Chapter 2

Ventricular Septal Defect

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Case Scenario

A 6-month-old child weighing 3.7 kg presents from home for bilateral inguinal hernia repair and circumcision. He was recently seen in the emergency room for an incarcerated hernia that was manually reduced. He was born at 26 weeks estimated gestational age with a birth weight of 1100 grams. He was intubated for 3 weeks and weaned from high-flow nasal cannula to room air prior to discharge from the neonatal intensive care unit at 34 weeks postconceptional age. The parents say that in the neonatal unit the baby was diagnosed with a "hole in his heart" and that he takes medicine for it every day.

Current vital signs are heart rate 140 beats/minute, respiratory rate 45 breaths/minute, blood pressure 70/38 mm Hg and SpO_2 98% on room air.

The patient has been scheduled for same-day surgery and the parents are requesting a spinal anesthetic, as they are concerned about neonatal apnea with general anesthesia.

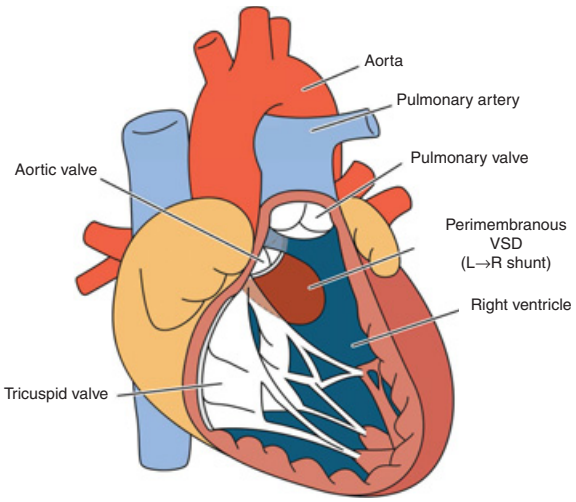

Figure 2.1 Perimembranous ventricular septal defect. Drawing by Ryan Moore, MD, and Matt Nelson.

Key Objectives

- Understand the physiology of a left-to-right shunt.
- Describe the preoperative workup and perioperative management of a premature infant with a ventricular septal defect.
- Identify an anesthetic plan in the context of balancing pulmonary vascular resistance and systemic vascular resistance.
- Outline postoperative management and appropriate discharge planning.

Pathophysiology

How are VSDs characterized?

Ventricular septal defects (VSDs) are the most common congenital heart defect, occurring in 50% of patients with congenital heart disease (CHD). It is estimated that 75%–80% of VSDs are ***perimembranous*** (see Figure 2.1), indicating the communication between ventricles occurs adjacent to the very small membranous septum. ***Inlet*** VSDs, also known as canal type, are located posteriorly beneath the septal leaflet of the tricuspid valve. (See Figure 2.2.) ***Muscular*** VSDs may occur anywhere within the muscular wall of the interventricular septum and can also exist as part of other more complex cardiac defects. (See Figure 2.3.) ***Subarterial*** (also called subpulmonary, supracristal, conal, or infundibular) VSDs lie beneath the pulmonary valve within the outlet septum. (See Figure 2.4.) A VSD can also be present in many other forms of CHD as part of a constellation of defects.

What are the hemodynamic effects of a VSD?

An isolated VSD results in the ability to shunt blood between the left and right ventricles. The size of the defect and pulmonary vascular resistance (PVR) determine the blood flow across the VSD. Left-to-right (L-to-R) shunting generally occurs predominantly during systole and this shunting results in an increased volume load to both ventricles.

