Congenital Esophageal Atresia with Tracheoesophageal Fistula

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Adapted from previous Creighton University Clinical Anatomy students' presentations

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Creighton University

Case Study

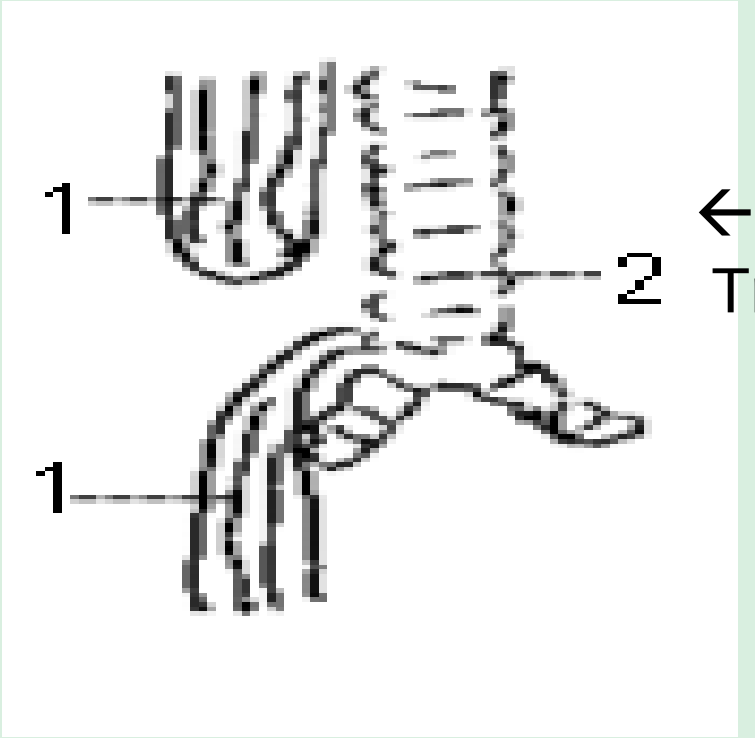
- 4 lb., 5-oz. Premature female infant
- Pregnancy complicated by maternal hydramnios
- Infant showed an excessive amount of oropharyngeal secretion
- Initial feeding 12 hours after birth resulted in vomiting, choking, and cyanosis
- Newborn’s abdomen was distended and became larger upon coughing and crying

Possibilities

- What could have caused vomiting, choking, and not enough O_2 in blood?
  - What happens when an individual vomits? Chokes? Does not have enough O_2 in blood?
- What could have caused abdomen expansion due to coughing and crying?
  - What happens when an individual coughs or cries?

