Isolated Noncompaction of the Myocardium

A Rarity or Missed Diagnosis?

J.A. McCrohon, FRACP, PhD; D.R. Richmond, MBChB, MSc, FRACP; D.J. Pennell, FRCP, MD; R.H. Mohiaddin, FRCR, PhD

A 31-year-old man presented to the emergency department with central chest tightness, shortness of breath, and presyncope. His admission ECG was abnormal (Figure 1), with inferior and lateral Q waves, left ventricular hypertrophy, repolarization changes in leads II, III, AVF, and V_4 to V_6, and nonspecific ST elevation in V_1 to V_3. A physical examination was unremarkable except for a fourth heart sound. Baseline blood studies showed mildly elevated troponin and creatine kinase-MB levels and hypercholesterolemia. A cardiac ultrasound showed the upper limit of normal wall thickness and normal valvular flows, biventricular size, and function. Cardiac catheterization revealed a mildly abnormal contraction of the anterobasal wall of the left ventricle and normal epicardial coronary vessels. Cardiac magnetic resonance imaging was performed to help exclude myocardial/pericardial disease.

Magnetic resonance documented intramyocardial recesses of the inferior and anterobasal left ventricular (LV) wall. These recesses were in communication with the LV lumen (Figure 2, A and B). In addition, prominent trabeculation extended into the LV cavity (Figure 3). Ventricular mass, size, and systolic function were normal. There was no evidence of myocardial hyperenhancement after gadolinium injection. Serology was normal.

These appearances are consistent with myocardial noncompaction, a congenital disorder of endomyocardial embryogenesis. This example is less florid than cases detailed in the limited number of echo and pathology series currently published and, in fact, would have been missed by ultrasound criteria. A diagnosis of noncompaction has important implications because of the need for familial screening and the possible association with other cardiac anomalies and/or muscle disorders, progressive LV dysfunction, risk of systemic embolism, and life-threatening arrhythmias. Techniques such as magnetic resonance imaging may improve detection rates and provide new insights into the prevalence, spectrum, and natural course of this potentially not-so-rare condition.

Acknowledgment

This work was supported by Coronary Artery Disease Research Association (CORDA; Marian Silcock Fellowship).

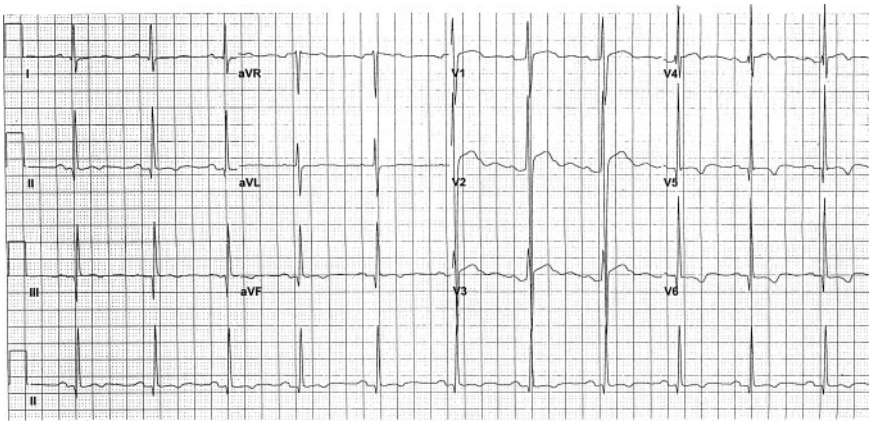

Figure 1. ECG showing left ventricular hypertrophy and T-wave abnormalities.

From the Department of Cardiovascular Magnetic Resonance, Royal Brompton Hospital, London, UK (J.A.M., D.J.P., R.H.M.); and Department of Cardiology, St George Hospital (J.A.M.), and Royal Prince Alfred Hospital (D.R.R.), Sydney, Australia.

