Congenital Diaphragmatic Hernia

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Congenital Diaphragmatic Hernias
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- 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.
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- Associated with genetic anomalies:
  - Smith-Lemli-Opitz syndrome
  - DiGeorge syndrome
  - Chromosome 15,18,13 and 21 anomalies
  - Fryns syndrome
  - Pallister-Killian syndrome
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- **Anatomy:**
  - Posterolateral: Bochdalek’s hernia, 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, 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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- Prenatal management:
  - 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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**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.
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.
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Factors with influence in the survival rate:

- Each increase 1 mmHg of pCO2 above 50 mmHg increases mortality by 1%
- Each decrease of 1 mmHg 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.
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- Approaches for surgical repair:
  - Abdominal subcostal
  - Thoracotomy
  - Laparoscopic vs Thoracoscopic
    - MIS ideal for Morgagni hernias but can be challenging because the pneumoperitoneum 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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- 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.