Congenital Diaphragmatic Hernia

A failure of closure of the diaphragm resulting in abdominal contents invading the hemithorax; as a result, the infant has pulmonary hypoplasia, presenting as hypoxemic respiratory distress or failure complicated by pulmonary hypertension

ANESTHETIC CONSIDERATIONS:

- Considerations of the neonatal/preterm patient
- Urgent, non-emergent procedure with time for preoperative optimization
- Risk of aspiration requires rapid airway control and gastric decompression
- Physiologic consequences of CDH:
  - Hypoxemia (secondary to pulmonary hypoplasia +/- R-L shunting)
  - Pulmonary hypertension (increased PVR due to underdeveloped pulmonary vascular bed)
  - Restrictive lung pattern (secondary to hypoplasia + bowel occupying chest)
  - Abnormal major vessel / cardiac orientation (displaced mediastinum)
- Ventilation strategy:
  - Avoid barotrauma (risk of life threatening PTX on good side)
  - Conventional ventilation vs. permissive hypercapnia vs. HFOV
  - +/- ECMO, surfactant, iNO, etc.
- Other congenital defects: ~25% have a cardiac defect
- Multidisciplinary care (ICU, NICU, surgery, anesthesia)

ANESTHETIC GOALS:

- Early airway management (avoid gastric distention):
  - Rapidly secure a/w - spontaneously breathing intubation vs. RSI
  - Avoid BMV with possibility of gastric / bowel distension further impairing oxygenation / ventilation
  - NG to decompress stomach
- Ventilatory Goals:
  - Avoid barotrauma to already compromised lungs (PIP < 25-30 cmH_2O)
  - PTX immediately life-threatening complication
  - Permissive hypercapnia is acceptable but must consider effects on PVR (pCO_2 60-65 mmHg, preductal SpO_2 > 90%)
  - Consider NO, HFOV, or ECMO
  - Avoid nitrous
- Avoid precipitants of ↑ PVR:
  - Hypoxia, hypercarbia, acidosis, atelectasis, hypothermia, elevated airway pressures, pulmonary vasoconstriction, light anesthesia, and polycythemia
- Patient should be medically stabilized prior to surgical repair (because pulmonary hypoplasia / hypertension is not relieved with evacuation of bowel from thorax)

HISTORY

- Plan for timing of repair
- Timing of diagnosis (antenatal, postnatal)
  - Often antenatal diagnosis based on routine U/S
- Classic findings: hypoxemia, scaphoid abdomen, bowel sounds in chest, bowel on CXR
- Postpartum course:
  - Immediate respiratory distress vs. more insidious (hypoxemia)
  - Already intubated?
  - Current ventilation strategy (low Vt / high RR / low a/w pressures, permissive hypercapnia, HFOV, +/- ECMO)
    - Conflict: minimizing pHTN (i.e. avoid hypercarbia) but want to avoid barotrauma, volutrauma, etc. associated with traditional ventilation strategies and very poorly compliant diseased lung
      - So, **despite our efforts to attempt to minimize precipitants of pHTN** (e.g. hypercarbia), **current recommendations are permissive hypercapnia ventilation strategies** as trauma to diseased lung more harmful than (hopefully) transient pHTN
- Severity of defect and resultant pulmonary hypoplasia +/- presence of cardiac hypoplasia
- Severity of pulmonary hypertension, R-L shunt with persistent fetal circulation
- Any other associated congenital defects
- Any complications of treatment (barotrauma, PTX, ECMO related – cannulation difficulties / heparinization / intracranial bleeds etc.)

PHYSICAL

- **VITALS** - HR, BP, $Sp0_2$ (pre and post ductal), temperature
- **GENERAL** - Cyanosis (peripheral / central) in nail bed and under tongue
- **RESP** - Unilateral breath sounds, bowel sounds in chest (usually left), increased WOB, respiratory distress / failure, barrel shaped chest
- **CVS** - Displaced apex & HS, murmur or separate cardiac defect, RV heave, RV volume overload

INVESTIGATIONS

- **Labs**
  - CBC, lytes, BUN, Cr, INR / PTT, ABG, group & screen
  - ABG / capillary gas (hypoxemia, acidosis)
- **Imaging**
  - CXR (primary defect R vs. L, line & tube placement, PTX)
  - Echo (clarify anatomy, assess obstruction of great vessels due to displacement) - PA pressures, shunting
  - U/S Abdomen
  - CT (assess anatomy, assess amount of lung parenchyma, r/o secondary complication, volume, aspiration, barotraumas etc.)

