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Anesthesia for Left-to-Right Shunt Lesions

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Overview

Introduction

Left-to-right shunt lesions are the most common congenital heart defects, accounting for approximately 50% of all lesions. They are defined by a communication between the systemic and pulmonary circulations that allows shunting of well-oxygenated (systemic) blood to the less oxygenated (pulmonary) circuit. This definition applies whether the associated structures are located on the left or right side anatomically

The degree of shunting through a left-to-right shunt lesion may be limited by the size of the defect or the resistance to blood flow on either side. For example, shunting between high-pressure systems like the ventricles through a large VSD is dependent on the ratio of pulmonary vascular resistance (PVR) to systemic vascular resistance (SVR). In contrast, left-to-right shunting through a large atrial septal defect (ASD) occurs primarily during atrial contraction and is dependent on the relative diastolic compliances of the right and left ventricles into which the atria eject.

Shunting at the level of the great arteries results in increased pulmonary artery blood flow, which increases pulmonary venous return to the left atrium, leading to increased left ventricular end-diastolic volume and left ventricular stroke work by the Frank–Starling mechanism. The left ventricle gradually dilates and hypertrophies, producing increased left ventricular end-diastolic pressure followed by increased left atrial pressure.

Shunting at the level of the great arteries also produces a decrease in diastolic blood pressure from run-off of blood into the low-pressure pulmonary circuit after closure of the aortic valve. Low diastolic pressures decrease coronary perfusion, potentially creating ischemia from decreased myocardial oxygen delivery in the setting of increased oxygen demand from the hypertrophied ventricle. The final result is pulmonary edema from pulmonary venous congestion and left heart failure. As PVR increases, there is an increased pressure burden on the right ventricle and eventual right heart failure.

Prolonged exposure of the pulmonary vasculature to increased flow and pressure results in a fixed increase in PVR. When PVR exceeds the SVR,shunt reversal occurs, resulting in cyanosis and erythrocytosis. Eisenmenger syndrome results when this level of PVR becomes irreversible.

Hemoglobin concentration is another contributing factor to the amount of left-to-right shunting. Elevated blood viscosity, which rises with increasing hemoglobin concentration, increases both PVR and SVR.

The physiologic decline in hemoglobin concentration in the first 3 months of life is thought to have a substantial role in the normal fall of PVR after birth.

The normal compensatory mechanisms that maintain systemic cardiac output and myocardial performance in the patient with a left-to-right shunt include the Frank–Starling mechanism, the sympathetic nervous system, and hypertrophy of the myocardium. Manifestations of these compensatory mechanisms include sweating and tachycardia. Infants are also often tachypneic from decreased lung compliance associated with increased pulmonary blood flow.

Anesthetic management for left-to-right shunt lesions should be individualized to the patient, but certain generalities do exist. Premedication with intravenous (IV) or oral drugs such as midazolam (0.05–0.1 mg/kg IV or 0.75–1.0 mg/kg PO) can be safely administered for the purpose of decreasing anxiety and providing more controlled induction of anesthesia. Standard American Society of Anesthesiologists (ASA) monitors along with the use of invasive arterial and central venous pressure monitoring and careful attention to urine output are recommended for all cases involving cardiopulmonary bypass (CPB).

Transesophageal echocardiography (TEE), cerebral oximetry, and cerebral blood flow monitoring are also useful monitoring adjuncts. Patients with severe, poorly controlled congestive heart failure (CHF) may be intolerant to the myocardial depressant effects of inhalational anesthetics, and for this group of patients, IV anesthesia with fentanyl and midazolam is preferred. In most situations, however, inhalation induction with sevoflurane is a viable option when IV access is not initially available

Additional anesthetic issues include avoidance of air bubbles in IV lines to prevent paradoxical emboli. The anesthesiologist must be cognizant of the pulmonary vasodilatory effect of oxygen and hypocarbia and manipulate ventilation in order to balance the PVR and SVR. Such measures generally include minimizing the FiO_2 and avoiding hyperventilation (maintaining $PaCO_2$ between 40 and 50 mmHg).

Patent ductus arteriosus

The incidence is higher for premature births and PDA is two to three times more common in females than in males. PDA is also found as part of other complex congenital heart defects and is usually the source of pulmonary or systemic blood flow in patients with a functional single ventricle before palliative repair.
The ductus arteriosus is a vascular communication between the descending aorta and the pulmonary artery.

The ductus arteriosus is an essential component in normal fetal circulation; it becomes functionally closed within 10–15 hours after birth and permanently closes by thrombosis, intimal proliferation, and fibrosis in the first 2–3 weeks. Functional closure is initiated by several mechanisms, including aeration of the lungs, removal of prostaglandins produced in the placenta, increased arterial PO_2, and release of vasoactive substances (bradykinin, thromboxanes, and endogenous catecholamines) .

