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Malignant arrhythmia associated with mitral annular disjunction — myocardial work as a potential tool in search for a substrate

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A 28-year-old woman with no history of cardiovascular disease survived witnessed sudden cardiac arrest, which happened at rest. She was successfully resuscitated and did not present any neurological deficit. The family history regarding sudden cardiac death was negative. Electrocardiogram showed no abnormalities specific for channelopathies. Transthoracic echocardiography revealed mitral valve prolapse with insignificant regurgitation, the morphology and function of both ventricles were normal. Cardiac magnetic resonance detected neither late gadolinium enhancement nor other features of structural heart disease. Coronary arteries were normal as assessed in computed tomography angiography. The patient underwent implantation of subcutaneous cardioverter-defibrillator.

Two years later she was admitted to our hospital due to ventricular fibrillation terminated by the device shock. Electrocardiogram (ECG) presented no relevant changes, corrected QT

interval was 444 ms (Figure 1A). The 24-hour ECG monitoring revealed premature ventricular complexes of several morphology and non-sustained ventricular tachycardia (Figure 1B). This arrhythmia was also induced during treadmill exercise test, especially in the recovery phase. In echocardiography the only pathological findings were left atrial dilation, mitral valve prolapse associated with mitral annular disjunction, mild mitral regurgitation and considerable tugging of papillary muscles (Figure 1C; Supplementary material, *Video S1*). Pickelhaube sign (high, spiked systolic velocity profile resembling German military helmet) was evident in tissue Doppler imaging (Figure 1D). The analysis of myocardial work (MW) demonstrated increased MW index in inferolateral wall of the left ventricle (Figure 1E–F). The therapy with beta-blocker was intensified. In over one-year follow-up no interventions of subcutaneous cardioverter-defibrillator were observed.

Mitral valve prolapse occurs in 2%–3% of the general population [1]. In most cases it is a benign entity but rarely it may be associated with malignant ventricular arrhythmias. The main risk factors include: leaflet thickness >5 mm, bileaflet prolapse, papillary muscle fibrosis and mitral annular disjunction [1, 2]. Zienciuk-Krajka et al. [3] reported recently that among patients with arrhythmic mitral valve prolapse, survivors of cardiac arrest had higher E/e' ratio. Myocardial work represents a relatively new echocardiographic parameter. Owing to the combination of speckle tracking technique assessing longitudinal strain and blood pressure measurements it reflects global as well as regional left ventricular work [4]. Application of this method provided insight into the possible substrate for arrhythmia in the presented patient. Excessive local contraction and stretch of cardiomyocytes may predispose to premature depolarization and trigger ventricular tachycardia or fibrillation via R/T phenomenon [2, 5]. Further studies are needed in order to evaluate the diagnostic and prognostic value of MW in patients with mitral valve prolapse.

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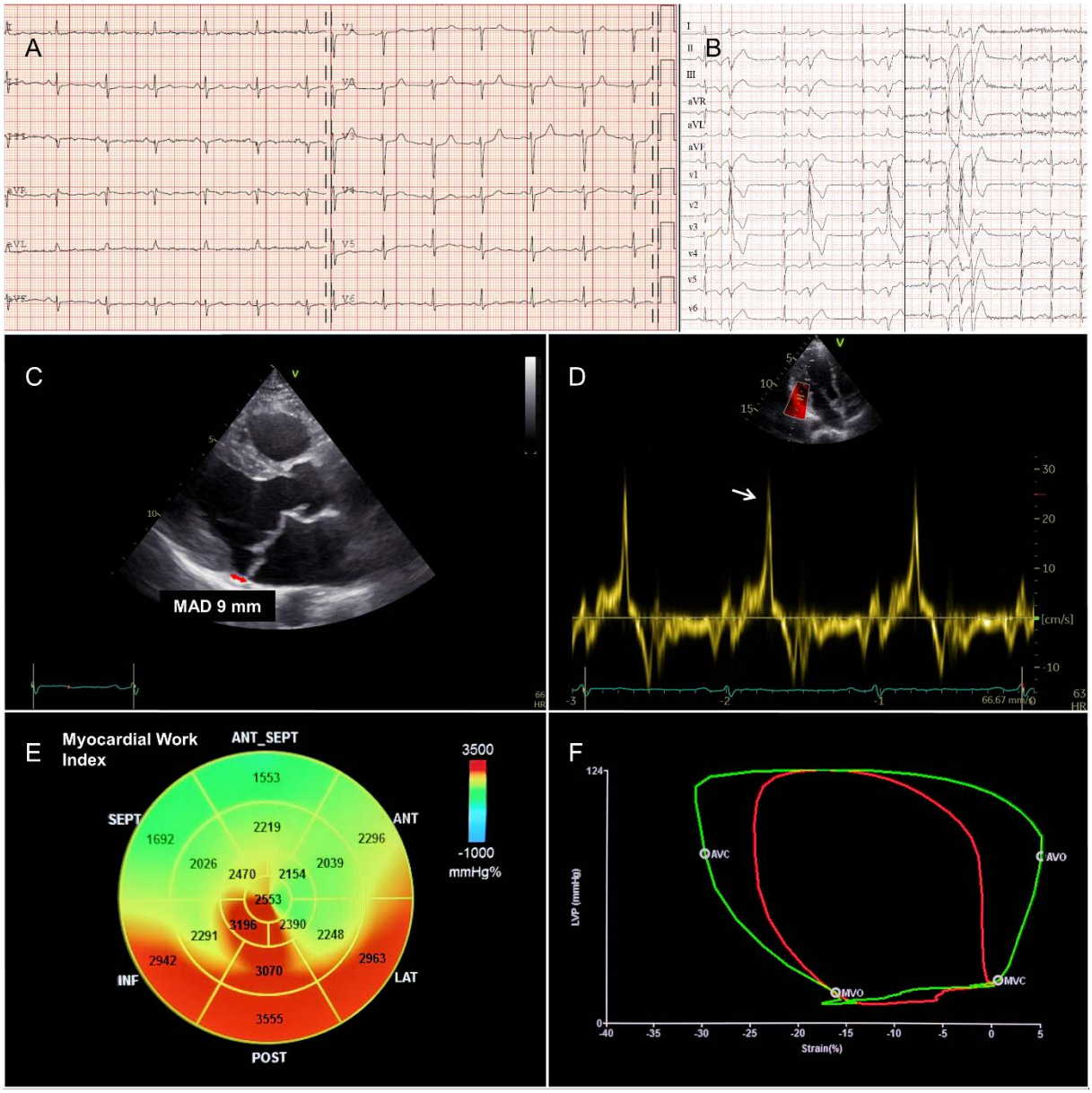

Figure 1. A. Electrocardiogram shows no relevant abnormalities. **B.** Holter electrocardiogram record of premature ventricular complexes and non-sustained ventricular tachycardia. **C–D.** Transthoracic echocardiogram presents mitral annular disjunction in 2D (red double arrow) and Pickelhaube sign in tissue Doppler imaging (white arrow). **E–F.** Myocardial work analysis demonstrates significantly increased MW index in inferolateral wall of the left ventricle depicted as bull's-eye plot and pressure-strain loop of the basal segment (green — basal posterior, red — average, blood pressure — 124/72 mm Hg).

Abbreviations: MAD, mitral annular disjunction; MW, myocardial work