Esophageal Atresia And Tracheo -esophageal Fistula Malformations

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Introduction

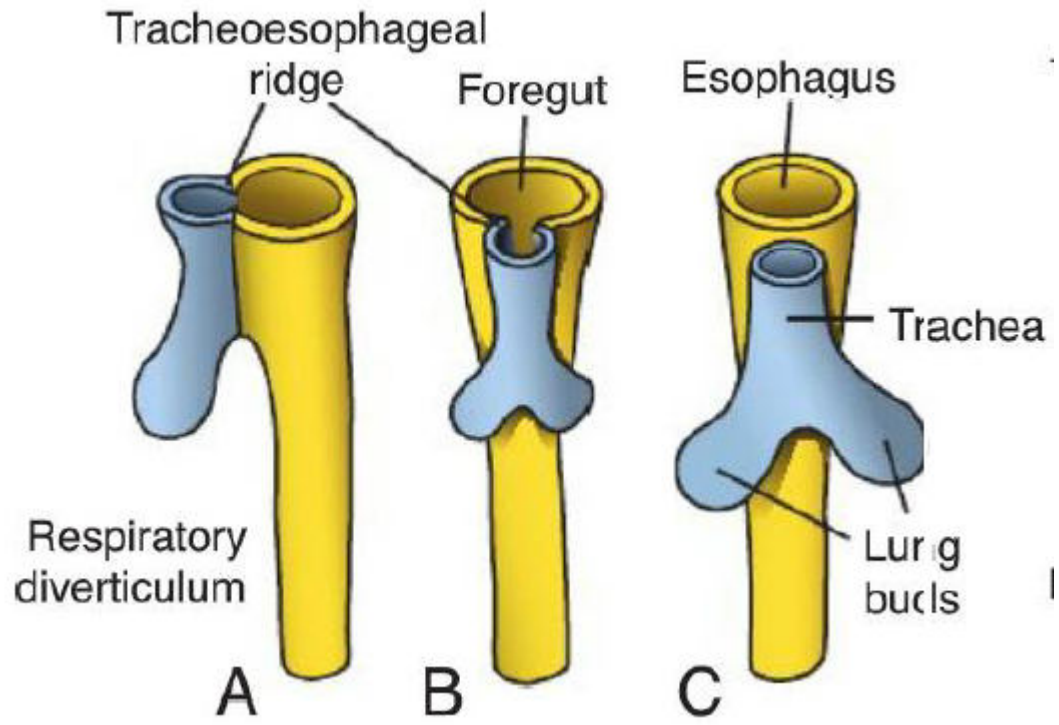
- To anastomose the ends of an infant's esophagus, the surgeon must be as delicate and precise as a skilled watchmaker. No other operation offers a greater opportunity for pure technical artistry.

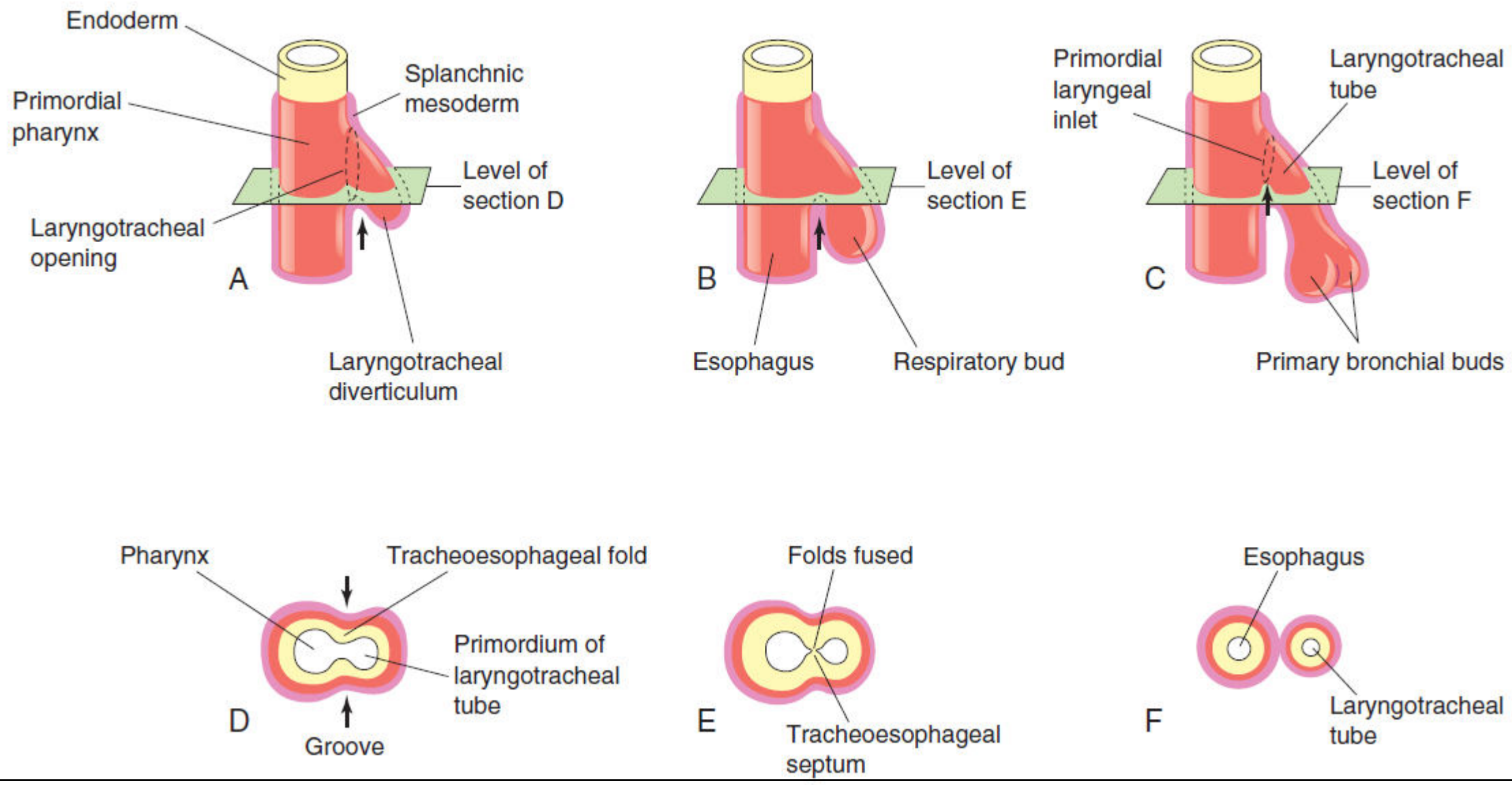
  -Dr. Willis Potts(1950)

- The first successful repair was done in 1940.
- Most-neonatal centres are performing repair now days with success up to 90%.

