus (pouch sign)
- Theoretically **reduce inadvertent newborn feeding** and aspiration pneumonitis

Clinical presentation

presents in the immediate post partum period

- **Excessive secretions**
- **Chocking** with attempts at feeding
- Respiratory **distress**, cyanosis
- **Failed attempts to advance a 10F tube** (NGT/OGT)
    - *Rarely tube may pass through the trachea-fistula -stomach giving a false impression of normal
- **Examine perineum** for ARM

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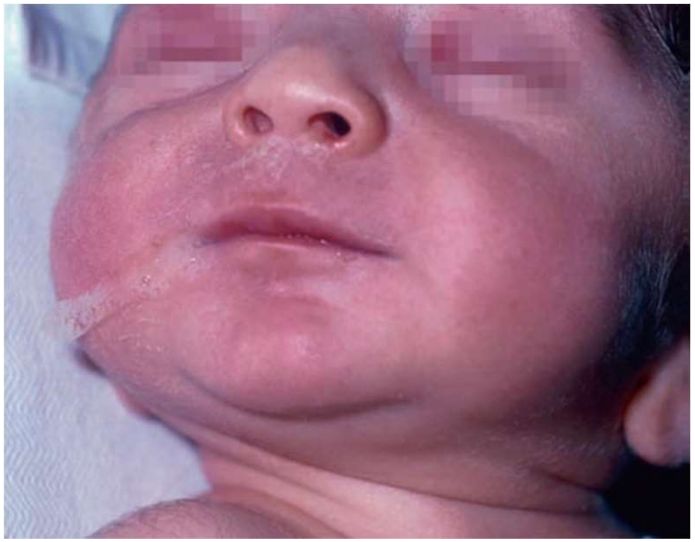

“mucousy baby”: salivate excessively because saliva accumulates in the blind upper esophageal pouch

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Radiology

- **Plain radiograph** including chest and abdomen (with downward pressure on stiff tube)
  - pouch-like gas filled lucency in neck. Tip of tube curled back / high in the neck
    - If tip passes beyond level of carina, the diagnosis should be questioned.
  - Fistula confirmed by presence of bowel gas
    - rarely, the distal fistula may be occluded or is a thin fibrous connection, leading to misdiagnosis
  - Vertebral anomalies and cardiomegaly
- **Contrast swallow** if suspect H-type fistula
- Some advocate **prone tube esopagogram** and **endoscopy** if contrast swallow is normal
- **Ultrasound** (head, spine, renal) can be deferred after surgery

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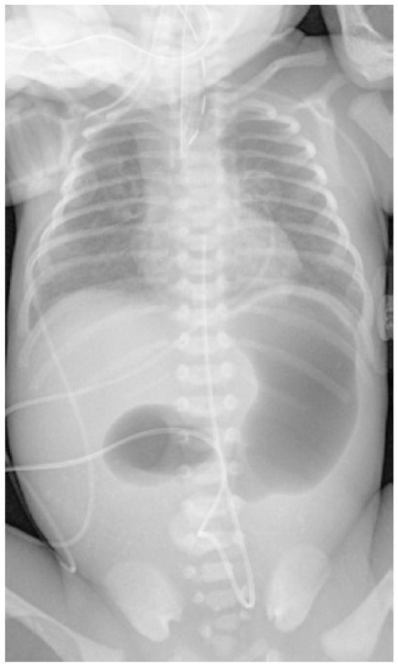
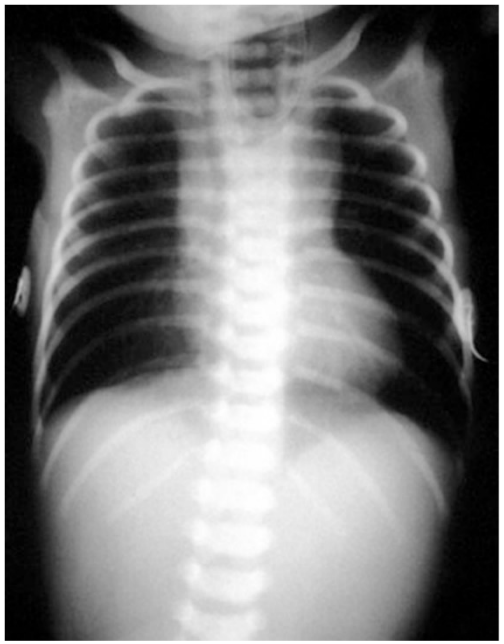


EA with TEF and duodenal atresia



Pure EA

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Echocardiography

Associated cardiac anomalies and laterality of aorta

- Associated **Right-sided aortic arch / right descending aorta** (~ 3%)
  - Occur frequently with **long gap**
  - **Usually undiagnosed** preop (may suspect on x-ray, Echo only has 20% accuracy)
  - May obscure TEF and lies at level anastomosis will be made
  - Approach through left thoracotomy (**High leak rate** ~40% when approached via right side)
  - If found at time of surgery: change sides if thoracoscopy. Continue on rt side if thoracotomy

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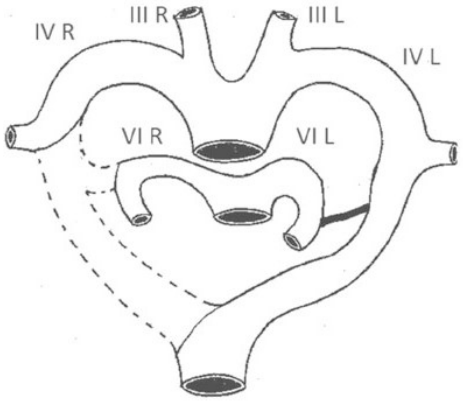

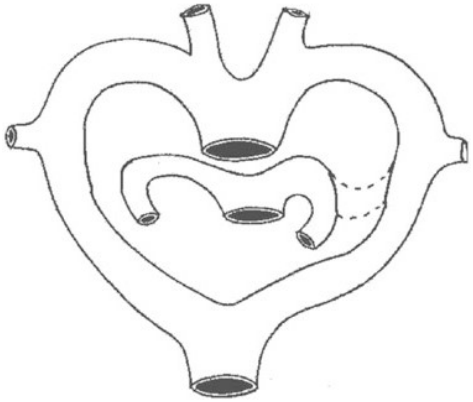

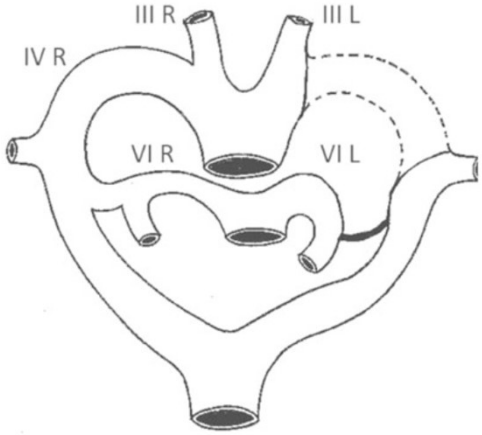

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Aortic arch anomalies: Normal left side arch, Double aortic arch, Right aortic arch with right descending aorta, Left aortic arch with right descending aorta

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Initial care

- Prevent **aspiration of unswallowed saliva**
  - Position head up in the lateral position
  - 10 Fr Replogle tube in the upper pouch
    - connect to continuous suction at low pressure (35-40mmHg)
    - Frequent irrigation of the air channel to prevent blockage

  *If Replogle not available use wide feeding tube but aspirated every 15 min during transfer
- Intravenous access with careful **fluid management** (aspirated saliva should be replaced)
- broad spectrum **antibiotics**, **Vitamin K**, prepare **blood**
- **Thermoregulation**

Stable Infant

- Surgery best performed
  - In first 24 h of life (avoid pneumonitis from reflux through fistula & aspiration of saliva)
  - scheduled procedure (during normal working hours)
- Additional pathology (duodenal atresia/ARM) can be dealt with under the same anesthesia
- Minimally invasive surgery
  - Advantage in scar & musculoskeletal morbidity (notablywinged scapula)
  - widely debated and best reserved for MIS master enthusiast

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The unstable infant

- **Significant distension** (diaphragmatic splinting and compromise ventilation)
  - Transpleural thoracotomy with fistula ligation (If stable continue with repair of esophagus)
    - second operation in 7–14 days because recanalization can occur
  - gastrostomy to decompress the stomach If the infant is extremely unstable
    - can result in a significant loss of tidal volume and may create respiratory issues as well.
- **gastric wall perforation** with sudden deterioration
  - Needle paracentesis to relieve tension pneumoperitoneum
  - laparotomy, repair of the stomach perforation and feeding gastrostomy

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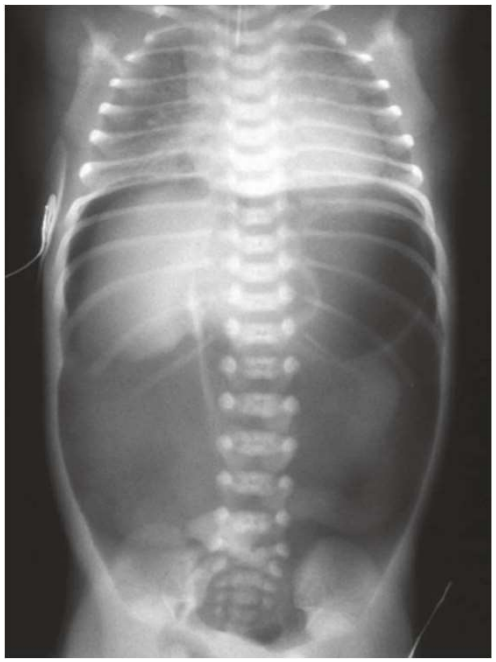

Premature infant with esophageal atresia and gastric perforation: note severe hyaline membrane disease and tension pneumoperitoneum

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Anesthesia

- Single lung ventilation if possible
    - *if not possible right lung collapse is achieved with CO2 insufflation for thoracoscopy.
- forceful ventilation must be avoided, to prevent gastric distention and perforation
- Control fistula prior to ligation
    - Pass fistula in mid-trachea with ETT
    - Fogarty prior to ligation

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Bronchoscopy

Routine use of rigid bronchoscopy is debated b/c simultaneous proximal and distal fistula is <5%.
However, many use 2.2 flexible bronchoscopy through the endotracheal tube or LMA.

