CDH

Congenital Diaphragmatic Hernia

WHAT IS 'CDH'?

A developmental discontinuity of the diaphragm..

that allows abdominal viscera to herniate into the chest (large bowel, small bowel, liver, stomach, spleen)

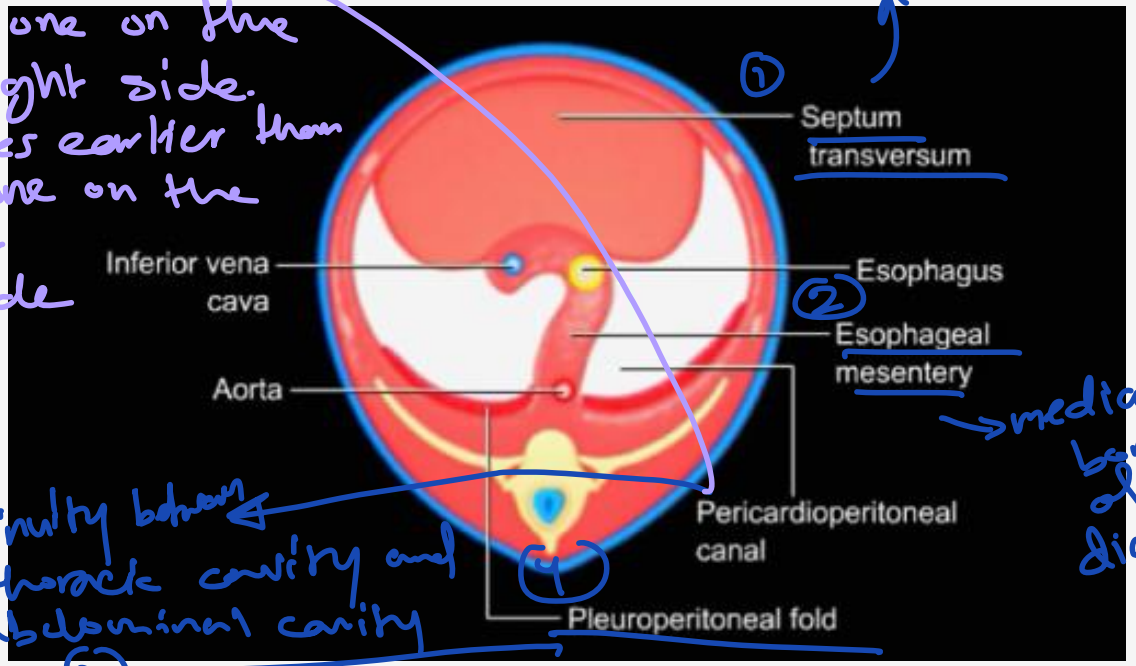
↓

pressure on the lung → hypoplastic

1 in 3,000 live births

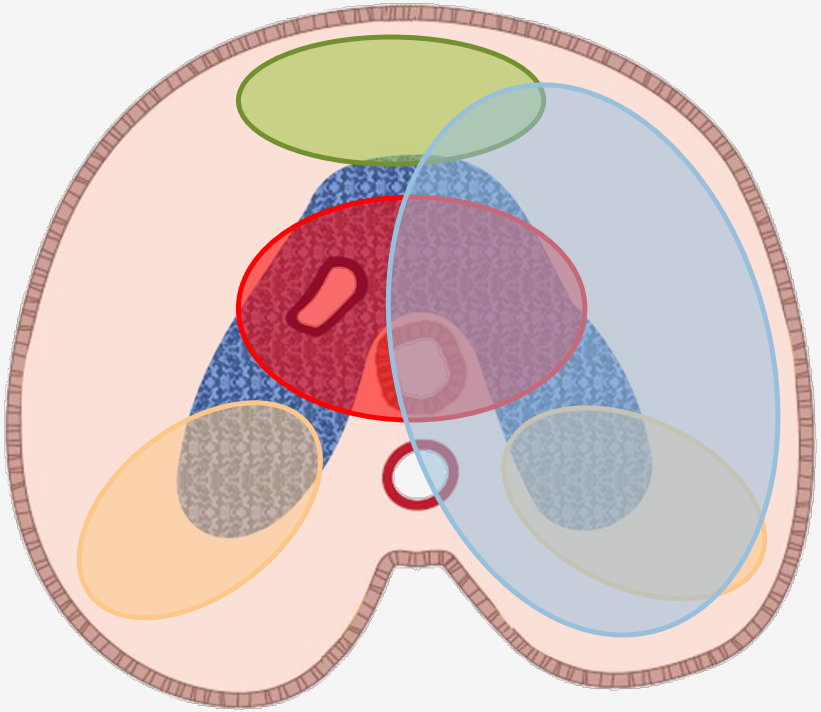
WHY DOES IT HAPPEN?

- Genetic
  - mostly sporadic
- Environmental
  - vitamin A deficiency
  - thalidomide
  - anticonvulsants
  - quinines



TYPES OF CDH

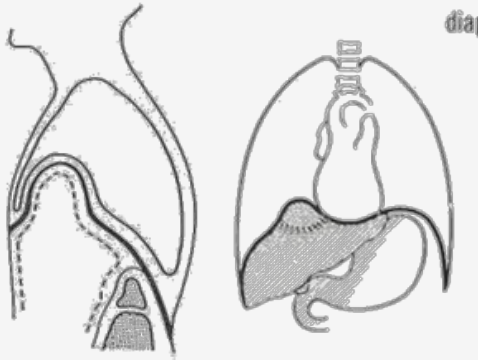
- Postero-lateral Hernia (Bochdalek)
  - most common (80-90%)
  - left side (80-85%)
- Anterior Hernia (Morgagni-Larrey)
  - 2% of all CDHs
- Central Hernia
  - extremely rare
  - involves the central tendon
- Diaphragm Agenesis

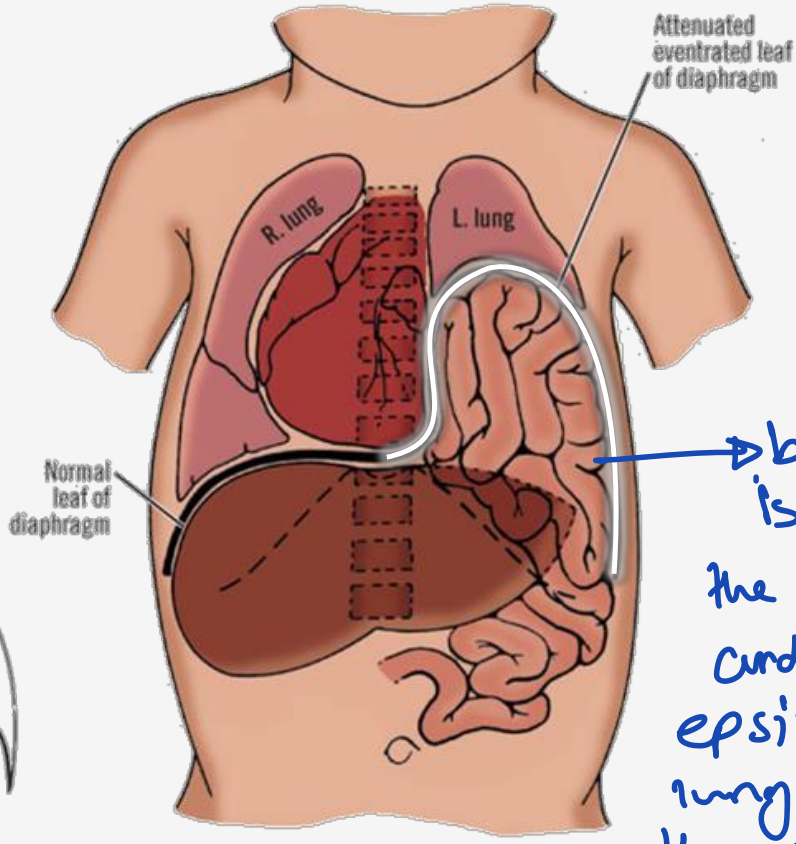


M. Lima, G. Ruggeri
Chirurgia Pediatrica
EdiSES

TYPES OF CDH

- Diaphragmatic Eventration
  - Abnormal elevation of congenitally thin, hypoplastic but intact diaphragm
  - Total or partial





bowel is pressing the diaphragm and the epsilateral lung as there's a CDH so it's considered

part of it

WHY TO CONCERN ABOUT?

- Fetal mortality (severe form described beside the figure)
  - Hydrops fetalis (as above)
  - Stillbirths
- Neonates
  - Pulmonary hypoplasia
  - Persistent Pulmonary hypertension
  - Right-to-left shunting
  - Hypoxemia & acidosis
  - Cardiorespiratory failure
  - Mortality
- Infants and children
  - Respiratory manifestations (Recurrent RTI)
  - GI manifestations
  - Asymptomatic (incidental)

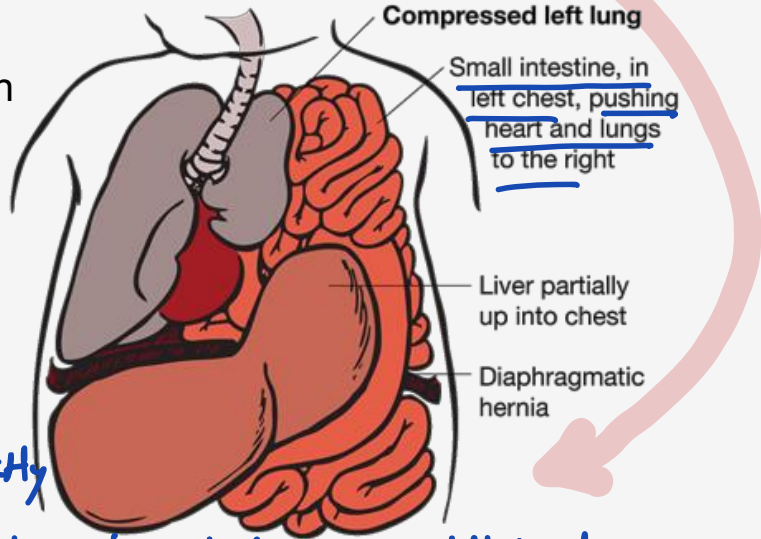
deoxygenated blood from the right side of the heart to the left side (aorta) by a shunt (PDA) → ↑tissue hypoxia → acidosis (cycle)

born with minimal defect so they

should be managed directly when born (↓shunting ↓pulmonary HTN ↓acidosis ↑O_2)

