Congenital Diaphragmatic Hernia

information for parents

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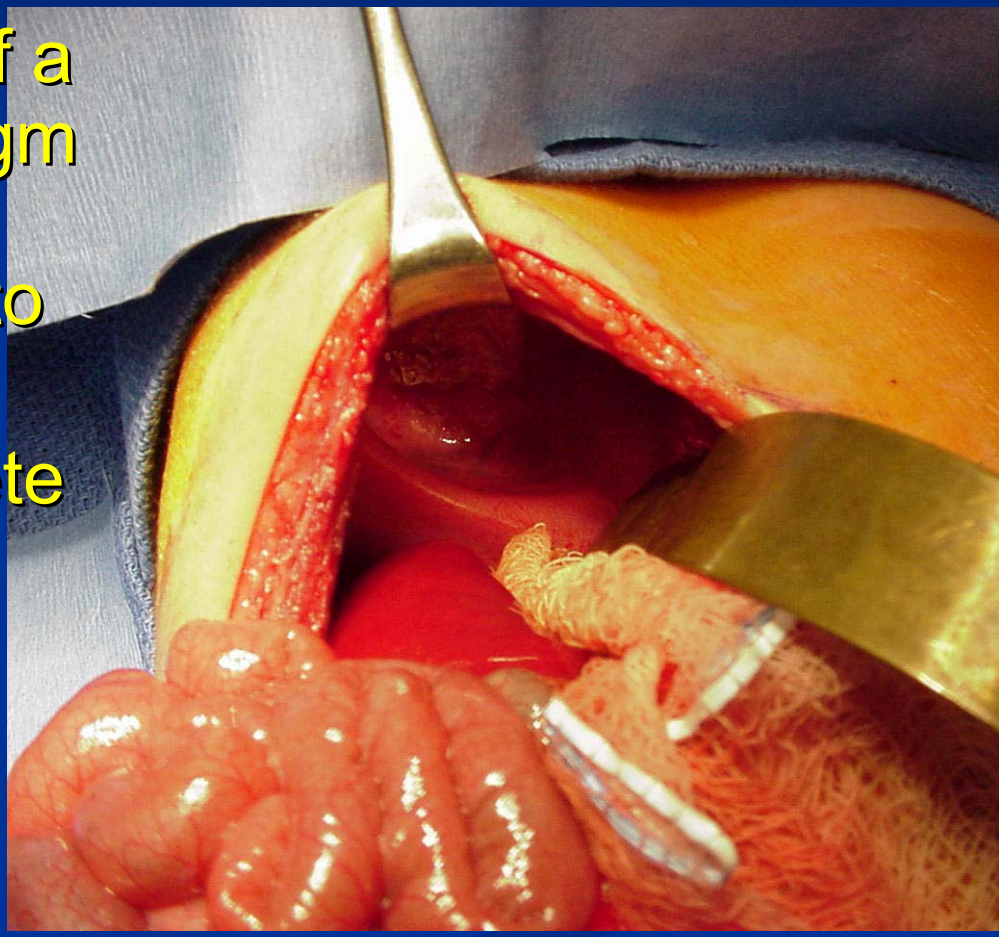
Pediatric Surgeons of Phoenix

CDH

- Congenital absence of a portion of the diaphragm allowing abdominal contents to migrate into the chest
- May be due to complete absence of the diaphragm
- Compare with Eventrated (thin) Diaphragm



How Common is CDH?

- 1 in 4000 births*
  - 1/3 of infants are stillborn
- Left = 80%
- Right = 20%
  - Bilateral is very rare
- 2% risk to first-degree relatives

*all births (alive and stillbirths) California population-based study
March of Dimes

Causes

- Unknown
- ?Genetic
- Drug/insecticides
  - Nitrofen
  - Phenmetrazine
  - Thalidomide
  - Quinine
- Vitamen A deficiency

