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IMAGING VIGNETTE

LV Noncompaction Cardiomyopathy or Just a Lot of Trabeculations?

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LEFT VENTRICULAR NONCOMPACTION (LVNC) IS CHARACTERIZED BY THE PRESENCE OF AN EXTENSIVE noncompacted myocardial layer lining the cavity of the left ventricle (LV) and potentially leads to cardiac failure, thromboembolism, and malignant arrhythmias (1). LVNC is a heterogeneous clinical condition, which often shows an overlap with other forms of established cardiomyopathy (2). Whether it is a distinct cardiomyopathy or a morphological variant of other types of nonischemic cardiomyopathy is still debated. There are uncertainties regarding its prevalence and incidence, natural history including prognosis, best diagnostic approaches, and management. Cardiac magnetic resonance plays an increasingly important role in detection of noncompacted myocardium and identification of prognostic markers such as late gadolinium enhancement.

We report 2 short cases of related individuals (**Figures 1**, **2**, and **3**, Online Videos 1, 2, 3, and 4) with increased LV trabeculations. The key learning points highlight the importance of LV systolic function and pre-test probability in formulating the management plan (**Figure 4**).

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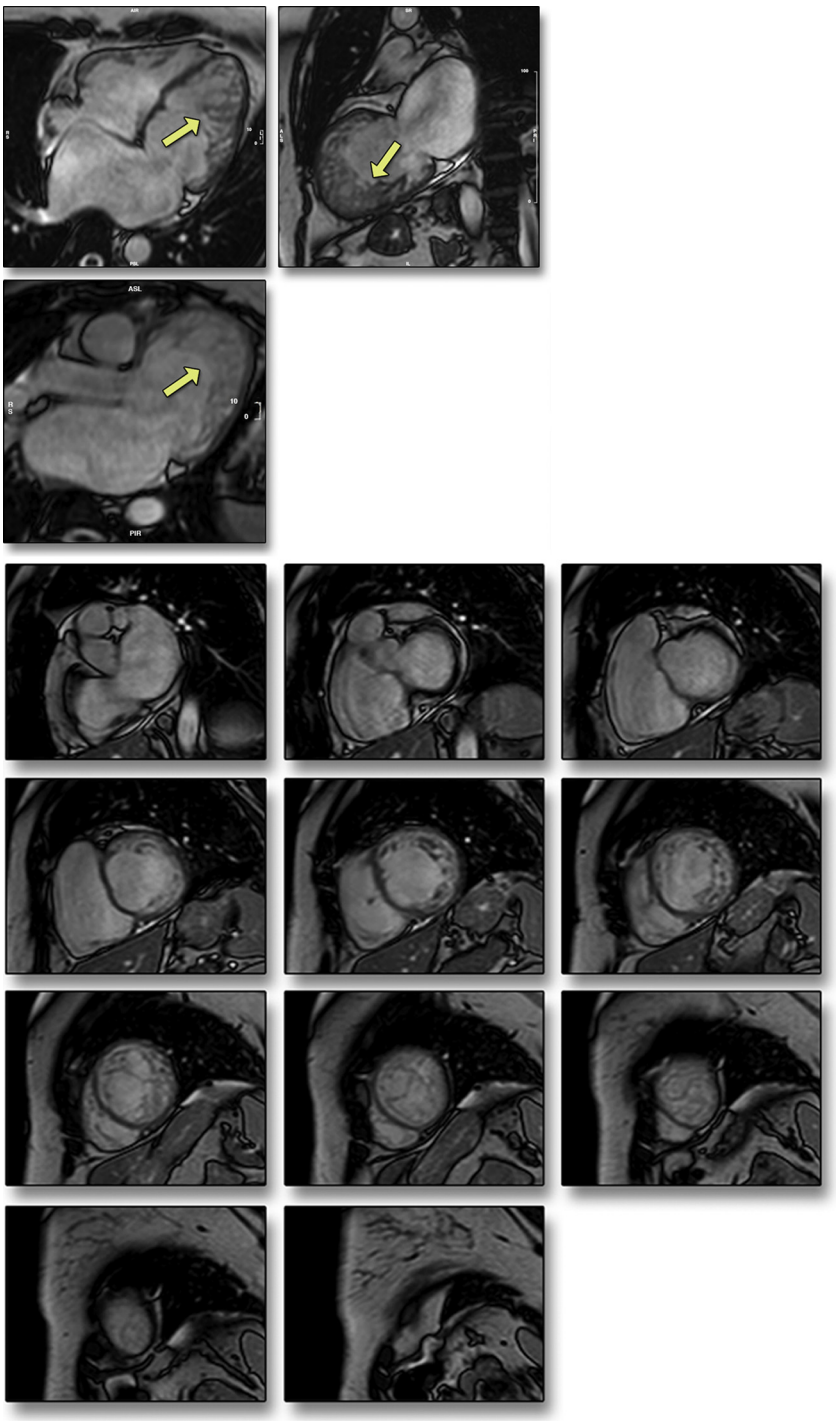
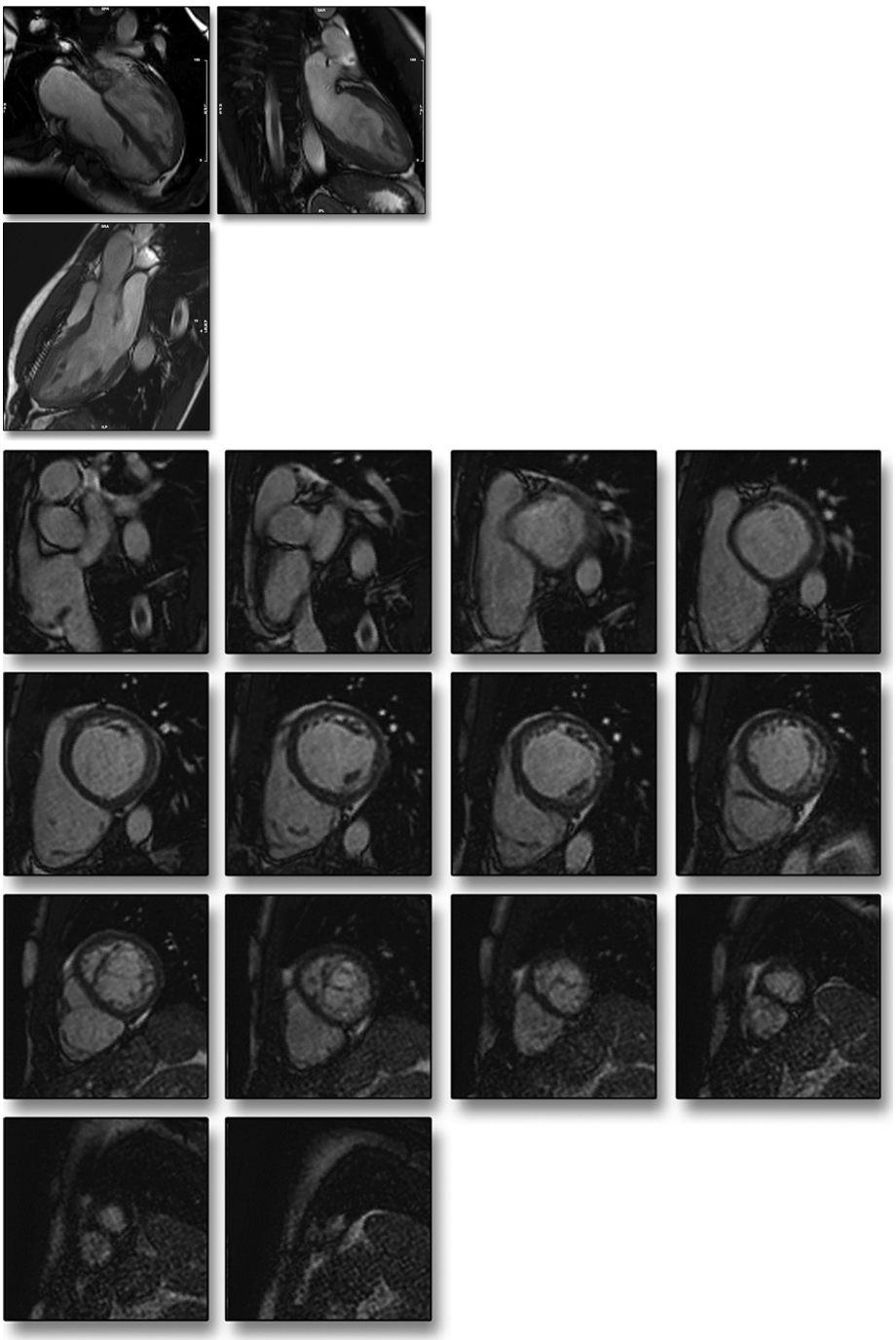
FIGURE 1 **CMR Images of Case 1 Demonstrating Prominent LV Trabeculations**



Case 1: A 51-year-old Caucasian woman with no previous medical history presented with a 10-min episode of severe dizziness while sitting. She had no other cardiac symptoms. Cardiac magnetic resonance (CMR) showed a dilated left ventricle (LV) with severe global systolic impairment (left ventricular end-diastolic volume [LVEDV] 233 ml; left ventricular end-systolic volume [LVESV] 166 ml; left ventricular stroke volume [LVSV] 67 ml; left ventricular ejection fraction [LVEF] 29%) and features of LV noncompaction based on the diastolic noncompaction to compaction ratio in the long-axis steady-state free precession cines of 4:1 **(yellow arrow)** (Online Videos 1 and 2). A diagnosis of dilated cardiomyopathy with features of left ventricular noncompaction (LVNC) was made, and she was treated with conventional heart failure medical therapy. An interval CMR 2 years later demonstrated progressive LV disease (LVEDV 210 ml, LVESV 155 ml, LVSV 55 ml, LVEF 26%). She subsequently received a primary prevention implantable cardioverter-defibrillator.

