

Figure 1.

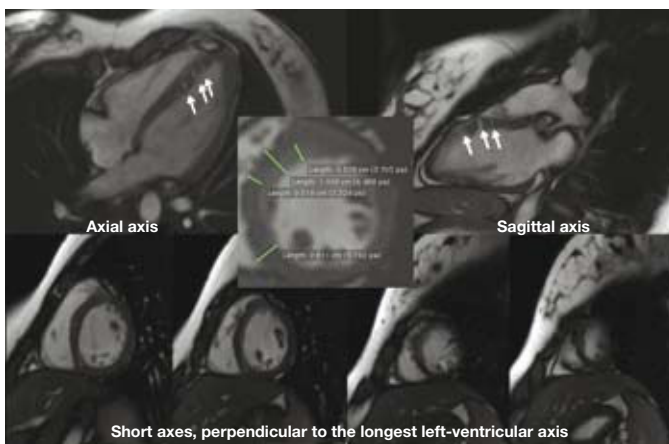


Figure 2.

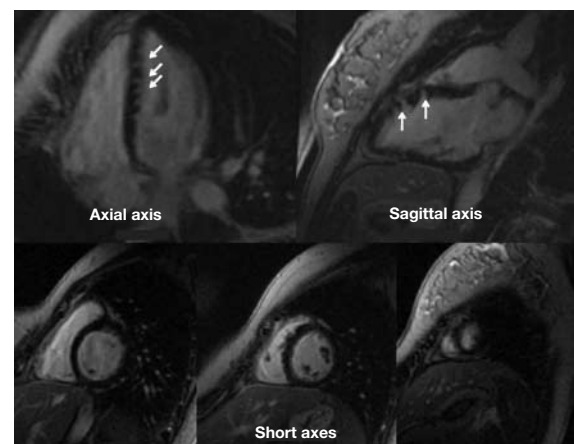


Figure 3.

Left Ventricular Hypertrabeculation or Noncompaction: Are New Diagnostic Criteria Required?

A 23-year-old asymptomatic woman was referred to rule out hypertrophic cardiomyopathy (HCM), which had already been diagnosed in her father by echocardiography. The electrocardiogram (ECG) was abnormal (Figure 1). Echocardiography showed increased apical trabeculation, and a relaxation alteration was seen on Doppler study.

Cardiac magnetic resonance (CMR) confirmed the increased anteroapical and septal trabeculation (Figure 2, arrows) in steady-state free precession sequences, and showed no contractility alterations. The mid-apical short-axis view presented a different signal intensity (SI) in the antero-septal region, disclosing an epicardial layer and another with a lower SI, which corresponded to scant myocardium density of the trabeculated stratum. Based on the current criteria for the diagnosis of noncompaction cardiomyopathy (NCC) by CMR (ratio of noncompacted to compacted myocardium >2.3 in diastole), this diagnosis could

not be established. The early and late phases of gadolinium enhancement (Figure 3) were normal.

In 68% of all post mortems and in recent studies using CMR, limited areas of noncompacted myocardium have been found in normal hearts and in patients with heart diseases, such as HCM, dilated cardiomyopathy (DCM), and even hypertensive cardiomyopathy.

Of note in this case was the considerable diagnostic value of the medical history (paternal background) and the ECG, which led us to suspect a partial phenotypic expression of NCC, considering the great variation in the manifestations of sarcomere mutations. Defects in the MYH7 gene are of particular interest here, being implicated in the family history of HCM and DCM and recently, in NCC.

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