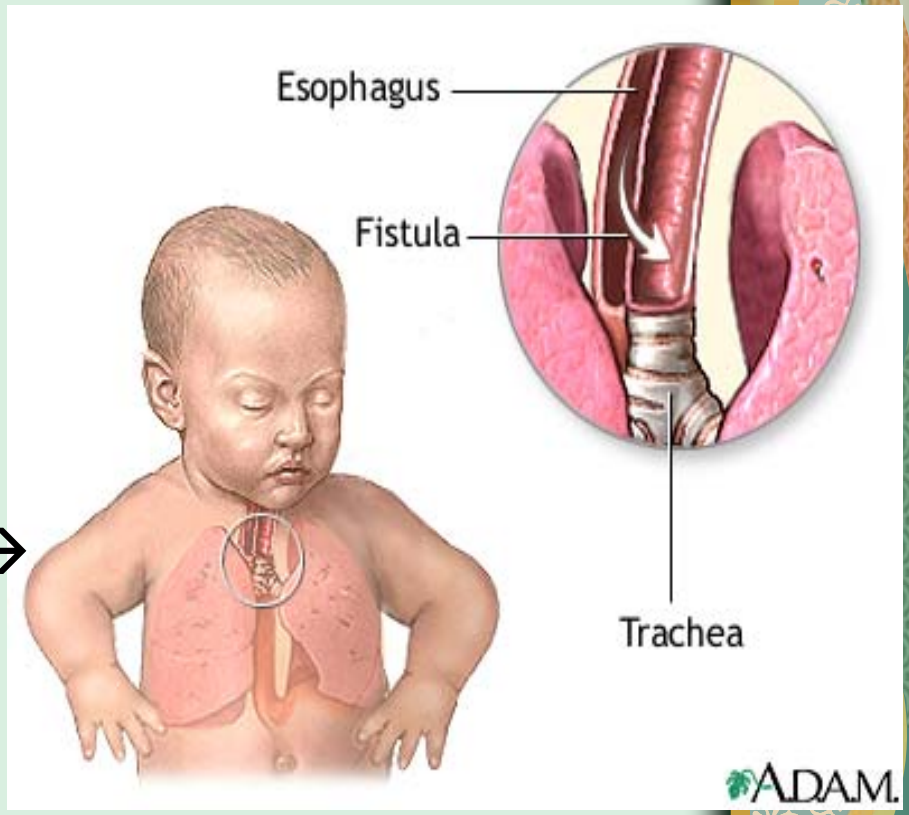
Examination and Diagnosis

- Radiopaque catheter introduced into the esophagus
  - stopped 9 cm from the upper gum margin
- Radiograph was made to determine the level of the obstruction in relation to the vertebrae and to estimate the length of the proximal esophageal pouch
- Full-length air contrast radiograph
  - determined the presence of air in stomach and intestines which indicated the presence of a fistulous communication between the trachea and lower esophagus



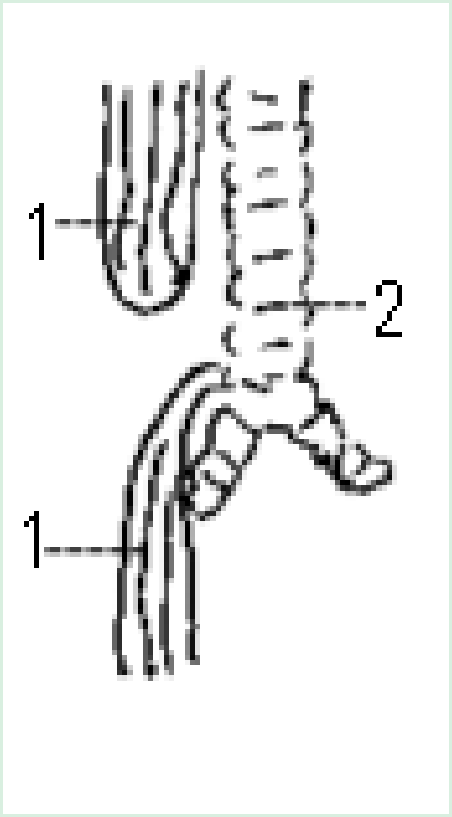
←Esophageal Atresia and Tracheoesophageal Fistula

Newborn with Tracheoesophageal Fistula →



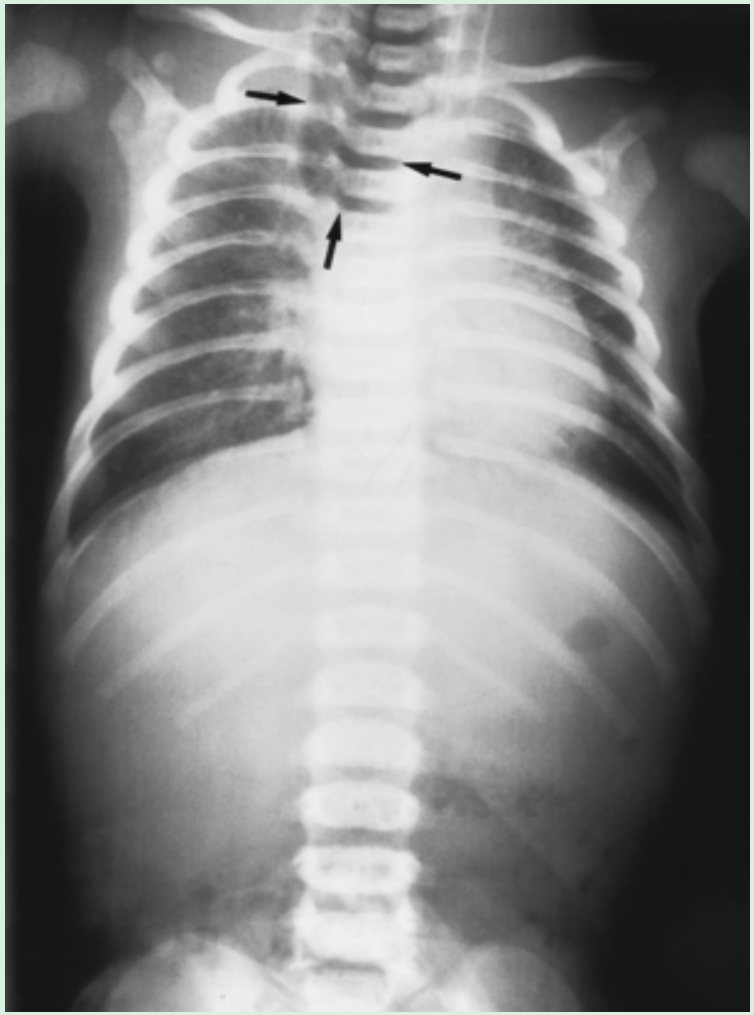
What is Congenital Esophageal Atresia with Tracheosophageal Fistula?

