

A developmental discontinuity of the diaphragm that allows abdominal viscera to herniate into the chest

**Diaphragmatic eventration:** abnormal elevation of congenitally thin, hypoplastic but intact diaphragm. Can be total or partial.

**Etiology:** mostly sporadic with environmental effects (vit.A deficiency, thalidomide, anticonvulsants, quinines).

- Types:**
- a. **Postero-lateral** hernia (Bochdalek): most common (80-90%), 80-85% on the left side
  - b. **Anterior** hernia (Morgagni-Larrey): 2% of all CDHs
  - c. **Central** hernia: extremely rare, involves the central tendon
  - d. Diaphragm **agenesis**

**Why so serious?**

- a. **Fetal mortality:** hydrops fetalis, stillbirths
- b. **Neonates:** pulmonary hypoplasia, persistent pulmonary hypertension, right-to-left shunting, hypoxemia and acidosis, cardiorespiratory failure, and mortality
- c. **Infants & children:** respiratory and GI manifestations, can be asymptomatic (incidental).

**Classification:**

1. **Isolated CDH:** 50-70% of cases, ↑ survival rate.
2. **Complex CDH:** 30-50%, ↓ survival rate. Associated with structural malformations, chromosomal abnormalities, and underlying syndromes.

**Diagnosis:**

- a. **Prenatally:** fetal ultrasound, and fetal MRI
- b. **Postnatally:** respiratory distress, PEx (scaphoid abdomen, absent breath sounds, bowel sounds in chest), CXR, +/- CT or MRI, +/- GI contrast study
- c. **Infants and children:** respiratory or GI symptoms, PEx, incidentally on CXR, (+/- CT, MRI, GI contrast study).

**Prognostic factors:**

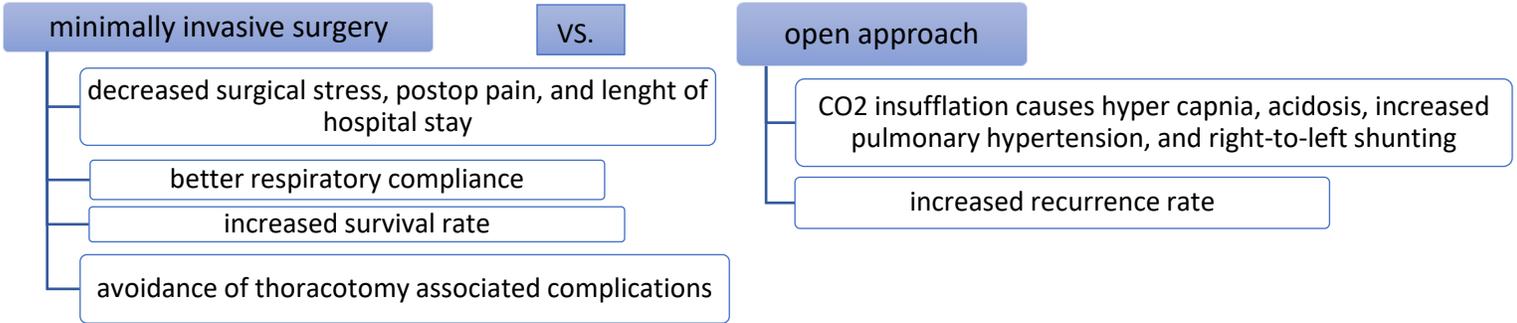
	Survival rate (75% normally)	
Associated malformations	↓	
Fetal lung volume	↓ if <30% of expected lung volume for GA	
Right-sided defects	↓ to 50%	↑ ECMO and patch rate
Liver herniation	↓ to 45%	
Lung area to head circumference ratio (LHR)	(↓ratio means small lungs) more indicative of morbidity than mortality	

