



ATRIOVENTRICULAR SEPTAL DEFECT REPAIR

Surgical Specialty:	Cardiac
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Background:

- **General Considerations**

Atrioventricular septal defect (AVSD), also known as endocardial cushion defect or atrioventricular canal defect, constitutes 4-5 % of congenital heart disease (CHD). It originates from a failure of normal endocardial cushion fusion during embryologic cardiac development. AVSDs are characterized by defects in the atrial and ventricular septa immediately above and below the atrioventricular (AV) valves (tricuspid and mitral) and abnormalities of AV valves. Based on the severity of septal defects and abnormality of AV valves, AVSDs are categorized as follows:

- *Partial AVSD or ostium primum atrial septal defect (ASD)*- when lesions are limited to a primum ASD and a cleft mitral valve.
- *Transitional AVSD or intermediate AVSD*- consists of a primum ASD, abnormal AV valves with two functionally separated orifices, and a very restrictive or absent ventricular septal defect (VSD).
- *Complete AVSD*- the most common type, combines deficiency of both the atrial and ventricular septa with an abnormality of the mitral and tricuspid valves, creating a common AV valve that serves both ventricles.
- *Other variants of AVSD*- include right or left ventricular dominant types, or unbalanced AVSDs, which are similar to the complex single-ventricle heart lesions and will not be discussed in this case-guide.

The pathophysiology of AVSDs ranges from mild left to right shunting, like a simple ASD, to severe volume overload of both the atria and ventricles. Persistent volume overload results in congestive heart failure (CHF) and potentially irreversible pulmonary hypertension. Complete surgical repair for patients with complete AVSDs is usually performed at infancy due to unrestrictive left to right shunting, while the repair of partial or transitional AVSDs is often performed at 2-5 years of age with close monitoring the signs of CHF or other associated lesions that may require earlier repair.

Additional cardiac lesions are present in about 25% of children with AVSDs, including subaortic stenosis, tetralogy of Fallot, and heterotaxy syndrome. There is a strong association with trisomy 21, with nearly 1/3 to 1/2 of the children with AVSDs diagnosed with trisomy 21. ⁽¹⁾

- **Disease Specific Considerations**

- The specific physiology of AVSDs caused by intracardiac shunts and abnormalities of common AV valves includes the following: ⁽²⁾
 - Left to right shunt(s) at atrial and/or ventricular level and AV valve regurgitation (see figure 1)
 - Increased pulmonary blood flow
 - Volume overloading of the right and left ventricles
 - The transmission of systemic pressures to the right ventricle and pulmonary arteries
 - Pulmonary arterial hypertension (PAH), bidirectional or even right to left shunt in AVSDs with large shunt, if left unrepaired.
- Children with partial or intermediate AVSD usually present with minimal symptoms, while those with large left to right shunts and severe AV valve regurgitation often present symptoms of CHF in infancy including failure to thrive, tachypnea, intercostal retractions, and recurrent respiratory infections. Infants with CHF are usually medically managed with diuretics +/- digoxin and are referred for early surgical repair.
- The surgical repair varies depending on the lesions and institutional factors, including patch closure of the primum ASD and/or the inlet VSD, closure of mitral valve cleft in the case of primum AVSDs, and septation of the common AV valve tissue into two separate competent left and right AV valves in complete AVSDs. In patients who have significant pulmonary over circulation or are not candidates for early complete repair, pulmonary artery banding can be utilized as a temporizing measure to limit pulmonary blood flow (PBF), and therefore, reduce the development of pulmonary vascular disease.

- **Associated Comorbidities/Syndromes**

- Patients with trisomy 21 are prone to the early development of PAH, potentially related to pulmonary hypoplasia, endothelial dysfunction, and obstructive sleep apnea. ⁽³⁾ Surgical repair of complete AVSD is generally performed at 3 to 6 months of age when there is sufficient maturation of AV valve tissue to facilitate good septation and before the development of pulmonary vascular changes and PAH ⁽⁴⁾
- AVSDs are also found in children diagnosed with other congenital syndromes, such as Noonan's syndrome and Ellis-van Creveld syndrome. ⁽⁵⁾ In addition to cardiac lesions, facial dysmorphism, vertebral anomalies, and coagulation factor deficits in Noonan's syndrome and characteristic orthopedic abnormalities in Ellis-van Creveld syndrome warrant additional anesthetic considerations.

