Congenital diaphragmatic hernia

Done by: Dana Alnasra

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
– screening for associated abnormalities – fetal ECG – genetic studies – family counseling – in utero fetal therapy: investigational procedures, patch closure, **fetoscopic endoluminal tracheal occlusion** – delivery planning	– optimizing cardiorespiratory status: reduce lung compression, ventilatory support, cardiovascular support, correction of acid-base status, **correction of pulmonary hypertension** – achieving hemodynamic stability (you need to know the details) – screening for associated malformation – increased survival rate up to 92%

Operative management: CDH is not a surgical emergency

Minimally invasive surgery is preferred more than the open approach

- advantages
  - decreased surgical stress, postop pain, and lenght of hospital stay
  - better respiratory compliance
  - increased survival rate
  - avoidance of thoracotomy associated complications
- VS.
- disadvantages
  - CO2 insufflation causes hyper capnia, acidosis, increased pulmonary hypertension, and right-to-left shunting
  - increased recurrence rate

Criteria for MIS:

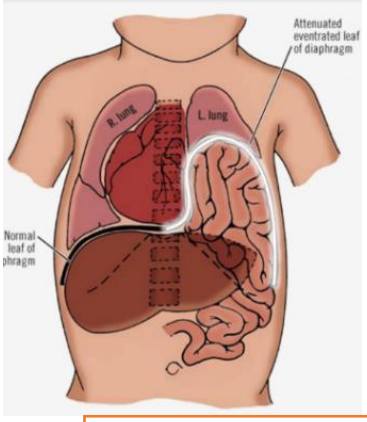
- **Neonates:** hemodynamic, radiographic and respiratory stability, and no severe associated cardiac malformations.
- **Infants & children:** late presenting or incidentally diagnosed

Outcomes: Mortality and morbidity are related to:

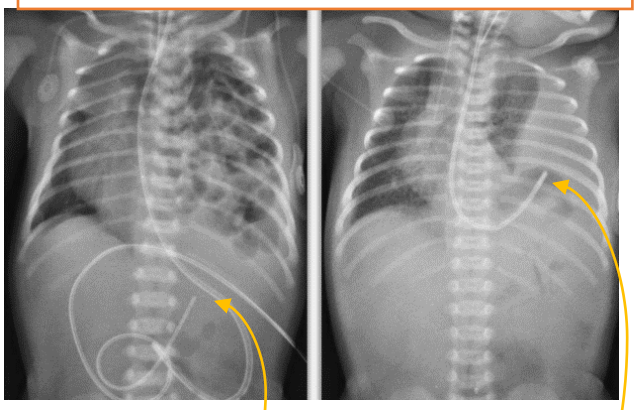
- Severity of lung hypoplasia
- Pulmonary hypertension
- Associated anomalies
- Prematurity

Appendix 1

Diaphragmatic eventration



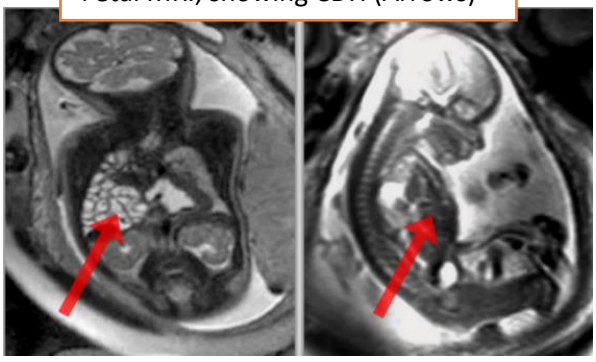
Chest x-ray in a neonate with CDH: bowel loops in the chest, mediastinum shifted to the right



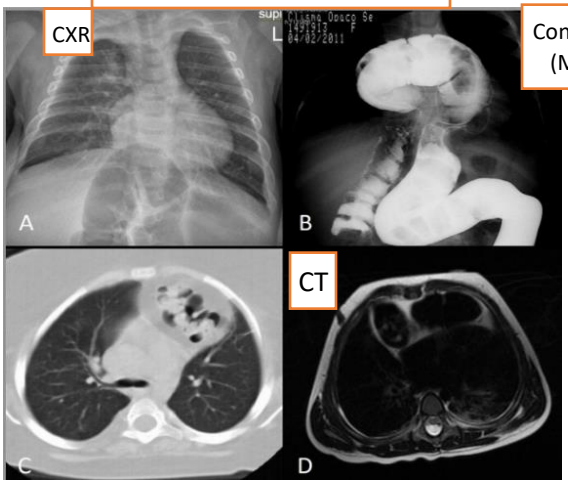
NG tube reaching the bowel; the stomach is not herniated

