SURGICAL VENTRICULAR SEPTAL DEFECT REPAIR

Surgical Specialty:	Cardiothoracic Surgery
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Background:

- General Considerations
  - A Ventricular Septal Defect (VSD) is the most common congenital defect in children. It constitutes more than 20% of CHD with an occurrence of 1.5 to 3.5 of 1000 live births.[1]
  - A VSD is an opening in the septum that allows communication between the right and left ventricles.
  - VSDs are classified according to their location in the septum, with many classification systems established.
  - A significant proportion of VSDs are small and will decrease in size or close spontaneously (30-40%) by the first few years of life.[2]
  - Surgical repair during infancy warrants patient-specific indications[3-4]:
    - Heart failure that is refractory to conservative medical management
    - Upper respiratory infections (URIs) beginning at 3 - 12 weeks of age
    - Failure to thrive
    - Pulmonary artery pressure above 60% of left ventricular pressure by 6 months of age
    - Aortic regurgitation with heart failure
  - Large VSDs are corrected early in childhood, and complete surgical repair restores cardiac and pulmonary function over the long term.
  - Older patients with severe pulmonary venous occlusive disease (PVOD) (PVR 6 to 9 Wood units/m^2) or Eisenmenger syndrome are not good candidates for surgical repair; however criteria are flexible[4]
    - Mortality is higher in older children.
    - VSD closure eliminates the right-to-left decompressive shunt, forcing all RV output through a fixed, high-resistance pulmonary circuit. This leads to a mismatch between increased right ventricular afterload and limited contractile reserve, often resulting in acute right heart failure.
    - Cardiac catheterization allows measurement of PVR to distinguish normal to slightly elevated PVR from substantially elevated PVR.[2]

- Patient Considerations
  - Disease Specific Considerations
    - The degree of physiologic compromise is determined by the degree of shunting
    - Two key determinants dictate the amount of shunting:

(1) The size of the VSD
(2) The dynamics between pulmonary and systemic vascular resistances

- Small VSDs (restrictive) have a large pressure gradient, limiting the amount of shunting. Patients are often asymptomatic.
- Large VSDs (nonrestrictive) have a small pressure gradient, allowing the relative pulmonary and systemic vascular resistance to determine the direction and degree of shunting.
- At birth, the high pulmonary vascular resistance (PVR) impedes left-to-right shunting.
- After birth, the decrease in PVR and physiologic anemia promotes left-to-right (L-R) shunting.
- L-R shunting increases pulmonary blood flow and triggers a sequence of events:[4]
    - Pulmonary vascular congestion
    - Decrease in pulmonary compliance
    - Airway compromise (increased work of breathing and airway resistance)
    - Myocardial stress. Chronic left-to-right shunting leads to both left and right ventricular myocardial stress. The left ventricle experiences volume overload, resulting in eccentric hypertrophy and increased wall tension, while the right ventricle undergoes pressure and volume loading due to pulmonary overcirculation. This biventricular strain increases myocardial oxygen consumption.
    - Pulmonary hypertension
    - Eisenmenger syndrome (reversal of the shunt)
- Congestive symptoms ensue when a ratio of pulmonary blood flow to systemic blood flow is greater than or equal to 1.5, or Qp:Qs > 1.5 [4]
- Surgery is indicated once medical therapy fails to control the congestive symptoms.[2]
- Pulmonary hypertension is reversible early on, but as PVOD develops, it becomes fixed.
- The progression to Eisenmenger's Syndrome is measured in years for a VSD rather than the decades expected for an ASD.