Anesthetic Planning:

- **Pre-Anesthetic Evaluation**

- History and physical exams
 - Partial AVSD- minimal abnormal findings on physical exam
 - Complete AVSD- signs and symptoms of CHF, including tachypnea, intercostal retractions, failure to thrive, and poor peripheral perfusion may present increased anesthetic risks.
 - Orofacial features leading to potential difficult airway management

- Additional labs/tests indicated during work-up
 - ECHO to assess the size and function of the heart, abnormalities of AV valve and severity of the regurgitation, the amount and direction of shunting, and additional intracardiac anomalies.
 - A 12-lead ECG for rhythm assessment
 - Chest radiograph for the presence of pulmonary edema or infiltrates
 - Cardiac catheterization is usually reserved for children with potential PAH. Review the catheterization report to assess PA pressures, pulmonary vascular resistance (PVR), ratio of pulmonary to systemic flow (Qp:Qs), and pulmonary vascular reactivity to selective vasodilators such as O₂ therapy, inhaled nitric oxide(NO), etc.
 - Routine preoperative laboratory testing, such as CBC, basic metabolic panel, and coagulation profile as per institutional protocol
 - Additional testing may be warranted for patients with syndromes. See last section for special consideration for children with trisomy 21

- Discussions to have with surgeon/family
 - Verify consent for blood products
 - Early extubation for eligible children
 - Prolonged mechanical ventilation and inhaled nitric oxide therapy for children with preoperative PAH
 - Code status and ECMO candidacy

- Specific or Unique Room Set-Up Requirements
 - Airway
 - Age-appropriate endotracheal tubes (ETTs) and airway equipment
 - Availability of smaller diameter ETTs for infants with trisomy 21
 - Advanced airway devices, such as video laryngoscope, for potential difficult airway in children with syndromes

 - Drugs/Infusions
 - Routine medications for cardiopulmonary bypass (CPB) cases per local practice, such as opioids (morphine, fentanyl), antifibrinolytics (tranexamic acid, aminocaproic acid), sedatives (midazolam, lorazepam, dexmedetomidine), heparin, and protamine.
 - Vasoactive medications (doses may vary), such as dopamine (3-10 mcg/kg/min), epinephrine (0.02-0.1 mcg/kg/min), vasopressin (0.0003-0.001units/kg/min), and milrinone (0.25-0.7 mcg/kg/min)
 - Esmolol (25-200 mcg/kg/min), nitroprusside (starting at 0.5 mcg/kg/min) infusion to treat potential immediate postoperative hypertension, if indicated.
 - Furosemide 0.5-1 mg/kg for children with CHF symptoms
 - Calcium chloride 10-20mg/kg via central line

 - Monitors
 - Standard ASA monitors with 5 lead EKG, invasive monitors (arterial blood pressure and central venous pressure) for CPB, pulmonary arterial pressure monitoring for children with PAH.
 - Monitors for measure tissue perfusion, such as Foley catheter, near infrared spectroscopy, etc.
 - Trans-esophageal echocardiography (TEE) probe and ECHO machine
 - Point-of-care viscoelastic tests, such as TEG, ROTEM per local resource

- Blood products available in the operating room
 - In addition to red blood cells or fresh whole blood, platelets, fresh frozen plasma, and cryoprecipitate are usually indicated in young children.
- Temporary pacemaker
- Postoperative PICU Admission. Confirm the availability of ventilatory support, including iNO.
- ECMO may be needed for children with PAH, increased PVR, and poor ventricular function.

Intraoperative Considerations:

- General
 - Anesthetic and Physiologic Implications of the Surgical Procedure
Anesthetic management depends on the degree of left to right shunting and the presence and severity of PAH. Balancing the ratio of PVR to SVR and thereby limiting the amount of pulmonary over circulation is paramount to successful management and is usually accomplished by manipulations in FiO₂ and ventilation.⁽⁵⁾
 - Patients with CHF and inadequate tissue perfusion have limited or exhausted cardiac reserve. Consider fentanyl, ketamine, and low concentrations of inhalational agent.
 - Hypotension may be caused by volume depletion due to preoperative fasting and diuretics. Consider judicious volume expansion with close monitoring of CVP.
- Induction
 - Inhalational induction with sevoflurane is reserved for infants and children with good cardiac function and minimal signs and symptoms
 - Consider an intravenous induction with midazolam, ketamine or etomidate, and fentanyl for children with limited cardiac reserve, symptomatic CHF, or increased PVR.
- Positioning
 - Supine position and avoid neck hyperextension in children with trisomy 21.
- Maintenance
 - Inhalational agent with sevoflurane or isoflurane combined with an opioid infusion for infants with good ventricular function and minimal preoperative CHF symptoms.
 - Combining an opioid and dexmedetomidine infusion with low dose inhalational agent is reserved for infants with limited cardiac reserve, PAH, and reversed shunting from right to left.
- Hemodynamic/Physiologic goals⁽²⁾
 - Pre-CPB:
 - Maintain heart rate, contractility, and preload to maintain cardiac output.
 - Avoid fluid overload, especially for children with CHF symptoms and on diuretics.
 - Avoid decreases in PVR: SVR ratio.
 - Avoid significant increases in PVR: SVR ratio in infants with increased PAH.
 - Post -CPB:
 - Maintain age-appropriate heart rate and sinus rhythm.

