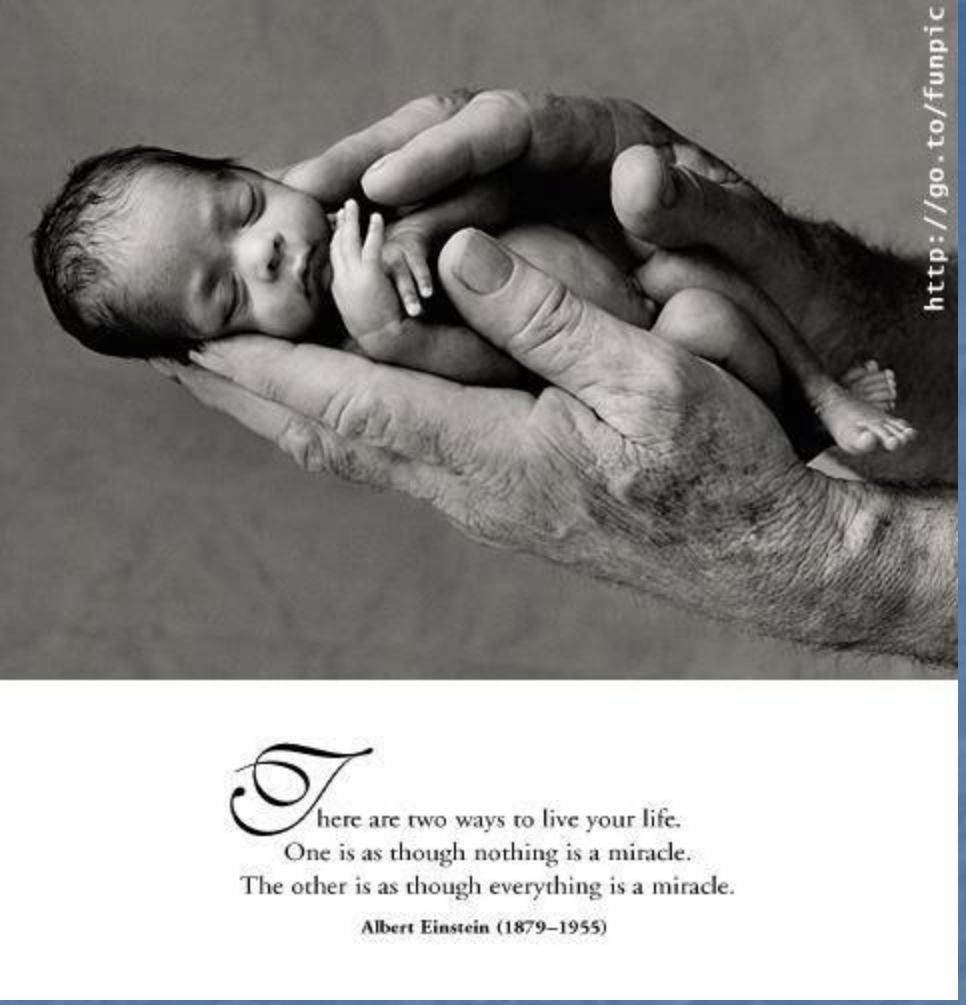
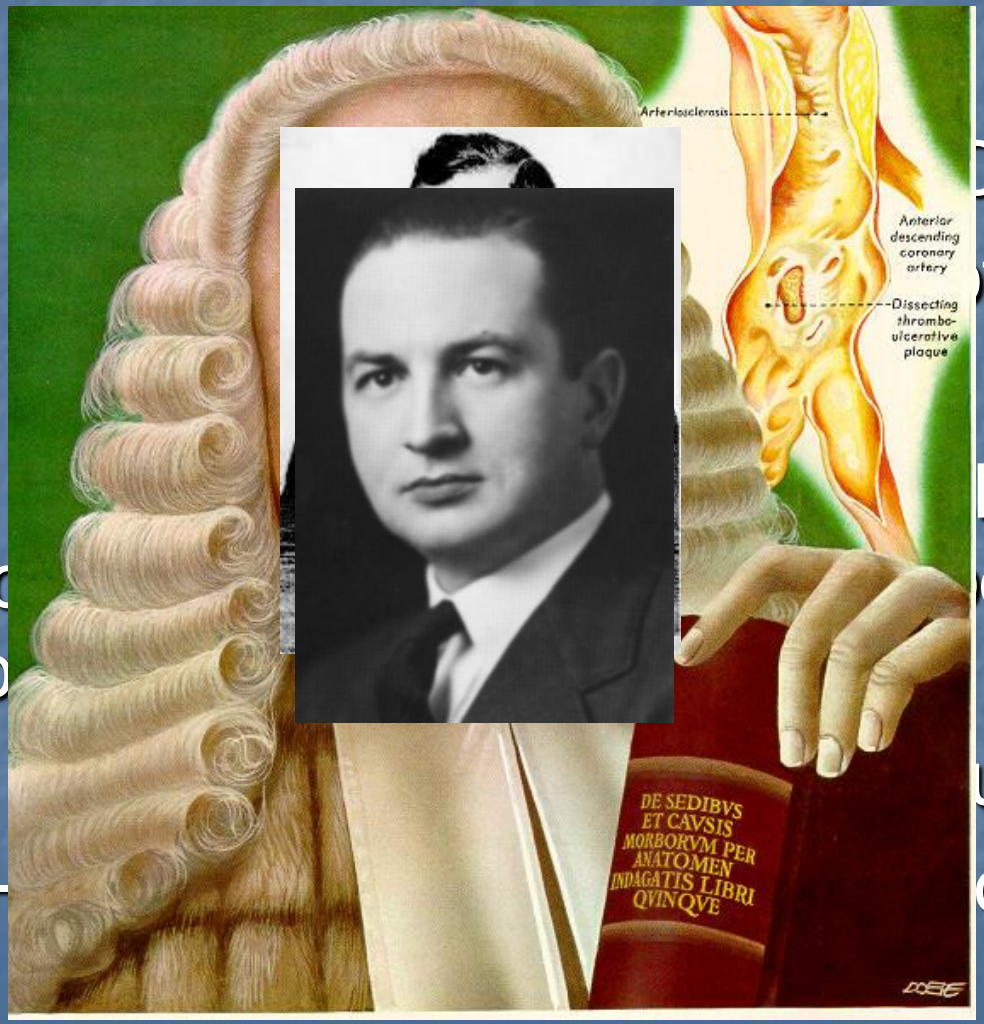


Pediatric Surgery Department

Congenital diaphragmatic hernia

History



- 1679
- 1761
- 1905
- 1925
- 1946
- 1980-

CDH

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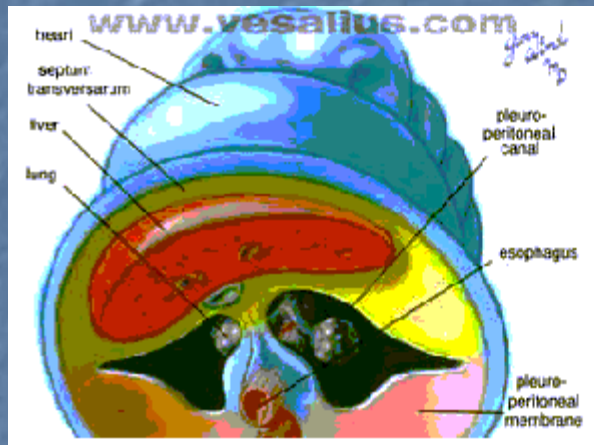
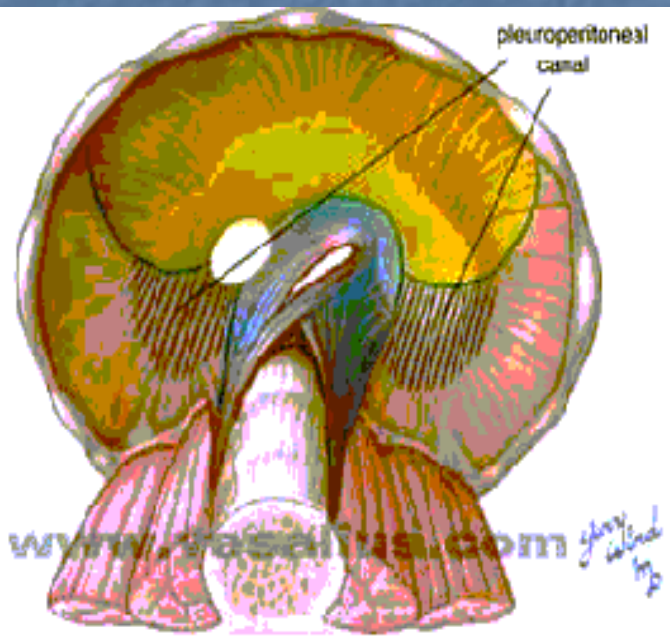
Incidence

