Atrial septal defects (ASDs) are one of the more common congenital lesions encountered perioperatively both in the adults and children. This review article describes ASD anatomy and pathophysiology with an emphasis on echocardiographic assessment of the lesions and their associated problems.

**Morphologically, four atrial septal defects exist: ostium secundum, ostium primum, sinus venosus, and coronary sinus.** ASD pathophysiology involves anatomic and physiologic shunting at the atrial level (1). Physiologic left to right shunting results in recirculation of oxygenated venous blood into the pulmonary artery and back into the left atrium (LA). Anatomically this involves blood flow from the higher-pressure atrium (LA), to the lower pressure right atrium (RA). Mean RA pressure is normally lower than mean LA pressure because right ventricular (RV) compliance exceeds left ventricular (LV) compliance. Usually, RV pressure is less than LV pressure, and pulmonary vascular resistance (PVR) is usually less than systemic vascular resistance (SVR). The left to right flow through the defect diminishes effective forward LV flow and results in volume overload of the RA, RV, and LA(2). Thus, right ventricular volume overload (RVVO) is the hallmark of large left to right atrial level shunts and is easily detected qualitatively with transesophageal echocardiography (TEE)(1,3). Leftward shifting of the interventricular septum during late diastole may be visualized with M-mode echocardiography and is characteristic of RVVO (2,4). Increased right-sided flow often results in dilation of the main pulmonary artery (MPA) and increased pulmonary venous flow. Color flow Doppler (CFD) assists detection of flow across
the interatrial septum. Bidirectional flow indicates elevated RA pressure, which is the consequence of RV dysfunction, tricuspid regurgitation, and/or impaired diastolic function. An important cause of these changes is RV hypertension from the pulmonary vascular remodeling that results from chronic pulmonary overcirculation due to a left to right shunt. These changes are more likely with a ratio of pulmonary to systemic flow (Qp/Qs) > 2:1.

Pulsed-wave or continuous-wave Doppler can be used to estimate the mean and peak pressure gradients between the two atria by the following formula: $\Delta P = (LAP - RAP) = 4 \text{(velocity of flow across septum)}^2$, where LAP is the left atrial pressure, RAP is the right atrial pressure and $\Delta P$ is the difference between the two (5).

**Ostium Secundum Defects**

Ostium secundum defects occur in the fossa ovalis and are the most common ASD (75%) (1). During embryologic development a thin pliable septum primum develops on the left atrial side of a thick muscular septum secundum (fatty limbus). During formation of these curtain-like septa, two gaps or foramen develop, the foramen primum and foramen secundum (6). Eventually both foramen close and the muscular, septum secundum lies on the right atrial side of the thin pliable septum primum, with the septum primum acting as the flap covering the fossa ovalis. Secundum ASDs represent a defect in the embryologic septum primum in the area of the fossa ovalis, which was the original fossa secundum; hence the name ostium secundum. Secundum ASDs represent a hole in the septum primum and this
distinguishes them from a patent foramen ovale that is simply failure of the septum primum (the flap of the fossa ovalis) to completely fuse with the septum secundum.

Morphologically four secundum subtypes exist (2):

- Virtual absence of the septum primum with the ASD representing the entire fossa ovalis
- Deficiency of a portion of the septum primum (ASD represents part of the fossa ovalis)
- Completely fenestrated septum primum where there are multiple small defects in the entire septum primum (“Swiss cheese appearance of fossa ovalis”)
- Partially fenestrated septum primum where part of the septum primum is missing and the remainder is fenestrated

Secundum ASDs are often oval shaped with a 2:1 ratio of the major to minor axis (2). The longest part of the oval or major axis runs cephalad to caudad from the superior vena cava (SVC) to inferior vena cava (IVC) as seen in a midesophageal (ME) bicaval view and usually ranges in size from 4 to 30 mm with a mean of 15 mm. Usually a tissue remnant or rim exists at both the anteroposterior and inferosuperior borders but in large defects the rim may be deficient.

