Chapter 5

Unbalanced Atrioventricular Septal Defect

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Case Scenario

A 1-day-old, full-term female weighing 3.1 kg with the characteristic facies of trisomy 21 vomits after her first oral feeding and an abdominal radiograph reveals duodenal atresia. She is made nil per os and a nasogastric tube is placed to aspirate secretions and gas. The patient is scheduled for repair of duodenal atresia with a laparoscopic approach.

Current vital signs are blood pressure 75/38 mm Hg, heart rate 138 beats/minute, respiratory rate 29 breaths/minute, and SpO_2 98% on room air.

Transthoracic echocardiogram revealed the following:

- *Complete atrioventricular septal defect (Rastelli type A) with a common atrioventricular valve unbalanced to the right*
- *Mild hypoplasia of the transverse aortic arch and aortic isthmus with peak gradient 21 mm Hg*
- *Moderate common atrioventricular valve regurgitation*
- *Moderately dilated right ventricle with normal biventricular function*

Key Objectives

- Review the pathophysiology of a complete atrioventricular septal defect.
- Compare the anatomic and physiologic differences between balanced and unbalanced atrioventricular septal defects.
- Formulate a perioperative anesthetic plan for an infant with an unbalanced atrioventricular septal defect.
- Discuss the implications of laparoscopic insufflation for patients with unrepaired atrioventricular septal defect.

Pathophysiology

What is a complete atrioventricular septal defect?

Also known as an endocardial cushion defect, a complete atrioventricular septal defect (AVSD) is a constellation of defects at the AV junction. Instead of two separate AV valves, mitral and tricuspid, there is a single common valve orifice serving both ventricles. The common AV valve is situated between the right and left sides of the heart and includes two leaflets that bridge the right and left sides; these are designated as the superior (anterior) bridging leaflet and the inferior (posterior) bridging leaflet. Just superior to the plane of the common AV valve is a primum atrial septal defect (ASD), and just inferior to this plane is an inlet ventricular septal defect (VSD). These defects collectively comprise a communication between the four chambers in the center of the heart. (See Figure 5.1.)

Clinical Pearl

With a septum primum ASD, an inlet VSD, and a common AV valve, a complete AVSD collectively comprises a communication between the four chambers in the center of the heart.

What are the primary physiologic issues in a patient with a complete AVSD?

The primary physiologic aberrations in patients with AVSD include:

- ***Left-to-right (L-to-R) shunting*** of blood through the ASD and VSD that can lead to congestive heart failure (CHF), increased cardiac work, and resultant ventricular hypertrophy
- ***AV valve regurgitation*** that can lead to volume overload of the ventricles and decreased cardiac output

How does L-to-R shunting occur in the context of an AVSD?

Left-to-right shunting occurs when blood moves from the left to the right side of the heart through the septal defects such that oxygenated blood meant to flow to the systemic circulation instead flows back through the right side of the heart to the lungs.

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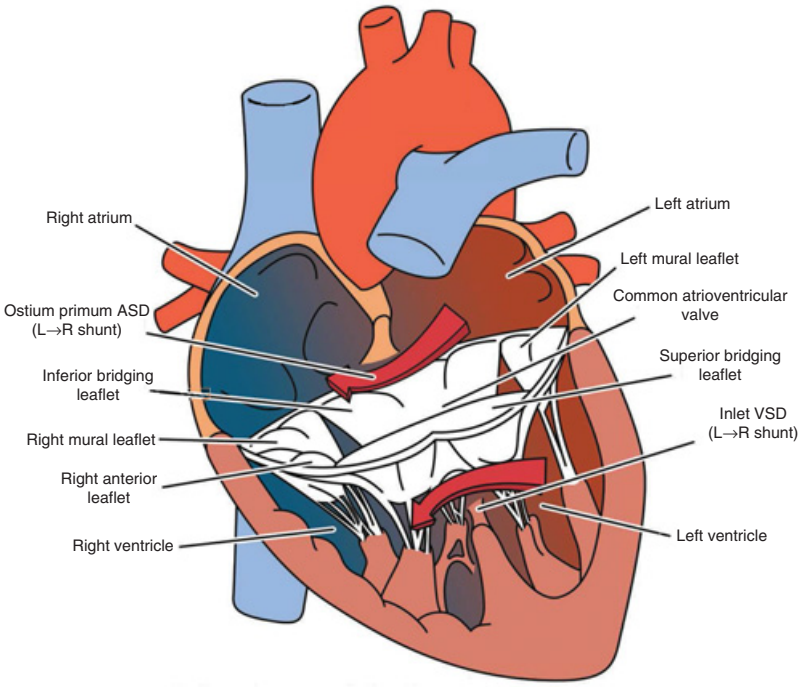