- **Esophageal Atresia** (absence of a normal opening) is a condition in which the proximal and distal ends of the esophagus fail to connect
- **Tracheoesophageal Fistula** (abnormal passage between two internal body structures) is a connection between the trachea and the esophagus
- The connection usually takes place between the distal end of the esophagus and the trachea

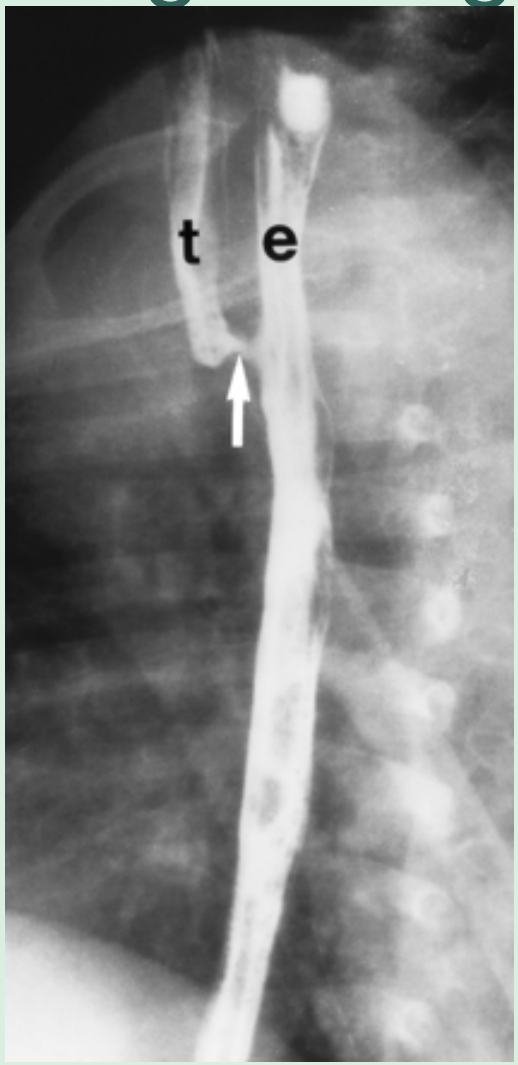


Diagnosing

Distended Proximal Esophageal Pouch



Diagnosing

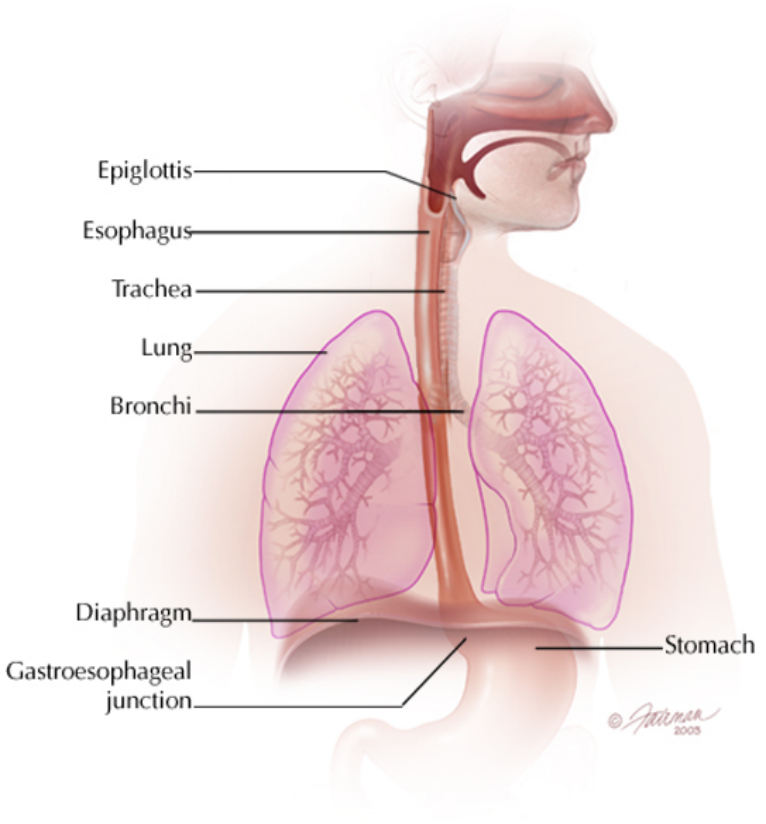


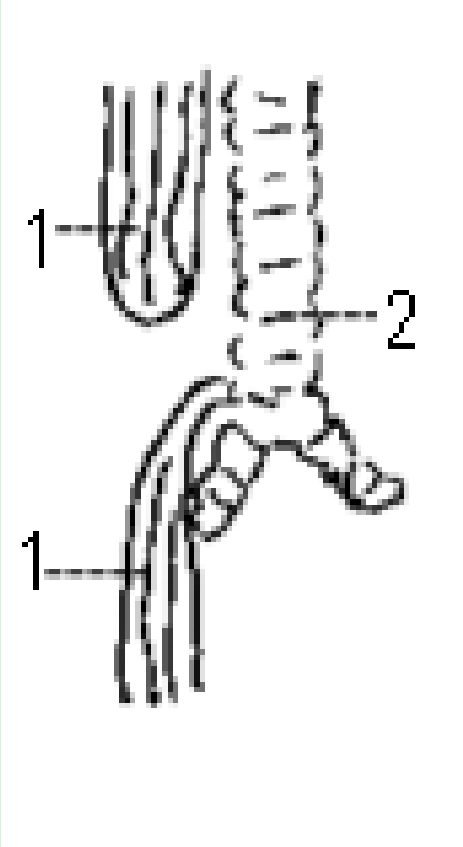
A

P

t=Trachea
e=Esophagus
Arrow is pointing to lower Tracheoesophageal fistula

