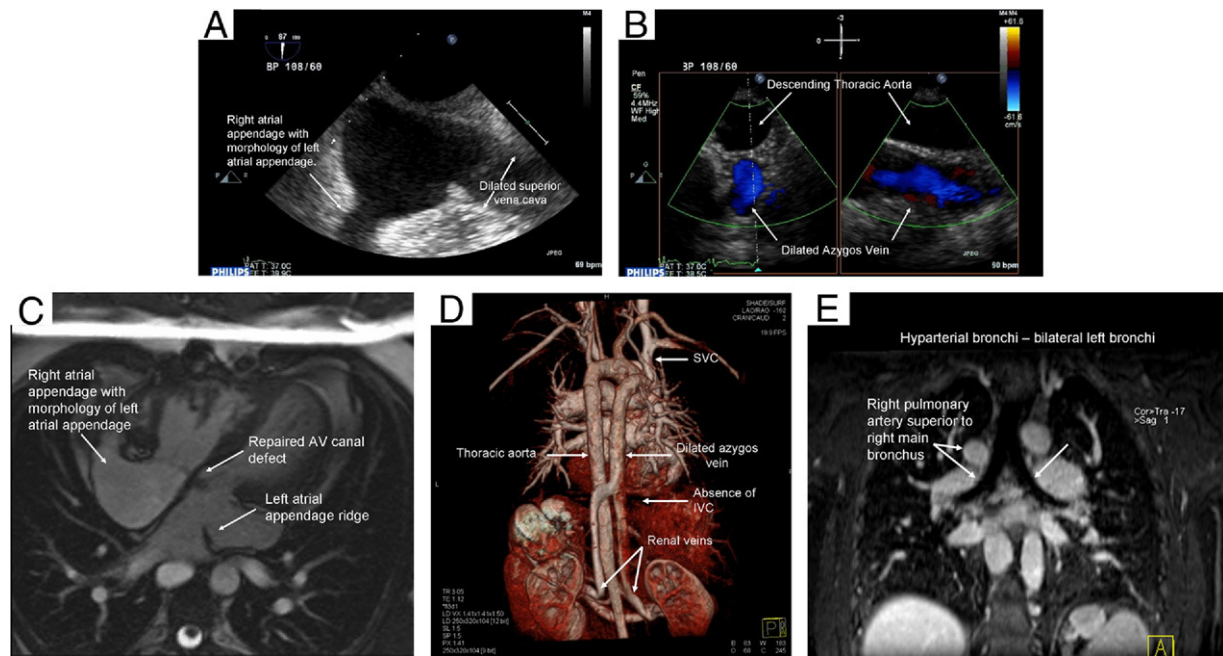


IMAGES IN CARDIOLOGY

## Bilateral Left-Sidedness Heterotaxy Syndrome

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**A** 25-year-old man with a history of atrioventricular (AV) canal defect repair in childhood underwent transesophageal echocardiography that revealed left atrial appendage morphology of the right atrial appendage (A) and a prominent azygos vein (B). Three-dimensional transesophageal echocardiography showed a cleft anterior mitral valve (Online Video 1). Cardiac magnetic resonance imaging confirmed the diagnosis of bilateral left-sidedness syndrome, including left atrial isomerism (C), absence of the intra-hepatic segment of the inferior vena cava (IVC), azygos continuation of the IVC (D, Online Video 2), and hyperarterial bronchi (E).

Heterotaxy syndrome represents a group of disorders of abnormal lateralization of the abdominal and thoracic organs. Sidedness of the heart is determined by the atrial appendage morphology. Cardiovascular findings of bilateral left-sidedness syndrome include bilateral left atrial appendage morphology, AV canal defects, azygos continuation of the IVC, as well as conduction abnormalities. Extracardiac findings include midline liver, polysplenia, hyperarterial bronchi, and bi-lobed lungs. SVC = superior vena cava.