Embyrology

- 4^{th} week of gestation:
- the foregut ---------->a ventral respiratory part
  ---------->a dorsal esophageal part





Incomplete fusion of the folds

defective trachea-esophageal septum

abnormal connection between the trachea and esophagus

Epidemiology

Incidence: 1 in 2500 to 3000 live births.

Slight male preponderance- 1.26:1

- Chromosomal associations:
  - DiGeorge syndrome
  - Trisomy 21, 13 and 18
  - Opitz syndrome
  - 13q, 17q and 16q24deletions.
- Single gene mutations:
  - Feingold syndrome
  - CHARGE syndrome
  - Fanconi anaemia
- Environmental associations:
  - Methimazole in early pregnancy
  - Prolonged use of contraceptive pills
  - Progesterone and estrogen exposure
  - Maternal diabetes
  - Thalidomide exposure
  - Fetal alcohol syndrome
  - Maternal phenylketonuria

Associated anomalies

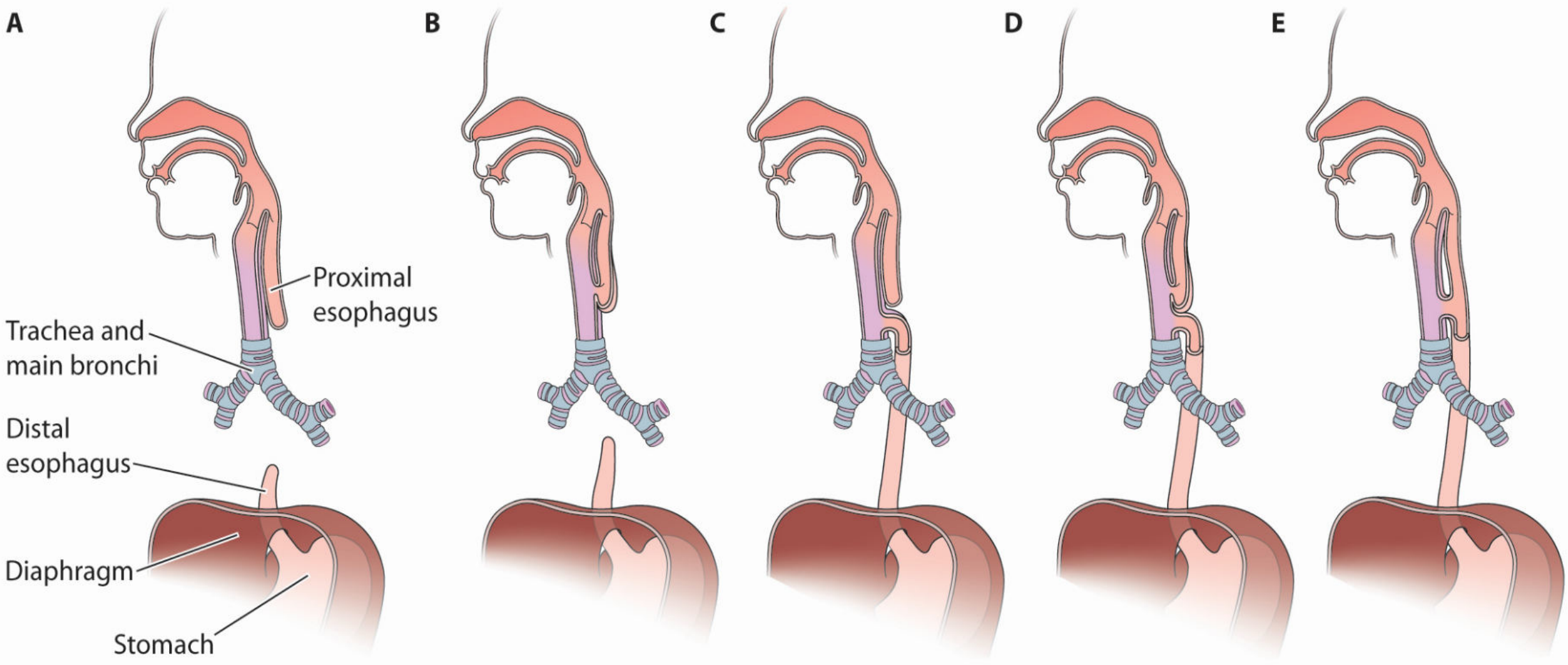
- The most frequent associated malformations encountered in syndromic EA are:
  - Cardiac (13–34%)
  - Vertebral (6–21%)
  - Limb (5–19%)
  - Anorectal (10–16%)
  - Renal (5–14%)

- Two syndromic associations:
  - VACTER-L: Vertebral, anorectal, cardiac, tracheo-esophageal, renal and limb

  - CHARGE: Coloboma, heart defects, atresia of the choanae, developmental retardation, genital hypoplasia and ear deformities.

Classification

- Many classification system has been proposed:
  - Vogt
  - Waterston
  - Ladd
  - Gross
- Gross classification is most commonly used.
  - Based on anatomic variation



Diagnosis

- Antenatal diagnosis: Ultrasonography
  - Polyhydramnios
  - Absent or small stomach bubble

