Chapter

61

Atrial Septal Defects

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A six-year-old female presents for annual physical examination.

Her current vital signs are: blood pressure 98/62 mmHg; heart rate 97/min; respiratory rate 15/min; SpO_2 100% on room air. On auscultation, there is a systolic murmur with a split S2. Upon further questioning, the mother reports that she does have some shortness of breath with strenuous activity.

Chest X-ray reveals prominent pulmonary vascular markings and mild enlargement of the right-sided cardiac silhouette. An EKG is performed showing right axis deviation and right ventricular hypertrophy. The patient is sent for transthoracic echocardiography revealing a 7 mm secundum type atrial septal defect.

What Is the Incidence of Atrial Septal Defects (ASDs)?

The incidence of atrial septal defects (ASDs) is approximately 50–100 per 100,000 live births. ASDs are the third most common congenital heart defect and have a female predominance. While most cases are sporadic, a familial inheritance appears in affected families. While most ASDs occur in isolation, they may occur in association with many genetic syndromes, e.g., Down, Holt–Oram, Noonan among others, or as part of more complex congenital heart disease (CHD).

What Is the Most Likely Age for Presentation?

ASDs may occur as isolated defects or as part of a larger continuum of congenital structural defects. It may take decades before an unrepaired ASD causes symptoms that necessitate cardiac evaluation. ASDs can often be diagnosed early in life by an astute clinician auscultating a murmur prompting further evaluation. ASDs associated with larger congenital heart defects, e.g., complete AV canal defects, are often diagnosed in the early neonatal period or prenatally.

Identify the Four Main Types of Atrial Septal Defects

ASDs are classified based on the anatomic location of the defect (Figure 61.1). The main types of atrial septal defects are:

- Primum
- Secundum
- Sinus Venosus (SV)
- Coronary Sinus

Describe the Formation of the Atrial Septum

The atrial septum primum begins to develop at around day 28 of gestation. It grows from the superior portion of the common atrium toward the central endocardial cushions. 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. The foramen ovale, a normally occurring atrial septal defect in fetal development, is a small opening in the septum primum.

Describe the Defect Associated with a Secundum Type ASD

A secundum ASD is really a defect in the septum primum. There are often one or more small holes in the septum primum in the center of the fossa ovalis. Secundum ASDs are the most common atrial septal defect (Figure 61.2).

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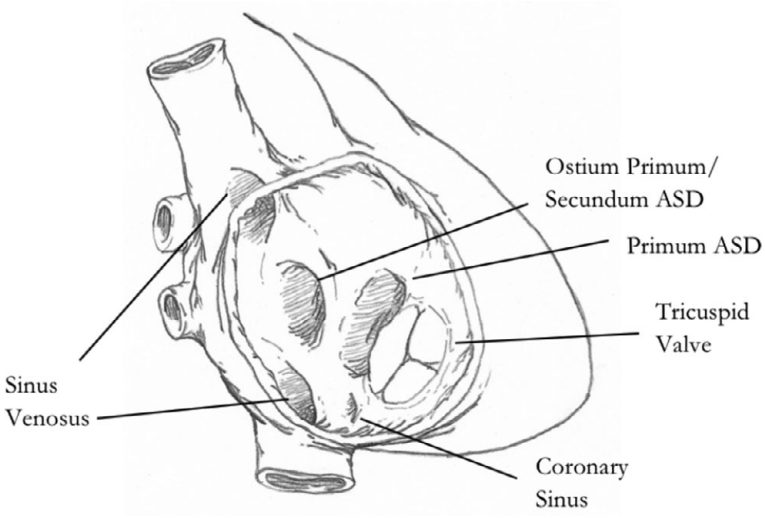
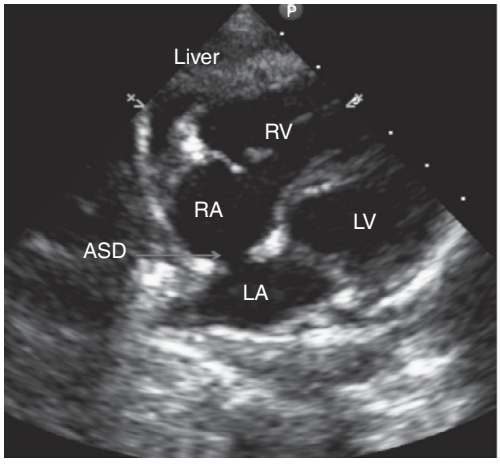