(supporting cardic + respiratory

most common type



hypoplastic lungs bilateral → respiratory insufficiency + Arteries are hypertrophied arteries (small lumen) →

ASSOCIATED ANOMALIES

- Isolated CDH
  - 50-70% of cases
  - ↑ survival
- Complex CDH
  - 30-50%
  - ↓ survival

associated with other anomalies

Structural Malformations
Neural Tube Defects
Cardiac Malformations
Bronchopulmonary Sequestration
Renal Malformations

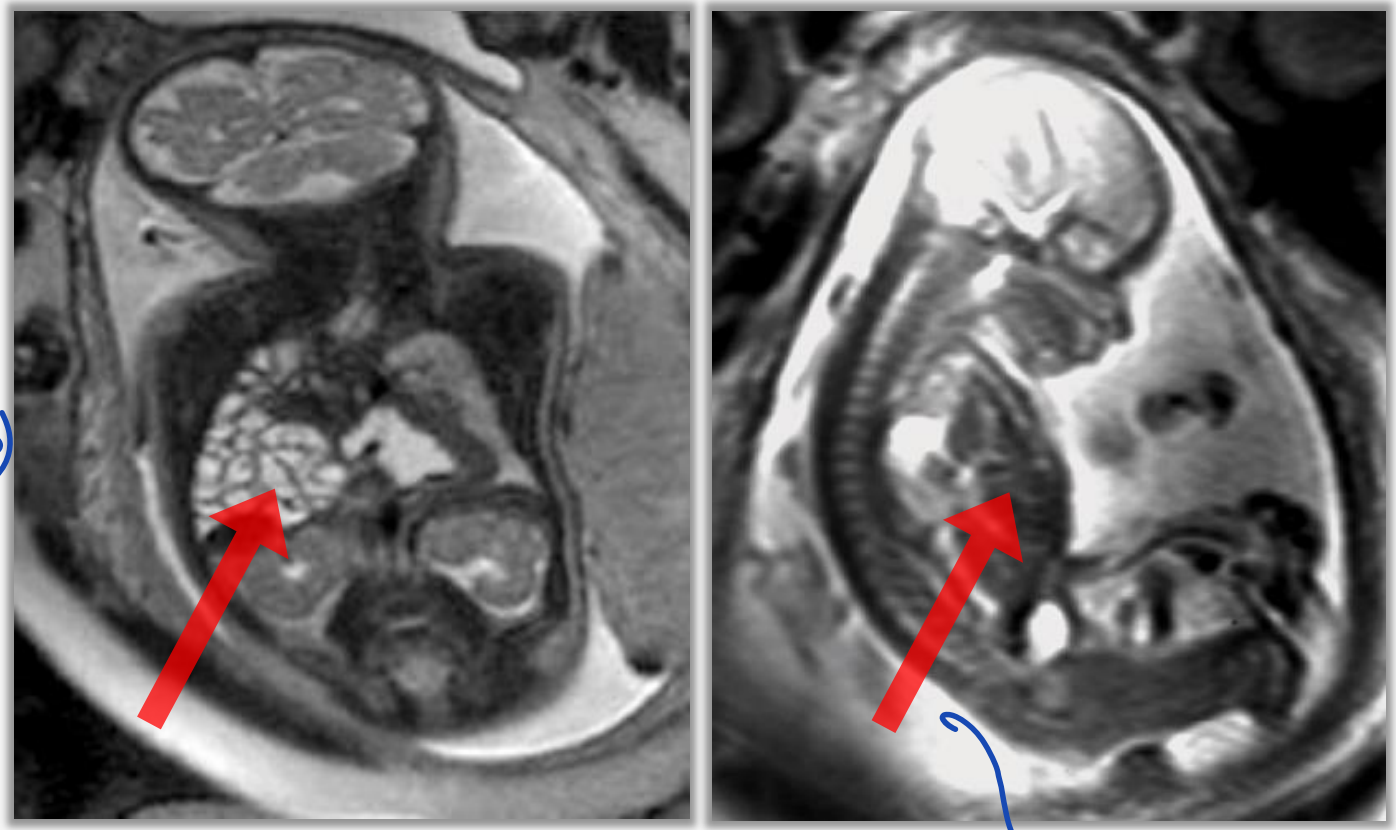
Chromosomal Abnormalities
Trisomies 18, 13, and 21
Karyotype abnormalities

Underlying Syndromes
CHARGE syndrome
Beckwith Wiedemann syndrome
Pentalogy of Cantrell

Don't memorize this slide

HOW TO DIAGNOSE?

- Prenatal
  - Fetal ultrasound screening
  - Fetal MRI (if suspicious)



→ Liver tissue is herniated towards the

thoracic cavity

HOW TO DIAGNOSE?

any baby tested with RD think of CHD as the first differential diagnosis

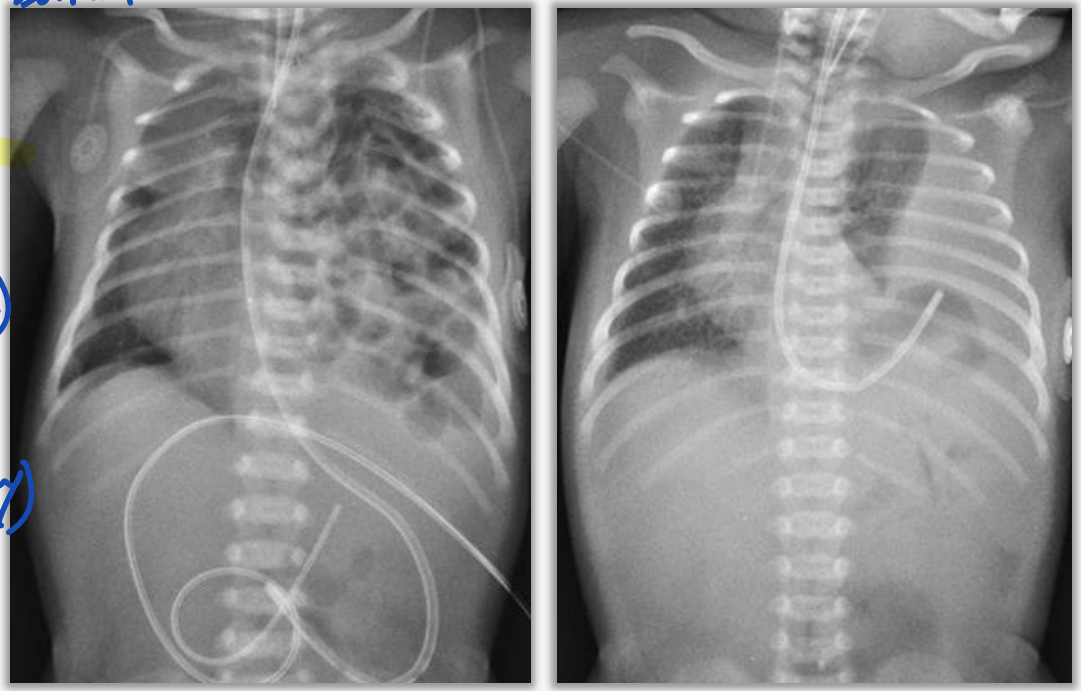
- Postnatal
  - **Respiratory distress**
  - Physical examination
  - **Chest x-ray** (diagnostic)
  - ± CT or MRI (query)
  - ± GI contrast study (query)

this triad is almost diagnostic

① scaphoid abdomen (empty of structures)

② absent or decreased breath sounds on the affected side

③ bowel sound when auscultating the chest



- shifted mediastinum
- bowel loops
- remnant lung tissue.

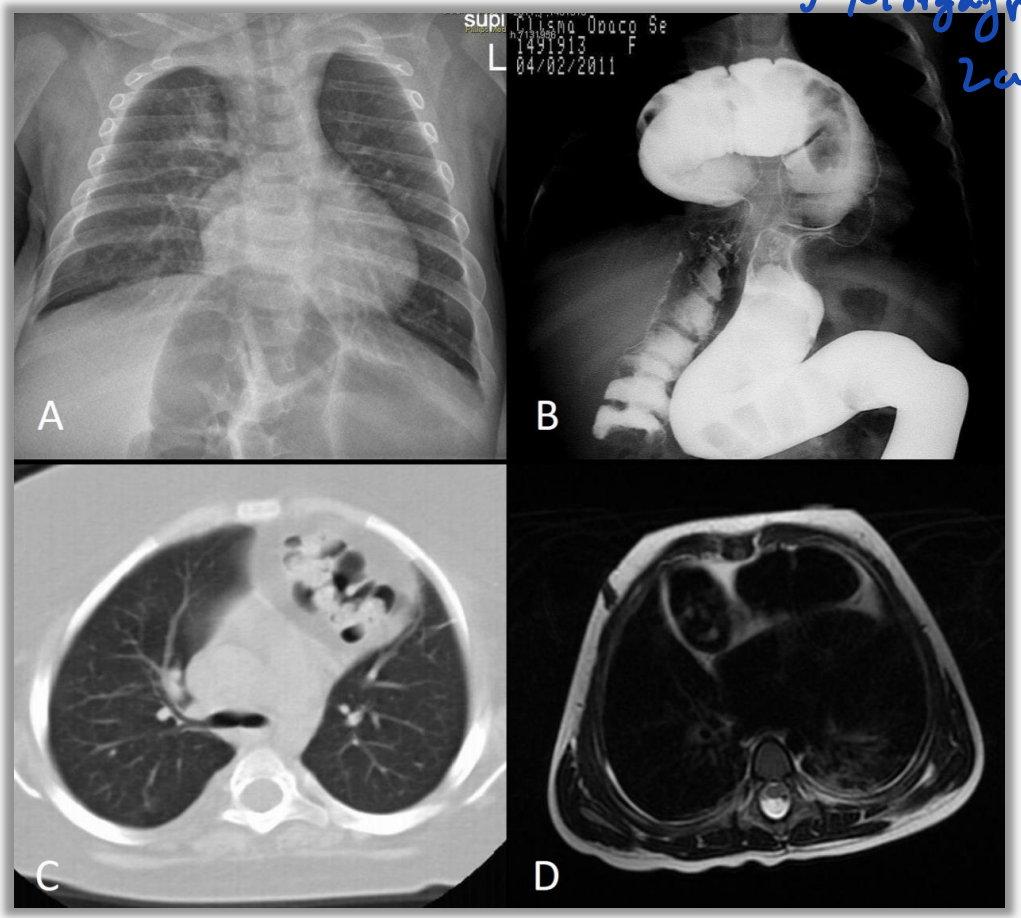
- NG tube → stomach isn't herniated with the herniated

