Congenital Diaphragmatic Hernia

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Congenital Diaphragmatic Hernias

Congenital Diaphragmatic Hernia

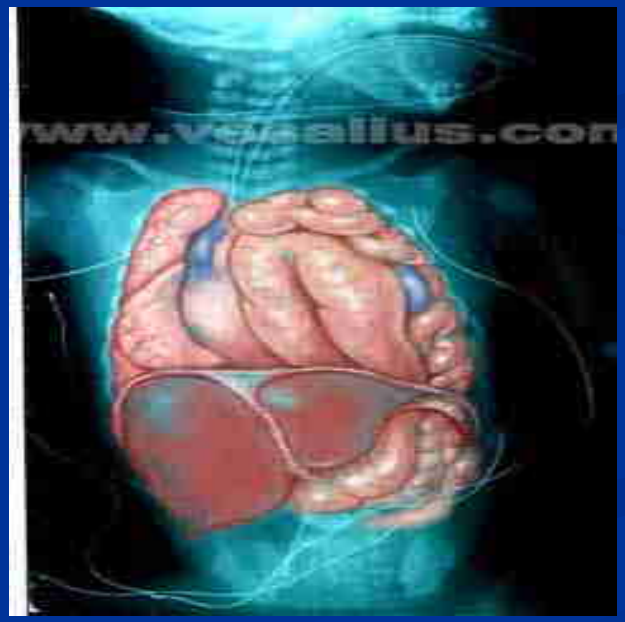
- Incidence 1 in 2000 to 5000 live births.
- 80% in the left side, 20% in the right. Rarely bilateral.
- Survival 60-70%
- Cause is unknown. Failure of normal closure of the pleuroperitoneal canal in the developing embryo. Abdominal contents herniate and compress the ipsilateral developing lung, causing pulmonary hypoplasia and hypertension.

Congenital Diaphragmatic Hernia

- Associated with genetic anomalies:
  - Smith-Lemli-Opitz syndrome
  - DiGeorge syndrome
  - Chromosome 15,18,13 and 21 anomalies
  - Fryns syndrome
  - Pallister-Killian syndrome

Congenital Diaphragmatic Hernia

- Anatomy:
  - Posterolateral: Bochdalek's henia, most common type.
  - Anteromedial: Morgagni's hernia.



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- Clinical presentation:
  - Dyspnea
  - Tachypnea
  - Cyanosis
  - Severe retractions of respiratory muscles
  - Scaphoid abdomen
  - Large antero-posterior diameter of the chest.
- Three general presentations:
  - Severe respiratory distress at the time of birth.(Severe hypoplasia)
  - Respiratory deterioration hours after delivery (honeymoon period). Benefit from correction of hypoxemia and pulmonary hypertension.
  - Feeding difficulties, chronic respiratory disease, pneumonia, intestinal obstruction 24h after birth. (10-20% of patients). Best Prognosis.

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- Diagnosis:
  - 59% antenatal detection with average age 24.2 weeks.(Garne et al, Ult Obst Gyn, 2002)
    - Polyhydramnios
    - Intrathoracic stomach or liver
    - Lung-to-head ratio and lung/transverse thorax ratio.
  - Usually at prenatal ultrasound (15 weeks).
  - Recently fetal MRI and fetal echocardiography, helpful to determine degree of pulmonary hypoplasia. (MRI lung volumetry, left ventricular mass and pulmonary artery diameter)
  - Postnatal diagnosis, on CXR with gastric air bubble or intestine in the chest.
  - Amniocentesis is recommended to provide information regarding possible chromosomal abnormalities.

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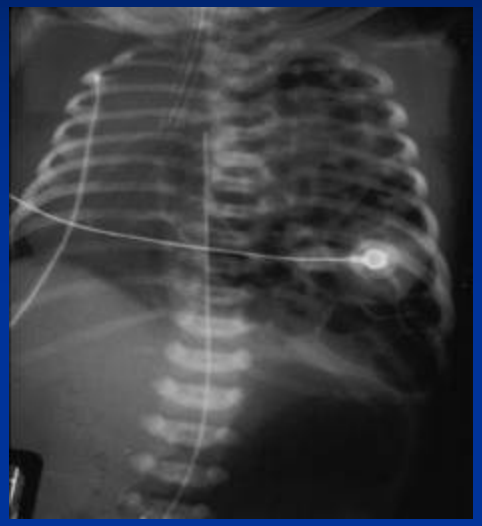