  } suggestive of EA

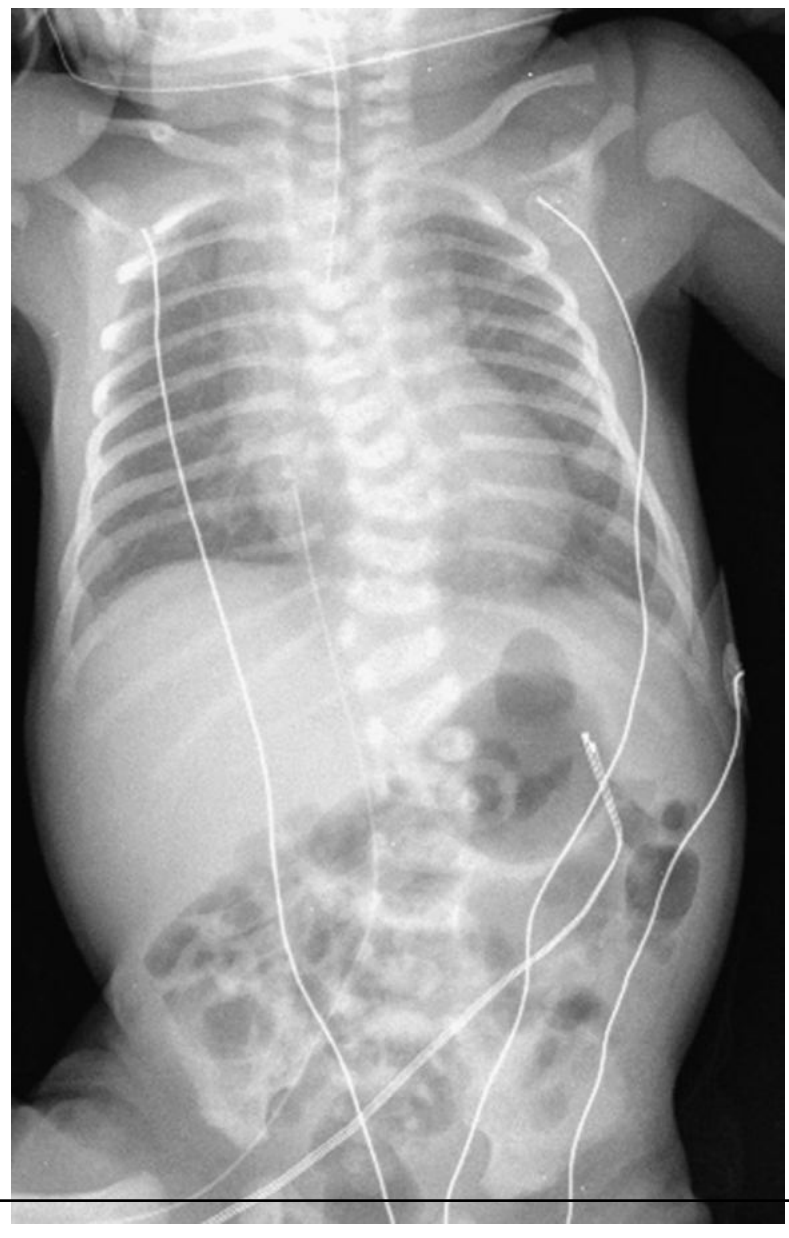
- Fetal MRI.

