

Percutaneous Management of Atrial Septal Defects

Percutaneous closure of ASDs yields results comparable to surgery in appropriate cases.

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Atrial septal defect (ASD) is one of the most common cardiac congenital anomalies. In the appropriate condition, ASDs can be closed surgically or percutaneously with excellent long-term prognosis and minimal morbidity. Recognition of the type of ASD is crucial for the selection of the appropriate treatment strategy. A brief description of the embryology, clinical presentation, diagnosis, and available therapeutic options is presented.

EMBRYOLOGY AND ANATOMIC CONSIDERATIONS

Understanding cardiac development is crucial for the recognition of ASDs and their associated conditions. By the end of the fourth week of gestation, the endocardial cushions fuse to form the left and right atrioventricular canal. The endocardial cushions form the inferior wall of both atria and the primordium of the atrioventricular valves. The common atrial chamber undergoes a complex

process of septation to become the right and left atrium. A simplified explanation of this process follows. The septum primum grows caudally toward the endocardial cushions, closing the interatrial communication (ostium primum). As the septum primum closes, the ostium primum cells in its superior portion undergo apoptosis, forming the ostium secundum. At this time, the septum secundum develops to the right of the septum primum and extends in the opposite direction until the process of septation is complete. In the area of overlap of both septa, a “flap-like” valve, the foramen ovale, is formed.¹

The sinus venosus gives rise to the structures that provide venous return into the right atrium. Throughout pregnancy, oxygenated placental blood flows through the inferior vena cava and is directed by the eustachian valve across the foramen ovale into the left atrium. Deoxygenated blood returns to the heart by way of the superior vena cava. Flow from the superior vena cava is directed away from the interatrial septum and toward

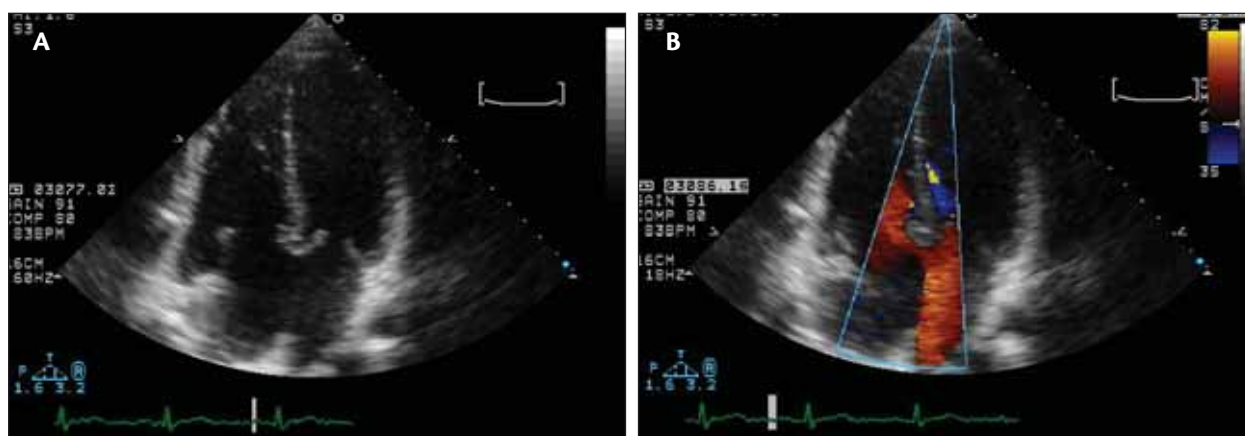


Figure 1. Four-chamber view of a transthoracic echocardiogram with (A) and without (B) color Doppler of an ostium primum ASD. The defect is inferior in location and abuts the atrioventricular valves.

the tricuspid valve by the crista interveniens. The right horn of the sinus venosus incorporates the superior and inferior vena cava into the right atrium.