Figure 5.1 Complete atrioventricular septal defect. Drawing by Ryan Moore, MD, and Matt Nelson.

In restrictive ASDs and VSDs, blood flows L-to-R due to the higher pressure in the LA and LV compared to the RA and RV.

With a nonrestrictive VSD, pressures are essentially equal in the LV and RV such that the direction of blood flow will proceed in the direction of lower resistance. Pulmonary vascular resistance (PVR) is usually lower than systemic vascular resistance (SVR) so blood preferentially flows away from the systemic circulation and toward the lungs. At birth, due to elevated PVR, the L-to-R shunt may not be as large, whereas several weeks later after PVR has fallen to postnatal levels the L-to-R shunt will be more significant. As PVR decreases, L-to-R shunting increases.

Clinical Pearl

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What is the Rastelli classification?

In 1966, Giancarlo Rastelli described a classification system to help identify different approaches for surgical repair of complete AVSD based on anatomic variation in the defects. This classification system is based on the morphology of the anterior (superior) bridging leaflet, the amount of bridging over the VSD, the location of chordal attachments, and the degree of associated hypoplasia of the tricuspid anterosuperior leaflet. Classification is not based on the inferior bridging leaflet or the posterior common leaflet.

- ***Rastelli type A***: This is the most common variant, with the anterior common leaflet separated into a right and left component by extensive attachment of chordae tendineae from the superior bridging leaflet to the crest of the ventricular septum. The left superior leaflet is situated directly over the left ventricle (LV) while the right superior leaflet is directly over the RV. The superior leaflet is divided and attached to the ventricular septum.
- ***Rastelli type B*:** The type B atrioventricular septal defect is rare, with an anomalous papillary muscle attached to the left side of the common anterior bridging leaflet from the right side of the ventricular septum.
- ***Rastelli type C***: In the type C AVSD, the ventricular septum is bridged by the anterior bridging leaflet, which is not divided and is without chordae attachments to the crest of the ventricular septum. The valve is therefore floating above the ventricular septum.

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What is the relationship between trisomy 21 and an AVSD?

Trisomy 21 is the most common genetic abnormality in infants and the most frequent chromosomal anomaly associated with CHD. Forty to fifty percent of patients with trisomy 21 have CHD, which is a major cause of morbidity and mortality for these patients. For children with both trisomy 21 and CHD, 45% have some type of AVSD, of which 75% are complete AVSDs. Additionally, 50% of patients found to have an AVSD have trisomy 21. However, if repaired in the first year of life, long-term results are unchanged.

What symptoms are associated with a complete AVSD in a neonate?

A neonate with an AVSD may exhibit signs of CHF. These signs may include tachypnea, tachycardia, fatigue when feeding, sweating, and poor weight gain. In a patient with a large VSD, severe AV valve regurgitation, or other cardiac anomalies, symptoms may occur in the first week of life. In less severely affected patients, symptoms may not become evident until the postnatal PVR falls and the L-to-R shunting increases, usually within the first few months of life.

Clinical Pearl

The degree of L-to-R shunting in a patient with an AVSD is a major determinant of both cardiac symptoms and the development of pulmonary hypertension.