HOW TO DIAGNOSE?

anterior part of the diaphragm
Morgagni - Larrey

- Infants and children
  - Respiratory, or GI symptoms (nonspecific)
  - Physical examination (nonspecific)
  - **Incidental finding** on chest x-ray
  - ± CT or MRI
  - ± GI contrast study

abnormal shadow



*Not all late presentation are anterior herniation

PROGNOSTIC FACTORS

- Associated malformations
  - ↓ SR
- Right-sided defects
  - ↓ SR (50% Vs 75% for left-sided)
  - ↑ ECMO rate
  - ↑ patch rate
- Liver herniation
  - ↓ SR (45% Vs 74% if not herniated)
- Fetal lung volume
  - ↓ SR if <30% of expected vol. for GA
- Lung area to head circumference ratio (LHR)
  - more indicative of morbidity than mortality

high variation normally

HOW TO MANAGE?

through mouth- larynx- trachea → inflating the balloon → ↑ pressure inside the lung over come the hernial pressure.

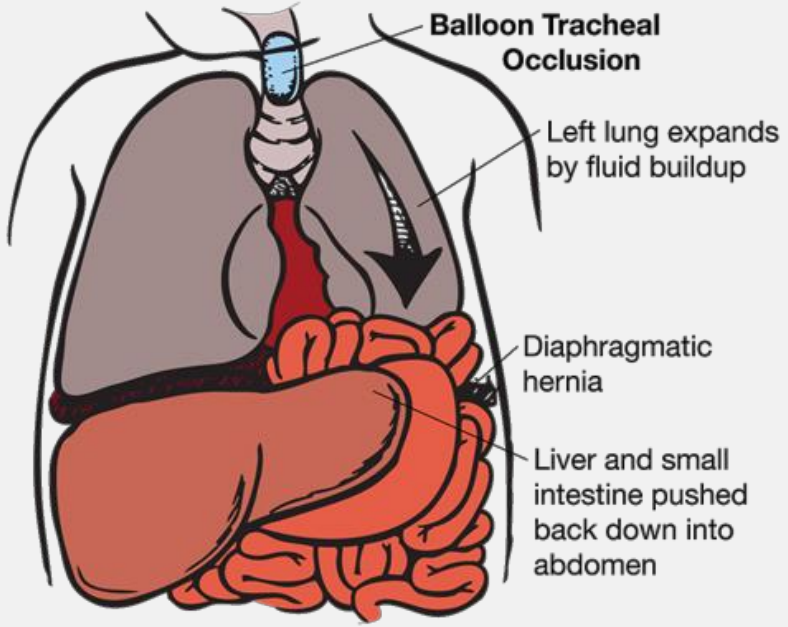
- Prenatal Management
  - Screening for associated abnormalities
  - Fetal echocardiography
  - Genetic studies
  - Family counselling
  - **In utero fetal therapy**
  - Delivery planning

Sample from amniotic fluid or cardic screening

- Investigational procedures

Before his delivery the baloon is removed

  - Patch closure (abandoned)
  - Fetoscopic Endoluminal Tracheal Occlusion (FETO)



HOW TO MANAGE?

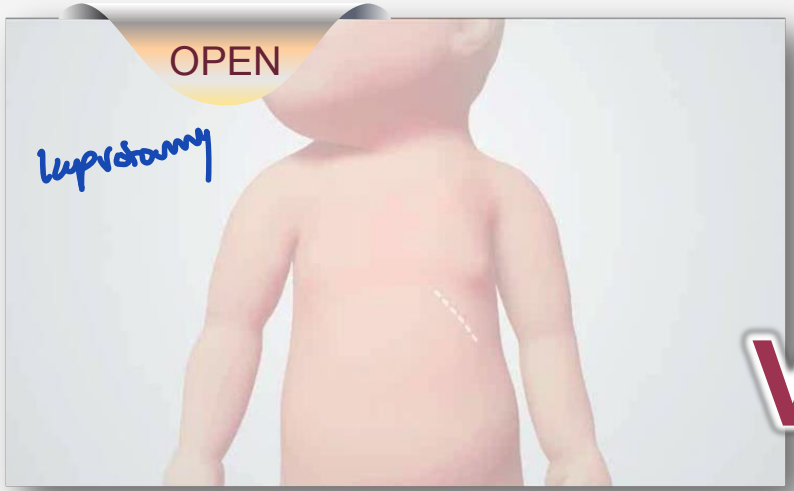
- Postnatal Management
  - Since mid-1980s - No more surgical emergency
  - Optimizing cardiorespiratory status
  - Achieving hemodynamic stability
  - Screening for associated malformations
  - ↑ SR up to 92% (instead of 50%)

- Main points:
  1. Reduce lung compression
     - Immediate intubation post-delivery
     - NGT
  2. Ventilatory support
     - HFOV in most cases
     - ECMO in sever cases
     - Liquid ventilation ?
  3. Cardiovascular support
     - Proper venous access
     - IV fluids
     - Inotropic agents
  4. Correction of acid-base status
  5. Correction of pulmonary hypertension
     - Maintain MAP ≥50 mmHg
     - Reverse right-to-left shunting
     - Inhaled Nitric Oxide (iNO)
     - Sildenafil
  - Surfactant ?
  - Hydrocortisone ?

HOW TO MANAGE?

- Operative Management

HOW ?



Vs

WHY 'MIS'?

- ↓ Overall surgical stress
- ↓ Postoperative pain
- Better respiratory compliance
- ↑ Survival rate
- ↓ Length of hospital stay
- Avoidance of thoracotomy-associated complications

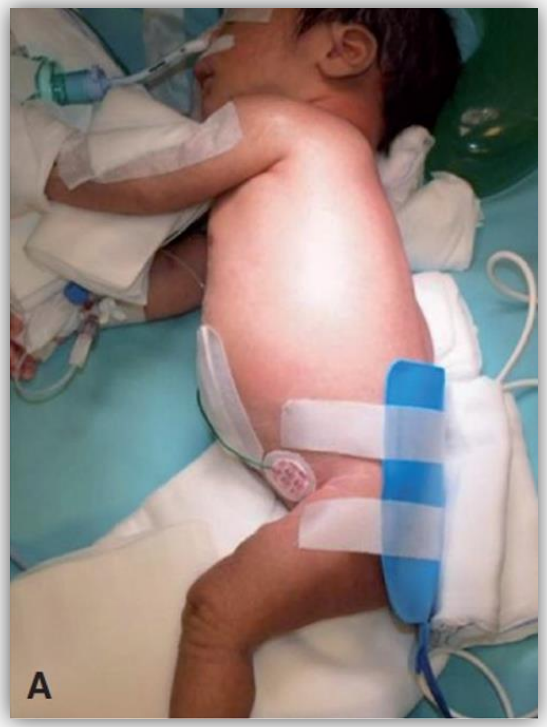
benefits > risks

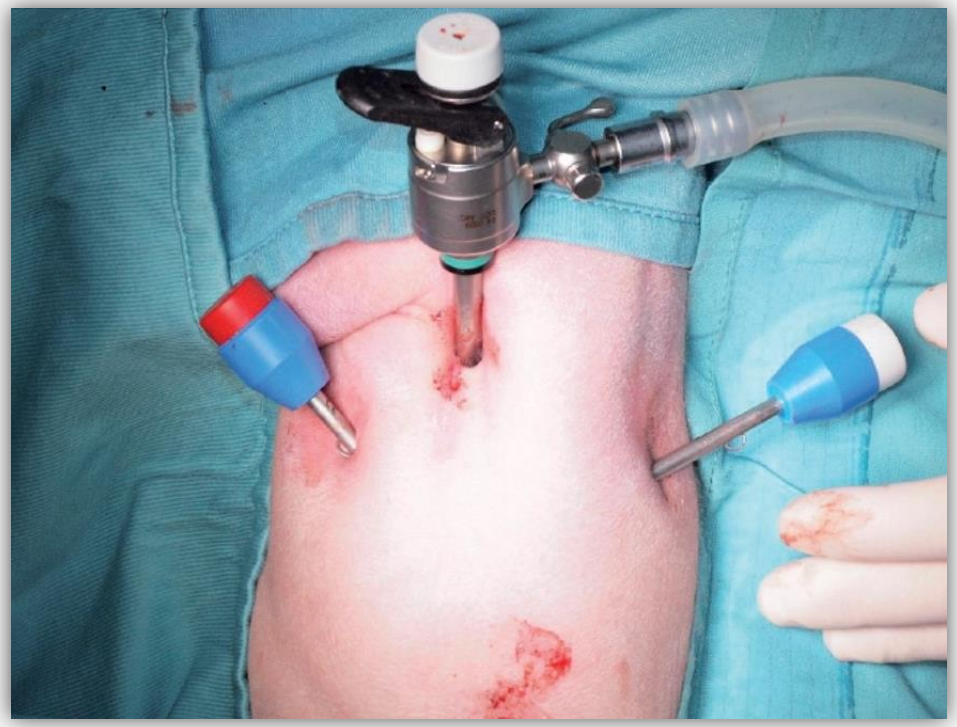
- CO_2 insufflation
  - Hypercapnia and acidosis
  - ↑ Pulmonary hypertension
  - ↑ Right-to-left shunting
- ↑ Recurrence rate