FIGURE 2 **Baseline CMR Images of Case 2**



Case 2: A 32-year-old Caucasian man was referred to the cardiology clinic with palpitations and a family history of LVNC. He had no other cardiac symptoms. Baseline CMR showed a nondilated LV with good systolic function (LVEDV 180 ml, LVESV 75 ml, LVSV 105 ml, LVEF 58%) (Online Video 3). Hypertrabeculation of the LV myocardium was noted with a noncompaction to compaction (NC/C) ratio of 2.3:1, and there was no late gadolinium enhancement. In the absence of any cardiac symptoms or LV impairment, no therapy was introduced at this point. A second CMR 2 years later demonstrated an increase in LV internal dimensions and decline in LV systolic function (LVEDV 219 ml, LVESV 128 ml, LVSV 91 ml, LVEF 48%) (Online Video 4). The maximal NC/C ratio was 3.4:1. In view of these interval changes in CMR findings, a diagnosis of LVNC was made and a long-term follow-up was offered. Abbreviations as in **Figure 1**.

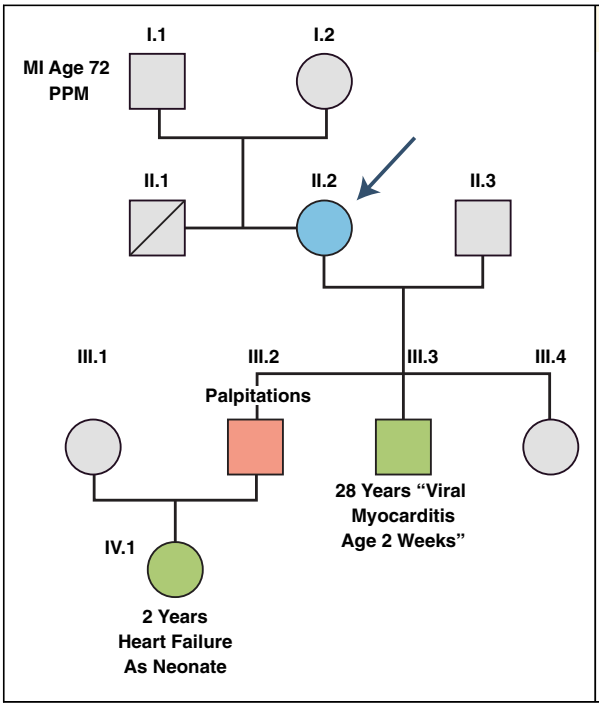


FIGURE 3 Family Pedigree

Case 1 is patient II.2. Case 2 is patient III.2.

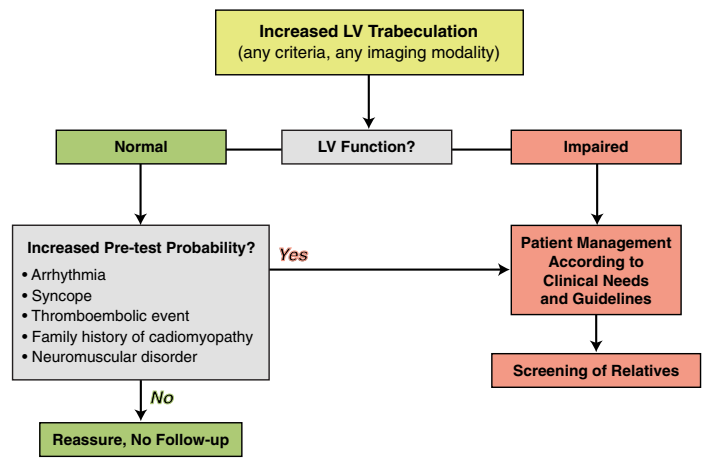


FIGURE 4 Clinical Algorithm Guiding Management of Patients With Increased LV Trabeculations

Case 1 follows the path of impaired LV systolic function **(pink boxes)** where the diagnosis is clear cut, but this case report highlights the current lack of evidence on whether the treatment should be different from the conventional management of dilated cardiomyopathy. Case 2's palpitations, along with normal systolic function would, in isolation, not have suggested a diagnosis of LVNC. However, Case 2 (III.2 in Figure 3) is the son of Case 1, and the follow-up CMR detected adverse cardiac remodeling over time. Case 2 illustrates the importance of a thorough family history and of pre-test probability. CMR is excellent at quantifying LV volumes, systolic function, and the extent of LV trabeculations. However, if marked LV trabeculations are not accompanied by suggestive clinical features or other CMR abnormalities associated with nonischemic cardiomyopathies, a diagnosis of LVNC should not be assumed, and patients should not be managed as having this poorly understood condition. Abbreviations as in Figure 1. (Zemrak F, Petersen SE. Spot the difference: LV trabeculation vs. LV non-compaction. Cardiology Today, February 2015. Reproduced with permission from SLACK Incorporated.)

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KEY WORDS cardiac magnetic resonance, left ventricular noncompaction, left ventricular trabeculations

APPENDIX For accompanying videos and their legends, please see the online version of this article.