

## Mitral annulus disjunction as an incremental risk factor for ventricular arrhythmia in young patient

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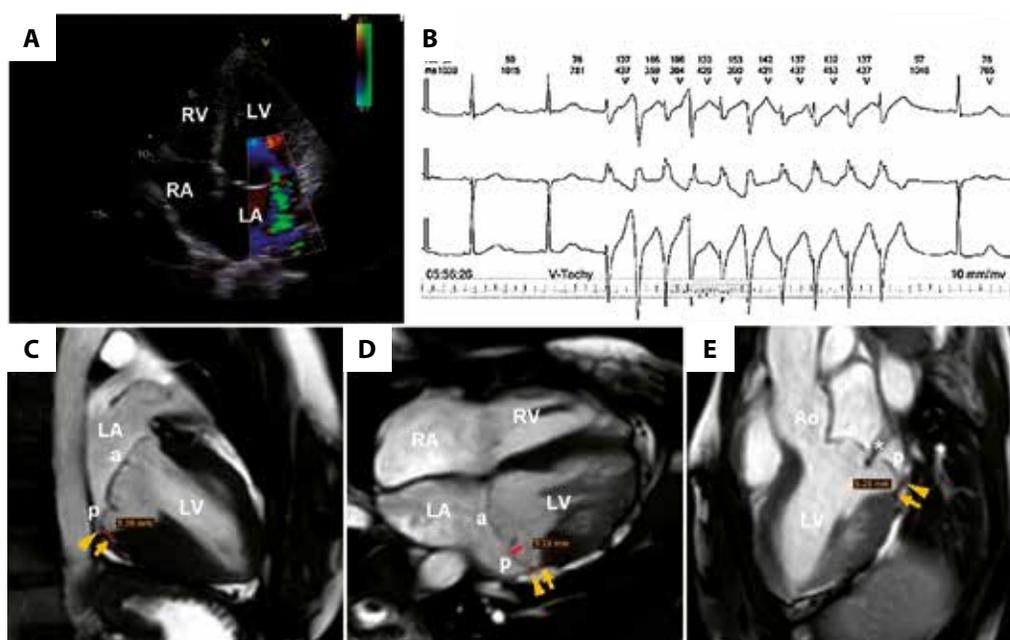
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Mitral annulus disjunction (MAD) is an abnormality defined as the distance of  $\geq 2$  mm between the mitral valve leaflet insertion point into the left atrial wall and the left atrium's connection point to the ventricular myocardium measured in systole in transthoracic echocardiography (TTE) or any distance between the abovementioned points in cardiac magnetic resonance (CMR) or coronary computed tomography angiography [1, 2]. MAD was found in 20%–32.6% of patients with mitral valve prolapse (MVP) [1, 3, 4]. On the contrary, MVP was found in 78% of patients with MAD [5].

A 23-years-old female patient was referred to our department with palpitations and presyncope recurring for a few months. There was no coronary artery disease, cardiomyopathies, channelopathies, arrhythmias, or sudden cardiac death in her family history. Resting electrocardiogram showed normal sinus rhythm without any specific abnormalities observed in channelopathies, corrected QT (QTc) 440 ms (Supplementary material, *Figure S1*). TTE revealed MVP with moderate mitral regurgitation (*Figure 1A*). 24-hour Holter recorded non-sustained ventricular tachycardia (VT) of frequency 140 bpm, varying in cycle length, lasting 4.2 seconds, preceded by bradycardia 50 bpm with normal QTc 402 calculated with Bazett's formula for the last sinus evolution before arrhythmia at night (*Figure 1B*), 27 premature ventricular contractions per hour, and median QTc interval 414 ms. Due to the strong suspicion of MAD, CMR was performed. CMR confirmed MAD and showed a longitudinal MAD distance of 5.2 mm (*Figure 1C–E*), posterior mitral valve leaflet billowing up to 7 mm, and moderate mitral regurgitation with a re-

gurgitant fraction of 21%; there was no late gadolinium enhancement in the left ventricle wall as well as papillary muscles, and left ventricle ejection fraction (LVEF) was as much as 54%. Clinical presentation, resting electrocardiogram, and 24-hours Holter allowed excluding common channelopathies as potential risk factors for VT. Besides, according to Schwartz et al. diagnostic criteria for congenital LQTS, the patient was of low probability of LQTS. In contrast, TTE and CMR allowed excluding cardiomyopathies.