Associated Anomalies

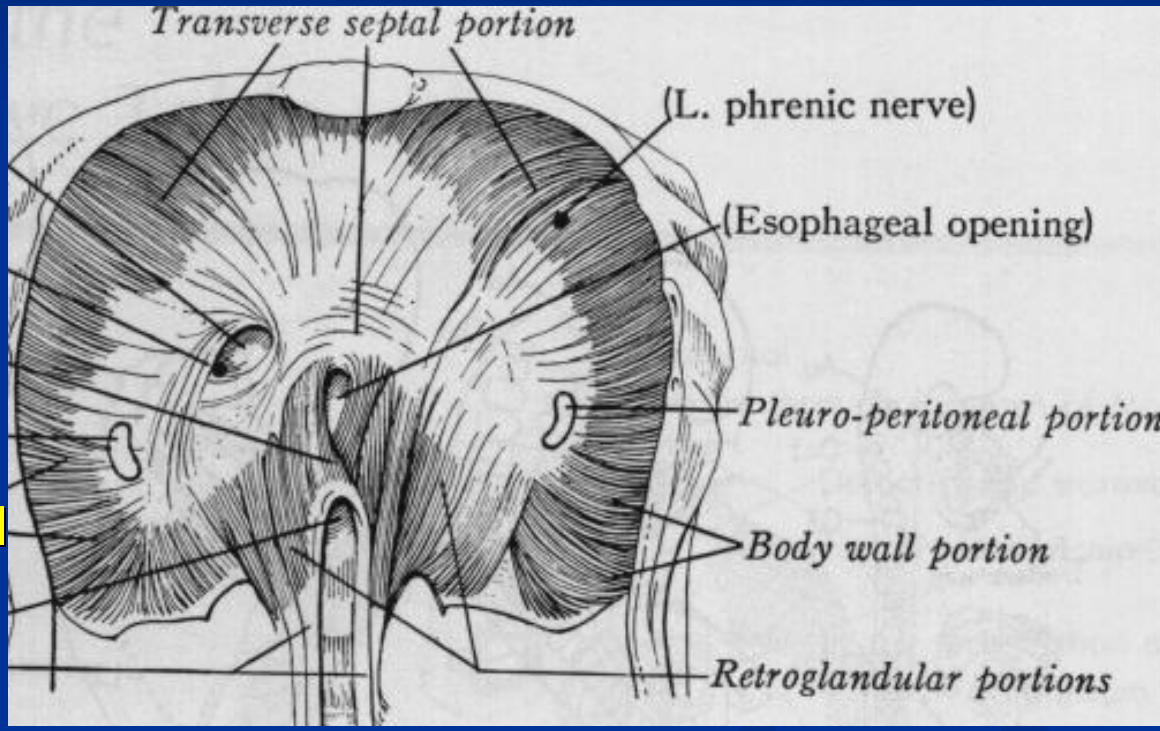
- 28% incidence
- Very common in stillborn infants
- Most common
  - Neural tube defects
  - Cardiac Defects
  - Esophageal atresia
  - Omphalocele
  - Trisomy 13, 18, 21

Embryology

4th week

4 components

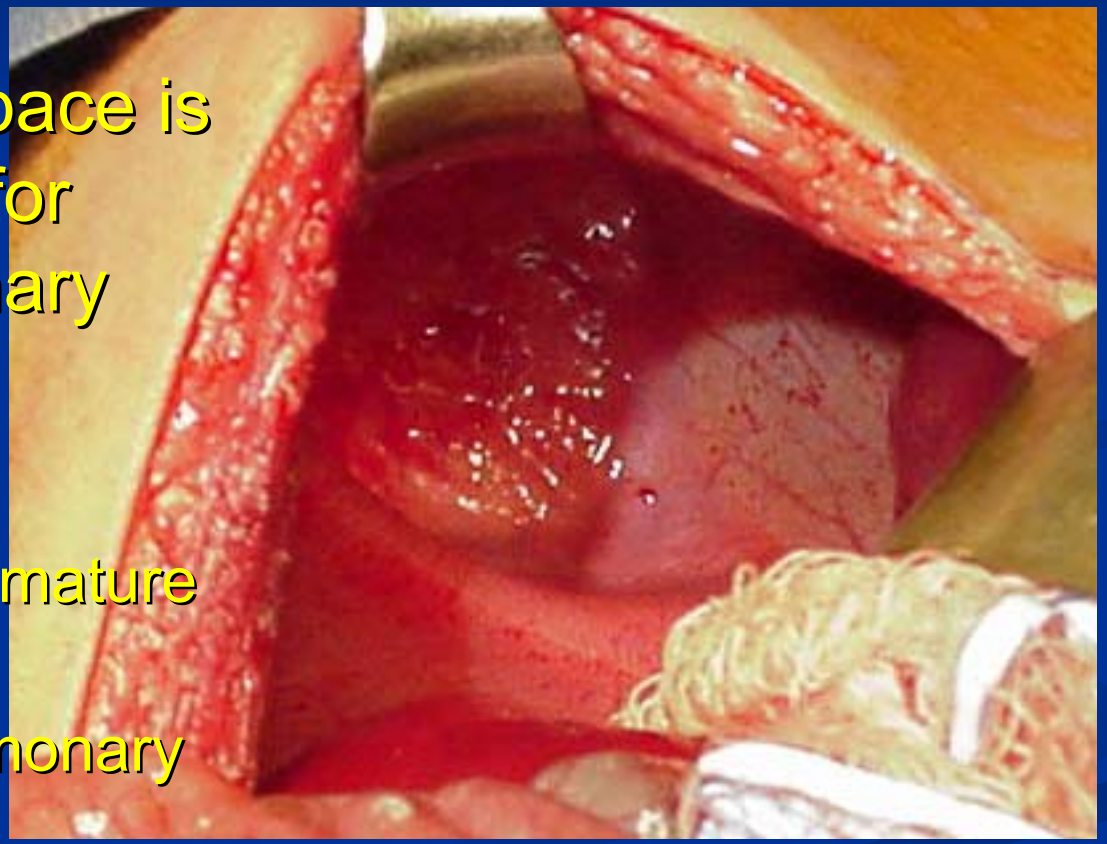
1. Septum transversum – Central Tendon
2. Pleuroperitoneal membranes – dorsolateral
3. Esophageal mesentery - doral crura
4. Intercostals – muscular portion



Complete closure by 8th Week

Pulmonary Vascular Development

- Adequate intrathoracic space is a prerequisite for normal pulmonary growth
  - Structurally immature lungs
  - Abnormal pulmonary vasculature



Hernia Contents

- Left CDH
  - Left lobe of liver
  - Spleen
  - Stomach
- Right
  - Liver
  - Everything else
  - Abnormal hepatic veins to right atrium
- Both
  - Large and Small bowel

Why are the infants sick at birth?

