Assessment of endocardial fibroelastosis by cardiac MRI

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A 17-year-old patient was referred to the Unit of Cardiovascular Medicine at the Leeds General Infirmary (Leeds, United Kingdom) for a cardiac magnetic resonance imaging (MRI) scan. At the age of two months, she had developed dyspnea and was found to have unexplained left atrial enlargement. By the age of six years, she had developed significant tricuspid regurgitation and pulmonary hypertension. Cardiac catheterization at that time confirmed a mean pulmonary artery pressure of 50 mmHg, with no evidence of pulmonary artery stenosis or shunt. A diagnosis of restrictive cardiomyopathy secondary to primary endocardial fibroelastosis was established after right ventricular endomyocardial biopsy.

Cardiac MRI using steady-state free precession cine imaging revealed the typical features of a restrictive cardiomyopathy (Figure 1A). The left ventricular cavity was small and showed normal systolic function. There was marked left atrial and right atrial dilation, global pericardial effusion, right ventricular hypertrophy and severe tricuspid regurgitation. The pulmonary arteries were dilated and the interventricular septum appeared flattened, consistent with right ventricular pressure overload (mean pulmonary artery pressure measured 80 mmHg at repeat cardiac catheterization at 17 years of age). In addition, the inferior vena cava was grossly dilated (Figure 1B) and a thrombus extended from the left atrial appendage into the left atrium (Figure 1C). Late gadolinium hyperenhancement imaging revealed a characteristic pattern of high-signal intensity confined to the endocardial surface of both ventricles and both atria (Figure 1D), representing fibrosis in these areas.

Cardiac MRI clearly demonstrates the pathognemonic features of endocardial fibroelastosis and can identify additional complications such as intracardiac thrombus.

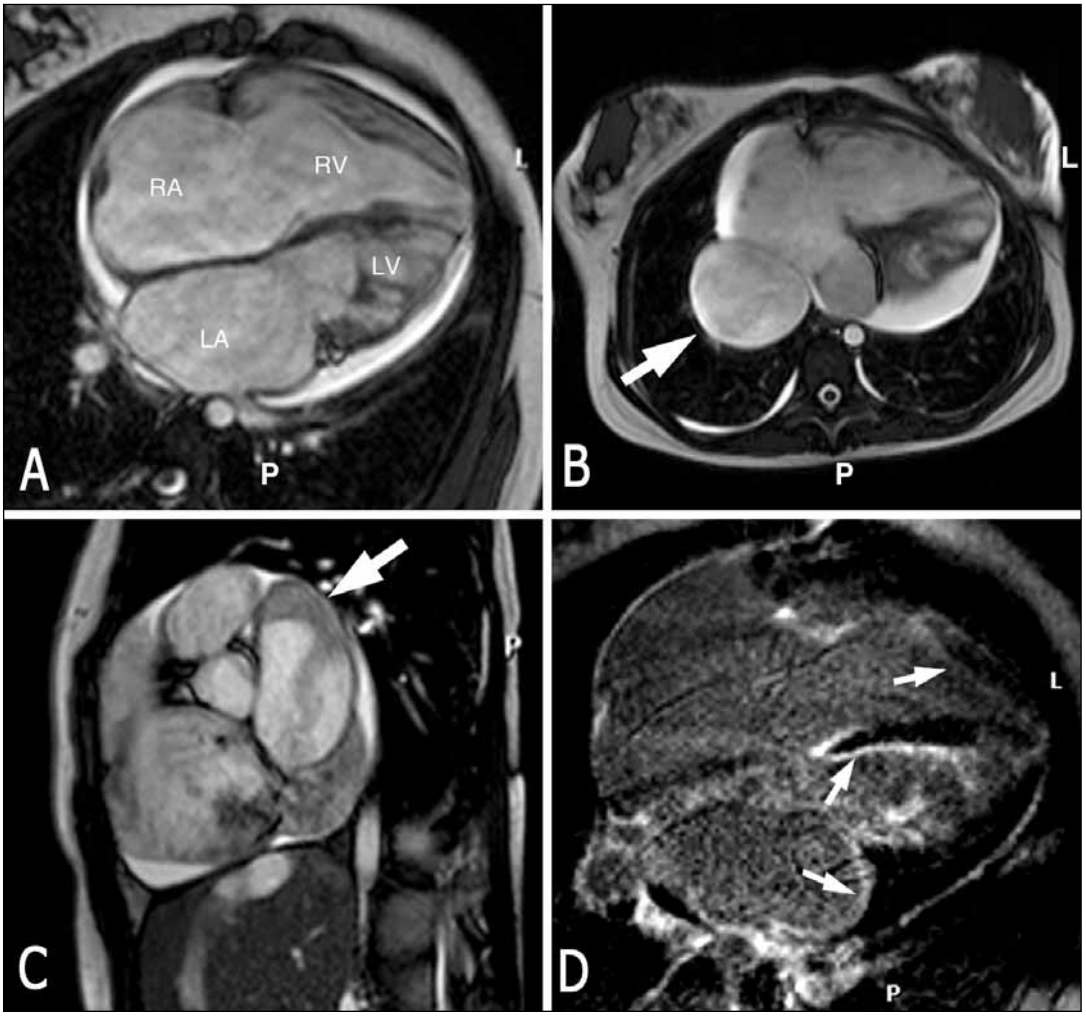

Figure 1) A *Four-chamber cine image demonstrating biatrial dilation, right ventricular hypertrophy, a small left ventricular cavity and pericardial effusion.* **B** *Cine image demonstrating dilation of the inferior vena cava (arrow).* **C** *Short-axis cine image demonstrating thrombus (arrow) extending from the left atrial appendage into the left atrium.* **D** *Four-chamber late gadolinium hyperenhancement image demonstrating high-signal intensity in the atrial and ventricular endocardium (arrows). L Left; LA Left atrium; LV Left ventricle; P Posterior; RA Right atrium; RV Right ventricle*

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