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Double-Chambered Right Ventricle and Situs Inversus With Dextrocardia

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We report the case of a 74-year-old woman with a history of complex congenital heart disease consisting of double-chambered right ventricle (DCRV) and situs inversus with dextrocardia. To our knowledge, this is the first reported case with the combination of these 2 congenital abnormalities. The patient was transferred to our hospital after experiencing abdominal discomfort, malaise, and cold sweats for 3 days. Initial examination revealed slightly elevated cardiac troponin T (peaked at 0.56 ng/mL). With her known history of dextrocardia, 12-lead surface electrocardiography (ECG) with both standard precordial leads (Figure 1A) and right-sided precordial

leads (Figure 1B) was performed. The standard ECG was notable for inverted P waves in the lateral leads (I and aVL), suggesting rightward atrial electric forces and poor R wave progression. The right-sided precordial lead ECG showed normalized R wave progression. These summations of ECG findings are suggestive of dextrocardia. There were also pseudo-Q waves in the limb leads and T wave inversions in I and aVL, which are also consistent with dextrocardia but less specific than the other findings. In addition, her right-sided ECG showed ST depression and T wave inversion in the anterior leads, which in the setting of troponin elevation was concerning for acute



Figure 1. Twelve-lead surface electrocardiography with standard left-sided precordial leads (A) and right-sided precordial leads (B) are consistent with dextrocardia.

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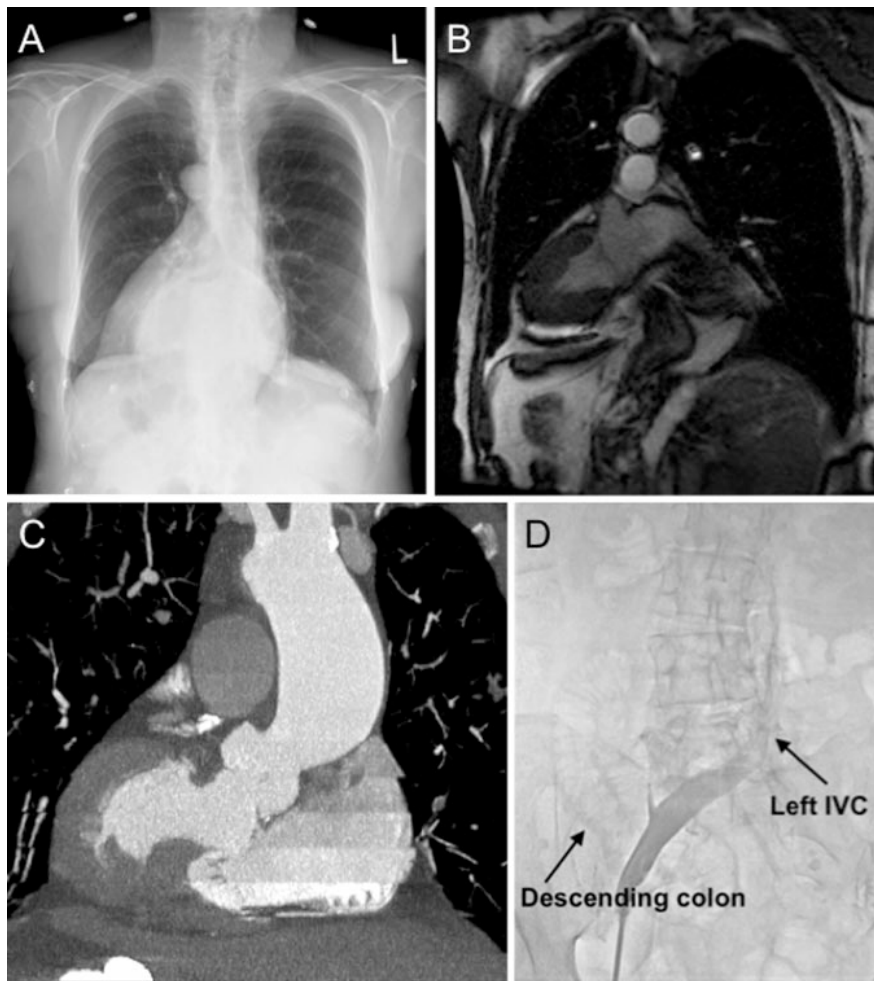


