

Atrial Septal Defect

Josh Kailin, Aimee Liou, Iki Adachi

Atrial septal defect (ASD) is a common congenital heart defect, comprising up to 10% of all congenital heart disease. ASDs frequently accompany other CHDs. This chapter focuses on isolated ASDs. Types of defects in the atrial septum include (Figure 8-1):

- **Patent foramen ovale:** persistent flap opening between the septum primum and the limbus of the fossa ovalis.
- **Secundum ASD:** deficiency, absence, or perforation of the septum primum, leaving shunting at the level of the ostium secundum.
- **Primum ASD:** an endocardial cushion defect resulting in deficiency of the AV septum.
- **Sinus venosus ASD:** an interatrial shunt resulting from abnormal incorporation of primitive venous structures into the atrium. This does not represent a deficiency of the atrial septum itself and is usually associated with partial anomalous pulmonary venous return.
- **Coronary sinus ASD:** a rare deficiency of the “roof” of the coronary sinus as it travels behind the LA that allows for interatrial shunting.

Pathophysiology and Clinical Presentation

ASDs allow for shunting between the left and right atria (Figure 8-2). The degree and directionality of shunting is typically determined by relative RV and LV compliance and defect size. Since the RV is typically more compliant than the LV, atrial-level shunting in the absence of other disease is typically left-to-right. Hemodynamically significant left-to-right shunts cause RA and RV dilation.

Patients with ASDs are typically asymptomatic. Large ASDs may occasionally present in the first few years of life with recurrent respiratory infections, generalized fatigue, failure to thrive and tachypnea; or in the 4th to 5th decade of life, when symptoms of right-heart failure develop secondary to chronic right-heart volume overload. Patients may develop atrial arrhythmias or pulmonary vascular disease in late adulthood.

Physical exam reveals a murmur of ejection quality in the pulmonary valve position, a result of extra flow across the pulmonary valve from the left-to-right shunt. The second heart sound may be widely split (fixed split S₂). The apical impulse may be diffuse and laterally displaced. Adult patients with a longstanding ASD may have findings of right-heart failure including hepatomegaly and edema.

Diagnosis

- **CXR.** Cardiomegaly is the result of RA and RV enlargement. The pulmonary vascular markings can be prominent secondary to the left-to-right shunt.
- **ECG.** RA enlargement, RV hypertrophy, and frequently an RSR' pattern in lead V1.
- **Echocardiogram (Figure 8-3).** Confirms diagnosis, size, and type of ASD.