MIS

3 port

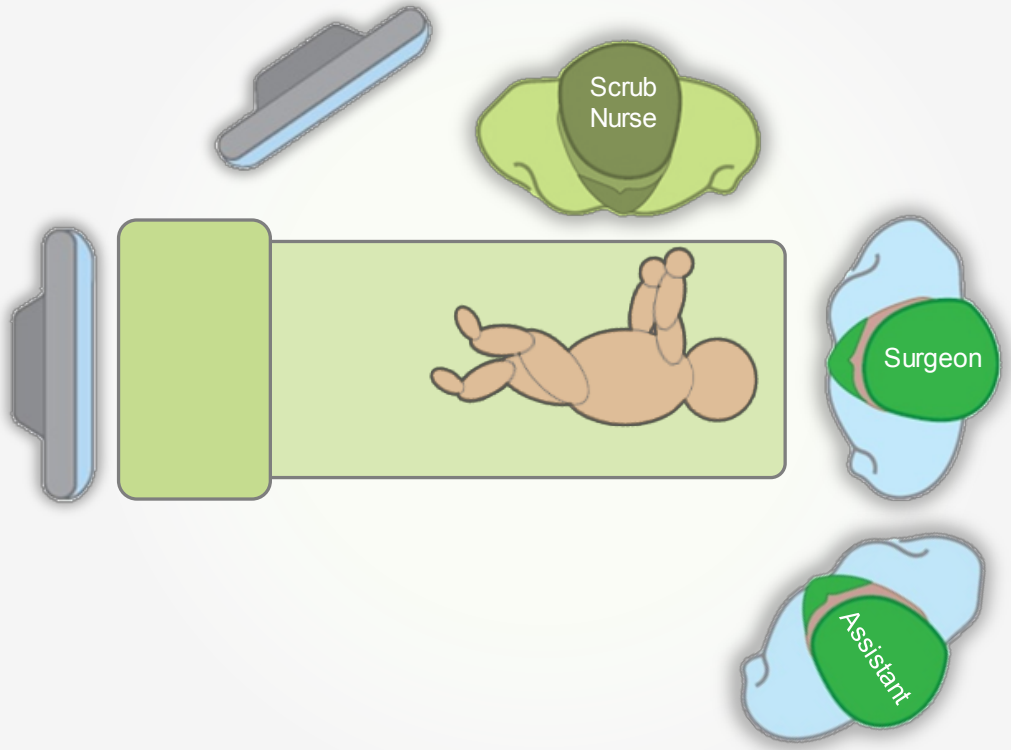
Thoracoscopy for Postero-Lateral (Bochdalek) Hernias





MIS

Thoracoscopy for Postero-Lateral (Bochdalek) Hernias



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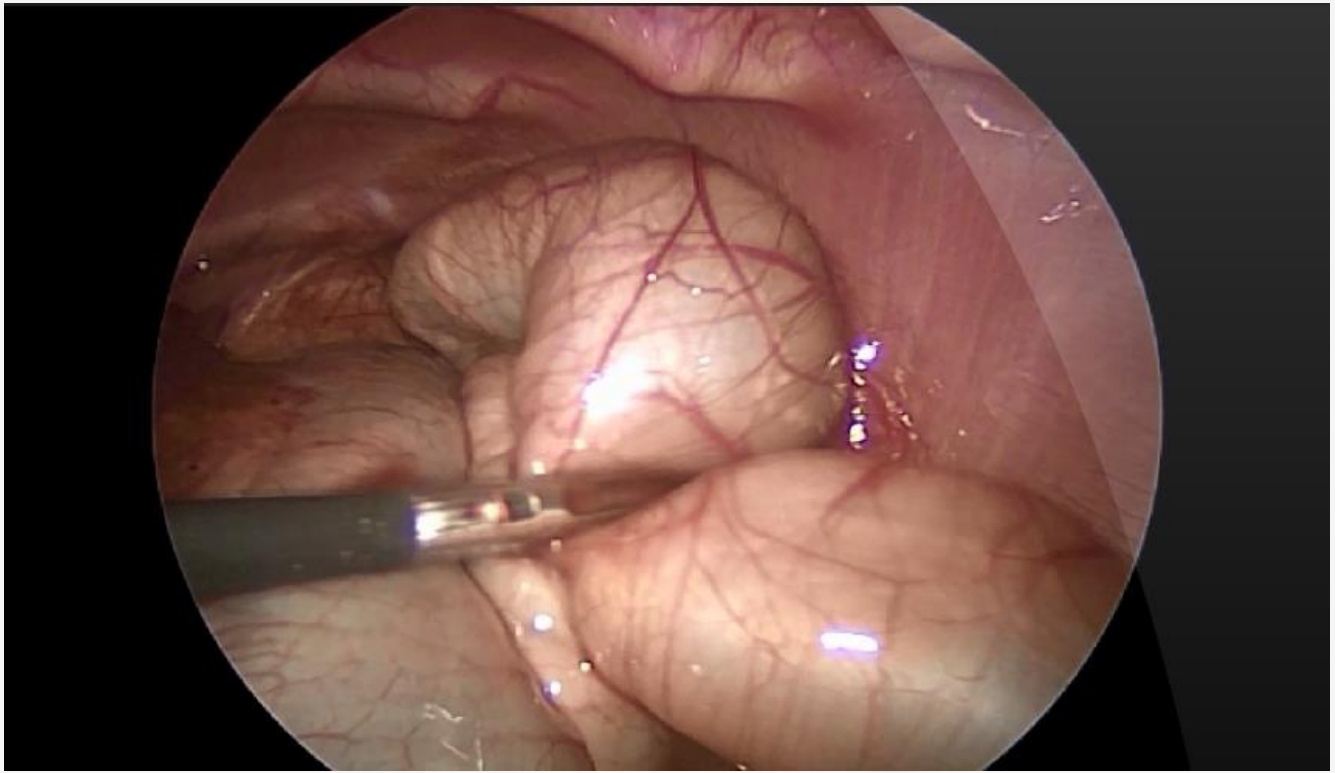
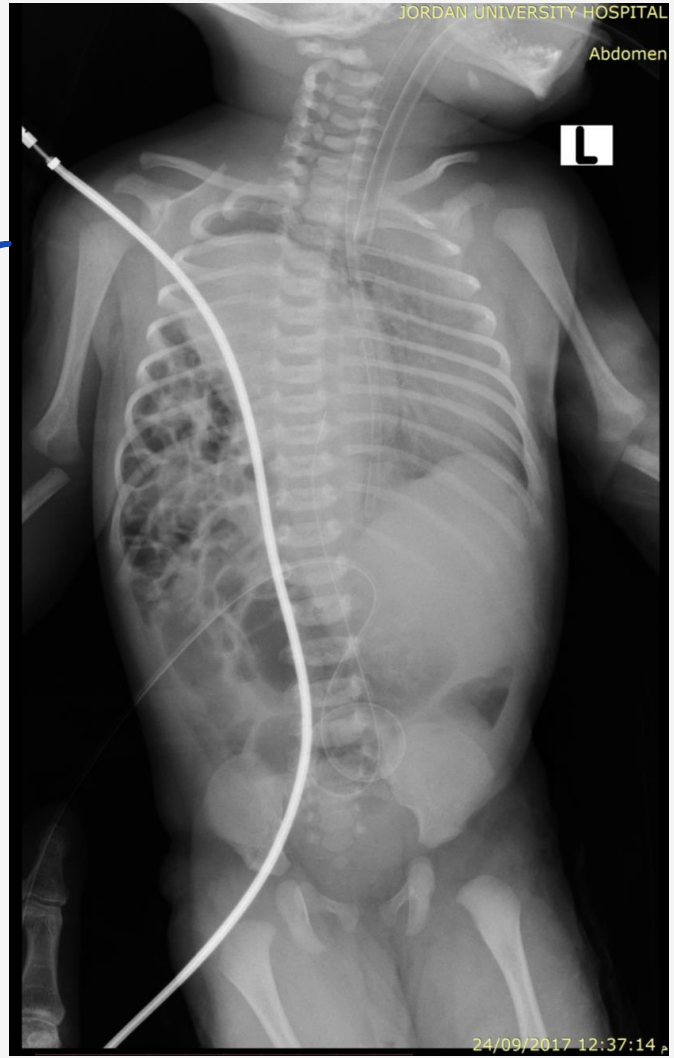
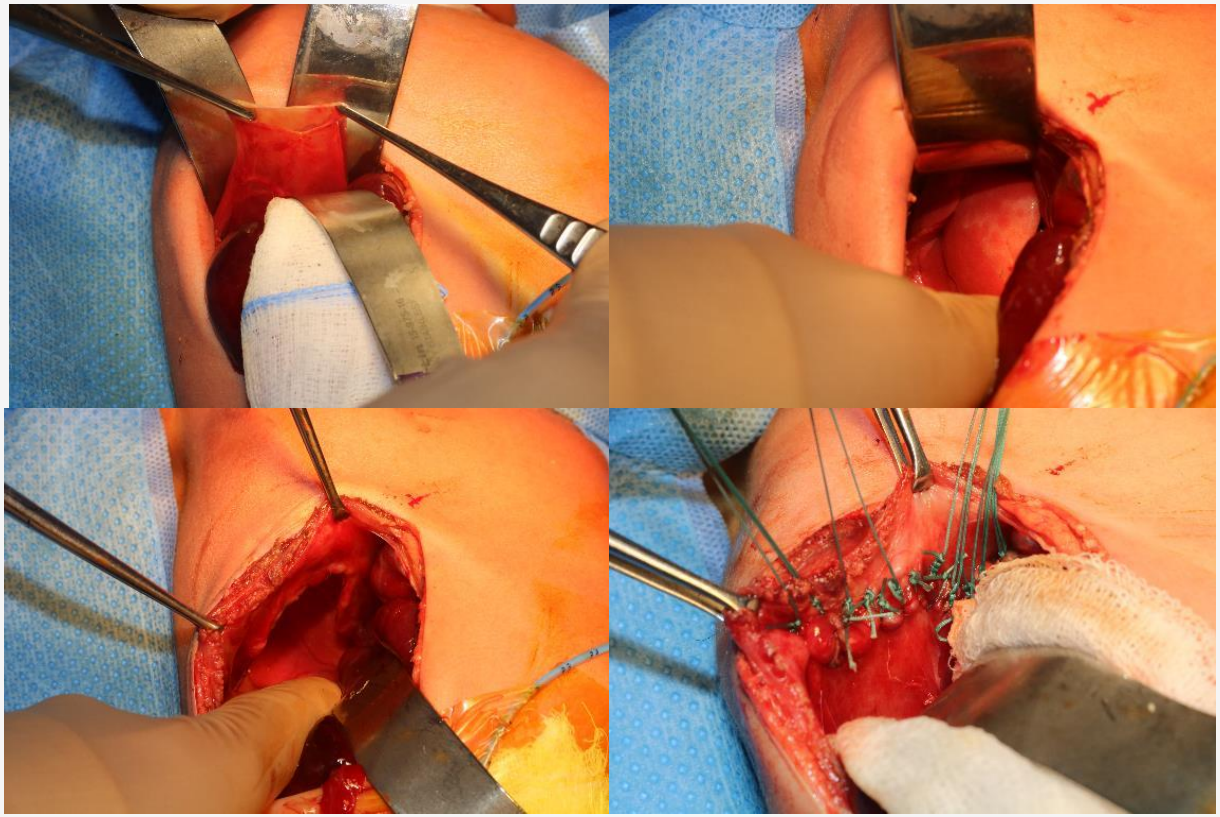