Correspondence to Dr Jane McCrohon, Department of Cardiology, St George Hospital, Gray St, Kogarah, Sydney, 2217, Australia. E-mail mccrohonj@sesahs.nsw.gov.au

The editor of Images in Cardiovascular Medicine is Hugh A. McAllister, Jr, MD, Chief, Department of Pathology, St Luke's Episcopal Hospital and Texas Heart Institute, and Clinical Professor of Pathology, University of Texas Medical School and Baylor College of Medicine.

Circulation encourages readers to submit cardiovascular images to the *Circulation* Editorial Office, St Luke's Episcopal Hospital/Texas Heart Institute, 6720 Bertner Ave, MC1-267, Houston, TX 77030.

(*Circulation.* 2002;106:e22-e23.)

***Circulation* is available at http://www.circulationaha.org** **DOI: 10.1161/01.CIR.0000028961.66881.2D**

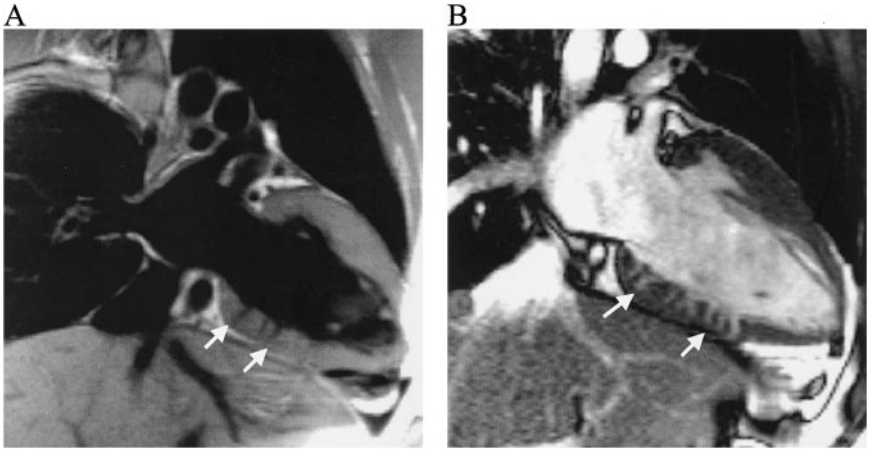

Figure 2. T_1-weighted turbo spin echo (A) and true fast imaging with steady-state free precession (true FISP) diastolic cine image (B) in the vertical long axis of the left ventricle, showing multiple inferior wall intertrabecular recesses in communication with the LV cavity (arrows).

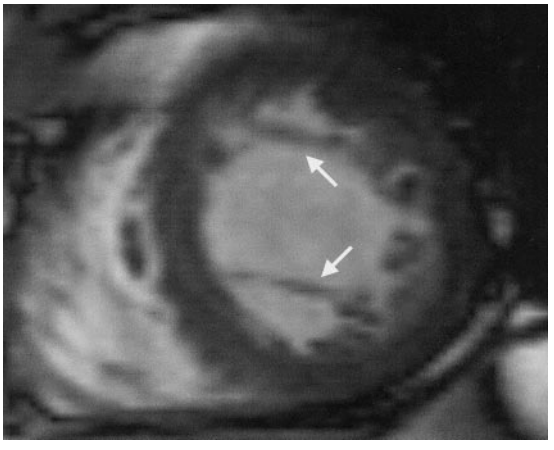

Figure 3. Short-axis true FISP cine image apical to the papillary muscles showing unusual muscle trabeculations extending into the LV cavity.

Isolated Noncompaction of the Myocardium: A Rarity or Missed Diagnosis?

J. A. McCrohon, D. R. Richmond, D. J. Pennell and R. H. Mohiaddin

Circulation. 2002;106:e22-e23
doi: 10.1161/01.CIR.0000028961.66881.2D
Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231

Print ISSN: 0009-7322. Online ISSN: 1524-4539