

## Rare case of mitral annulus disjunction and noncompaction-like myocardium

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A 74-year-old man presented to the cardiology office with exertional dyspnea. Transthoracic echocardiography showed mild left ventricular (LV) dilation, LV hypertrophy, LV ejection fraction 50%, mitral valve prolapse and bicuspid aortic valve. Stress echocardiography was nonrevealing. Coronary angiography showed non-obstructive coronary artery disease. Cardiac magnetic resonance imaging was performed, and showed a 9.1 mm atrial displacement of the posterior mitral valve leaflet at the hinge point during systole; the finding was consistent with mitral annulus disjunction (Fig. 1A, **Suppl. Video 1**). Cardiac magnetic resonance imaging also showed spongy/noncompacted myocardium in the LV, with a noncompaction-to-compaction ratio of 1.86 during end-diastole (Fig. 1B). Late gadolinium enhancement images did not show myocardial fibrosis or presence of LV thrombus. 24-hour Holter monitoring showed occasional premature ventricular complex, but no ventricular tachycardia.

Mitral annulus disjunction and LV noncompaction are rare arrhythmogenic cardiac anomalies. Mitral annulus disjunction is most commonly seen in patients with mitral valve prolapse. Mitral annulus disjunction distance > 8.5 mm, and myocardial fibrosis of LV papillary muscles and basal inferior wall are high risk features for ventricular arrhythmias. For patients with LV noncompaction, LV systolic impairment is associated with increased risk of sudden cardiac death. Competitive sports should be refrained in individuals with LV noncompaction and systolic impairment.

According to the current guidelines, implantable-cardioverter defibrillator is not indicated in patients with mitral annulus disjunction or LV noncompaction unless there is a history of sustained ventricular tachycardia, ventricular fibrillation, or aborted sudden cardiac death. To our best knowledge, this is the first reported case describing a patient with concomitant mitral annulus disjunction and LV ‘spongy’ noncompaction-like myocardium on cardiac magnetic resonance imaging.

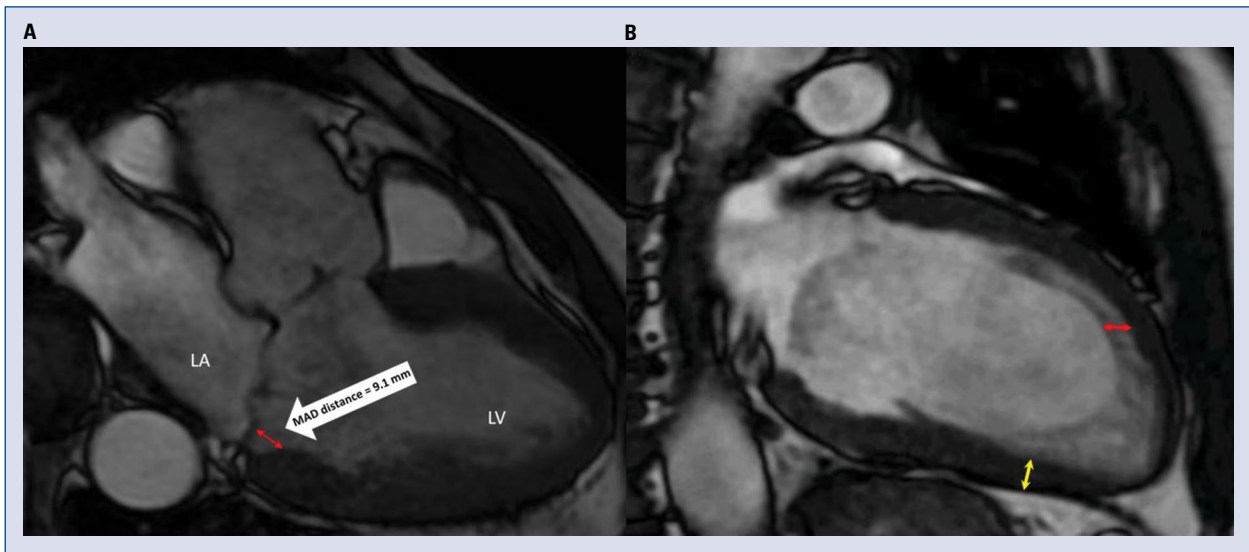
**Conflict of interest:** None declared

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**Figure 1. A.** Cardiac magnetic resonance imaging (MRI) showed mitral annulus disjunction (MAD) with MAD distance of 9.1 mm during systole (red arrow); **B.** Cardiac MRI showed noncompacted myocardium at the apical cap, all apical segments, and mid-lateral wall with noncompacted to compacted myocardial thickness ratio of 1.86 during end-diastole. Red arrow — noncompacted myocardium; yellow arrow — compacted myocardium; LA — left atrium; LV — left ventricle.