OPEN REPAIR

right sided diaphragmatic hernia:
Liver is totally elevated
(↓ survival rate)

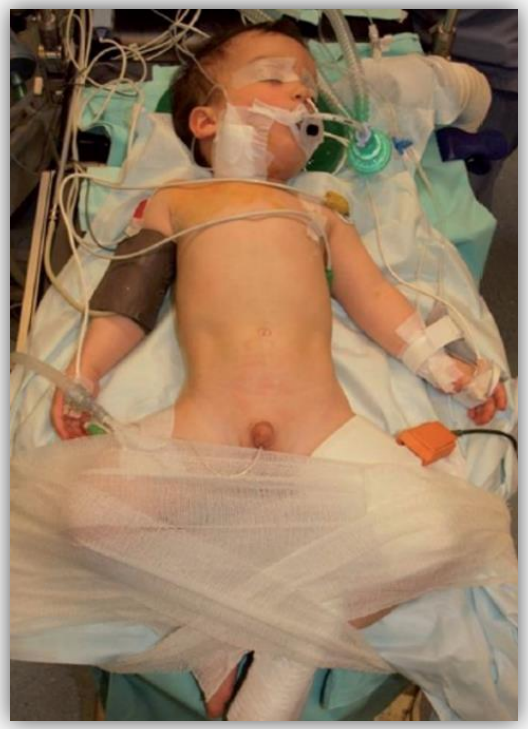


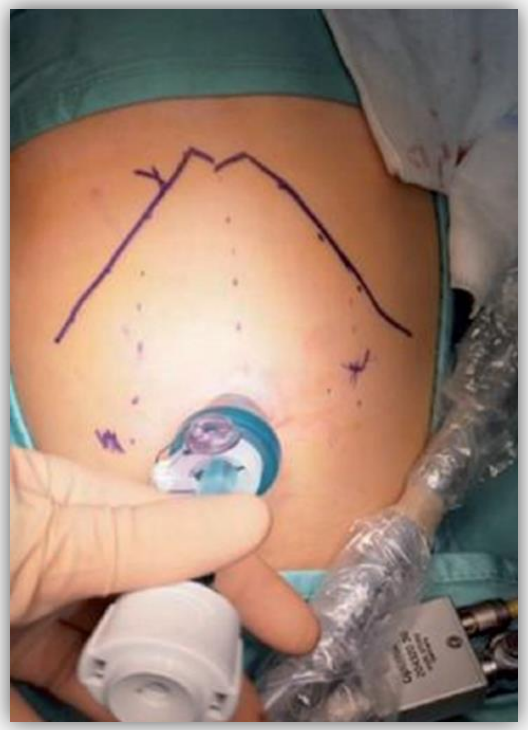
OPEN REPAIR



MIS

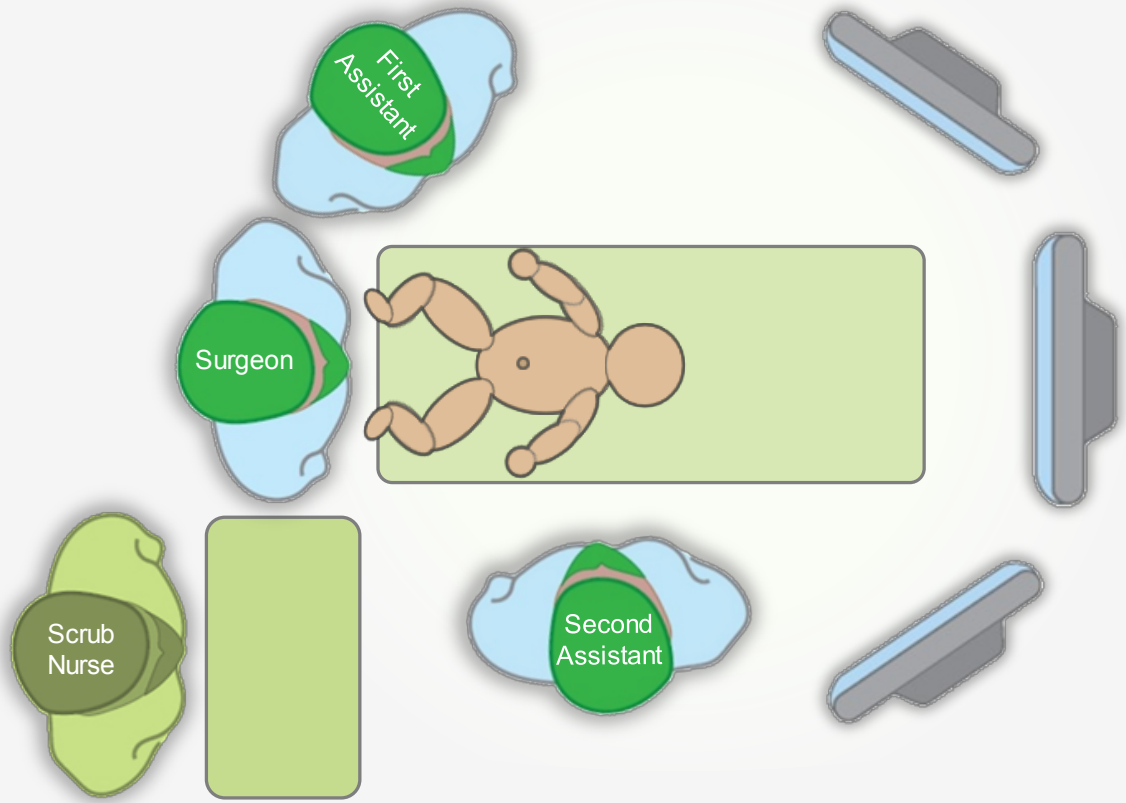
Laparoscopy for Anterior (Morgagni-Larrey) Hernias





MIS

Laparoscopy for Anterior (Morgagni-Larrey) Hernias



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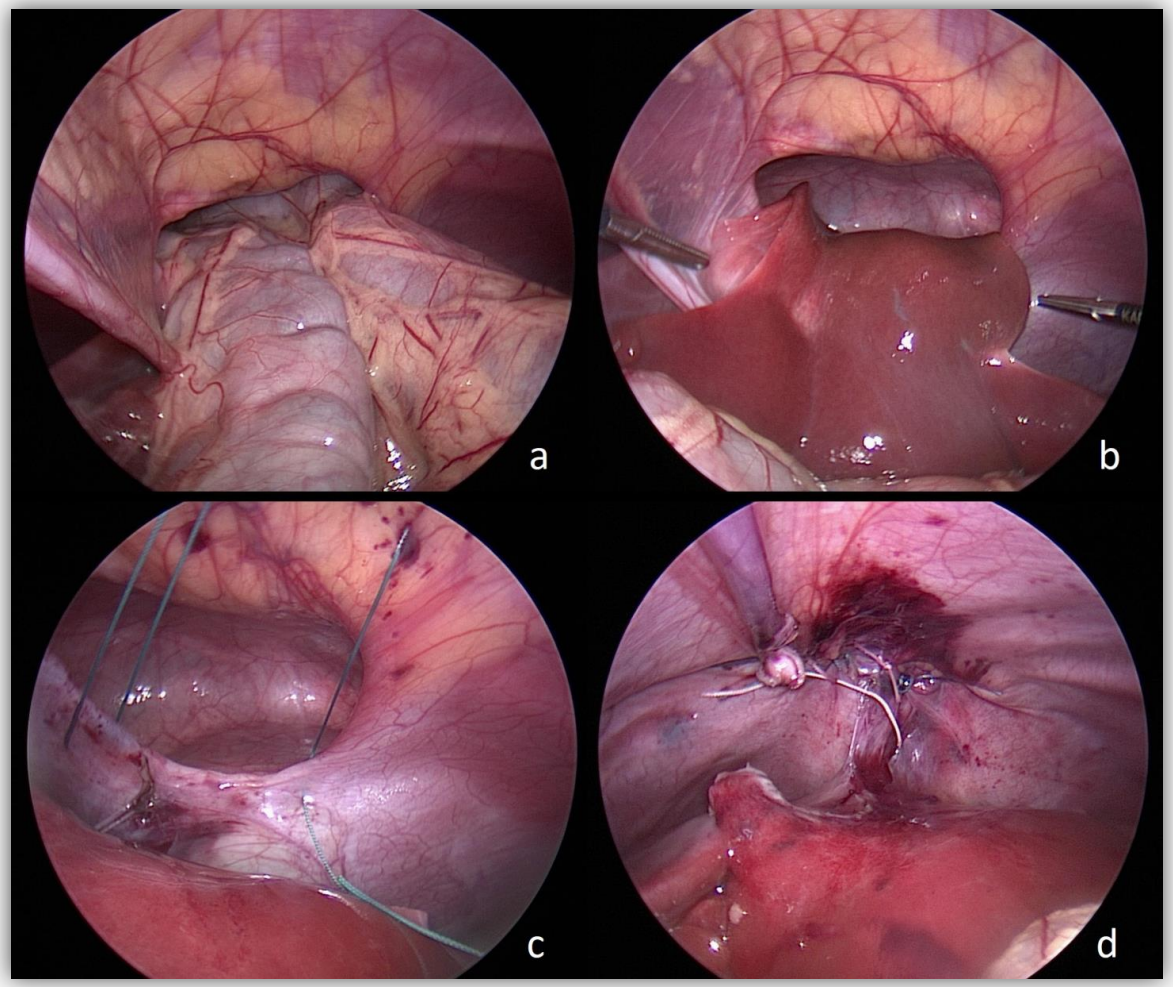