Pathophysiology

The degree of left-to-right shunting depends on several factors, including the size of the PDA and the ratio of PVR and SVR. In patients with large PDAs, the diastolic runoff into the pulmonary artery results in lowered aortic diastolic pressure, which may increase the risk of myocardial ischemia, especially in the presence of anemia or lowered SVR.

Surgical and transcatheter approaches and outcomes

In premature newborns, initial management of a PDA is typically pharmacological closure using cyclo-oxygenase inhibitors such as ibuprofen or indomethacin. Surgical treatment is usually reserved for patients who fail medical therapy. Surgical options include posterolateral thoracotomy with ligation or division of the PDA.

Complications of surgical treatment include bleeding, chylothorax, vocal cord paralysis (injury to recurrent laryngeal nerve), pneumothorax, atelectasis, recurrence of patency, and inadvertent ligation of the pulmonary artery or descending aorta.

Video-assisted thoracoscopic surgery is increasingly popular due to decreased pain, decreased hospital cost (secondary to decreased hospital stay), and avoidance of post-thoracotomy syndrome (rib fusion, chest wall deformities, scoliosis, and compromise of pulmonary function). Disadvantages of VATS include intraoperative oxygen desaturation and hypercarbia.

Anesthetic considerations

The anesthetic management for PDA ligation depends on factors such as the patient's clinical condition, prematurity, coexisting disease, body weight, and surgical technique. Large volume venous access (which may be a 22- or 24-gauge IV in a premature infant) and forced air-warming devices are recommended. Pulse oximetry of both upper and lower extremities will assist in detecting inadvertent ligation of the descending aorta. For patients with coexisting disease, intra-arterial pressure monitoring provides a method of assessing arterial blood gases, electrolytes, hematocrit, and acid–base status.

Whether by cuff or arterial line, blood pressure should be monitored in both an upper and lower extremity and observed carefully before and after ductal occlusion. Proper ductal occlusion will typically be accompanied by an increase in diastolic blood pressure consistent with elimination of pulmonary runoff. Significantly decreased or absent blood pressure in the lower extremity indicates aortic rather than ductal occlusion. A gradient between the systolic pressure in the upper and lower extremities indicates creation of an aortic coarctation.

Although inhaled anesthetics can be safely used for many patients undergoing PDA ligation, neonates are prone to hemodynamic instability with exposure to inhaled anesthetics and benefit from an IV anesthetic technique using opioids, such as fentanyl, and possibly a benzodiazepine along with muscle relaxation.

Lung isolation improves surgical exposure, especially for VATS surgical techniques, but may require ventilation with 100% inspired oxygen to maintain acceptable oxygenation. Prior to lung isolation, FiO_2 should be minimized and hypocarbia avoided in order to maintain pulmonary vascular tone and limit the degree of left-to-right shunting. Lung isolation is usually unnecessary for small infants; gentle retraction and packing during open thoracotomy, or gentle CO_2 insufflation and retraction during VATS are generally sufficient.

Infants having thoracotomy or VATS often require postoperative mechanical ventilation, especially if they are premature. Older patients or patients undergoing transcatheter closure are often extubated at the conclusion of the case.

Atrial septal defects

The right and left atria are normally divided by the fusion of two septa: the septum primum and the septum secundum.
Five different types of ASDs exist: secundum, primum, sinus venosus, patent foramen ovale (PFO), and coronary sinus.