- 1 : 2000-5000 live birth
- 8 % of all major congenital anomalies
- mortality rate nearing 70 percent
- CDH accounts > 1% of total infant mortality in USA

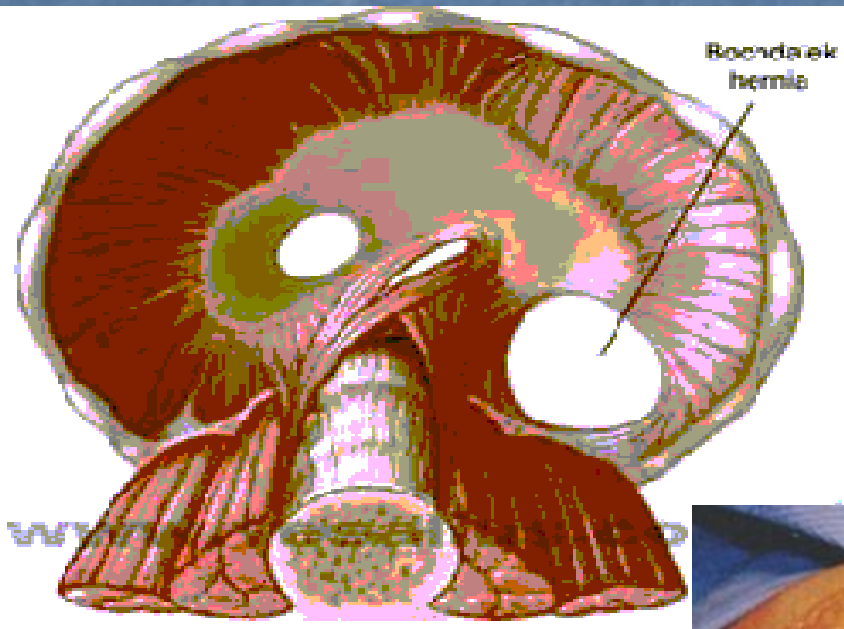
- Cost per new case CDH = 250 000 $

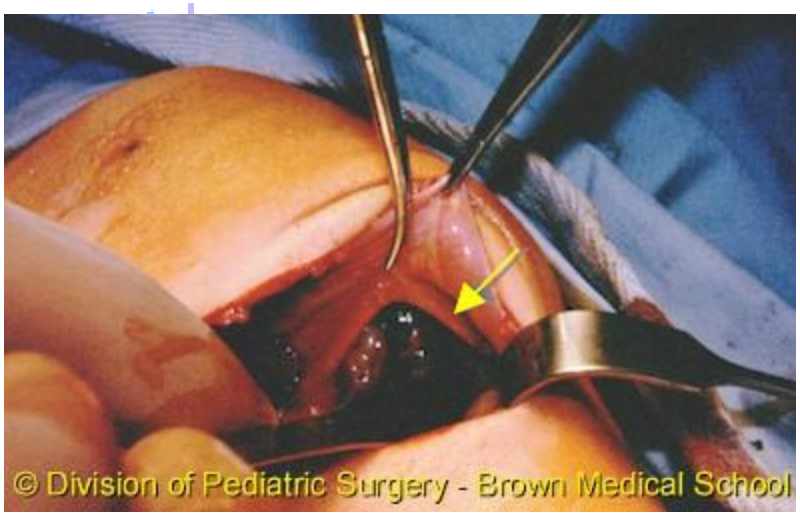
Diaphragm Development



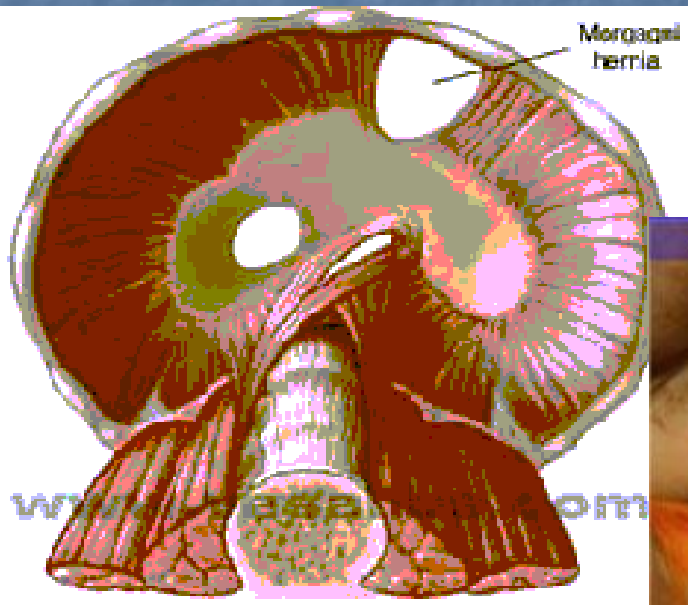


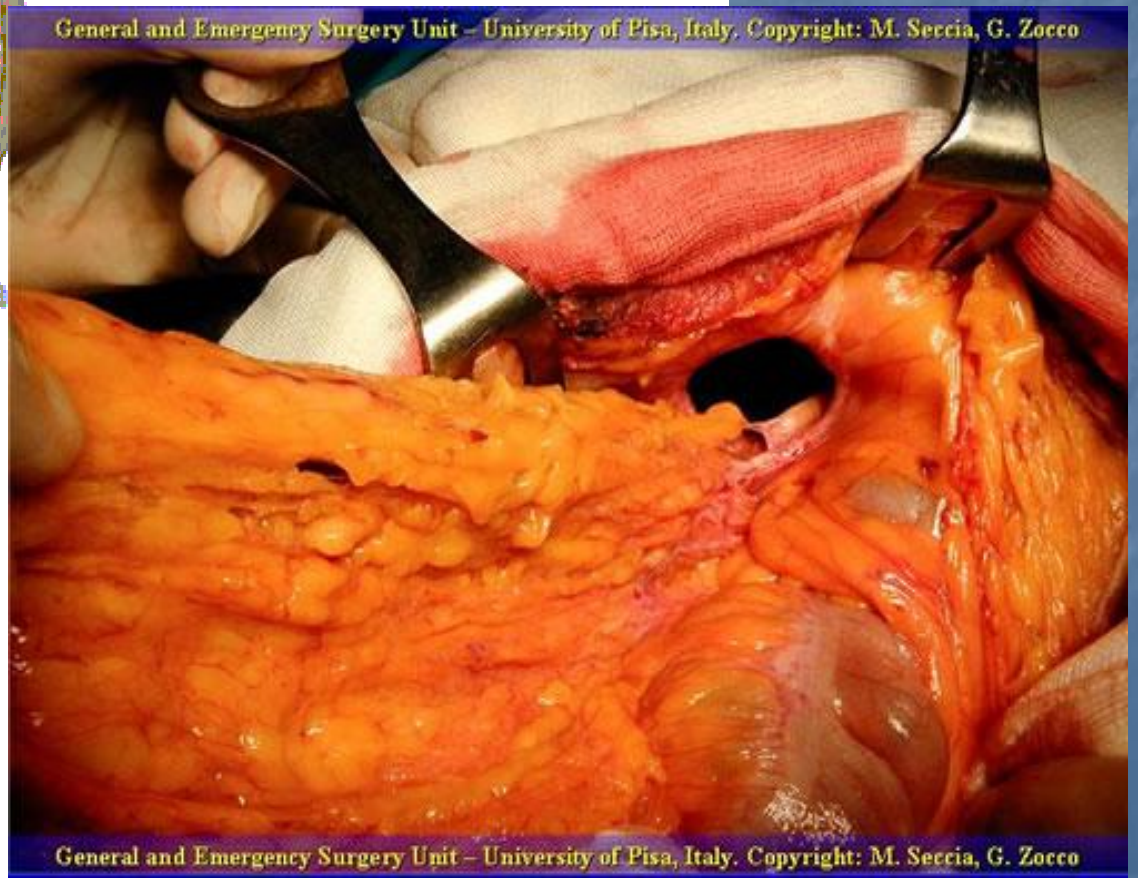
Diaphragm Development





Diaphragm Development





Causes

- The cause of CDH is largely unknown
- CDH can occur as part of a multiple malformation syndrome in up to 40% of infants (cardiovascular, genitourinary, and gastrointestinal malformations)
- Karyotype abnormalities have been reported in 4% of infants with CDH, and CDH may be found in a variety of chromosomal anomalies including trisomy 13, trisomy 18, and tetrasomy 12p mosaicism

