A 63-year-old man was referred for cardiac evaluation after a chest radiograph for an upper respiratory tract infection demonstrated prominent mediastinal borders (Figure 1). He had a normal clinical examination. A dilated coronary sinus was identified on transthoracic echocardiography. On subsequent transesophageal echocardiography, agitated saline contrast injected into the left antecubital vein first appeared in the coronary sinus, consistent with a persistent left-sided superior vena cava (Figure 2A and Movie 1). Interestingly, saline injected into the right antecubital vein also first appeared in the coronary sinus (Figure 2B and Movie 2). Imaging of the atria confirmed the absence of a right superior vena cava (Figure 2C). Cardiovascular MRI demonstrated that blood from the right side of the body passed via an innominate vein, anterior to the aortic arch (Figure 3A), to the left-sided superior vena cava (Figure 3B and 3C and Movie 3) and then, via the dilated coronary sinus, into the right atrium (Figure 3D and Movie 4). There was also drainage via a prominent left-sided azygos venous system (Figure 4). The aorta was normal and left-sided. There was no left-to-right intracardiac shunt, and pulmonary venous drainage was normal.

Absence of the right superior vena cava is rare. During development, blood usually passes preferentially to the right-sided cardinal venous system via a vessel between the anterior cardinal veins that becomes the left brachiocephalic vein. The left-sided veins involute to become the coronary sinus and the ligament of Marshall. In this case, the left common cardinal vein has persisted and the common and proximal anterior cardinal veins on the right have regressed. The azygos system develops from the supracardinal venous system and again usually the right-sided azygos vein is predominant with hemi-azygos and accessory hemi-azygos veins on the left. Reversal of the pattern of persistence and regression may occur if flow is restricted on the right during embryological development, similar to the mechanism described for correct patterning of the branchial arch arteries.1

In the absence of other congenital anomalies, long-term prognosis from abnormal superior vena caval development is thought to be good, and clinical relevance relates to technical problems during pacemaker implantation,2 insertion of central lines, and cardiopulmonary bypass. The techniques of chest radiography, transthoracic and transesophageal echocardiography, and cardiovascular MRI offered complementary approaches for diagnosis and delineation of anatomy in this case. Use of bilateral contrast injections during transesophageal imaging elegantly demonstrated absence of the right superior vena cava, whereas cardiovascular MRI provided detailed assessment of upper body venous drainage and exclusion of possible associated congenital abnormalities.

Sources of Funding
This work was supported by the Oxford Partnership Comprehensive Biomedical Research Centre, with funding from the Department of Health’s National Institute for Health Research Biomedical Research Centres funding scheme.

Disclosures
None.

References

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The online-only Data Supplement is available at http://circimaging.ahajournals.org/cgi/content/full/2/5/e34/DC1.

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(Circ Cardiovasc Imaging. 2009;2:e34-e36.)
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Circ Cardiovasc Imaging is available at http://circimaging.ahajournals.org
DOI: 10.1161/CIRCIMAGING.108.828558
Figure 1. Chest radiograph demonstrates normal cardiac size but prominence of the aortic shadow.

Figure 2. Transesophageal echocardiography views of the right atria (RA). The probe in the lower esophagus at 0° demonstrates the coronary sinus after agitated saline contrast injection into the left antecubital fossa (A) and after injection into the right antecubital fossa (B). In both images, the saline bubbles first appear in the coronary sinus. C, Mid-esophagus at 110° provides a bicaval view. A right-sided superior vena cava should be evident on the right-hand side of the view. RV indicates right ventricle; LA, left atrium.
Figure 3. Cardiovascular MRIs of upper body venous drainage. A and B, Images taken at the level of the aortic arch and at the level of the pulmonary artery (PA) demonstrate the innominate vein, left superior vena cava, and absent right superior vena cava. C, Images of a sagittal plane through the left-sided superior vena cava. D, Cardiac short-axis view at the level of the mitral valve annulus demonstrates dilated coronary sinus. LV indicates left ventricle; RA, right atrium; LA, left atrium.

Figure 4. Three-dimensional surface shaded display reconstruction from multislice transverse steady-state free procession images of the upper thorax. Color has been used to highlight the aortic arch, left-sided superior vena cava, and left-sided azygos venous system.
Absent Right Superior Vena Cava: Multimodality Imaging of Upper Body Venous Drainage via Left-Sided Superior Vena Cava and Azygos Venous System
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doi: 10.1161/CIRCIMAGING.108.828558
Circulation: Cardiovascular Imaging is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 1941-9651. Online ISSN: 1942-0080

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