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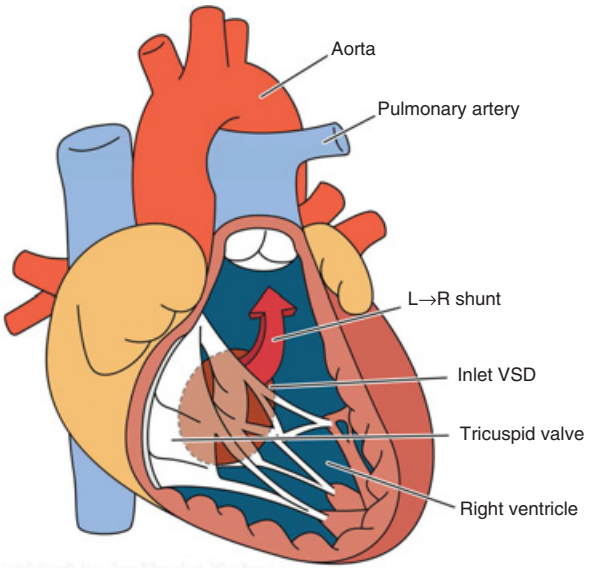

Figure 2.2 Inlet ventricular septal defect. Drawing by Ryan Moore, MD, and Matt Nelson.

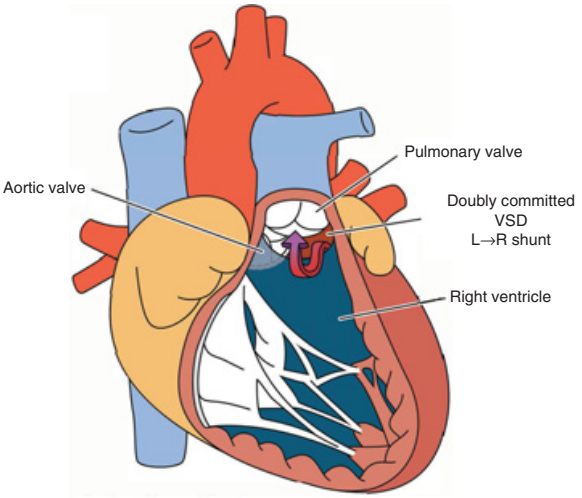

Figure 2.4 Subarterial ventricular septal defect. Drawing by Ryan Moore, MD, and Matt Nelson.

What is the difference between a restrictive and a nonrestrictive VSD?

The term "nonrestrictive" implies that the size of the VSD approximates the size of the aortic annulus, allowing equalization of pressures in the right and left ventricles. In reality, small pressure gradients may exist due to the relative resistances of the systemic and pulmonary vascular beds.

Infants with nonrestrictive VSDs often display signs and symptoms of congestive heart failure (CHF) as the PVR falls in the first postnatal months and the degree of L-to-R shunting increases. Nonrestrictive VSDs should be closed in the first 2 years of life to avoid the development of pulmonary vascular changes that can lead to the development of pulmonary vascular occlusive disease (PVOD) and Eisenmenger's syndrome. Given the history of prematurity this infant is at increased risk for early development of such changes. A patient's symptomatology depends on his age, the size of the VSD, the degree of L-to-R shunting, and other factors impacting PVR, such as a history of prematurity.

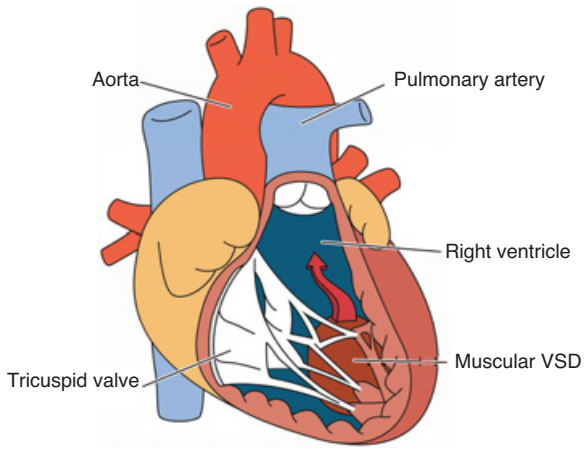

Figure 2.3 Muscular ventricular septal defect. Drawing by Ryan Moore, MD, and Matt Nelson.