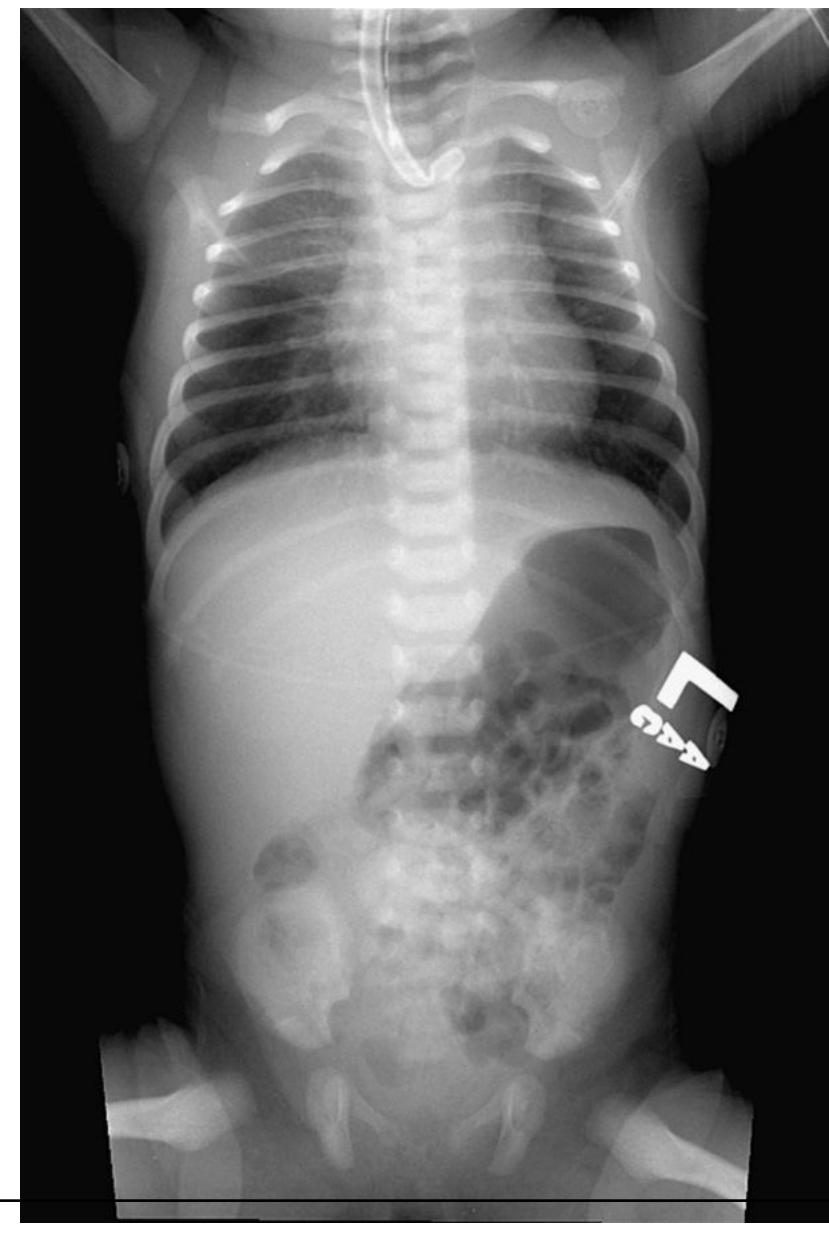
Diagnosis

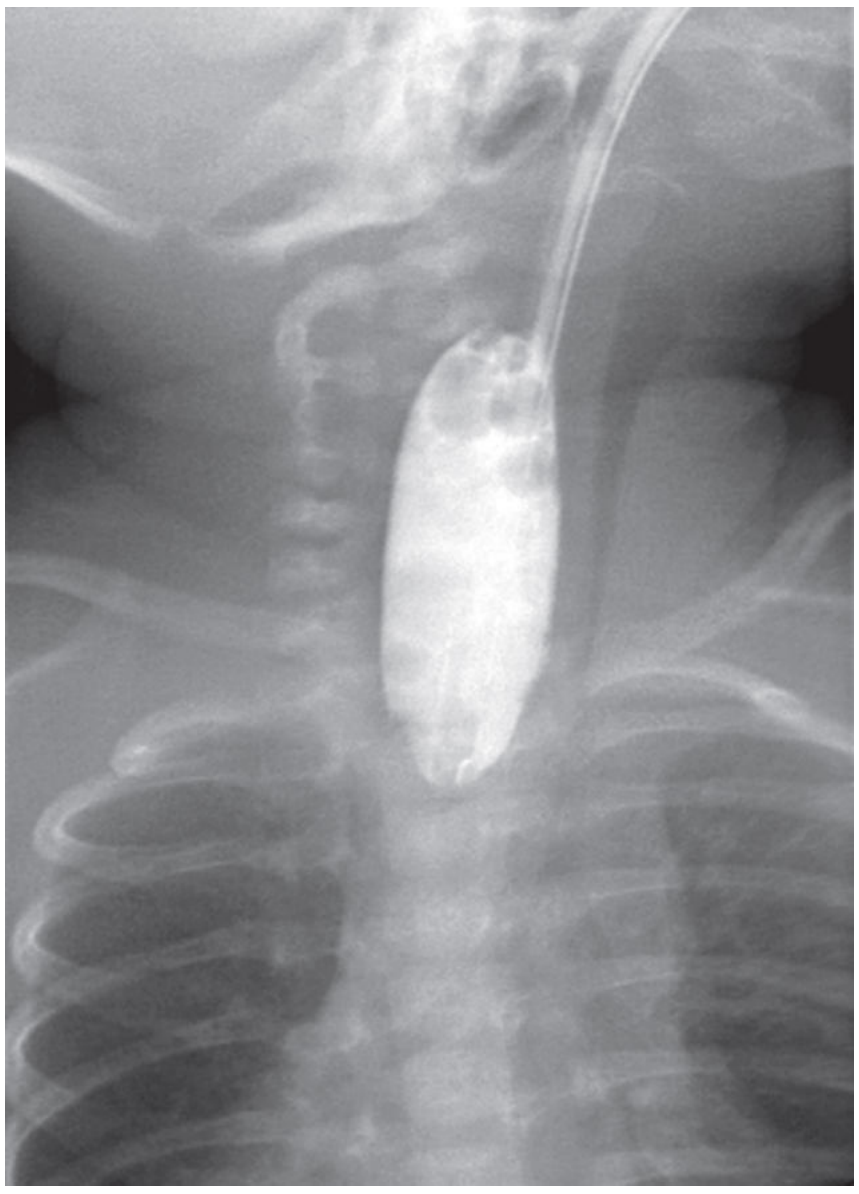
- Postnatal:
  - Premature babies.
  - Symptomatic within first few hours of life.
  - Variants A, B, C and D- excessive drooling of saliva.
  - Choking, regurgitation and coughing on oral feeds.
  - Variants C and D- as the child cries, air goes into the stomach and causes abdominal distension and respiratory distress.
  - Variants C, D and E- gastric juice may regurgitate through the fistula causing chemical pneumonitis.

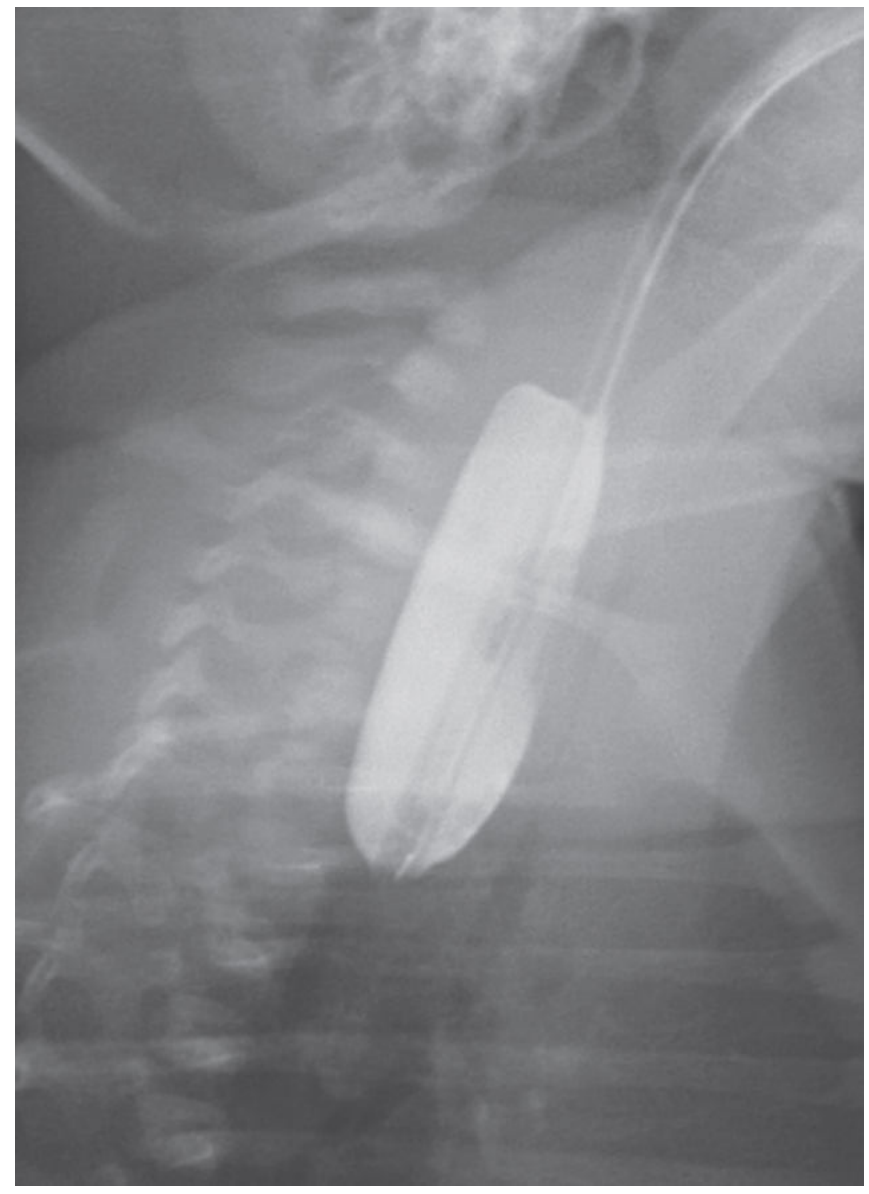
- Inability to pass an orogastric tube into the stomach.
- X-ray:
  - Plain chest radiograph: dilated upper pouch.