**Management:**

Prenatal management	Postnatal management
<ul style="list-style-type: none"> <li>- screening for associated abnormalities</li> <li>- fetal ECG</li> <li>- genetic studies</li> <li>- family counseling</li> <li>- in utero fetal therapy: investigational procedures, patch closure, <b>fetoscopic endoluminal tracheal occlusion</b></li> <li>- delivery planning</li> </ul>	<ul style="list-style-type: none"> <li>- optimizing cardiorespiratory status: reduce lung compression, ventilatory support, cardiovascular support, correction of acid-base status, <b>correction of pulmonary hypertension</b></li> <li>- achieving hemodynamic stability (you need to know the details)</li> <li>- screening for associated malformation</li> <li>- increased survival rate up to 92%</li> </ul>

**Operative management:** CDH is not a surgical emergency

Minimally invasive surgery is preferred more than the open approach



**Criteria for MIS:**

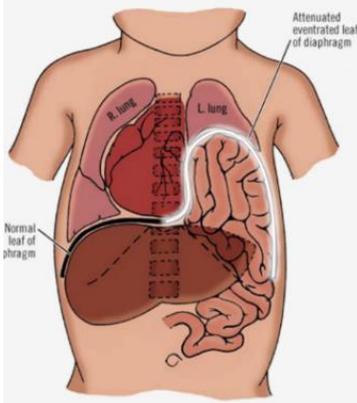
- **Neonates:** hemodynamic, radiographic and respiratory stability, and no severe associated cardiac malformations.
- **Infants & children:** late presenting or incidentally diagnosed

**Outcomes:** Mortality and morbidity are related to:

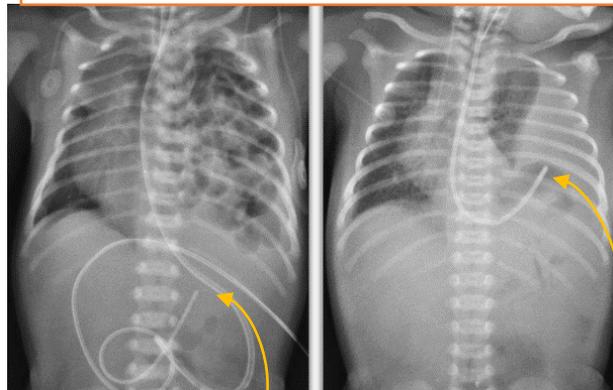
- Severity of lung hypoplasia
- Pulmonary hypertension
- Associated anomalies
- Prematurity

**Appendix 1**

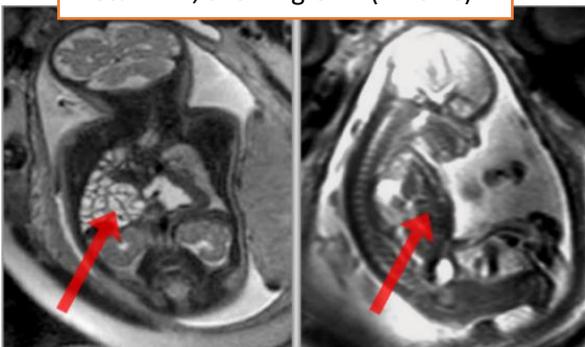
Diaphragmatic eventration



Chest x-ray in a neonate with CDH: bowel loops in the chest, mediastinum shifted to the right



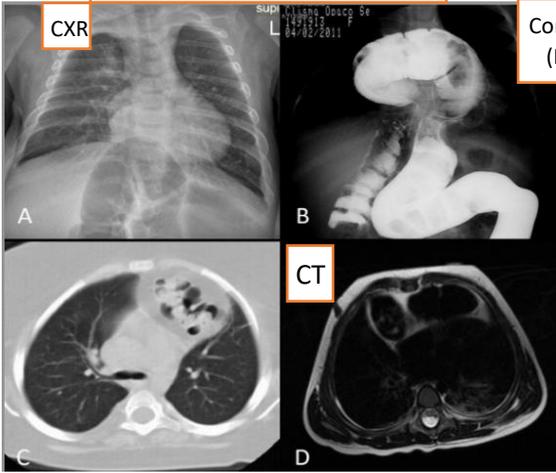
Fetal MRI, showing CDH (Arrows)



