Anomalous vena caval return to the left atrium (LA) is a rare congenital anomaly. The most common variant is a persistent left superior vena cava (SVC), estimated to occur among 2.1% to 4.3% of hearts with congenital defects.1 Much rarer, an anomalous right SVC connects to the LA.2 Similarly rare is inferior vena caval (IVC) return to the LA, which has been reported as a congenital anomaly often occurring with atrial septal defect (ASD),3 as well as a postcardiac surgical complication, typically caused by incorrect closure of the IVC after repair of an inferiorly located ASD.4 We present multimodality imaging of a case of a right SVC connecting to the LA and a case of an IVC connecting to the LA.

Case Presentation
The first patient is a 45-year-old woman who was referred for evaluation of her increasing shortness of breath and nearly syncopal episodes over a 1-year period. Physical examination revealed a resting hypoxia of 90% on room air and a systolic murmur. Her ECG and chest x-ray were normal (Figures I and II in the online-only Data Supplement). A transesophageal echocardiogram (Figure 1 and Movies I and II in the online-only Data Supplement) revealed a sinus venosus ASD and anomalous right superior pulmonary venous return. Cardiac magnetic resonance imaging demonstrated a right SVC draining the majority of its contents to the LA, an anomalous right upper pulmonary vein draining into the SVC, and an atypical sinus venous defect (Figures 2 and 3 and Movies III and IV in the online-only Data Supplement). Phase-contrast magnetic resonance imaging of the pulmonary arterial and aortic flow found the shunt ratio (Qp/Qs) to be 0.81 (Figure 4). The patient underwent a Warden procedure that involved transection of the SVC above the highest pulmonary vein, closure of the cardiac end, closure of the ASD, and anastomosis of the free end of the SVC to the right atrial appendage (Figure 1 and Movie II in the online-only Data Supplement). She tolerated the procedure well and was discharged to rehabilitation a few days later, asymptomatic and with normalized oxygen saturation.

The second patient is a 62-year-old man transferred to our hospital for open surgical ASD repair. He had a reported history of exertional dyspnea in childhood leading to an open ASD repair in 1964 at 16 years of age at an outside hospital with no records available. Three years after ASD repair, he was diagnosed with polycythemia. An extensive evaluation that included a bone marrow biopsy did not identify a cause. He has been undergoing phlebotomy on a weekly basis for 43 years. His examination revealed oxygen saturation of 89% on room air; scleral, lip, and digital cyanosis without clubbing; and a pansystolic murmur. ECG and chest x-ray were normal (Figures III and IV in the online-only Data Supplement). More recently, he had developed symptomatic paroxysmal atrial fibrillation and underwent an attempted ablation, which...
was aborted as a result of catheter malposition through a suspected ASD. At this time, he was referred to our institution for open closure of what was believed to be a large ASD, but on admission, a cardiac computed tomography (performed before redo sternotomy to scout poststernotomy anatomic landmarks) demonstrated IVC drainage to the LA (Figure 5). Transesophageal echocardiography (Figure 6 and Movie V in the online-only Data Supplement) and cardiac magnetic resonance imaging were performed for quantitative shunt calculations (Figure 4) and additional anatomic clarification (Figure 7 and Movie VI in the online-only Data Supplement). Preoperative right and left heart catheterization demonstrated filling of the LA with contrast injection in the IVC (Figure 8 and Movie VII in the online-only Data Supplement). A brain magnetic resonance imaging was without evidence of any subclinical cerebral infarcts. He has continued warfarin for stroke prophylaxis and is considering surgery.

Discussion

Clinically, most patients with congenital anomalous vena caval return present most commonly at a young age with symptoms of right-to-left shunt. Clinical signs and symptoms include hypoxia, exertional dyspnea, cyanosis, pansystolic murmur, and clubbing. However, these congenital findings have been reported in asymptomatic patients referred for evaluation of incidentally noted hypoxemia or cyanosis.3

Most patients with vena caval drainage to the LA diagnosed either congenitally or after incorrect ASD repair undergo surgery to correct hypoxemia or to reduce the theoretical risk of future arterial embolization. Although a previous diagnostic evaluation including bone marrow biopsy was unable to diagnose the cause of polycythemia, we believe that the second patient developed secondary polycythemia as a result of right-to-left shunt as a complication of the ASD repair in 1964. Less likely possibilities include a missed congenital IVC to LA at the time of his original surgery. Another possibility in this case is a partially dehisced ASD patch, allowing preferential IVC flow to the LA. Finally, his atrial fibrillation, although a common arrhythmia, may also be related to hypoxemia or volume overload secondary to his chronic shunt.
Conclusion

Anomalous vena caval return to the LA is rare, but may occur congenitally or as a complication after cardiac surgery. Magnetic resonance imaging and cardiac computed tomography demonstration of these rare defects is not widely reported.

Disclosures

None. The opinions and assertions contained herein are the authors’ alone and do not represent the views of the Walter Reed Army Medical Center, the US Army, or the Department of Defense.

References

Figure 8. Contrast angiography demonstrating flow from the inferior vena cava (IVC) to the left atrium and ventricle.
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