CONGENITAL DIAPHRAGMATIC HERNIA (CDH)

Surgical Specialty:	General Surgery
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Background:

- General Considerations
  - Congenital Diaphragmatic Hernia (CDH) is a developmental discontinuation of the diaphragm that allows the viscera of the abdomen to herniate into the chest.
    - Herniation occurs during the period of lung development when the pulmonary arteries and lung bronchi begin branching approximately between 6 and 12 weeks post-fertilization
    - The importance of the defect is variable from A (small muscular defect) to D (agenesis of the diaphram)(international classification). Large defects are closed with a tissue patch (e.g. Gore-Tex®).
    - In developed countries, the diagnosis is often made at prenatal screening
  - The most common and largest diaphragmatic defect occurs through the left posterolateral pleuroperitoneal canal (foramen of Bochdalek) and accounts for > 75% of all cases. The remainder of the defects occur at the right posterolateral foramen of Bochdalek, at the anterior foramen of Morgagni, and in the paraesophageal sites. Bilateral cases are very rare.
  - Pulmonary hypoplasia and pulmonary hypertension are the major sequelae, presenting with the classic triad of cyanosis, dyspnea, and scaphoid abdomen
  - On physical examination, typical findings include a scaphoid abdomen, bulging chest, decreased breath sounds, distant or laterally-displaced heart sounds, and (rarely) bowel sounds in the chest
  - Herniated contents can include the stomach, small bowel, pancreas, and liver.
  - Severe cases of lung hypoplasia and pulmonary hypertension require early initiation of Extracorporeal Membrane Oxygenation (ECMO) followed by delayed surgical repair
  - The timing of surgical repair is institution and patient dependent. Typically, repair t occurs within the first week of life following medical stabilization. It is no longer considered a surgical emergency
  - The decision to operate is a multidisciplinary decision based oninstitutional criteria. For example, the following criteria are used by the CDH EURO Consortium:
    - Normal mean arterial blood pressure for age
    - Preductal SpO_2 between 85% à 95% with a FiO_2 < 50%
    - Blood lactates < 3 mmol/L
    - Urine output > 1 ml/kg/h
  - Late presentation: With mild muscular defect or anterior hernia and minor pulmonary hypoplasia, CDH may be asymptomatic at birth and remain undiagnosed for days or weeks. The diagnosis is usually made following an acute episode of respiratory distress (due to airway infection), asthma-like episodes or fortuitously. The prognosis

of these so-called " late forms" is much better than that of the classic neonatal forms. They are managed as a semi-urgent laparotomy or laparoscopy

- Associated Comorbidities/syndromes
  - The majority of CDH cases are *not* part of a genetic syndrome and typically present as an isolated defect in 50-70% of cases[2].
  - There are non-isolated CDH associated with specific chromosomal anomalies (trisomy 13, 18, or 21), and CDH has been observed in over fifty syndromes (not covered in this review). A congenital cardiac (ASD, VSD, Fallot, coarctation) or pulmonary (sequestration) anomaly can be associated.
  - Reduction of abdominal contents into the small abdominal cavity may acutelyincrease intraabdominal pressure, decrease venous return, impair cardiac output, decrease GI, liver and renal perfusion (abdominal compartment syndrome)
  - Pulmonary hypoplasia is present on both sides but is more severe on the ipsilateral lung, ~~I~~mpaired pulmonary vascular development results in pulmonary hypertension. Neonates with CDH are at risk for pulmonary hypertensive crises
  - The degree of pulmonary hypoplasia and hypertension is predictive not only of mortality but of the development of long-term sequelae in the survivor
  - Cardiovascular collapse may occur due to any combination of mechanical factors discussed aboveand physiologic sequelae such as pulmonary hypertensive crises
  - Adrenal insufficiency is not uncommon and should be kept in mind in case of poor response to inotropes or vasoconstrictors

Anesthetic Planning:

- Pre-Anesthetic Evaluation
  - The patient's status (e.g. prematurity) and comorbidites significantly affect the anesthetic plan and preparation
  - A complete blood count, complete metabolic panel, lactate, coagulation assays, arterial blood gas, chest X-ray, echocardiography, and a head ultrasound are part of the postnatal workup performed by Neonatal Intensive Care Unit (NICU) and should be available for review
  - Review all pertinent ventilator parameters during handoff in the NICU, including FIO_2, PIP, V_T, and PEEP
  - In severe cases, the patient may have surgical repair while on ECMO.
    - Anesthesia may or may not be involved in the care of these patients. Specific considerations for the care of the patient on ECMO are not covered in this review.

