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SURGICAL ATRIAL SEPTAL DEFECT (ASD) REPAIR

Surgical Specialty:	Cardiothoracic
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Background:

- General Considerations
  - Atrial septal defects (ASDs) account for approximately 10% of congenital heart disorders and occur about twice as often in females than in males.[1]
  - ASDs can occur in different portions of the atrial septum and the location reflects the abnormality during embryogenesis that led to the anomaly.
  - Four main morphologic types exist: ostium secundum defects, ostium primum defects, inferior and superior sinus venosus (SVASD) defects, and coronary sinus (CS) defects.
  - Physiologic consequences of ASDs reflect the shunting of blood across the atria; the direction and magnitude of the shunt are determined by the size of the ASD and the relative compliance of the ventricles.
  - Secundum defects account for approximately 80% of all ASDs.[1]
  - Small ostium secundum ASDs can be primarily closed while large defects are patched (usually with pericardium).
  - Non-operative transcatheter device closure of small secundum ASDs can be done in the cardiac catheterization laboratory.
  - Primum ASDs usually require patch closure, with concurrent suture closure of the anterior leaflet mitral cleft when present.
  - SVASD defects without partial anomalous pulmonary venous return (PAPVR) can be primarily closed with a patch.
  - In the setting of PAPVR, the superior vena cava (SVC) is transected and oversewn above the origin of the anomalous vein(s). A pericardial patch is used to direct the SVC orifice across the ASD into the left atrium (LA). The distal end of the SVC is anastomosed to the roof of the right atrial appendage, recreating an SVC-RA continuity.[2]
  - When the anomalous vein(s) enter the SVC remote from the SCV/RA junction, a baffle within the SVC to direct pulmonary venous blood across the ASD into the LA may be necessary. A patch is used to enlarge the anterior wall of the SVC to allow unrestricted blood return to the RA.[2]
  - Correction of CS defects depends on the presence/absence of a left SVC (LSVC). In the absence of an LSVC, the ostium of the CS is oversewn allowing coronary sinus blood to drain into the LA (small physiologic R-L shunt). When an LSVC is present

and drains into the CS, the LSVC is ligated below the vein connecting the 2 cavae and the CS is oversewn.[2]

- Patient Considerations
  - Spontaneous closure is most likely to occur in patients with small secundum ASDs; the majority close spontaneously by two years but some as late as five years of age.
  - Moderate to large secundum ASDs, types other than secundum ASDs, and ASDs detected later in childhood/adolescence are unlikely to close spontaneously; some may increase in size with time.
  - ASD surgical repair is usually done for children between the ages of 3 and 5 years.[1]
  - ASD surgical repair is recommended for children who have pulmonary over-circulation, enlargement of the right heart, and evidence of severe L-R shunting through the ASD (ie, ratio of pulmonary-to-systemic flow [Qp/Qs] >2:1 or other evidence of a large L-R shunt by echocardiography).
  - Associated Comorbidities/Syndromes
    - Primum ASDs are usually not isolated, typically being associated with cleft anterior mitral valve leaflet and atrioventricular canal defects (AVCDs).
    - Secundum ASDs typically present as isolated cardiac defects but they may coexist with other types of ASDs. Some patients with secundum ASD have functional mitral valve prolapse.
    - In some cases, patients with secundum ASDs may have a family history due to a specific genetic mutation. These genetic disorders can be associated with other congenital cardiac and/or extra-cardiac abnormalities (i.e. Holt-Oram Syndrome, Noonan syndrome, Treacher Collins syndrome, and thrombocytopenia-absent radii syndrome.)[3]

Anesthetic Planning:

- Pre-Anesthetic Evaluation
  - Physical exam to assess for the presence of signs and symptoms indicating right heart failure and pulmonary hypertension (PH).
  - Review of medications, particularly heart failure medications.
  - Pre-operative electrocardiogram (ECG), particularly to assess for any possible arrhythmias.
  - A comprehensive transthoracic echocardiographic evaluation (TTE) is performed. Important aspects of the exam include:
    - Accurate measurement of the size of the defect and degree of L-R shunting
    - Identification of other cardiac defects
    - Assessment for PH
  - Cardiac catheterization is considered the gold standard for measuring Qp/Qs, but noninvasive techniques are accepted alternatives, and cardiac catheterization before ASD repair is not routinely done. Pulsed Doppler echocardiography and cardiac MRI are now routinely used and provide an acceptable correlation with catheterization laboratory estimates of shunt flow.
  - Discussions to have with surgeon/family
    - Risks/benefits/alternatives/surgical vs percutaneous closure with parents and surgeon
    - Code Status
    - Cardiopulmonary By-Pass (CBP) use and its risks and possible complications
    - ECMO possibility in case of failure to wean off-CPB