Normal vs. Abnormal





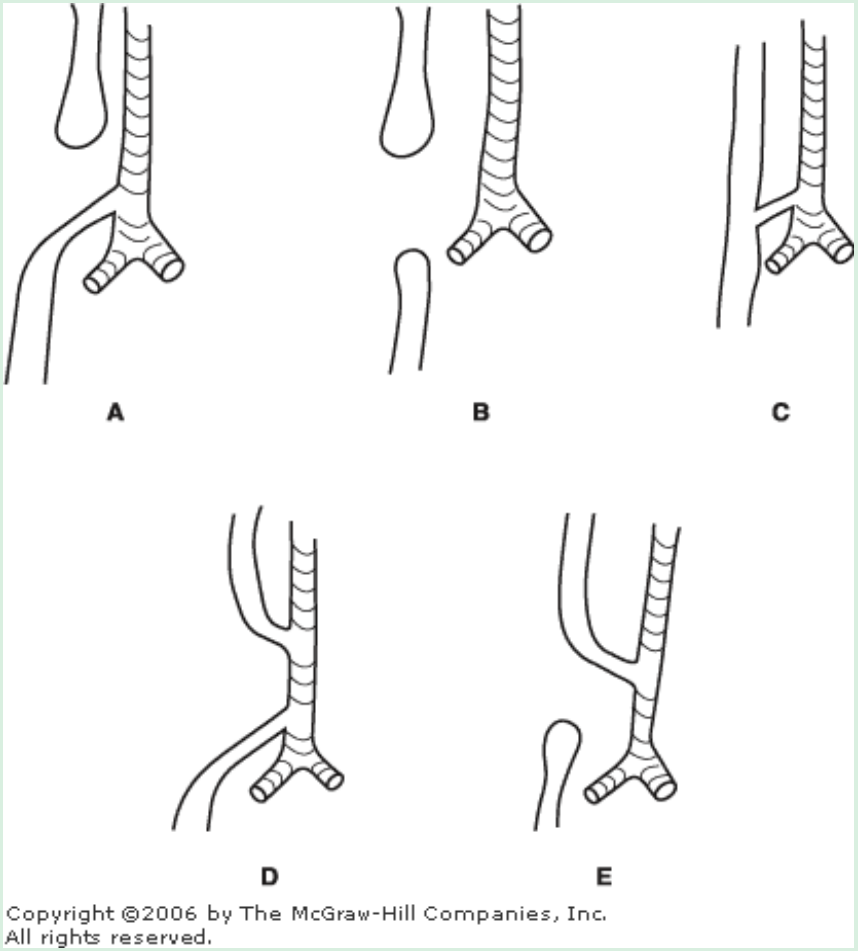
Embryology of the Defect

- Esophagus and trachea form from the primitive foregut
- At 22 days' gestation, the median pharyngeal groove develops in the ventral aspect of the foregut and elongates
- Epithelium proliferation
- Lateral mesodermal ridges form in the proximal esophagus during the fourth week and the fusion of these grooves separates the esophagus from the trachea at 26 days
- Lumen forms following a process of mucosal proliferation and vacuole formation
- EA and TEF believed to develop from an improper separation of the respiratory and digestive divisions during 3-6 weeks of gestation