- Reduce PVR with O₂ and paralytics for children with PAH. Consider nitric oxide.
 - Inotropic support (e.g., dopamine 3-10 mcg/kg/min, epinephrine 0.02-0.1 mcg/kg/min, milrinone 0.25-0.7 mcg/kg/min, vasopressin 0.0003-0.001 units/kg/min)
 - Arrhythmias, including complete heart block, are common; consider temporary pacing.
 - Pulmonary hypertension crisis (increased PA pressure, decreased right ventricular function, and systemic hypotension) is treated with moderate hyperventilation with 100% O₂, correcting acidosis (respiratory and metabolic), neuromuscular blockade, and inhaled pulmonary vasodilators (iNO or prostacyclin analogs).
- Surgical Considerations
 - Surgical issues, such as residual ASD or VSD, persistent left AV valve regurgitation or stenosis, surgical induced heart block, and other arrhythmias, may exist and cause difficulty to separate from bypass. TEE for assessment of surgical repair and cardiac function is extremely useful in these situations.
 - Both coagulopathy and surgical bleeding may exist post bypass; Consider using TEG or ROTEM to guide the treatment
- Emergence/Disposition and postoperative Care
 - Early extubation is reserved for infants with adequate surgical repair, minimal post bypass bleeding, and minimal vasoactive medication support. This is commonly seen in patients with partial AVSDs and some minimally symptomatic complete AVSDs. ICU admission after extubation is required.
 - Delayed extubation is considered for children with severe preoperative CHF symptoms, decreased ventricular function, required substantial blood products for post bypass bleeding, large doses of vasoactive medication support, complete heart block, and those who developed PH crisis. These patients should remain intubated postoperatively.
 - Multimodal pain control is essential.
 - Intraoperative local anesthetic infiltration or regional blocks (caudal, intrathecal opioids) depend on local practice.
 - Acetaminophen intravenous or PO, q6h around the clock (doses vary based on patient's age, please consult pharmacists)
 - Non-steroidal anti-inflammatory drugs can be considered when adequate hemostasis is achieved in infants older than 6 months
 - Opioid administration, including oral (hydrocodone, oxycodone) and intravenous such as continuous morphine or fentanyl infusion or nurse-controlled analgesia.
 - Dexmedetomidine infusion can be used as an adjuvant.

Case Specific Complications/Pitfalls

Special consideration for children with trisomy 21

- Routine preoperative C-spine imaging is not necessary for asymptomatic children and is reserved for children with symptoms of cervical myelopathy⁽⁶⁾
- Check preoperative thyroid function and continue thyroid hormone replacement in children with hypothyroidism.

- Difficult intravenous access is frequently encountered, and some patients may require ultrasound-guided peripheral intravenous access.
- Pay close attention to minimal neck manipulation during induction, intraoperative positioning, and post-operative transportation.
- Bradycardia and hypotension during inhalational induction with sevoflurane- in addition to decreasing the sevoflurane concentration, treatment with intravenous atropine (0.01-0.02 mg/kg, max 0.5mg), or glycopyrrolate (4 mcg/kg, max 0.1mg), and/or epinephrine (titrate to effect) may be necessary ⁽⁷⁾
- Higher incidence of hypotonia and obstructive sleep apnea in these children. Consider appropriate ventilatory support after extubation, such as CPAP, BiPAP, and high flow nasal cannula.

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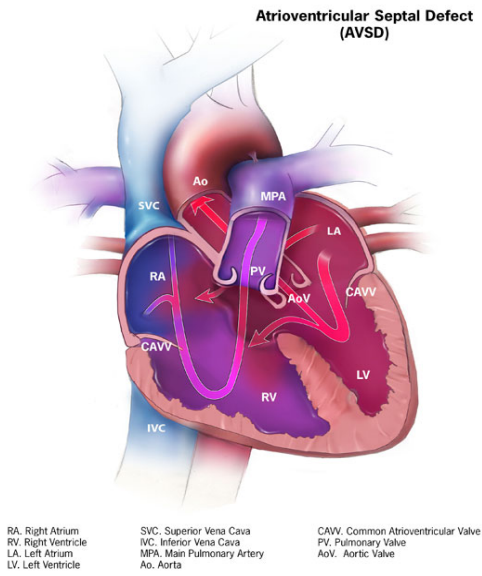


Figure 1. Pathophysiology of complete atrioventricular septal defect
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