Figure 61.1 Drawing highlighting the types and locations of the four types of atrial septal defects. © Adam C. Adler, MD, drawn by J. Daynes



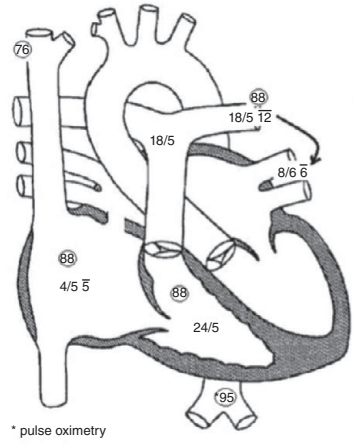


Figure 61.2 Left: Subcostal transthoracic echocardiography demonstrating a secundum type ASD. Right: Cardiac catheterization pictorial of a patient with a moderate ASD. There is a "step up" in saturation from the superior vena cava (76%) to the right atrium (88%).

Describe the Defect Associated with a Primum ASD

A primum ASD, because of its association with the endocardial cushions, may involve a cleft within the anterior leaflet of the mitral valve. If severe, this defect may be addressed during surgical intervention (Figure 61.3).

Describe the Defect Associated with a Sinus Venosus ASD

This defect does not actually involve the atrial septal tissue. Sinus venosus ASDs are actually a connection between the right-sided pulmonary veins and the superior vena cava (SVC) or right atrium. Sinus venosus ASDs account for <10% of ASDs. The most common defect involves connection between the right upper pulmonary vein and the SVC. This defect is a left-to-right shunt at the atrial level as oxygenated blood returning from the lungs is directed to the right atrium instead of the left.

Describe the Defect Associated with a Coronary Sinus ASD

Similar to a SV ASD, a coronary sinus ASD is not a defect in the atrial septal tissue. Rather, the coronary

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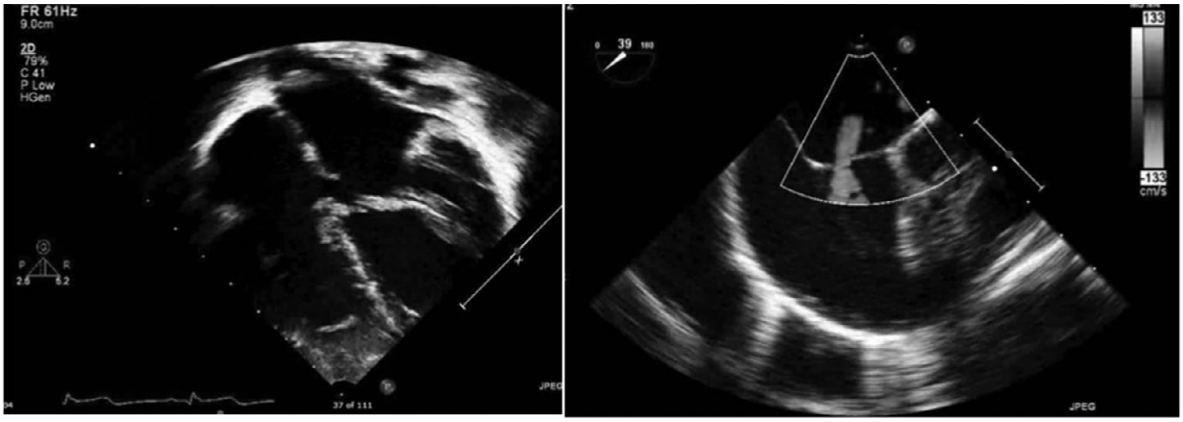