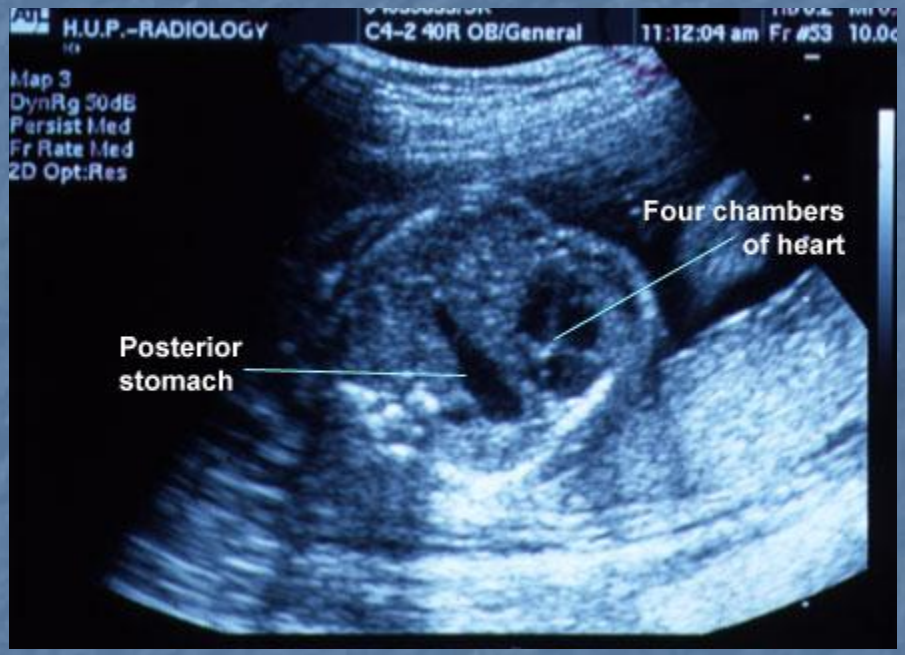
Prenatal Diagnosis

- ultrasonography diagnosis (as early as the second trimester)

Mediastinal shunt
Viscera herniation (stomach, intestines, liver*, kidneys, spleen and gall bladder)
Abnormal position of certain viscera inside the abdomen
Stomach visualization out of its usual position
Intrauterine growth retardation*
Polyhydramnios*
Fetal hydrops*

*** bad prognosis**

Fetal diafragmatic hernia: Ultrasound diagnosis



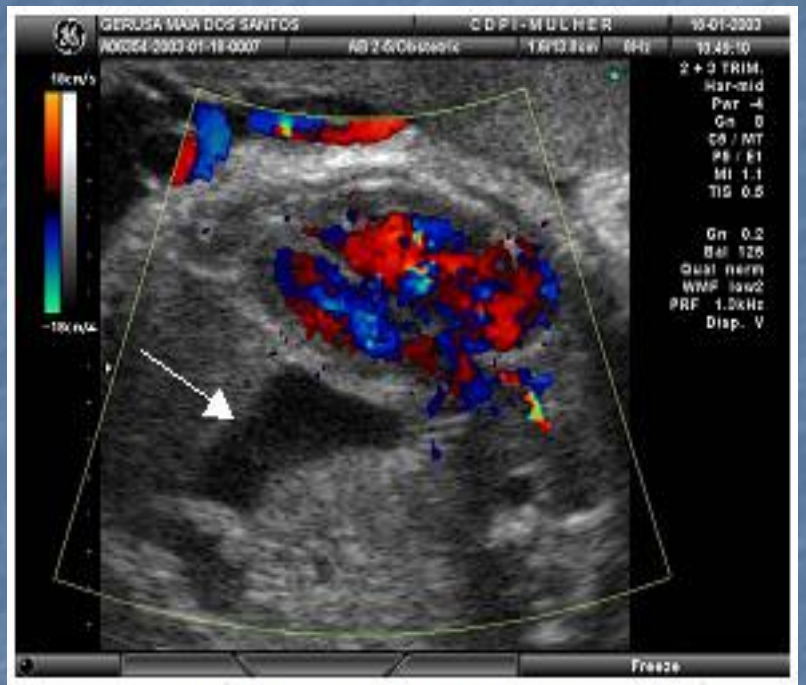


Figure 1: Ultrasound axial view showing the typical image of the left diaphragmatic hernia. Note the heart placed to the right side and the stomach in the same plane (arrow).

Prenatal MR Imaging - single-shot turbo spin-echo (HASTE)- of congenital diaphragmatic hernia

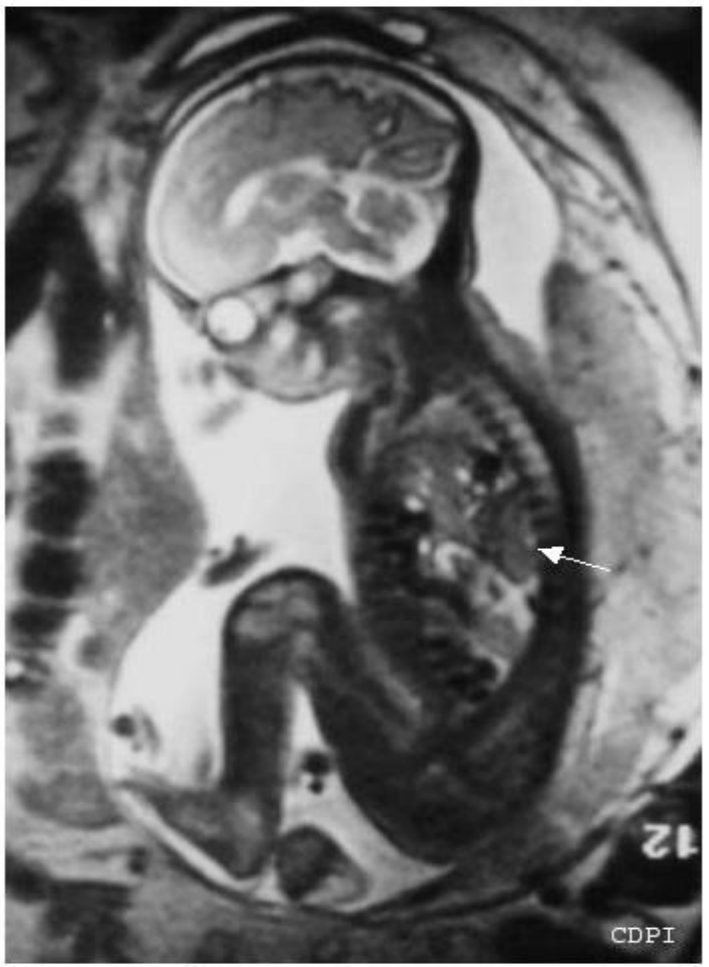


Figure 3: Sagittal T2-weighted MR image show the left diaphragmatic hernia. Note the herniation of the left lobe of the liver (arrow).