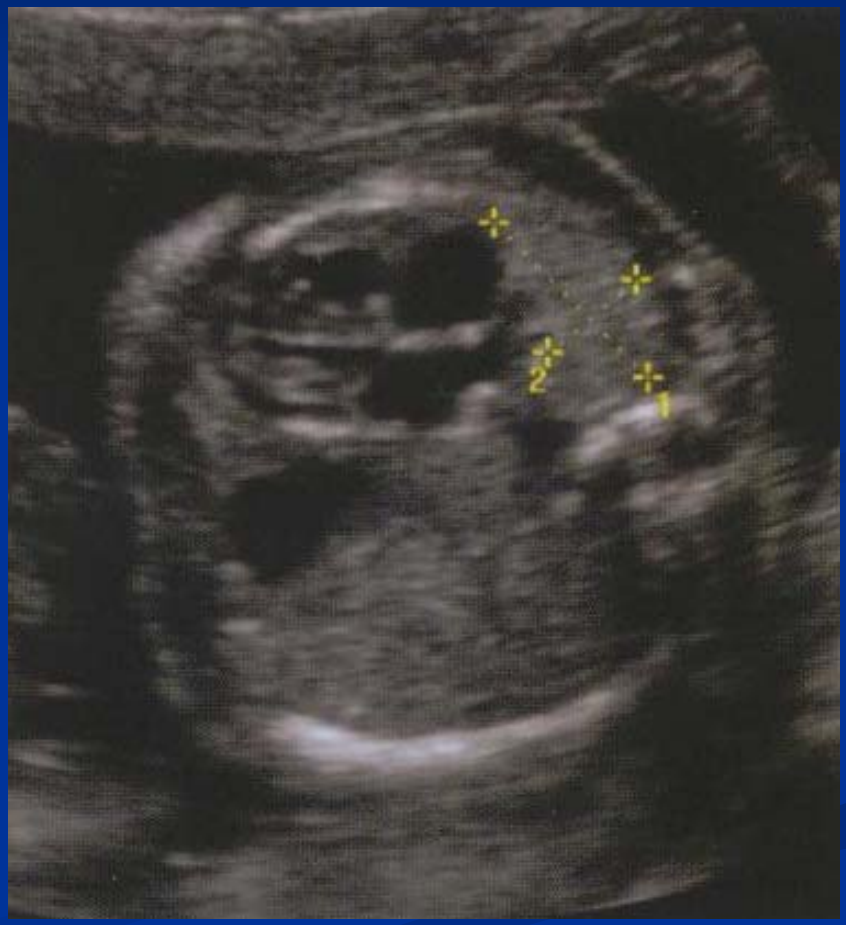
- Pulmonary Hypoplasia
  - Small lungs
  - Caused by compression
  - Prevented growth and maturation
- Pulmonary Hypertension
  - Abnormal Lung Vasculature
  - The high pressure in the lungs limits blood flow to the lungs

Both Sides Effected

- Hernia is on one side
  - Both sides are underdeveloped
  - Fewer than normal alveoli (sacs for oxygen transfer)
- Abnormal pulmonary blood vessels on both sides
  - Pulmonary hypertension

Diagnosis

- Prenatal ultrasound
  - Polyhydramios (80%)
  - Stomach at level of heart
  - Absence of stomach in abdomen