Figure 2. Dextrocardia with situs inversus. Chest radiography in the anteroposterior view (A), cardiac magnetic resonance imaging with balanced steady-state free precession sequence (B), and contrast-enhanced electrocardiogram-gated cardiac computed tomography in the coronal view (C), showing dextrocardia with a right-sided aortic arch and situs inversus. Fluoroscopy during cardiac catheterization (D), showing situs inversus with a left-sided inferior vena cava (IVC) and right-sided descending colon.

coronary syndrome. She subsequently underwent cardiac catheterization (Movies I and II of the online-only Data Supplement), which revealed nonobstructive coronary artery disease, and her troponin elevation was attributed to demand ischemia. Multimodality imaging studies were performed to assess her anatomy and its functional significance. Owing to her inability to maintain adequate breath-holds during cardiac magnetic resonance imaging, she also underwent contrast-enhanced ECG-gated cardiac dual-source computed tomography for visualization of her anatomy.

We confirmed the presence of dextrocardia with situs inversus by chest radiography, cardiac magnetic resonance imaging, computed tomography, and fluoroscopy (Figure 2). In addition to the dextrocardia, she had a DCRV with right ventricular outflow tract (infundibular) stenosis (Figure 3, Movies III–V in the online-only Data Supplement). The peak and mean instantaneous gradients across the infundibular stenosis were 56 and 32 mm Hg by transthoracic echocardiography (Figure 4A), and a peak–peak systolic pressure gradient between the pulmonary artery and right ventricle of 56 mm Hg was measured by cardiac catheterization (Figure 4B). Phase-contrast cardiac magnetic resonance imaging confirmed the infundibular stenosis with a peak velocity of 3.8 m/s, corresponding to a right ventricular intracavity peak gradient of 58 mm Hg.

Double-chambered right ventricle is a form of subvalvular right ventricular outflow tract obstruction caused by anomalous muscle bundles that divide the right ventricle into a high-pressure proximal chamber and a lower-pressure distal chamber.¹ The anomalous muscle bundles, which can range from 1 to many in number, usually arise from the body of the septal band (septomarginal trabeculation) and pass through the chamber of the right ventricle to the anterior free wall. DCRV is exceptionally rare as an isolated anomaly. Most commonly (in approximately 90% of affected individuals) it is associated with a membranous-type ventricular septal defect. Other coexisting lesions include subaortic stenosis, pulmonary valve stenosis, double-outlet right ventricle, tetralogy of Fallot, or anomalous pulmonary venous drainage, among others.^{2–4} We are unaware of any report of DCRV with dextrocardia. Although case reports of isolated DCRV have been published,^{5,6} we herein describe the first case, to our knowledge, of DCRV and situs inversus with dextrocardia, and we show images of this combination of complex congenital lesions and of its hemodynamic significance using multimodality imaging, including chest radiography, transthoracic echocardiography, cardiac computed tomography, cardiac magnetic resonance imaging, and cardiac catheterization.