- Specific or Unique Room Set-Up Requirements
  - Airway
    - The infant with CDH is typically intubated immediately at delivery and presents with an existing endotracheal tube (ETT). To ensure adequate ablity to ventilate and minimal leak around ETT, reintubation with a cuffed ETT may be required
    - Ventilation may be conventional or high frequency ventilation (HFOV). Discuss considerations for availability of different ventilators, transport from ICU, and setup in OR.
  - Drugs/Infusions
    - Consider inhaled Nitric Oxide (iNO) if pulmonary hypertension is present. Equipment to deliver iNO in the inspiratory limb of the ventilator should be present and ready to use

- Opioids
- Muscle Relaxation
- Volatile is available with anesthesia ventilators but not for HFV or ICU ventilators. (In these cases, total intravenous anesthesia (TIVA) is used Pharmacologic options includeopioid, benzodiazepine, ketamine, and muscle relaxants
- Have emergency drugs available
    - Epinephrine (1-10mcg/kg)
    - Phenylephrine (0.5-2mcg/kg)
    - Calcium chloride (10-20mg/kg) / Calcium gluconate (30-40mg/kg)
    - Atropine (10-20mcg/kg)
- Infusions: (recommended initial doses)
    - Epinephrine (0.05mcg/kg/min)
    - Vasopressin (20milli-unit/kg/hr)
    - Norepinephrine (0.05mcg/kg/min)
    - Milrinone (0.5mcg/kg/min)
- Consider having albumin drawn up and ready to bolus

- Monitors
    - Standard ASA monitors + Arterial line
    - Pre- and post-ductal pulse oximeter
    - Consider Near-Infrared Spectroscopy (NIRS)
        - NIRS changes may be an early indicator of impaired perfusion
        - Somatic (flank) oximetry values typically drop before cerebral values and are an early indicator of impaired perfusion
        - Cerebral oximetry can be used to ensure cerebral perfusion is kept near baseline values
            - Over-ventilation can cause cerebral vasoconstriction
            - Cerebral oximetry changes are a later sign of impaired perfusion due to cerebral autoregulation
- Blood Availability
    - Type and cross (prepare) 15-20mL/kg
- ICU Bed Availability
    - These patients are frequently critically ill and come to the operating room from the NICU
    - Ensure availability of post-op ventilation
- Other Indicated Resources (eg. ECMO standby)
    - If the patient is an ECMO candidate, make sure perfusion team is aware and ready
    - Refer to your institutional guidelines regarding ECMO candidacy
    - Discuss ECMO candidacy and intra-op ECMO criteria with the surgical team before starting
    - If the patient deteriorates, everyone involved should know the next step. This is not a discussion you want to start while coding the patient
    - (a bit redundant?)*Note:* If there is a significant chance of peri-operative death, this should be clearly communicated to the family and all staff present
- Cases performed in the ICU
    - There are instances when a case will be booked and performed at the bedside in the ICU. This is often indicative of how “sick” the patient is

- Discuss the case with the surgeon and specifically identify what makes the patient "too sick" to transport
    - Evaluate whether the risks of transport (mobilization, changes in ventilation, risk of hypothermia etc) are outweighed by the benefit of being in the OR if an emergency occurs
- Managing a critically ill patient outside the OR has disadvantages that necessitate early planning. Plan ahead! Considerations include:
    - Help is often far away. *Identify potential help and contact them in advance. If able, consider requesting additional anesthesia providers*
    - Emergency supplies are also often far away. *Bring emergency equipment and drugs in advance*
        - Airway, Code, and line carts if possible
        - Ultrasound if additional lines anticipated
        - Infusion medications and pumps
    - (discussed earlier)Unfamiliar environment. *If time allows, orient you and your team to a typical bedside setup and clearly define positions, roles, and equipment*
    - Charting of vitals may not be automatic. *Bring the required equipment for documentation. Consider assigning a team member to assist in charting, minimizing distractions from direct patient attention and care*

