SECUNDUM ATRIAL SEPTAL DEFECT

Kamran Shaikh, M.D.; Faisal Nabi, M.D.

Methodist DeBakey Heart & Vascular Center, The Methodist Hospital, Houston, Texas

A 38-year-old Hispanic female with a history of hypertension and progressive shortness of breath underwent evaluation that included transthoracic and transesophageal echocardiograms. Images from both identified a secundum atrial septal defect (ASD) with a mildly dilated right ventricle (RV) and preserved RV systolic function. The left upper and lower and right lower pulmonary veins were reportedly identified. The patient was referred to The Methodist Hospital's cardiovascular magnetic resonance (CMR) laboratory for further anatomic and hemodynamic assessment of the defect and possible other associated congenital abnormalities.

A 2.0 x 1.6 cm ASD (Figures 1 and 2, white arrows) was seen in the anterosuperior aspect of the interatrial septum with at least 1 cm rim of tissue in all planes except the retroaortic rim, where no rim of tissue was seen. The RV was mildly dilated at 207 ml but with a normal systolic function (50%).

Quantitation of right- and left-sided cardiac outputs revealed a Qp:Qs ratio of 2.0:1.0 with a total net left-to-right shunt of 4 L/min (Figure 3). However, direct volumetric assessment through the ASD revealed a flow of only 2.1 L/min.

Upon further review of the anatomic images and the magnetic resonance angiography (MRA) of the chest, we identified an anomalous left superior pulmonary vein emptying into the left innominate vein (Figure 4, white arrows). This contributed the remaining 1.9 L/min of the total left-to-right shunt.

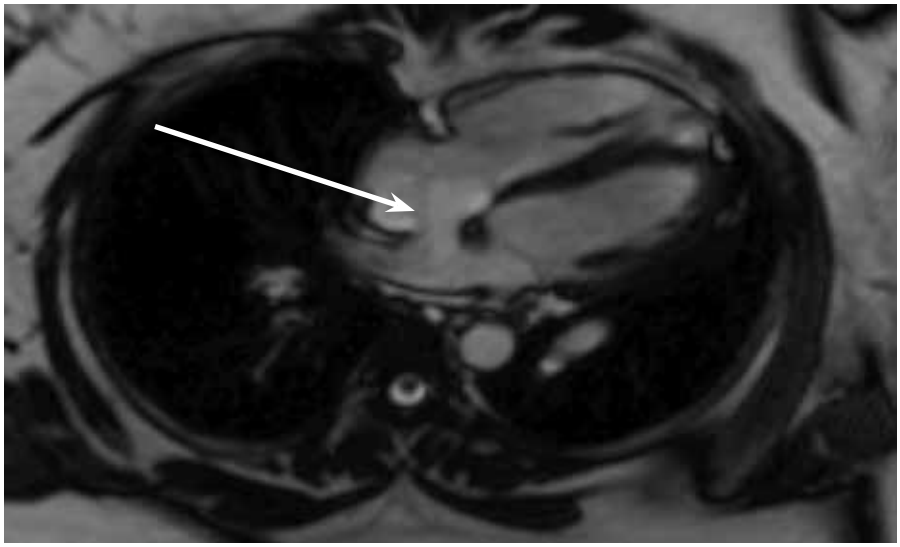

Figure 1. Single-frame SSFP image showing secundum atrial septal defect.
SSFP: steady state free precession

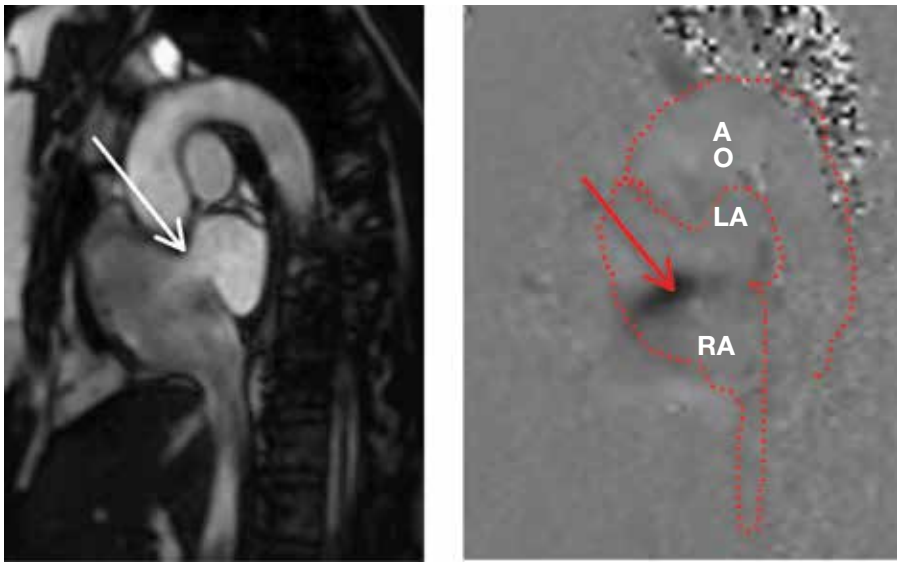

Figure 2. Single frame SSFP and phase-contract images showing secundum ASD (white arrow) with predominant left-to-right atrial shunting (red arrow).
SSFP: steady state free precession; LA: left atrium; RA: right atrium; AO: aorta; ASD: atrial septal defect.