NG tube reaching the bowel; the stomach is not herniated

CDH + NG tube only reaching the chest; stomach is herniated

Dx in Infants & children



CXR

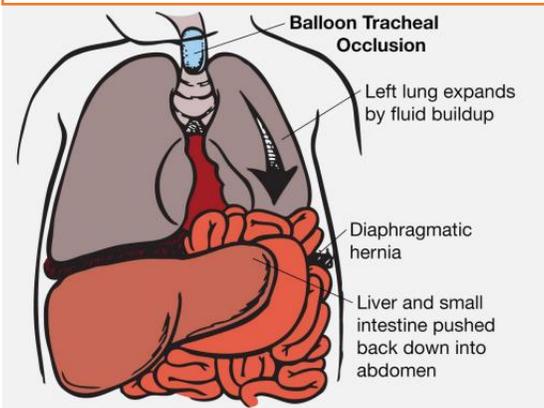
Contrast study (Morgagni)

CT

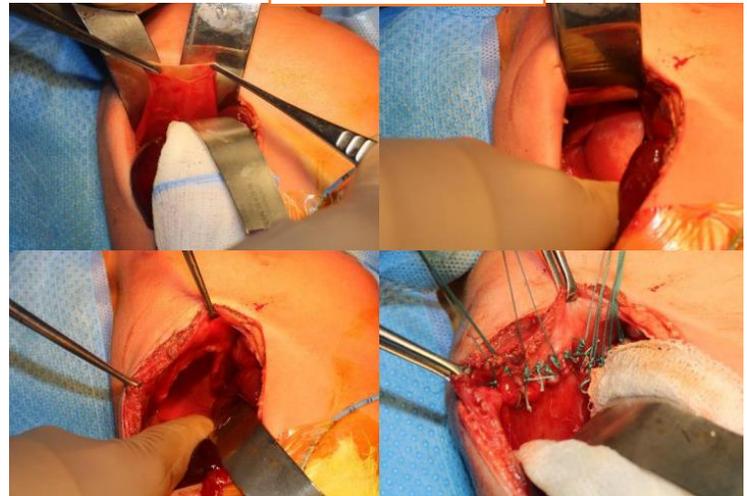


Right sided CDH; note the liver is also herniated

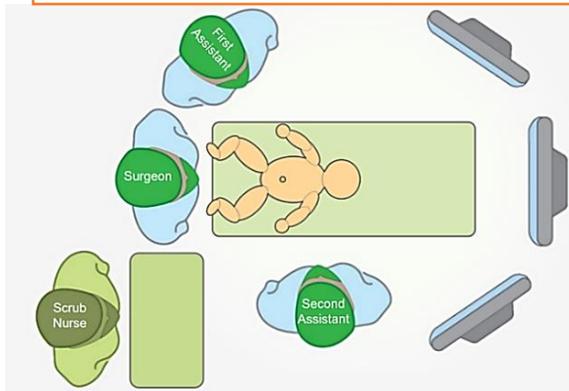
Fetoscopic Endoluminal Tracheal Occlusion; prevents lung hypoplasia