Clinical Pearl

A patient's symptomatology depends on his age, the size of the VSD, the degree of L-to-R shunting, and other factors impacting PVR, such as a history of prematurity.

What is the Q_p:Q_s ratio and what is its significance?

The Q_p:Q_s ratio is the ratio of the pulmonary blood flow (Q_p) to systemic blood flow (Q_s) and denotes the magnitude of a cardiovascular shunt. Normally this ratio equals 1, with the entire preload to the RV eventually becoming the preload to the LV. In cardiac lesions with a L-to-R shunt the Q_p:Q_s is >1, and in lesions with a resultant R-to-L shunt the Q_p:Q_s is <1.

In lesions with the capability for intracardiac shunting it is important to determine the percentage of recirculating blood. Often this ratio is calculated by comparing blood saturation levels obtained during cardiac catheterization.

The following formula may be utilized to calculate the ratio of pulmonary to systemic blood flow:

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$$Q_p{:}Q_s = \text{Ao sat} - \text{MV sat}/\text{PV sat} - \text{PA sat},$$

where *Ao sat* is aortic oxygen saturation; *MV sat* is mixed venous oxygen saturation; *PV sat* is pulmonary venous saturation; and *PA sat* is pulmonary arterial saturation. Small VSDs will have a $Q_p{:}Q_s$ ratio of <1.5:1, medium VSDs a $Q_p{:}Q_s$ of 2–3:1, and large VSDs can have $Q_p{:}Q_s$ ratios exceeding 3:1.

Clinical Pearl

While it is not essential to know the $Q_p{:}Q_s$ ratio for an isolated VSD, the $Q_p{:}Q_s$, if available, can provide valuable information to assess the degree of shunting occurring with a VSD. A larger $Q_p{:}Q_s$ is indicative of greater pulmonary overcirculation.

What factors influence the $Q_p{:}Q_s$ ratio and the degree of shunting?

The $Q_p{:}Q_s$ ratio is affected by changes in pulmonary and systemic vascular resistance (PVR and SVR).

Acidemia, hypercarbia, hypoxemia, hypothermia, and pain are known to elevate PVR, thereby decreasing pulmonary blood flow (PBF). Additionally, significant atelectasis or high inspiratory pressures and/or tidal volumes can contribute to reductions in PBF.

Conversely, increased FiO_2, hyperventilation, alkalosis, and the use of inhaled nitric oxide (iNO) can reduce PVR and promote PBF. Nitric oxide should be available in the operating room for high-risk patients with evidence of pulmonary hypertension.

Is there a "typical" age for repair of an isolated VSD?

Timing for the surgical repair of a VSD varies based on patient age and symptomatology as well as the size and location of the lesion. Most nonrestrictive VSDs are closed within the first few years of life to avoid long-term pulmonary sequelae that can lead to eventual shunt reversal and cyanotic R-to-L shunting (Eisenmenger syndrome). If the patient is <6 months old and has failure to thrive and CHF refractory to medical management, then surgical repair of the VSD is generally considered. If medical management allows continued growth and appropriate milestone acquisition, surgery is often delayed until the child is older to avoid cardiopulmonary bypass during infancy.

Clinical Pearl

Poor weight gain and continued tachypnea and dyspnea, particularly with exertion or feeding, often signify pulmonary overcirculation.

Should the cardiac lesion be repaired prior to the hernia surgery?

Ventricular septal defects may decrease in size over the first year of life and may become relatively asymptomatic. Spontaneous closure of small perimembranous and muscular VSDs occurs in up to 50% of patients. With careful surveillance and medical management some patients may be able to avoid surgical intervention altogether. However, the incidence of inguinal hernias is much higher in low birthweight and premature infants compared to full term infants, affecting up to 30% of preterm infants. Overall, it is also thought that risk of incarceration is much higher in this population. As hernias can compromise intestinal blood supply, repair should be performed on a semielective basis to avoid the risk of emergency surgery should the hernia become incarcerated. In this case scenario, the patient has already experienced one episode of incarceration and should therefore undergo definitive hernia sac closure to prevent a potentially life-threatening recurrence.

