Atrial Septal Defects in Adults

Todd Greenspoon, Medicine 2010 and Aiman Alak, Medicine 2011
Reviewed by Dr. Keith Finnie

Atrial septal defects (ASDs) are abnormal communications between the left and right atria allowing mixing of the blood between these two compartments. ASDs are the second most common congenital lesions in adults. Embryologically, the septum between the atria is made of septum primum covering the ostium primum orifice, and the septum secundum covering the ostium secundum orifice. In 70% of the population, these septa fuse. A patent foramen ovale exists if the space is covered but the septa are not fused. An ASD exists when an open communication exists between atria. A large ASD can cause extra blood to accumulate in the right atrium and right ventricle. Eventually, the shunted flow causes right side dilatation, main pulmonary artery enlargement, and increase in pulmonary vasculature. Most ASDs are asymptomatic in infancy and present upon routine physical examination. The time it takes for symptoms such as heart failure, generalized edema, exercise intolerance or dyspnea is inversely related to the size of the ASD. On physical examination, the most common findings are a precordial bulge, abnormal murmurs, and extraneous heart sounds. The most useful test in the identification and quantification of the ASD is an echocardiogram (transthoracic or transesophageal, with or without Doppler); however, other investigations including ECG and chest radiography may be utilized. Mechanical closure is indicated for patients that develop symptoms or have a large degree of left-to-right shunt. Historically, surgical closure has been the mainstream treatment, but recently percutaneous devices have come to the forefront in the repair of ostium secundum ASDs because of their excellent outcomes and decreased perioperative morbidity. The transcatheter approach has allowed elderly patients and those with co-morbidities to undergo closure of their ASDs and thrive afterwards.

Case
A 19 year old student presented with an incidentally noted irregular heart beat. Patient worked out regularly, but his aerobic endurance was not that great. He was not a smoker. He had no family history of congenital heart disease. He did not take any medications, and had no allergies. He had no problems with palpitations, light-headedness, or syncope. He had no medical history. He weighed 76 kg. His blood pressure was 110/70 mmHg. His heart rate was 62 beats per minute and regular. His second heart sound was widely split and fixed. His murmur was described as a midsystolic pulmonary ejection murmur.

Chest x-ray revealed cardiomegaly due to right ventricle enlargement. Echocardiography revealed that his right atrium and right ventricle were moderately dilated. His right ventricle had mildly reduced systolic function, and color flow Doppler suggested left-to-right shunting. Transesophageal ECG found a dilated right atrium and right ventricle, moderate right ventricle global hypokinesis, and a 1 cm ostium secundum atrial septal defect with left-to-right shunting.

Patient underwent a percutaneous device closure. Arrangements were made for follow-up in 2 months.

Introduction
Atrial septal defects (ASDs) are the second most common congenital lesions in adults. A patent foramen ovale (PFO) exists in 30-40% of the normal adult population. In an ASD, a defect in the interatrial septum of the heart allows mixing of the blood in the right and left sides of the heart. The extent of hemodynamic and clinical significance depends on the extent of shunting. Embryology: The mother’s placenta provides oxygen for the foetus, so blood can bypass the lungs. In early weeks of gestation, an orifice, ostium primum, exists between the atra. Beginning in the 5th week, the septum primum begins closing the orifice. Within this septum, an orifice called ostium secundum forms, which in turn becomes closed by the septum secundum. The septum secundum does not completely seal, leaving the foramen ovale. The septum primum forms a flexible flop on the left side of the foramen ovale. However, since the right atrial pressure is higher than the left side, the flexible
flap is pushed aside holding the foramen ovale open. At birth, expansion of the lungs and increase in systemic vasculature resistance reverses the atrial pressure gradient; the flap is held down and the interatrial shunt ceases. In approximately 70% of the population, the septa fuse after birth. A “probe patent” or “patent” foramen ovale (PFO) exists if the space is covered but the septa are not fused. A reversal of the interatrial pressure gradient or an intercardiac catheter can open the PFO. An ASD exists when an open communication exists between the atria.

**Pathophysiology:** Many types of ASDs exist, but the pathophysiology is very similar. Normally, the left side of the heart has a higher pressure than the right side. A large ASD can cause extra blood to accumulate in the right atrium and right ventricle. Pulmonary to systemic flow ratio can be as high as 8:1, while some patients can have a 5:1 ratio but be asymptomatic. Eventually, the shunted flow causes right size dilatation, main pulmonary artery dilation, and increase in pulmonary vasculature. Untreated, the increases the size of the right side of the heart can lead to heart failure. Any increase in the pressure in the left ventricle, such as by hypertension and coronary artery disease, worsens the left-to-right shunt. Overload of the right side of the heart, in turn, overloads the pulmonary vasculature, which leads to pulmonary hypertension. The pulmonary hypertension further overload the right ventricle, called afterload, and results in the right ventricle to generate higher pressures to overcome the pulmonary hypertension. Potentially, the right ventricle fails or the pressure in the right side of the heart becomes higher than the left side. As the normal pressure differential between the right and left sides of the heart decreases, the shunting decreases. Eisenmenger’s syndrome describes the situation where severe fixed pulmonary hypertension caused by the shunt leads to elimination of the left to right shunt and bi-directional or right to left shunting leading to arterial desaturation and cyanosis.

**Types of ASDs:** Many types of ASDs exist. Primum ASDs are typically associated with ventricular septal defects and/or AV valve malformations. Poor growth of the secundum septum or excessive absorption of the primum septum is called secundum type ASD. This type accounts for 70% of all ASDs, and is more common in females. It can be associated with other ASDs. Some patients have a family history of this defect. An abnormality in the insertion of the superior or inferior vena cava can form an interatrial communication outside the fossa ovalis. This defect is called sinus venosus ASD. When a part of the wall between the coronary sinus and left atrium is absent, the defect is called coronary sinus ASD. A PFO is not a real ASD. It can be detected in 25 to 40% of normal adult hearts. PFO may be a familial trait.

