



## EDITORIAL COMMENT

## The dark side of mitral annular disjunction

## O lado negro da disjunção do anel mitral

Ana Teresa Timóteo<sup>a,b</sup><sup>a</sup> Cardiology Department, Santa Marta Hospital, Centro Hospitalar Universitário Lisboa Central, Lisbon, Portugal<sup>b</sup> NOVA Medical School, Lisbon, Portugal

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Mitral valve prolapse (MVP) is a very common condition, affecting 2–3% of the general population.<sup>1</sup> In most cases, it is benign. However, a small subset of patients are at high risk of malignant ventricular arrhythmias (VAs).<sup>1</sup> The mechanisms underlying arrhythmias in patients with MVP are incompletely understood, but there is clearly an association with bileaflet MVP, papillary muscle fibrosis, and mitral annular disjunction (MAD).<sup>2</sup> MAD is defined as an abnormal atrial displacement of the hinge point of the mitral valve away from the ventricular myocardium, with loss of mechanical annular function, and is usually observed at the insertion of the posterior leaflet.<sup>1,2</sup> However, cardiac magnetic resonance imaging (CMRI) studies show that MAD is a circumferential continuum, interspersed with regions of apparently normal mitral annulus.<sup>2</sup> The reported prevalence is highly variable (between 20% and 58%), depending on the clinical context as well as on the imaging method used.<sup>1</sup> A population study using CMRI found disjunction in 76%, most commonly at the anterior and inferior ventricular wall. Prolapse was more frequent in subjects with disjunction.<sup>3</sup> MAD is thus a common finding on CMRI. Nevertheless, disjunction at the inferolateral ventricular wall was rare, supporting the notion that only extensive inferolateral disjunction warrants consideration of further investigation and that disjunction elsewhere in the annulus should be considered a normal finding.<sup>3</sup> A previous study showed a high prevalence of VAs in patients with MAD, independently of concomitant MVP. One fifth did not have MVP and one-tenth had life-threatening arrhyth-

mic events.<sup>1</sup> Frequent premature ventricular contractions (PVCs) are usually reported.<sup>2</sup>

The arrhythmic mitral valve complex is defined by the presence of MVP (with or without MAD), combined with frequent and/or complex VAs in the absence of any other well-defined arrhythmic substrate.<sup>1</sup> A recent study monitored patients with arrhythmic MVP, defined by the concomitant presence of MAD or VAs, using an implantable loop recorder (ILR) or an implantable cardioverter-defibrillator (ICD) for secondary prevention. During a three-year follow-up, severe VAs were recorded in 12% of patients with an ILR and 20% of those with an ICD, with a high yearly incidence of VAs, even in patients without previous severe arrhythmias (in the ILR group, annual incidence of 4% per person/year).<sup>4</sup>

There are various determinants of arrhythmias, such as syncope, T-wave inversion (mostly in the inferior and lateral leads), QT prolongation, fragmented QRS, premature ventricular contractions, VAs on Holter monitoring, severe mitral regurgitation, presence and distance of MAD, severe myxomatous degeneration, and fibrosis within the mitral apparatus by CMRI.<sup>1</sup> Therefore, risk stratification for VAs is essential in patients with MVP and a complete clinical assessment, electrocardiogram (ECG), Holter monitoring, echocardiography and CMRI (when echocardiography is equivocal and in patients with documented severe VAs) are warranted in these patients.

The paper published by Faria et al. in the current issue of the *Journal* reports two clinical cases of patients with MAD, and highlights the risk of VAs, as well as the difficulties of management.<sup>5</sup> The first case describes a patient

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with sustained ventricular tachycardia who required emergent electrical cardioversion, while the second case is of a young woman with very frequent PVCs; both had documented fibrosis at the MAD level. The article stresses the importance of risk stratification in patients with MAD.

Guidelines for this entity recommend that frequent PVCs should be treated medically, for instance with beta-blockers.<sup>1</sup> A primary prevention ICD is indicated in symptomatic heart failure with an ejection fraction  $\leq 35\%$  after three months of optimal medical therapy.<sup>1</sup> A secondary prevention ICD is indicated in patients with MVP and a documented history of sudden cardiac arrest with ventricular fibrillation/tachycardia without reversible causes. An ICD should be a strong consideration in patients presenting with unexplained syncope and high-risk VAs detected by ECG, Holter, ILR or exercise testing. Other options, such as PVC and VA ablation and mitral valve surgery, may also be considered in more severe and refractory cases.

Therefore, MAD is a common finding, not always associated with MVP, but with a potential risk for VAs, and a thorough assessment is warranted in these patients to identify those who would benefit most from invasive treatment options.

### Conflicts of interest

The author has no conflicts of interest to declare.

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