Secundum defects are visualized in the ME bicaval view and in a modified ME aortic valve short axis view between 30 and 60 degrees. The defect is limited to the fossa ovalis and usually a rim is seen surrounding the defect. (Videos 1 & 3)
In the ME bicaval view the defect is seen clearly in the fossa ovalis and this helps distinguish it from a superior sinus venosus defect, which lies cephalad (superior) to the crista terminalis (Figure 1, Videos 1 and 3). Surgical repair of a secundum ASD often involves a patch with pericardium. Alternatively homograft material or woven Dacron patch material is utilized. Small defects may be closed primarily. In patients with a prominent Eustachian valve care must be taken to ensure this is not mistaken for the inferior end of the ASD. Should this mistake occur, the IVC would be baffled to the LA creating a right to left shunt, which may be visualized on perioperative TEE.

Secundum defects often have a surrounding “rim” of septal tissue making them amenable to closure with percutaneous devices (1,7,8). Percutaneous device closure of an ASD is a generally safe and effective alternative to open surgical closure with decreased trauma and shorter hospital length of stay(7). Embolization following transcatheter ASD device placement is a rare (<2%(8), 0.55%(7) ) but potentially lethal complication. Other possible problems following transcatheter ASD device closure include: device erosion (0.1%), residual shunts (<4%), atrial arrhythmias (<5%), device size mismatch (<5%), cerebral infarcts (rare), infective endocarditis, and vascular access complications. (8) The ideal ASD for transcatheter closure is small (<20 mm) with large (>5 mm) firm rims of septal tissue separating the ASD from the surrounding structures (atrioventricular valves, superior and inferior vena cavae, right upper pulmonary vein, coronary sinus). Very large ASDs (>40mm) have been closed with devices, but the catastrophic risk of erosion
(cardiac perforation) increases as device size increases(8). Although the mechanism for erosion is not fully understood, the only known independent risk factor from a survey of all reported erosions was an oversized device (8). Device embolization can cause significant ventricular arrhythmias, obstruction to flow, and valvular insufficiency. Although percutaneous retrieval of stray devices has been reported (9,10), many consider this a cardiac surgical emergency. TEE is useful for determining the location of the stray device, detecting associated pathology (such as LVOT obstruction) and ensuring appropriate ASD closure and de-airing postbypass.

**Sinus Venous Defects**

Sinus venous ASDs are located near either the SVC (superior defects) or IVC (inferior defects) entrance into the RA (Video 3, figure 1). Superior defects are much more common than inferior defects which are rare. Sinus venous defects are often associated with partially anomalous pulmonary venous connections. The distinction between an *anomalous connection* (drainage of a pulmonary vein into a structure other than the LA) and *anomalous drainage* (drainage of normally positioned pulmonary veins across the defect into the SVC/RA junction or IVC/RA junction) must be made.

Both superior and inferior defects are located posterior to the fossa ovalis. Strictly speaking these are not defects in the true atrial septum. These defects are believed to result from a deficiency in the common wall, which normally separates the pulmonary veins from the RA and the SVC rather than a defect in the atrial septum or a change in the position of the pulmonary veins. Unroofing of the posteriorly located pulmonary vein(s) produces drainage of the LA into the SVC and
RA creating a left to right shunt. The interatrial communication is an orifice of the pulmonary veins rather than a defect in the atrial septum. The most common type of anomalous pulmonary connection is the right upper pulmonary vein (RUPV) draining directly into the lateral wall of the SVC above the SVC/RA junction at the level of the right pulmonary artery (figure 2, video 3).

Imaging superior sinus venosus defects is facilitated by multiplane TEE, which is superior to transthoracic echocardiography for visualizing this lesion(1,2). In the ME bicaval view, the superior aspect of a superior sinus venosus defect appears to be the right pulmonary artery due to the absence of the fatty limbus just posterior to the orifice of the SVC (fig 2, video 3). If pulmonary veins are normally positioned the defect may be closed with a pericardial patch. If the pulmonary veins enter the SVC anomalously, then a Warden procedure may be performed(11,12). With the Warden procedure the SVC is transected above the origin of the anomalous vein(s), the proximal end of the SVC is oversewn and the orifice of the SVC is baffled into the LA with a pericardial patch. The distal end of the SVC is then anastomosed end-to-end to the roof of the RA appendage (RAA)(Figure 3)(11,12).