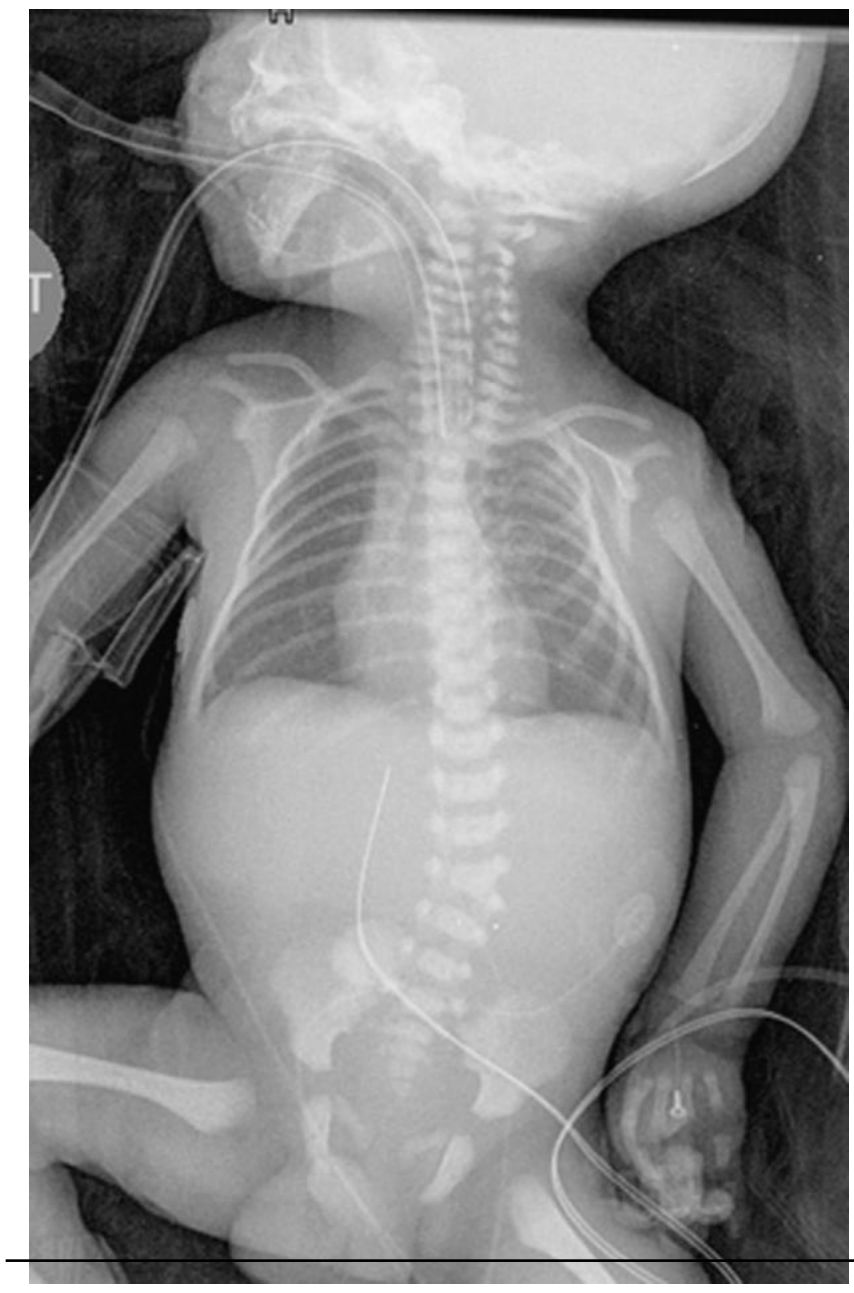
  - If a soft feeding tube is used, the tube will coil in the upper pouch.