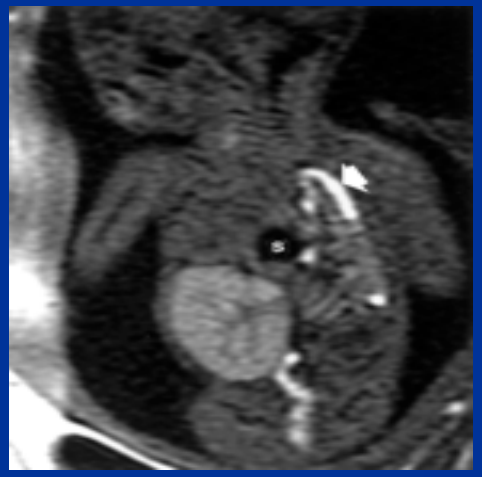

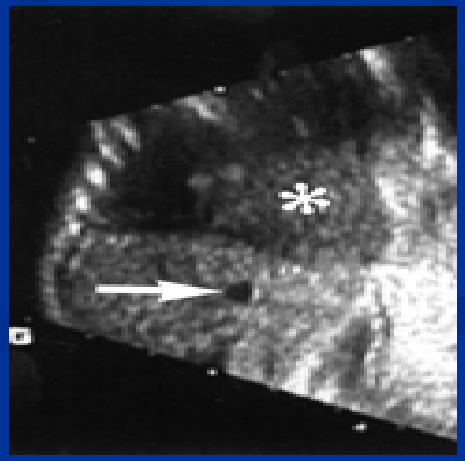

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- Prenatal management:
  - Open fetal surgery: remove the compression of the abdominal viscera. High risk for fetus and the mother. No survival advantage. (Harrison et al, J Ped Surg, 1997)
  - Fetal tracheal occlusion: stimulation of lung growth with accumulation of fluid. Result in larger but persistent abnormal lung. (Flake et al, Am J Obst Gyn, 183, 2000).
  - Steroids therapy weekly to improve lung function is controversial (risk of brain and body development problems). (Ford et al., Ped Surg Int, 2002)

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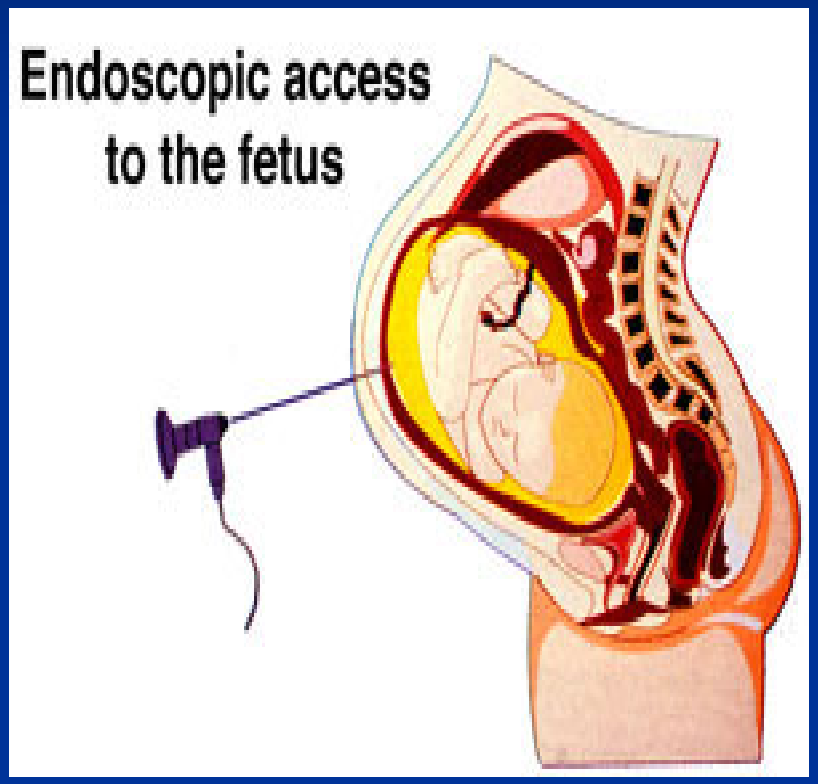