Any abnormality in the process of septation, incorporation of the sinus venosus into the right atrium, or fusion of the endocardial cushions will generate an ASD. Because the components involved in atrial septation also contribute to the development of other cardiac structures, ASDs may be accompanied by other cardiac anomalies depending on the structure involved. Incomplete fusion of the endocardial cushions will yield an ostium primum ASD (Figure 1). These defects are located inferiorly and are commonly associated with a cleft mitral valve and ventricular septal defects. If fusion of the endocardial cushions is halted in a very early stage, an atrioventricular canal defect results. Excessive resorption of the septum primum or incomplete growth of the septum secundum will give rise to an ostium secundum ASD. Characteristically, these defects are located in the midportion of the interatrial septum involving the fossa ovalis (Figure 2). Excessive resorption in multiple areas of the atrial septum gives rise to a fenestrated septum. Inappropriate blending of the right horn of the sinus venosus will generate a sinus venosus ASD (Figure 3). Sinus venosus ASDs are primarily located in the superior caval-atrial junction. Due to the close relationship of the superior interatrial septum and drainage of the right pulmonary veins, these defects may be accompanied by partial anomalous pulmonary venous return. Inferior caval-atrial ASDs have been described but are rare. Unroofed coronary sinus is another type of ASD that is exceedingly rare and involves an abnormal communication between the coronary sinus and the left atrium.

CLINICAL PRESENTATION

The clinical presentation of patients with an ASD will depend on the degree of shunting and associated cardiac anomalies. Larger defects and those with other malformations will present earlier. Typically, patients with ASDs are asymptomatic during childhood. If large defects are present, children may experience exertional dyspnea, easy fatigability, recurrent heart failure, or repetitive respiratory infections.² In the adult population, ASD with a significant shunt may present with atrial arrhythmias, heart failure, paradoxical embolization, and pulmonary arterial hypertension. If asymptomatic, they may be suspected by the findings of cardiac examination.

DIAGNOSIS

The findings on physical examination typically initiate the evaluation for a suspected ASD. These findings



Figure 2. Bicaval view obtained by transesophageal echocardiogram of a patient with a secundum ASD. The defect lies in the area of the fossa ovalis.

include hyperdynamic precordium, prominent pulmonic component of the second heart sound, pulmonary outflow murmur, or a fixed split second heart sound. Ostium primum ASDs are typically accompanied by tricuspid and/or mitral regurgitation. The electrocardiogram supports the clinical suspicion and suggests the type of ASD present. Lengthening of the PR interval secondary to conduction delay or right atrial enlargement is seen in all types of ASDs.³ Right axis deviation, right ventricular hypertrophy, and an rSR pattern in the right precordial leads without QRS lengthening are suggestive of an ostium secundum ASD. Left axis deviation is typically seen in the ostium primum ASD, whereas inverted P waves in lead III are common in sinus venosus type.

ECHOCARDIOGRAPHY

Echocardiography has become the diagnostic modality of choice to confirm the presence of an ASD. A transthoracic echocardiogram with saline contrast study should be done first. The defect may be visualized directly by transthoracic echo imaging, particularly from a subcostal view of the interatrial septum, but a transesophageal study will provide much better anatomic characterization of the septum and the adjoining structures.^{4,5} Typical echocardiographic findings include right atrium and right ventricle enlargement, increased pulmonary artery pressures and Doppler interrogation demonstrating the presence of continuous flow across the atrial septum.⁶ Negative contrast in the right atrium after agitated saline injection corroborates the diagnosis (Figure 4). Intracardiac echocardiography can also be used to provide guidance for percutaneous closure.

CARDIAC CATHETERIZATION

Cardiac catheterization is performed when a discrepancy exists between clinical and echocardiographic findings, or during a planned percutaneous closure. Oximetry run and assessment of cardiac output will help estimate the size of the shunt and also the location of the defect. Angiography (left atrial injection) will further demonstrate the presence of shunt and other associated anomalies.⁷ The presence of a defect in the atrial septum is usually obvious when the guidewire or the catheter crosses the midline (atrial septum) into the left atrium. The site at which the catheter crosses provides diagnostic clues for the defects. Secundum defects are midseptal, whereas in sinus venosus and primum defects, the catheter crosses the interatrial septum at a high and low level, respectively.

INDICATIONS AND CONTRAINDICATIONS FOR ASD CLOSURE

In stable patients without contraindications for surgery, a pulmonary to systemic flow ratio (Qp:Qs) >1.5:1 is widely accepted as an indication for repair. Patients with smaller ASDs (pulmonary to systemic shunt flow ratios <1.5:1) are referred for closure in the setting of paradoxical embolization or evidence of right ventricular volume overload. It is reasonable to close ASDs in asymptomatic patients with evidence of a dilated right ventricle to reduce the risk of atrial arrhythmias.⁸ Closure is contraindicated in patients with pulmonary vascular resistance >7-8 Woods units.⁹