Anesthetic Implications

What are the important preoperative considerations for this child?

History The focus of the preoperative evaluation should center on this patient's current medical status as related to his major concerns of prematurity and an unrepaired VSD. While failure to thrive may be noted in infants for a variety of reasons, the most likely etiology in this patient is the presence of the VSD, resulting in chronic pulmonary overcirculation. In addition to history from the parents, efforts should be made to review the most recent cardiology evaluation and echocardiogram, chest radiograph, and electrocardiogram.

Due to this patient's prolonged neonatal intubation it is important to identify airway concerns, including any known or suspected airway stenosis. The results of any prior evaluation of the airway by otolaryngology should be reviewed. Additionally, significant prematurity can also predispose patients to laryngotracheomalacia. The patient history should elucidate episodes of noisy breathing or stridor.

Physical Examination On physical examination the presence of increased work of breathing and/or significant chest retractions at baseline should be noted along with the rate and character of respirations. Recent symptoms of any recent respiratory infections should be elicited, as these would place the child at increased anesthetic risk due to potential increases in airway irritability and PVR. Preoperative baseline hemoglobin–oxygen saturation should be noted. Signs and

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symptoms of CHF may be present, including hepatomegaly and pulmonary congestion.

Laboratory A preoperative hemoglobin/hematocrit should be obtained as anemia may place this baby at increased risk for postoperative apneic events. If significant anemia is present, it may be prudent to obtain a type and screen. Since the infant is taking furosemide it may be appropriate to obtain an electrolyte panel as well.

Regarding this child's cardiac condition, what other information should be sought?

The current medication regimen, compliance with medications, and the response to therapy should be evaluated. The presence of tachypnea, especially with feeding and/or poor weight gain, can signify chronic pulmonary overcirculation and CHF. Additionally, severely preterm infants are at risk for bronchopulmonary dysplasia due to failure of arborization of distal alveoli and their vasculature. This is a common cause of elevated pulmonary vascular pressures in formerly preterm children and can lead to varying degrees of pulmonary hypertension. Generally, these patients are seen relatively frequently by the pediatrician and cardiologist to ensure adequacy of medical management. If a change in symptoms is noted during the preoperative evaluation, this should raise concerns that the patient's medical management is not currently optimized.

The most recent cardiology evaluation should be reviewed along with recent echocardiographic findings. Echocardiographic findings to review include

- Type, number, location, and size of VSDs
- Estimates of pulmonary artery pressure and signs of RV hypertension
- The presence of any valvular abnormalities/regurgitance
- Biventricular function
- Presence of other cardiac anomalies

What potential perioperative issues exist for a patient on diuretic therapy?

Diuretics are frequently used to medically manage symptoms associated with CHF. The goal of treatment is euvolemia achieved by fluid restriction and diuretic therapy. Diuretics such as furosemide may result in electrolyte disturbances and/or preoperative hypovolemia, particularly in a patient who has been nil per os (NPO) for an extended time period.

What are the effects of anesthesia on systemic and pulmonary vascular resistances?

An understanding of factors impacting PVR and SVR is vital to the anesthetic management of the patient with a large VSD. Most anesthetic agents will decrease SVR to a greater degree than PVR, especially during induction and prior to surgical stimulation. This imbalance may initially reduce L-to-R shunting through the VSD, particularly if pulmonary vascular pressure is elevated. With the initiation of surgery an increase in SVR may augment L-to-R shunting through the VSD.

The optimal anesthetic is one in which PVR and SVR are relatively balanced with respect to the patient's preoperative baseline. Both PVR and SVR are impacted by multiple factors including ventilation strategy, anesthetic agents used, and the depth of anesthesia. A ventilation strategy utilizing a low FiO_2, mild hypercarbia, and acidosis (pH 7.30) is the primary modality used to limit pulmonary overcirculation during anesthesia and surgery. It is important to remember common causes of hypotension and oxygen desaturation during anesthesia. In addition to issues with airway management, the differential diagnosis for hypoxemia in a patient with a large VSD should include increases in PVR and an awareness of the factors that can adversely impact PVR. It is important to recognize that the use of phenylephrine to treat hypotension may actually increase the degree of L-to-R shunting by increasing SVR, and although systemic blood pressure may increase, there may be a reduction in systemic cardiac output.

