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FONTAN PROCEDURE

Surgical Specialty:	Cardiac Surgery
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Background:

- General Considerations
    - The Fontan procedure is the third and final stage of single ventricle palliation, connecting the inferior vena cava (IVC) to the pulmonary artery (PA), which completes the separation of pulmonary and systemic circulations.
        - Ventricular preload is dependent on passive flow across the pulmonary vascular bed.
            - Circulation is a series circuit: Systemic veins → lungs → pulmonary veins → atria →ventricle → body
    - Multiple anatomic variations exist.
        - Lateral tunnel Fontan- an intra-atrial baffle is placed in the atrium to direct blood flow from the IVC to the right pulmonary artery (RPA).
            - The benefit of this procedure is that it allows for growth as the child grows but may result in an increased incidence of arrhythmias due to some incorporation of the atrium.
        - Extracardiac Fontan- the IVC is separated from the atrium, and a conduit is utilized to create an anastomosis to the RPA, allowing the atrium to be bypassed completely.
            - The benefit here is the decreased incidence of arrhythmias since the atrium is excluded, but the synthetic conduit does not have growth potential and therefore must be large enough to accommodate increased IVC blood flow over time.
        - Fenestrated Fontan- a surgical opening is created between the intra-atrial baffle or conduit and the atrium, which allows for "pop off" right-to-left shunting in the event of increased pulmonary vascular resistance (PVR). This allows cardiac output to be maintained but will result in decreased oxygen saturation and increases the risk for paradoxical emboli.[1]
- Patient Considerations
    - The primary benefit of Fontan completion is the subsequent improvement in oxygenation. The age at Fontan completion has varied across surgical periods and among institutions from early Fontan at 1 year old to 5 years old.
    - Immediately performing the Fontan operation for single ventricle palliation is not recommended because the acute decrease in ventricular volume coupled with diastolic dysfunction in infancy is poorly tolerated in single ventricle physiology. Therefore, a second-stage operation (superior cavopulmonary anastomosis, also

termed a bidirectional Glenn shunt) is typically performed around 3-6 months of age. By performing a superior cavopulmonary anastomosis before the Fontan procedure, the volume reduction in the heart is smaller, and there is time to allow for ventricular remodeling. However, there is most likely an age at which progressive desaturation necessitates Fontan completion.

- Although the Fontan surgical approach results in stable physiology, there are consequent circulatory disadvantages, including elevated central venous pressures, reduced ventricular filling, and overall decreased cardiac output. Elevation of venous pressures exerts deleterious effects upon renal, intestinal (protein-losing enteropathy), and hepatic function. The drawbacks of long-standing Fontan circulation support delaying the total time that a patient is exposed to the imperfect physiology imposed by the total cavopulmonary connection.
- Pre-operative oxygen saturations typically range from 75-85%.
- Associated Comorbidities/Syndromes
    1. Pulmonary arteriovenous malformations or venovenous collaterals may form. This abnormal angiogenesis is thought to be due to a lack of hepatic effluent perfusing the pulmonary arteries. Instead, IVC blood (with hepatic factor) returns to the systemic ventricle, bypassing the lungs. This can result in worsening cyanosis, hypoxemia, and activity tolerance preoperatively. These typically regress following Fontan completion.[2]
    - The primary congenital cardiac abnormality may vary in single ventricle palliation (i.e., hypoplastic left heart syndrome, tricuspid atresia, unbalanced atrioventricular canal defect, heterotaxy). Therefore, a knowledge of the cardiac defect at birth is essential to understanding the patient's anatomy.
    - http://www.pted.org/?id=list#1

Anesthetic Planning:

- Pre-Anesthetic Evaluation
    - Review prior anesthetic history paying attention to previous airway management.
    - Review of cardiac history, including an echocardiogram (with particular attention to ventricular function and atrioventricular valve competence), cardiac catheterization (with particular attention to pulmonary artery pressure and PVR), cardiac MRI, and EKG.
        - Pulmonary artery pressure (PAP) should be low (ideally mPAP<15mm Hg), and PVR should be <4 Wood units/m^2. [3]
            - Patients with higher mPAP or PVR should be considered at higher perioperative risk, as this would represent increased resistance to passive pulmonary blood flow.
    - Non-cardiac history should be thoroughly reviewed. Recent upper and lower respiratory tract infections can have significant hemodynamic implications by increasing PVR.
    - Preoperatively, patients may be on several medications, including antiarrhythmic agents, diuretics, anticoagulants, anti-platelet agents, and antihypertensives.
    - Hematocrit is typically 40-45% as a compensatory mechanism from persistent cyanosis.
    - Discussions to have with surgeon/family
        - Code status
        - ECMO candidacy

- Specific or Unique Room Set-Up Requirements
  - Airway
    - Standard oral endotracheal tube
  - Drugs/Infusions
    - Consider oral premedication
    - Anti-fibrinolytic (Tranexamic acid, amicar) bolus and infusion
    - Heparin, protamine
    - Calcium and emergency medications
    - Inotropic support should be readily available following cardiopulmonary bypass.
      - Commonly used inotropes include dopamine, epinephrine and milrinone.
      - Afterload reduction (if necessary) can be achieved with nicardipine. Milrinone can also be helpful in this regard.
    - Consider a sedative (dexmedetomidine) and narcotic infusion for transport to ICU.
  - Monitors
    - Standard ASA monitors prior to induction
    - Post-induction
      - Arterial line- if the patient had a prior Blalock-Taussig shunt, the ipsilateral subclavian artery may be compromised.
      - Central line- location may vary dependent on institutional preference (femoral, internal jugular, intra-cardiac).
      - Multisite near-infrared spectroscopy (NIRS) to detect low system oxygen delivery (DO_2)
      - Trans-esophageal echocardiography (TEE)
  - Blood Availability
    - These patients are usually at their second or third sternotomy. Obtain the best IV access possible. Have a rapid infusion system available for fluids and blood transfusion.
      - Consider lower extremity IV for redo sternotomy, as the IVC is the most direct route to the atrium.
  - Cardiac ICU bed
  - Other Indicated Resources: Perfusion, cardiology for intra-operative TEE.

Intraoperative Considerations:

- General
  - There is a higher risk of bleeding with repeat sternotomy. Blood products should be in the OR and checked prior to incision.
  - External defibrillator pads should be placed, given repeat sternotomy.
  - Upon completion, blood flow to the pulmonary circulation is an entirely passive process. Therefore, it is essential to optimize hemodynamics post-cardiopulmonary bypass to prevent a low cardiac output state.
    - Blood flow is dependent on the transpulmonary gradient (TPG)
      - TPG =mean pulmonary artery pressure minus common atrial pressure
      - Target TPG is <10mm Hg
    - SVC and IVC pressures will increase following surgery and are correlates of the mean pulmonary arterial pressure (typically 12-16mm Hg).
- Induction