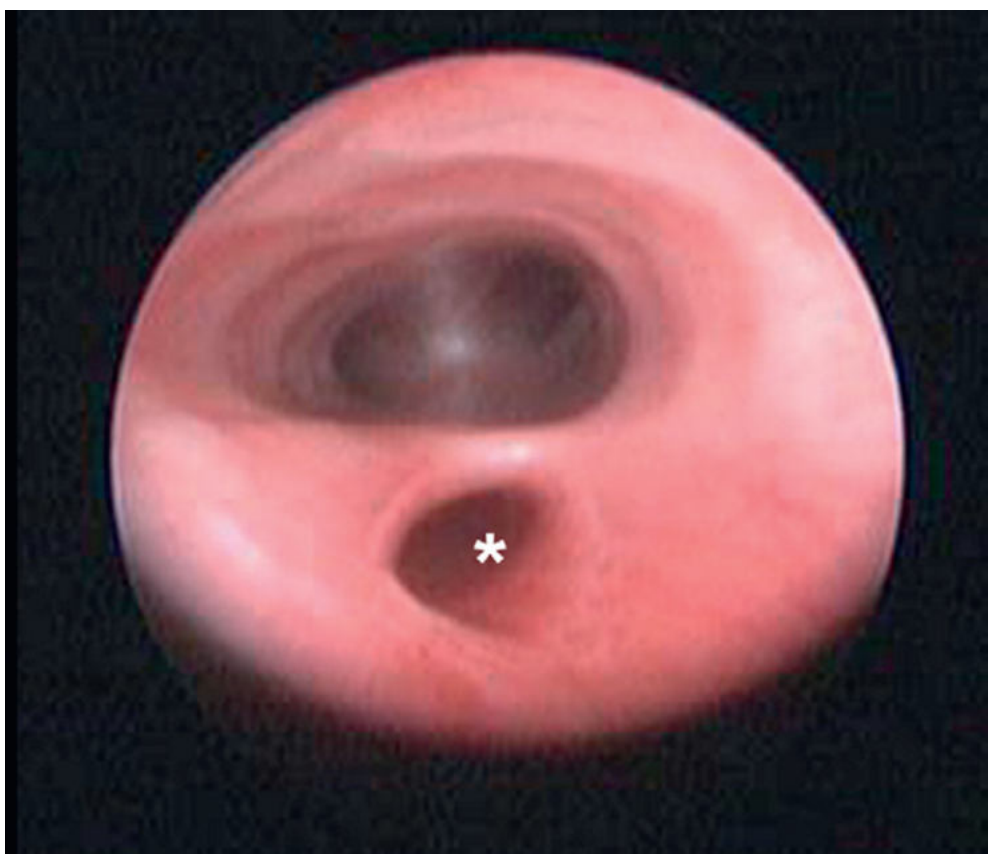


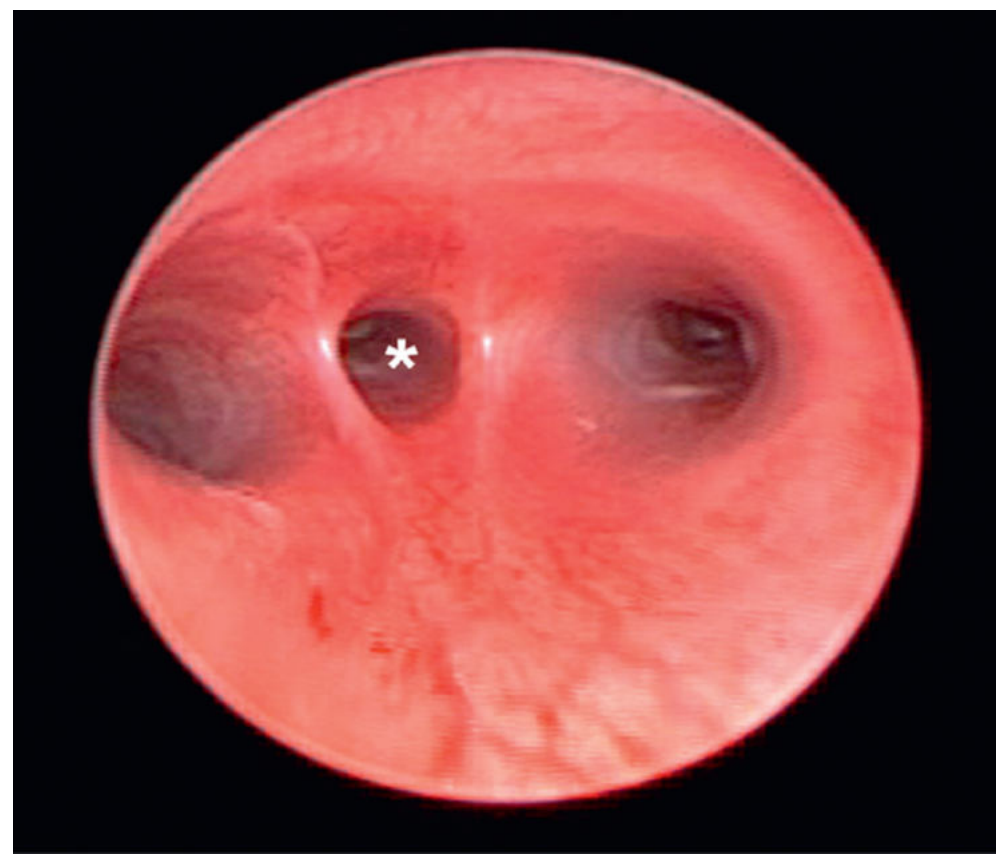


Gasless abdomen:
Pure esophageal atresia and proximal fistula

No route for gas to enter abdomen

- Plain chest radiographs help in assessment of the gap between the proximal and distal end.
- Helps to plan for surgical management.
  - Absence of air in abdomen is associated with long gap.
  - Gap < 2 vertebrae - Primary anastomosis
  - Gap > 2and < 6 vertebrae - Delayed anastomosis
  - Gap > 6 vertebrae - Esophageal replacement