Embryology of the Defect

- Numerous theories postulated concerning the embryogenesis of EA and TEF
  - EA: asymmetric growth of the esophageal mesenchyme/grooves and epithelial lining; notochord or neural crest abnormalities
  - TEF: failure of the grooves to fuse in the midline, disturbances in epithelial proliferation
  - Vascular deficiencies
  - Chromosomal defects and gene associations have been suggested to play a role in the separation of the primitive foregut

Five Types of Esophageal Atresia with Tracheoesophageal Fistula



- (A) Esophageal Atresia with distal Tracheoesophageal Fistula (84%)
- (B) Isolated Atresia with no fistula (8%)
- (C) Tracheoesophageal Fistula with no atresia (“H-type TEF”) (4%)
- (D) Esophageal Atresia with proximal and distal fistulas (3%)
- (E) Esophageal Atresia with a proximal fistula (1%)

Associated Malformations

- Congenital anomalies exist in 50% of the cases of EA; overall 25% of patients have other congenital defects
- **VACTERL** association describes commonly associated combination of defects
  - **V**ertebral
  - **A**norectal (imperforate anus, duodenal atresia)
  - ****C**ardiac (VSD, ASD, patent ductus arteriosus, tetralogy of Fallot, right-sided aortic arch)
  - **T**racheal (and anomalies of the respiratory tree)
  - **E**sophageal
  - **R**enal (polycystic kidney, renal agenesis)
  - **L**imb (polydactyly, lower-limb defects, rib defects, scoliosis)

