INTRAOPERATIVE MANAGEMENT FOR PATIENTS WITH PULMONARY HYPERTENSION UNDERGOING NON-CARDIAC SURGERY

Surgical Specialty:	Other Topics
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Background:

- General Considerations
  - Children with pulmonary hypertension (PH) have a **significantly** increased risk of perioperative morbidity and mortality
    - Anesthetic management for patients with PH requires an in-depth understanding of the pathophysiology of PH and careful planning for perioperative optimization, management, and disposition
    - Potential complications include arrhythmias, hemodynamic instability, right heart failure, myocardial ischemia, respiratory failure, unanticipated ICU admission, CV collapse and death
  - Diagnosis of PH in the pediatric population[1]
    - Defined as a resting mean pulmonary artery pressure (mPAP) >20mmHg in individuals > 3 months of age at sea level
    - Cardiac catheterization (gold standard) with acute vasoreactivity testing with oxygen and nitric oxide
    - Other diagnostic tools: echocardiogram (TTE), cardiac MRI
  - Classifications of PH & common pediatric examples [1]
    - Group 1: Pulmonary arterial hypertension (PAH) - thickened arteries in the lung
      - Idiopathic PAH
      - Heritable PAH
      - Persistent PH of the newborn (PPHN) – syndrome
      - Associated PAH (CHD, HIV, portal hypertension, etc)
    - Group 2: PH caused by left heart disease
      - LV dysfunction (systolic or diastolic)
      - Valvular disease
      - Congenital/acquired left heart inflow/outflow tract obstruction
      - Pulmonary vein stenosis

        - Group 3: PH caused by lung disease and/or hypoxia
            - Bronchopulmonary dysplasia (BPD)
            - Congenital diaphragmatic hernia (CDH)
            - Trisomy 21
            - Sleep-disordered breathing
        - Group 4: PH due to pulmonary artery obstructions
            - Chronic thromboembolic PAH
            - Pulmonary artery obstructions either congenital or acquired after cardiac surgery
        - Group 5: PH due to unclear/multifactorial mechanisms
            - Hematologic disorders (sickle cell disease, myeloproliferative disorders)
            - Systemic disorders (sarcoid, pulmonary histiocytosis)
            - Metabolic disorders (glycogen storage disease, Gaucher disease)
            - Other (tumor, chronic renal failure, complex CHD)
- Treatment Modalities for PH - treatment course must take into consideration the underlying etiology of PH and the presence of PAH[1-3.]
    - Diuretics
    - Oxygen therapy
    - Calcium channel blockers (diltiazem, amlodipine)
    - Phosphodiesterase (PDE-5) inhibitors (sildenafil, tadalafil)
    - Endothelin-receptor antagonists (bosentan, ambrisentan)
    - Prostacyclin analogs and receptor agonists (epoprostenol, treprostinil)
    - Inhaled Nitric Oxide
    - ECMO
    - Surgical/Interventional-Catheter Based Shunts (ie Potts Shunt)
    - Lung transplant

Anesthetic Planning:

- Pre-Anesthetic Evaluation
    - A detailed history and physical exam should be performed with a focus on functional status and identification of any acute comorbidities.
    - Disease progression and the presence of pulmonary arterial remodeling are important considerations for risk stratification.
    - Risk stratification – factors associated with increased perioperative morbidity and mortality[2]
        - Systemic or suprasystemic PAP
        - Presence of syncope
        - Elevated mean RA pressure
        - Depressed RV function
        - Young age (<1 year old)
        - Home oxygen use
        - Requiring multiple anti-pulmonary hypertension medication classes
        - Present or absence of interventional shunts
    - Additional labs/tests to be considered during work-up
        - Hemoglobin and hematocrit
        - Complete metabolic panel (to assess renal and liver function)
        - Arterial blood gas
        - EKG

    - Echocardiogram – tricuspid regurgitation (TR) velocity can be used to estimate RVP; septal flattening can be used as a surrogate if no TR is present; evaluation of shunt directions and velocities (if present); RV/LV function
    - Chest X-ray
    - Most recent cardiac catheterization data
    - Pulmonary function testing
- Discussions/considerations
    - AHA Guidelines: "Elective surgery for patients with pediatric PH should be performed at hospitals with expertise in PH and in consultation with the pediatric PH service and anesthesiologists with experience in the perioperative management of children with PH."[3]
    - Recommend multidisciplinary planning with other specialties involved in the patient's care (i.e. pulmonology and/or cardiology) and the surgeon prior to surgery to ensure consideration of the indications/benefits of surgery as well as the potential risks.
    - Pre-operative goals include; treating comorbidities, optimizing fluid balance, adjusting pulmonary vasodilator management, and optimizing RV function prior to surgery, when possible.
    - Continue all oral, inhaled, or intravenous pulmonary vasodilator medications with a dedicated IV line for intravenous therapies for uninterrupted delivery
    - Continuation of diuretics, anticoagulation, and antihypertensive agents should be specifically discussed with the cardiology and surgical teams.
    - Careful consideration of premedication to alleviate anxiety and associated increased PVR while avoiding oversedation and hypoventilation
- Other considerations and available resources[4]
    - Pediatric cardiac anesthesiologist
    - Monitors as outlined below
    - Inhaled pulmonary vasodilators such as nitric oxide
    - Inodilators such as milrinone
    - Vasopressors such as norepinephrine and vasopressin
    - Inotropes such as epinephrine and dobutamine
    - Consider ECMO standby (blood products available)
    - NICU/PICU/PCICU bed availability