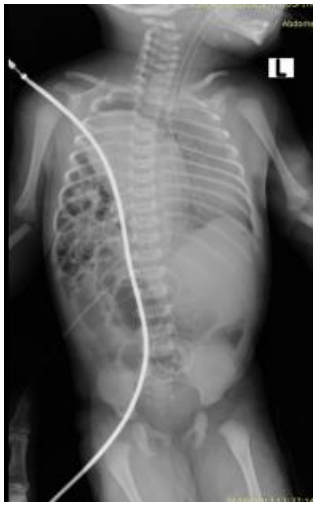
CDH + NG tube only reaching the chest; stomach is herniated

Fetal MRI, showing CDH (Arrows)



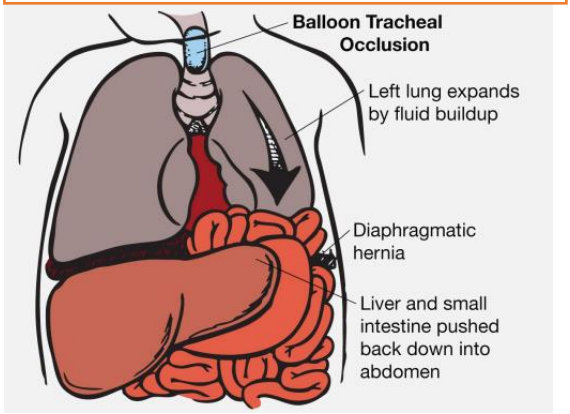
Dx in Infants &children



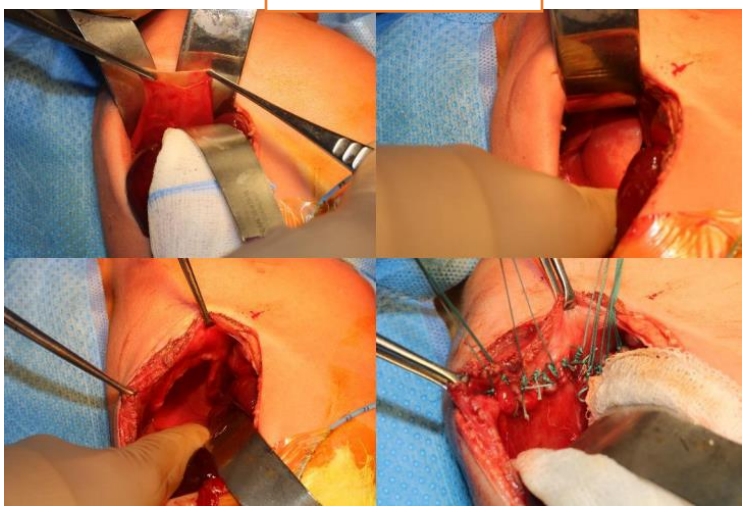


Right sided CDH; note the liver is also herniated

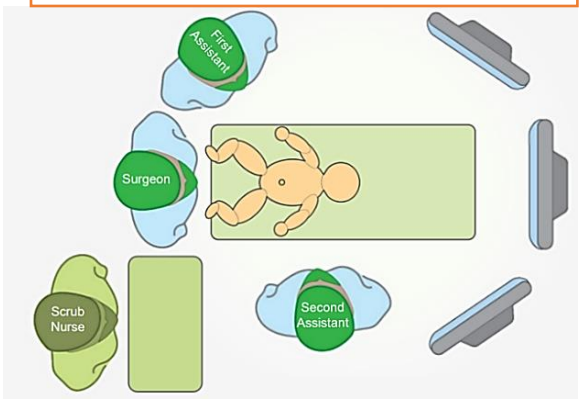
Fetoscopic Endoluminal Tracheal Occlusion; prevents lung hypoplasia