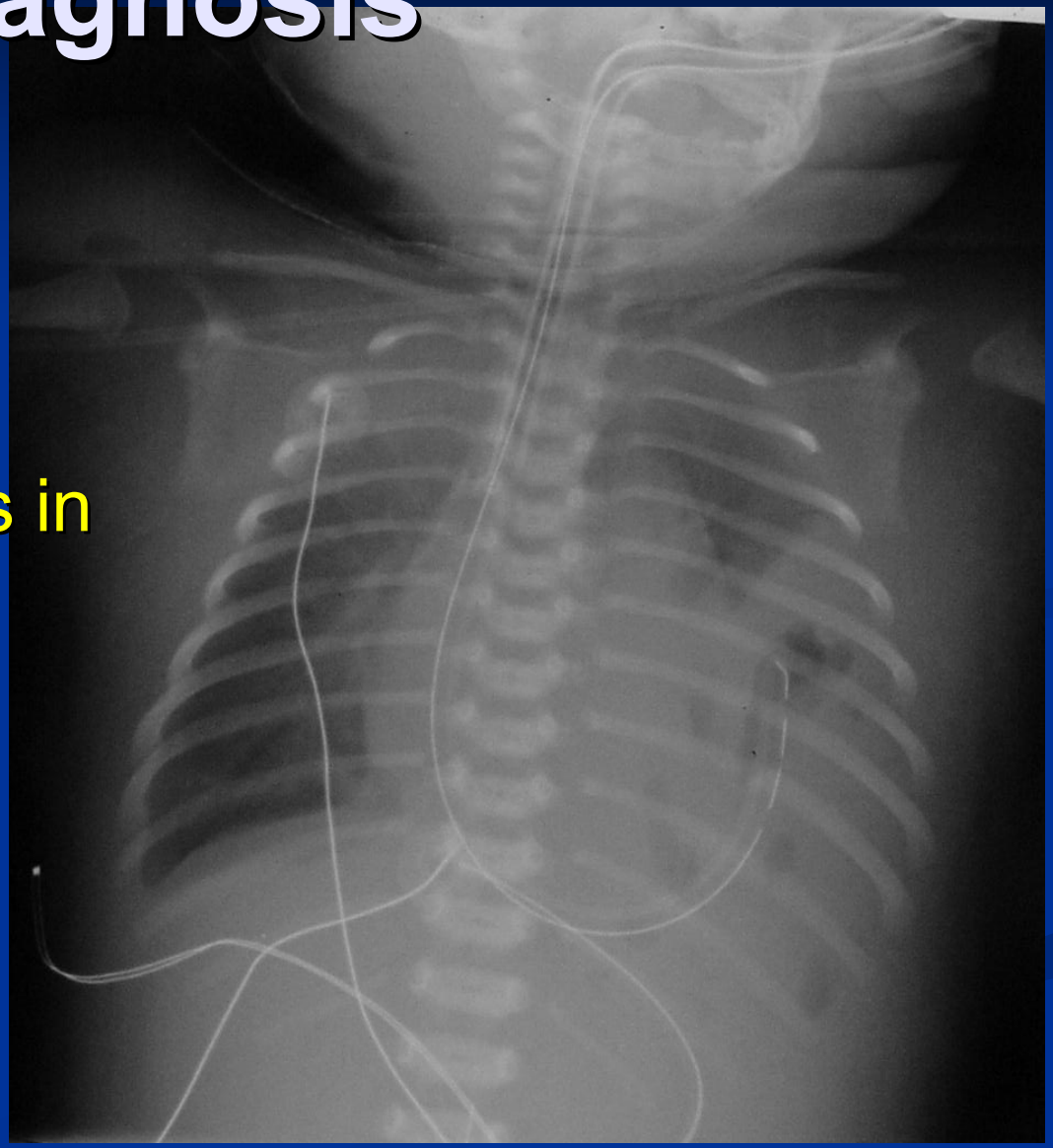


Diagnosis

- Narrow abdomen
- Large chest
- Respiratory distress

Diagnosis

- CXR
- Ultrasound only if diagnosis in doubt
- GI Contrast studies in difficult cases



Possible Misdiagnosis

- Eventration of Diaphragm
  - Elevated hemidiaphragm
- Congenital Pulmonary Adenomatous Malformations (CPAMs, or CCAMs)
- Lung agenesis

Prenatal Prognosis

- Associated anomalies
  - Cardiac (excl. single ventricle)
    - 47% survival
  - Single ventricle
    - 5% survival
- Ultrasound Studies
  - Lung to Head ratio (LHR)*
    - <0.8 fatal
    - <0.8 but >1.0 poor prognosis
    - >1.0 good prognosis

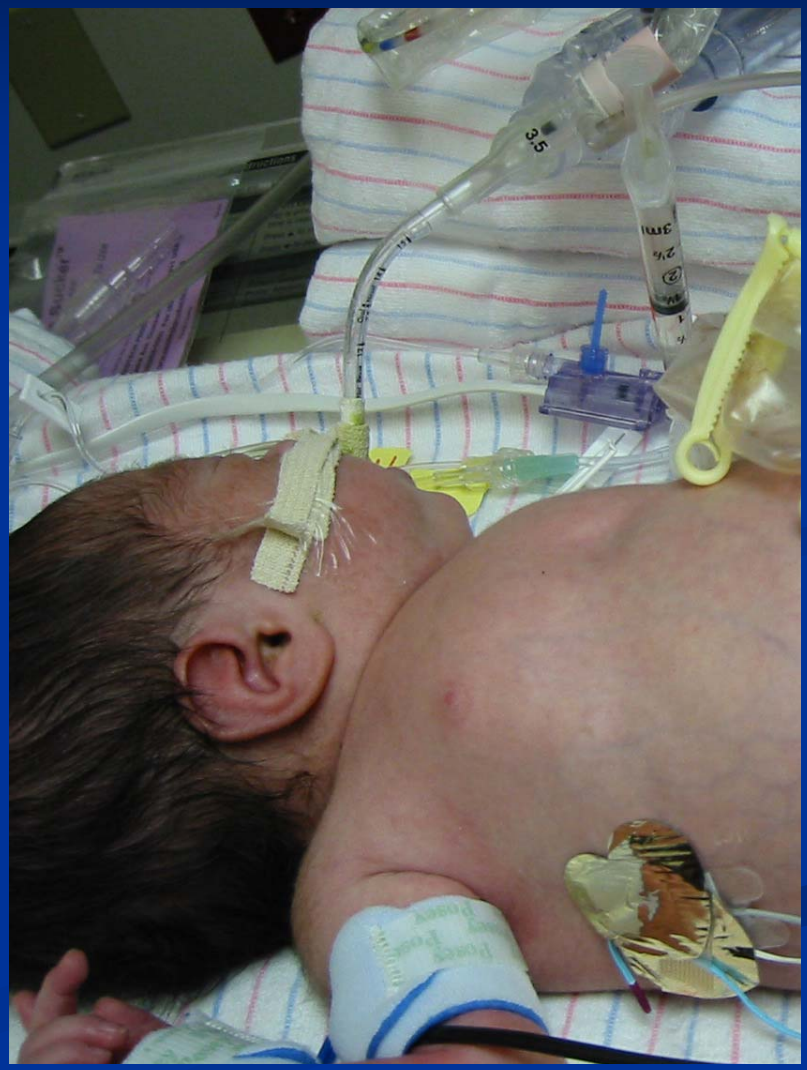
*LHR has not been prospectively validated

