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Long-Term Prognostic Value of Cardiac Magnetic Resonance in Left Ventricle Noncompaction

A Prospective Multicenter Study

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ABSTRACT

BACKGROUND Cardiac magnetic resonance (CMR) is useful for the diagnosis of left ventricular noncompaction (LVNC). However, there are limited data regarding its prognostic value.

OBJECTIVES The goal of this study was to evaluate the prognostic relevance of CMR findings in patients with LVNC.

METHODS A total of 113 patients with an echocardiographic diagnosis of LVNC underwent CMR at 5 referral centers. CMR diagnostic criterion of LVNC (noncompacted/compacted ratio >2.3 in end-diastole) was confirmed in all patients. We performed left ventricular (LV) and right ventricular quantitative analysis and late gadolinium enhancement (LGE) assessments and analyzed the following LVNC diagnostic criteria: left ventricular noncompacted myocardial mass (LV-ncMM) >20% and >25%, total LV-ncMM index >15 g/m^2, noncompacted/compacted ratio ≥3:1 ≥1 of segments 1 to 3 and 7 to 16 or ≥2:1 in at least 1 of segments 4 to 6 of the American Heart Association model. Outcome was a composite of thromboembolic events, heart failure hospitalizations, ventricular arrhythmias, and cardiac death.

RESULTS At a mean follow-up of 48 ± 24 months, cardiac events (CEs) occurred in 36 patients (16 heart failure hospitalizations, 10 ventricular arrhythmias, 5 cardiac deaths, and 5 thromboembolic events). LV dilation, impaired LV ejection fraction, and LV-ncMM >20% was significantly more frequent in patients with CEs. LV fibrosis was detected by using LGE in 11 cases. CMR predictors of CEs were LV dilation and LGE. LGE was associated with improved prediction of CEs, compared with clinical data and CMR functional parameters in all 3 models. No CEs occurred in patients without dilated cardiomyopathy and/or LGE.

CONCLUSIONS In patients with LVNC evaluated by using CMR, the degree of LV trabeculation seems to have no prognostic impact over and above LV dilation, LV systolic dysfunction, and presence of LGE. (J Am Coll Cardiol 2016;68:2166–81) © 2016 by the American College of Cardiology Foundation.

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Left ventricular noncompaction (LVNC) is a rare myocardial disorder characterized by prominent left ventricular (LV) trabeculations, deep intertrabecular recesses communicating with the ventricular cavity, and a thin and compacted epicardial layer (1). The disease may be asymptomatic or may have clinical manifestations, including heart failure (HF), malignant arrhythmias, and systemic thromboembolic events. It is unclear whether LVNC is a distinct cardiomyopathy or a morphological trait shared by different types of cardiomyopathies (2).

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LVNC can be isolated or associated with other cardiomyopathies, congenital heart diseases, and complex syndromes. The LVNC trait may be familial or sporadic (3). Sporadic LVNC can be acquired in response to a mechanical load (4), whereas LV trabeculations are influenced by race and/or ethnicity (5). The genetic bases of familial LVNC are still a matter of research (2). Although there is no current gold standard for LVNC diagnosis, cardiac imaging is the best tool available.

Echocardiography and cardiac magnetic resonance (CMR) are the most commonly used imaging modalities. Echocardiography is the first-line tool for diagnosis (1), whereas CMR adds anatomic and functional information about noncompacted and compacted myocardium, along with fibrosis detection using the late gadolinium enhancement (LGE) technique (6). The ratio of the thickness of noncompacted to compacted myocardial layers measured at end-diastole is the main CMR criterion for a diagnosis of LVNC (7). Left ventricular noncompacted myocardial mass (LV-ncMM) >20% of total mass is another criterion (8). Since Chin et al. (9) reported the first 8 cases in 1990, several studies have been published on LVNC, but these studies have limitations (1,4-8). They included small and highly selected cohorts of patients who often have a severe phenotype, they used different criteria for LVNC diagnosis, and they were frequently single-center studies with conflicting results.

The present prospective, multicenter study included a large cohort of patients with LVNC; the goal was to evaluate the long-term prognostic relevance of several traditional and newly proposed diagnostic CMR criteria, as well as the presence of LV fibrosis as detected by using LGE.

ABBREVIATIONS AND ACRONYMS

CE = cardiac event

CMR = cardiac magnetic resonance

DCM = dilated cardiomyopathy

HF = heart failure

HR = hazard ratio

IDI = integrated discrimination improvement

LGE = late gadolinium enhancement

LV = left ventricular

LVEDV = left ventricular end-diastolic volume

LVEF = left ventricular ejection fraction

LV-MM = left ventricular myocardial mass

LVNC = left ventricular noncompaction

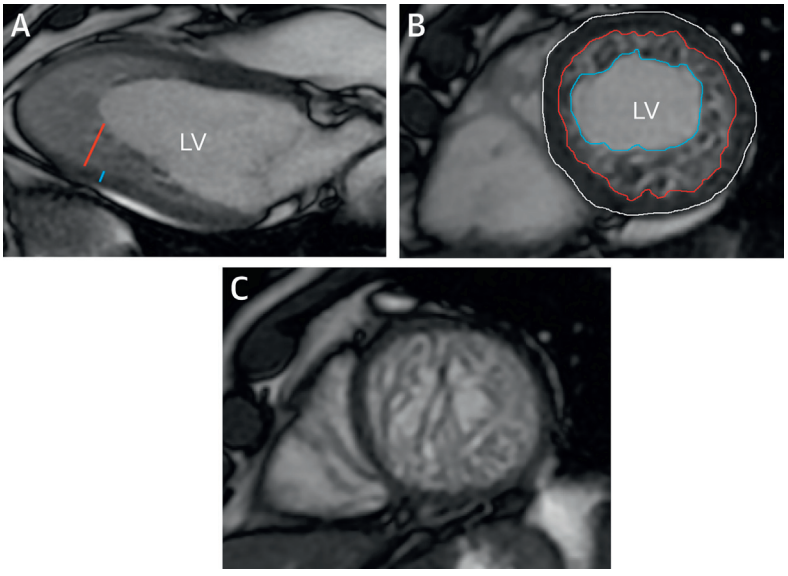
LV-ncMM = left ventricular noncompacted myocardial mass

LVSV = left ventricular stroke volume

NRI = net reclassification improvement

RV = right ventricular

FIGURE 1 LVNC Evaluation by Cardiovascular Magnetic Resonance



(A) Steady-state free precession (SSFP) sequence in long-axis orientation showing an extensive noncompacted myocardial layer lining the cavity of the mid-apical portions of the left ventricle (LV). The **orange line** identifies the thickness of the trabeculated layer; the **blue line** identifies the thickness of the compacted layer. These 2 measurements were used to calculate the end-diastolic noncompacted/compacted ratio. **(B)** SSFP short-axis acquisition confirming the hypertrabeculation of the mid portions of the LV. The **blue** and **orange lines** identify the drawn endocardial contour of the trabeculated and compacted layer, respectively. The **white line** identifies the epicardial contour. These measurements were used to calculate left ventricular noncompacted myocardial mass (LV-ncMM) and global left ventricular myocardial mass (LV-MM). **(C)** SSFP short-axis acquisition showing the apex of the LV completely filled by trabeculated myocardium.