Advantage

- to help plan the repair (gap estimation)
- identify distal fistula (pass guidewire)
- occlude the fistula (placement of a “blocker”)
- Identification of proximal fistula
- Identification of laryngo-treacheo-bronchial variant anomalies

Disadvantage

- Prolong the procedure
- infant can decompensate prior to ligation of the fistula
- gastric fluid may reflux through a fistula into the trachea during manipulations

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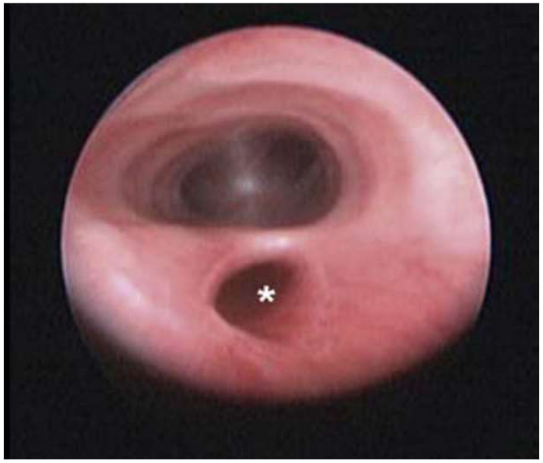
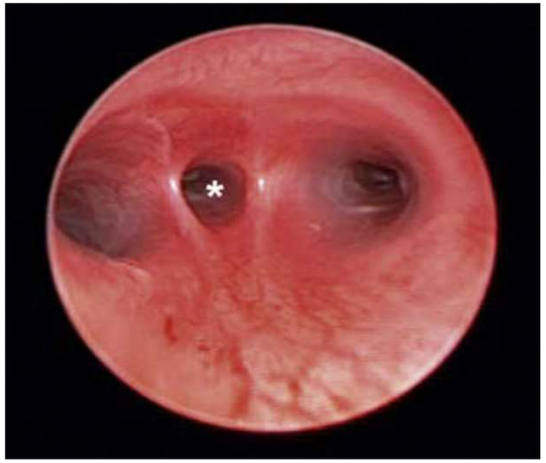
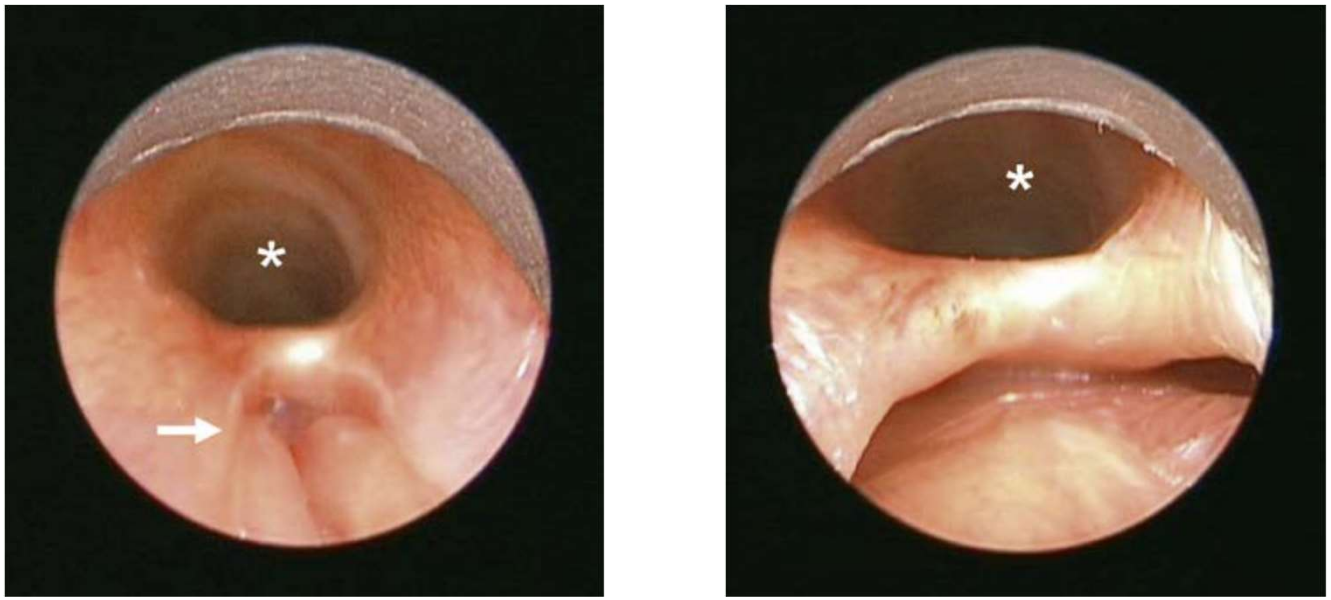




Bronchoscopy showing TEF: TEF at mid trachea and TEF at carina



Bronchoscopy showing laryngotrachoesophageal cleft

4. Operative Details

“To anastomose the ends of an infant’s esophagus, the surgeon must be as delicate and precise as a skilled watchmaker. No other operation offers a greater opportunity for pure technical artistry.”

Willis Potts

Anatomic considerations

- **Innervation of esophagus**
  - Autonomic – splanchnic, vagus
- **Blood supply of esophagus**
  - Upper segment - branches of the inferior thyroid artery (branch of thyrocervical trunk) running vertically downward (can be fully mobilized without ischemia)
  - Lower segment - segmental esophageal branches from the aorta (deficient in EA), ascending branch of the left gastric artery

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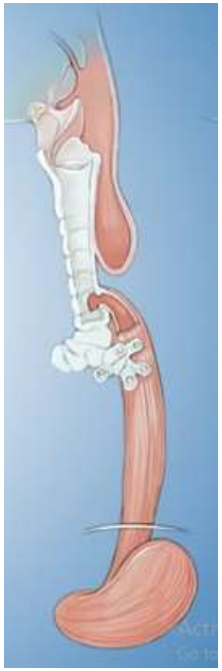
Anatomic considerations: EA with TEF

- **Proximal blind ending pouch**
  - very dilated, thick walled
  - Found between T2-T4, within 1 cm of the arch of the azygos vein
  - identified with anesthetist introducing a stiff catheter into the esophagus.
- **Distal segment with fistula on posterior trachea**
  - Slender, thin wall
  - enters trachea posteriorly at carina or 1–2 cm higher (mid trachea)
  - identify by recognition of vagus over its surface & rhythmic distention with ventilation
    - *Rarely difficulty can be encountered in locating the distal esophagus, and it is quite possible to mobilize the descending aorta in the erroneous impression that it is the esophagus.*
- **Trachea =** Deficiency of cartilage and increase width of the membranous (tracheomalacia)



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Intra-operative considerations

- **Extra-pleural approach**
  - aid in exposure as it is easier to retract the lung when it is incased in the pleura
  - possibility of avoiding chest drain insertion
  - potential containment of any leak/soiling within the extra-pleural space
- **Division of Azygus**
  - some surgeons elect to preserve it (association with anastomotic leak has been suggested)
  - some advocate temporary occlusion before ligation (venous return may rarely be dependent on azygos)
- **Trans anastomotic tube**
  - helps protect the lumen from inadvertent closure and allows for gastric decompression.
  - Others feel it causes an increased risk of esophageal stricture and choose not to use it.

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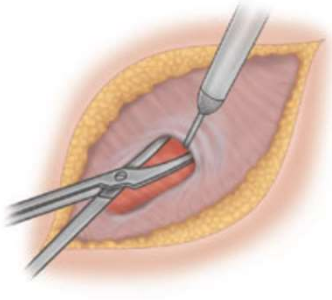

Classical operation with muscle sparing thoracotomy: lateral decubitus position on side opposite the turn of aortic arch. A small axillary role is placed under the chest to enlarge intercostal spaces. Ipsilateral arm is positioned over the head of the patient. A slightly curved 4–5 cm incision is made 1 cm below the tip of the scapula. Auscultatory triangle is opened and the muscles are retracted (latissimus dorsi posteriorly and the serratus anterior anteriorly). If serratus muscle needs to be transected, this should be done as low as possible to preserve the long thoracic nerve.

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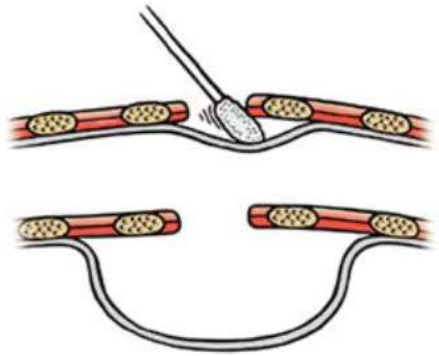

The fourth or fifth intercostal space is entered: A peanut or sterile cotton swab used to gently push the pleura away from the chest wall. Finochettio rib retractor inserted. Great care as it is easy to create a pleural tear in the anterior aspect. If a significant pleural tear occurs during the dissection it is wise to open the pleura widely converting to a transpleural approach.