When anatomically describing an AVSD what does the term "unbalanced" imply?

An unbalanced AVSD occurs when the common AV valve sits disproportionately in one ventricle such that the atrioventricular junction is misaligned. The contralateral ventricle and outflow tract typically have varying degrees of hypoplasia, with the inflow to that ventricle often diminished or restricted. While only approximately 10% of AVSDs are unbalanced, this anatomic variant is more likely to be associated with poor outcomes and increased mortality [1]. (See Figure 5.2.)

Clinical Pearl

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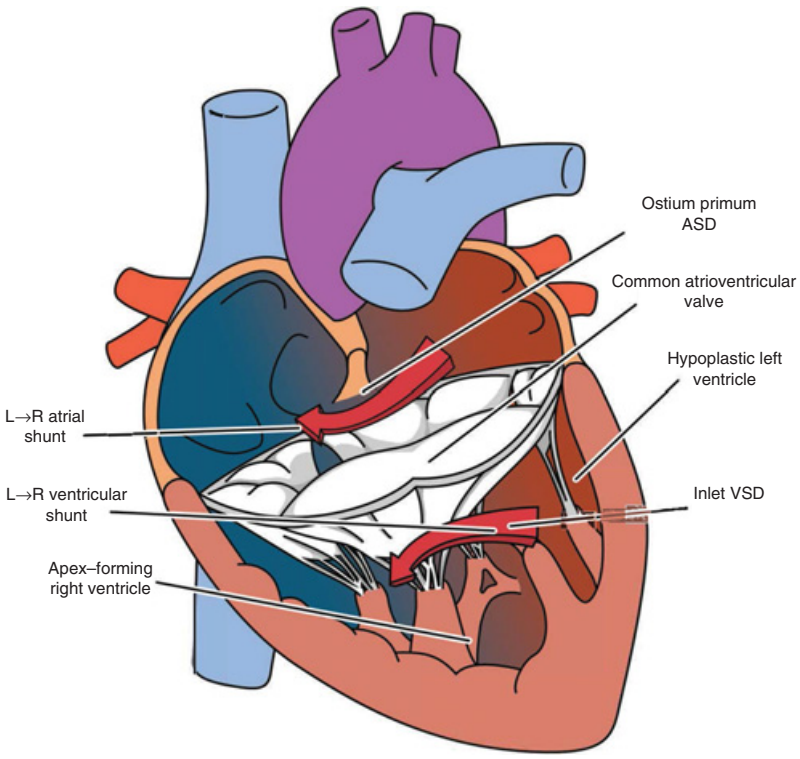

Figure 5.2 Unbalanced atrioventricular septal defect. Drawing by Ryan Moore, MD, and Matt Nelson.

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In unbalanced AVSD anatomy, what is right dominance versus left dominance?

Dominance reflects the ability of blood to traverse through the valve inlet to fill the ventricle. The side contralateral to the dominant ventricle often has some degree of hypoplasia and filling may be inadequate through the diminished area of the corresponding valve leaflets. When more of the common AV valve is apportioned over the RV, this is known as ***right dominance***. Those with the common AV valve distributed more over the LV have ***left dominance***.

Why is it important to understand the degree to which the AVSD is unbalanced?

The degree to which the AVSD is unbalanced will guide treatment of the cardiac disease.

If common AV valve unbalance is mild, the patient may be treated similarly to a patient with a balanced AVSD, where the predominant concern is to mitigate the L-to-R shunt and decrease the risk of heart failure and pulmonary hypertension. These patients may be discharged from the hospital after birth on medical therapy (diuretics and antihypertensives) to decrease the risk of heart failure. Cardiac surgery to repair the AVSD will likely occur when the child is several months old. Early surgical intervention may be required during the neonatal period if there is significant pulmonary overcirculation or inadequate pulmonary blood flow (PBF).

If common AV valve unbalance is severe, the child generally requires single ventricle palliation as the structures on the nondominant side of the heart are hypoplastic, including the ventricle and outflow tracts.