Intraoperative Considerations:

- General
    - The anesthetic should aim to avoid additional pulmonary complications that include
        - Pulmonary hypertensive crises with R to L shunting and cyanosis
        - Pneumothorax due to excessive ventilation pressure
- Induction
    - Patients typically have an ETT in place
    - Mask ventilation is ideally avoided as insufflation of the intrathoracic stomach will decrease pulmonary compliance. However, if performing mask induction, avoid nitrous oxide as it may expand in herniated tissue
    - Nasogastric tube is instituted early following intubation to help decompress the stomach
    - Caution should be used with positive pressure ventilation. See ventilation strategies below
- Positioning
    - Supine vs lateral (surgical repair frequently performed via subcostal incision with ipsilateral chest tube placement)
- Maintenance
    - Balanced anesthesia with volatile anesthetic, opioids, and muscle relaxation
    - Total IV if on HFOV or ECMO, or in the NICU
- Hemodynamic/Physiologic goals
    - Avoid pulmonary hypertensive crisis by mitigating catecholamine surges with adequate anesthesia and analgesia, avoiding hypoxia and hypercapnia as much as possible
    - avoid volutrauma and barotrauma
    - Goal MAP = gestational age if newborn, postconceptual age if older or normative data from Zadelhoff et al.

    - Ventilation goals
        - Preductal saturations between 80% and 95%
        - Post-ductal saturation >70%
        - Partial pressure of carbon dioxide ($PaCO_2$) levels between 50 and 70 mmHg
        - Cerebral NIRS may help guide ventilation
            - If $PaCO_2$ is significantly decreased due to overventilation, NIRS will show decreased cerebral perfusion
    - Initial ventilator settings
        - Peak Inspiratory Pressure (PIP) <25 cm H_2O
        - Positive End-Expiratory Pressure (PEEP) of 3–5 cm H_2O,
        - Respiratory Rate (RR) of 40–60 breaths/min to maintain a $PaCO_2$ between 50 and 70 mm Hg
            - *Note: Inability to ventilate with a PIP of 30 cm H_2O and rate of 60 breaths/min should prompt initiation of HFOV if available*
- Surgical Considerations
    - Emerging modalities for surgical repair are being investigated and increasingly practiced. Maintain close communication with the surgeon regarding surgical approach and patient-specific concerns
    - Laparoscopic approach may be used in cases with asmall defect andno pulmonary hypertension
    - If attempts at abdominal closure result in ventilation difficulties, the abdominal muscle layers should not be closed. Definitive abdominal wall closure is delayed for days to weeks
- Emergence/Disposition
    - Patients remain intubated and transported to NICU or Pediatric Intensive Care Unit (PICU). Consider using a transport ventilator with $EtCO_2$ monitoring over manual bagging to avoid hypoventilation and barotrauma.
- Post-op Care
    - IV opioids and sedatives while intubated. Wean as able in NICU.

Case-Specific Complications/Pitfalls

- Reduction of intrathoracic abdominal viscera back into the abdomen may have acute consequences:
    - Decrease venous return leading to impaired cardiac output
    - Increase intrabdominal pressure resulting in abdominal compartment syndrome and decreased thoracic compliance
- Hypoplasia of the affected lung impairs pulmonary vascular development leading to pulmonary hypertension
    - These patients are prone to pulmonary hypertensive crises
- Aggressive ventilation may cause barotrauma on the contralateral lung: consider the presence of a contralateral pneumothorax in case of acute deterioration
- Any combination of the above may precipitate cardiovascular collapse
- Left Ventricular (LV) dysfunction may be seen particularly in left-sided herniations secondary to LV hypoplasia. Consider milrinone to improve lusitropy and reduce LV afterload.

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