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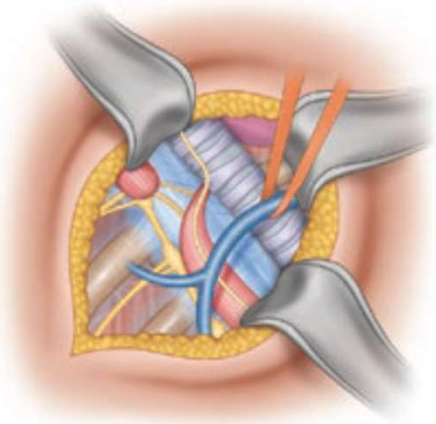

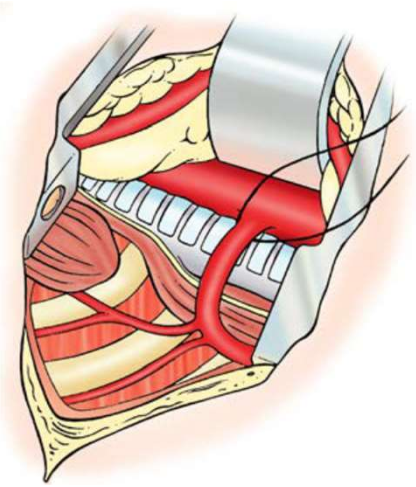

Retract the posterior mediastinal pleura forwards with a malleable retractor: Azygus vein is visualized as it enters the superior vena cava. Ligation of the azygos vein between 3-0 or 4-0 absorbable ligatures or simply cauterized and divide. If fistula originates more cephalad on the trachea, the vein can be left intact.

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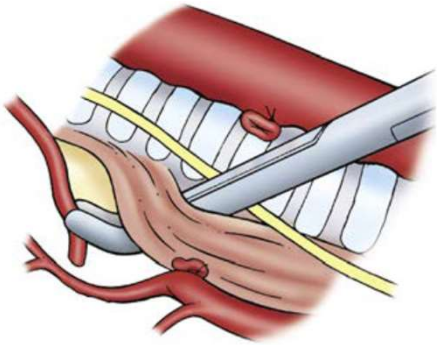
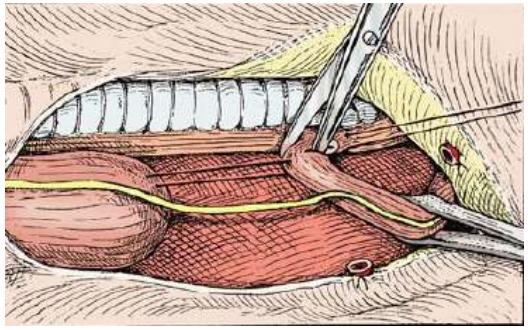
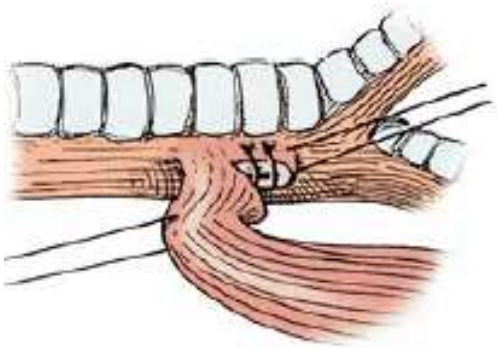
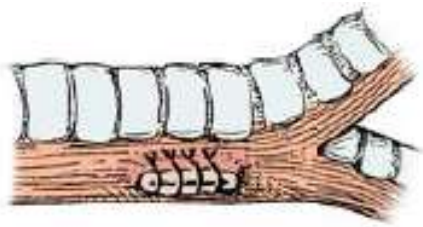

Distal esophagus is identified and looped: Traction on the loop controls the fistula and enables its junction with the trachea to be located precisely. The distal esophagus is secured with a stay suture. Divide the fistula sharply close to the trachea, to spare as many vagal nerve branches as possible. It is possible to suture ligate the fistula or to divide the fistula in stages and apply interrupted 5-0 or 6-0 monofilament prolene sutures. Adjacent tissue, if available, is tacked over the closure. The integrity of the TEF repair is checked by instilling saline into the thoracic cavity and exerting airway pressure.

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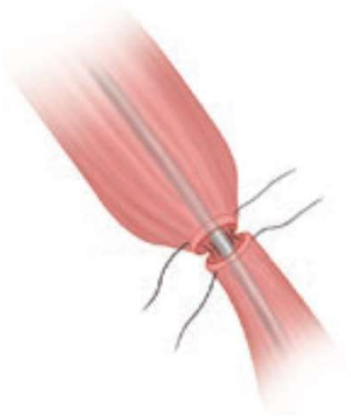

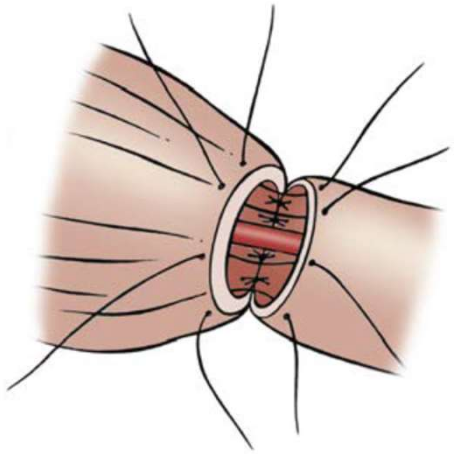

Stay suture taking a good bite of the top of the muscular proximal esophageal pouch: Take great care to avoid excessive tissue handling and trauma with fine forceps. Using traction the proximal pouch can be freed posteriorly and laterally by blunt dissection. Anteriorly, dissect sharply, staying on the esophageal side. Extensive dissection is not warranted, unless there is a long gap. The tip of the pouch is amputated so that the lumen and mucosa become visible. End-to-end anastomosis is performed with 5-0, absorbable sutures, including both the mucosa and the muscular wall. Posterior and lateral wall sutures have knots inside. Pass NGT through anastomosis before anterior wall anastomosis.

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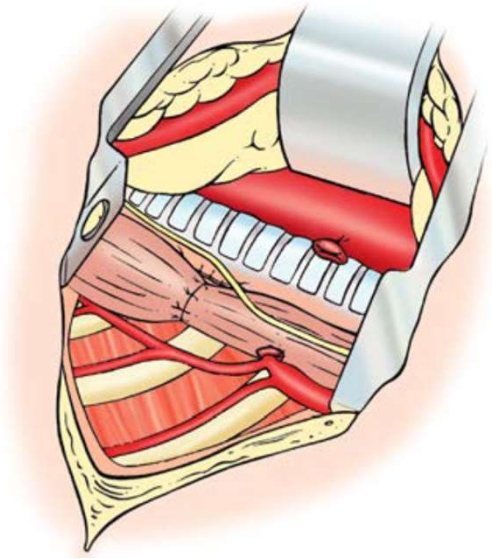

Chest incision is then closed in layers. The ribs should be approximated with one 3-0 absorbable suture.

5. Pure TEF (“H” type)

Has different symptoms from esophageal atresia because the esophagus is patent. However has presumed common etiology.

Diagnosis: “H-type”

- Often present **immediately after birth** with feeding.
    - Choking
    - Abdomen is distended with air, and flatulence may be present
    - Symptoms subside when the child is fed by a nasogastric tube.
- Can present at **older age** with recurrent pneumonia
- A water-soluble, low osmolar **contrast**
- Diagnostic **Bronchoscopy** (contrast studies misses 50%)

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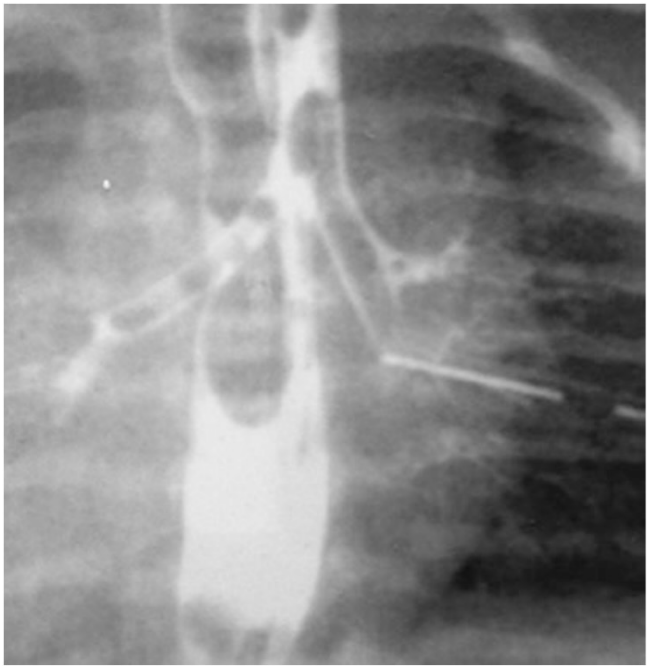
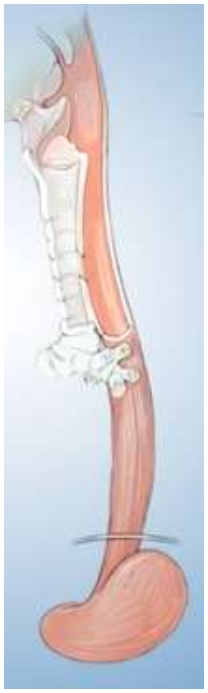


Tube esophagogram in H-type

Surgical anatomy: “H-type”

- Fistula is usually higher in the neck (C5-C7) or at thoracic aperture (T1-T3)
- Fistula is short
- Fistula runs from the trachea proximally, downward to the esophagus