Treatment

- Gentle lung ventilation
- “Physiologic Emergency” not a Surgical Emergency
- Survival depends on
  - Uncorrectable pulmonary hypoplasia
  - Reversible pulmonary hypertension

Gentle Lung Ventilation

- Place ETT early
- Avoid high pressures
- HFV
  - Jet Ventilator
  - Oscillating Vent



Treatment

- Surfactant
  - No Difference in many studies
- iNO
  - Mixed results

Timing of Repair

- High mortality for emergent surgery
- Best survival is delayed repair
- Optimal timing
  - Unproven
- **Serial Echocardiography**
  - Evidence of resolution of pulmonary hypertension

Extracorporeal Life Support ECLS

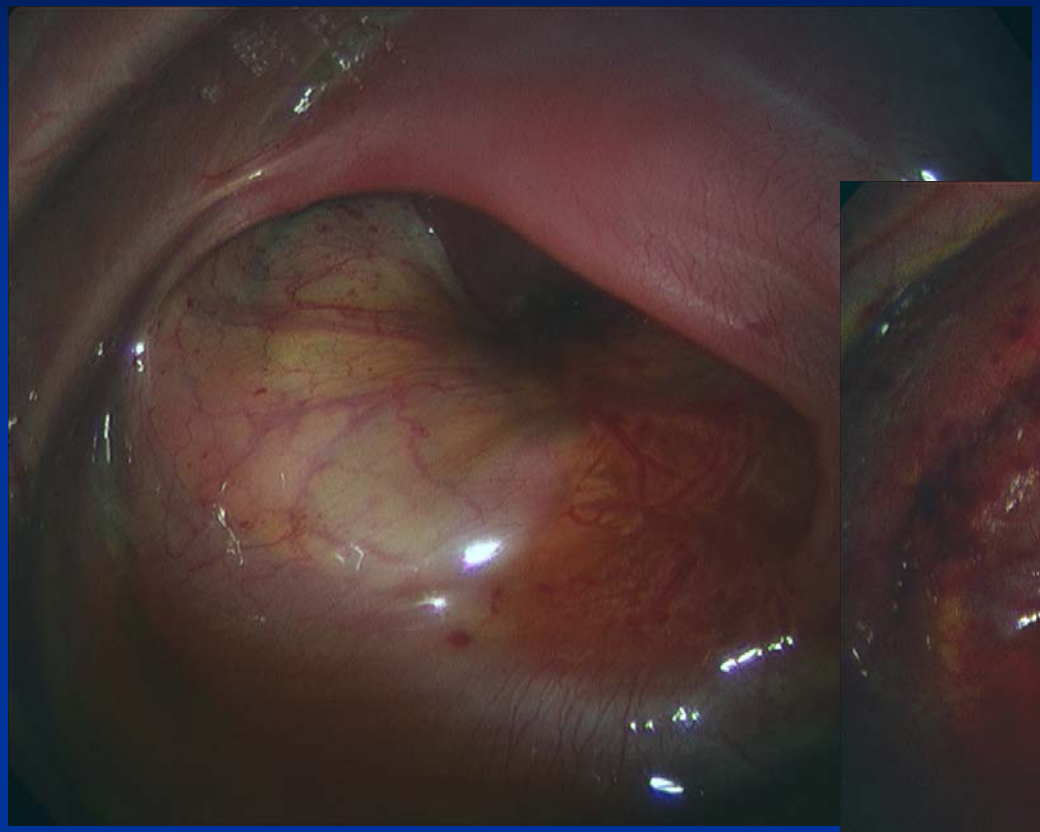
- Introduced in 1977
- Formerly called “ECMO”
- Criteria for use
  - Inadequate delivery of Oxygen
  - Predicted 80% mortality without ECLS