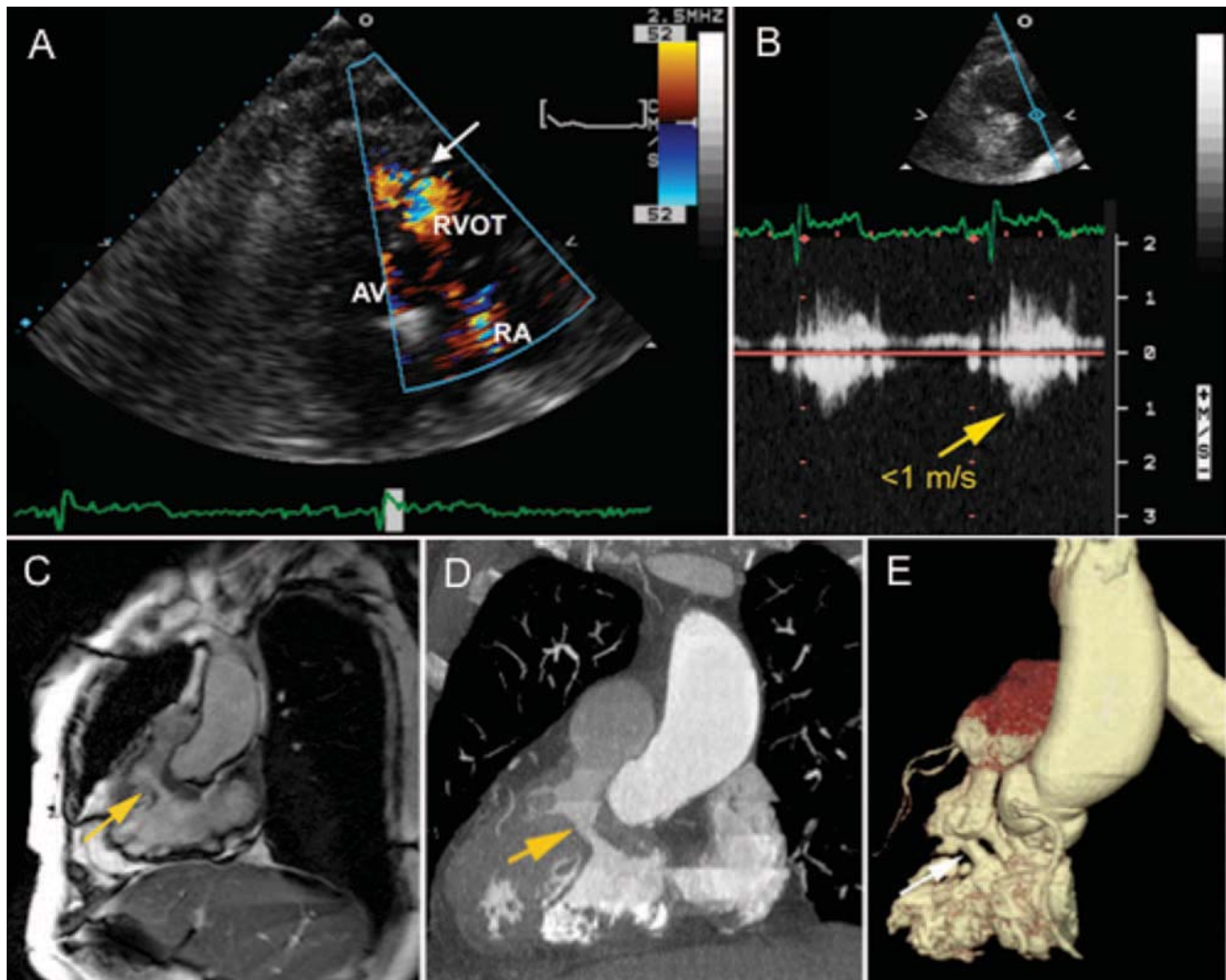


Figure 3. Anatomic assessment of the double-chambered right ventricle with infundibular stenosis. A, Transthoracic echocardiogram in the parasternal short-axis view, showing the right atrium (RA) and right ventricular outflow tract (RVOT) located to the right of the aortic valve (AV) and aorta with color flow acceleration across the infundibular stenosis (arrow). B, Continuous-wave Doppler across the pulmonary valve, demonstrating lack of a significant gradient (<1 m/s) and no evidence of pulmonic stenosis. C, Cardiac MRI with balanced steady-state free precession sequence, demonstrating infundibular stenosis (arrow). Maximum-intensity projection image in the coronal view (D) and volume-rendered 3-dimensional image (E) obtained from contrast-enhanced cardiac computed tomography, showing infundibular stenosis (arrow).

