

## Cardiac imaging high-risk features of malignant mitral valve prolapse

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Mitral valve prolapse (MVP) is an underappreciated cause of sudden cardiac death (SCD). There are some imaging test signs that can help identify high-risk cases which may result in serious outcomes.

A 35-year-old woman suffered sudden loss of consciousness at home. Emergency services observed a ventricular fibrillation rhythm, and after the delivery of electric shocks, sinus rhythm returned, showing negative T waves in inferolateral leads (Fig. 1A). Two-dimensional echocardiogram showed prolapse and thickness of mitral leaflets and mitral annulus disjunction (separation of mitral leaflet and left atrial junction from left ventricular posterior wall during systole) (Fig. 1B). Doppler echocardiography showed tissue supranormal longitudinal strain in left ventricle posterolateral wall (Fig. 1C). Cardiac magnetic resonance (CMR) confirmed mitral annulus disjunction and showed

pathological late gadolinium enhancement (LGE) in papillary muscles and inferolateral left ventricular wall (Fig. 1D, E). Pickelhaube sign (mitral annulus velocity > 16 cm/s) was also observed in the Doppler study (Fig. 1F). This sign is so called because of its similarity to an old Prussian soldiers' helmet (Pickelhaube, in German) (Fig. 1G). Coronary artery disease was ruled out, and an implantable cardioverter-defibrillator was implanted. The substrate for ventricular arrhythmia is the presence of fibrosis, identified by LGE in CMR, in papillary muscles and inferobasal wall, due to an abnormal systolic mechanical stretch of the myocardium adjacent to the valve. Fortunately, most patients with these signs do not experience such a malignant course, but the finding of the described characteristics in MVP patients should alert us to the risk of SCD and lead to a careful monitoring and evaluation of the patients.

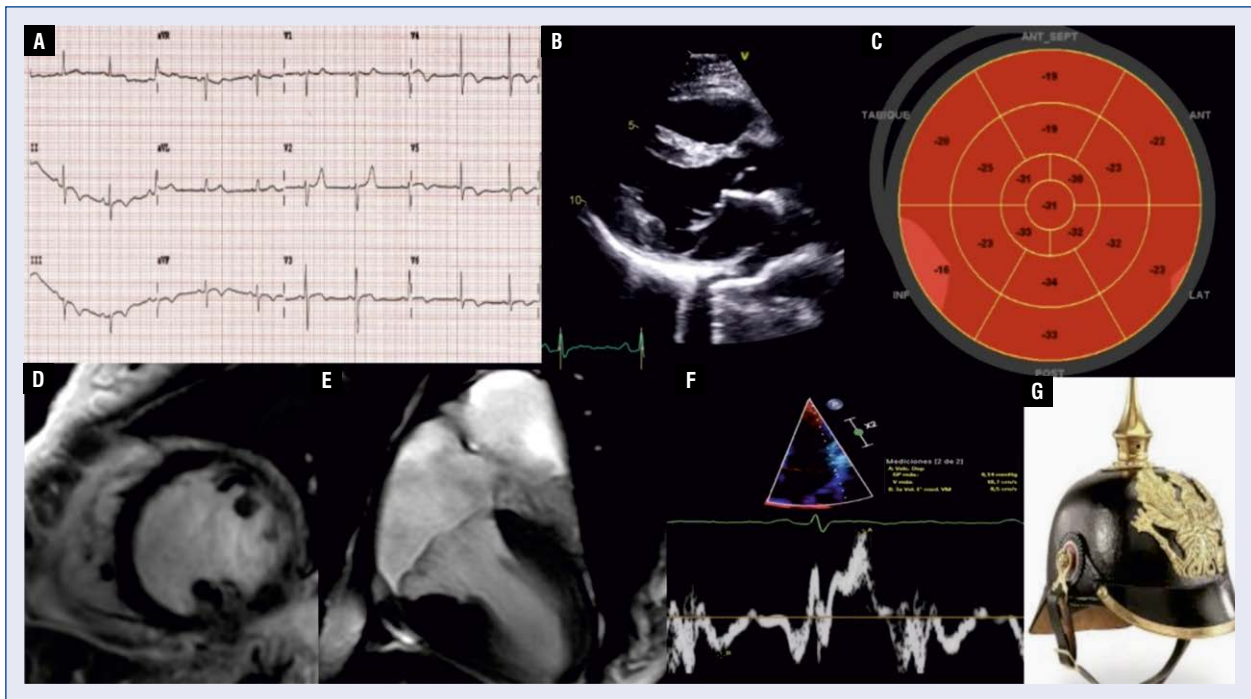
**Conflict of interest:** None declared

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**Figure 1.** **A.** Baseline electrocardiogram in sinus rhythm, showing negative T waves in inferior and lateral leads; **B.** Two-dimensional echocardiogram (parasternal long-axis view), showing prolapse and thickness of both mitral leaflets and mitral annulus disjunction; **C.** Doppler echocardiography showing tissue supranormal longitudinal strain in left ventricle posterolateral wall; **D, E.** Cardiac magnetic resonance, showing pathological late gadolinium enhancement in papillary muscles and inferolateral left ventricular wall (**D**) and mitral annulus disjunction (**E**); **F.** Doppler study with the Pickelhaube sign (mitral annulus velocity > 16 cm/s), so called because of its similarity to an old Prussian soldiers’ helmet (Pickelhaube, in German) (**G**).