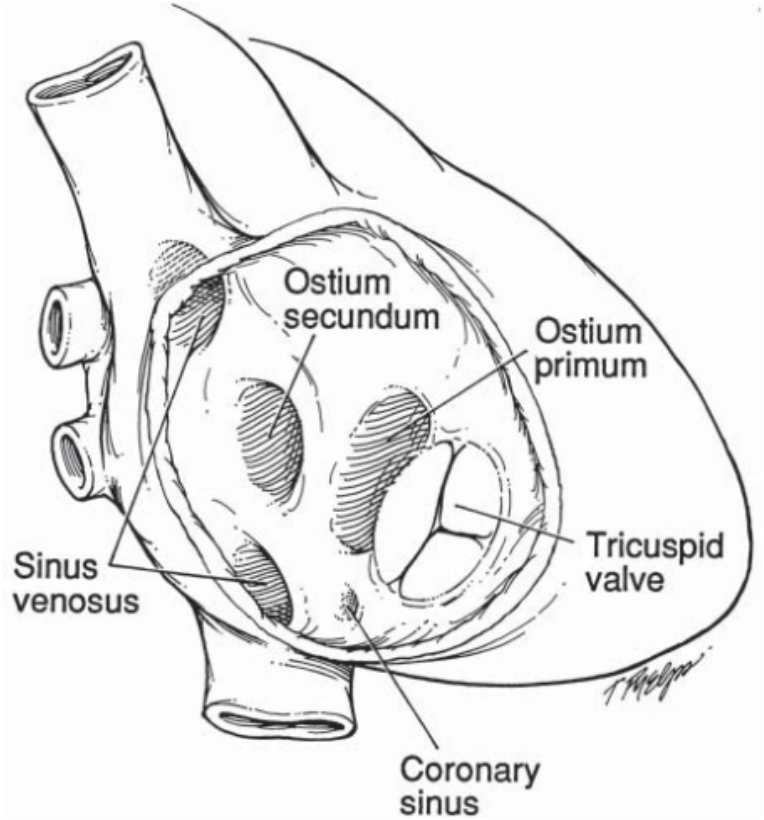
Types of atrial septal defects

- Secundum ASD
  It results from an abnormal reabsorption of the septum primum or defective formation or shortening of the septum secundum.
- Primum ASD
  The primum ASD results from abnormalities in formation of the septum primum. It is frequently associated with atrioventricular canal (AVC) defects.
- Sinus venosus ASD
  Sinus venosus defects result from abnormal development of the septum secundum or the sinus venosus (the primitive venous collecting chamber). The most common type is located near the superior vena cava (SVC) orifice and is associated with partial anomalous pulmonary venous return (PAPVR).

Types of atrial septal defects...

- Patent foramen ovale
  Patent foramen ovale results from failure of fusion of the septum primum to the limbus of the septum secundum. Patency of the foramen ovale is normal during fetal life and allows right-to-left shunting of blood in order to bypass the lungs in fetal circulation. Following birth, PVR decreases and SVR increases. Subsequent higher pressure in the left atrium causes the septum primum to close over the foramen ovale.
- Coronary sinus ASD
  Coronary sinus ASD, also called an unroofed coronary sinus, results from an absence in the wall between the coronary sinus and the left atrium.

Types of atrial septal defects



Natural history

Isolated ASDs are usually asymptomatic during infancy and childhood, despite the increased volume load on the right ventricle. CHF usually occurs after the second or third decade of life due to chronic right ventricular volume overload. Pulmonary hypertension can occur in up to 13% of unoperated patients younger than 10 years of age; however, progression to Eisenmenger syndrome is unusual.

An ASD is sometimes discovered during a neurological work-up for transient ischemic attacks or strokes from paradoxical emboli.

Pathophysiology

The amount of left-to-right shunting at the atrial level is dependent on two factors: the size of the defect and the relative compliance of the right and left ventricles. Shunting occurs primarily during diastole and produces a volume burden on the cardiovascular system that is proportionate to the degree of shunting.

Anesthetic considerations

Patients with an isolated ASD are generally asymptomatic and do not have pulmonary hypertension. Therefore, the induction of anesthesia can be safely accomplished with either inhalation or IV techniques. Whenever possible, patients should have an intraoperative TEE performed prior to incision, because transthoracic echocardiographic studies are sometimes unable to exclude the possibility of PAPVR due to difficulty in visualizing all four pulmonary veins.

During surgery, TEE can be helpful in assessing de-airing of the left heart and adequacy of the repair. Most patients have good myocardial function and do not require inotropic support perioperatively. Maintenance of anesthesia typically consists of a combination of inhaled and IV agents. Adjunct regional techniques are favored by some, and may facilitate early extubation. Tracheal extubation in the operating room has been shown to decrease patient charges without compromising patient care when compared with extubation in the ICU. Whatever technique is chosen, the primary goals for the uncomplicated ASD patient should include preparation for an early extubation either in the operating room or within the first 4 hours postoperatively

Ventricular septal defects

Ventricular septal defect is the most common congenital heart defect, occurring in 50% of all children with congenital heart disease (CHD) and in 20% as an isolated lesion. Reported incidence ranges from 1.56 to 53.2 per 1,000 live births. VSD is associated with a variety of inherited conditions, including trisomy 13, 18, and 21.