- In a ***LEFT dominant*** unbalanced AVSD, the RV and outflow tract may be hypoplastic and PBF may depend on a patent ductus arteriosus (PDA), requiring a prostaglandin (PGE_1) infusion to maintain ductal patency.
- In a ***RIGHT dominant*** unbalanced AVSD such as this patient, the LV, left ventricular outflow tract, and aorta may be hypoplastic and therefore systemic blood flow may depend on a PDA.

The decision to repair a mildly unbalanced AVSD with a two-ventricular repair or a severely unbalanced AVSD with SV palliation may be clear. However, the optimal surgical approach for a patient with a moderately unbalanced AVSD may not always be apparent.

What cardiac surgical treatment options exist for a moderately unbalanced AVSD?

Overall, surgical options are suboptimal. In a patient with a ***left dominant***, moderately unbalanced AVSD the RV is usually hypoplastic to varying degrees. Depending on the degree of hypoplasia, a two-ventricle repair may be attempted because some RV hypoplasia may be tolerated. If a two-ventricle repair is not well tolerated, a superior cavopulmonary shunt (Glenn shunt) may be added as a way to offload volume from the small RV. This anatomic configuration is referred to as a 1 ½ ventricle repair and is an option only for a left dominant unbalanced AVSD.

There is no way to offload the LV in a ***right dominant*** unbalanced AVSD. The options for this type of defect are dichotomous: either a two-ventricle repair or SV palliation. In this case, the patient has a gradient across the aortic arch and isthmus but has normal LV systolic function and will ultimately have a two-ventricle repair.

What is the expected long-term survival for patients with unbalanced AVSDs?

Long-term survival is dependent on the degree of AVSD imbalance and the planned surgical interventions. A patient with a mildly unbalanced AVSD who undergoes two-ventricle repair may be expected to have survival rates close to those of patients with a balanced AVSD repair. Alternatively, patients with a severely unbalanced AVSD requiring single ventricle palliation (due to one ventricle being excessively diminutive) have lower long-term survival compared to patients with two-ventricle repairs. The difference in outcome may be attributable to either coexisting congenital defects or severe dysfunction of the AV valve, or both, in a patient with severely unbalanced AVSD. There is a 30% incidence of reoperation being required to address AV valve regurgitation in these patients [2].

With regard to single-ventricle palliation, patients with a single left ventricle tend to fare better than those with a single right ventricle.

Anesthetic Implications

Patients with trisomy 21 are known to be at risk for early onset of pulmonary hypertension. What are the implications for these patients?

Rapid development of pulmonary arterial hypertension (PAH) within the first year of life has been described in trisomy 21 patients. Although the mechanisms are not clearly understood, this makes it imperative to repair lesions with large L-to-R shunts early in life before PAH can develop. Additionally, medical therapy aimed to limit the excessive PBF should be undertaken in the time preceding the cardiac surgery to lessen the risk of PAH. Some patients with trisomy 21 may not have the expected

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postnatal decrease in PVR seen in patients without trisomy 21. When the PVR remains high in such patients, they may not exhibit signs of CHF, since the PBF would be impeded by elevated PVR. Interestingly, patients with trisomy 21 without CHD are still at risk for developing PAH. This suggests that other pathologies such as airway obstruction (tonsillar hypertrophy or laryngotracheomalacia) may contribute to the development of PAH in these patients [2]. Because PVR is often elevated in patients with trisomy 21, single ventricle palliation is not the preferred long-term option in patients with this genetic disorder and a two-ventricle repair may be more aggressively pursued.

Should other diagnostic tests be done prior to surgery?

Preoperative laboratory studies should include hemoglobin/hematocrit, electrolyte panel, and a type and screen for blood.

A chest radiograph may show an enlarged cardiac silhouette and increased vascular markings consistent with pulmonary fluid overload. The electrocardiogram (ECG) will often show first degree AV block. Congenital heart block is also a risk in a patient with an AVSD.