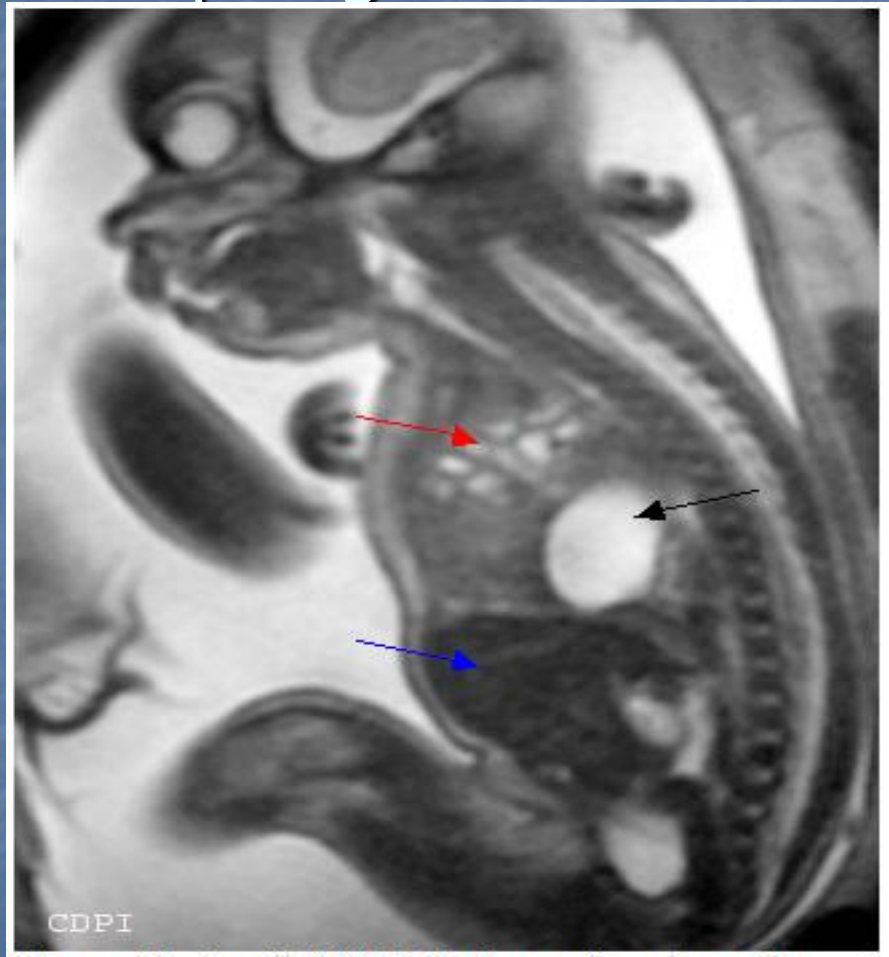


Figure 30: Sagittal HASTE shows the stomach (black arrow), bowel loops (red arrow) and the liver-down (blue arrow).

Prenatal MR Imaging of congenital diaphragmatic hernia

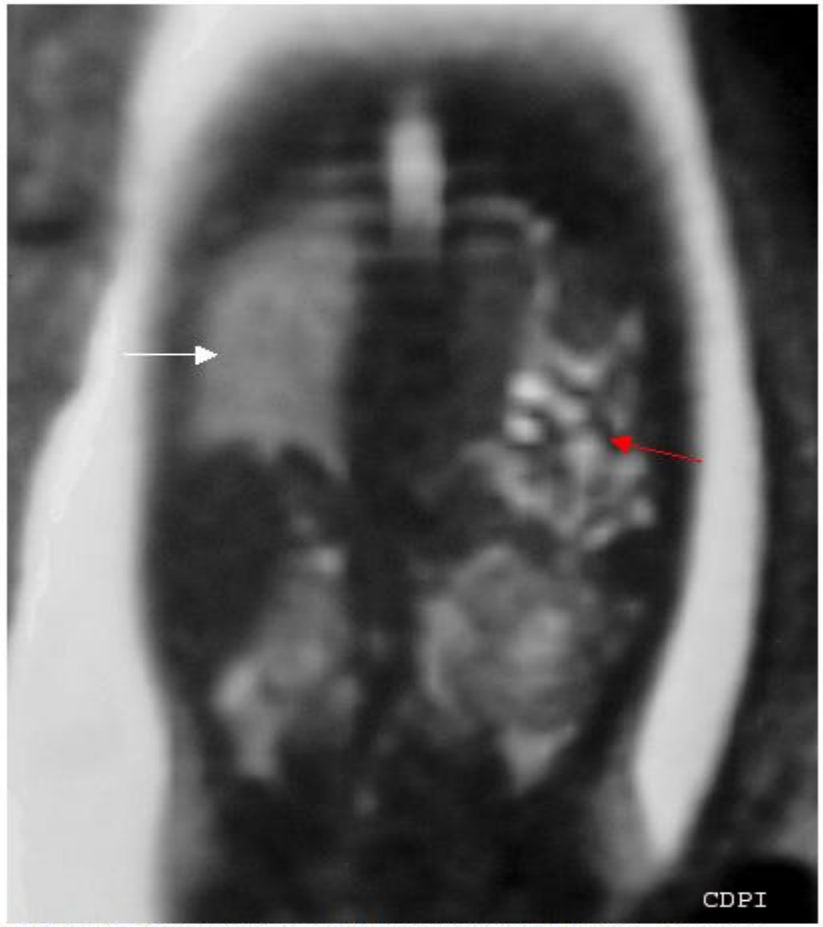


Figure 13: Coronal view on T2-weighted MR image, show the normal right lung (white arrow) and hypoplasic left. Note the intestinal loops on the left side (red arrow).

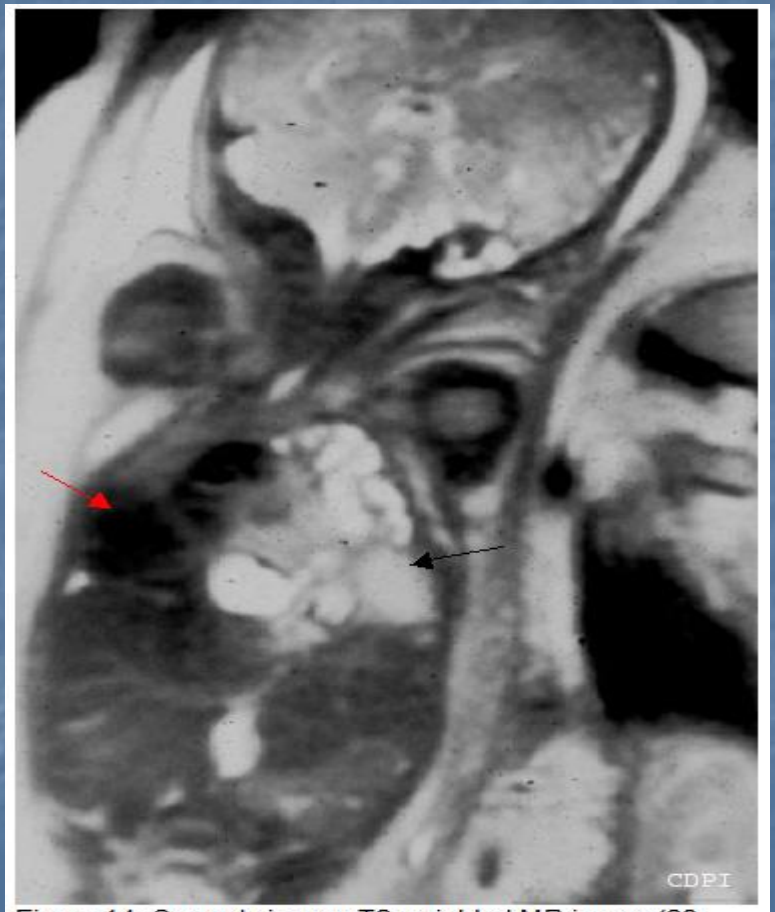
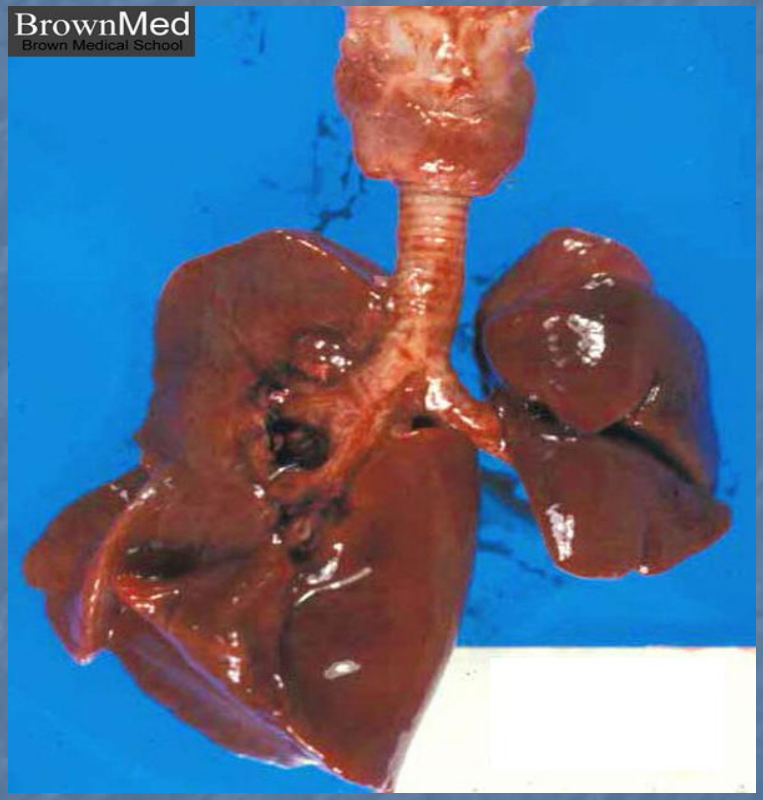
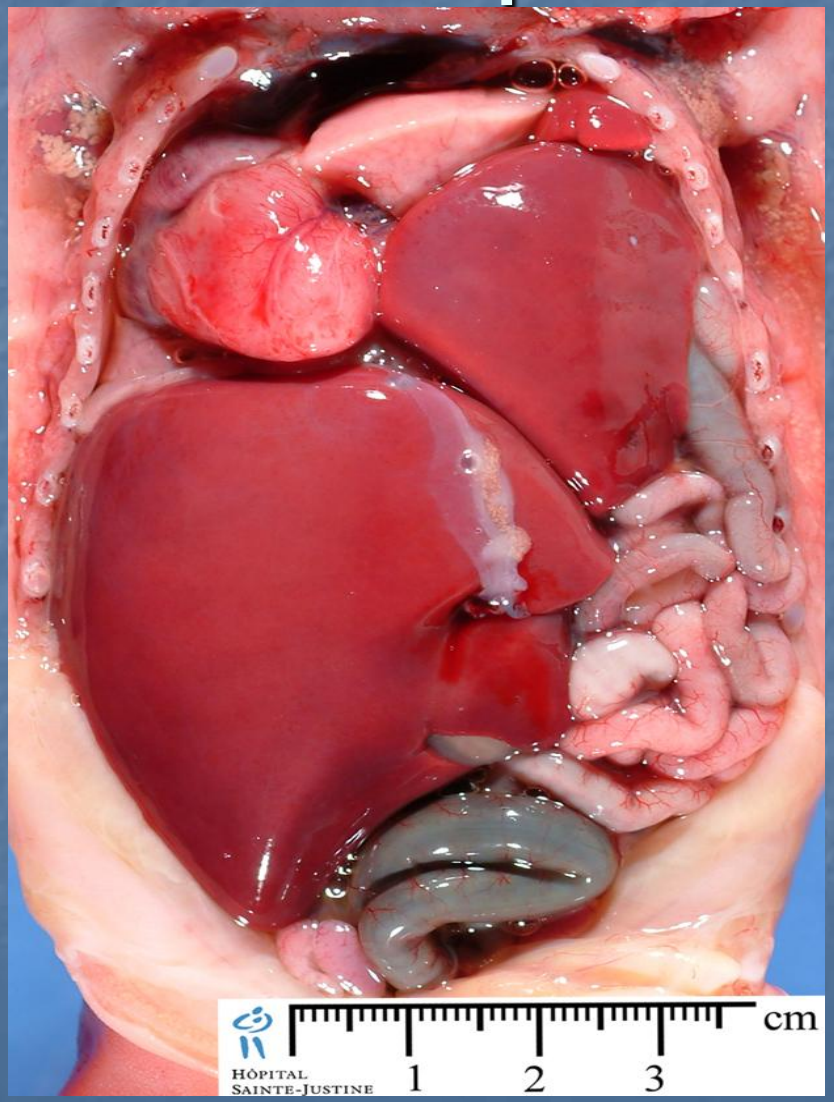


