

Diagnosis of Isolated Noncompaction of the Myocardium by Magnetic Resonance Imaging

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A 15-year-old asymptomatic, physically active man, who had a history of syncope at the age of 12 and a family history of arrhythmogenic right ventricular dysplasia (father), presented for evaluation. Physical examination revealed no abnormal findings. The ECG demonstrated sinus rhythm, -20° QRS, incomplete right bundle-branch block, and left ventricular hypertrophy; the 24-hour Holter monitoring excluded ventricular arrhythmias. Normal exercise capacity (15 metabolic equivalent) without arrhythmias was confirmed by treadmill exercise testing. The echocardiogram showed apical hypertrophy of the left ventricle and an interatrial pseudo-aneurysm without shunt (Figure 1). To rule out right ventricular dysplasia, an ECG-gated cardiac MRI was performed. Surprisingly, the “black-blood” cardiac MRI examination demonstrated thickened apical segments of the myocardium and areas suspicious for deep intertrabecular recesses that were not observed at echocardiography (Figure 2). The diagnosis of isolated left ventricular noncompaction was confirmed through the use of a high-resolution cine gradient echo sequence that is particularly sen-

sitive to slow flow (Figure 3). The presence of deep recesses filled with blood and trabeculations of the myocardium were evident during diastole, whereas during systole, the recesses collapsed and the myocardium appeared compacted. The right ventricle was normal.

Noncompaction of the ventricular myocardium is a rare congenital cardiomyopathy characterized by an altered structure of the myocardial wall resulting from intrauterine arrest in normal endomyocardial embryogenesis and compaction of the myocardial fibers in absence of any coexisting congenital lesions. The morpho-pathological findings consist of multiple, prominent myocardial trabeculations and deep intertrabecular recesses communicating with the left ventricular cavity. The clinical manifestations are not specific for this cardiomyopathy and include heart failure, tachyarrhythmias (which predispose to sudden death), and cardioembolic events.

This case provides evidence of the potential of cardiac MR imaging in the evaluation of the right ventricle and of cardiac anomalies such as noncompaction.

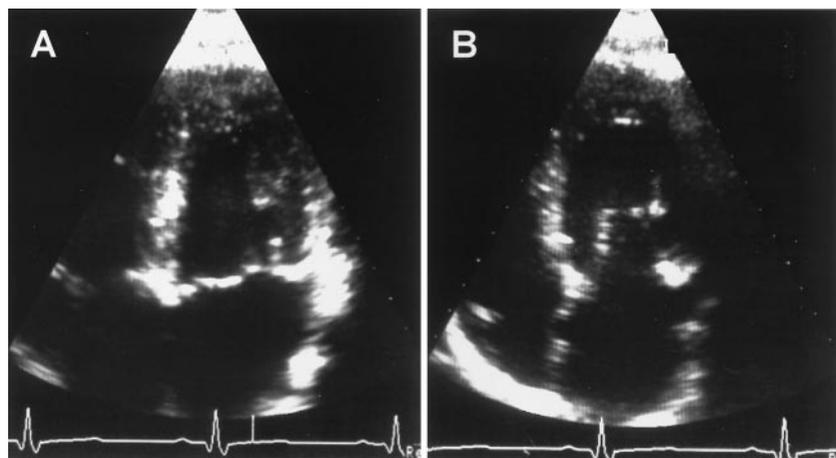


Figure 1. Two-dimensional echocardiography in a 4-chamber view. A, Systole; B, diastole.

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Movies I, II, and III are available in an online-only Data Supplement at <http://www.circulationaha.org>.

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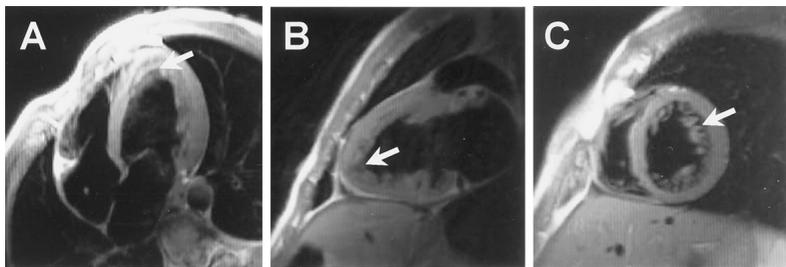


Figure 2. MR double inversion recovery images in diastole in 4-chamber (A), 2-chamber (B), and left ventricular (LV) short-axis (C) views. Arrows indicate thickened noncompacted myocardial wall.