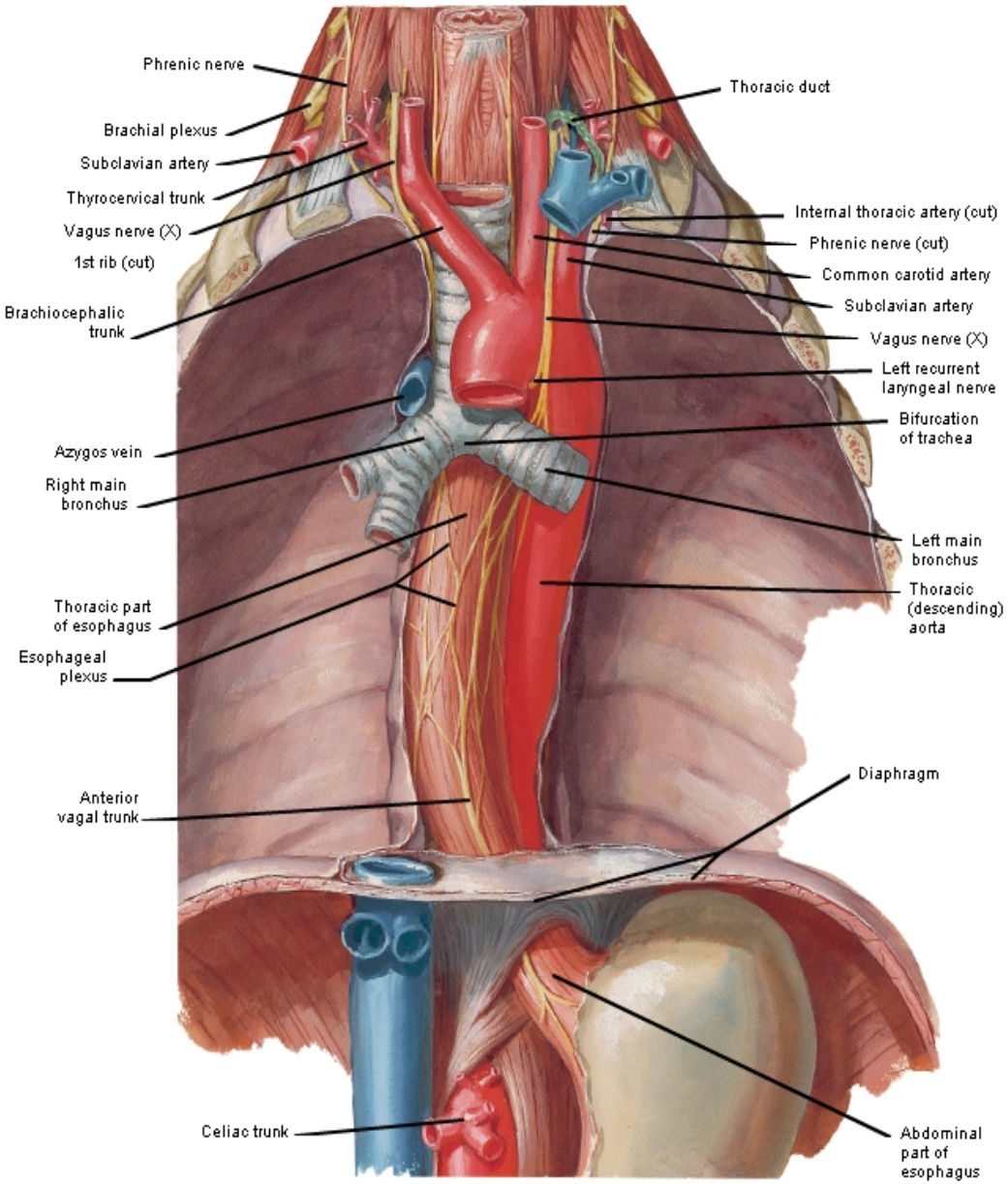
Essentials of Diagnosis

- Polyhydramnios (prenatal sonography/MRI)
- Excessive drooling, fine frothy bubbles
- Coughing, cyanosis, vomiting or difficulty breathing
- Association with VACTERL
- Imaging studies:
  - Unable to pass feeding tube/catheter
  - Chest/Abdominal x-ray
    - Shows tube curled up in upper chest or neck
    - Intestinal gas indicates esophageal atresia with a tracheoesophageal fistula; no gas represents isolated esophageal atresia
- Echocardiogram

Relevant Anatomy

- Esophagus divided into segments based on blood supply
  - Cervical: inferior thyroid artery and vessels derived from the common carotid, subclavian, vertebral, ascending pharyngeal, superficial cervical
  - Thoracic: segmented blood supply from bronchial arteries
  - Lower thoracic and abdominal: unpaired esophageal and ascending branch of left gastric artery and branches of left inferior phrenic
  - Azygos vein serves as a good landmark during surgery
- Esophagus innervated largely by the autonomic nervous system
  - Sympathetic (pharyngeal plexus and the stellate ganglia)
  - Parasympathetic (vagus, recurrent laryngeal nerves)

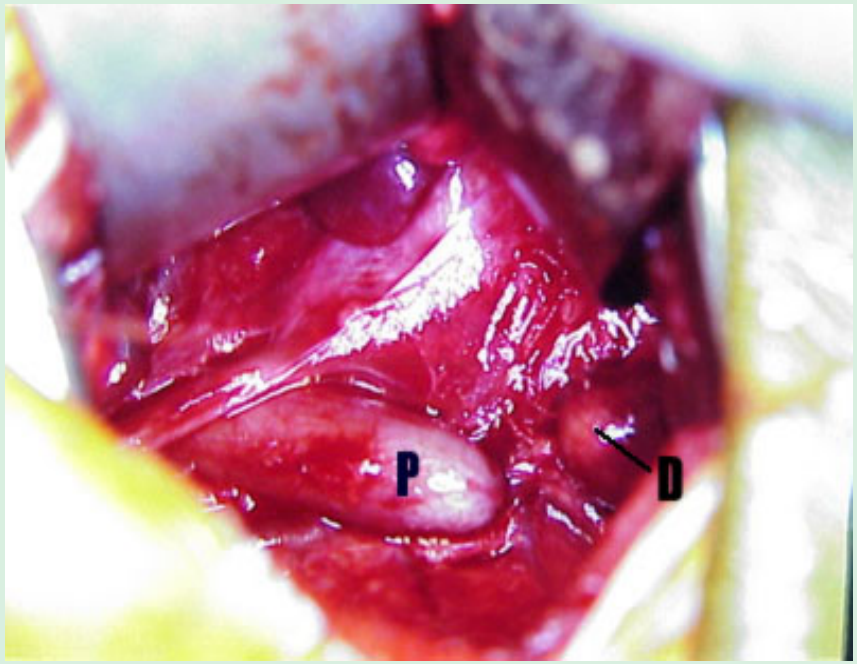
Esophagus In Situ



Treatment

- All children with EA/TEF require surgical intervention shortly after birth
- Operative repair through a right posterior lateral thoracotomy at the fourth intercostal space
- Performed extrapleurally, transpleurally and recently, thoracoscopically
- Esophagus dissected from the membranous portion of trachea
- Primary anastomosis performed whenever possible
- Drainage catheter placed in retropleural space

