Imaging of Adult Atrial Septal Defects With CT Angiography

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ATRIAL SEPTAL DEFECTS (ASD) ACCOUNT FOR 5% TO 10% OF ALL CASES OF CONGENITAL heart disease and as many as 30% of cases of congenital heart disease presenting in adulthood. These defects make up a spectrum of interesting and distinct entities. The ostium secundum ASD accounts for 70% to 80% of all adult ASDs. Other less common forms of adult ASDs include ostium primum ASD (15% of ASDs), sinus venosus ASD (10% of ASDs), and the unroofed coronary sinus (<1% of ASDs) (1). The complex anatomy of the heart may make differentiation of type, size, and extent of an ASD difficult. The modern post-processing techniques of computed tomography angiography (CTA) are ideally suited for such complicated anatomy and pathology. The type, size, and extent of ASDs can be exquisitely discerned with CTA. Our aim is to provide a practical and image-based resource for review of ASDs in adults, including the rarer variations (Figs. 1 to 7).

Figure 1. Patient #1: Secundum ASD

By far the most common type of ASD is the secundum type, comprising 70% to 80% of all ASDs. This type of defect occurs when there is failure of the septum secundum to cover the ostium secundum (1). These defects always occur at the site of the ostium secundum, which is located centrally in the septum primum (the eventual site of the fossa ovalis). During development, the interatrial septum arises as 2 sequentially appearing septa that eventually fuse to form one septum. In the fetus, the centrally located ostium secundum allows oxygenated blood from the placenta to bypass the lungs by shunting from the right atrium (RA) to the left atrium (LA). In normal development, the second appearing septum covers the ostium secundum and fusion of the two after birth closes this conduit. Patient #1 has a secundum ASD (yellow arrows), allowing concentrated contrast from the LA to shunt into the RA. (A) Four-chamber, (B) short axis, and (C) sagittal computed tomography angiography (CTA) images are shown. A = aorta; AR = aortic root; ASD = atrial septal defect; L = liver; LV = left ventricle; PA = pulmonary artery; PV = pulmonary vein; RV = right ventricle; RVOT = right ventricular outflow tract.

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Figure 2. Patient #2: Superior Sinus Venosus ASD

Compared with the centrally located secundum ASD, the sinus venosus is located eccentrically either superiorly or inferiorly in the interatrial septum at the sites of inflow of the superior vena cava (SVC) and inferior vena cava (IVC), respectively. The superior sinus venosus ASD is far more common than the inferior type. (A) Four-chamber, (B) short axis, (C) sagittal, (D) high 5-chamber, (E) right 2-chamber, and (F) axial CTA images are shown from Patient #2. This figure shows a superior sinus venosus ASD as a defect in the superior aspect of the interatrial septum at the level of entry of the SVC (A to D, yellow arrows). (C) The superiorly eccentric position of the defect (yellow arrow) along the interatrial septum (blue arrow), allowing communication between the LA and SVC, is clearly depicted. (D) The superior location of the ASD (yellow arrow) is further demonstrated as it is seen at the same level of the aortic root. The defect in this patient resulted in volume overload of the right heart chambers, leading to enlargement of the RA, RV, and PA. Partial anomalous pulmonary venous return from the right lung is commonly associated with sinus venosus ASD. The partial anomalous pulmonary venous returns, in this case, drain into the SVC (E, orange arrows). An additional anomaly in this case, although not typical of superior sinus venosus ASD, was a left-sided SVC (F, green arrow) draining into a dilated coronary sinus (A and D, red arrows). LAA = left atrial appendage; other abbreviations as in Figure 1.

Figure 3. Patient #3: Inferior Sinus Venosus ASD

The inferior sinus venousus ASD is depicted in (A) 4-chamber, (B) short axis, and (C and D) sagittal CTA images from Patient #3. A defect is present in the inferior portion of the interatrial septum that forms the floor of the LA (A to D, yellow arrows) near the site of inflow from the IVC, allowing shunting of concentrated contrast from the LA into the inferior portion of the RA and inferiorly into the IVC. (A) This low 4-chamber view shows the inferior location of the ASD as it is seen at the same level of the orifice of the coronary sinus (red arrow), which courses beneath the floor of the LA but anterior to the IVC and ASD. (B) This short axis view depicts ASD flow into the orifice of the IVC (yellow arrow). (C and D) These sagittal views exquisitely depict the location of the defect in the inferior interatrial septum that forms the floor of the LA (yellow arrow). Patient #3 has both a secundum ASD and an inferior sinus venosus ASD, allowing for direct comparison of their differences (D). The smaller secundum ASD (red arrow) is located centrally in the interatrial septum at the site of the ostium secundum and, as the name implies, the inferior sinus venosus ASD (yellow arrow) is eccentrically located inferiorly. Incidentally present is an anomalous left circumflex artery (B, blue arrow) that originates from the right sinus of Valsalva and courses posteriorly to the aortic root. Abbreviations as in Figures 1 and 2.
Figure 4. Patient #4: Unroofing of the Coronary Sinus