- Associated Comorbidities/Syndromes
    - Emphasis on comorbidities with significant anesthetic implications
        - VSD can be an isolated finding, but in 50% of cases, additional congenital cardiac anomalies may be found:[4]
            - Inlet VSDs are most often associated with AV canal defects.
            - In malalignment VSD, there is malalignment of the infundibular septum that separates the ventricular outflow tracts,
                - Anterior malalignment leads to RVOT obstruction.
                - Posterior malalignment leads to left ventricular outflow tract (LVOT) obstruction. Tetralogy of Fallot is the classical example.
        - VSDs can be associated with genetic syndromes [2]
            - Trisomy 13, 18, and 21 are the most common

Anesthetic Planning:

- Pre-Anesthetic Evaluation
    - Preoperative assessment to determine the extent of systemic circulatory compromise
    - Children who are asymptomatic without anticipated surgical complications can be considered for fast tracking, aiming at an early extubation[1]
    - System-based approach[3]
        - Cardiovascular
            - Detailed history of symptoms of CHF
            - Physical exam
                - Loud holosystolic murmur and thrill on auscultation
                - Look for signs of tachycardia, diminished pulses, decreased peripheral perfusion, mottling, cyanosis
                - Tachypnea, grunting, flaring, retractions
                - Hepatomegaly due to hepatic congestion
            - Diagnostic tests
                - CXR may show pulmonary congestion
                - Echocardiogram
                - Cardiac catheterization to diagnose pulmonary HTN
        - Respiratory
            - Airway examination
            - Patients with CHF have frequent URIs, and history should include questions about recent symptoms of URI, and physical exam should include auscultation of the chest determine crackles, wheezing, or rhonchi.
        - Hematology
            - Hct
                - L to R shunt leads to anemia
                - R to L shunt (Eisenmenger syndrome) leads to polycythemia
        - Nutrition
            - Poor feeding, poor growth, failure to thrive
        - Musculoskeletal
            - Signs of Eisenmenger syndrome include cyanosis and clubbing of the digits
    - Preoperative Optimization[3]
        - Infants with heart failure are medically treated for their CHF with digoxin and furosemide. ACE inhibitors are controversial due to their potential to cause hemodynamic instability, especially during anesthesia induction and cardiopulmonary bypass.
            - Management of CHF may require aggressive diuresis to the point that the infant is intravascularly volume-depleted and may become hemodynamically compromised with the induction of anesthesia and initiation of positive pressure ventilation.
            - A clue to the intensity of diuresis preoperatively is the serum bicarbonate, as furosemide causes metabolic alkalosis.
        - URIs are recurrent, and delay of surgery may not be possible
    - Discussions to have with the surgeon/family
        - Benefits and risks of the procedure, as well as alternatives to surgical treatment
        - Code Status

- ECMO Candidacy

- Syndrome-specific or Unique Room Set-Up Requirements
  - Airway
    - Standard ETT
    - Associated syndrome-specific considerations
      - For example, airway considerations for Down syndrome: Macroglossia, tonsillar/adenoidal hypertrophy, micrognathia, short neck, OSA, cervical instability, small trachea
  - Drugs/Infusions
    - Inhalation anesthesia
      - Sevoflurane
      - N_2O carries the risk of paradoxical air embolization and is therefore avoided by some practitioners
    - Emergency drugs for patients < 10kg
      - Epinephrine diluted to 100mcg/ml,10mcg/ml and 1 mcg/ml
      - Phenylephrine 40mcg/ml in TB syringe
      - Atropine 20 mcg/kg dosing
      - Succinylcholine 2 mg/kg for children
    - Muscle relaxant
      - Rocuronium
    - Opioids
      - Fentanyl
    - Dexmedetomidine
      - Typical dosing of 0.2-0.4mcg/kg/hr
    - Antibiotics
      - Cefazolin 30mg/kg q3h
        - Neonates <30 days old dose at 25mg/kg
      - Vancomycin 15mg/kg given over 1 hour for Cephalosporin or severe PCN allergy.
      - Cefazolin and Vancomycin 15mg/kg over 1 hr for MRSA + or unknown status
    - Heparin 350-400units/kg with an ACT goal of >400; dosing is size, age, and institution dependent
    - Tranexamic Acid

      Contraindications: h/o seizure, h/o CVA, renal insufficiency; some institutions avoid it for patients > 18 years of age

      Dosing is institution-dependent, for example:

