A 20-year-old man was referred for the assessment of palpitations and associated dyspnea during exertion. He had no other associated cardiovascular symptoms. Physical examination revealed a healthy male with stable vital signs and a normal cardio respiratory examination. ECG, (Figure 1A), chest x-ray (Figure 1B), and blood counts as well as electrolyte panel were normal.

Transthoracic echocardiography revealed a dilated coronary sinus (Figure 2); in addition, a persistent left superior vena cava was noted (Figure 3). A small coronary-pulmonary arterial fistula was noted (Figure 4). Two-dimensional subcostal echocardiography view demonstrated absence of the hepatic portion of the inferior vena cava with the hepatic veins draining directly into the right atrium (interrupted inferior vena cava). The right suprasternal view revealed venous flow into the right superior vena cava from a dilated azygous vein (Figure 5). Left and right ventricles were normal in size and function.

Figure 1. A, Twelve-lead ECG with normal sinus rhythm, cardiac axis, and normal intervals. B, Chest x-ray postero-anterior and lateral views showing normal cardiac silhouette, but gastric air noted on the right subcostal in this case compared with the usual location.
Computerized axial tomographic angiography and magnetic resonance imaging confirmed the extracardiac venous anomalies diagnosed by detailed 2-dimensional echocardiography (see Figures 2B, 3B, 4B, 4C, 5B, and 5C). In addition, computerized axial tomographic angiography showed right-sided polysplenia, a centric liver, and a right stomach fulfilling the diagnostic criteria for heterotaxy with abdominal viscera situs ambiguus (Figure 6 and online-only Data Supplement Movies I to IV).

Electrophysiological study identified the cause of the exertional palpitations as an atrioventricular nodal reentrant tachycardia, which was successfully managed with an uncomplicated radiofrequency ablation procedure. At 1-year follow-up, the patient is doing well and is asymptomatic.

**Background**

Human external morphometry is usually that of symmetry, in contradistinction to the internal organs where established left to right asymmetry is the norm. When disturbance of this asymmetry occurs during embryonic development, the terminology of heterotaxy is applied. Heterotaxy is the nomenclature used to describe any malposition (discordance) of the thoracoabdominal organs and vessels, complex congenital heart disease, and extra cardiac defects or defects involving midline structures. Although the term is used to describe complex congenital anomalies, it also applies to isolated anomalies such as persistent left superior vena cava (SVC) and interrupted inferior vena cava. An association exists between these anomalies and splenic anomalies. Before the use of advanced echocardiography and noninvasive radiological imaging, most reports about heterotaxy were derived from autopsy series.

When a patient is found to have a cardiac anomaly, a systemic approach to other systems, including the spleen and its function, as well as assessment of ciliary function to rule out primary ciliary dyskinesia, is necessary. Complete diagnosis of these complex congenital abnormalities can now be achieved with integrated information from complementary multimodality noninvasive imaging technologies.
Management

The clinical course and management of extracardiac venous heterotaxy syndrome depends on symptoms related to the involved structures. Care includes regular follow-up, endocarditis prophylaxis when indicated, and the use of anticoagulation if abnormal venous flow or venous stasis is noted. In rare cases, surgical correction of anomalous venous drainage may be necessary.7

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Disclosures

None.

References


Figure 4. A small coronary fistula arising from left anterior descending branch to pulmonary trunk shown by: 2-dimensional short-axis view (A) and by computerized axial tomographic angiography, 3-dimensional reconstruction images (B and C). LAD indicates left anterior descending artery; PA, pulmonary artery; PV, pulmonary valve; PR, pulmonary regurgitation; Ao, aorta; RCA, right coronary artery; and CX, circumflex artery.
Figure 5. Abnormally dilated azygous vein draining into right SVC shown by right suprasternal color Doppler view (A), computed tomography with contrast showing the dilated azygous vein mimicking aortic arch draining into the right SVC (B), and other computed tomographic image showing the azygous vein giving picture similar to double aortic arch (C). R.SVC indicates right superior vena cava; AZ V, azygous vein; and Ao, aorta.
Figure 6. Computed tomographic image of abdomen with intravenous contrast showing visceral situs ambiguous. Where liver and portal vein are in the midline, right-sided splenules are seen in this image (2 out of 4 could be identified) and right-sided stomach. PV indicates portal vein; Ao, aorta; AZ V, azygous vein; and H-AZ V, hemiazygous vein.
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