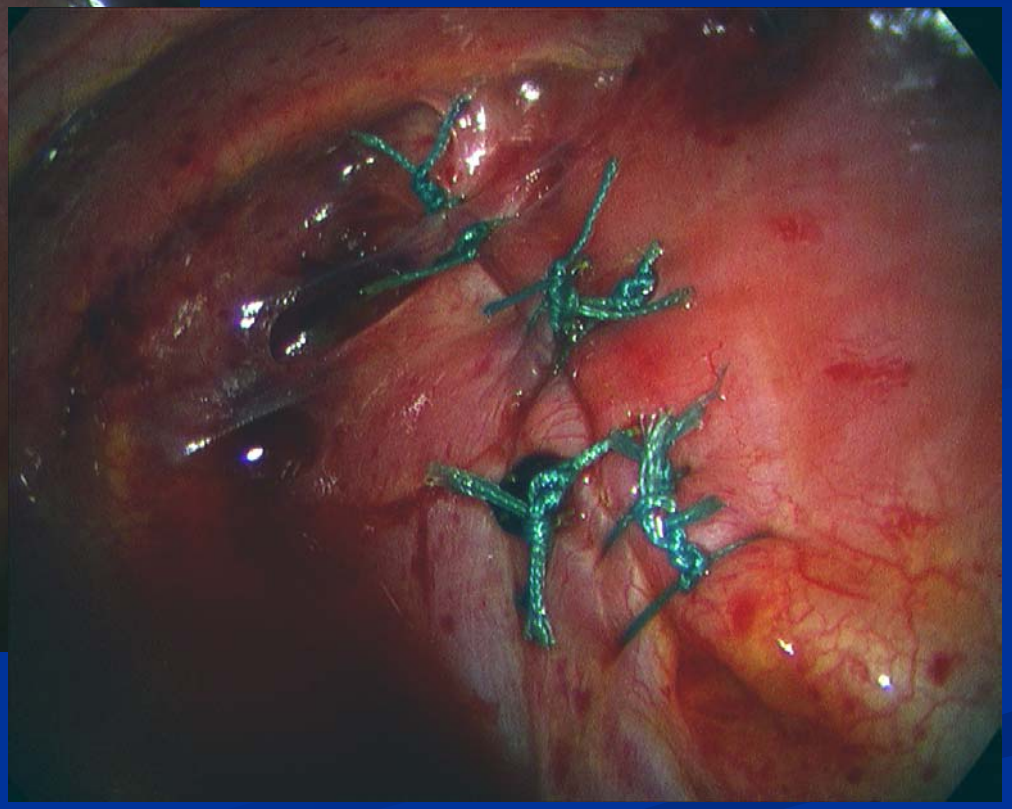
Oxygenation Index (OI) is no longer a sole criteria

Options for Repair

- Thoracosopic Repair
  - Minimally invasive repair
  - Best suited for stable, near-term patients
  - Surgery is done through small incisions in the chest
- Open repair through abdomen
  - Can be done on less stable patients
  - Well-suited for complete absence of the diaphragm