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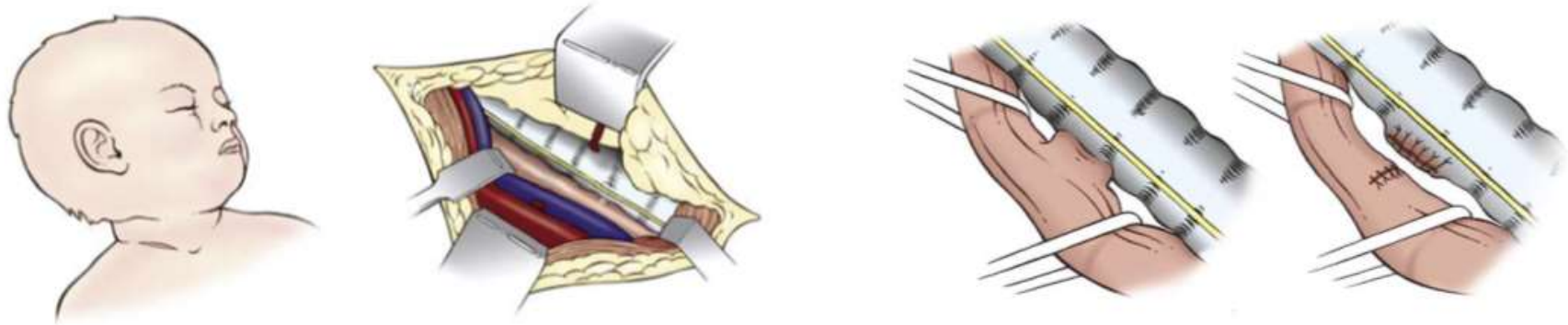
Intra-operative considerations: “H-type”

- **Guide wire** is placed through the fistula at tracheoscopy. Light of flexible scope can also aid identification
- A **cervical approach** can be used in most (80%)
    - lower fistulas can be pulled up and approached through the neck as well.
- **Thoracoscopically** repaired H-fistulas have been described
    - Depending on the location of the fistula (preoperative imaging), and the surgeon /parent preference.



Classic approach through low right cervical incision (finger's breadth above clavicle): positioning the neck in extension with a roll placed under the shoulders. Sternal head of the SCM is transected/split. Internal jugular vein and carotid artery mobilise laterally to expose the cervical esophagus and trachea. Esophagus is dissected from trachea and slung with vascular sloops both above and below the fistula tract. Visualize RLN to avoid injury (Lt RLN is also vulnerable).Traction sutures are placed at the upper and lower ends of the fistula. Fistula is transected close to the esophagus. Trachea is closed longitudinally with 5-0 or 6-0 prolene and the esophagus transversely with interrupted 5-0 or 6-0 PDS. Sternal head of the SCM can be interposed in-between.

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6. Pure EA (long gap)

“Biological solutions are the best because in pediatrics we need the best end result… An esophagus that child can use 70+ years “

John Foker

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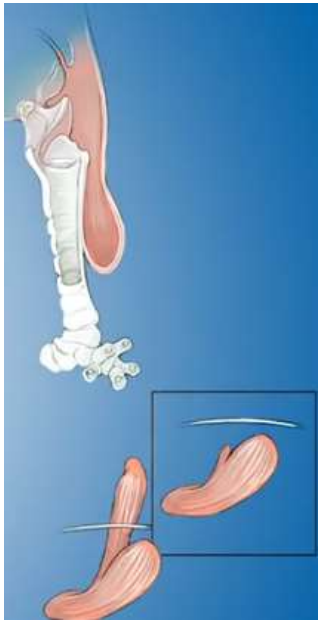
Surgical anatomy: Pure EA

- **Upper esophagus**
  - Ends at the level of the azygos vein.
- **distal esophagus**
  - is short and often suspended by a fibrotic band.
  - May even be below diaphragm



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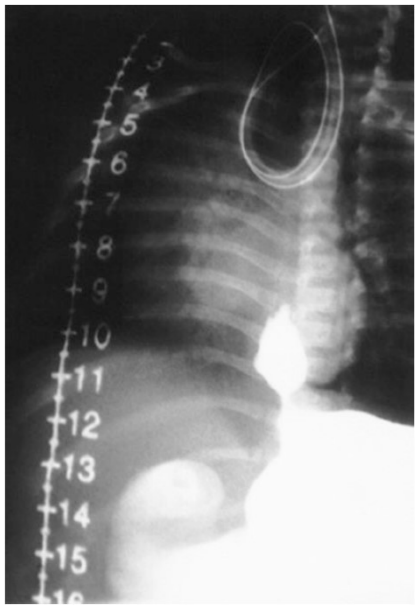
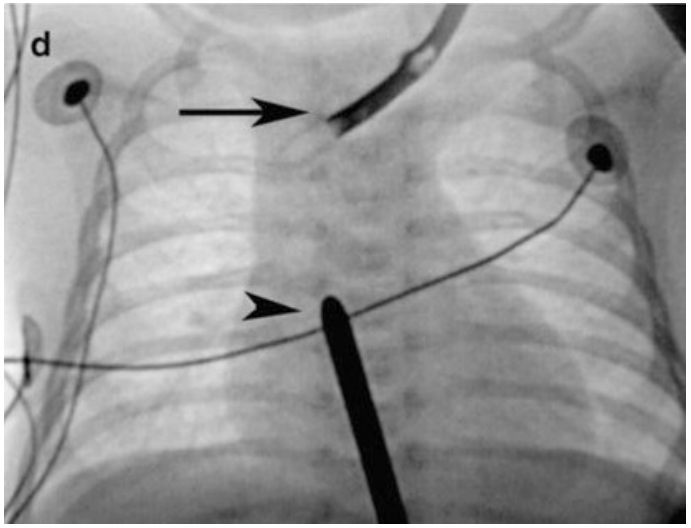
Initial surgery

- **Bronchoscopy + Gastrostomy**
  - Incidence of proximal fistula in absence of distal fistula is high (50%)
- **Thoracoscopic Exploration**
  - Can be considered if upper pouch appears long
  - Gap assesment, mobilization and Foker procedure can also be performed via thoracoscopy
- **Assessment of the gap** (at time of gastrostomy or 7-10d later)
  - ? at time of gastrostomy, after 7-10d, after 3-6 wk
  - **Radiologically** (gapogram)





Gapogram: Gap assessment in pure EA using contrast instilled via the gastrostomy. Metal sound or Flexible endoscopy (under general anesthesia) can also be used once tract is sufficiently mature. This procedure can be repeated 2–3 weekly intervals, to assess whether the ends of the esophagus are sufficiently close together. Gap is expressed in terms of the number of vertebral bodies, thus taking the child's length into account. Distance of < 2 vertebral bodies is ideal for anastomosis.

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Subsequent approach

Delayed primary anastomosis (use native esophagus)

- Avoid cervical esophagostomy = demands meticulous nursing care, regular suctioning, chest physiotherapy.
- inability to establish oral feeds (aversive feeding behavior)
- Don't delay surgery > 3 mo
  - Try intraop lengthening techniques
  - Cervical esophagostomy ("Spit stoma") if it fails
  - Delayed replacement at 18 mo

Early Esophagostomy (delayed replacement)

- Spitz has recommended assessing the gap at time of gastrostomy
  - > 6 vertebral bodies prompt esophagostomy
- permits early sham feeding,
  - development of the learning skills needed for feeding and later speech acquisition.

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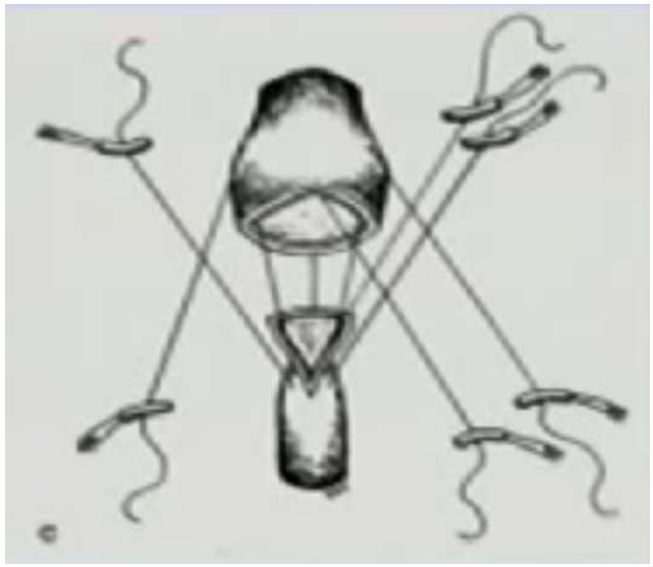
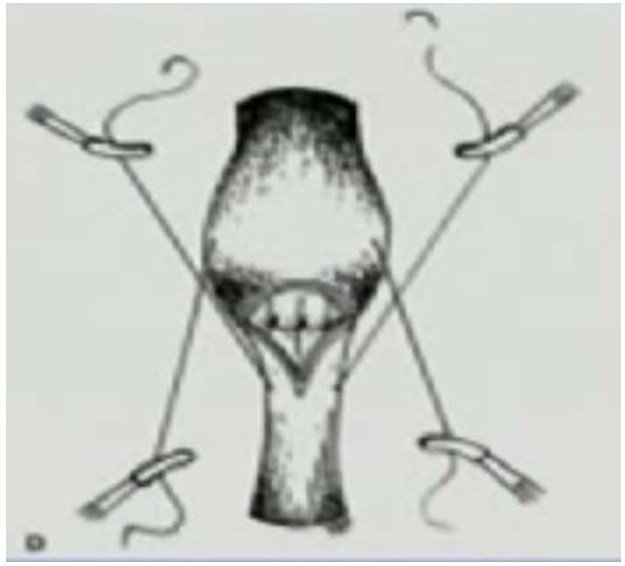
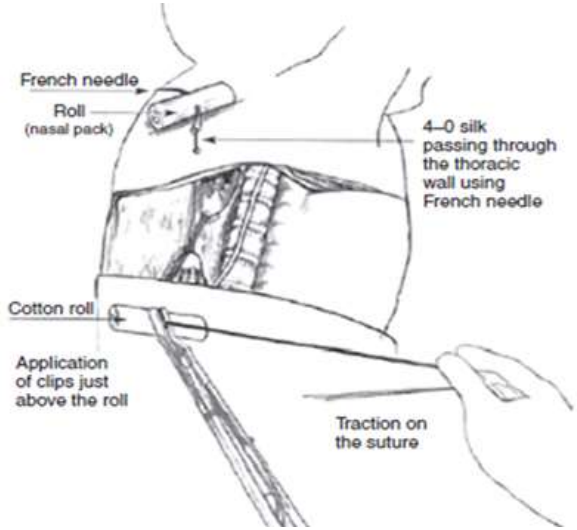
Esophageal lengthening procedures

