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ANESTHESIA FOR PATIENTS WITH SINGLE VENTRICLE FOR NONCARDIAC SURGERY (INCLUDING LAPAROTOMY)

Surgical Specialty:	Multiple
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Background:

- General Considerations
    - The anesthetic management of children with a single ventricle requires a clear understanding of the following:
        - The concepts of shunting, single ventricle physiology, and intercirculatory mixing
        - Types and dynamics of anatomic shunts
        - Effects of lesions on the development of the pulmonary vascular system
        - Factors that influence pulmonary and systemic vascular resistance
        - Factors responsible for producing myocardial ischemia
    - Patients can often present for non-cardiac surgeries related to their underlying single ventricle physiology, such as laparoscopic gastrostomy for feeding intolerance, PICC or central venous catheter placement for necessary infusions or total parenteral nutrition (TPN), imaging studies, airway evaluations or laparotomy for necrotizing enterocolitis.[1]
    - Elective procedures are best planned to follow the Glenn/hemi-Fontan operation (usually 4-5 months of age) as these patients have more stable physiology than patients who have had the first stage of the single ventricle palliation and overall highest risk.[1]
- Patient Considerations
    - Disease Specific Considerations
        - You need to know the patient's current physiology.
            - Review the patient's current stage of single ventricle palliation and the underlying physiology of that stage.
            - It is essential to know the ratio of pulmonary to systemic blood flow (Qp:Qs) from prior catheterizations and baseline saturations, as these are good measures of shunting.[2]
        - Intraoperative management of Qp:Qs
            - Management of FiO_2
                - Dependent on the stage of repair.
                - Patients with some cardiac lesions are poorly tolerant of higher O_2 saturations than their normal range for significant periods of time. This is particularly true of single ventricle patients who are unrepaired or have had a Stage 1 palliation and who have not yet proceeded to the Glenn/hemi-Fontan palliation.

- Other Modifiable Factors
  - Ventilation can be manipulated to reduce intrathoracic pressures and control $PaCO_2$
  - The patient's acid-base status should be monitored and adjusted as indicated.
  - Volume and systemic vascular resistance (SVR) adjustments can be made as indicated.
- Preoperative intravascular volume status
  - Decreases should be avoided in all single ventricle patients, especially in patients who have undergone Glenn/hemi-Fontan or Fontan palliation.[2]
- Associated Comorbidities/Syndromes
  - There is most often sporadic inheritance.
  - If comorbidities are present, clinicians should prepare for the related anesthetic implications.

Anesthetic Planning:

- Pre-Anesthetic Evaluation
  - It is best practice to review prior echocardiography, ECG for history of arrythmias, and any cardiac catheterizations to understand the patient's specific anatomy and circulation (Qp:Qs) and for overall risk assessment.
    - Stage 1 (Norwood, hybrid Norwood) patients
      - Assess for valvular regurgitation, aortic arch obstruction, ventricular function, and any shunt (modified Blalock–Taussig–Thomas Shunt /Sano, PDA stent with PA bands for hybrid procedure, or any other aortopulmonary shunt) stenoses contribute to the risk of perioperative hemodynamic instability.[2]
    - Stage 2 (Glenn) and 3 (Fontan)
      - Assessment of intravascular volume status is important.
      - Ventricular systolic and diastolic function should be assessed prior to anesthesia. Single ventricle patients with ventricular dysfunction are at high risk of significant anesthetic morbidity and mortality. These patients may also have liver or kidney dysfunction.[3]
      - Any evidence of elevated pulmonary vascular resistance (PVR) by echocardiography or diagnostic catheterization makes these patients higher risk for anesthesia. Pulmonary vasodilators should be continued throughout the perioperative period.[4,5]
      - Evidence of protein losing syndromes (protein losing enteropathy, plastic bronchitis, ascites, and pleural effusions) are often harbingers of single ventricle palliation failure. Patients with protein losing syndromes also may have liver dysfunction.[4]
      - Patients with Fontan circulation, especially those who have had a lateral tunnel Fontan, are at risk for dysrhythmias.[4]
      - Post-Fontan patients are at higher risk of thrombosis at baseline than the general population.[4]

- Determine the patient's baseline oxygen saturations.
  - Normal oxygen saturations for pre-palliation and Stage 1 patients are 75-85% and for Stage 2 patients is >80%. Stage 3 patients may have baseline SpO_2 of as low as 85% if the Fontan has been fenestrated or if there is significant collateral circulation and as high as 99% if not fenestrated.
- Review preoperative laboratory tests, including hemoglobin, hematocrit, liver function, and electrolytes.
- Discussions to have with surgeon/family
  - It is vital to know a patient's Code Status.
  - ECMO candidacy should be established, especially prior to high-risk surgeries.
  - An ICU bed must be available for patients who are pre-Stage 1 or post-Stage 1 and prior to Glenn/hemi-Fontan.
  - If the procedure is elective, it is best to wait until after Glenn/hemi-Fontan surgery.