Currently, closure of ASDs can be performed percutaneously or surgically. Percutaneous closure can only be performed in patients with an ostium secundum ASD. In order to qualify for percutaneous ASD closure, the defect must have an appropriate rim of tissue that allows the closure device to firmly attach to the atrial septum. The

rim of tissue stabilizes the device, preventing migration or erosion into the aorta or atrial wall. Percutaneous closure should not be performed if the closure device interferes with the function of the atrioventricular valves or impedes the drainage from the coronary sinus, vena cava, or pulmonary veins. Closure of multiple ASDs can be done percutaneously with more than one device as long as the devices remain stable, do not interfere with surrounding structures, and obliterate shunt flow. If multiple ASDs are close together, a thin-stalked device with a large left atrial disk may be placed to cover these fenestrations.¹⁰

Surgical closure is the preferred treatment for all patients with multiple congenital cardiac anomalies and ASDs other than the ostium secundum variety. Patients with ostium secundum ASDs that cannot be closed percutaneously in a safe or effective manner should be treated surgically. For properly selected patients, percutaneous closure has been shown to be as effective as surgery and is accompanied by significantly lower morbidity, complication rate, and in-hospital length of stay.¹¹

PERCUTANEOUS CLOSURE

Percutaneous ASD closure has proven to be reliable, safe, and effective.^{11,12} At the present time, only two devices are currently approved by the FDA for percutaneous closure of ASD: the Amplatzer Septal Occluder (AGA Medical Corporation, Plymouth, MN) and the CardioSeal Septal Occlusion System (NMT Medical, Inc., Boston, MA).¹³ The choice of the device is dictated by operator experience and preference. The Amplatzer Septal Occluder is a self-expandable, double-disc device made from nitinol wire mesh. The two discs are linked by a connecting waist, which corresponds to the size of the ASD. The device's discs and the waist are filled with poly-



Figure 3. Subcostal view of a transthoracic echocardiogram showing echo dropout in the superior portion of the interatrial septum consistent with a superior sinus venosus ASD (A). Color interrogation demonstrates turbulent flow across the interatrial septum (B).

ester fabric that promotes defect closure. Device sizes range from 4 mm to 38 mm. Selection is based on the stretched diameter of the defect. In distinction to the PFO occluder, the left atrium disk is larger than the right atrium disk. The reported success rate of these devices in closing ASD is as high as 98%.^{14,15} The proposed advantages of the Amplatzer device include ease of use, deliverability using smaller-diameter catheters, and ability to retrieve and reposition prior to complete deployment. Furthermore, the device design permits it to self-center across the defect (Figure 5).

The CardioSeal device has a unique design in which two self-expanding umbrellas adhere to the atrial septum by means of spring tension. These umbrellas range in size from 17 mm to 40 mm. The device is conformed by four metal arms covered by Dacron patches that are attached to each other in the center. Each arm is hinged and spreads from the center of the device to support each umbrella. The hinges are designed to relieve stress during cardiac contraction. Initial models suffered from arm fractures (14%) and protrusion of one arm through the defect (32%), thus prompting its revision.¹⁶ The new version sports a self-centering mechanism composed of nitinol springs that join the two umbrellas and a flexible core wire with a pin-pivoting connection. The modified device has been reported to have a lower incidence of arm fractures and is currently being marketed in Europe under the name StarFlex.¹⁷ The CardioSeal device has been associated with embolization and migration when used for large ASDs.

Procedural Details

Femoral venous access is typically used. Sheath size will depend on the size of the device and delivery system utilized. A 6-F, multipurpose catheter is used to direct a .035-inch, 1.5-mm, J-tipped guide across the defect. If the septal defect is more inferior, a Judkins Right catheter may be used to change the angle of approach. Once the defect is crossed, patients should be anticoagulated with unfractionated heparin to maintain an ACT >200 seconds. The guidewire is exchanged for a 260-cm, 1.5-mm, J-tipped Amplatzer Super Stiff guidewire (Boston Scientific Corporation, Natick, MA) and placed in the left upper pulmonary vein. The defect diameter is measured with a dedicated sizing balloon. The stretched diameter of the balloon waist, sufficient to occlude flow across the interatrial septum, is measured. This dimension can be obtained by fluoroscopy or echocardiography. Devices are generally oversized by 1 mm to ensure an appropriate fit. Aggressive oversizing should be avoided because it may increase the chances of subsequent erosion. After the device is selected, the appropriate delivery sheath is

placed. Subsequently, the device is deployed under direct fluoroscopic guidance. Meticulous attention should be paid to avoid suction of air into the delivery system, which is particularly seen when using large-caliber delivery sheaths. Before device release, confirmation of appropriate device seating is imperative. Lack of interference with the function of the mitral and tricuspid valve, as well as drainage from the coronary sinus, vena cava, and pulmonary veins, is corroborated by echocardiography, and the device is released.