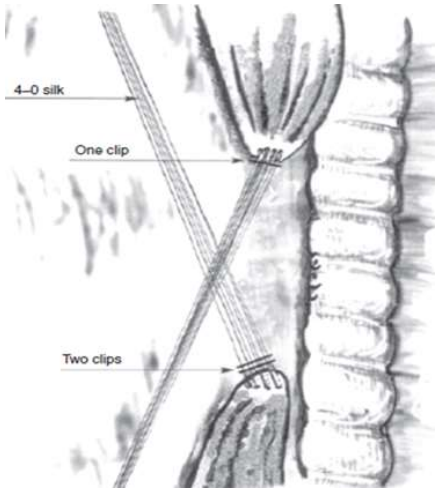
- **Prior to definitive surgery**
  - Spontaneous growth (4–8 weeks)
  - Bougienage
  - External traction (Foker process)
  - Extrathoracic esophageal elongation (Kimura technique)
- **During definitive surgery**
  - extensive mobilization of the proximal and distal esophagus
  - Myotomy of upper pouch
  - Flap lengthening of upper pouch
  - Elongation of the lesser curvature





Anastomosis under tension: pre-place all sutures in back wall and slowly bring them together. Then tie individual sutures off tension. (distribute tension gradualy). A well constructed anastomosis withstands tension, rarely leaks.





Fokers Method (External Traction): Mechanism is stimulating growth, not stretch. If distal esophagus below diaphragm, follow vagus nerve to left side to develop the hiatus. 4 Traction sutures in wall (outside lumen) of esophagus. Follow location of clips. Repair when cross each other. Can grow upto 50-70x

Moderate gap = traction in OR ~10 min.

Longer gap = internal traction 5-6 d.

very long gap = Bring outside chest wall for external traction (1-3 wk)

no apparent distal esophagus = traction > 3wk (the smallest primordium, even 3 mm grow).

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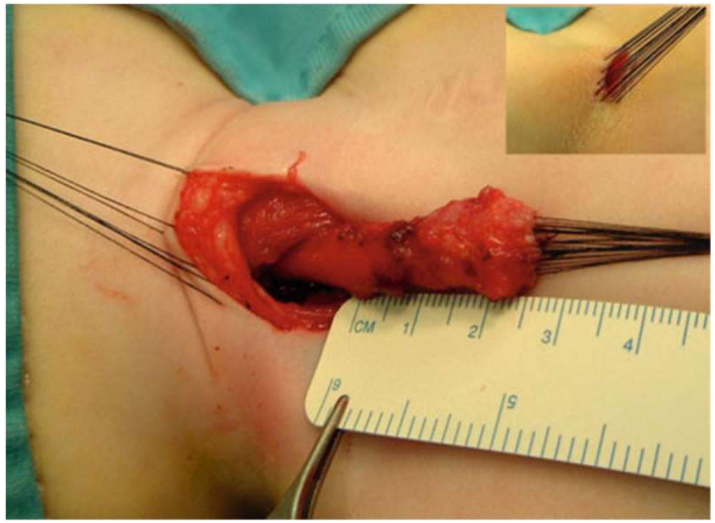

Kimura (Extrathoracic Esophageal Elongation of upper pouch) : upper esophagus brought to subcutaneous tissue of anterior chest wall. After 30 days the procedure is repeated—the esophagostomy is dissected, mobilized, and exteriorized a few centimeters below the previous one. Goretex® sheet is wrapped around the upper esophagus to help subsequent dissection. At each elongation, the neck is fl exed and the esophagus gently stretched caudally and anchored to the pectoralis major fascia with 2 or 3 absorbable stitches. Thoracotomy, prolonged sedation, and muscle paralysis are not needed. Usually used as an esophageal “rescue” in patients who have been treated with cervical esophagostomy.

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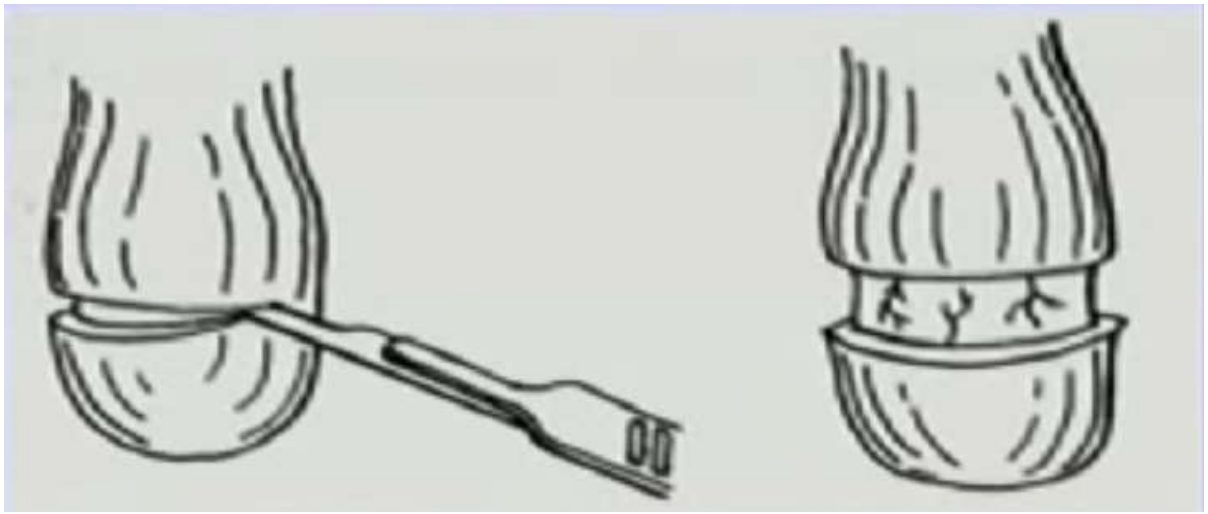
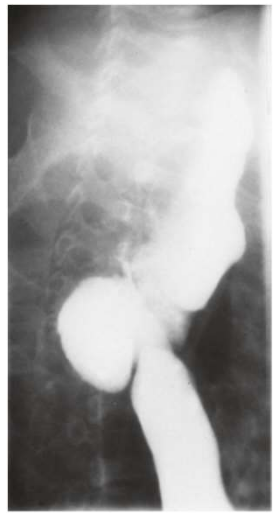
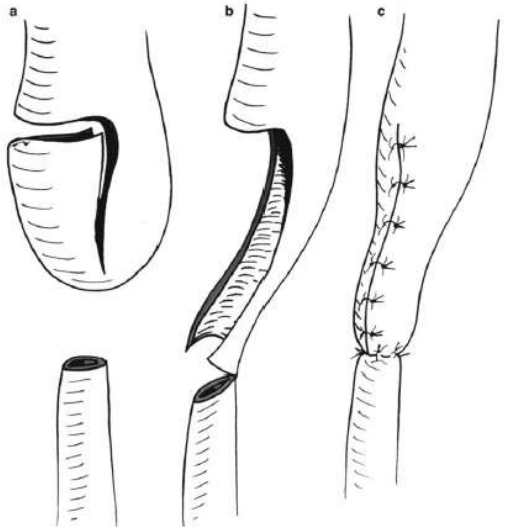
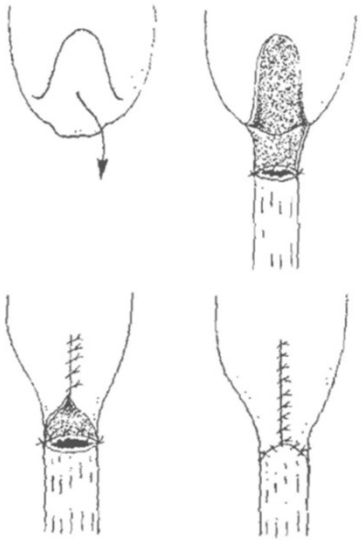




Upper pouch circular myotomy: Foley catheter passed by the anaesthetist via the mouth to the upper pouch. myotomy over the inflated balloon of complication is large diverticulum in unsupported esophagus





Upper esophageal flap: Anterior horizontal incision of the upper pouch prolonged caudally on both sides. The flap is reversed, brought down and anastomosedto the lower pouch.

