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NORWOOD PROCEDURE

Surgical Specialty:	Cardiac
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Background:

- General Considerations
  - The first procedure in staged palliation for hypoplastic left heart syndrome and variants
  - Typically performed in the first week of life
  - The goal is to provide systemic circulation from the right ventricle as well as a source of pulmonary blood flow
    - Two variations for providing pulmonary blood flow[1]:
      - Systemic to Pulmonary:
        - Modified Blalock-Thomas-Taussig (mBTT) shunt: Shunt between innominate artery and proximal branch PA (most commonly right-sided)
        - Central Shunt: Shunt between ascending aorta and main pulmonary artery
      - RV to Pulmonary (e.g., Sano)
    - To provide systemic circulation, a neo-aorta is formed from the proximal main PA and reconstructed aortic arch
  - Alternatively, or prior to the Norwood procedure, patients may undergo a hybrid procedure where pulmonary blood flow continues from the RV, but constricting bands are placed on branch PAs to control pulmonary flow. Systemic circulation is provided via a stented open ductus arteriosis or by continuing intravenous prostaglandin E1 (PGE1) infusion to keep the ductus arteriosus patent. Balloon atrial septostomy is done if there is a restrictive ASD/PFO.
- Patient Considerations
  - Usually pre-natally diagnosed but may be diagnosed post-natally
  - Ductal dependent lesion so will be started on intravenous PGE1 immediately after birth
  - Clinical status depends on the presence of patent atrial communication. Symptoms can progress rapidly, especially in the absence of an atrial communication for left atrial egress. Surgical or catheter-based atrial septostomy may be indicated in these patients.
  - Extremely high mortality rate (~95%) if left untreated in first few weeks of life[2]
- Associated Comorbidities/Syndromes
    - Can be associated with genetic syndromes but this is the minority of cases
    - Greater chance of being born small for gestational age or prematurely due to poor fetal growth

Anesthetic Planning:

- Pre-Anesthetic Evaluation
  - Labs/tests indicated during work-up

        - Echocardiography to evaluate the size of atrial level communication, degree of tricuspid regurgitation, RV function, and size of ascending aorta
        - ECG – Expect to see features of RV predominance
        - Type & Screen, CBC, BMP, Coagulation labs
    - Discussions to have with surgeon/family
        - Code Status
        - ECMO Candidacy
- Specific or Unique Room Set-up Requirements
    - Airway
        - Some institutions perform nasotracheal intubation since there is a high likelihood that the patient will remain intubated post-operatively. This helps with oral readiness and eventual oral feeding.[3]
    - Access
        - 2-3 large peripheral IVs for large-volume transfusion and rapid resuscitation
        - Arterial line for BP monitoring and arterial blood gas collection
            - Using existing umbilical artery catheter or placing one may be considered. If an upper extremity arterial line is needed and the patient will be undergoing a mBTT shunt, consider placing it on the side opposite the planned shunt
        - Central line for transfusion of inotropic and vasopressor medication
            - An umbilical venous catheter (UVC) placed pre-operatively or a direct right atrial (RA) line placed intraoperatively by the surgeon may be considered. Some institutions avoid IJ access because SVC patency is essential for future bidirectional cavopulmonary anastomosis, but this is institution dependent.
    - Drugs/Infusions
        - Induction
            - Several options but the goal is to keep Qp:Qs as close to 1 as possible. Institutional practices may vary
                - Ketamine 1-2 mg/kg boluses
                - Fentanyl 3-5 mcg/kg boluses
                - Inhalational induction may cause a drop in SVR and alteration of Qp:Qs so be prepared with pharmacologic support.
        - Infusions for post-CPB:
            - Epinephrine
                - Begin infusion at 0.02-0.1 mcg/kg/min and titrate to effect
            - Dopamine
                - Begin infusion at 2-5 mcg/kg/min
            - Milrinone
                - Loading dose of 100 mcg/kg/min then start infusion at 0.2-0.7 mcg/kg/min[4]
            - Vasopressin for vasoplegia
                - 0.0003 U/kg/min – 0.0012 U/kg/min

    - Monitors
        - Standard ASA monitors: pulse-oximetry, EKG, non-invasive blood pressure, capnography, temperature (recommend two separate locations for temperature when cooling to deep hypothermia)
        - Invasive monitoring: invasive arterial pressure, invasive central venous pressure
        - Neuromonitoring: NIRS, EEG
    - Blood Availability
        - CPB prime
            - Whole blood preferred over pRBC[5]
            - If whole blood is not available, prime with a combination of irradiated pRBC and fresh-frozen plasma to avoid dilution of clotting factors
        - Post-CPB transfusion

        - Blood products: cryoprecipitate, platelets, FFP, possibly Factor VII
        - Consider thromboelastogram for targeted management of blood product administration and hemostasis[6]
    - ICU Bed Availability
    - Other Indicated Resources ECMO standby

Intraoperative Considerations:

- Induction
    - Ketamine 2 mg/kg for induction due to stable hemodynamic profile. Avoid induction agents that will reduce SVR
    - Glycopyrrolate 10 mcg/kg is useful for preventing an increase in oral secretions from ketamine
    - Limit pre-oxygenation, and return to room air as soon as the airway is secured to avoid decreasing PVR
- Maintenance
    - Volatile agents are appropriate for maintenance but need to be titrated to avoid decreases in SVR. Other adjuncts such as fentanyl, midazolam, dexmedetomidine, and ketamine can be used to allow a lower MAC of volatile agent. Nondepolarizing neuromuscular blockers dosed throughout surgery
    - Antifibrinolytics. Tranexamic acid or aminocaproic acid infusion to mitigate bleeding
- Hemodynamic/Physiologic goals while coming off cardiopulmonary bypass
    - Maintain Qp:Qs as close to 1:1 by aiming for an oxygen saturation between 75%-85%. Avoid large changes in SVR and PVR.
    - An arterial pO_2 less than 35 mmHg in the immediate post-op period should trigger evaluation and concurrent management of hypoxemia. These would include:
        - Ensuring adequate lung expansion and ventilation
        - Assessment of patency of mBTT shunt/Sano conduit and branch pulmonary arteries using intraoperative echocardiogram or cardiac catheterization
    - Closely maintaining central venous pressures within physiologic limits during blood product resuscitation in the post-bypass period in order to not over-distend the myocardium
    - Arrhythmia during the post-bypass period is most likely related to coronary insufficiency
- Surgical Considerations (such as neuromonitoring, muscle relaxation, anticipated blood loss)
    - Cerebral oximetry is useful for the management of cardiopulmonary bypass (CPB) and antegrade cerebral perfusion (ACP)
    - If ACP is utilized for arch reconstruction, a right upper extremity arterial line is preferred
- Emergence/Disposition
    - Most centers elect to keep these patients intubated and paralyzed for a few days post-op. Some surgeons may leave the chest open.
- Post-op Care
    - Hemodynamic goals and management depend on the procedure (Sano vs BT shunt)
        - BT shunt – avoid decreases in PVR (avoid 100% FiO_2, hyperventilation, etc.)
        - Sano – typically better hemodynamic stability in the immediate post-operative period

Case-Specific Complications/Pitfalls.

- Pulmonary over-circulation prior to CPB will cause myocardial ischemia. Baseline SpO_2 should be 75%-85%, and saturations over this may indicate pulmonary over-circulation and poor systemic perfusion. Crucial to keep Qp:Qs near baseline during the pre-bypass period.

References

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