OUTCOME

- Mortality and morbidity are related mainly to:
  - severity of lung hypoplasia
  - pulmonary hypertension
  - associated anomalies
  - prematurity

EA ± TEF

Esophageal Atresia ± Tracheoesophageal Fistula

WHY DOES IT HAPPEN?

- 4th week of gestation → foregut starts to differentiate (ventral respiratory and dorsal esophageal)

- Formation of lateral tracheoesophageal folds → tracheoesophageal septum

- 6-7 weeks of gestation → separation is complete

WHY DOES IT HAPPEN?

- Genetic factors

- Environmental factors
  - ✓ methimazole
  - ✓ OCPs
  - ✓ progesterone and estrogen exposure
  - ✓ maternal diabetes
  - ✓ thalidomide exposure
  - ✓ fetal alcohol syndrome
  - ✓ maternal phenylketonuria

- Chromosomal anomalies (trisomy 18 & 21)

EPIDEMIOLOGY

- Incidence: 1 in 2500-3000 live births
- Slight male preponderance (1.26:1)

ASSOCIATED ANOMALIES

- Isolated EA (50%)

- Syndromic EA (50%):
  - ✓ Cardiac (m.c)
  - ✓ Vertebral
  - ✓ Limb
  - ✓ Anorectal
  - ✓ Renal

ASSOCIATED ANOMALIES

VACTERL

ASSOCIATED ANOMALIES

Vertebral, **A**norectal, **C**ardiac, **T**racheo-**E**sophageal, **R**enal, and **L**imb abnormalities

ASSOCIATED ANOMALIES

CHARGE

ASSOCIATED ANOMALIES

just read

Coloboma

Heart defects

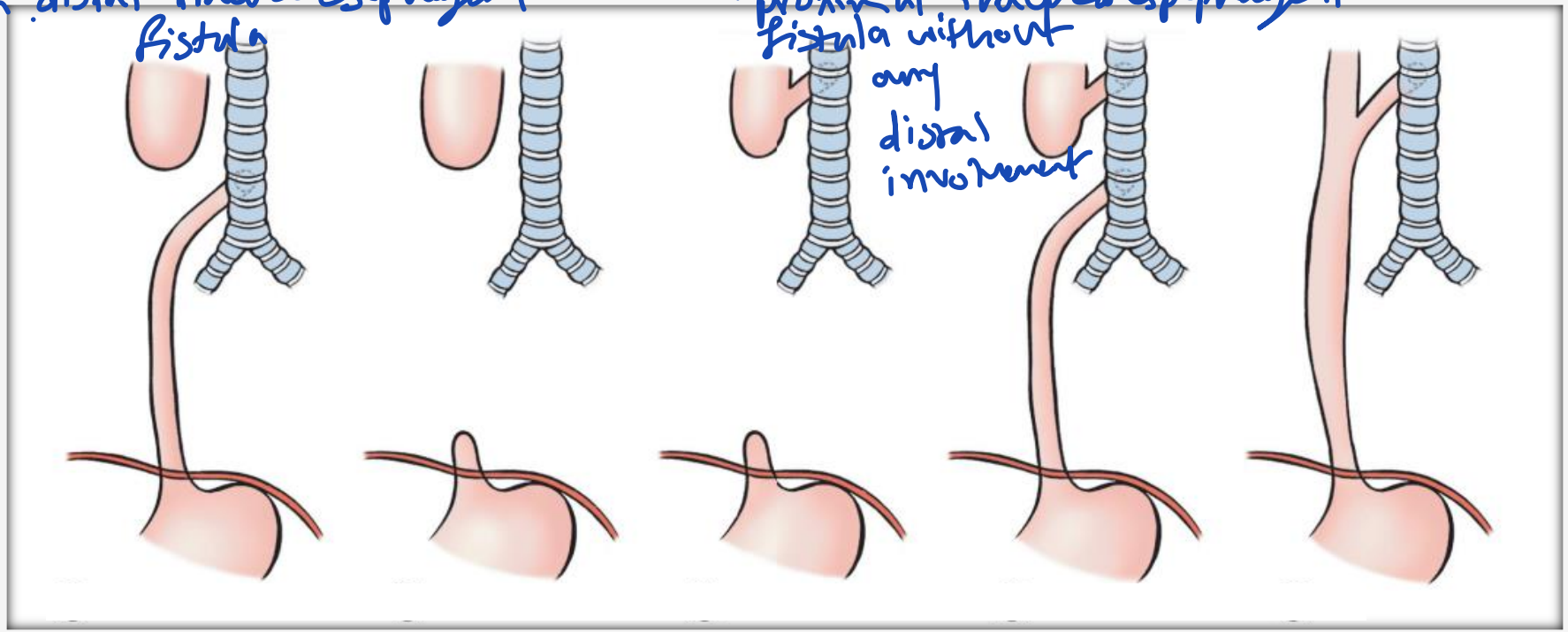
Atresia of the choanae

developmental **R**etardation

Genital hypoplasia

Ear deformities

CLASSIFICATION



85% | 7% | 2% | <1% | 4%

proximal atresia

proximal

without any fistula

fistula with distal fistula

ANTENATAL DIAGNOSIS

Two nonspecific signs:

- ✓Polyhydramnios → gi obstruction
- ✓Absent or small stomach bubble

POSTNATAL DIAGNOSIS

- Excessive salivation (because of closed esophagus)