Mitral annulus disjunction arrhythmic syndrome is a clinically significant diagnosis evidenced by several clinical studies. In the study by Dejgaard et al. [5], ventricular arrhythmias (VA) (non-sustained VT, sustained VT, or aborted cardiac arrest) are postulated to occur in 34% of patients with MAD. In the same study, 71% of patients reported palpitations, 41% demonstrated presyncope, and 13% experienced syncope [5]. Young age, lower LVEF, papillary muscle fibrosis [5], and disjunction distance  $> 8.5$  mm [3] are markers for the prediction of VA. Sudden cardiac death might occur in up to 3.8% of patients with MAD [4]. MAD should be considered in younger patients with no other cause for VA or presyncope/syncope of uncertain etiology [5]. Our patient VA occurred despite preserved LVEF, absence of papillary muscles late gadolinium enhancement, and MAD distance  $\leq 8.5$  mm. We administered metoprolol succinate 25 mg once a day in VT prevention and referred the patient for subcutaneous loop recorder implantation. In further studies, in patients with the MAD arrhythmic syndrome, pharmacological or device therapies should be evaluated [5].



**Figure 1.** **A.** Transthoracic echocardiography presenting moderate mitral regurgitation and mitral valve posterior leaflet prolapse in apical four-chamber view. **B.** Non-sustained ventricular tachycardia, varying in cycle length, preceded by bradycardia with normal corrected QT recorded at night in 24-hour Holter. **C–E.** Cinematographic sequences of cardiac magnetic resonance in **C.** vertical long axis (two chambers) view; **D.** horizontal long axis (four chambers) view, and **E.** sagittal left ventricle outflow tract (three chambers) view showing mitral annulus disjunction distance of 5.2 mm measured between mitral valve posterior leaflet insertion point into the left atrial wall (orange arrowhead) and the left atrium's connection point to the ventricular myocardium (orange arrow), mitral valve posterior leaflet billowing of 7 mm (red two-headed arrow in panel E) and moderate mitral regurgitation jet (white asterisk in panel F).

Abbreviations: a, mitral valve anterior leaflet; Ao, ascending aorta; LA, left atrium; LV, left ventricle; p, mitral valve posterior leaflet; RA, right atrium; RV, right ventricle

## Article information

**Conflict of interest:** None declared.

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## REFERENCES

1. Konda T, Tani T, Suganuma N, et al. Mitral annular disjunction in patients with primary severe mitral regurgitation and mitral valve prolapse. *Echocardiography*. 2020; 37(11): 1716–1722, doi: 10.1111/echo.14896, indexed in Pubmed: 33091171.
2. Wunderlich NC, Ho SY, Flint N, et al. Myxomatous mitral valve disease with mitral valve prolapse and mitral annulus disjunction: clinical and functional significance of the coincidence. *J Cardiovasc Dev Dis*. 2021; 8(2), doi: 10.3390/jcdd8020009, indexed in Pubmed: 33498935.
3. Bennett S, Thamman R, Griffiths T, et al. Mitral annulus disjunction: A systematic review of the literature. *Echocardiography*. 2019; 36(8): 1549–1558, doi: 10.1111/echo.14437, indexed in Pubmed: 31385360.
4. Putnam AJ, Kebed K, Mor-Avi V, et al. Prevalence of mitral annulus disjunction in patients with mitral valve prolapse and severe regurgitation. *Int J Cardiovasc Imaging*. 2020; 36(7): 1363–1370, doi: 10.1007/s10554-020-01818-4, indexed in Pubmed: 32221771.
5. Dejgaard LA, Skjølsvik ET, Lie ØH, et al. The mitral annulus disjunction arrhythmic syndrome. *J Am Coll Cardiol*. 2018; 72(14): 1600–1609, doi: 10.1016/j.jacc.2018.07.070, indexed in Pubmed: 30261961.