Disclosures

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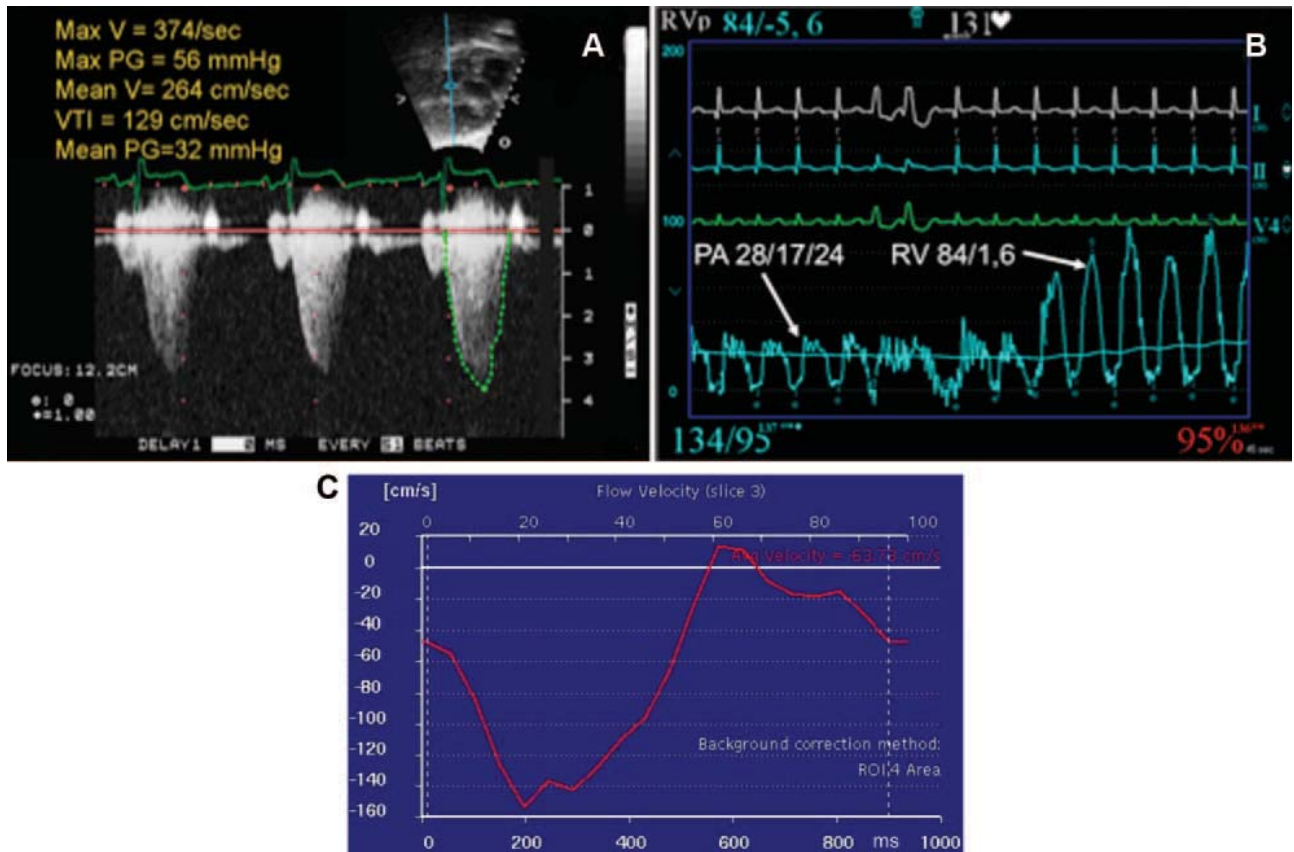


Figure 4. Hemodynamics of the gradient of infundibular stenosis. A, Transthoracic echocardiogram in the subcostal view with continuous-wave Doppler interrogation across the infundibular stenosis, depicting a peak and mean gradient of 56/32 mm Hg. B, Hemodynamic tracing from catheterization of the right side of the heart during pulmonary artery (PA) to right ventricle (RV) pullback, showing a peak systolic pressure gradient of 56 mm Hg, from peak PA pressure of 28 mm Hg to peak RV pressure of 84 mm Hg as the catheter crosses the infundibular stenosis. C, Flow curve derived from phase contrast cine MRI data showing mean velocity over time through the infundibular stenosis (a negative value indicates flow direction is cephalad).