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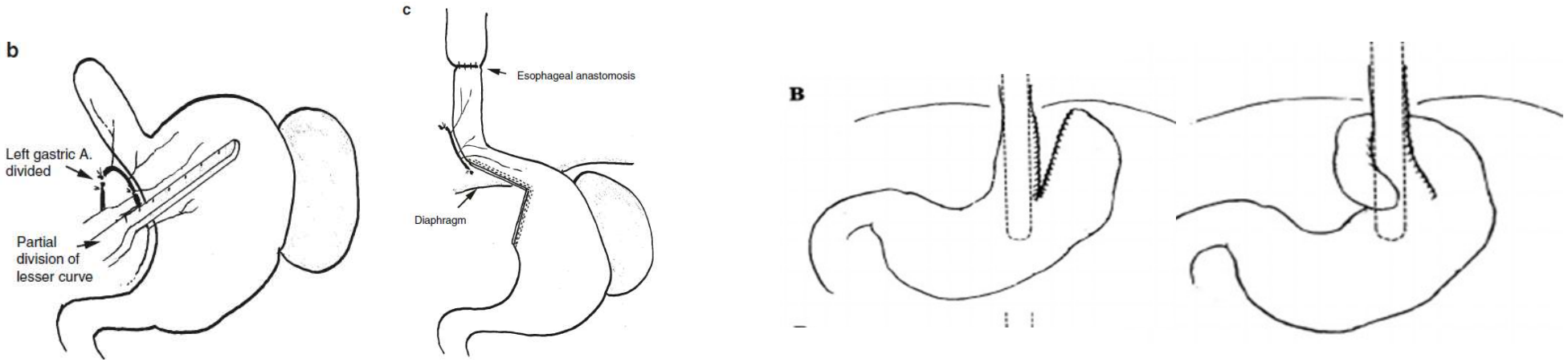

Scharli and Collis gastroplasty: Laparatomy and lesser curve lengthening procedures

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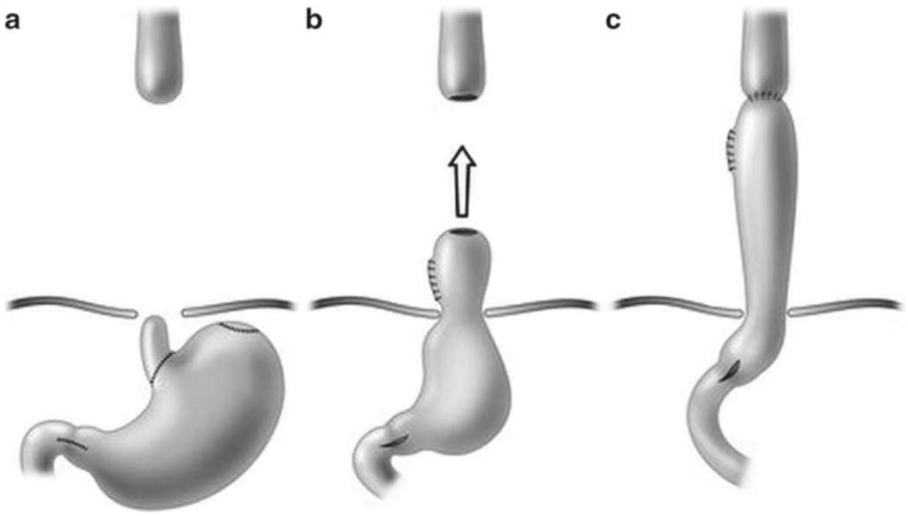
Esophageal replacement

- Colon
- Stomach
  - Gastric tube
  - Gastric transposition
- Jejunum
  - Pedicle graft
  - Free graft



Gastric transposition (pull-up): popularized by Spitz and has the merit of simplicity. vascular supply is based on the right gastric and gastroepiploic vessels. After pyloroplasty, the stomach is routed through the posterior mediastinum via the diaphragmatic hiatus and the esophago-gastric anastomosis completed in the neck. Complications are dec reservouir function, dumping, poor nutrition, anemia, cervical esophageal reflux and baretts esophagus, atrophic gastritis. Leakage and stricture from the neck anastomosis is reported in 12%

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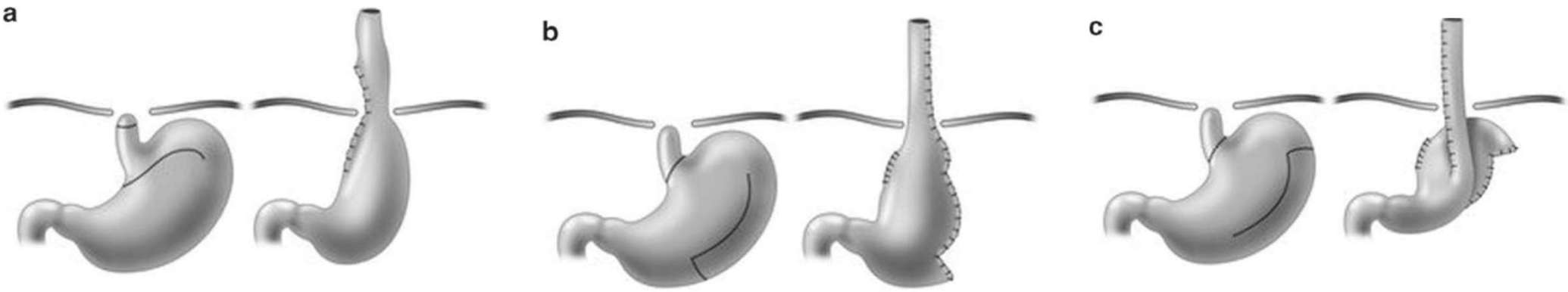

Gastric tube interposition (tube fashioned from lesser or greater curvature) : based on the left gastroepiploic (ante-peristaltic) or right gastroepiploic arcades (iso-peristaltic) preferentially using the posterior mediastinum as a route to the neck anastomosis. Complications are Reflux, cervical baretts esophagus, Long suture lines are susceptible to bleeding and leaking.

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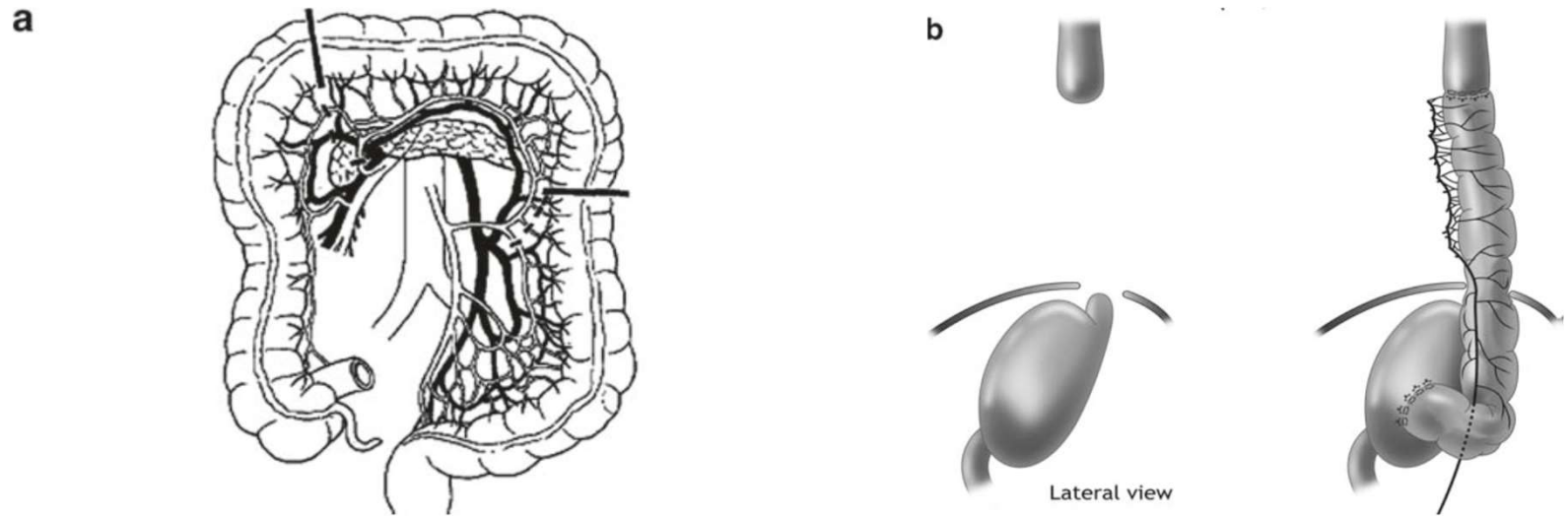

Colon interposition (commonly used) : Colon conduit may be isoperistaltic (left colon based on the left colic vessels), or anteperistaltic (right colon based on the ileocolic vessels). The colon is best routed via the posterior mediastinum (via the hiatus), or through right or left chest cavities to the neck. Complications are No peristalisis tend to dilate and kink. Recurrent aspiration as the new food pipe created with colon is 'non refluxing'. Cervical anastomosis is prone to leak.

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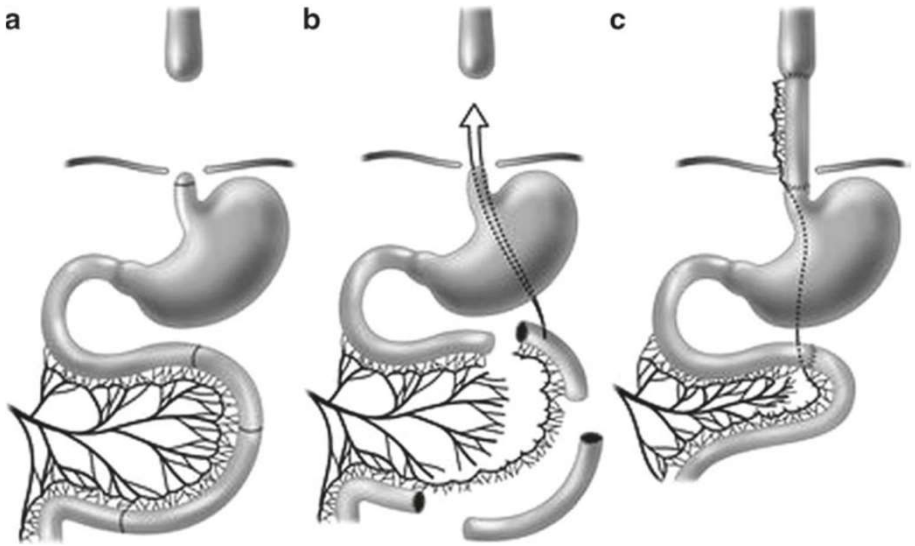

Jejunal interposition: divide aracades to straighten. Free graft requires microvascular anastomosis (Supercharged). Technically difficult, has motility but there's still problem with redundancy and dilation. do not dilate excessively and retain good peristaltic function which may explain why GER does not appear problematic for some of the patients

7. Outcome

EA/TEF used to be a condition incompatible with life but by the 1980s pediatric surgery units in the developed world were achieving spectacular outcomes with 85–90% survival. Current focus is on reducing morbidity and improving quality of life.