Echocardiography should provide adequate information regarding cardiac anatomy and additional imaging should not be necessary prior to the abdominal surgery. Details that should be gleaned from the echocardiogram include:

- The degree of L-to-R shunting
- The degree of unbalance of the AV valve
- Is one ventricle dominant, and if so, to what degree?
- Evidence for and severity of pulmonary hypertension
- Size and function of the ventricles
- Any existence of outflow tract obstruction

What anesthetic concerns exist in a patient with trisomy 21?

Anesthetic concerns in patients with trisomy 21 focus on three main areas: risk for bradycardia, airway issues, and other comorbidities. Owing to their high vagal tone trisomy 21 patients are at increased risk for bradycardia during anesthetic induction with sevoflurane so prompt recognition and treatment – either by decrease in volatile agent concentration and/or administration of atropine – are indicated. Patients often have anatomic airway abnormalities including macroglossia, crowding of the midface, high arched palate, subglottic stenosis, wide, short neck, and adenotonsillar hypertrophy that can increase the risk of upper airway obstruction and difficult intubation. Owing to the risk of subglottic stenosis it is often prudent to downsize the endotracheal tube. Other comorbidities frequently found in patients with trisomy 21 that should be addressed in the preoperative assessment are the potential for C1–C2 subluxation causing cervical spine instability, developmental delay, hypothyroidism, difficult intravenous (IV) access, and gastrointestinal anomalies.

Clinical Pearl

Anesthetic concerns in trisomy 21 patients include bradycardia with sevoflurane induction, airway obstruction, and cardiac disease.

What considerations exist regarding arterial and central line monitoring in this patient?

For patients with all but the most mildly unbalanced AVSD (which may effectively be treated as a large VSD or balanced AVSD) an arterial line may be useful to provide blood pressure monitoring and blood gas analysis, particularly for prolonged cases or in cases where large fluid shifts and/or blood loss are anticipated. However, it is important to realize that placement of an arterial line in infants with Down syndrome can be challenging. The patient's preoperative condition and proposed length of surgery will assist with decision making. With a laparoscopic surgical approach planned in this patient, it can also prove helpful to closely monitor the effects of insufflation on blood pressure and ventilation so that the surgical plan may be modified if necessary.

In the neonate with a severely unbalanced AVSD and significant outflow tract obstruction, a PGE_1 infusion will likely be required for maintenance of the PDA. In this situation a central line is indicated in order to administer vasoactive medications. A peripherally inserted central catheter (PICC) may have already been placed prior to surgery and can be useful for administration of PGE_1 and other vasoactive drugs. When possible, it is best to avoid placing an internal jugular vein line in a neonate who may require a future superior cavopulmonary anastomosis (Glenn procedure). Peripheral IV access will suffice for administration of anesthetic drugs and blood products if needed.

What concerns exist regarding utilizing a laparoscopic surgical approach in this particular case?

Abdominal insufflation is known to decrease cardiac output, increase SVR, increase peak inspiratory pressures, and

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decrease lung volumes at a given pressure. Insufflation with CO_2 may elevate right-sided pressures and increase $PaCO_2$, resulting in decreased PBF and increased R-to-L shunting. A preoperative discussion with the surgeon prior to a laparoscopic procedure in a patient with CHD should include the potential need to decrease insufflation pressure or abandon laparoscopy and open the abdomen if the patient cannot tolerate pneumoperitoneum.

Although the same study has not been repeated in children with CHD, outcome studies in patients with CHD having laparoscopic surgeries have demonstrated that as the severity of the heart disease increases, so does the morbidity and mortality.

Clinical Pearl

Laparoscopic surgery in patients with complex CHD is most successful when there is continual communication between the surgical and anesthesia teams about the effects of insufflation on the patient's hemodynamics. It may be necessary to decrease insufflation pressures or modify the original surgical plan if ventilation and hemodynamics are negatively impacted.