        - Either an intravenous or inhaled induction is appropriate, with primary hemodynamic goals of maintaining mean arterial pressure (MAP) and minimizing increases in PVR.
            - Inhalational induction may be slowed due to right-to-left shunting.
- Positioning
    - Supine, shoulder roll
- Maintenance
    - Volatile anesthetic, drugs as above
        - Analgesic strategy should target facilitating early extubation
- Hemodynamic/Physiologic goals
    - Pre-cardiopulmonary bypass
        - Alterations in cerebral blood flow (CBF) and PVR (hypoxia, hypercarbia, acidosis, hypothermia, pain stimuli) will significantly impact oxygen saturation.
    - Post-cardiopulmonary bypass
        - Maintenance of adequate preload is of utmost importance, paying careful attention to bleeding.
            - Bleeding post-bypass is typically not a major issue, but pRBC, FFP, and platelets should be readily available.
            - Thromboelastography (TEG) or rotational thromboelastrometry (ROTEM®) may be useful to assess coagulation status.
        - Perform maneuvers to reduce V/Q mismatch
            - Suction endotracheal tube
            - Utilize low PEEP (4-5cm H_2O) to prevent atelectasis; too high PEEP will impair venous return and thus decrease pulmonary blood flow.
        - Continue to optimize conditions to minimize PVR.
            - Inhaled nitric oxide is rarely effective in this population.
            - Maintain mechanical ventilation at a point that avoids hypo or hyperventilation. Maintain low mean airway pressures and return to spontaneous ventilation when feasible.
            - Milrinone is an effective pharmacologic agent, given its inotropic and inodilator properties.
        - Consider shortening inspiratory time to allow greater time in the respiratory cycle for passive pulmonary blood flow.
        - Maintain sinus rhythm, institute atrioventricular (AV) pacing if necessary
            - Tachyarrhythmias are poorly tolerated because it impairs pulmonary venous blood flow, which is most brisk during diastole.
        - Inotropic support may be necessary due to ventricular dysfunction following cardiopulmonary bypass. As stated earlier, epinephrine and milrinone are most commonly used. [1]
- Surgical Considerations (such as neuromonitoring, muscle relaxation, anticipated blood loss)
    - Redo sternotomy incurs a higher risk of massive hemorrhage.
    - Cannulation strategy (bicaval vs. single) is usually planned by the surgeon and depends on the surgical approach. Some may prefer deep hypothermic circulatory arrest.
    - If there is pulmonary artery (PA) stenosis, pulmonary arterioplasty may be performed to ensure adequate pulmonary blood flow.
    - Surgically placed atrial line can be used to measure the atrial pressure, which can be used to monitor the transpulmonary gradient in the perioperative period.
    - Intra-operative TEE is used to assess ventricular function, AV valve competence, ensure non-obstructed pulmonary venous return, and exclude outflow

tract obstruction.

- Emergence/Disposition
  - Early extubation should be considered, assuming hemodynamic stability and adequate oxygenation, and spontaneous ventilation. Positive pressure ventilation can decrease flow through the cavopulmonary circuit and pulmonary venous return.
  - If a fenestrated Fontan is performed, expect oxygen saturations of 88-95% in the early postoperative period.
  - If the Fontan is not fenestrated, oxygen saturations may still be less than 100% due to veno-venous collaterals.
- Post-op Care
  - Pain control- intravenous or single injection caudally administered epidural opioids are effective methods of pain control.
  - Vigilant assessment of intravascular volume and replace any ongoing bleeding.
  - Systolic dysfunction is rare following surgery, but potential etiologies could be inadequate myocardial preservation, coronary perfusion, or a metabolic process.
  - Tachyarrhythmias are rare, though they can have significant hemodynamic effects when they do occur.
    - A-V pacing may be beneficial for any junctional rhythm that results in hemodynamic consequences
  - Patients are often initiated on anti-platelet therapy, secondary to the risk for stasis with passive venous return

Case-Specific Complications/Pitfalls

- Low cardiac output can be secondary to:
  - Hypovolemia
  - Low pulmonary blood flow (Fontan obstruction, PA obstruction, pulmonary vein obstruction), resulting in inadequate preload.
    - Saturation may be normal (if not fenestrated, and there is no other physiologic right to left shunting)
    - Common signs include hypotension, tachycardia, poor distal perfusion, and acidosis
    - mPAP will be elevated, and common atrial pressure will be low
    - Treatment: volume replacement, decrease PVR, consider catheterization to create a fenestration
  - Ventricular dysfunction
    - mPAP and common atrial pressures are elevated
    - Pulmonary edema may develop and result in hypoxemia, which is further exacerbated if a fenestration is present.
    - Treatment: judicious increase in PEEP, inotropic support with afterload reduction, consider takedown to superior cavopulmonary anastomosis. [4]
- Arrhythmias, as described above.
- Pleural effusion/chylothorax- secondary to impaired lymphatic drainage due to the high central venous pressure.
- Persistent hypoxemia due to persistent blood flow across the fenestration or the presence of collateral circulation.

References

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