Outcome in Ethiopia

NEONATAL GASTROINTESTINAL SURGICAL EMERGENCIES: A 5- YEAR REVIEW IN A TEACHING HOSPITAL ADDIS ABABA, ETHIOPIA

Endale Tefera,MD[1], Telahun Teka, MD[1], Milliard Derbew,MD[2]

- 1997-2001, 12 patients
  - 3-repair, 6-gastrostomy, 3-pre-op
  - **91% mortality** (11/12)

Assessment Of The Outcomes Of Neonates With The Diagnosis Of Esophageal Atresia And/Or Tracheoesophageal Fistula Admitted To Tikur Anbessa Specialized Hospital-Neonatal Intensive Care Unit

MOGES AMARE, MD*, ABEBE HABTAMU, MD*

- 2008-2013, 34 patients
  - 17-repair, 4-gastrostomy, 13-pre-op
  - **85% mortality** (29/34)

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Post operative care

- **Mechanical ventilation** with muscle relaxation for 3-5 days
  - For an anastomosis performed under considerable tension. (evidence is lacking)
  - For H-type fistula (high risk of tracheal edema and RLN injury)
- **H2-antagonists** (ranitidine)
  - prophylactic therapy in an effort to reduce the risks of anastomotic stricture (author's opinion)
- **Chest drain** is optional
  - For transpeural and For anastomosis under tension
- **Feeding** can be started early (48 h) through NGT and slowly inc as tolerated
  - if no evidence of a leak
- **Contrast esophagogram** is optional (5–7 days to evaluate the anastomosis)
- **Antibiotics, Analgesics**
- **Discharge** when able to eat enough to maintain weight

Risk factors for poor outcome

- Low birth weight
- Associated anomalies
- Respiratory distress syndrome and pneumonia
- preoperative ventilator dependence
- Variant anatomy (pure atresia / long gap)
    - Quality of life metrics were also more favourable in patients who had native esophageal repair

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Waterston's risk stratification (outdated due to advances in NICU)

	Birth weight, pneumonia and congenital anomalies	Survival?
A	- >2.5 kg and healthy	95 %
B	- 2.0-2.5 kg and healthy - >2.5 kg and moderate pneumonia / anomalies	68 %
C	- < 2.0 kg - Severe pneumonia - severe anomalies	6 %

	Moderate	Severe
Anomalies	Limb anomalies, cleft lip/palate	Associated GI atresia, Severe renal anomalies
	ASD or small PDA	Transposition of great arteries
Pneumonia	Limited to one lobe	Changes in multiple lobes

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Spitz risk stratification (for the modern era)

	Birth weight and congenital heart disease	Survival
A	- >1.5 kg and no major CHD	97 %
B	- < 1.5 kg or major CHD	59 %
C	- < 1.5 kg and major CHD	22 %

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Anastomotic leak (3.5 – 17%)

Risk factors

- **Operative technique**
  - Incorrectly placed /Insecure sutures
  - Sutures “cut through” esophagus, Suture material (Silk)
- **Ischemia**
  - Excessive mobilization/ tension (Gap >4 cm)
  - Esophageal myotomy
- **Infection**

Anastomotic leak (3.5 – 17%)

- **‘radiological’ leaks**: Incidental finding on postoperative contrast study, no clinical symptoms
  - no clinical significance (do not preclude the infant from being offered feeds)
- **Minor leak**: pneumothorax/ saliva in chest drain, but infant is well
  - Cease oral feeds, Antibiotics, TPN
  - Will close spontaneously
- **Major leak**: major disruption of anastomosis on imaging, mediastinitis, abscess, pneumothorax, empyema
  - Cease oral feeds, Antibiotics, TPN
  - Some prefer conservative with prolonged chest tube if transanastomotic tube is in place
    - If fails (uncontrolled sepsis) cervical esophagostomy is essential (committed to replacement)
  - Others advocate early re-exploration (<48 h) with satisfactory drainage and direct repair if possible

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Anastomotic Stricture (17-60%)

- Most common reason for further surgery to the esophagus
- **Risk factors** are similar to anastomotic leakage, with the addition of GER
    - elective esophageal dilatation and the routine use of anti-reflux medications have been proposed
- **Diagnosis**
    - Some degree of anastomotic narrowing is invariably seen in all postoperative esophogram
    - Narrowing (>50%) detected on a contrast study, or at esophagoscopy in combination with symptoms
    - Prolonged feeding (“slow feeders”), regurgitation, respiratory difficulty, Food impaction in esophagus in older (2-5 yr)

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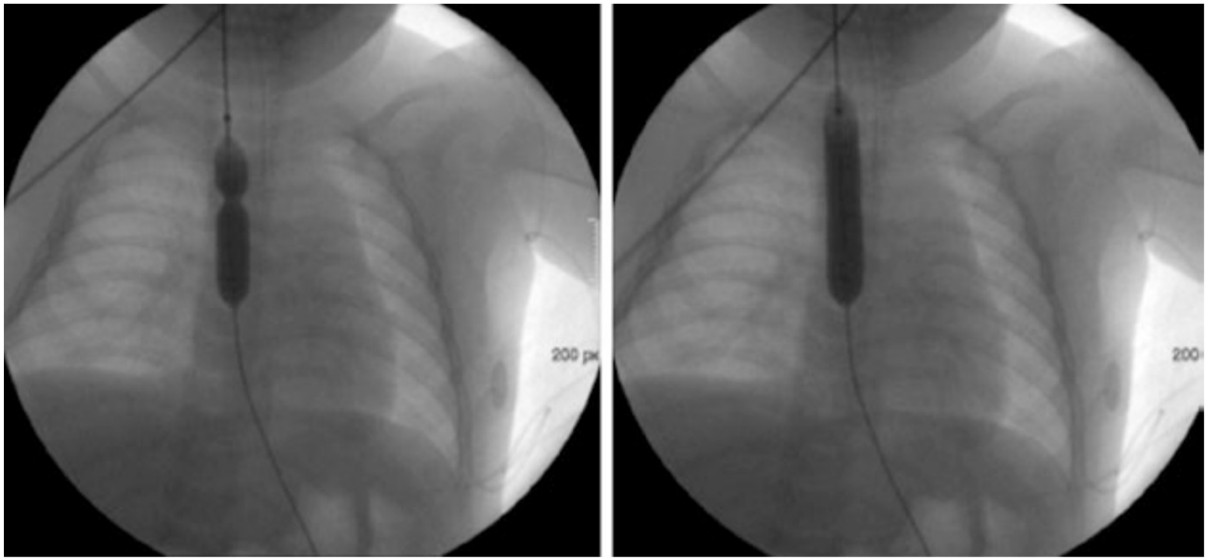
Anastomotic Stricture (17-60%)

- **Balloon dilation** under fluoroscopic or enodoscopic guidance (less traumatic than bougienage)
  - One or two dilatations may be all that is required to treat patients with mild narrowing
  - If a stricture requires frequent dilatation GER should be investigated
- **Resection-anastomosis** and fundoplication
  - Resistant, Recalcitrant strictures
  - Foker's method for long strictures (resect as much as possible and anastomoses under tension to stimulate growth. Resect more and anastomose again after 1 wk)
- Some surgeons deploy **steroid injections or mitomycin C** to manage esophageal strictures



Balloon dilatation esophageal anastomotic stricture: ‘wasting’ appearance shows the region of stricture

Respiratory Morbidity

- 50% hospitalized with a respiratory illness
- Different contribution (assess swallowing-video fluoroscopy, pH monitoring, contrast studies, bronchoscopy)
  - **susceptibility** to respiratory infections (24% reccurent infection)
  - **aspiration** episodes (stricture, dysmotility or GER)
  - **Tracheomalacia, fistula**
  - **obstructive and restrictive** abnormalities seen in spirometry (>50%)
  - **Bronchial inflammation** and airway narrowing seen in bronchoscopy (30%)
- Treatment does not prevent the persistent cough (15%)

Recurrent TEF

- 3% - 15%
- Related to leak
- cough during feeding, apneic or cyanotic episodes, repeat respiratory infections.
- **esophagram** using a water-soluble contrast medium
- **Bronchoscopy** is usually needed to confirm the diagnosis.
  - site of the original fistula carefully examined.
  - methylene blue dye carefully instilled into the fistula pit. Synchronous flexible esophagoscopy to check
  - esophagus is filled with water and positive pressure ventilation applied to check.