What medications could be used to induce and maintain anesthesia?

An IV induction is appropriate in this scenario. Care should be taken to avoid acute changes in either SVR or PVR during induction. Ketamine, etomidate, midazolam, fentanyl, and/or dexmedetomidine along with a neuromuscular blocking agent can be utilized. Propofol, if chosen, should be given in incremental or reduced doses to avoid significant decreases in SVR.

Anesthetic maintenance may be achieved with a combination of inhalational agents and narcotics. The use of neuromuscular blocking agents is helpful during intubation and generally requested by the surgeon to achieve optimal surgical conditions. Although for most infants the goal should be to extubate the trachea at the conclusion of the procedure, consideration should be given to the length of the procedure and the infant's preoperative condition.

How should ventilation be managed in this patient?

The degree to which blood shunts and the direction in which it shunts can be heavily influenced by ventilatory techniques. Hypocarbia and the use of high inspired concentrations of oxygen can result in decreases in PVR which promote L-to-R shunting and increased PBF at the expense of systemic blood flow. These patients should be placed on low inspired oxygen concentrations and the $PaCO_2$ should be maintained near 40 mm Hg to avoid pulmonary overcirculation. However, during the period of insufflation, the increased abdominal pressure may decrease gas exchange and require increased minute ventilation and/or oxygen concentration to maintain the desired hemoglobin oxygen saturation and $ETCO_2$. Monitoring of arterial blood gases during this time may help guide ventilatory strategies as they may require frequent attention and manipulation.

Clinical Pearl

Adjusting ventilatory strategies can prove helpful in balancing cardiac shunting during abdominal insufflation.

What are the considerations for intraoperative hypotension and how should it be managed?

In addition to surgically related causes of hypotension such as bleeding and insufflation, cardiac related causes should also be considered. Intracardiac shunting can occur in either direction (L-to-R or R-to-L) and will vary throughout surgery depending on the relative systemic and pulmonary vascular resistances. Factors lowering SVR (e.g., anesthetic agents) will result in increases in systemic blood flow at the expense of PBF, and similarly factors that lower PVR and augment PBF (hypocarbia, increased FiO_2) will consequently decrease systemic blood flow. If the infant is hypotensive, it is imperative to adjust any factors which may be augmenting PBF at the expense of systemic blood flow.

In general, these patients may prove sensitive to the myocardial depressant and vasodilatory effects of inhalation and intravenous anesthetics. If an arterial line is present, an arterial blood gas should be assessed for anemia, acidosis, and calcium levels, and any abnormalities treated. Fluids should be judiciously replaced to account for blood loss and third spacing. If appropriate fluid replacement has occurred and hypotension persists, an epinephrine or dopamine infusion should be available if needed to support the patient's cardiac function. Calcium chloride 10 mg/kg or calcium gluconate 30 mg/kg can also be useful for augmenting cardiac output. Calcium gluconate is preferable for patients who have only peripheral IV access.

If laparoscopy is abandoned for laparotomy and the patient remains hypotensive, inotropic support should be instituted, and plans made to keep the infant intubated postoperatively.

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Clinical Pearl

Anesthetic and/or surgical interventions that affect PVR or SVR will directly impact the relative flows of blood to the pulmonary or systemic circulations. Care must be taken throughout surgery to maintain a relative balance of blood flow between the two circulations.

What are the postoperative considerations for this patient?

If the procedure is completed via laparoscopic approach and the patient has been hemodynamically stable throughout the procedure, it may be appropriate to extubate the patient in the operating room, albeit with the usual caution utilized for a 1-day-old infant. Even so, recovery should occur in the appropriate intensive care environment, either neonatal or cardiac. In a patient with ductal-dependent physiology, recovery (with or without extubation) should preferably take place in an intensive care unit where practitioners are familiar with cardiac physiology and management (e.g., the cardiac intensive care unit). If the patient has been unstable during the procedure, it is advisable to defer extubation.

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