Intraoperative Considerations:

- General Considerations
    - General anesthesia is a common approach utilized for pediatric patients with pulmonary hypertension. Careful consideration must be made to optimize pain management, avoid arrhythmias, optimize oxygenation and ventilation, and avoid hypotension and direct myocardial depression[4].
    - Ensure no air is present in IV lines and syringes
    - Utilize forced air-warming blankets and fluid warmers to avoid hypothermia
    - Monitors to consider
        - ASA standard monitors
        - Consider arterial line for close hemodynamic monitoring, especially in patients with PAH and systemic/supra-systemic RVP
        - Consider a central venous catheter to monitor central venous pressures and for more targeted therapy with vasopressors, vasodilators, and fluids.

        - Consider near-infrared spectroscopy (NIRS) monitoring (cerebral +/- flank), if available.
        - Transesophageal echocardiography (TEE) - largely used to evaluate the right ventricle and to calculate the pulmonary artery pressure and evaluate volume status
        - Mid-esophageal four-chamber view is ideal for measuring the dimensions of the RV
        - A failing RV will shift the septal wall into the LV cavity, causing it to look like the letter "D"
- Surgical Considerations
    - Insufflation during laparoscopic procedures can increase abdominal pressure leading to decreased venous return and/or decreased lung compliance.
    - Exogenous CO_2 used for insufflation may increase pulmonary vascular resistance.
    - Orthopedic procedures utilizing cement may lead to profound hypotension
    - Procedures with the potential for significant blood and/or insensible losses can lead to rapid fluid shifts drastically affecting the loading condition of the RV
- Induction
    - Induction can precipitate a pulmonary hypertensive crisis due to noxious stimuli leading to catecholamine release and PVR spike, hypoxia/hypercarbia during intubation, myocardial and SVR depression from medications, or decreased RV preload due to prolonged NPO times
    - Choose induction technique that will minimize impact on cardiac function and SVR, and blunt sympathetic response to laryngoscopy
    - Careful titration of medications. Ketamine and dexmedetomidine have not been shown to increase PVR [5, 6]
    - The most skilled personnel should perform airway management/laryngoscopy to limit hypercarbia, hypoxemia, or acidosis.
- Hemodynamic/Physiologic goals
    - Overall goals:
        - Avoid hypoxia as it can lead to pulmonary vasoconstriction
        - Avoid hypercarbia, acidosis and, hypothermia which can also increase PVR
        - Avoid high airway pressures
        - Maintain sinus rhythm
        - Optimize RV preload
        - Reduce RV afterload
        - Maintain coronary blood flow
    - Ventilation:
        - Avoid hypoventilation, hypercarbia and hypoxia
        - Avoid excessive PEEP (goal 5-10mmHg)
        - Liberal oxygen use (outside of the neonatal period and when appropriate for surgical procedure)
        - Maintain ETCO2 30-35mmHg
        - Avoid one lung ventilation if possible
- Post-op Care
    - Consider fully monitoring patients in the ICU post-operatively, even after an uneventful surgery, as clinical deterioration can occur rapidly and lead to death.
    - Avoid hypoxia, hypothermia, hypotension, fluid shifts, and noxious stimuli
    - Consider regional blocks, epidurals, and non-opioid medications for pain management to minimize sympathetic activation

Disease-Specific Complications/Pitfalls

- Pulmonary Hypertensive Crisis
    - PH crisis occurs when there is a sudden rise in PVR leading to acute right heart failure. Decreased cardiac output compromises blood flow to the coronary arteries which worsens right ventricular function in a cyclical manner
    - Can be precipitated by hypercarbia/acidosis, hypoxia, hypothermia, noxious stimuli, and systemic hypotension
    - Presentation:
        - May be gradual or acute CV collapse
        - Decreased $ETCO_2$, arterial desaturation, cerebral NIRS desaturation, ECG changes, and hypotension
    - Immediate treatment is critical and should include
        - Administration of 100% FiO2
        - Mild hyperventilation
        - Correction or removal of noxious stimulus – deepen anesthetic depth
        - Correcton of acidosis (sodium bicarbonate)
        - Inotropic support to the RV/LV with epinephrine, calcium
        - Consider vasopressin to support SVR (does not increase PVR)[7]
        - Pulmonary vasodilator therapy (iNO)
        - Consider fluid bolus if hypovolemia is suspected but use caution if significant RV failure is suspected.
        - Consider ECLS (eCPR) in the setting of circulatory collapse
- Laryngospasm in a patient with PH
    - Use special caution when treating suspected laryngospasm in patients with PH.
        - High CPAP can precipitate acute RV failure secondary to an acute increase in PVR and a simultaneous decrease in RV preload.
        - Large doses of sedatives with subsequent drops in SVR can result in myocardial depression due to decreased CPP
        - Prolonged lack of ventilation with associated hypercarbia and hypoxia will further increase PVR.

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