Figure 61.3 Transthoracic echocardiogram demonstrating a primum ASD (left) with large left to right shunting across the defect (right)

sinus is unroofed to drain into the left atrium. Normally, the coronary sinus directs the venous return from the heart into the right atrium. In coronary sinus ASDs, the coronary sinus is opened to the left and right atria allowing the two atria to communicate through this structure. This generates left-to-right shunting at the atrial level.

When a Coronary Sinus ASD Is Identified, What Other Anomaly Must Be Excluded?

Coronary sinus ASDs are associated with a persistent left superior vena cava (LSVC). An LSVC, is a normal structure during fetal life generally in involutes during fetal development. When present, it often communicates with the coronary sinus and/or left atrium. If the coronary sinus communicates directly with the left atrium, a right-to-left shunt exists. Often, a bridging vein between the right and left SVC exists. Identification of a persistent LSVC is important when planning the strategy for cardiopulmonary bypass as the LSVC can provide unwanted venous return and obscure the surgeon's view. Typically, the surgeon will choose to snare the LSVC, especially in the presence of a bridging vein, and direct the flow toward the SVC cannula.

What Is the Pathophysiology of an ASD?

An ASD serves as a left-to-right shunt. The degree of shunting is determined by the size of the defect, relative diastolic compliance of the ventricles, presence of pulmonary valve stenosis, and pulmonary vascular resistance.

What Are the Major Hemodynamic Consequences of an Unrepaired ASD?

With an ASD, both atria and the right ventricle are subjected to chronic volume overload. This results in atrial and ventricular dilation resulting in right ventricle (RV) dysfunction and atrial arrhythmia. The pulmonary vasculature also undergoes changes from the chronic volume overload resulting in muscularization of the pulmonary vascular bed and elevated pulmonary vascular resistance. Over time, elevated pulmonary vascular resistance results in RV pressure elevations and hypertrophy. In severe cases, this can result in reversal of flow (right-to-left shunting) across the ASD, a condition known as Eisenmenger syndrome.

Describe the Appearance of a Chest X-Ray in a Patient with a Large and Unrepaired ASD

Generally, these patients will have right ventricular hypertrophy and enlargement of the cardiac silhouette, especially of right-sided structures. There may be prominence of pulmonary vasculature due to pulmonary overcirculation.

What Is the Characteristic Murmur in Patients with an ASD?

The most characteristic murmur in patients with an ASD is the fixed split S2. The S2 sound (closure of the semilunar valves) is physiologically split during inspiration. During inspiration, the increase in venous return

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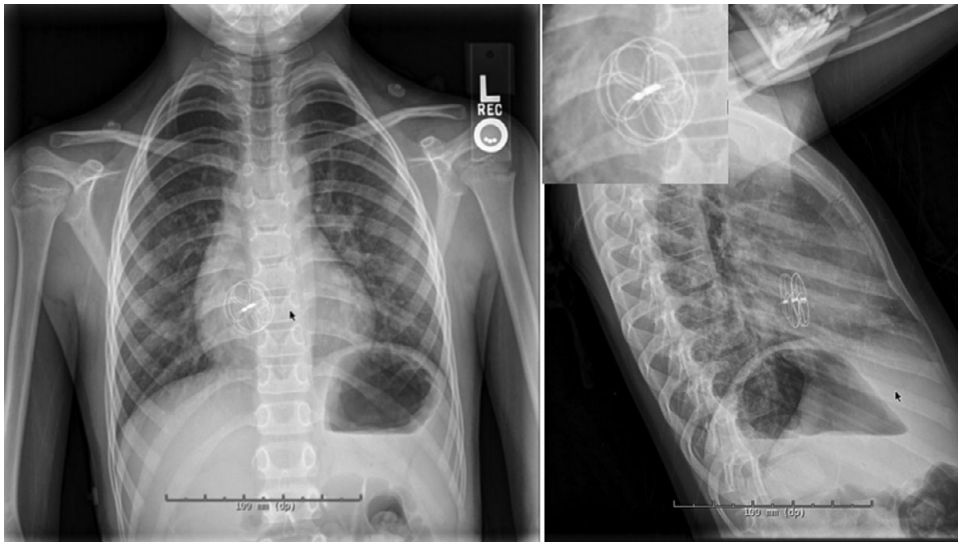