- Specific or Unique Room Set-Up Requirements
  - Airway
    - Standard airway equipment is appropriate but important to review prior anesthetics and examine patients for evaluation of a potentially difficult airway.
      - Important to note any stridor on exam and review history for potential vocal cord dysfunction, subglottic stenosis, or tracheolaryngomalacia related to prior intubation or related comorbidities/ syndromes.
    - Stage 1 patients tend to have limited physiologic reserve and are poorly tolerant of repeated attempts to secure the airway.
  - Drugs/Infusions
    - If an invasive procedure, one should plan to have emergency vasoactive medications for inotropic support and to counteract shunting and fall in systemic vascular resistance (SVR). Vasopressin can be especially effective in Fontan patients with normal ventricular function as it improves the drop in SVR caused by anesthesia without increasing PVR.
    - Single ventricle patients do poorly with intravascular volume depletion, so pre-hydration should be considered and intraoperatively, balanced crystalloid administration should be used. Use caution in patients with diastolic ventricular dysfunction.
    - Anesthesia providers should treat inhaled oxygen as a drug that increases Qp:Qs and, by virtue, competitive flow, which can cause systemic hypoperfusion in Stage 1 patients placing coronary artery perfusion at risk.
    - Patients with any evidence of pulmonary hypertension should have their anti-pulmonary hypertensive medications continued throughout the perioperative period, and the anesthesiologist should be prepared to treat pulmonary hypertension with medications such as inhaled nitric oxide (iNO).
  - Monitors
    - Standard ASA monitors, five-lead ECG, and arterial BP monitor are indicated for major surgeries or any surgery requiring insufflation (laparoscopic or thoracoscopic) where hemodynamic changes are likely to occur.
      - Clinicians can consider the use of cerebral and somatic near-infrared spectroscopy (NIRS) to monitor organ perfusion in major surgery.

- Blood Availability
  - Should be available for major surgeries with a high likelihood of significant blood loss.
  - Cyanotic patients compensate with higher baseline hematocrits; thus, they have less tolerance to anemia.
  - It is particularly important for post-Fontan patients that are sensitive to hypovolemia.[2]
- PICU Bed Availability
  - A bed should always be available, especially for major surgery, as postoperative ventilatory and hemodynamic support may be necessary.
- Other Indicated Resources
  - Patients with evidence of a protein-losing syndrome or ventricular dysfunction should be evaluated by a general cardiologist or heart failure specialist prior to any non-emergent procedure.

Intraoperative Considerations:

- General
  - Anesthetic and Physiologic Implications of the Surgical Procedure
    - It depends on the type and invasiveness of the surgery.
      - Intrathoracic and intrabdominal procedures that may affect ventilation and hemodynamics are higher risk for any single ventricle patients during any phase of the single ventricle palliation pathway.[6]
      - Factors that increase PVR, such as acidosis, atelectasis, hypoxia, severe hypercapnia, and hypothermia, must be avoided.[6]
- Induction
  - Mask induction can be used during any stage of repair for non-cardiac surgery in patients with normal ventricular function, though intravenous (IV) access should be quickly established to allow for volume and vasoactive administration, if needed. Patients post-Stage 1 are especially unlikely to tolerate prolonged, high concentrations of inhaled anesthetic, especially in combination of high FiO_2 and hyperventilation.
  - IV medications with more stable hemodynamics, such as fentanyl and ketamine, are preferrable to prevent significant decreases in SVR and changes in the SVR/PVR ratio.
  - Prior to Stage 1 repair and after Stage 1 procedure, it is important to minimize FiO_2 to reduce pulmonary over circulation and hypotension. As the patient grows after Stage 1 repair or if shunt stenosis occurs, it may be necessary to titrate FiO_2 to maintain $SpO_2 > 75\%$.
- Positioning
  - Anesthesia providers should ensure appropriate padding of pressure points as needed based on the type of surgery.
  - There are no specific, special precautions.
- Maintenance
  - Volatile vs. TIVA
    - In pre- and post- Stage 1 patients, a common strategy is to use IV opioids or ketamine as the predominant anesthetic—either alone or in combination with low dose propofol or inhaled volatile agents—to maintain SVR and maintain preoperative Qp:Qs .
    - In Glenn/hemi-Fontan and Fontan patients, both can be utilized, though changes in SVR will require treatment to maintain preload.

- Use of neuromuscular blockade in patients during the maintenance phase is more common in patients with congenital cardiac disease than in the general pediatric population. It allows volatile agents and propofol—which lower SVR—to be minimized, resulting in improved hemodynamics while still maintaining adequate anesthetic depth.