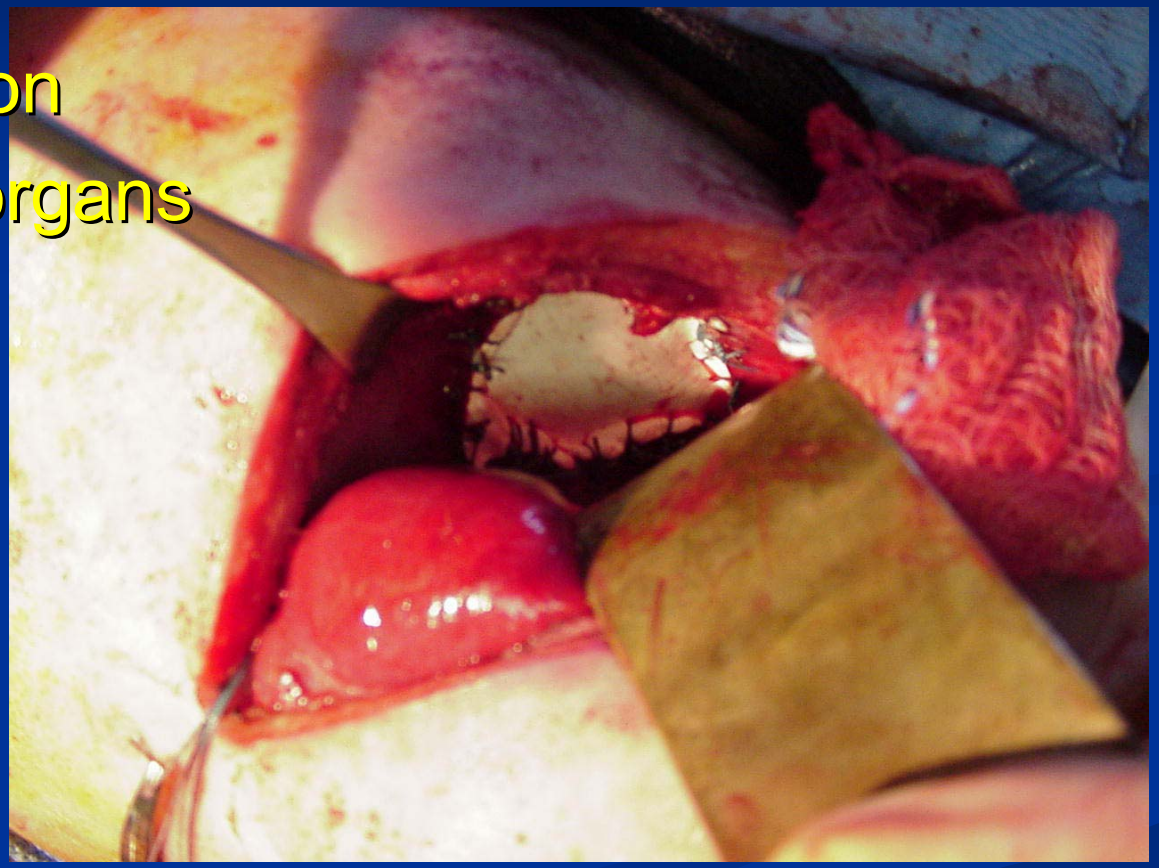
Thoracoscopic Repair



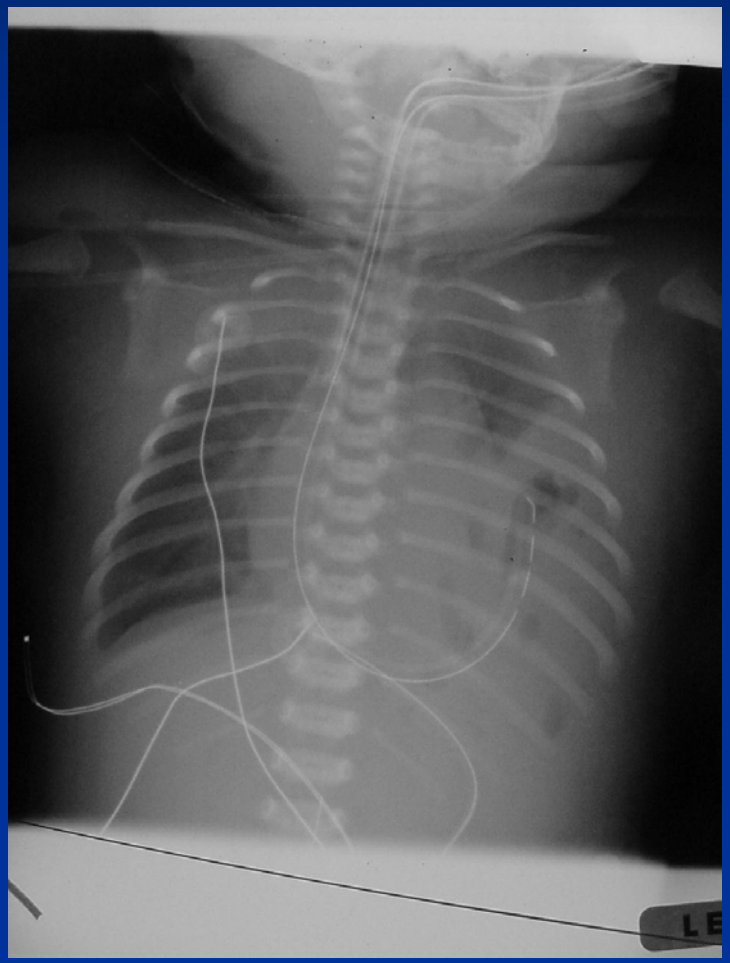


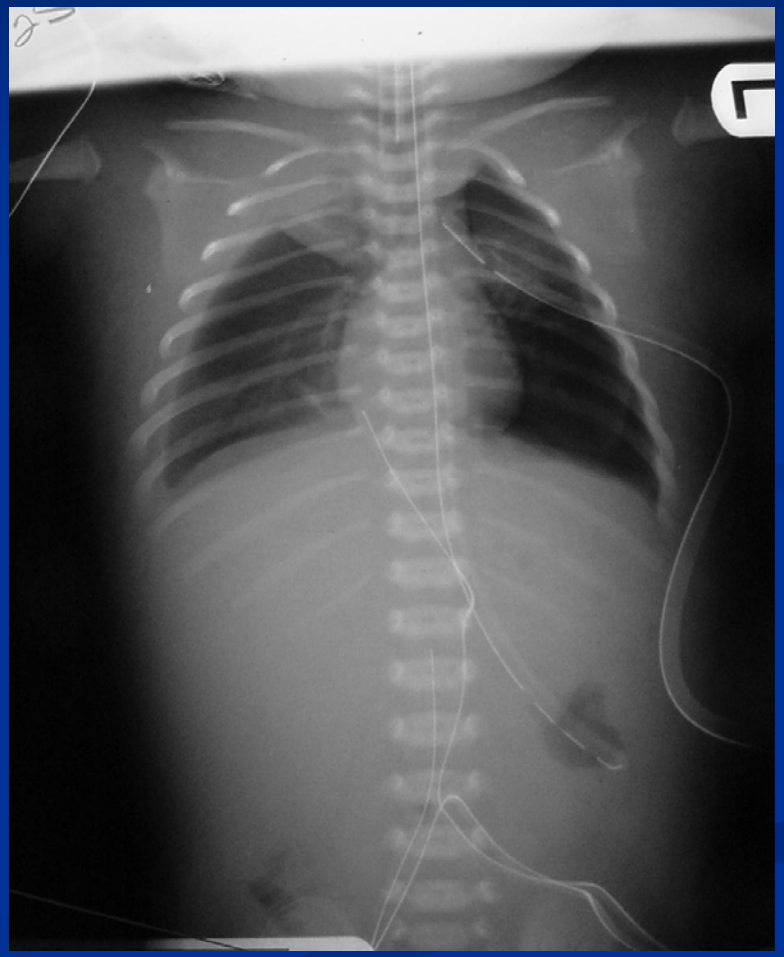
Open Surgery

- Subcostal Incision
- pull abdominal organs back into the abdomen
- Close hole
- Patch Repair of defect
  - Gortex
  - Surgisis



Repair





Results

- Congenital Diaphragmatic Hernia Study Group
  - 67% Survival
  - 71% if no CHD
  - Decreasing survival with ECLS due to better results with ventilators, and only sicker patients going on heart lung bypass

Long Term Results

- Children over 8 yrs of age
  - Total Lung Capacity
    - 99% of expected volume
  - Forced Expired Volume in 1 second
    - 89% of expected volume
  - Normal diffusion coefficient

- ECLS
  - Overall, developmental delay is seen in 45% of patients requiring ECSL. May be due to poor oxygenation prior to ECSL or ECSL itself.