OPTIMIZATION

- Diagnosis is usually made antenatally, therefore plans should be made for delivery in a facility capable of treating neonate
- Supplemental O_2, airway control (avoid BMV)
- Plan for immediate post delivery intubation (**awake spontaneous breathing**, no drugs), assess ventilation strategy on the degree of hypoxemia and lung compliance
- **Gastric decompression**
- Transfer to ICU setting for management of respiratory failure, pulmonary hypertension
- Neonatology, surgery consultation
- Cardiology consult if congenital cardiac lesion or hypoplastic LV
- May require **ECMO** due to persistent respiratory failure (hypoxemic / hypercarbic)
- Consider **surfactant, HFOV, iNO, sildenafil,** etc. (yet evidence not clear)

ANESTHETIC OPTIONS

- GA
    - Usually already intubated, therefore narcotic + relaxant technique (common, safe & easy)
    - Goals: facilitate surgical repair, **minimize precipitants of pHTN**, avoid secondary lung injury with ventilation strategy
    - Minimize opiates, consider regional anesthesia for pts with small hernias and minimal respiratory distress with potential for early extubation

ANESTHETIC SETUP

- **Drugs**
    - Emergency drugs
- **Equipment**
    - CAS + **arterial line (pre-ductal**, right radial)
    - **Pre- and post-ductal SpO_2 monitors**
    - $EtCO_2$ may be unreliable due ventilation strategy (esp. not feasible during HFOV, rely on ABG)
    - Temperature monitoring
    - **Avoid venous access in lower extremity** as venous return may be impaired by tight abdominal closure
    - Warming blanket, heated room
    - Fluid warmer
    - +/- CVP (for monitoring pressure / inotropes)
    - ICU ventilator / HFOV

MANAGEMENT OF ANESTHESIA

- **Induction**
    - ETT and NGT usually in situ, otherwise **awake NGT and ETT vs. RSI** (no / minimal BMV)
    - Consider pre intubation anticholinergics
    - IV / **Inhaled agents acceptable**, observe hemodynamic goals of co-existing cardiac disease
    - **Avoid precipitation of pulmonary hypertension**, observe protective ventilation strategies (PIP < 25-30 cmH_2O)
- **Maintenance**
    - Ventilation strategy to minimize barotrauma yet avoid of hypoxemia or hypercapnia
        - Generally small Vt, high rate, +/- PEEP
    - **Opioid / relaxant technique common**
        - **Avoid N_2O** (potentially distend bowel already in chest)
    - **Inhaled anesthetics** (low concentrations) also **ok**
    - Any that observes goals for oxygenation, ventilation and pulmonary hypertension
    - NMB required for closure of abdomen (**potential for intra-abdominal compartment syndrome**)
        - Can monitor with **pulse oximeter on lower extremity**
- **Emergence**
    - **Do not inflate hypoplastic lung post-reduction**
    - PICU / NICU for post op ventilation, pulmonary hypoplasia is not reversed with surgery
    - Morphine infusion for analgesia

DISPOSITION & MONITORING

- ICU, continued respiratory support
- Consider HFOV, ECLS, ECMO

COMPLICATIONS

- **PTX** – life threatening hypotension / hypoxemia (increased a/w pressures) – immediate decompression required
- **Great vessel kinking** – hypotension secondary to displacement of mediastinum / heart
- **Pulmonary HTN** – acute crisis
- **Abdominal compartment syndrome**

PATHOPHYSIOLOGY

- Incidence 1:2000-4000 neonates
- Incomplete embryologic closure of diaphragm
- Commonly defect is in the foramen of Bochdalek, resulting in herniation through the L side of the diaphragm into the L hemithorax
- Uncommonly through foramen of Morgagni into the R hemithorax
- Herniation of abdominal contents into hemithorax of affected side, resulting in hypoplasia of the lung due to compression in utero
- Severity of hypoplasia related to timing of defect and herniation in utero → earlier herniation → more severe pulmonary hypoplasia and poorer prognosis
- Possible hypoplasia of left ventricle → post-natal cardiac insufficiency
- **Mortality related to pulmonary hypertension, pulmonary hypoplasia** & hypoxemia

- Survival rates 40-50%, but with delayed closure and pre-operative stabilization survival can be as high as 90%
- **Interventions and Outcomes:**
    - Prenatal surgical interventions have been described and include:
        - Open fetal surgical repair
        - Open surgical occlusion
        - Endoscopic external tracheal occlusion
        - Endoscopic fetal endoluminal tracheal occlusion (FETO)
            - FETO accelerates fetal lung growth, alveolar and capillary growth and remodeling of pulmonary arterioles
            - Observation: occlusion of the larynx results in decreased egress of intraluminal lung fluid causing rapid pulmonary growth (as observed in neonates with laryngeal atresia and hyperplastic lungs)
    - **Delayed repair has shown to improve survival** and decrease need for ECLS compared to early repair
    - **Permissive hypercapnia / protective ventilation strategies have shown improved survival,** particularly when combined with delayed repair (post-ductal PaO_2 30-40mmHg, $PaCO_2$ 50-80 mmHg)
    - Inhaled NO prior to repair is not effective, and has been shown by some to increase the need for ECLS
        - There may be a role for iNO following delayed repair or following ECLS with persistent pulmonary hypertension
    - Lung Transplant options are limited in pediatric patients, some cases of lung segment transplantation to allow contra lateral lung to develop and pulmonary hypertension to improve, then transplant pneumonectomy at later age

REFERENCES

- Stoelting Co-Existing Disease 5th Ed.
- Bosenberg, et al. Management of Congenital Diaphragmatic Hernia. Curr Opin Anes 2008;21:321-331.
- Ivascu, F & Hirschl, R. New Approaches to Managing Congenital Diaphragmatic Hernia Seminars in Perinatology; 2004, 28 (3) pp 185 – 198
- Liu MP et al Pediatric Emergencies, Neonatal Surgical Emergencies Anes Clinics of NA, Vol 19 N2 June 2001
- Miller 7th Ed
- Barash 6th