CLINICAL IMAGE

When echocardiography misleads: criss-cross heart in an adult

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Criss-cross heart (CCH) is a rare congenital anomaly characterized by crossing of the atrioventricular inflow streams due to abnormal spatial orientation of the ventricles.[1-4] Adult presentation is exceptional, particularly in the absence of significant associated structural abnormalities.[4,5]

A 35-year-old man with no prior cardiovascular history presented with several hours of stabbing retrosternal chest pain and severe hypertension (systolic blood pressure of up to 195 mm Hg). Twelve-lead electrocardiography showed sinus rhythm with first-degree atrioventricular block and an indeterminate frontal QRS axis (FIGURE 1A). The high-sensitivity troponin T level was mildly elevated but stable; creatine kinase and its muscle-brain isoform levels were normal. The level of N-terminal pro–B-type natriuretic peptide was slightly elevated.

Transthoracic echocardiography was technically limited by poor acoustic windows and an unusual cardiac position, with the apex displaced posteriorly within the left hemithorax (FIGURE 1B). Off-axis imaging and foreshortening likely contributed to the underestimation of left ventricular (LV) systolic function, which was visually assessed as reduced (LV ejection fraction [LVEF], 35%–40%), raising suspicion of cardiomyopathy. Given the discordance between the echocardiographic findings and the atypical cardiac position and electrical axis, cardiac magnetic resonance (CMR) was performed as the next diagnostic step.

CMR demonstrated crossing atrioventricular inflow streams with abnormal ventricular alignment, consistent with CCH, and preserved ventricular size (FIGURE 1C and 1D). No ventricular septal defect or other major structural abnormalities were identified. CMR excluded active myocarditis (no myocardial edema or late gadolinium enhancement) and showed preserved LVEF (52%).

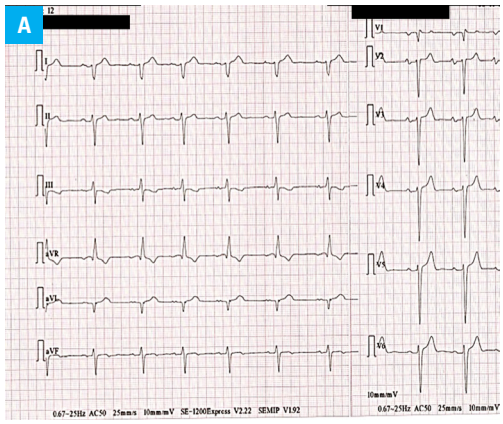

FIGURE 1 Criss-cross heart (CCH): multimodality imaging findings; **A** – 12-lead electrocardiography showing sinus rhythm with first-degree atrioventricular block and an indeterminate frontal QRS axis; **B** – transthoracic echocardiography (parasternal long-axis view) demonstrating unusual cardiac orientation and suboptimal imaging conditions