Figure 61.4 Chest X-ray demonstrating an Amplatzer device in situ

overloads the right ventricle and delays the closure of the pulmonary valve. In patients with an ASD, the RV is continuously overloaded from left-to-right shunting, producing a widely split S2.

What Is the Natural History of Unrepaired ASDs?

Secundum ASDs <5 mm may close spontaneously. Primum, large secundum, sinus venosus, and coronary sinus defects generally do not close and usually require intervention.

Describe the Two Main Invasive Approaches for Closure of ASDs

Common approaches for closure of ASDs include surgical closure or catheter-based device closure.

Secundum type ASDs are generally the only type that are amenable to catheter-based device closure at present, most commonly using an Amplatzer device (bilobed disk) (Figure 61.4). A preintervention echocardiogram is performed which is often a transesophageal echocardiography (TEE) to evaluate the rims on either side of the defect. Intraoperatively, a TEE is used throughout the procedure to: evaluate the regions surrounding the defect, guide device positioning, evaluate the atrial septum for evidence of residual shunting, and assure surrounding structures, e.g., AV valves, are intact.

Surgical intervention is done using cardiopulmonary bypass most commonly with bicaval cannulation to optimize visualization. The ASD is approached by right atriotomy and closed using an autologous pericardial patch.

Repair of a sinus venosus defect requires the surgeon to baffle the anomalous venous drainage across the ASD to the left atrium. This baffle is generally accomplished using either autologous pericardium or the atrial tissue itself to create a baffle shunting this blood across the area of the ASD without suturing the pulmonary vein itself. The ASD is closed to the right of the baffle, reestablishing atrial isolation.

A coronary sinus defect (unroofed coronary sinus) is closed by a "roofing" procedure. The coronary sinus is exposed through the right atrium and generally requires excision of the atrial septum. The coronary sinus, which is freely open to both the right and left atria, is closed or "roofed" on the left atrial side using pericardium. The atrial septum is closed leaving the coronary sinus draining solely to the right atrium as intended.

What Are the Major Anesthetic Considerations for Catheter-Based Repair of ASDs?

Aside from the routine risk associated with any catheter-based intervention, the primary risks include damage to the atrial septum (and creation of a larger ASD), device migration/embolization, and damage to the aortic valve. Prior to placing a transcatheter ASD device, an echocardiogram is performed to evaluate

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the retro-aortic rims. Insufficient rims are associated with long-term erosion into the aortic valve. Long-term issues following transcatheter device closure include: local tissue erosion, arrhythmias, endocarditis, and in situ device thrombosis.

What Are the Major Anesthetic Considerations for Surgical Repair of ASDs?

Generally, patients with ASDs are asymptomatic and do not have pulmonary hypertension. These patients may be managed with diuretics if significant pulmonary congestion is present. Mask induction is usually performed with premedication as needed. Presence of an LSVC should be noted as it may interfere with cannulation. If the surgeon snares the LSVC, careful and continued examination of the head for signs of venous congestion should be monitored and communicated with the surgeon.

In general, the repair of routine ASD is uneventful. The patient should be prepared for extubation either in the operating room or shortly afterwards in the intensive care unit. Repair of coronary sinus ASDs may result in damage to the conduction system.

Suggested Reading

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