- Coiled feeding tube in the blind upper pouch around T2–T4 on chest x-ray

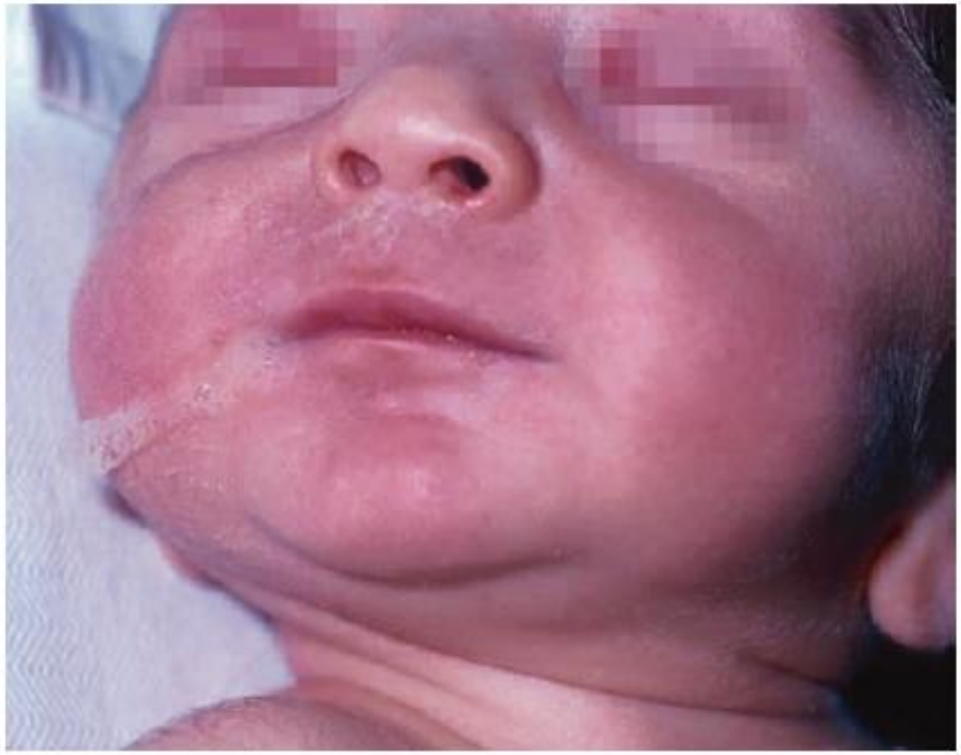
POSTNATAL DIAGNOSIS

- Presence/absence of gas in the stomach and bowel on abdominal x-ray

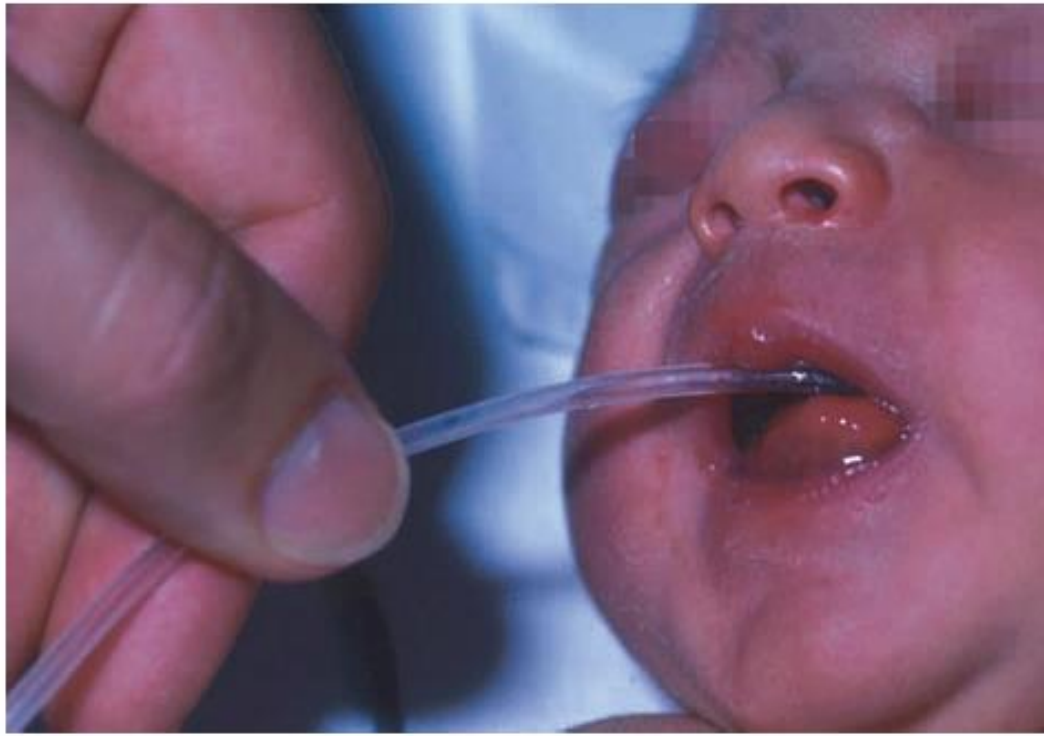
  → *assign the type of EA*

- +/- Contrast study (usually not needed)

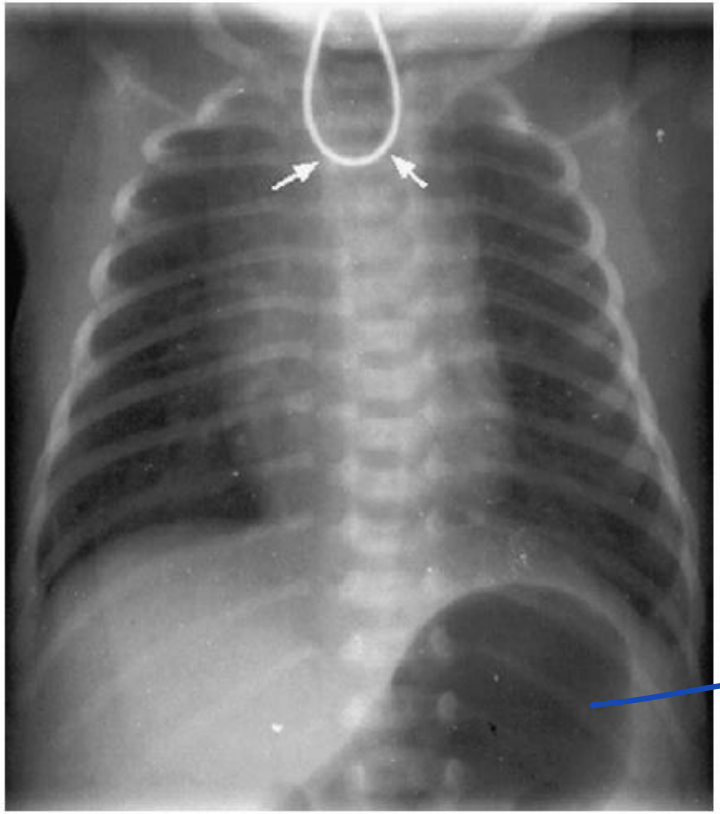
POSTNATAL DIAGNOSIS



excessive salivation



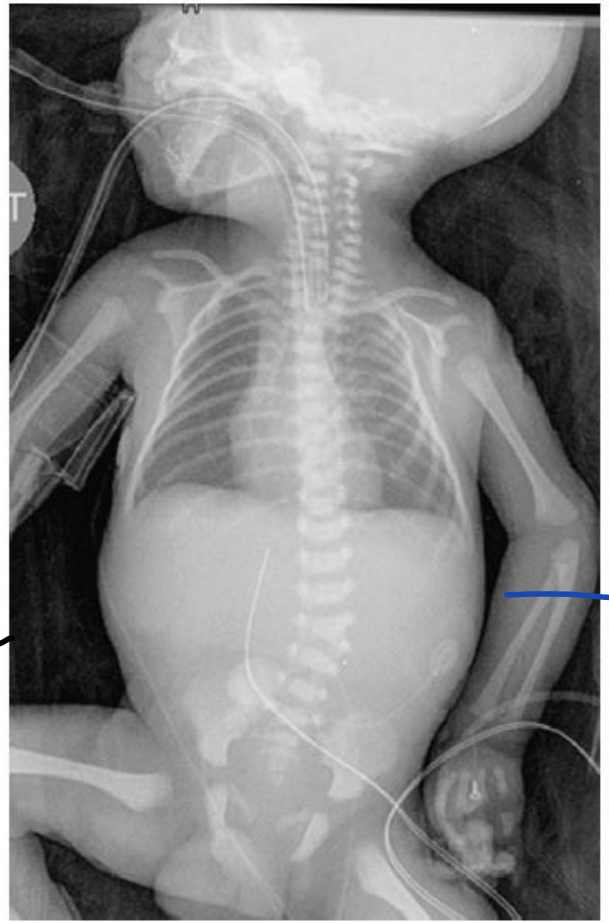
POSTNATAL DIAGNOSIS



gas in stomach
so there's a
fistula

complete esophageal atresia without any fistula

POSTNATAL DIAGNOSIS



* Another diagnostic sign: the patient is born without anus

→ think of VATREL

→ opacity in the abdominal cavity

MANAGEMENT

- Operative treatment of EA/TEF is not an emergency procedure.

There is usually time to confirm the diagnosis and to assess for associated anomalies

MANAGEMENT

- Preoperative measures:

  - ✓ Continuous suctioning tube in the upper esophagus → to avoid aspiration and lung infections (aspirating pneumonia)
  - ✓ Head-up position & on the side
  - ✓ If in respiratory distress → gentle low-pressure ventilation

MANAGEMENT

- Preoperative work up:

  - ✓ Echocardiography (to r/o cardiac &/or aortic arch anomalies)
  - ✓ Renal ultrasound
  - ✓ Spine radiographs

MANAGEMENT

- Operative repair depends on the gap between esophageal ends (on xray):

Gap	Surgical option
< 2 vertebrae	Primary anastomosis
2-6 vertebrae	Gastrostomy + delayed primary anastomosis
> 6 vertebrae	Gastrostomy + esophagostomy + esophageal replacement later on

without tension

↳ bowel

MANAGEMENT

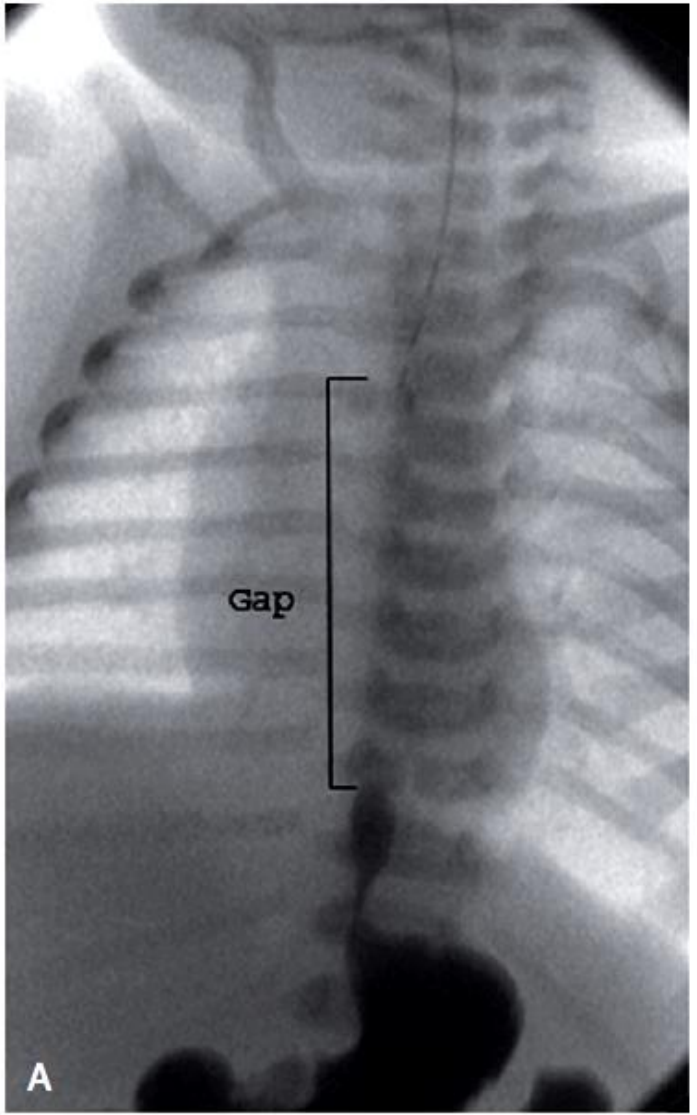
6 vertebrae ←

↓

most likely we're unable to do direct anastomosis

↓

replacement surgery



MANAGEMENT

Open (thoracotomy)