**Ostium Primum Defects (figure 4, Videos 2 and 4)**

Ostium primum ASDs often occur in conjunction with a common atrioventricular valve orifice. Primum defects are associated with inlet ventricular septal defects (VSD), and cleft atrioventricular (AV) valve leaflets. These lesions are commonly seen in patients with trisomy 21. Primum defects result from a defect in the endocardial cushions. A primum ASD with no associated VSD is a partial AV
canal. A primum ASD plus a restrictive inlet VSD constitutes a transitional AV canal, and a primum ASD plus a nonrestrictive inlet VSD constitutes a complete AV canal (CAVC) defect. The ostium primum defect lies posterior and inferior to the fossa ovalis near the AV valves. A characteristic finding of this lesion is insertion of the septal portions of both atrioventricular valves to the IVS at the same level. Normally insertion of the tricuspid valve (TV) to the IVS is inferior (more apical) to that of the mitral valve (MV), producing a ventriculoatrial septum, which separates the RA from the LV. As a consequence, with primum ASDs and atrioventricular (AV) canal defects the valves appear in the same plane and the defect is very close to these valves (Figure 4). Close proximity to the AV valve leaflets helps explain the association of cleft septal tricuspid and anterior mitral valve (MV) leaflets. The cleft anterior MV leaflet is the result of partial fusion of the antero-superior and infero-posterior bridging leaflets, which normally fuse to form the anterior MV leaflet. Occasionally the posterior MV leaflet has a cleft or both mitral valve leaflets contain clefts (13-15). These clefts cause mitral insufficiency and complicate repair of this lesion. The ME 4-chamber view will reveal a defect in the posterior-inferior aspect of the interatrial septum extending to the junction of the AV valves (figure 4). The cleft MV can be appreciated as discontinuity of the anterior MV leaflet in any of the short or long axis views, which optimize imaging of this leaflet. Three-dimensional echocardiography of an en face view of the mitral valve and/or the transgastric basal short axis view assist identification of the mitral valve cleft(s) (13-15). Color flow Doppler imaging can be used to assess the severity of the associated insufficiency by measuring the width of the vena contracta, and jet area as described...
Repair usually requires patch closure of the primum ASD and repair of the cleft AV valve leaflet(s). This mitral repair is normally done with a few interrupted sutures to create continuity of the mitral leaflet(s). Occasionally primum ASDs may be closed with transcatheter devices (video 2), but this is often not feasible due to the proximity of this lesion to the AV valves.

**Coronary Sinus Defects**

Coronary sinus ASDs result from an unroofing of the posterior aspect of the coronary sinus (CS) such that LA blood drains into the CS and through the orifice of the CS into the RA creating a left to right shunt. Coronary sinus ASDs are usually associated with a persistent left SVC and a large dilated coronary sinus (Figure 5 and Video 1). A persistent left SVC is easily detected with transverse plane imaging in a modified view between the ME four-chamber view and the transgastric basal short axis view. The probe is advanced posteriorly or retroflexed and a dilated coronary sinus becomes visible (figure 5). From the mid esophagus (ME), the persistent left SVC can be seen between the LA appendage and the left upper pulmonary vein. A persistent left SVC can be diagnosed with a contrast study with saline contrast injected into the left arm. With a persistent left SVC the contrast will be seen entering the coronary sinus prior to the right atrium (video 4). In the ME two-chamber view and/or ME long axis view, the coronary sinus is seen in short-axis as an echolucent circle on the post aspect of the LA and communication between the dilated CS and the LA may be detected (17,18). If a persistent left SVC is present, repair requires patch closure of the unroofed coronary sinus, which directs
left SVC flow into RA. In the absence of a left SVC, the orifice of the coronary sinus may be oversewn resulting in closure of the LA to RA communication while leaving a very small (<5%) right to left coronary sinus to LA shunt (12). As shown by Joffe et al. (18) the ME 2-chamber view often nicely illustrates the communication of the coronary sinus with the left atrium.

