Chapter 4

Transitional Atrioventricular Septal Defect

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Case Scenario

A 4-year-old child weighing 14 kg arrives to the emergency department after being attacked by a dog. He ate lunch shortly before the incident occurred. The child presents with multiple facial lacerations resulting in significant loss of tissue around his nose and upper lip and is scheduled for urgent debridement, washout, and repair.

His family recently emigrated from Central America and he presented to the pediatrician several weeks ago with shortness of breath. Cardiology evaluation included a chest radiograph that was remarkable for cardiomegaly and increased pulmonary markings.

Transthoracic echocardiogram revealed the following:

- *Large ostium primum atrial septal defect with a left-to-right (L-to-R) shunt*
- *Moderate left-sided atrioventricular valve regurgitation*
- *Restrictive ventricular septal defect*
- *Dilated right atrium and ventricle with preserved function*

The child was diagnosed with a transitional atrioventricular septal defect and started on furosemide.

Examination reveals a mildly tachypneic child who is comfortable after receiving morphine. Auscultation reveals a splitting of the second heart sound along with a holosystolic murmur radiating from the apex to the base. Bibasilar rhonchi are present. Vital signs are respiratory rate 42 breaths/minute, blood pressure 80/39 mm Hg, and SpO_2 100% on room air.

Key Objectives

- Identify the anatomic subtypes of atrioventricular septal defects.
- Review the pathophysiology and clinical presentation for atrioventricular septal defects.
- Formulate an anesthetic plan for children with atrioventricular septal defects.

Pathophysiology

What are the common anatomic characteristics of atrioventricular septal defects?

Atrioventricular septal defects (AVSDs) are common congenital heart defects affecting the interatrial and interventricular septa as well as the atrioventricular valves (AVV). There are three common "types" or descriptions for AVSD defects: partial, transitional, and complete. (See Figure 4.1.) An ostium primum type atrial septal defect (ASD) exists in all types of AVSD defects. (See Chapter 5.)

Atrioventricular septal defects occur due to the failure of endocardial cushions to properly develop and migrate to septate the heart during embryonic development; therefore they are also referred to as ***endocardial cushion defects***. The atrial septum primum begins to develop at around day 28 of gestation, growing from the superior portion of the common atrium toward the central endocardial cushions. The septum secundum develops as an atrial infolding to the right of the septum primum. The inferior segment of the atrial septum is an upward extension of the endocardial cushions. It is this inferior segment that fails to extend, leading to the type of ASD found in patients with AVSD defects.

In addition, there is an abnormal, common level of insertion of the AVVs, as well as an elongation of the left ventricular outflow tract (LVOT) due to the anterior displacement of the aortic valve. The anatomic defects associated with the various subtypes of AVSDs are summarized and compared in Table 4.1.

What are the physiologic differences between partial, transitional, and complete AVSD defects?

The physiologic implications of a ***partial AVSD*** are dependent on the patient's age, the size of the atrial defect, and the presence and degree of other medical issues. Isolated partial AVSD defects will, over time, lead to right-sided overcirculation (L-to-R shunting) and pulmonary overload. In the

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Table 4.1 Anatomic Defects Associated with Subtypes of Atrioventricular Septal Defects

Atrioventricular Septal Defect Type	Defect Location	Associated Issues
Partial AV septal defect	***Atrial:*** Primum ASD ***AVV:*** Cleft anterior mitral valve leaflet Two distinct AVV orifices	Tricuspid valve often abnormal Mitral regurgitation due to cleft leaflet
Transitional AV septal defect	***Atrial:*** Primum ASD ***AVV:*** Cleft anterior mitral valve leaflet AV valve anomaly *Ventricular*: VSD (restrictive)	Mitral regurgitation due to cleft leaflet
Complete AV septal defect	***Atrial:*** Primum ASD ***AVV:*** Common AV Valve *Ventricular*: VSD (nonrestrictive)	Mitral regurgitation due to cleft leaflet Classified according to Rastelli types A, B, C

Adapted from Adler A. C., Chandrakantan A., and Litman R. S. (eds.), *Case Studies in Pediatric Anesthesia*, 1st ed. Cambridge University Press. With permission.