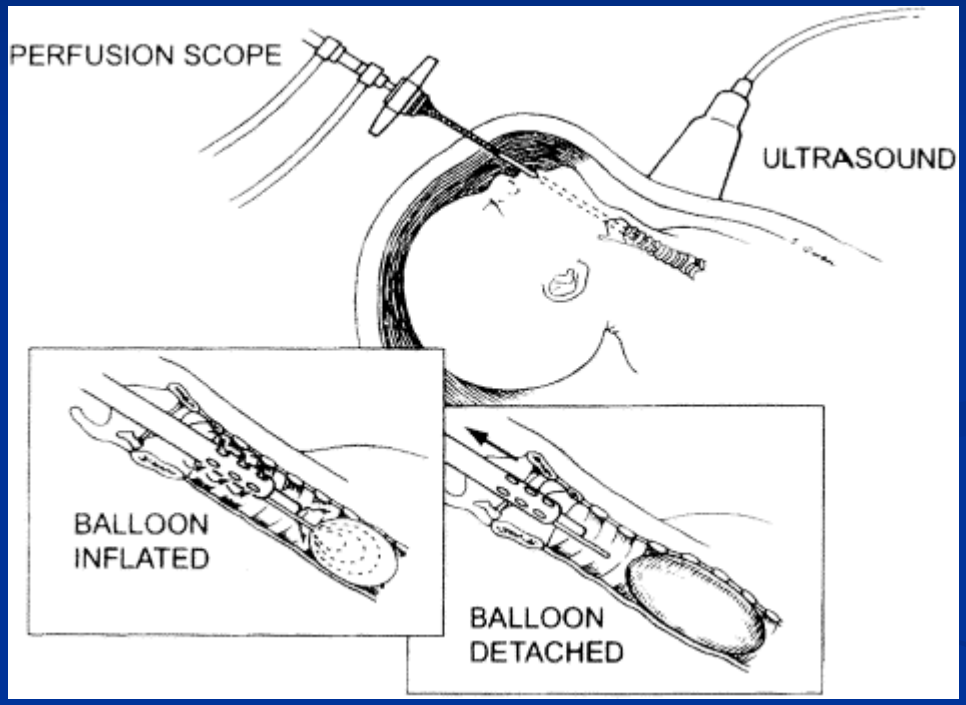
Fetal Surgery

- Fetoscopic tracheal occlusion (FETO)
- Developed at UC San Francisco

- Criteria
  - LHR <0.8 or <1.0 with liver up
  - 28 weeks EGA
  - PROM, prematurity common
  - 50% survival

- *In Utero* repair has been abandoned

Tracheal Occlusion UCSF



J Pediatr Surg. 2003 Jul;38(7):1012-20. Fetoscopic temporary tracheal occlusion for congenital diaphragmatic hernia: prelude to a randomized, controlled trial. Harrison MR, Sydorak RM, Farrell JA, Kitterman JA, Filly RA, Albanese CT.
Department of Surgery, The Fetal Treatment Center, University of California, San Francisco, CA 94143-0570, USA.

Congenital Diaphragmatic Hernia Links

- CHERUBS
  - http://cdhsupport.blogspot.com/2009/06/national-congenital-diaphragmatic.html
- Congenital Diaphragmatic Study Group
  - http://cdhsg.net/

Thanks