A patent foramen ovale (PFO), the most common defect in the interatrial septum, occurs in 25% of the population (19,20) (Figure 6, Video 1). This may be distinguished from an ostium secundum ASD by the presence of a flap. A secundum represents a hole in the septum primum whereas a PFO represents failure of the septum primum to completely fuse with the septum secundum.

Transesophageal echocardiography (TEE) assists in PFO detection (19,20). Every comprehensive TEE examination should include interrogation of the interatrial septum for a PFO with color flow Doppler and, if necessary, contrast echocardiography. Color flow Doppler examination of the septum should be performed in multiple views, with the color flow Doppler scale decreased to 20-40 cm/s since flow across the septum is low velocity. If color flow Doppler is negative, or inconclusive and ruling out a PFO essential (for example in a patient undergoing left ventricular assist device placement), then a contrast exam should be performed. Agitated saline may function as echo contrast and it should be injected intravenously while imaging the septum. In ventilated patients, this is performed with and without the release of 20-30 cm H2O positive airway pressure. The release of positive airway pressure provokes a transient increase in right atrial pressure, (increasing RAP >LAP) which forces the contrast medium against the septum and
across the defect, if present. Visualization of contrast medium crossing into the left atrium within 3-5 cardiac cycles is consistent with a positive contrast study (20). An agitated saline contrast study with release of positive pressure (as described above) significantly improves PFO detection and should be conducted if color flow Doppler examination is negative and ruling out a septal defect is essential (20). Release of a Valsalva maneuver results in similar findings in non-ventilated patients and is often used with transthoracic echocardiography. Of note, absence of leftward bulging of the interatrial septum during opacification of the right atrium with saline contrast was the most frequent characteristic of a false-negative injection (21).

A patent foramen ovale is associated with a Chiari network and aneurysmal interatrial septum. The prevalence of an aneurysmal interatrial septum is 1%-2.2% and they are associated with a PFO in 50%-89% of patients (19). Patients with an aneurysmal interatrial septum and a PFO are at high-risk for paradoxical cerebral embolus (3-5 times higher risk than patients with a PFO alone) (22-24). Although there is a good study showing that an incidental PFO probably should not be repaired during cardiac surgery (25), patients with an aneurysmal interatrial septum may still benefit because they are at very high risk for paradoxical cerebral embolus(22-24).

Conclusion:

Periprocedure echocardiography assists in confirmation of the diagnosis, placement of catheter closure devices and postoperative assessment of adequate
surgical or device closure. Echocardiography continues to improve and evolve, enhancing our understanding of these lesions in children and adults.

Figure 1
Figure 1 shows a comparison of an ostium secundum ASD and a superior sinus venosus ASD seen in a bicaval view. LA = left atrium, RA = right atrium, ME = midesophageal, Sec ASD = ostium secundum ASD, SV ASD = sinus venosus ASD, ASD = atrial septal defect.
Figure 2
Figure 2 shows a ME Bicaval view showing a superior sinus venosus ASD. LA = left atrium, RA = right atrium, ME = midesophageal, RPA = right pulmonary artery, RUPV = right upper pulmonary vein.
Figure 3 shows the surgical approach for a Warden procedure. With the Warden procedure the SVC is transected above the origin of the anomalous vein(s), the proximal end of the SVC is oversewn and the orifice of the SVC is baffled into the LA with a pericardial patch. The distal end of the SVC is then anastomosed end-to-end to the roof of the RA appendage (RAA). RUPV = right upper pulmonary vein, SVC = superior vena cava, RAA = right atrial appendage, RA = right atrium, RV = right ventricle, LA = left atrium, LV = left ventricle B = area above incision, anastomosed to the right atrial appendage. A = area below incision.
**Figure 4** shows an ostium primum ASD in a midesophageal four chamber view. RA = right atrium, LA = left atrium, RV = right ventricle, LV = left ventricle, ASD = atrial septal defect.

**Figure 5** shows a dilated coronary sinus (CS) in a patient with a persistent left superior vena cava. RA = right atrium, RV = right ventricle, LV = left ventricle.
Figure 6
Figure 6 shows a patent foramen ovale (PFO). RA = right atrium, LA = left atrium, ASD = atrial septal defect.

References:

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