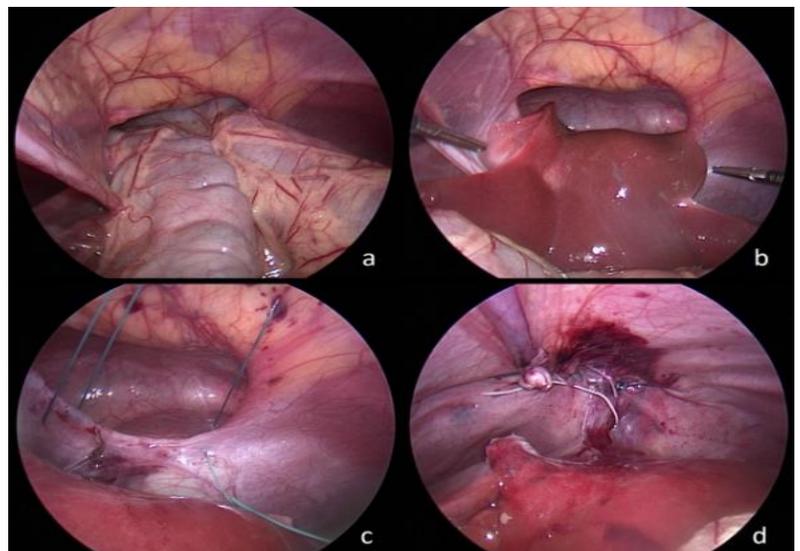
Open CDH repair



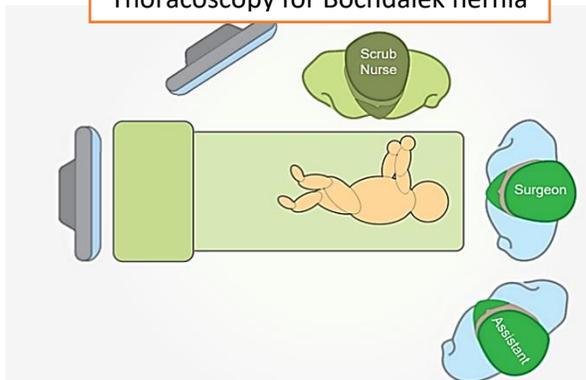
Thoracoscopy for Morgagni-Larrev hernia



Minimally invasive surgery



Thoracoscopy for Bochdalek hernia



## Esophageal atresia (EA) and tracheoesophageal fistula (TEF)

### Etiology:

- Genetic factors.
- Environmental factors: methimazole, OCPs, progesterone & estrogen exposure, maternal diabetes, thalidomide, fetal alcohol syndrome, maternal phenylketonuria.
- Chromosomal anomalies (trisomy 18&21).

### Normal embryogenesis of the trachea and esophagus:

At the 4<sup>th</sup> week of gestation, the tracheoesophageal septum separates the foregut into ventral (respiratory) and dorsal (esophageal) parts. Separation of the septum occurs at 6-7<sup>th</sup> week.

Occurs 1 in 2500-3000 live births with slight male predominance (1.26:1)

**Associated anomalies:** (Isolated EA in 50% of cases)

Syndromic EA (50%): VACTERL and CHARGE

### VACTERL:

V: vertebral  
A: anorectal  
C: cardiac (m/c)  
T: tracheal  
E: esophageal  
R: renal  
L: limb abnormalities

### CHARGE:

C: coloboma  
H: heart defects  
A: atresia of the choana  
R: developmental retardation  
G: genital hypoplasia  
E: ear deformities

**Classification:** (check appendix 2 for pics)

- 85% proximal atresia with distal fistula
- 7% atresia without fistula
- 4% fistula without atresia
- 2% proximal atresia with proximal fistula
- <1% proximal atresia with proximal and distal fistulas (N or H type).

### Diagnosis:

- Antenatal:** polyhydramnios, absent/small stomach bubble (both are nonspecific)
- Postnatal:** excessive salivation, coiled feeding tube on CXR, +/- contrast study.

Presence/absence of gas in the stomach and bowel on AXR is to help determine the type of EA,

**Management:** EA & TEF are not surgical emergencies.

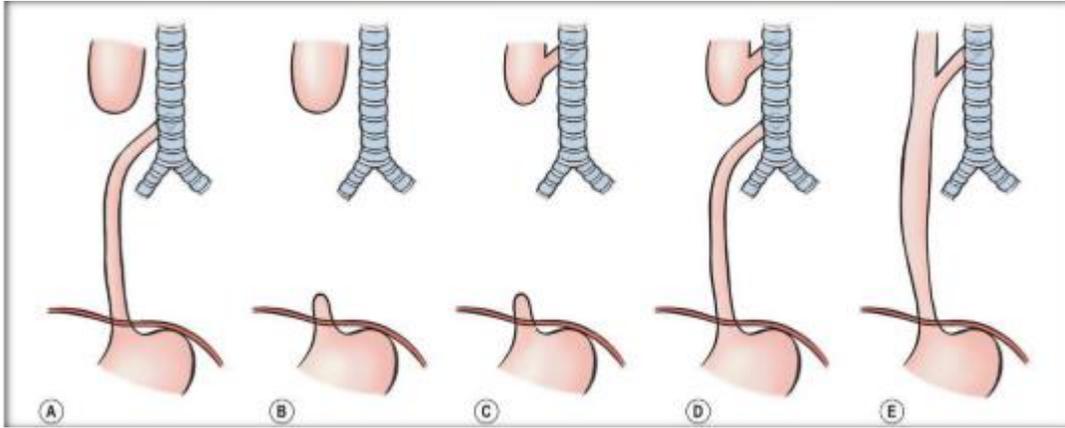
- **Preoperative preparation:**
  - Continuous suctioning tube in the upper esophagus.
  - Head-up position or on the side.
  - Gentle low-pressure ventilation if baby is in respiratory distress.
- **Preoperative workup:** ECG (to rule out cardiac and aortic arch anomalies), renal ultrasound, and spine radiographs.
- Operative repair depends on the gap between esophageal ends:
  - <2 vertebrae → primary anastomosis
  - 2-6 vertebrae → gastrostomy + delayed primary anastomosis
  - >6 vertebrae → gastrostomy + esophagostomy + esophageal replacement later on
- Open (thoracotomy) vs. MIS (thoracoscopy)