      For patients <1 yo

      Loading dose 30mg/kg over 20 mins after induction

      Continuous infusion 10mg/kg/hr

      For patients >1 yo

      Loading dose 10mg/kg over 20 mins after induction

      Continuous infusion 10mg/kg/hr
  - Inotropes
    - Epinephrine 0.01 – 0.5 mcg/kg/min
      - For neonates and infants (<10kg), Epinephrine will be used as the primary inotrope
    - Dobutamine 2-15 mcg/kg/min
    - Dopamine 2-10 mcg/kg/min

        - Milrinone 0.25-0.75 mcg/kg/min
        - Vasopressin 0.0003 – 0.0012 mcg/kg/min
    - Additional medications when early extubation is planned
        - Sugammadex is most commonly used or Neostigmine/Glycopyrrolate
        - Ondansetron
        - IV Acetaminophen
        - Ketorolac in patients > 6 months, after discussion with the surgeon regarding the safety of use
        - Consider long-acting narcotic (Morphine or Dilaudid) for pain control after extubation.

    - Monitors
        - Invasive blood pressure monitoring with an indwelling arterial catheter
        - Central venous line access and pressure monitoring
        - ASA standard monitoring: 2 pulse oximetry, 5-lead ECG, capnograph, two-site temperature monitoring
        - Cerebral oximetry
        - TEE
    - Blood products

      Standard of care to have pRBC available for bypass cases

      Autologous whole blood may be taken pre-bypass, stored in anticoagulant CPD, then administered back-to-back (with filter), after heparin reversal with protamine
    - PICU Bed Availability
        - Patient transferred postoperatively to the Cardiothoracic Intensive Care Unit.

Intraoperative Considerations:

- General
    - Anesthetic and Physiologic Implications of the Surgical Procedure
        - In addition to the implications discussed above, the anesthesiologist should be vigilant concerning the following key points:[3]
            - Worsening of L-to-R shunt with hyperventilation and increased FIO_2
            - Paradoxical embolus
            - Hypothermia
            - Post–CPB pulmonary hypertension and right ventricular failure
- Induction
    - Inhalation induction

      Patients without CHF can tolerate inhalation induction
    - IV induction

      Patients with CHF benefit from IV induction

      Options:
        - Etomidate, ketamine, low-dose propofol
        - Fentanyl or sufentanil due to better hemodynamic stability
    - IM induction
        - IM ketamine can be used for patients who cannot tolerate inhalation induction or if IV access cannot be established.
- Positioning

  Supine

  Head turned to the left to expose the neck vessels in case quick surgical access is needed

- Maintenance
    - Drugs and agents as mentioned above
- Hemodynamic/Physiologic goals [1-4]
    - Management Goals Before Cardiopulmonary Bypass:
        - Maintain cardiac output by maintaining HR, contractility, and preload
        - Maintain a balance between PVR and SVR
            - Avoid maneuvers that decrease the PVR-to-SVR ratio
            - A reduction in the PVR-to-SVR ratio increases pulmonary blood flow and compromises systemic perfusion
        - Maintain normal oxygenation and normal to mild hypercarbia.
            - Limit FiO_2 to restrict pulmonary blood flow with goal oxygen saturations low to mid 90s.
            - Patients who have persistently elevated pulmonary blood flow despite these measures may benefit from increased systemic perfusion prior to CPB with additional volume expansion, inotropes, or both.
            - Once surgical exposure is established, partial or complete occlusion of the right pulmonary artery mechanically hinders the amount of L-R shunt, facilitating better systemic perfusion.
        - Avoid hemodilution with a large amount of crystalloids or colloids
        - Avoid large increases in the PVR-to-SVR ratio. A significant increase can lead to right-to-left shunting
            - Loss of the airway due to apnea, inadvertent extubation, or obstruction will increase PVR
            - High opioid doses can blunt the increase in PVR upon surgical stimulation.
        - In the case of right-to-left shunting, measures should be taken to decrease it.
            - Controlled Ventilation is the most reliable way to manipulate PVR by controlling carbon dioxide, avoiding hypo- or hypercapnia.
            - SVR must be maintained or increased
    - Management Goals After Cardiopulmonary Bypass [1-4]
        - Maintain HR (sinus rhythm preferred) corresponding to age normal range
            - Post-CPB, stroke volume is often diminished, so maintaining HR becomes critical to preserve CO.
        - Decrease PVR through ventilation control.
            - Ventilation strategies play a pivotal role in controlling PVR after cardiopulmonary bypass. Optimizing oxygenation, avoiding hypercarbia and acidosis, and maintaining ideal lung volumes and airway pressures can significantly reduce PVR.
            - This is especially critical in the presence of PVOD, where elevated PVR can compromise cardiac output.
        - In the circumstance where PVR is high, inotropic support of the right ventricle may be warranted.
            - After VSD closure, the right ventricle will face a high afterload
            - Dobutamine (5–10 mcg/kg/min) or dopamine (5–10 mcg/kg/min) provides inotropic support without the expense of an increased PVR
            - Milrinone (0.5–1.0 mcg/kg/min) after a loading dose of 50 mcg/kg has more direct PVR-decreasing effects in addition to inotropic, lusitropic, and SVR-reducing effects.