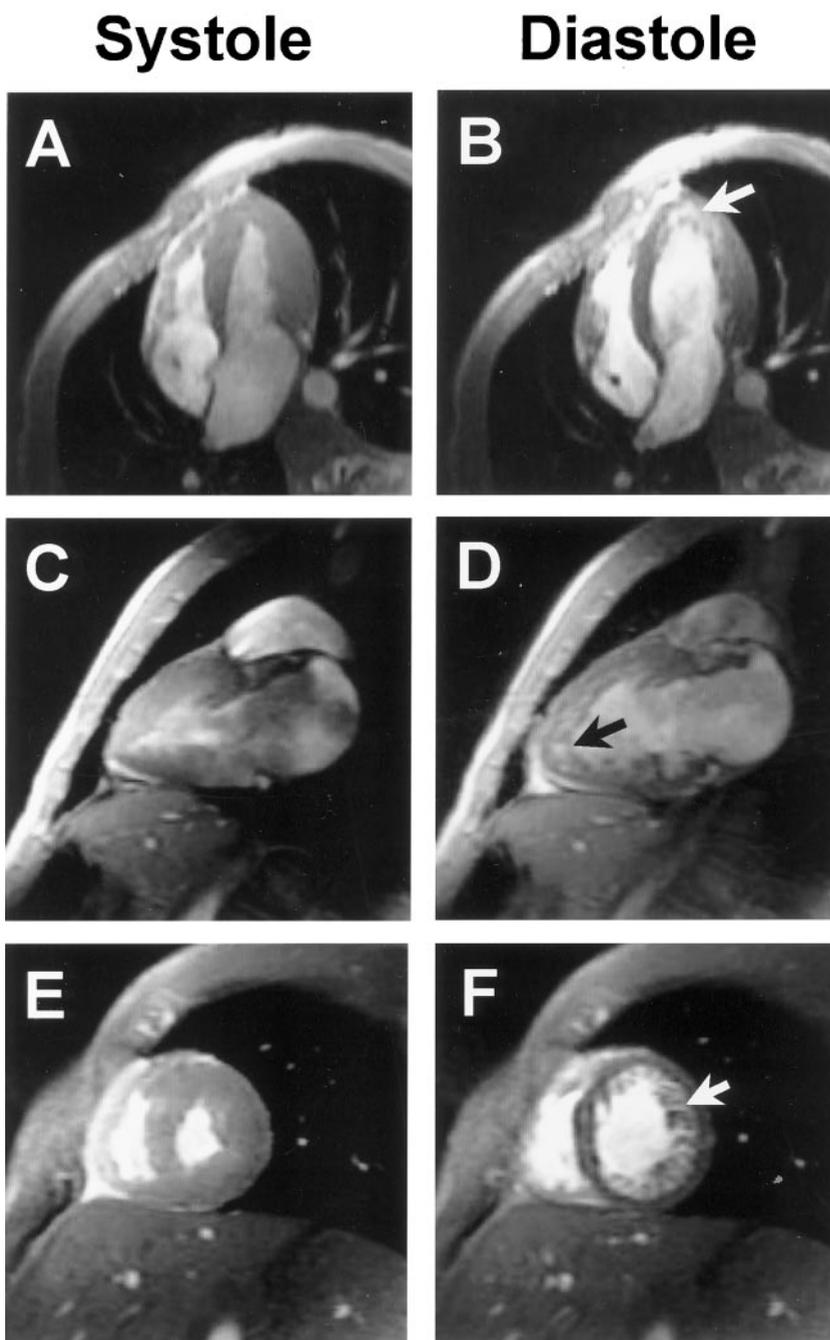


Figure 3. MR gradient echo images are very sensitive to slow flow. The presence of deep apical recesses filled with blood that communicate with the ventricle (bright areas within the myocardium) is diagnostic of non-compacted myocardium (arrow). A and B, 4-chamber view; C and D, 2-chamber view; and E and F, short-axis view. In contrast to diastolic images (B, D, and F), systolic images (A, C, and E) do not show flowing blood within the myocardial wall because the trabeculations are very close to each other, and recesses are collapsed. A also demonstrates the interatrial pseudo-aneurysm without shunt.

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