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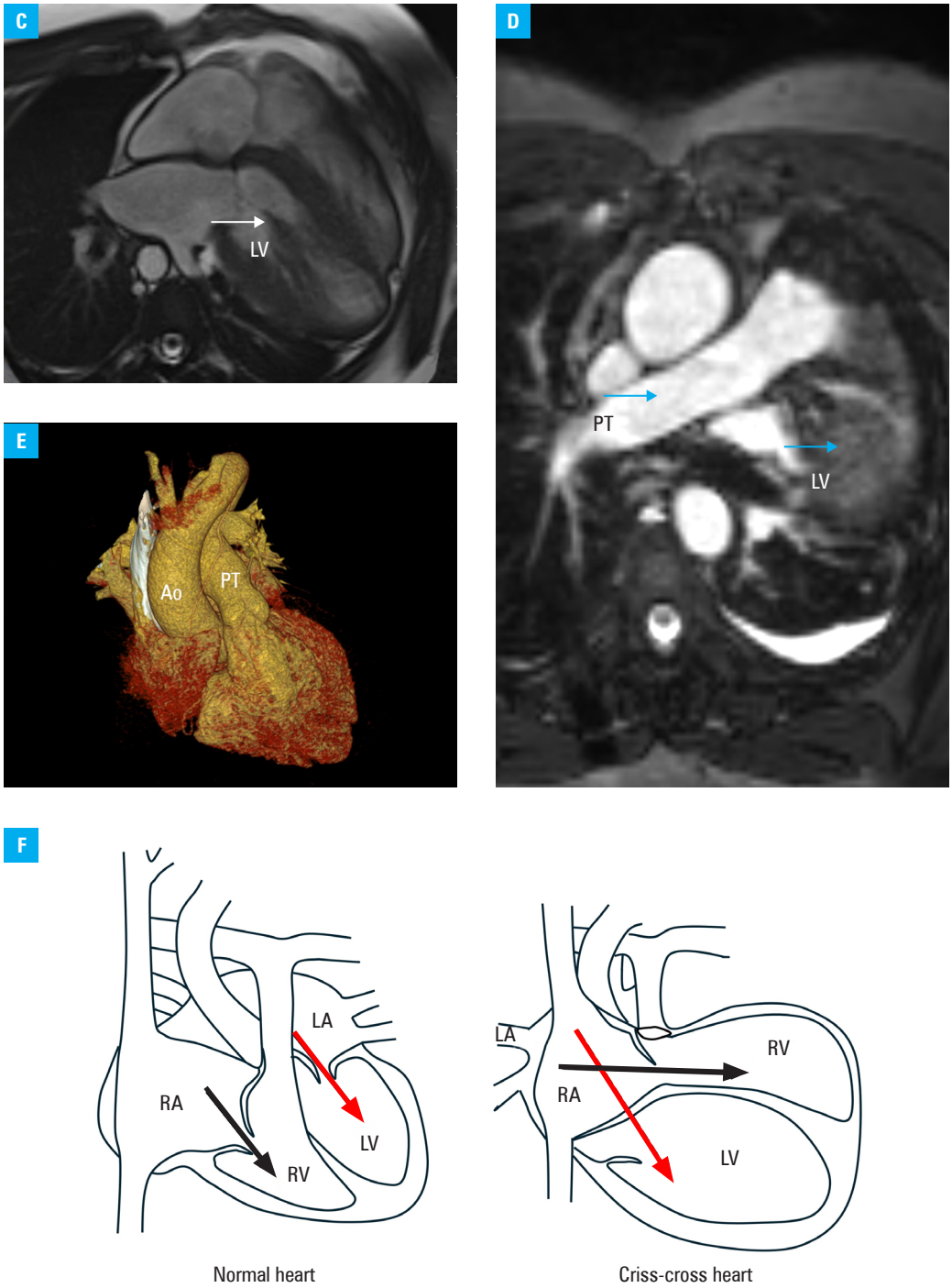

FIGURE 1 Criss-cross heart (CCH): multimodality imaging findings; **C** – cine cardiac magnetic resonance (CMR; long-axis view) demonstrating abnormal ventricular alignment; **D** – CMR, short-axis view, showing atypical spatial relationship between the ventricles and great vessels; **E** – coronary computed tomography showing normal aortic arch morphology with a tortuous descending thoracic aorta; **F** – a schematic comparison of normal cardiac anatomy and CCH, illustrating the orientation of atrioventricular inflow streams (black arrows: RA to RV; red arrows: LA to LV)
Abbreviations: Ao, aorta; LA, left atrium; LV, left ventricle; PT, pulmonary trunk; RA, right atrium; RV, right ventricle

Coronary computed tomography excluded obstructive coronary artery disease and confirmed normal aortic arch morphology with a tortuous descending aorta (FIGURE 1E). The patient was managed conservatively with antihypertensive therapy and scheduled for outpatient follow-up.

This case illustrates that CCH in adults may be hemodynamically uncomplicated and remain undiagnosed until incidental detection. It also highlights that reduced LVEF on echocardiography may result from suboptimal imaging conditions, including foreshortening and off-axis views, rather than true systolic dysfunction. Multimodality imaging, particularly CMR, plays a key role in clarifying cardiac anatomy and ventricular function in such cases, and may prevent misdiagnosis of cardiomyopathy and unnecessary invasive testing (FIGURE 1C, 1D, and 1F).

ARTICLE INFORMATION

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