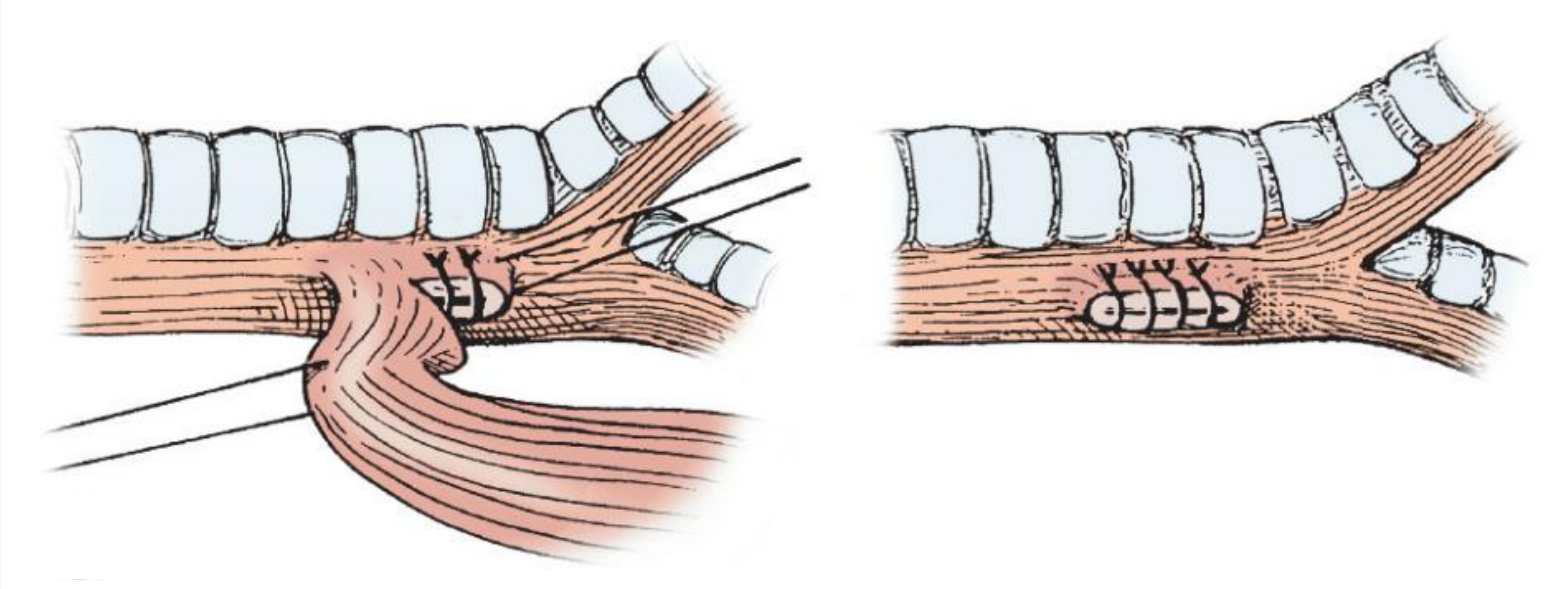
Vs

MIS (thoracoscopy)

(better but harder)

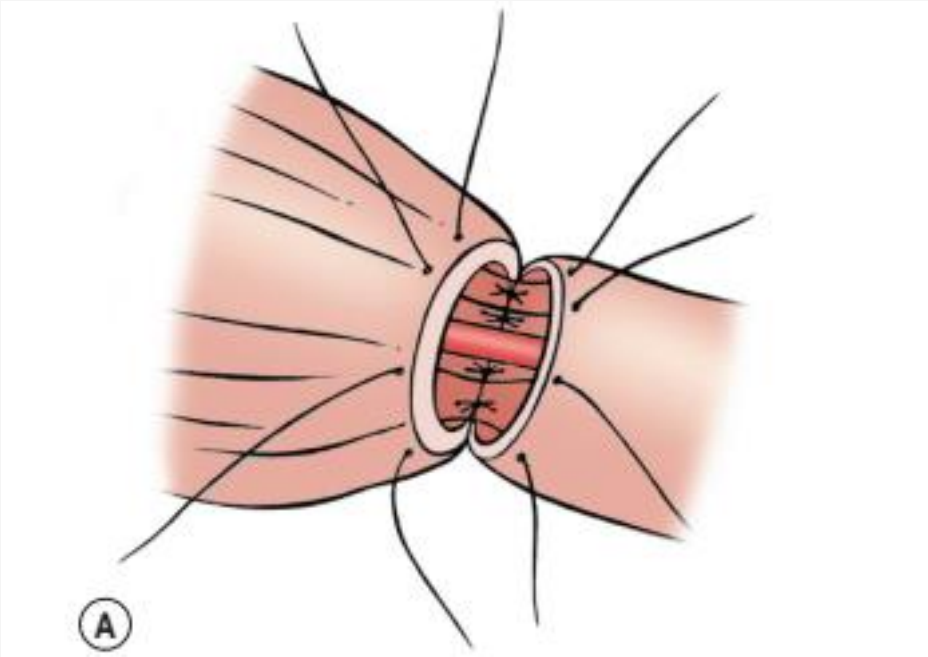
MANAGEMENT

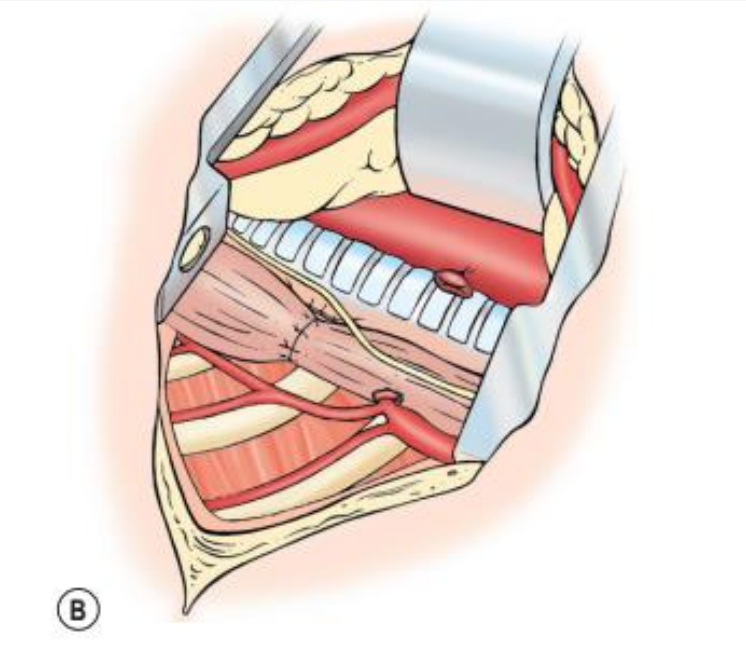
- Main principles



MANAGEMENT

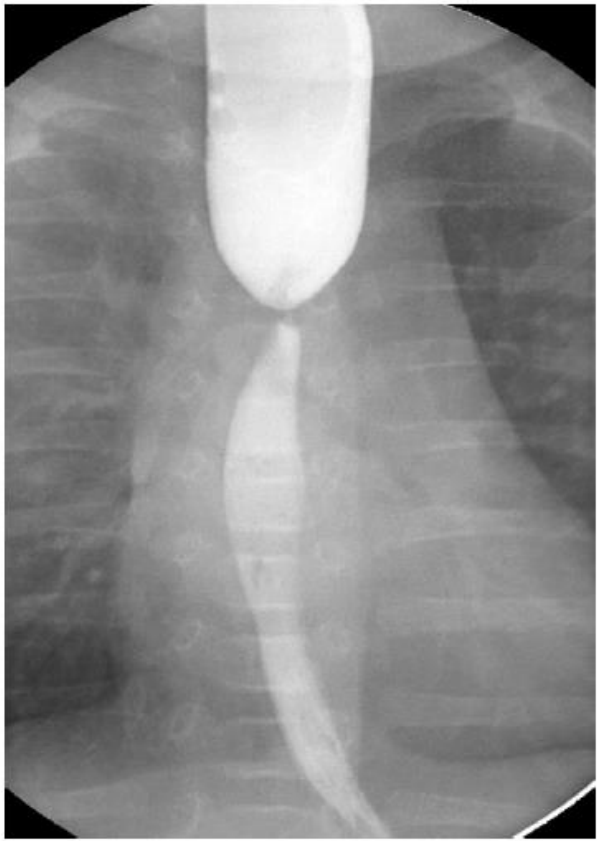
- Main principles

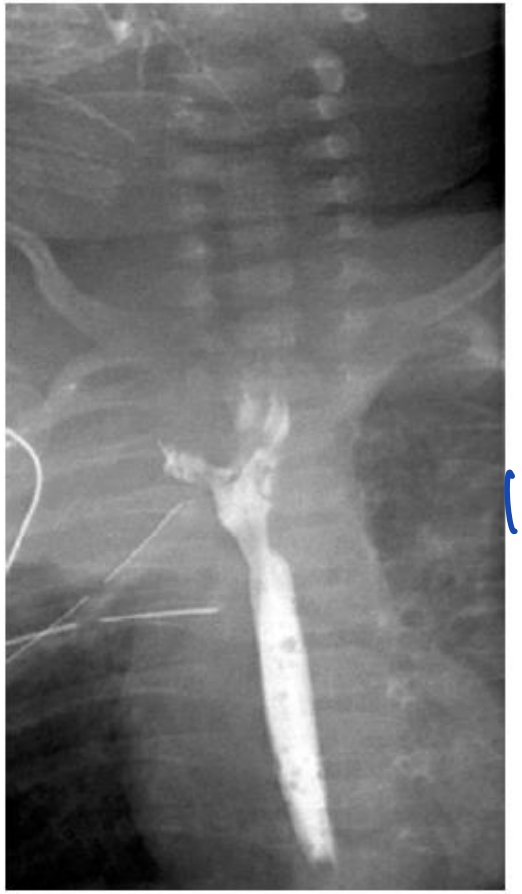




COMPLICATIONS

Stricture formation between upper and lower parts





leaking out

COMPLICATIONS

- Anastomotic Leaks (3.5-17%)
- **Anastomotic Stricture (17-60%)**
- Recurrent Tracheoesophageal Fistula (3-15%)
- Tracheomalacia
- Disordered Peristalsis → GERD → ?! Esophageal Cancer
- Vocal Cord Dysfunction
- Respiratory Morbidity
- Thoracotomy-Related Morbidity