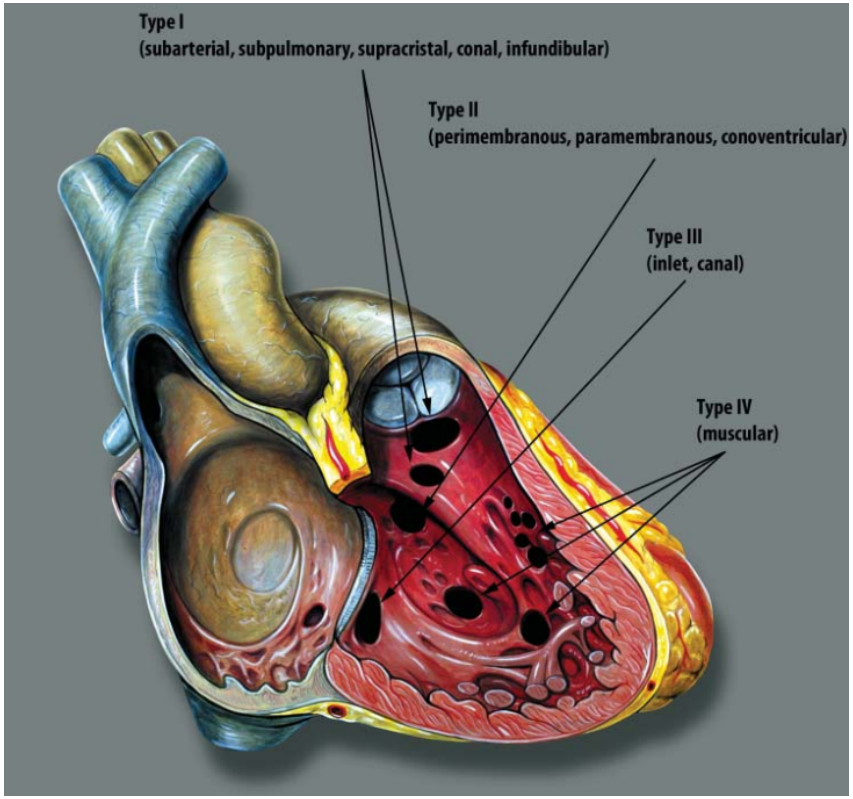
Types of ventricular septal defect

- Type I: subarterial VSD
  The subarterial (also called subpulmonary, supracristal, conal, or infundibular) VSD is located just beneath the pulmonary valve.
  As a result of the location of this defect, a Venturi effect may be produced by the jet of blood flowing through the VSD, causing the right or non-coronary aortic cusp of the aortic valve to prolapse toward the defect, producing aortic insufficiency. This type of lesion is more common in the Asian population
- Type II: perimembranous VSD
  The perimembranous (also called paramembranous or conoventricular) VSD is a communication adjacent to a portion of the membranous septum and the fibrous trigone of the heart, where the aortic, mitral, and tricuspid valves are in fibrous continuity.

Types of ventricular septal defect...

- Type III: inlet VSD Inlet VSDs (also called canal type) are located in the posterior region of the septum beneath the septal leaflet of the tricuspid valve. These defects account for approximately 10% of VSDs.
- Type IV: muscular VSD Muscular VSDs are located anywhere within the muscular portion of the interventricular septum. These defects can be multiple and represent approximately 2–7% of VSDs.

Types of ventricular septal defect



Natural history

Patients presenting with VSD may be asymptomatic or exhibit signs and symptoms of CHF in varying degrees.The rate and extent of progression of symptomatology depend on the patient's age, the size of the defect, and the degree of left-to-right shunting. Infants who have non-restrictive VSDs typically develop symptoms of CHF by 3 months of age, because of the physiologic decline in PVR that occurs during early postnatal life. If left untreated, 15% of patients with large VSDs develop pulmonary hypertension that will progress to the development of pulmonary vascular obstructive disease, shunt reversal, and cyanosis (Eisenmenger syndrome) by the age of 20 years.

Therefore it is important to close large non-restrictive VSDs within the first 2 years of life.

Spontaneous closure of small perimembranous and muscular VSDs occurs in as many as 50% of patients , and such patients are typically asymptomatic.

Anesthetic considerations

Anesthetic management for the patient with VSD is similar to that of ASD. Pulmonary hypertension may develop early, especially in patients with trisomy 21, and preoperative chest radiograph revealing decreased pulmonary vascular markings is indicative of pulmonary hypertension. Such patients may respond to the use of inhaled nitric oxide prior to termination of CPB and/or in the postoperative period. Right heart failure with decreased cardiac output may result if pulmonary hypertension is not controlled, and may require the use of dopamine, milrinone, dobutamine, or isoproterenol.

Conduction disturbances may be transient or permanent. Atrioventricular block, formerly reported to occur in up to 10% of patients post-VSD repair, is now a rare complication occurring in less than 1% of patients after VSD closure. If heart block develops, treatment with atrioventricular synchronous pacing using temporary pacing wires is indicated. Junctional ectopic tachycardia is sometimes observed in patients younger than 1 year after surgery for lesions that involve VSD repair, most commonly after tetralogy of Fallot correction. Treatment includes cooling to 35°C, increasing anesthetic depth, paralysis, procainamide, esmolol, or amiodarone.

Intraoperative use of TEE will allow recognition of residual VSDs and intracardiac air, as well as providing an assessment of ventricular volume and function.

Patients with uncomplicated VSDs are good candidates for extubation in the operating room or early after arrival in the ICU.

The End