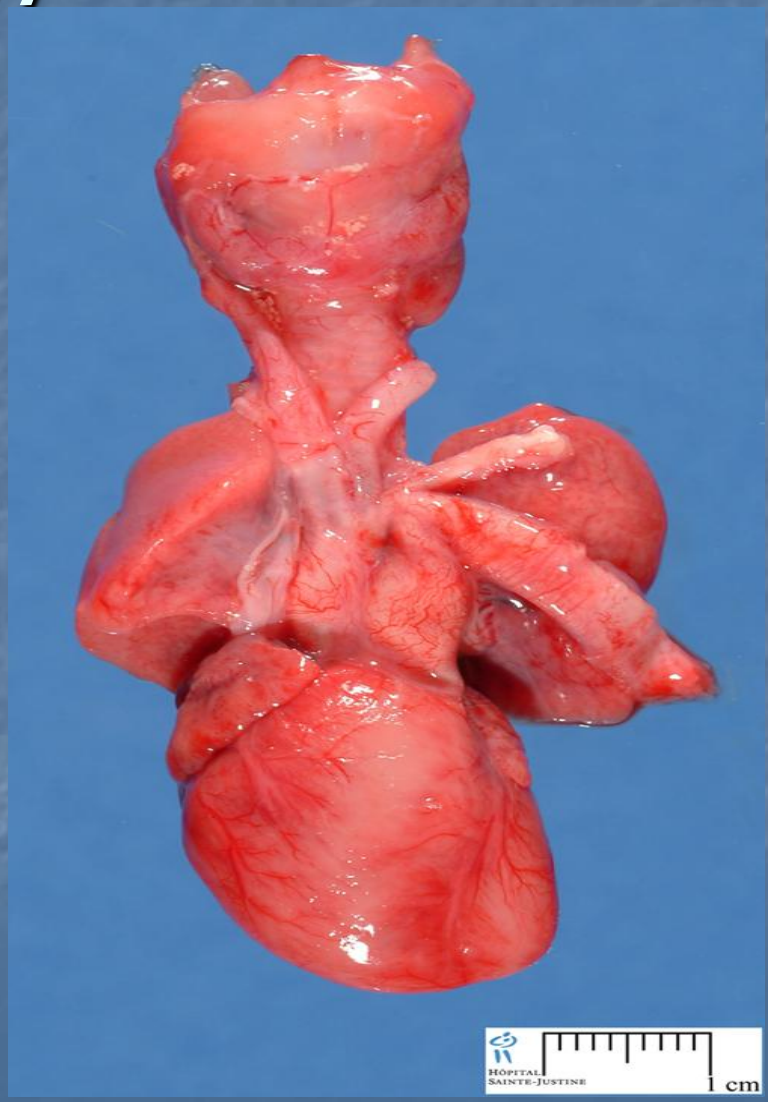
Figure 14: Coronal view on T2-weighted MR image (28 weeks), show the intestinal loops on the left side of the thorax (dark arrow) and the heart placed on the right side (red arrow).

Pulmonary hypoplasia



Anatomopathology show of CDH





Prenatal Counseling
multidisciplinary team

- patient's obstetrician
- perinatologist
- geneticist
- surgeon
- social worker

Prenatal management

- Glucocorticoids
- Thyrotropin-releasing hormone
- Fetal surgical therapy (Antenatal surgical intervention, In utero tracheal occlusion)

Delivery Room Management

- affected infants should be delivered in a center that has experienced personnel and available therapies.
- the team in the delivery room consist of personnel experienced in the immediate resuscitation and stabilization of critically ill neonates
- affected patients in any respiratory distress require positive pressure ventilation in the delivery room.
- To prevent distension of the gastrointestinal tract and further compression of the pulmonary parenchyma, a double-lumen nasogastric or orogastric tube of large caliber is placed to act as a vent.
- Early intubation is preferable to bag-mask ventilation or continuous positive airway pressure via mask or nasal prongs

Postnanal Diagnosis

- Respiratory distress
- Scaphoid abdomen
- Auscultation of the lungs reveals poor air entry
- Shift of the heart to the side opposite

Lab Studies

- Arterial blood gas
  - Obtain frequent arterial blood gas (ABG) measurements to assess for pH, PaCO2, and PaO2.
  - Note the sampling site because persistent pulmonary hypertension (PPHN) with right-to-left ductal shunting often complicates CDH. The PaO2 may be higher from a preductal (right-hand) sampling site.
- Chromosome studies
  - Obtain chromosome studies because of the frequent association with chromosomal anomalies.
  - In rare cases, chromosomal disorders that can only be diagnosed by skin biopsy may be present. If dysmorphic features are observed on examination, a consultation with a geneticist is often helpful.
- Serum electrolytes: Monitor serum electrolytes, ionized calcium, and glucose levels initially and frequently. Maintenance of reference range glucose levels and calcium homeostasis is particularly important

