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.

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The azygos system develops from the supracardinal venous system and again usually the right-sided azygos vein is predominant with azygos and accessory 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.

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, 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.

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Disclosures

None.

References

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, Midesophagus 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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