        - Infants with large VSDs, particularly in association with trisomy 21, are at high risk for the development of severe pulmonary hypertension and hypertensive crises after CPB.
            - Inhaled nitric oxide (iNO) can be started when pulmonary hypertension, RV dysfunction, and low cardiac output persist.
- Surgical Considerations [3]
    - TEE diagnostic imaging will demonstrate the adequacy of repair after CPB. It will rule out residual VSD.
    - Maintain Hct >25% to 30%
    - Coagulopathy, particularly in very small children.
    - Point-of-care measurements, including ACT, thromboelastography, and quantitative platelet counts, can guide treatment of inadequate hemostasis.

- Emergence/Disposition
    - The majority of patients without significant comorbidities who undergo VSD repair can be extubated immediately after the surgery ends.
    - Mechanical ventilation and sedation are reserved for patients who are at risk of pulmonary HTN crisis.

- Post-op Care
    - Children in cardiac failure may require inotropic support postoperatively[1]
    - Infective endocarditis prophylaxis for 6 months[3]
        - In case there is a residual defect, prophylaxis is continued indefinitely.

Case Specific Complications/Pitfalls [2]

- Post-op ventricular dysfunction is more likely with ventriculotomy
    - Ventriculotomy during VSD repair is associated with a higher risk of postoperative ventricular dysfunction due to risk of myocardial injury, impaired electrical conduction, reduced compliance due to scarring and inflammation, and increased risk of aneurysm formation
- Incomplete closure of the VSD may prevent separation from CPB
    - TEE can assess for residual VSD
- Tricuspid regurgitation may result if the tricuspid valve sustains an insult during a difficult transatrial approach.
- Subaortic or subpulmonic obstruction may result after patch closure of the VSD.
- Transient heart block may ensue as the atrioventricular node and the bundle of His are in near proximity to the surgical site.
    - Temporary epicardial AV sequential pacing may be required to end CPB

References

1. Coté Charles J., Lerman J, Anderson BJ. anesthesia for children undergoing heart surgery. In: *A Practice of Anesthesia for Infants and Children*. Philadelphia, PA: Elsevier; 2019.
2. Nasr VG, DiNardo JA. Ventricular Septal Defect. In: *The Pediatric Cardiac Anesthesia Handbook*. Hoboken, NJ: John Wiley & Sons Inc; 2017.
3. Fleisher LA, Roizen MF, Roizen JD. Ventricular Septal Defect (Congenital). In: *Essence of Anesthesia Practice*. Philadelphia, PA: Elsevier; 2018.
4. Davis PJ, Cladis FP, Smith RM. anesthesia for congenital heart disease . In: *Smith's Anesthesia for Infants and Children*. Amsterdam: Elsevier; 2022.

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