- Specific or Unique Room Set-up Requirements
    - Airway
        - Size-appropriate endotracheal tube (ETT)
    - Drugs/Infusions
        - Emergency drugs
            - Epinephrine diluted to 100mcg/ml,10mcg/ml, 1 mcg/ml
            - Phenylephrine 10 mcg/ml, 1 mcg/ml
        - Muscle relaxant
            - Rocuronium (induction dose: 0.6-1.2 mg/kg) is most often used, also used in incremental doses to ensure immobility.
            - Vecuronium or Cisatracurium may be used instead
        - Midazolam (0.05-0.2 mg/kg)
        - Opioids
            - Fentanyl: Up to 15mcg/kg used for patients planned for extubation, higher doses can be used for patients with anticipated post-operative mechanical ventilation
            - Fentanyl infusion (1-2 mcg/kg/h)
            - Morphine infusion 10-30 mcg/kg/hr
        - Dexmedetomidine (0.2-1 mcg/kg/hr)
        - Antibiotics
            - Cefazolin (30mg/kg every 4 hours)
            - Bactroban ointment applied to nares if MSSA-positive
            - Cefazolin and Vancomycin (15mg/kg over 1 hr) for MRSA-positive or unknown status
        - Heparin 350-400 units/kg with ACT goal of >400
        - Protamine: Dosing is institution-dependent and typically based body weight (3 to 4 mg/kg) or in a ratio to the heparin dosage (mg-to-mg) of 0.7:1 up to 1.3:1, with target ACT after protamine administration ~10% of pre-CPB baseline
        - Antifibrinolytics:
            - Tranexamic Acid (TA) (Dosing is institution-dependent)
                - For patients <1 yo
                    - Loading dose 30mg/kg
                    - Continuous infusion 10mg/kg/hr
                - For patients >1 yo
                    - Loading dose 10mg/kg
                    - Continuous infusion 10mg/kg/hr
            - Aminocaproic Acid (EACA)
                - For patients < 2 months old:
                    - Loading dose 40 mg/kg
                    - Continuous infusion 30 mg/kg/hr
                - For patients > 2 months old:
                    - Loading dose 75 mg/kg
                    - Continuous infusion 75 mg/kg/hr
        - Inotropes
            - Epinephrine (0.05-1 mcg/kg/min)
            - Milrinone (0.5-1 mcg/kg/min)
            - Dopamine (5-10 mcg/kg/min)
            - Dobutamine (5-10 mcg/kg/min)
            - Vasopressin (3–30 mU kg/h)

    - Inhaled nitric oxide (iNO) can be considered when PH, RV dysfunction, and low cardiac output persist.
    - Additional medications when early extubation is planned
        - Sugammadex or Neostigmine/Glycopyrrolate for reversal of muscle paralysis dosed according to train-of-four
        - Ondansetron (0.15 mg/kg)
        - IV Acetaminophen (10-15 mg/kg)
        - Consider titration of long-acting narcotics (Morphine 0.05-0.2 mg/kg or Hydromorphone 0.005-0.015 mg/kg) for pain control after extubation
        - Ropivacaine 0.25% (2-3 mg/kg) ± Dexmedetomidine (1 mcg/kg) can be used for neuraxial/regional blocks for patients with anticipated extubation
        - Regional or field block – see below

- Monitors
    - Pulse oximetry x 2
    - 5-lead EKG
    - Age/size appropriate non-invasive blood pressure cuff
    - Arterial line
    - Central line for central venous pressure (CVP) and other invasive pressure monitoring.
        - Some institutions opt for no central pressure monitoring
    - Temperature monitors x2: nasopharyngeal/oropharyngeal/esophageal AND urinary/rectal.
    - Cerebral oximetry: Near-infrared spectroscopy (NIRS) and/ or transcranial Doppler are utilized by many centers
    - Warming device(s)
    - Bladder catheter for volumetric urine collection
    - Transesophageal Echocardiography (TEE)
    - Activated clotting time (ACT) point-of-care monitor
    - Viscoelastic measurements of hemostasis with thromboelastography (TEG) or rotational thromboelastometry (ROTEM) have been increasingly used to guide decisions concerning blood product administration

- Blood Availability
    - Packed red blood cells, platelets, fresh frozen plasma
    - Cryoprecipitate and Factor VII can be considered in cases with complicated medical history and excessive blood loss
    - Some institutions harvest autologous whole blood from the patient before bypass (collected into a citrate-phosphate-dextrose (CPD) storage bag to prevent clot formation) and transfuse it post-bypass after protamine administration. Autologous blood may decrease the need for donor blood products. Typical volumes of autologous phlebotomy are 10-20% estimated blood volume.
- PICU Bed Availability
    - Cardiac ICU bed is needed for post-op monitoring
- Other Indicated Resources
    - CPB machine
    - ECMO may be needed in case of failure to wean off CPB

Intraoperative Considerations:

- General
  - Considerations for ASD repair include general considerations for CPB and its risks and potential complications.
  - Arterial blood gases are measured to ensure the adequacy of ventilation and oxygenation, as well as to ensure normal electrolytes, hemoglobin, lactate, and calcium; measurements are done before, during, and post-CPB.
  - In general, a reduced FiO_2, a PaO_2 of 50–60mmHg, a $PaCO_2$ of 45–55mmHg, and the application of PEEP, can be used to maintain pulmonary vascular resistance (PVR).
- Induction
  - Patients frequently don’t have intravenous access preoperatively and may require a mask induction.
  - Most children with ASDs have good cardiac reserve and tolerate inhalation induction.
  - Good ventilatory management is important to allow manipulation of the PVR and prevent shunt reversal.
- Positioning
  - Supine with careful consideration for pressure points
- Maintenance
  - Anesthesia is maintained with isoflurane (0.5–1.0%) or sevoflurane (1.0–2.0%).
  - Muscle paralysis is given to ensure patient immobilization.
  - Dexmedetomidine (0.2-1mcg/kg/hr) &/or fentanyl (1-3 mcg/kg/h) or morphine drips can be used to supplement inhaled anesthetics.
  - Vaporizer on the CPB circuit along with supplemental opioids and muscle relaxants can be used for maintenance during CPB.
- Hemodynamic/Physiologic goals
  - Pre-Bypass:
    - Maintenance of the heart rate, contractility, and preload to maintain cardiac output and systemic perfusion
    - Avoidance of large increases in the PVR:SVR ratio which may induce a R-L shunt with the potential for reduced arterial oxygen saturation
    - Avoidance of a decrease in the PVR:SVR ratio which may cause an increase in L-R shunt with a consequent increase in pulmonary blood flow and potential reduction of systemic perfusion and oxygen delivery
  - Post-Bypass
    - Maintainance of heart rate to maintain cardiac output
    - Reduction of PVR, especially in cases where an elevated PVR exists
    - Support to the RV with inotropes if needed
  - Ensure adequate hemostasis post-CPB; arterial blood gas, ACT, and TEG or ROTEM can help guide the administration of anticoagulation/antifibrinolytics and blood products.
- Surgical Considerations
  - Cerebral NIRS is used during CPB to assess the adequacy of oxygen delivery to the brain which may lead to improved neurologic outcomes after cardiac surgery; a drop of 20% from baseline is considered significant and protective measures to increase cerebral saturation should be made.[4]

    - Adequate muscle paralysis with neuromuscular blocking agents is necessary to ensure immobility and diaphragmatic paralysis to create a completely still surgical field and to decrease subclinical shivering and oxygen consumption.
- Emergence/Disposition
    - In most cases, prolonged ventilation is not needed and early extubation can be done after adequate titration of anesthesia and muscle paralysis.
    - Early extubation offers advantages in terms of decreased costs and morbidity associated with ICU admissions.
    - Some clinicians advocate extubation in the OR, others advocate waiting until the child is in ICU which could save OR time and decrease risks of cardiovascular instability, hypothermia, and bleeding.[5]
    - Research into regional and neuraxial anesthesia for pediatric cardiac surgery is limited:
        - The use of neuraxial anesthesia remains controversial with the main concerns being the risk of bleeding and the potential for neurologic complications.
        - Caudal morphine has been used to provide postoperative analgesia; produced good analgesia for approximately 6 hours and reduced analgesic requirements for up to 24 hours.[6]
        - The rate of adverse effects was less with thoracic catheter epidurals compared with various caudal, lumbar epidural, and spinal approaches.[7]
        - Bilateral paravertebral blocks (PVB) have been used for post-op pain relief but evidence for their use in children is limited.[8]
        - Bilateral transversus thoracis plane (TTP) blocks have recently been studied and showed potential efficacy and safety for use in pediatrics undergoing cardiac surgery.[9]
- Post-op Care
    - Clinical evaluation after surgical closure should assess for signs of embolic events, arrhythmia, and chest pain.
    - Patients are monitored for complications such as pericardial and pleural effusions and post-pericardiotomy syndrome.
    - Echocardiography is essential to detect complications such as residual shunting and pericardial effusions and to assess ventricular function and pulmonary artery pressure.
    - Electrocardiographic monitoring is performed to detect any possible atrial arrhythmias.
    - In patients with repaired ASD requiring a prosthetic device, prophylactic antibiotics are recommended for dental and respiratory tract procedures within the first 6 months after surgery.
    - In patients with repaired ASD with a residual defect at/adjacent to the site of a prosthetic device, antibiotic prophylaxis is recommended for dental and respiratory tract procedures.

Case-Specific Complications/Pitfalls

- During primum ASD repair, heart block may occur due to patch suturing to the crest of the interventricular septum. Heart block may be transient due to myocardial edema at the conducting system but may also be permanent. Heart block can be managed with a temporary or permanent pacemaker.
- Residual or new iatrogenic mitral regurgitation can occur.

- A residual L-R shunt may exist after surgical repair in case PAPVR goes unrecognized during SVASD defect closure.
- In case of elevated PVR, pulmonary vasculature can be hyper-reactive post-CPB; ventilatory measures to reduce post-CPB PVR should be employed to prevent RV dysfunction due to increased afterload.

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