Imaging Studies

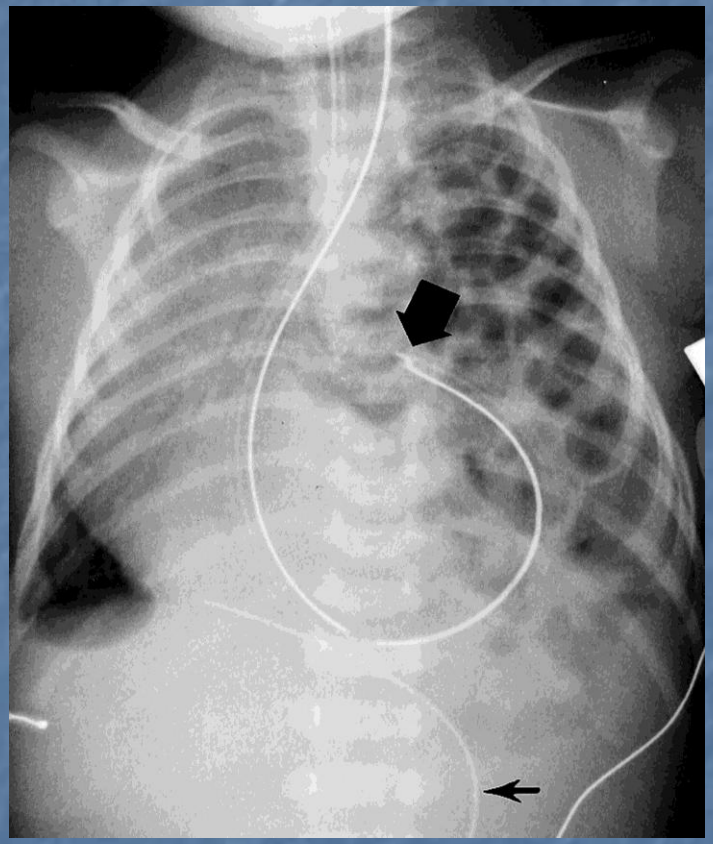
- Cardiac ultrasonography
  - Perform ultrasonographic studies to rule out congenital heart diseases.
  - Because the incidence of associated cardiac anomalies is high (up to 25%), cardiac ultrasonography is needed to evaluate for associated cardiac anomalies.
- Renal ultrasonography: A renal ultrasonographic examination may be needed to rule out genitourinary anomalies.
- Cranial ultrasonography
  - Perform cranial sonography if the infant is being considered for extracorporeal support.
  - Ultrasonographic examination should focus on evaluation of intraventricular bleeding and peripheral areas of hemorrhage or infarct or intracranial anomalies.

Other Tests

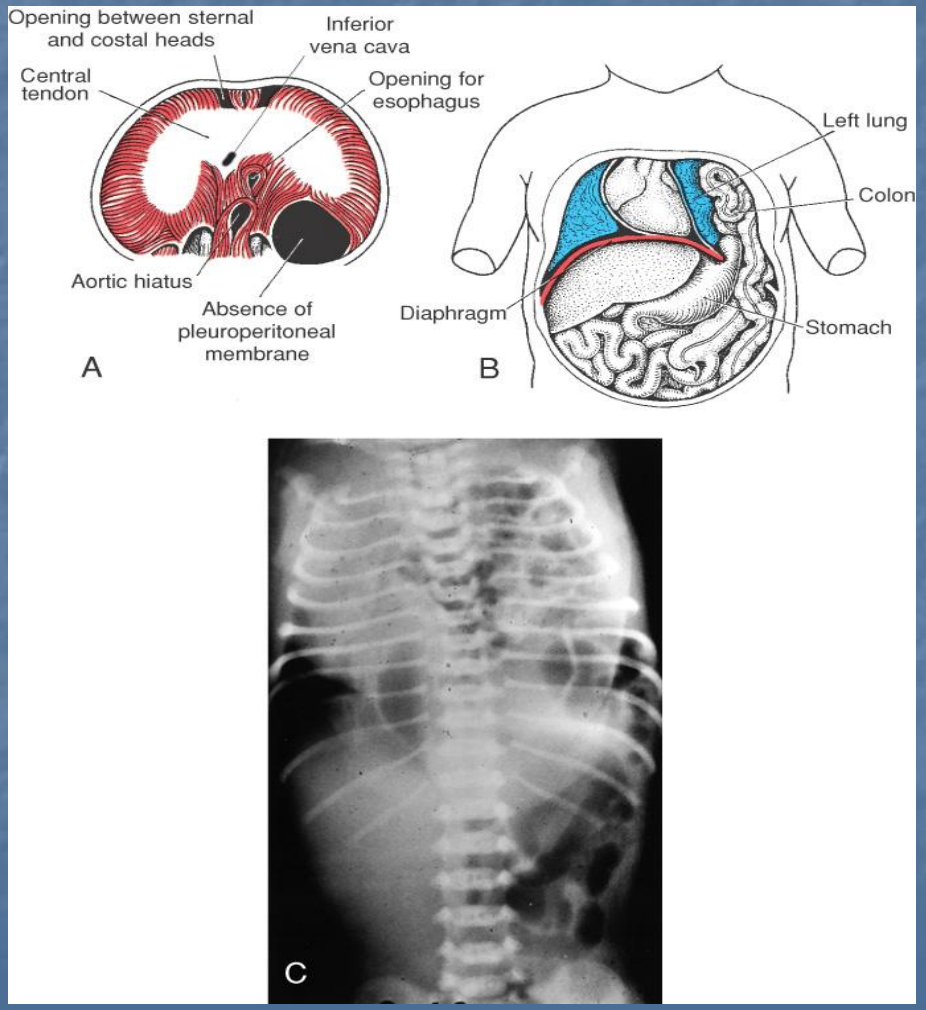
Pulse oximetry

- Continuous pulse oximetry is valuable in the diagnosis and management of PPHN.
- Place oximeter probes at preductal (right-hand) and postductal (either foot) sites to assess for a right-to-left shunt at the level of the ductus arteriosus

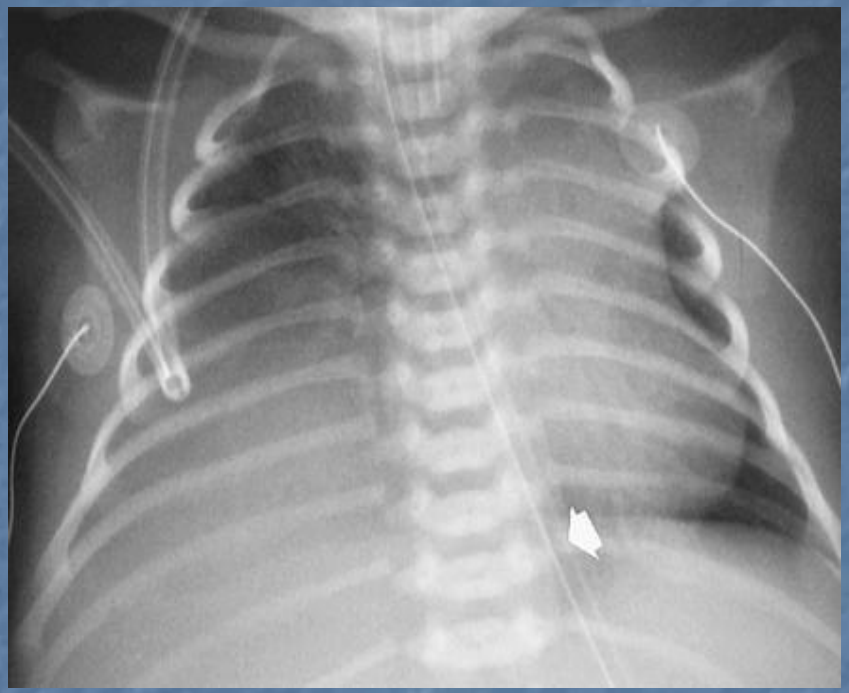
Postnanal Diagnosis left-sided CDH



- Radiograph in a male neonate shows the tip (large arrow) of the nasogastric tube positioned in the left hemithorax. Note the marked apex leftward angulation of the umbilical venous catheter (small arrow).



Right congenital diaphragmatic hernia



- Radiograph in a male neonate shows that the nasogastric tube (arrow) deviates to the left of the thoracic vertebral bodies as it passes through the inferior portion of the thorax

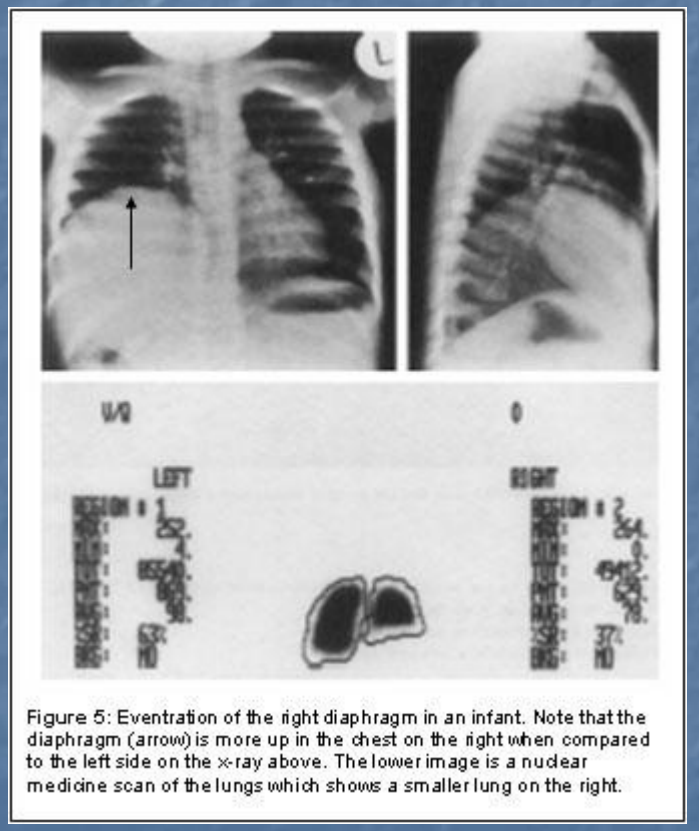


Figure 5: Eventration of the right diaphragm in an infant. Note that the diaphragm (arrow) is more up in the chest on the right when compared to the left side on the x-ray above. The lower image is a nuclear medicine scan of the lungs which shows a smaller lung on the right.