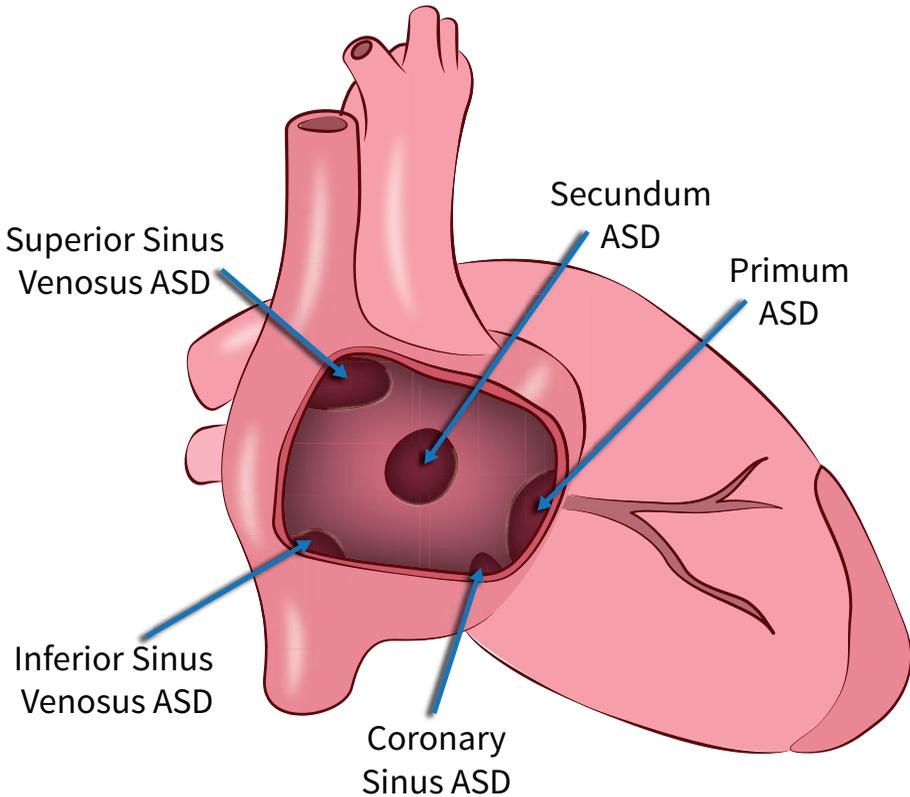


Figure 8-1. Types of ASD. Image courtesy of Dr. Josh Kailin, www.pedecho.org.

Indications/Timing for Intervention

Because of asymptomatic presentation and a high rate of spontaneous closure of smaller defects in young children, referral is typically delayed until patients are 4 to 5 years of age. Additional factors may prompt earlier referral. Indications for referral include:

- Moderate-to-large ASD with right-heart enlargement and diastolic flattening on echocardiogram
- $Q_p:Q_s \geq 1.5:1$
- Special circumstances when ASD is associated to other problems such as pulmonary hypertension, mitral stenosis, etc.
- Comorbidities such as chronic lung disease and prematurity
- Frequent respiratory infections
- Sequelae of transient right-to-left shunting including cyanosis, embolism, recurrent transient ischemic attacks (TIA)
- Symptoms including fatigue and exercise intolerance (rare)

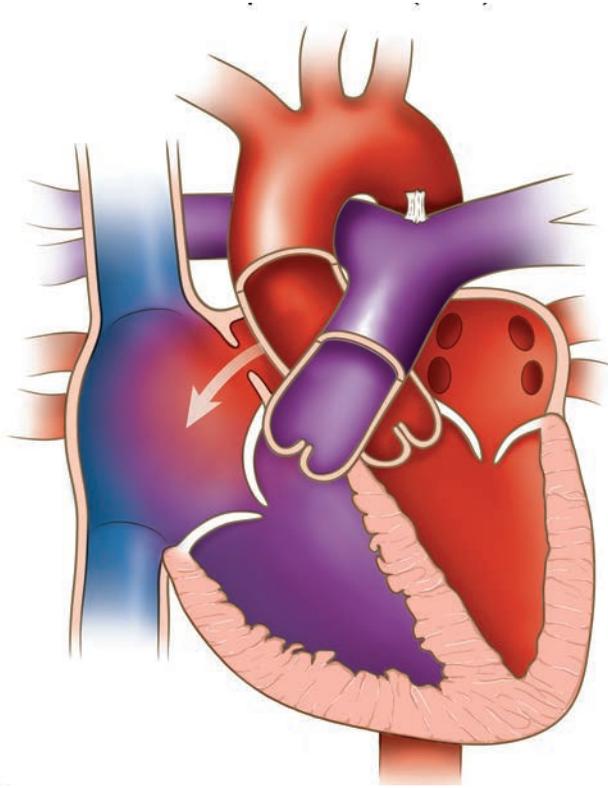


Figure 8-2. Pathophysiology of ASD.

Catheter-Based Intervention

Currently, catheter-based device occlusion is reserved for select patients with secundum ASDs. The transcatheter approach can be used in infants when clinically indicated. In asymptomatic patients without pressing clinical symptoms, allowing the patient to reach a weight of 20 kg can confer additional ease to the procedure.

TEE is performed to fully assess the features of the defect and the feasibility of transcatheter closure; features evaluated include defect size, shape, septal rims, and location of defect. Patients with defects determined by TEE to be unsuitable for catheter-based closure are referred for surgical closure.