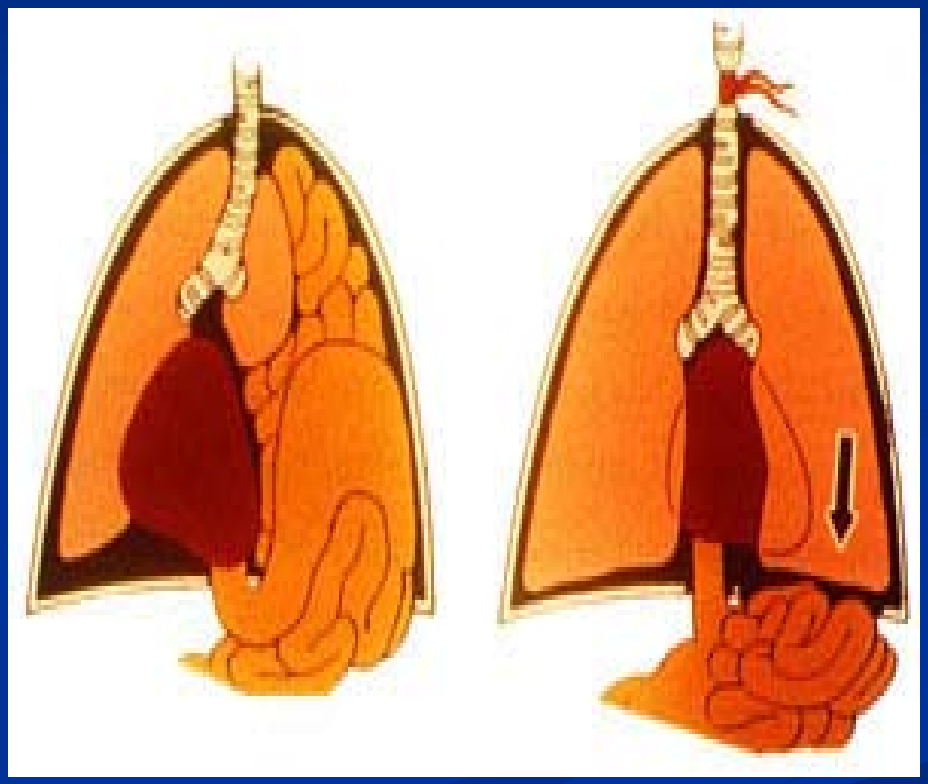

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- Postnatal management:
  - Stabilization of cardiorespiratory system. Endotracheal intubation is critical
  - Nasogastric or Orogastric tube placement.
  - Permissive hypercapnia and stable hypoxemia (>80%), associated with survival of 76%. (Boloker et al, J Ped Surg, 2002).
  - Delayed repair (24 to 72 hours) improves survival when compared with early emergent repair. Allows stabilization of the infant before surgical repair. (Andrew et al., J Ped Surg, june 2004).
  - 1/3 of patients will require ECMO. Head US to r/o hemorrhage prior to ECMO.

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- Reports of 90%survival by Bohn with early use of HFOV and virtually no ECMO. (Bohn. Am J Resp Crit Care Med, 2002)
- Nitric oxide or ECMO for patients unable to be stabilize on reasonable ventilatory setting (pH.7.25, Peak Pressures <30 cm H2O, pre-ductal SO2 >90% with FiO2<60%). iNO beneficial in isolated PPHN. Careful attention to right sided heart failure increases survival by 10%.
- Preliminary results from CDH Study Group: the use of surfactant may worsen outcome.

Congenital Diaphragmatic Hernia

- Factors with influence in the survival rate:
  - Each increase 1mmHg of pCO2 above 50 mmHg increases mortality by 1%
  - Each decrease of 1mmHg in the pO2 below 40% the mortality rate increases by 1%
  - Cardiac disease 3 times most likely to die.
  - Renal disease 6 times more likely to die.