**Management**

**History and Physical Examination**

The vast majority of small ASDs are asymptomatic in infancy and childhood, and they are found secondary to hearing an incidental murmur upon auscultation of the chest. The minority of newborns with small ASDs may present with mild cyanosis with right to left shunting across the ASD. Typically small ASDs are not associated with a significant shunt and so may not be associated with a murmur or any abnormal findings. Infants and children with larger ASDs may present with heart failure, failure to thrive, or recurrent respiratory tract infections. Patients with moderately sized ASDs, if not corrected, will develop symptoms such as heart failure, hepatomegaly, generalized edema, atrial arrhythmias, exercise intolerance, or dyspnea later in life (usually before age 40) as the degree of left to right shunting increases with age.

There are numerous physical findings associated with ASDs. Children with an ASD may be small for their age, even in the absence of complications such as heart failure. Upon palpation of the precordium there may be a precordial bulge, a right ventricular heave, or a palpable pulmonary artery at the second left interspace. During auscultation of the heart, commonly a widely spaced second left heart sound is present hypothesized to stem from the increase in pulmonary arterial flow and delayed closure of the pulmonic valve. This, however, is usually absent in newborns because of a minimal left to right shunt. If the ASD is associated with
pulmonary hypertension the pulmonary component of the second heart sound is louder than usual. The usually split first heart sound might be even more pronounced in patients with ASDs because of increased blood flow needed to occur across the valve.

A variety of murmurs may be associated with ASDs primarily or secondary to pulmonary hypertension. It is important to realize that the blood flow across the ASD contains insufficient turbulence to be auscultated. The primary murmurs include: a midsystolic pulmonary ejection murmur because of increased flow thorough the pulmonic valve, a mid diastolic murmur secondary to increased flow across the tricuspid valve, a diastolic murmur consistent with pulmonary regurgitation because of pulmonary trunk dilatation, and a systolic ejection murmur best heard over the lungs consistent with increased flow through the smaller pulmonary arteries. Additionally, with pulmonary hypertension patients with ASDs may have additional murmurs and extraneous heart sounds.

Other Investigations: In the evaluation of a patient with an ASD the most utilized tests are the echocardiogram, the electrocardiograph, and the chest x-ray. Echocardiography is the most useful test for the diagnosis of ASD. Transthoracic echocardiography is usually diagnostic; however, transesophageal echocardiography can provide additional information regarding the size of the defect and the other associated congenital anomalies. The volume of blood shunted, the shunt ratios, as well as pulmonary artery pressures can be measured with Doppler flow echocardiography.

Treatment: The two main indications for surgical or percutaneous closure of the ASD are the development of symptoms or a large left to right shunt. The American Heart Association recommends closure when the Qp/Qs is greater than 1.5:1, while the Canadian Cardiac Society recommends closure when the Qp/Qs is greater than 2:1, or greater than 1.5:1 in addition to reversible pulmonary hypertension.

The traditional surgery performed for ASD repairs has been a median sternotomy with Pericardial or Dacron patches, although a right anterolateral submammary subpectoral approach may be preferred by women for cosmetic reasons. There has been an increased use of minimally invasive surgery recently, as an alternative to surgery or percutaneous repair. Intraoperative transesophageal echocardiography may be used to ensure proper closure of the defect. Postoperatively, beta blockers and anticoagulation reduce the risk of atrial fibrillation and stroke respectively. The perioperative mortality of such procedures approaches 0%.

Over the long term, patients still have quite successful outcomes, especially with ongoing medical therapy, with minor differences in survival compared to the population, and minimal risk of needing additional surgery. Minimally invasive approaches may reduce the perioperative morbidity and decreased hospital stay. Dr. K. Finnie adds that closure of the ASD does not reduce the incidence of recurrent atrial arrhythmias such as atrial fibrillation. (Personal communication, 2008)

The FDA has approved two percutaneous devices, Amplatzer Septal Occluder and the CardioSEAL Septal Occlusion System that may be used as an alternative to treatment. The benefits of a transcatheter closure are that the patient avoids cardiopulmonary bypass, thoracotomy, and atriotomy. The outcomes are excellent and the transcatheter closure is largely replacing surgical closure in some centers for the management of appropriate ASDs. Additionally the perioperative morbidity, rate of complications, and hospital stay may be reduced. The percutaneous method of ASD closure will become the more preferred method of closure, especially in elderly patients or those with significant comorbidities.

This nonoperative device closure only can be used in patients with ostium secundum ASDs (about 50-70% of all ASDs) and is not used in patients with ostium primum, sinus venosus, or coronary sinus atrial septal defects. The ASD must also be small or moderately sized, ideally under 20 mm. Dr. K. Finnie adds that defects up to 39 mm can now be repaired. (Personal communication, 2008)

Conclusion
Atrial septal defects (ASDs) are abnormal communications between the left and right atria
allowing mixing of the blood between these two compartments. ASDs are the second most common congenital lesions in adults. Transcatheter closure of ASDs with the Amplatzer Septal Ocluder and the CardioSEAL Septal Occlusion System are streamlining the way ASDs are managed. Decreased hospital stay, decreased morbidity, and outcomes on par or better than with surgery are popularizing the minimally invasive procedure for the repair of specific types of ASDs.

References
1. Weigers SE, Sutton MGJ. Pathophysiology and clinical features of atrial septal defects in adults. UpToDate. 2007 Sep 21; 15.3.