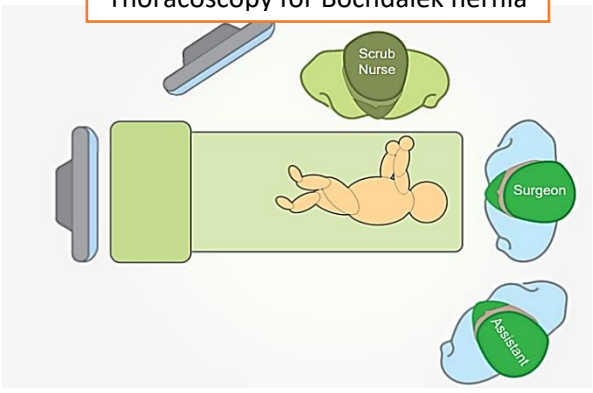
Open CDH repair



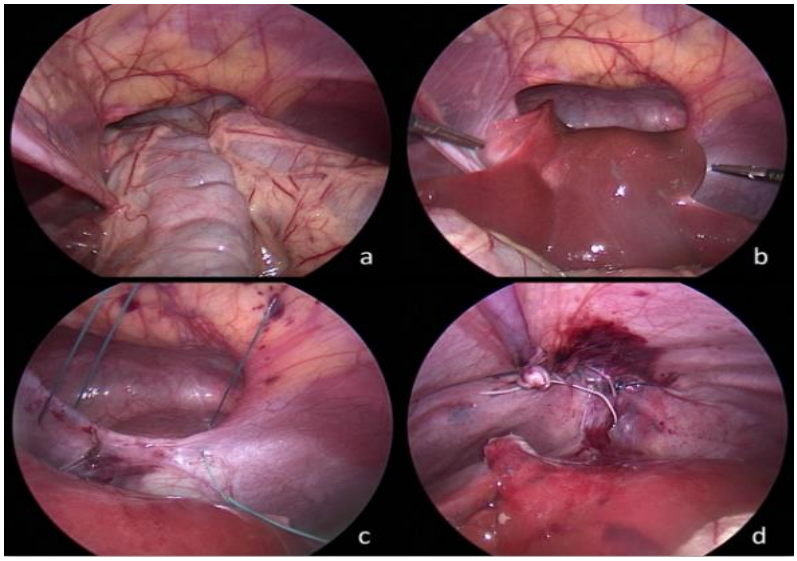
Thoracoscopy for Morgagni-Larrey hernia



Thoracoscopy for Bochdalek hernia



Minimally invasive surgery



Esophageal atresia (EA) and tracheoesophageal fistula (TEF)

Etiology:

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

Normal embryogenesis of the trachea and esophagus:

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

1. **Antenatal:** polyhydramnios, absent/small stomach bubble (both are nonspecific)
2. **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.

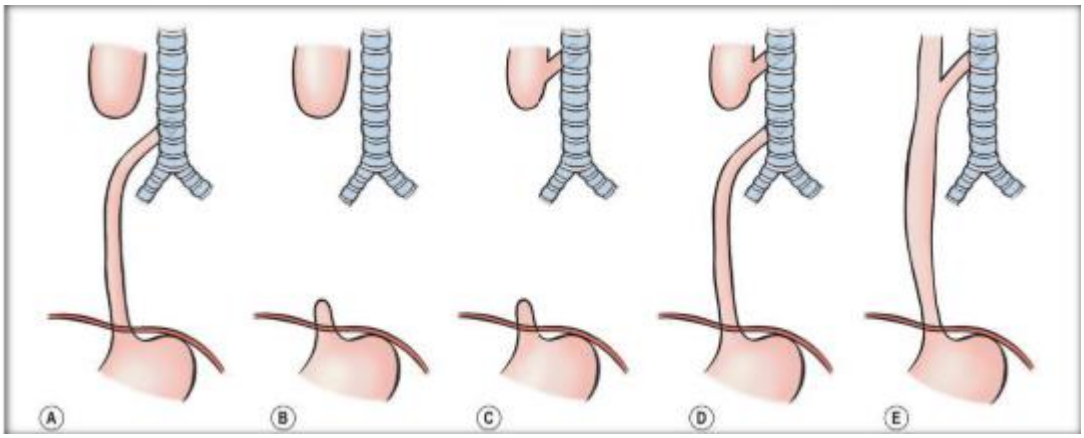
- **Preoperative preparation:**
  a. Continuous suctioning tube in the upper esophagus.
  b. Head-up position or on the side.
  c. Gentle low-pressure ventilation if baby is in respiratory distress.
- **Preoperative workup:**
  ECG (to rule out cardiac and aortic arch anomalies), renal ultrasound, and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