- Hemodynamic/Physiologic goals
  - It is important to maintain a balanced circulation in Stage 1 patients with close vigilance for adequate coronary artery perfusion pressure.
  - Clinicians should maintain adequate pulmonary circulation and prevent increases in PVR in Glenn and Fontan patients.
  - Avoid hyperventilation/ hypocapnia in Glenn/hemi-Fontan patients as this leads to decreased cerebral perfusion and venous return to the pulmonary circulation. Hypercapnia increases PVR, which also decreases venous return to the pulmonary circulation. Optimal $PaCO_2$ to balance cerebral circulation and venous return (cerebro-pulmonary interaction) is approximately 45 mmHg.
  - Glenn/hemi-Fontan and Fontan patients will do better with spontaneous negative pressure than positive pressure ventilation due to cardio-pulmonary interactions, though healthy patients will likely tolerate positive ventilation if SVR and preload are maintained.[5]
    - Positive pressure ventilation should attempt to keep mean airway pressure as low as possible while preventing atelectasis. PEEP of 5 with higher tidal volume and lower respiratory rate with prolonged I:E ratio is a typical ventilation strategy. This type of strategy is used because pulmonary blood flow predominantly occurs during low intrathoracic pressure (such as exhalation during positive pressure ventilation), and by extending the expiratory time, the mean airway pressure is reduced, and there is increased time for venous return to the pulmonary circulation.
    - It is important to monitor end-tidal CO_2 or arterial CO_2 in spontaneously breathing patients during anesthesia, as hypoventilation will cause hypercapnia and increase PVR.
  - Erythrocyte transfusion thresholds are dependent on patient age, underlying physiology, stage of palliation, and baseline oxygen saturations. Patients at any stage with lower baseline saturations will require higher hematocrits than those with higher baseline saturations. For asymptomatic children with cyanotic heart disease, a transfusion threshold of 9.0 g/dL has been proposed.[6] Intraoperatively, factors such as rate of blood loss, hemodynamics, saturations, urine output, serum lactate, and cerebral and somatic NIRS should be considered along with the patient's hemoglobin level to decide whether erythrocyte transfusion is warranted.
- Surgical Considerations (such as neuromonitoring, muscle relaxation, anticipated blood loss)
  - Any surgery that requires insufflation (such as laparoscopic and thoracoscopic) will have an exaggerated effect on the physiology of single ventricle patients at any phase of the single ventricle palliation due to effects on venous return and PVR. These surgeries should be considered high risk in these patients and open alternatives to laparoscopic and thoracoscopic procedures should be discussed with the surgeon. Invasive arterial access should be considered for these procedures.[7,8]
    - One of the most common non-cardiac surgeries performed in single ventricle patients who have received a Stage 1 palliation is laparoscopic gastrostomy tube placement. A common intraoperative complication in these patients

during laparoscopic surgery is acute hypoxemia. Pneumoperitoneum decreases cardiac index by increasing intrathoracic and mean airway pressure, thereby decreasing preload and increasing pulmonary capillary wedge pressure.[8] The CO_2 used to insufflate the abdomen also increases PVR if ventilation is insufficient to maintain normocarbia and can rarely cause CO_2 embolism.[8] These factors can increase PVR and decrease pulmonary perfusion. If acute hypoxemia occurs, the surgeons should immediately stop CO_2 insufflation, adequate ventilation and intravascular volume must be ensured, and it may be necessary to increase systemic blood pressure with vasoactive medications or increase FiO_2 to ensure adequate shunt flow and pulmonary perfusion. If it is decided that it is safe to attempt to continue the procedure, the surgeon should insufflate to 10 mmHg or less of intraabdominal CO_2 pressure and should minimize the duration of insufflation; if the patient does not tolerate these measures, an open approach to gastrostomy tube placement should be considered.

- Emergence/Disposition
  - Providers must ensure adequate analgesia to prevent splinting and hypoventilation that may lead to hypercapnia and hypoxemia post-extubation.
- Post-operative Care
  - If requiring ventilatory or hemodynamic support, the patient should be transferred to ICU until stable.
  - Many centers recover some or all Stage 1 patients in the ICU following general anesthesia. This practice is institution dependent, however.

Case Specific Complications/Pitfalls

- Any ventricular dysfunction, arrhythmia, or ischemia will be poorly tolerated by single ventricle patients.
- Maintain intravascular volume and SVR but be cautious of excess volume administration in patients with diastolic ventricular dysfunction and large increases in SVR in patients with systolic ventricular dysfunction.
- Increases in PVR must be avoided in these patients; maintain normothermia, normoxemia, neutral pH. Vasopressin is an excellent choice to maintain SVR without increasing PVR in patients with normal ventricular function.
- Insufflation during laparoscopic or thoracoscopic procedures can cause significant derangements in physiology in these patients and is a time when the anesthesiologist must be vigilant.

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