The delivery sheath is exchanged for a short sheath, which is pulled once the ACT normalizes. A chest x-ray to confirm device placement is obtained 24 hours after implantation. Patients should receive intravenous antibiotics prior to and after the procedure. Patients may be discharged after a 24-hour admission with mild limitations of physical activity. Patients should receive 325 mg of aspirin prior to the procedure, which should be continued daily for 6 months. Seventy-five milligrams of clopidogrel is given daily for 1 month. Standard subacute bacterial endocarditis prophylaxis should be followed for 6 months. Repeat transthoracic echocardiography should be performed at 6 months to confirm complete closure.

Complications

Procedural complications after ASD closure occur in 4% to 7% of cases but are usually mild.^{16,17} A procedural learning curve and significant device modifications have transformed ASD closure into an effective and safe procedure. Periprocedural complications include air embolism, device migration, cardiac tamponade, device erosion, atri-



Figure 4. Four-chamber view of a transthoracic echocardiogram illustrating the phenomenon of negative echo contrast. As saline contrast injected into the venous system drains into the right atrium, noncontrasted blood enters the right atrium via a secundum ASD, yielding an area of lack of contrast in the right atrium (arrow).

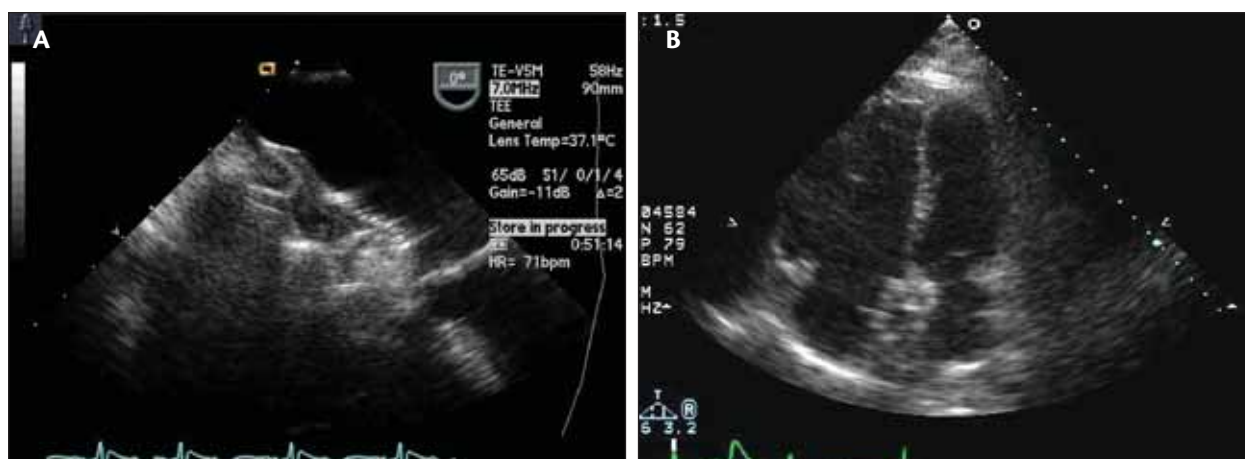


Figure 5. Transesophageal (A) and transthoracic (B) echocardiography depicting appropriate placement of an Amplatzer Septal Occluder. The device does not interfere with the function of the mitral or tricuspid valve.

al arrhythmias, and those related to vascular access.¹⁸ Device-related complications include device migration, device fracture, device thrombosis, and device erosion. Device erosion into the aorta is a dreaded complication that has been associated with a lack of tissue rim surrounding the septal occluder, which prevents the device from abutting the aorta.^{19,20} Device thrombosis and fracture have been described in the older generation of CardioSeal devices. Newer iterations have not been associated with this complication. The presence of device thrombosis, fracture, or migration is treated by surgical removal, with subsequent ASD closure. Bacterial device infection or colonization may prove catastrophic. Surgical excision is warranted if endocarditis ensues.

CONCLUSIONS

Percutaneous closure of secundum ASD has changed our approach to structural heart disease. Percutaneous closure of ASDs is an excellent therapeutic option for the appropriate patient, yielding comparable results to surgery with minimal morbidity and complications. Surgical closure of septal defects is preferred for patients with nonsecundum ASDs and in those with a secundum ASD in which the occluding device interferes with the function of the surrounding structures (pulmonary veins, vena cava, coronary sinus, or atrioventricular valves). ■

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