Recurrent TEF

- **Redo fistula repair (**via thoracotomy)
  - interposition of a biosynthetic patch or Native tissue such as pleura, pericardium, or intercostal muscle
  - Recurrence reported between 10 - 20%
- **Endoscopic management** (cautery, fibrin glue, chemocauterization, and small intestine submucosa)
  - success rate around 55%

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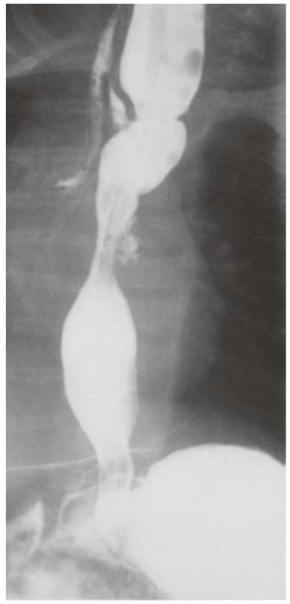
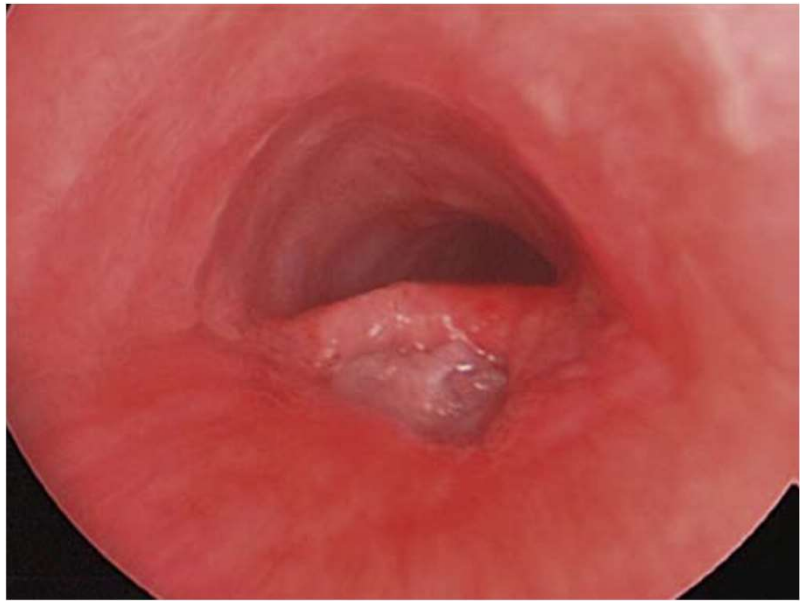


Recurrent TEF



Surgisis placed into a recurrent TEF

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Missed upper fistula (“double fistula”)

- Usually found above thoracic inlet
- **Clues** to alert the surgeon
  - a short upper pouch
  - upper pouch more adherent than usual to airway.
  - Trachea entered during dissection of proximal pouch
- **Diagnosis**
  - Recurrent chest infections and aspiration on oral feeding in the early weeks
  - May be diagnosed on a postoperative esophogogram to evaluate the primary anastomosis.
- **Repair** at a later date (to allow primary anastomosis to heal)

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Tracheomalacia

- **Weakness of the trachea** that allows the anterior and posterior walls to come together during expiration
  - Usually affect lower half (in region of fistula), but can extend beyond main bronchus
- **Cause**
  - Inherent tracheomalacia in esophageal atresia (severity and extent is variable).
  - ? external pressure from the dilated proximal esophageal segment in utero
  - Aberrant major vessels may exacerbate the severity of tracheomalacia locally in the adjacent trachea
- **Presentation**
  - expiratory stridor, loud “barking” TEF cough, cyanosis or apnea.
  - difficulty in reducing mechanical ventilation (‘failure of extubation’)

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Tracheomalacia

- **Diagnosis**
  - combination of flexible bronchoscopy and bronchography.
  - **maintain spontaneous ventilation** to demonstrate dynamic collapse of the trachea during expiraiton
  - stenting effect of the ETT is minimized by carefully withdrawing the tube to the immediate subglottic region
  - significantly compressed antero-posteriorly and assumes a appearance during expiration

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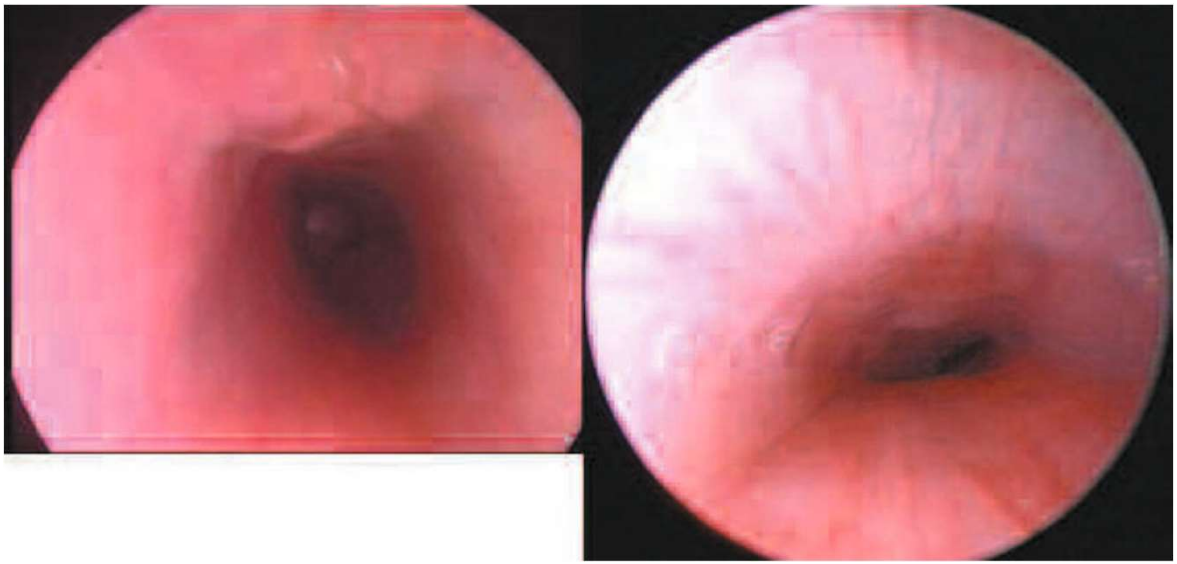

Bronchoscopic view of the trachea in an infant with tracheomalacia: The trachea is open during inspiration, and collapses on itself during expiration (narrow slit like)

Tracheomalacia

- **CPAP**
  - surgical intervention is best indicated for patients with life-threatening symptoms
- **Tracheostomy**
  - traditional view has been that tracheobronchomalacia Is self-limiting, so put on tracheostomy
- **Aortopexy** (hitching the aorta forwards and thereby pulling the trachea open)
  - Definitive treatment
- **Tracheal stenting**
  - Failure of aortopexy

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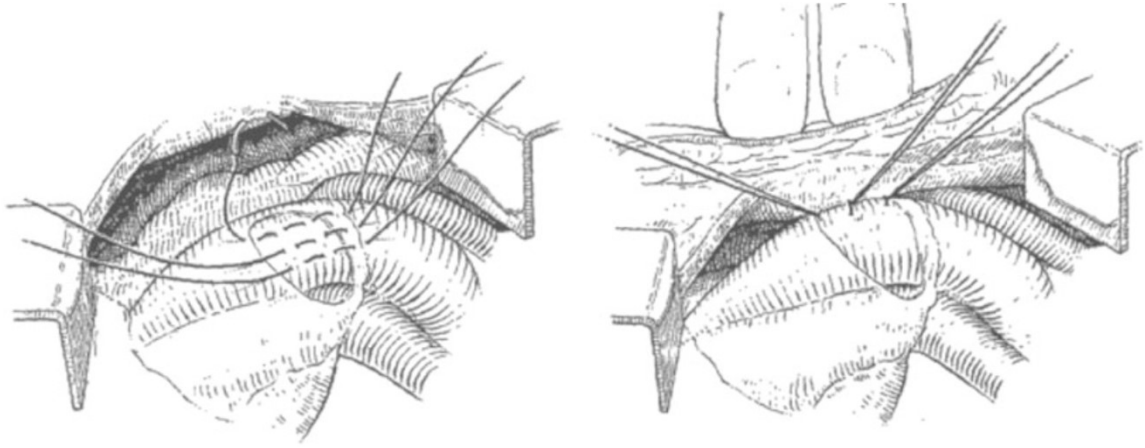

Aortopexy: Approached hrough left anterolateral thoracotomy or median sternotomy, or low cervical incision, or thoracoscopically. Left lobe of the thymus gland is excised taking care not to damage the phrenic nerve.Non-absorbable pledgetted sutures are placed through the adventitia coat of the ascending aortic arch and then passed to the undersurface of the periostium of the sternum and tied. Tracheal patency is confirmed bronchoscopically during the procedure

Esophageal motility disorders

- Lower segment peristalsis uncoordinated. Predisposes to GERD
- **Cause**
  - Intrinsic innervation abnormalities of the esophagus
  - Injury to vagus nerve
- **Diagnosis**
  - dysphagia, episodes of foreign body impaction, heartburn, vomiting, and various respiratory disorders
  - **endoscopy** reveals no significant anastomotic narrowing.
  - **Manometry**

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GERD (up to 50%)

- **Diagnosis**
  - Failure to thrive, recurrent aspiration, esophagitis and stricture
  - Early normal pH values in the esophagus do not exclude significant reflux on followup
- **Aggressive medical therapy** (successful in only about half, doesn't help bile reflux)
  - Postural therapy
  - close attention to feeding regimes, with calorie supplementation
  - Feed thickeners (Carobel),
  - antacid (Gaviscon), H2-antagonists (Ranitidine), PPI (Omeprazole),
  - pro-kinetic agents (Domperidone)
- **Fundoplication** (have a higher failure rate 15-38% but indicated if medical fails)
  - some advocate partial wraps (thal) not to aggravate dysphagia (consequence of underlying dysmotility)
  - many prefer a short (1.5–2.0 cm), floppy, 360° Nissen wrap for its effectiveness

Esophageal Cancer

- **Metaplasia** was found in 15% (lag time for development ~ 10 years)
- **Barrett esophagus** was found in 6.4%
- **Esophageal Cancer** risk uncertain (5 reported so far)
- The question remains whether adults who underwent EA repair as an infant should be screened

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Vocal cord dysfunction

- 3% - 12% (higher in isolated EA an H-type – 50%)
- etiology difficult to assess
    - Inherent problem
    - Injury to the recurrent laryngeal nerves during EA repair
- preoperative laryngoscopy or bronchoscopy to identify infants with congenital vocal fold paralysis .

Musculoskeletal complications

- Pain
- winged scapula
- elevation or fixation of the shoulder
- asymmetry of the chest wall, rib fusion, scoliosis
- breast and pectoral muscle maldevelopment