### Complications:

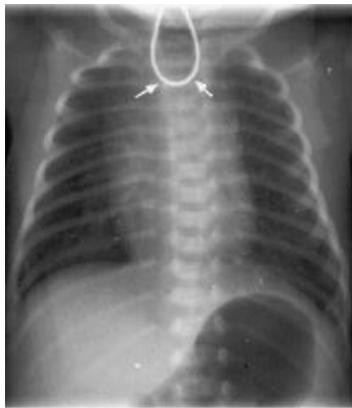
- **anastomotic strictures (17-60%)**
- anastomotic leaks (3.5-17%)
- recurrent TEF (3-15%)
- tracheomalacia
- disordered peristalsis → GERD → esophageal cancer ???
- vocal cord dysfunction
- respiratory morbidity
- thoracotomy-related morbidity

# Appendix 2

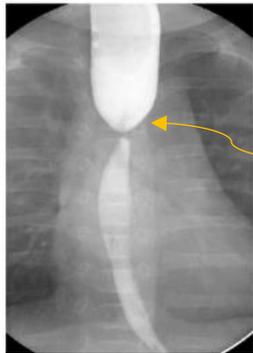
## Classification of EA & TEF



Coiling of tube in the blind upper pouch around T2-T4. Note that there's gas in the stomach, indicating a TEF (type A).



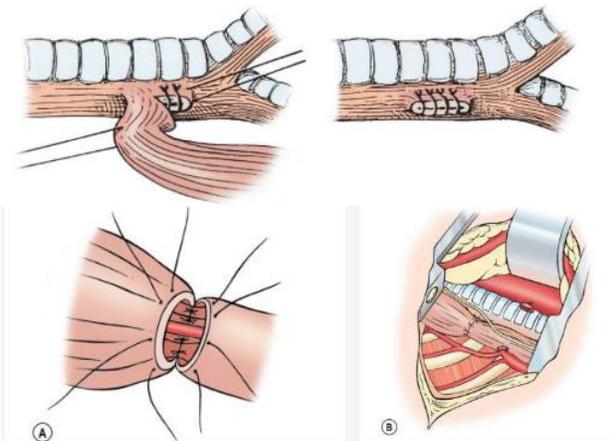
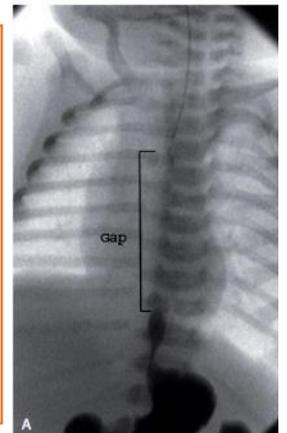
Opacity of the abdomen + NG tube stopped at the upper esophagus → proximal atresia without TEF



Surgery complications:  
Anastomotic stricture  
Anastomotic leaks



Contrast through gastrostomy goes up to the lower esophagus, the NG tube is stopped at the upper esophagus, and the gap is measured 6 vertebral bodies.



Main principle of surgery: to remove the fistula and anastomose the two ends of the esophagus.

Note that the proximal part of the esophagus is dilated due to saliva collection.