Congenital Diaphragmatic Hernia

- Approaches for surgical repair:
  - Abdominal subcostal
  - Thoracotomy
  - Laparoscopic vs Thoracoscopic
    - MIS ideal for Morgagni hernias but can be challenging because the peumoperitoneum widens the defect. Laparoscopy for Bochdalek's has a high failure rate and is associated with ↑pCO2 and acidemia
    - Contraindicated if very high pCO2.
    - Thoracoscopy is better approach for Bochdalek hernias with recurrence of 14%. Open approach 3-22%.(Marjorie et al, J Ped Surg, Nov 2003.)
- Small defect can be repaired primarily. Large defect will require abdominal or thoracic muscle flaps, or prosthetic patch (tension free).

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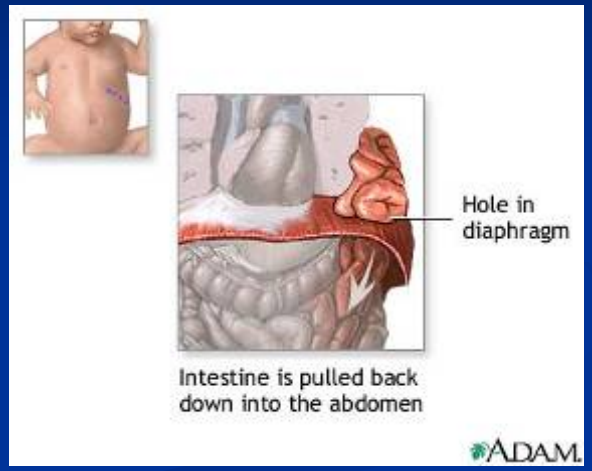

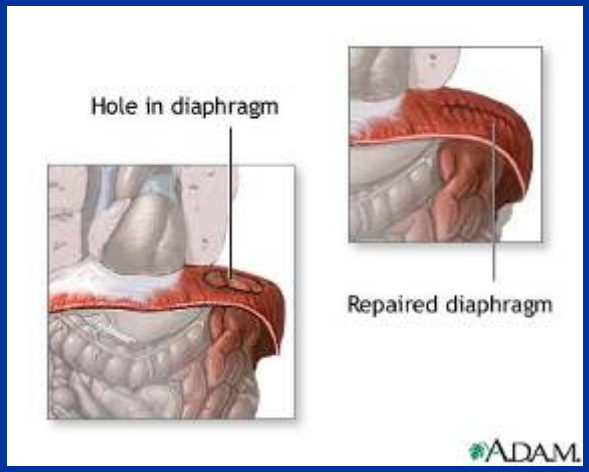

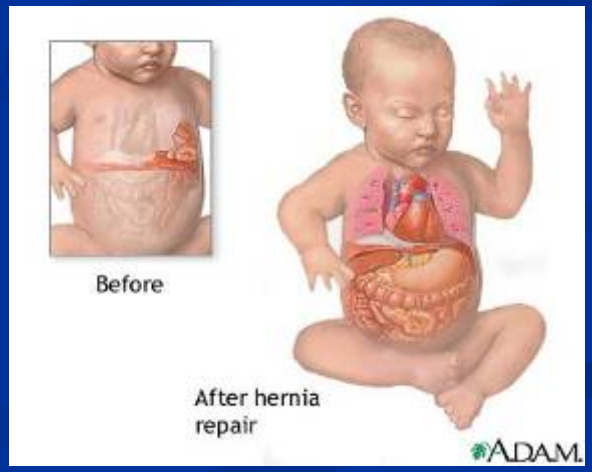

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- Long term follow up:
  - GERD
  - Foregut dysmotility
  - Chronic lung disease
  - Scoliosis
  - Pectum excavatum
  - Cognitive skills, developmental delay, seizures, hearing loss if severe respiratory distress.

Conclusions

- CDH is a congenital anomaly with a high mortality. Usually associated with pulmonary hypoplasia and hypertension.
- Surgical repair is only treatment. Delayed surgery until the patient is stable is associated with better outcomes.
- Congenital cardiac and renal diseases, hypoxemia and hypercapnia increases mortality.
- HFOV, ECMO, iNO has improved the survival of CDH.
- Permissive hypercapnia with acceptable pO2 has shown to improve survival.
- Long term follow up is necessary to detect complications.
- Tracheal occlusion in utero, keeps lung expanded but is a abnormal lung.
- Primary repair is small defect, patch if large defect, to prevent tension.