Complications:

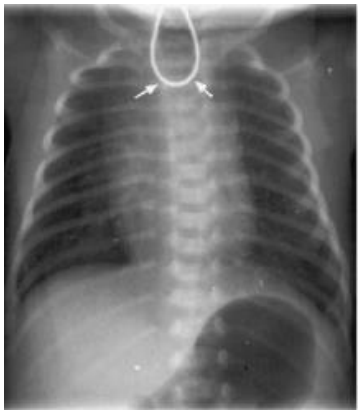
- **anastomotic strictures (17-60%)**
- anastomotic leaks (3.5-17%)
- recurrent TEF (3-15%)
- tracheomalacia
- disordered peristalsis → GERD → esophageal cancer ???
- vocal cord dysfunction
- respiratory morbidity
- thoracotomy-related morbidity

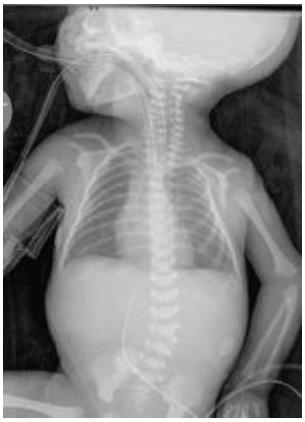
Appendix 2

Classification of EA & TEF

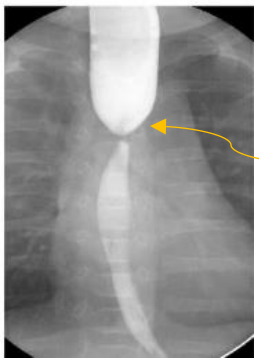


Coiling of tube in the blind upper pouch around T2-T4. Note that there's gas in the stomach, indicating a TEF (type A).





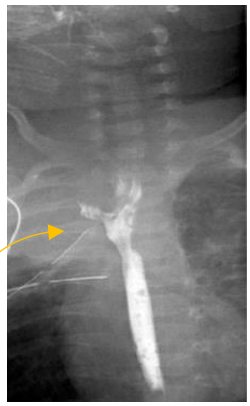
Opacity of the abdomen + NG tube stopped at the upper esophagus → proximal atresia without TEF



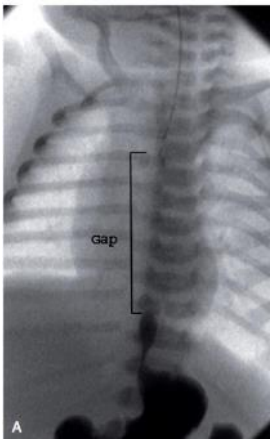
Surgery complications:

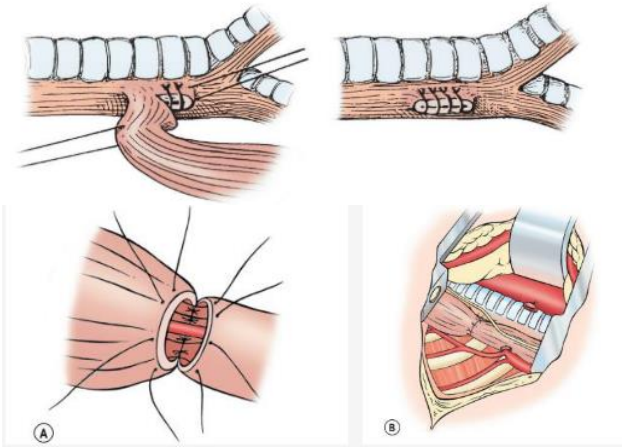
Anastomotic stricture

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.