The procedure includes a right-heart catheterization with determination of the degree of left-to-right shunting ($Q_p:Q_s$). The defect is then balloon-sized using the “stop flow” technique (Carlson et al. 2005).

A device is placed within the defect using TEE, fluoroscopy, and angiography (Figure 8-4). These imaging techniques allow for guidance and assessment for device malposition, residual shunt, or impingement on neighboring cardiac structures. Following device closure, patients are treated with aspirin for 6 months, after which the device is

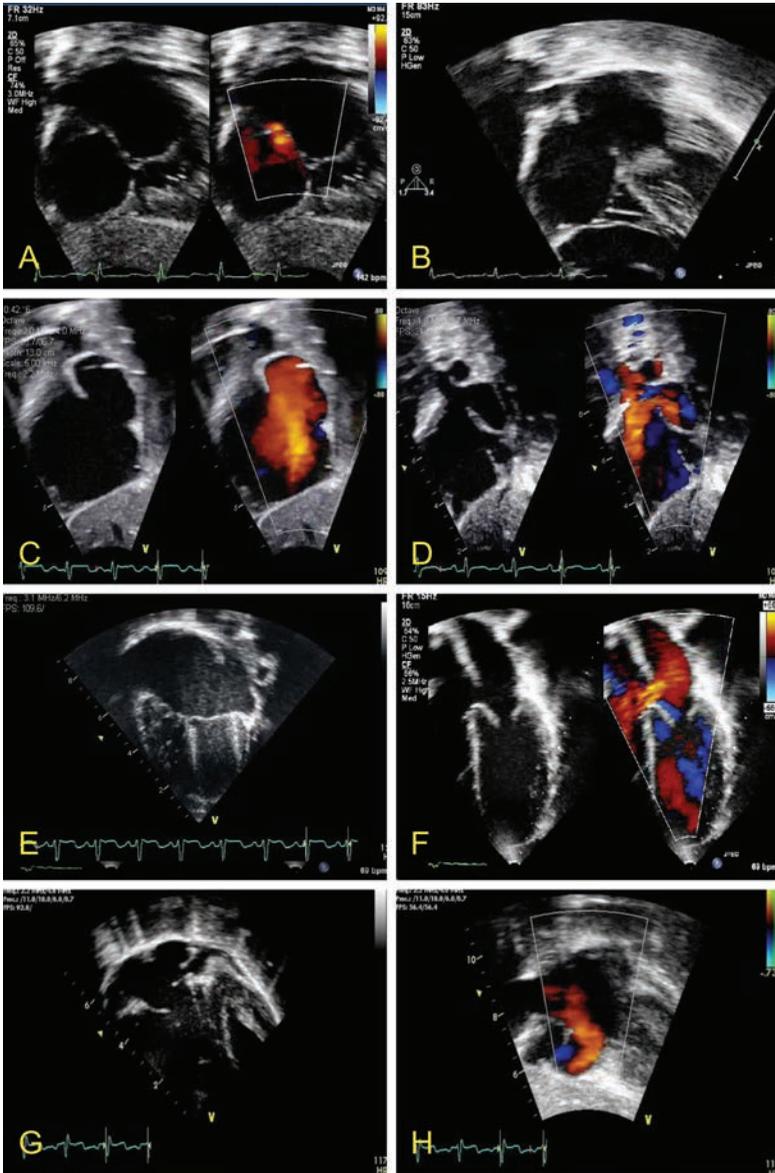


Figure 8-3. Echocardiographic images of ASDs. A) Color-compare image of a PFO with left-to-right shunting, B) 2D image of a large secundum ASD, C) Color-compare image of a large secundum ASD with left-to-right shunting, D) Color-compare image of a superior sinus venosus ASD with left-to-right shunting, E) 4-chamber view demonstrating a large primum ASD, F) 4-chamber view with color-compare showing left-to-right shunting across a primum ASD, G) 4-chamber view demonstrating a coronary sinus ASD due to an unroofed coronary sinus, H) color Doppler image of a coronary sinus ASD showing left-to-right shunting. Images courtesy of Dr. Josh Kailin, www.pedecho.org.