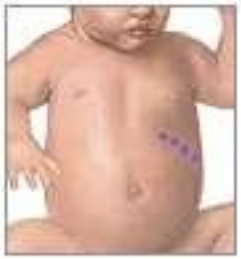
Procedures

- Intubation and mechanical ventilation
  - Endotracheal intubation and mechanical ventilation are required for all infants with severe CDH who present in the first hours of life.
  - Avoid bag-and-mask ventilation in the delivery room because the stomach and intestines become distended with air and further compromise pulmonary function.
  - Avoid high peak inspiratory pressures and overdistension. Consider high-frequency ventilation if high peak inspiratory pressures are required.
- Arterial catheter placement: Place an indwelling catheter in the umbilical artery or in a peripheral artery (radial, posterior tibial) for frequent ABG monitoring.
- Central venous catheter placement
  - Place a venous catheter via the umbilical or femoral vein to allow for administration of inotropic agents and hypertonic solutions such as calcium gluconate

Postnatal management

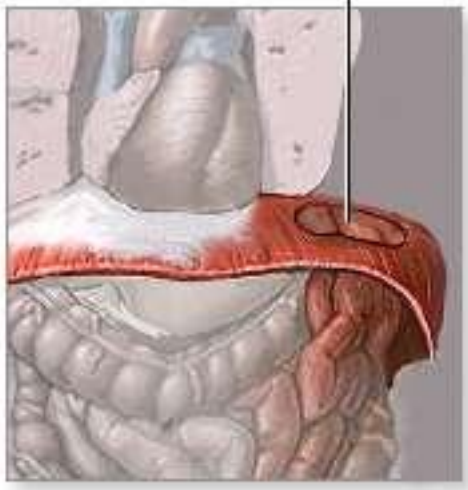
- Mechanical ventilation
- Nitric Oxide
- Surfactant
- ECMO
- surgery