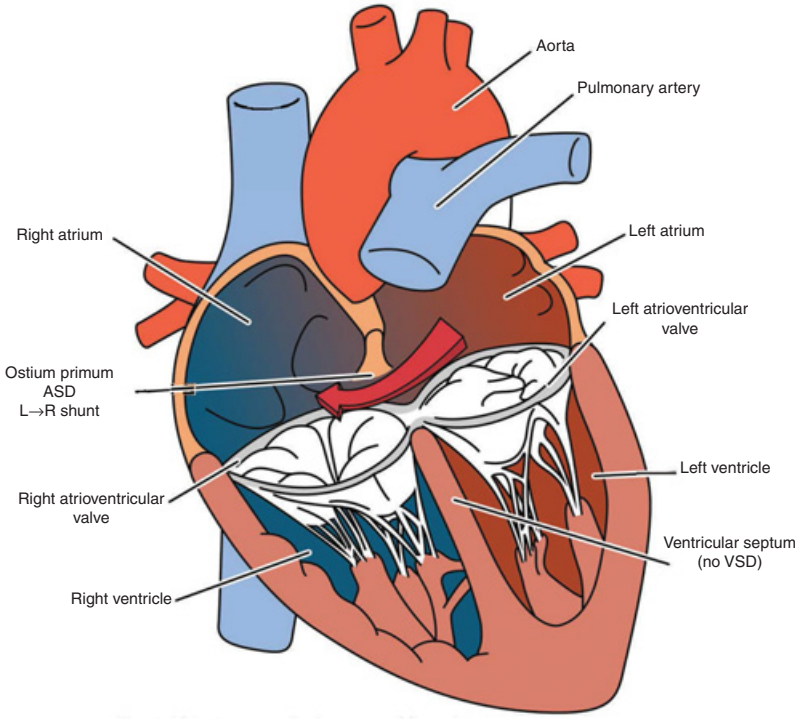

Figure 4.1 **Partial atrioventricular septal defect.** Drawing by Ryan Moore, MD, and Matt Nelson.

presence of pulmonary hypertension or right ventricular outflow tract (RVOT) obstruction, the degree of L-to-R shunting may be reduced or eliminated. (See Figure 4.1.)

In addition to a partial AVSD defect, ***transitional AVSDs*** include a small and restrictive VSD that is occluded by chordal attachments. Generally, a transitional AVSD is not associated with physiologic implications of greater significance than those seen with a partial AVSD. The right atrium and ventricle are typically enlarged and, with time, the increased pulmonary blood flow (PBF) may produce changes in the pulmonary vasculature. As a response to prolonged and excessive PBF, changes occur in the pulmonary vascular musculature that can result in pulmonary vascular occlusive disease (PVOD). If untreated, flow reversal ultimately occurs when right-sided pressures exceed left-sided pressures. This is known as Eisenmenger syndrome

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and patients become cyanotic due to the right-to-left (R-to-L) shunting.

In ***complete AVSD*** the VSD is large and unrestrictive, and therefore significant L-to-R shunting occurs at both the atrial and ventricular levels, with right ventricular and pulmonary artery pressures that are the same as systemic pressures. This produces symptoms earlier in the course of disease and hastens both diagnosis and need for treatment.

What cardiac anomalies of the AV valves and LVOT are associated with AVSD defects?

In patients with AVSDs the left and right AVV have the same level of insertion. This occurs due to a downward displacement of the left AV valve to the level of the septal leaflet of the right AV valve. Secondary to this, the distance between the cardiac crux to the left ventricular apex is decreased and the distance between the apex and the LVOT increases giving the characteristic "goose-neck" deformity. The elongation and narrowing of the LVOT in conjunction with the chordal attachment to the septum increases the risk of subaortic stenosis and coarctation of the aorta.