An unroofed coronary sinus is a rare cause of communication between the right and left atria in an adult. This defect does not involve the interatrial septum but rather unroofing of the coronary sinus as it courses beneath the floor of the LA. (A) Four-chamber, (B) short axis, and (C) sagittal CTA images are shown for Patient #4. In these images, there is a defect in the floor of the LA at the site where the coronary sinus (CS) courses beneath it (B and C, yellow arrows). The high pressure of the LA forces blood into the low-pressure CS and into the RA, creating a left-to-right shunt. (A) The CS (red arrows), which normally courses beneath the LA to empty into the RA, is shown. Similar opacification of the CS and the LA is a result of direct communication. Abbreviations as in Figure 1.

Figure 5. Patients #5 and #6: Patent Foramen Ovale and Probe Patent Foramen Ovale

A patent foramen ovale (PFO) differs anatomically from an ASD in that it consists of a persistent tunneled fetal-type communication between the atria due to failure of fusion of the septum primum (blue arrows) with the septum secundum (red arrows) rather than a simple defect in the septum. Up to 25% of adults have a PFO (2). A PFO is best visualized on CTA images in the axial plane or in the vertical plane perpendicular to the interatrial septum, where the 2 embryologic components of the interatrial septa are separated and surrounded by intravenous contrast. The axis of the PFO tunnel is always directed toward the IVC, because the foramen ovale (green arrows) serves to conduct oxygenated blood from the IVC into the LA during fetal development. (A) Four-chamber, (B) short axis, and (C) sagittal CTA images are shown for Patient #5, and (D) 4-chamber, (E) short axis, and (F) sagittal CTA images are shown for Patient #6. The images for Patient #5 show the left-to-right shunting of contrast (yellow arrows) through the PFO from the LA to the RA. (B) The entry site of the PFO at the persistent ostium secundum is shown. (C) The sagittal view demonstrates the expected inferior direction of shunting into the RA toward the IVC. Occasionally, there is partial fusion of the septum primum and septum secundum without shunting between the atria. This phenomenon, often called a probe patent PFO, is present in Patient #6. This entity results in visualization of 2 embryological components of the interatrial septum with contrast insinuating between the 2 but without contrast extending into the RA. The images for Patient #6 show intravenous contrast separating the septum primum (blue arrows) and the septum secundum (red arrows); however, there is no shunting of contrast into the RA because this represents incomplete fusion of the septa. Contrast enters this space via the foramen ovale (green arrow). Abbreviations as in Figures 1 and 2.
Another rare type of ASD encountered in the adult is the ostium primum defect. When the atrial septum develops in the fetus, the first septum that arises, the septum primum, grows from the basal wall of the atria toward the apical wall (toward the ventricles). In the ostium primum ASD, this septum fails to reach its destination, and there is persistent communication between the atria with a variable amount of intact septum attached to the basal wall. This defect commonly presents with a proximal ventricular septal defect (VSD). The combination of this ASD and VSD is referred to as the "endocardial cushion defect" because the endocardial cushion is the site where the atrial septum meets the ventricular septum and the mitral valve meets the tricuspid valve. It has also been referred to as the "atrioventricular septal defect" and "atrioventricular canal defect." This type of defect is the most common congenital heart defect associated with Down syndrome.

Figure 6. Patient #7: Primum ASD

As with other disease spectrums, there is occasionally overlap of findings of multiple ASDs in the same patient. The ASD in Patient #8 is complex; (A) mid-level 4-chamber, (B) inferior 4-chamber, (C) short axis, and (D) sagittal CTA images are shown. (A) This mid-level 4-chamber view shows a centrally located ASD (yellow arrows). Basal-sided (green arrow) and apical-sided (red arrows) rims are present. The intact apical-sided rim differentiates this defect from the ostium primum ASD; however, this view alone could be confused for a typical large ostium secundum ASD. (B) This 4-chamber view is more inferiorly located than that in A. At this inferior level, there is no longer a basal-sided rim. The case becomes more complex in the (C) short axis and (D) sagittal views. There is a superior septal rim (blue arrow); however, no real inferior rim is present, typical of inferior sinus venous ASDs. As is also typical of inferior sinus venous ASDs, there is inferiorly directed left-to-right shunting into the IVC oriﬁce. Abbreviations as in Figures 1 and 2.

Figure 7. Patient #8: Complex ASD

REFERENCES