Bronchoscopy

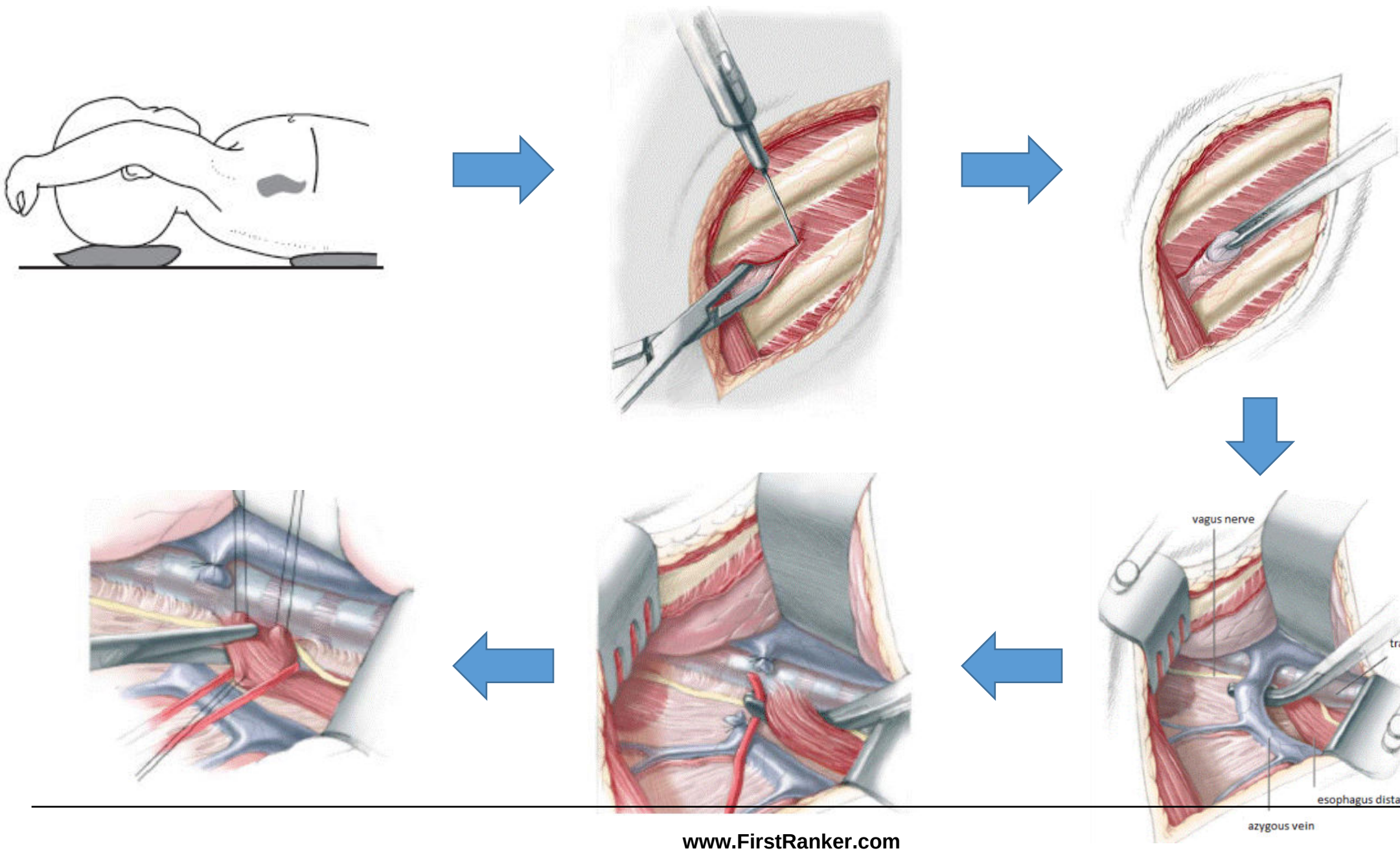
Pre-operative management

- Oral suctioning through 10F double lumen Replogle tube.
- Position of child: Propped up position- least gastric reflux.
- Intravenous access and I. V. antibiotics.
- I. V. fluids (10% dextrose and hypotonic saline)and Vitamin K administration.
- If respiratory distress is present due to pneumonitis, mechanical ventilation may be done at low pressures.
  - High pressure ventilation may cause gastric dilatation and gastric perforation as well as diaphragmatic splinting.

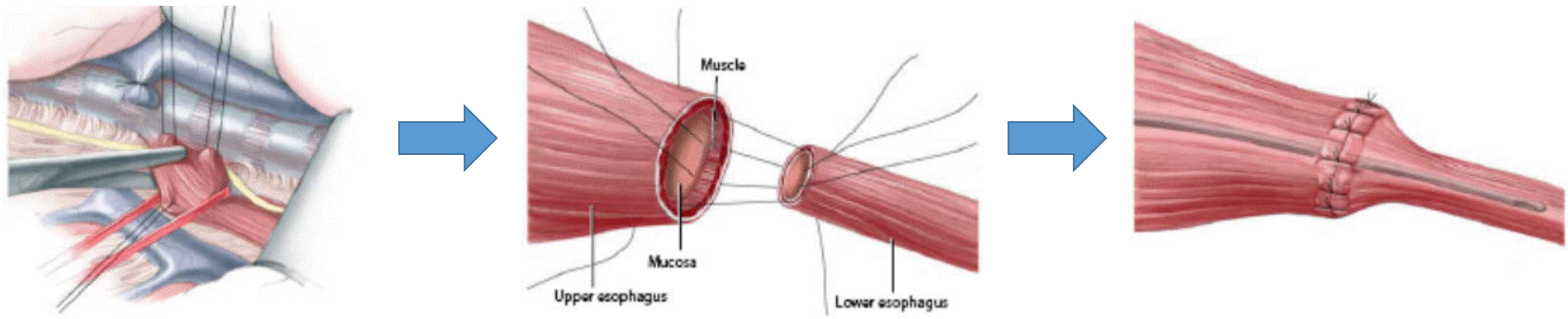
- Investigations for associated anomalies:
- Echocardiography- to look for cardiac defects
- USG abdomen- to look for any renal abnormality.
- X-rays spine- to look for any vertebral anomaly
- X-ray upper limbs- to look for any radial anomaly.

Operative management

- Open thoracotomy:
  - Through 4th intercostal space
  - Extrapleural approach
  - Posterior mediastinum is exposed.
  - Azygous vein is divided to reveal underlying TEF.
  - TEF is dissected circumferentially.