Clinical Pearl

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If the surgeon prefers a laparoscopic approach, what are the physiologic implications for this patient?

Laparoscopic abdominal insufflation increases intraabdominal pressure and may reduce preload. In turn, the pressures required to maintain adequate ventilation increase, as does the degree of atelectasis. Additionally, insufflation with carbon dioxide raises $PaCO_2$. When taken together, this cascade tends to reduce PBF and the degree of L-to-R shunting through the VSD which may be beneficial for this patient. Release of the pneumoperitoneum (i.e., insufflation) at the end of the laparoscopy can lead to a sudden increase in L-to-R shunting and should be monitored closely. Treatment may consist of elevating PVR by reducing minute ventilation or anesthetic depth. If at any point during the laparoscopic approach the hemodynamics become compromised, there should be

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a discussion with the surgeon regarding conversion to an open surgical approach.

What is an appropriate anesthetic plan for this patient?

In the absence of poor ventricular function, pulmonary hypertension, or other major comorbidities, most patients with isolated VSDs can safely undergo an inhalational induction if appropriately NPO and without risk factors necessitating a rapid sequence induction of anesthesia. However, as this patient has a history of prematurity, vascular access could prove challenging and this should be factored into decision-making regarding the appropriate method of anesthetic induction.

Assuming this patient does not currently have symptoms of heart failure or evidence of pulmonary hypertension, it would be appropriate to proceed with a laparoscopic approach. Although spinal anesthesia is successfully used in some centers, this technique is dependent on surgeon and institutional preference and the surgical approach chosen. A spinal block is not generally used for a laparoscopic approach given the impact of insufflation on the child's ventilatory status and a general anesthetic with endotracheal intubation is appropriate.

A caudal block is helpful to minimize both intraoperative anesthetic requirements and to promote postoperative pain control while minimizing the need for opioids. Young children undergoing regional or neuraxial anesthesia do not experience the sympathectomy seen in adults, and therefore concern for preload reduction is not warranted. While a penile block and/or bilateral ilioinguinal blocks would be reasonable, a caudal block will provide analgesia to all surgical sites. The patient's weight of 3.7 kg in this case precludes multiple blocks due to the limited maximum allowable local anesthetic dosage that can be used.

Fluid management is a final major anesthetic consideration. Intravenous fluids should be administered judiciously, as excessive volume loading may promote postoperative symptoms of CHF and pulmonary edema.

Clinical Pearl

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Can this patient be discharged following an uncomplicated repair?

This patient is at risk for apnea of prematurity which is known to occur more commonly in premature infants (<60 weeks postconceptional age [PCA]) and those with anemia (hemoglobin <10 g/dL). Apnea can be centrally or peripherally mediated, although most cases at this age are of mixed etiology. At-risk patients should be observed following surgery and have at least 12 apnea-free hours prior to discharge. Institutional guidelines may vary regarding the precise PCA required for discharge.

If a spinal anesthetic is performed, it does not obviate the need for postoperative monitoring for apnea. This patient's PCA is approximately 50 weeks, and he should be admitted postoperatively to a unit with the capability to continuously monitor heart rate and pulse oximetry.

Where should the patient be monitored postoperatively and for how long?

The patient may be monitored in the post-anesthesia care unit provided he has an uneventful operative course. Postoperative admission for observation due to the concern for apnea of prematurity is required and should, at minimum, include continuous heart rate and pulse oximetry monitoring. Home medications and diet should be resumed as soon as possible, and unnecessary administration of intravenous fluids should be avoided.

Suggested Reading

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