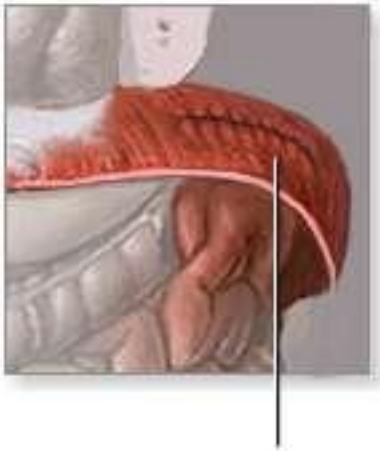
Operative approach



Intestine is pulled bac
down into the abdome

Hole in diaphragm

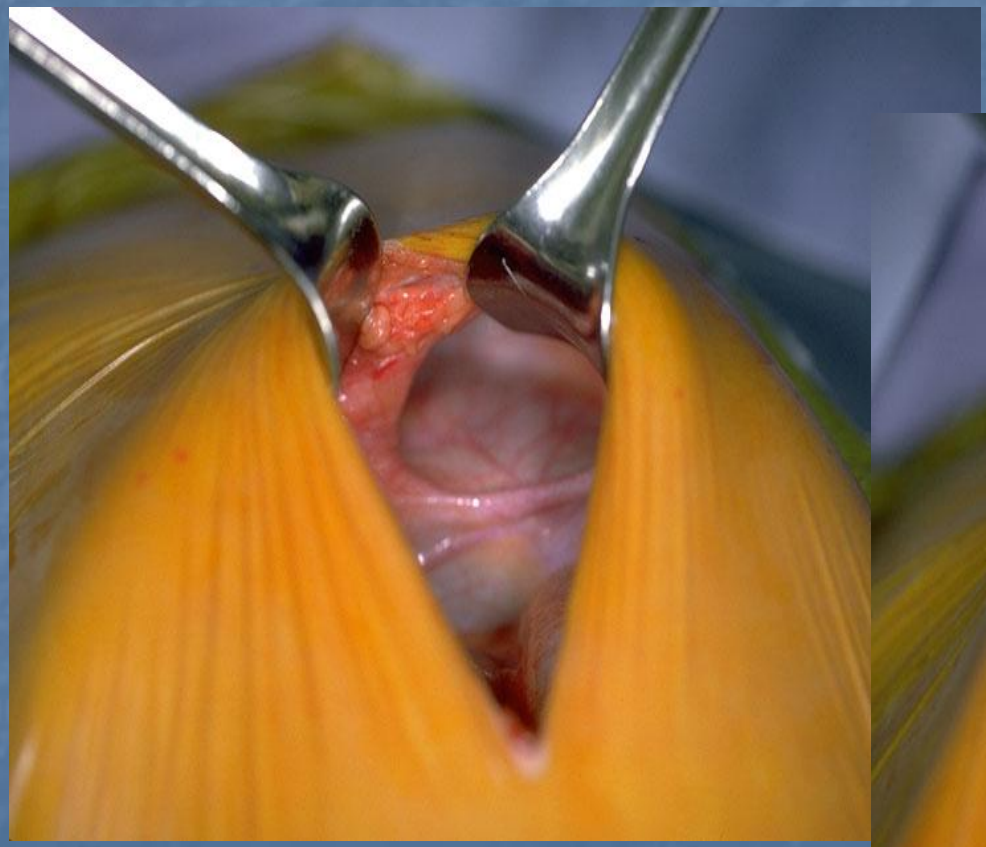


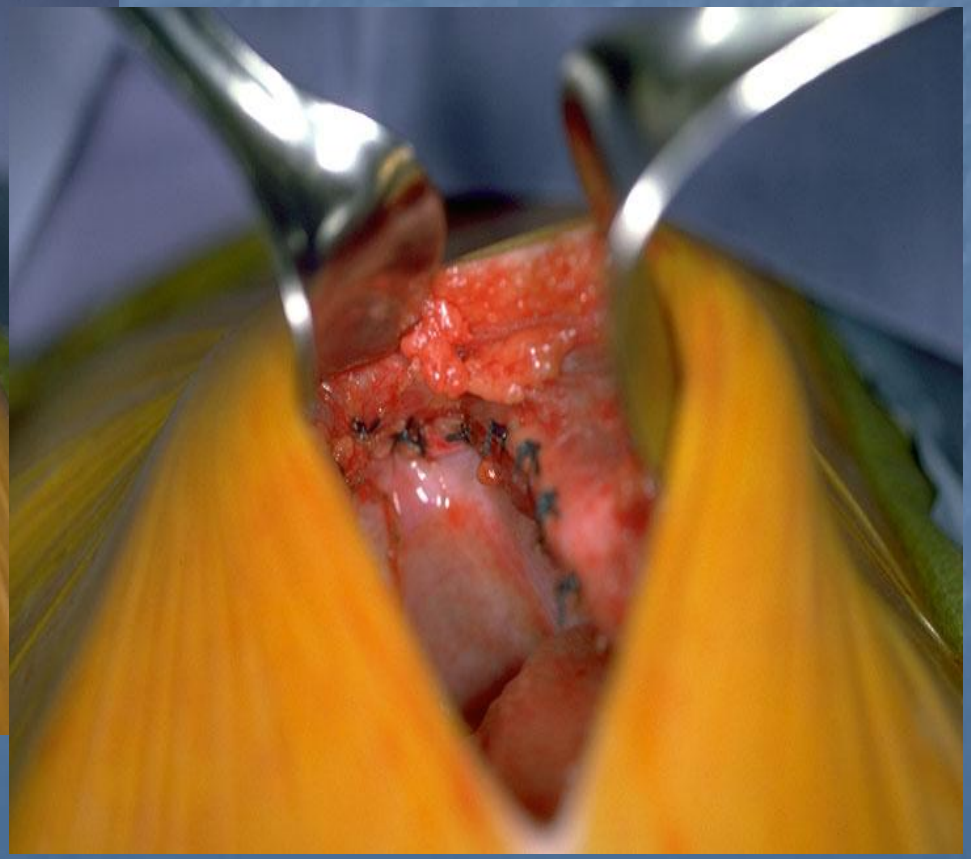


Repaired diaphragm

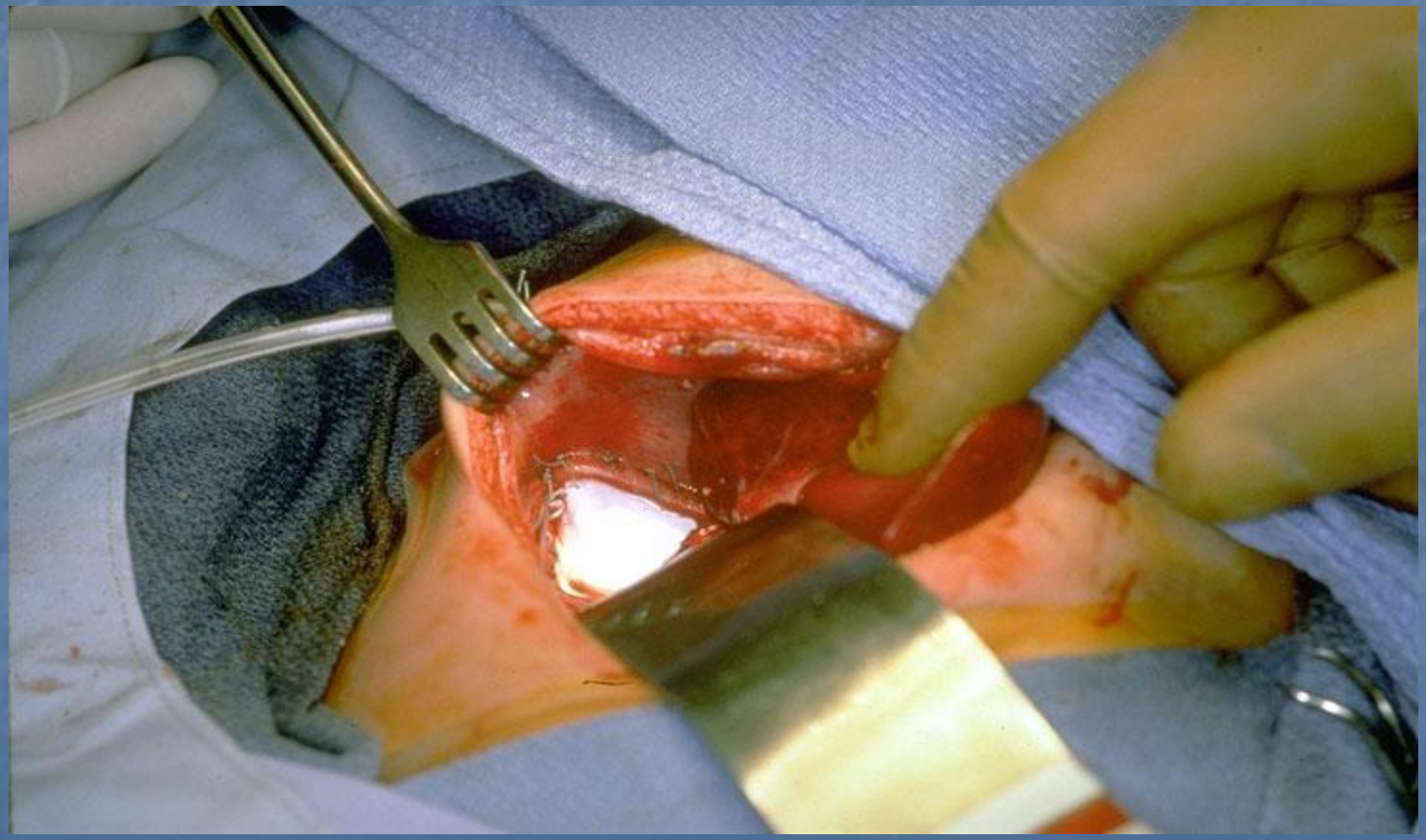
adam.com

The defect in the diaphragm

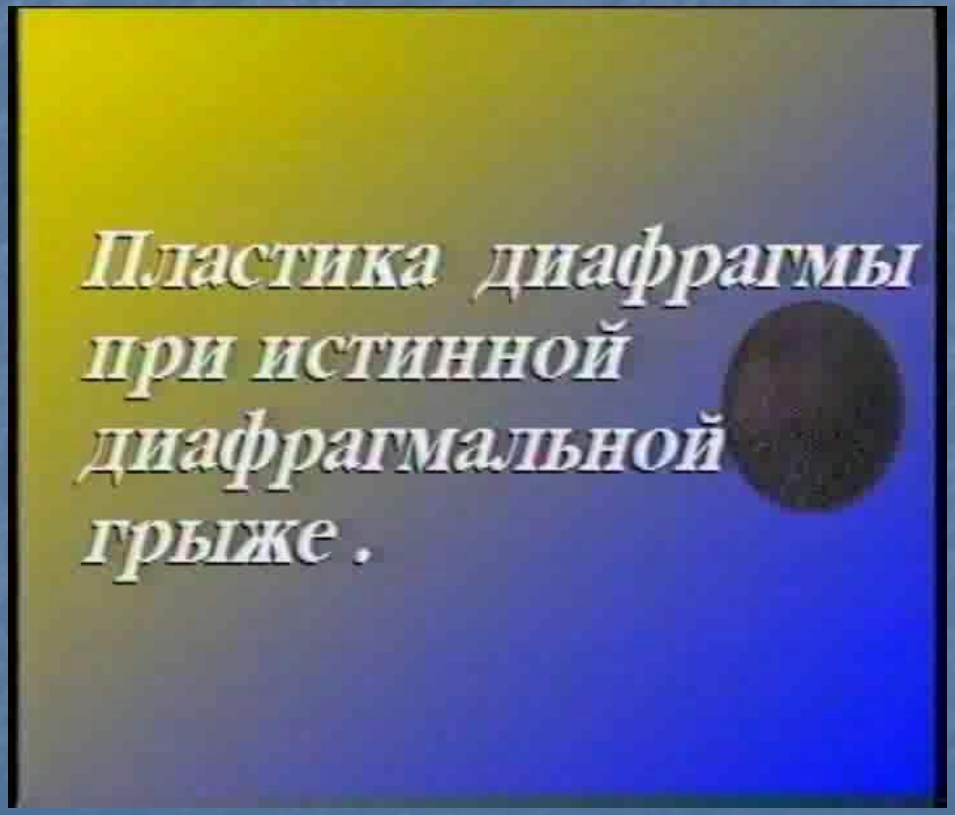




Patch repair of a large defect



Repair of congenital diaphragmatic hernia by VATS



Further Inpatient Care

- Pulmonary care
  - Some severely affected infants have chronic lung disease. These infants may require prolonged therapy with supplemental oxygen and diuretics, an approach similar to that for bronchopulmonary dysplasia.
  - The use of steroids, particularly high doses for prolonged periods, is controversial and may actually hinder appropriate lung and brain development.
- Neurologic evaluation
  - Following recovery, a neurologist or developmental pediatrician should examine the patient, including an evaluation for CNS injury by head CT scanning.
  - Because the incidence of hearing loss is high, perform an automated hearing test prior to discharge.
- Feeding: Incidence of significant gastroesophageal reflux is very high. While most infants can be managed medically, surgical intervention with Nissen or Thal procedures is sometimes required.

Further Outpatient Care

- **Growth**: Failure to thrive is common in a significant percentage of survivors and is most common in severely affected infants. Possible causes include increased caloric requirements because of chronic lung disease, poor oral feeding because of neurologic delays, and gastroesophageal reflux.
- **Developmental follow-up**
  - Because of the risk for CNS insult and sensorineural hearing loss, infants should be closely monitored for the first 3 years of life, preferably in a specialty follow-up clinic.
  - Reassess hearing at 6 months of life (and later if indicated) because late sensorineural hearing loss occurs in a high percentage of patients.
  - Evaluate the patient prior to entering school to determine if any subtle deficits may predispose the patient to learning disabilities.

Prognosis

- Pulmonary recovery: When all resources, including ECMO, are provided, survival rates range from 40-69%.
- Long-term morbidity: Significant long-term morbidity, including chronic lung disease, growth failure, gastroesophageal reflux, and neurodevelopmental delay, may occur in survivors.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