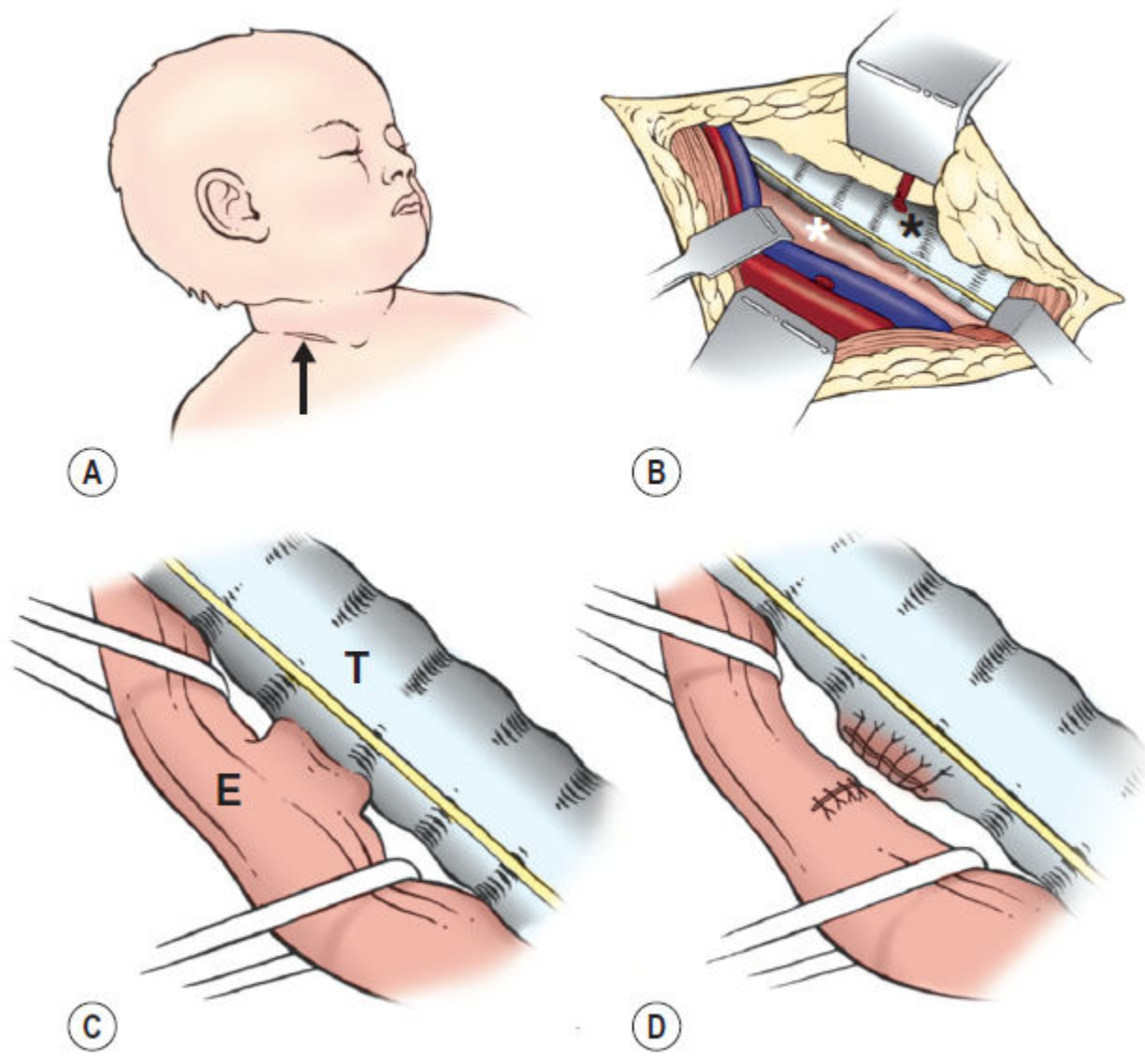


- Attachment of the fistula to the membranous portion of the trachea is taken down.
- Tracheal opening is closed with non absorbable interrupted sutures.
- Upper pouch of the esophagus is mobilised as much as possible.
- The distal end of the upper pouch is opened and a nasogastric tube is passed through the nares to upper esophagus to the lower esophagus and then to the stomach.
- Anastomosis of the ends of the esophagus are done over the nasograstric tube with absorbable sutures.
- Wounds is closed after placement of chest drain.



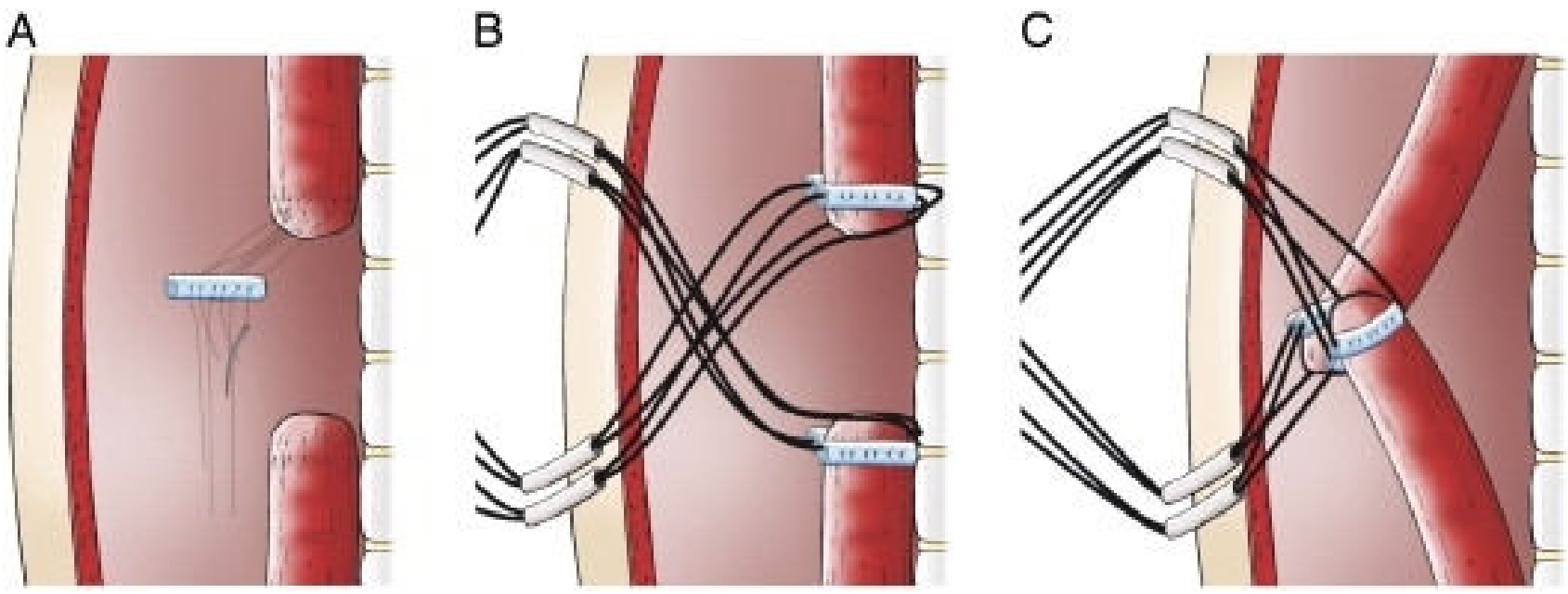
- Thoracoscopic approach is also being used.

- Special cases:
  - H type fistula: division of the fistula done through the cervical approach.



Long gap atresia

- Delayed closure with lengthening procedure (at about 12weeks of life)
  - Spontaneous growth
  - Bougienage
  - Upper pouch mobilisation
  - Kimura’s extrathoracic elongation technique
  - Foker traction suture technique
  - Upper pouch myotomy and flaps
- Esophageal replacement
  - Stomach
  - Colon
  - Jejunum
  - Ileum



Complications

- Anastomotic leaks
- Anastomotic stricture
- Recurrent tracheoesophageal fistula
- Tracheomalacia
- Disordered peristalsis/ gastroesophageal reflux/ esophageal cancer
- Vocal cord dysfunction
- Respiratory morbidity