Treatment

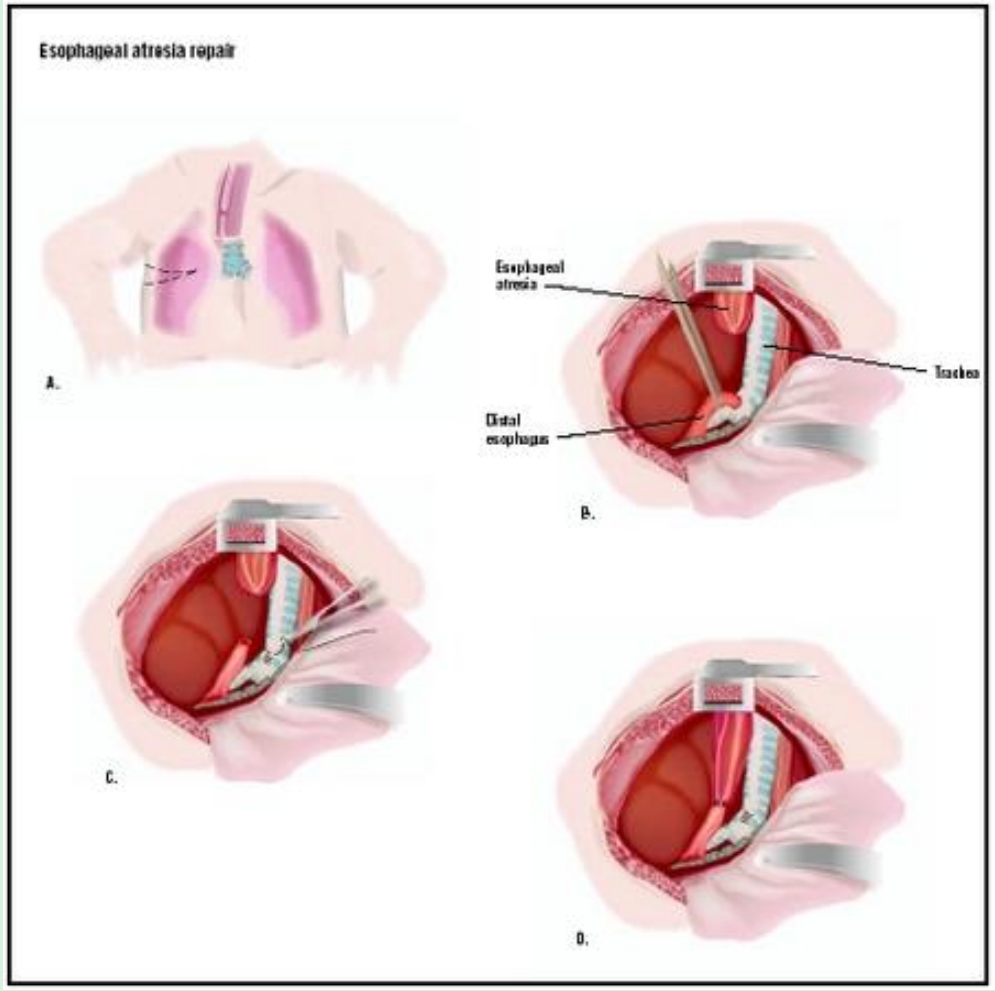


- Intraoperative photograph during repair of esophageal atresia. The head is to the left. The azygous vein has been divided. The proximal (P) and distal (D) esophageal pouches can be easily observed.