Clinical Pearl

Patients with AVSDs have an anterior displacement of the aorta, increasing the elongation and narrowing of the LVOT. It is important to evaluate for the presence of LVOT obstruction and subaortic stenosis by echocardiography.

What is the characteristic defect of the AVV in patients with AVSD defect?

The AVV is abnormal, with a coaptation defect frequently referred to as a cleft. The presence of a cleft results in regurgitance, occurring mainly at the left-sided AVV.

What are the cardiac surgical considerations for a patient with an AVSD?

In general, the timing of cardiac surgical intervention is dependent on the type of AVSD and patient symptomatology. The presence of congestive heart failure (CHF) and/or failure to thrive not amenable to medical management are indications for surgery. Surgery requires the use of cardiopulmonary bypass (CPB), allowing for patch closures of the atrial and ventricular level defects and repair of the AVV. The most technically challenging part of this procedure is addressing the common AVV in cases of complete ASVD. The valve is suspended and divided into right and left AV valves. Lesion-specific complications include residual atrial or ventricular septal defects, conduction pathway anomalies, and valvular regurgitation/insufficiency.

What are the clinical implications of AVSDs?

In both partial and transitional AVSDs, medical and surgical treatment is dictated by the degree of L-to-R shunting and the presence of other cardiac anomalies. Generally, there is significant flow from the left atrium to the right atrium, but cardiac catheterization usually demonstrates RV pressures that are less than 60% of systemic pressures. Patients are often managed with diuretics and fluid restriction and typically remain asymptomatic for years. The most common scenario involves deferral of repair until the patient is older, avoiding the necessity for CPB early in development.

A subset of patients with AVSDs, especially those with complete defects, may present with early symptoms of heart failure, necessitating more urgent repair. The severity of clinical presentation depends on the size of the L-to-R shunt, the degree of regurgitant volume at the level of the AVV, and the presence of other cardiac anomalies. These patients have higher mortality after repair and generally require more reoperations on the left AVV. Patients with trisomy 21 are at particular risk for early development of pulmonary hypertension (PH) and resultant PVOD. Early surgical management may include placement of a pulmonary artery band to restrict PBF and reduce L-to-R shunting by reducing the pressure gradient between the right and left ventricles. This procedure is more likely to be performed on smaller, sicker infants as a palliative procedure until complete repair is possible.

Interestingly, the presence of syndromic associations (trisomy) is more common in patients with complete AVSDs when compared to those with partial AVSDs. The majority of AVSDs are complete (56%–75%).

Clinical Pearl

Partial and transitional AVSDs have similar physiologic implications and are often diagnosed later in life as L-to-R shunting is not as significant. Patients with complete AVSDs have more significant L-to-R shunting with more severe pulmonary overcirculation, usually necessitating earlier intervention. Patients with trisomy 21 are at particular risk for early development of PH and resultant PVOD.

What findings are commonly seen in patients with AVSDs?

In general, the presence of an isolated ostium primum ASD is discovered by auscultation of a holosystolic murmur due to high pulmonary flow, and a mid-diastolic rumble may be noted as well depending on the degree of AVV

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regurgitation present. Due to pulmonary overcirculation patients may appear dyspneic or tachypneic, especially with exertion or during feeding.

Electrocardiography may demonstrate a prolonged P-R interval due to the downward displacement of the conductive tissue (AV node) and the enlargement of the right atrium. A chest radiograph may show cardiomegaly with increased pulmonary markings and a characteristic "goose-neck" appearance due to the anterior displacement of the aorta.

What are the important echocardiographic and angiographic findings?

Echocardiography is the gold standard for diagnosis of AVSDs. The ASD is usually large and appreciated from the apical and subcostal four-chamber views. Despite cavity enlargement, biventricular function in these patients is generally normal. The degree of regurgitation of the AV valves should be evaluated as well as an estimate of the RV pressures from the right-sided AV valve insufficiency jet. The position, size, and degree of shunting at the ventricular level should be well characterized, and systematic evaluation of the LVOT for the detection of narrowing, obstruction, or the presence of subaortic stenosis should be well defined. Three-dimensional echocardiography may be employed to determine the mechanism of AVV lack of coaptation in order to facilitate surgical planning.