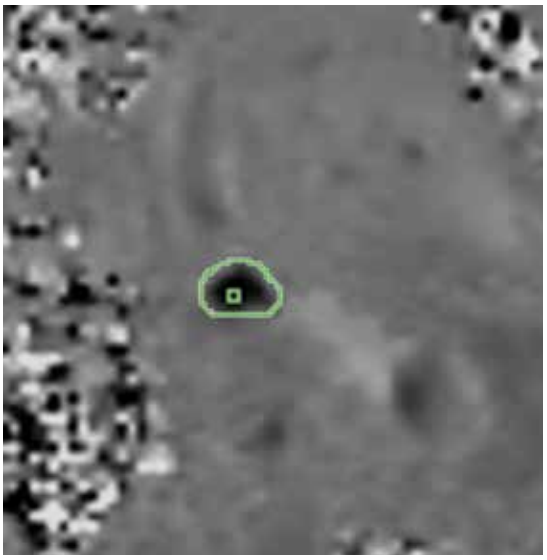

Figure 3. Direct thru-plane phase-contrast assessment of ASD net flow. ASD: atrial septal defect.

Figure 4. Volume-rendered reconstruction of gadolinium enhanced 3D- showing anomalous left superior pulmonary venous drainage.

Discussion

CMR is correctly suited to accurately assess complex congenital heart disease. With its excellent spatial resolution and large field of view (~400 mm), CMR is able to accurately delineate the entire cardiac and thoracic anatomy. As opposed to being limited to specific imaging planes with either transthoracic or transesophageal echocardiography, the ability to visualize in any desired imaging plane with CMR allows for a more complete assessment of structure and anatomic relationships. Highly accurate and reproducible assessment of atrial and ventricular volumes and ventricular function also greatly aid medical decision making. Furthermore, contrast-enhanced MRA allows the accurate, noninvasive description of arterial and venous structures.

Flow quantification techniques also make CMR an ideal imaging modality to assess congenital shunt lesions. Conventional methods to assess Qp:Qs ratios noninvasively measure volume/flow across the aortic and pulmonic valves. This ratio, however, reflects a total shunt ratio. In situations where there may be multiple shunts present, echocardiography may not be able to determine the individual contribution of each lesion. On the other hand, the exact contribution of each individual shunt can be quantified by phase-contrast CMR by direct assessment of flow through each shunt.

As illustrated in this case, direct assessment of a known shunt prompted further investigation for additional shunts since the total shunt fraction and direct flow assessment were not concordant.[1] This resulted in the new identification of a rare anomalous pulmonary vein to the left innominate vein. This had important management implications, as the combination of both a secundum ASD without sufficient rim margins and anomalous pulmonary venous drainage encouraged a surgical corrective approach over a percutaneous approach.[2]

CMR imaging is deemed appropriate for the assessment of complex congenital heart disease including anomalies of coronary circulation, great vessels, and cardiac chambers and valves.[3]

References

1. Thomson LE, Crowley AL, Heitner JF, Cawley PJ, Weinsaft JW, Kim HW, et al. Direct en face imaging of secundum atrial septal defects by velocity-encoded cardiovascular magnetic resonance in patients evaluated for possible transcatheter closure. Circ Cardiovasc Imaging. 2008 Jul;1(1);31-40.
2. Warnes CA, Williams RG, Bashore TM, Child JS, Connolly HM, Dearani JA, et al. ACC/AHA 2008 Guidelines for the Management of Adults with Congenital Heart Disease. A report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (Writing Committee to Develop Guidelines on the Management of Adults With Congenital Heart Disease): Developed in Collaboration With the American Society of Echocardiography, Heart Rhythm Society, International Society for Adult Congenital Heart Disease, Society for Cardiovascular Angiography and Interventions, and Society of Thoracic Surgeons. Circ. 2008; 118:e714.
3. Hendel RC, Patel MR, Kramer CM, Poon M, Hendel RC, Carr JC, et al. ACCF/ACR/SCCT/SCMR/ASNC/NASCI/SCAI/SIR 2006 appropriateness criteria for cardiac computed tomography and cardiac magnetic resonance imaging: a report of the American College of Cardiology Foundation Quality Strategic Directions Committee Appropriateness Criteria Working Group, American College of Radiology, Society of Cardiovascular Computed Tomography, Society for Cardiovascular Magnetic Resonance, American Society of Nuclear Cardiology, North American Society for Cardiac Imaging, Society for Cardiovascular Angiography and Interventions, and Society of Interventional Radiology. J Am Coll Cardiol. 2006 Oct 3;48(7):1475-97.