Invasive Treatment

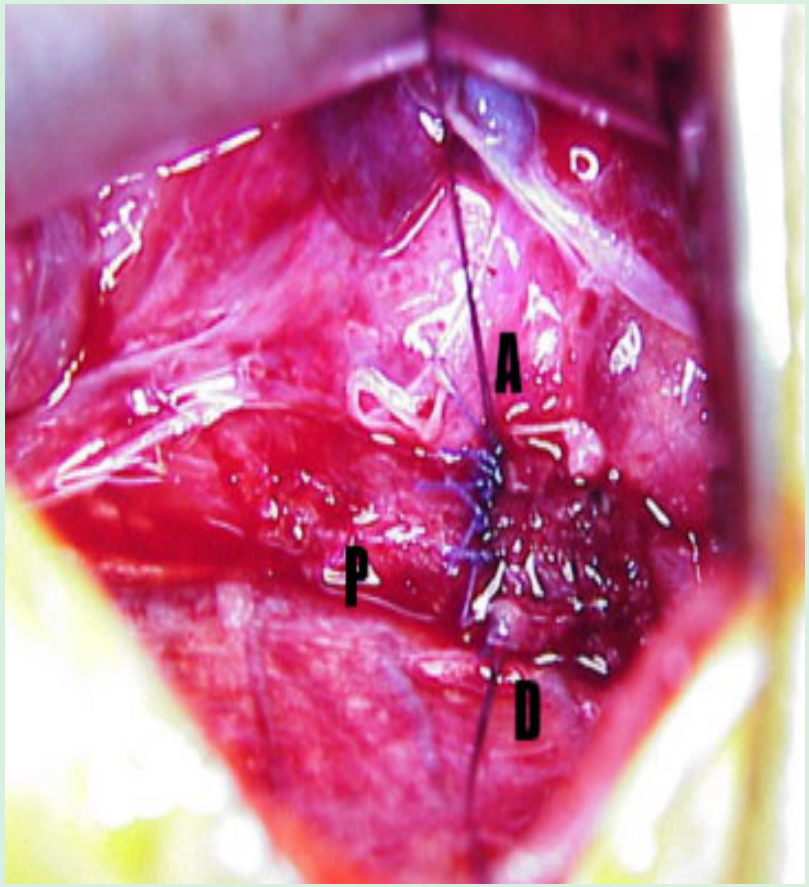
- Surgical correction in one stage
- Extrapleural approach used
  - Fourth rib removed, parietal pleura reflected from thoracic wall and posterior mediastinum entered
  - Vagus nerve, esophageal pouches and the fistulous connection were identified
  - Connection between lower pouch and trachea separated and suture line tested
  - Gap between portions of esophagus overcome by mobilization of the upper pouch
  - Upper portion of lower lumen excised
  - Anastomosis made to prevent recurrence of a fistula

Treatment



www.answers.com/topic/esophageal-atresia-repair

Treatment



- Intraoperative photograph showing primary anastomosis (A) between the proximal and distal esophagus.

Outcome

- Surgery was well tolerated
- Postoperative feedings of glucose and saline by dropper
- On the 4th day, an aqueous contrast swallow was done to determine if any leakage was present-none was found
- Feedings moved on to glucose and boiled skim milk with full caloric intake reached by the 14th day
- Result:

  With the exception of several dilations of the anastomotic site, the patient made an uneventful recovery

  At the age of 2 the patient is well developed and free from any symptoms

Postoperative Complications

- Anastomotic leak occurs in 10-20% of patients
- Anastomotic stricture (narrowing) presents in 25% of cases
- Gastroesophageal Reflux Disease (GERD)
  - Can contribute to stricture and occurs in 50% of patients
- Patients with the VACTERL association have a poorer prognosis due to presence of other anomalies
- Current survival rate of post surgical repair is >90%

References

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(Image) www.answers.com/topic/esophageal-atresia-repair

Glossary of Terms

- **Agenesis** – absence of an organ, usually referring to such an absence resulting from failure of its appearance in primordia in embryonic development
- **Anastomosis** – a connection between two vessels or to create a connection between two formerly separate structures
- **Atelectasis** – incomplete expansion of a lung or a portion of a lung
- **Atresia** – congenital absence or closure of a normal body orifice or tubular organ
- **Congenital** – conditions existing at and usually before birth; hereditary
- **Cyanosis** – bluish discoloration, esp. of the skin, due to excessive concentration of deoxyhemoglobin in the blood
- **Distended** – enlarged; to expand outward owning to pressure from within
- **Dyspnea** – shortness of breath; difficult or labored respiration
- **Empyema** – abscess containing pus

Glossary of Terms

- **Esophagus** – the muscleomembranous passage (cervical, thoracic and abdominal) extending from the pharynx to the stomach
- **Etiology** –the causes or origin of a disease or disorder
- **Fistula** – an abnormal passage or communication, usually between two internal organs, or leading from an organ to the surface of the body
- **Hydramnios (polyhydramnios)** – excess of amniotic fluid usually exceeding 2,000 mL
- **Hypertrophy** – the enlargement of an organ due to an increase in the size of its constituent cells
- **Imperforate anus** – not open; abnormally closed
- **Pneumonitis** – inflammation of the lungs
- **Polycystic kidney** – made up of many cysts
- **Thoracotomy** – surgical incision through the wall of the chest in the plural space