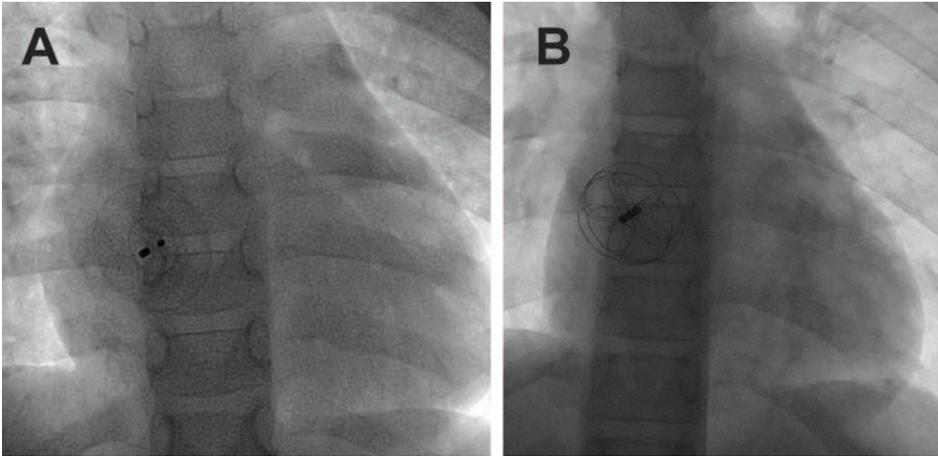


Figure 8-4. Angiographic images of two different ASD devices: A) the Amplatzer™ Septal Occluder (St. Jude Medical Inc., St Paul, MN) and B) the Gore® Cardioform ASD Occluder (W.L. Gore & Associates, Flagstaff, AZ).

likely to have endothelialized; an echocardiogram 6 months after the procedure helps to rule out residual shunt.

Postcatheterization Complications

- **Device embolization.** Can occur secondary to device undersizing or inadequate tissue rims. The device may be retrieved via transcatheter approach or retrieved surgically, with concomitant surgical ASD repair.
- **Device erosion.** May occur secondary to device oversizing or device proximity to the atrial roof or the aorta.
- **Atrial arrhythmias.** Isolated ectopic beats or nonsustained atrial tachycardia can occur but are uncommon.

Surgical Repair

The standard approach for surgical ASD repair is via median sternotomy, although a minimally invasive approach (partial low sternotomy) can be performed in selected secundum ASD cases. CPB is established with bicaval cannulation. The heart is arrested by cross-clamping the ascending aorta while protecting the myocardium with cold cardioplegia administered into the aortic root. The ASD is exposed by opening the RA. Typically, the ASD is closed with a fresh autologous pericardial patch, unless the defect is small, in which case primary closure may be an option.

Surgical Complications

A relatively common complication seen after ASD repair is the development of a pericardial effusion, also known as postcardiotomy syndrome. Patients are often asymptomatic and may present with an increased cardiac silhouette on CXR. Echocardiography is

used to confirm the diagnosis. Depending on the size of effusion and presence or absence of tamponade physiology, treatment includes fluid evacuation (surgical vs. catheter-based) or medical management (diuretics and nonsteroidal anti-inflammatory drugs).

ASD procedures at TCH (2013-2017)

Median number of procedures per year: 37 (33-42)

Perioperative mortality: 0

Long-Term Follow-Up

Patients with unrepaired ASDs require follow-up with a cardiologist. Patients with small ASDs without significant right-heart dilation may follow up every 2 to 3 years. Patients with moderate-to-large ASDs with right-heart dilation should follow up at least annually and sometimes more frequently if <2 years of age to assess for symptoms. Following surgical repair or device closure, patients typically follow up annually with ECG and echocardiogram.

Suggested Reading

Carlson KM, Justino H, O'Brien RE, et al. Transcatheter atrial septal defect closure: modified balloon sizing technique to avoid overstretching the defect and oversizing the Amplatzer septal occluder. *Catheter Cardiovasc Interv* 2005;66:390-6.