Angiography and cardiac catheterization are generally reserved for patients in whom there is a clinical or echocardiographic suspicion of PH or PVOD in order to determine the likelihood of successful closure of the septal defect.

Clinical Pearl

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Anesthetic Implications

What are the anesthetic implications for patients with unrepaired AVSDs?

Preoperative evaluation should focus on the type of AVSD, the degree of pulmonary overcirculation, and a detailed review of echocardiographic findings. The presence of coexisting anomalies (e.g., trisomy 21) should be considered. Functional capacity should be elucidated, as it helps to understand the degree of heart failure due to pulmonary overcirculation.

Intraoperative management is also dictated by the degree of pulmonary overcirculation, biventricular function, and comorbidities. Generally, an inhalation induction of anesthesia is well tolerated in the absence of PH. In the presence of severe heart failure, an intravenous (IV) induction is preferred, with the agent of choice depending on the degree of heart failure and severity of AVV regurgitation. In the presence of normal ventricular function, anesthetic agents that reduce systemic vascular resistance (SVR), such as sevoflurane and propofol, are usually well tolerated. In the patient with a large L-to-R shunt, the use of a low inspired oxygen concentration and mild hypoventilation increases PVR and improves systemic cardiac output.

A most important consideration is the meticulous de-airing of IV lines and the use of filters, as air bubbles may enter the arterial circulation. This risk for paradoxical embolism is increased if the shunt reverses to R-to-L. Intravenous fluids should be administered judiciously to avoid pulmonary edema from increased pulmonary flow while also recognizing that many of these patients are on chronic diuretic therapy and that commonly used anesthetic agents lead to vasodilation and reduced preload. Additionally, special consideration is required during periods of desaturation as intraoperative desaturation in patients with complete AVSDs may be the result of pulmonary pressure elevation promoting R-to-L shunting. Finally, patients with large left AVV coaptation defects and significant regurgitation benefit from reduced afterload.

In general, the use of standard American Society of Anesthesiologists recommended monitoring is sufficient. Consideration may be given to the use of invasive arterial blood pressure monitoring in cases where large fluid shifts or blood loss are anticipated, and/or for patients with ventricular dysfunction.

Clinical Pearl

Intraoperative management is also dictated by the degree of pulmonary overcirculation, ventricular function, and comorbidities. Important considerations include the meticulous de-airing of IV lines to avoid the risk of air emboli.

What specific considerations exist for this patient?

This patient, having eaten immediately prior to the incident, has not met the nil per os (NPO) standard and should

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therefore have an IV rapid sequence induction of anesthesia. A specific and detailed examination of the child's airway and injuries is important prior to induction of anesthesia, realizing that it may be difficult to get a good mask fit due to the extent of the injuries. As the child has received morphine in the emergency department, a peripheral IV is likely already present and may be utilized to provide additional anxiolysis if necessary, at the time of parental separation.

As the echocardiogram shows preserved ventricular function, propofol could be used for induction along with either succinylcholine or rocuronium for neuromuscular blockade. A multimodal approach to analgesia would be appropriate, with the use of intravenous acetaminophen and local anesthetic by the surgeons.

Are there special considerations for postoperative care?

Following an uneventful intraoperative course, patients with unrepaired partial or transitional AVSDs without signs of CHF may recover in the post-anesthesia care unit with standard monitoring and postoperative discharge or admission. Home medications should be continued. If large fluid shifts occurred or a large volume of fluid or blood transfusion was required, cardiology consultation may be prudent.

In patients with complete and unrepaired AVSDs, or those with preoperative signs of heart failure or indicators of PVOD, admission to the intensive care unit may be warranted, particularly after major surgeries. Following prolonged surgical procedures or procedures with expected large fluid shifts, patients should be followed closely because of the risk of pulmonary volume overload and an increased likelihood of postoperative oxygen requirements in these patients.

Suggested Reading

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