Congenital Diaphragmatic Hernia

- 1 in 2000 to 500 live births
- Slight male preponderance
- 80% Left, 20% Right and <1% bilateral
- Bilateral has high incidence of assoc anomalies
- Bochdalek (posterolateral) and Morgagni (anterior)
- Isolated CDH likely to be male, macrosomic and prem
- Associated anomalies in 40-60% of cases: Cardiac (15%), Neural Tube, Chomosomal, Renal, Genital, Pulmonary sequestrations, Malrotation and Duodenal atresia (40%)

Pathogenesis

- Diaphragmatic hernia thought to be secondary to lung hypoplasia, not the other way round
- Hypoplasia ipsilateral lung with possible contralateral as well
- Lung: Fewer alveoli, thickened walls and interstitial tissue, decreased gas exchange, decreased vascular density with resulting PHT (potentially fixed)
- Degree of PHT and hypoplasia are determinants of outcome

Diagnosis

- By prenatal ultrasound at 24 weeks. Bowel loops in chest
- Prognosticate by measuring right lung size, thoracic volume and total lung volume. Commonest is head to lung ratio for prognosis
- Once delivered: Scaphoid abdomen, unilateral breath sounds, compressed mediastinum, deviated trachea

Prognosis

- Mortality 21-48%
- Anatomical predictors of prognosis:
  - Associated anomalies
  - Liver herniation May need patch and ECMO
  - Lung:head ratio < 1 has high mortality
- Physiological parameters
  - Ventilatory Index (RR x (PIP-PEEP)) of < 1000 to keep $PaCO_2$ < 40mmHg has 100% survival
  - Modified Ventilatory Index (RR x PIP x $PaCO_2$)/1000 of < 40, 96% survive BUT > 80, all die
  - Oxygenation Index (MAP x FiO_2 x 100/PaO_2) < 6, 98% survive BUT > 17.5, all die **[MAP is Mean Airway Pressure]**
- Pulmonary Function Parameters
  - If pre-op compliance > 0.25ml/cmH_2O/kg and Vt > 3.5ml/kg/L don't need ECMO
- Other poor features: Major cardiac defects, ECMO > 2 weeks, PHT for > 3 weeks, LBW, prematurity, low 1&5 minute APGARS, right-sided defect

Management

- Intrauterine Foetal Endoscopic Tracheal Occlusion no longer done as increased lung size but not doesn't help hypoplasia
- Decompression of stomach by NGT/OGT
- Avoidance of PHT as much as possible

Perioperative:

- Physiological emergency not surgical
- Gentle bagging to avoid distention of gut and lung protection
- Permissive hypercapnoea
- Preductal PaO_2 > 66mmHg waned SpO_2 > 85%. Postductal SpO_2 > 70mmHg
- Sats of less than 80 on Fi 0.6 indication for "other ventilation" (ECMO, HFOV and Inhaled NO)
- PHT major concern
- Opiate based and N_2O avoidance
- Prepare to manage systemic hypotension and PHT crisis
- A-Line highly recommended
- Surgery: Thoracoabdominal incision/Laparoscopic/Thoracoscopic

Outcomes:

- Survival rates 60-90%
- Sequelae: Chronic lung disease, bronchopulmonary dysplasia, reactive airway disease, neurological injury
- Persistent pulmonary hypertension a major issue

Indications for ECMO (EURO Consortium Consensus):

- Hypoxia – Defined as preductal saturations less than 80-85%
- Acidosis – Defined as metabolic (lactate > 5 or pH < 7.2) or respiratory (pH < 7.2 due to hypercarbia)
- Hypercarbia – Defined as $PaCO_2$ > 70mmHg
- Hypotension – Defined as poor tissue perfusion, urine output < 0.5ml/kg/hr or unresponsive to inotropic support

Many facilities also use ventilatory measures:

- PIP exceeding 26cmH_2O
- HFOV needing to exceed MAP of 14-15cmH_2O