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EDITORIAL COMMENT

Mitral Annular Disjunction and Fibrosis in Bileaflet MVP

High-Risk Phenotype for Sudden Cardiac Arrest

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The recognition of arrhythmic mitral valve prolapse (AMVP) has evolved significantly over decades. Early reports in the 1960s, following Barlow's description of the valvular origin of midsystolic clicks and murmurs, included sporadic cases of ventricular arrhythmias and sudden cardiac death (SCD) in patients with mitral valve prolapse (MVP). With better understanding of the mitral annulus saddle shape and standardized diagnostic criteria,[1] past cases could not be formally linked to AMVP, and MVP without significant mitral regurgitation (MR) was largely deemed benign.[2] Subsequent large-scale studies[3] revealed that severe ventricular arrhythmias occur in only a small proportion of patients with MVP, explaining why this subset went largely unnoticed amid the broader, predominantly asymptomatic population.[4] However, these rare cases can have devastating consequences, including SCD, prompting renewed interest in identifying high-risk phenotypes through advanced imaging and risk stratification.[5,6] This evolution underscores the shift from viewing MVP as uniformly low risk to recognizing AMVP as a circumscribed, distinct entity warranting targeted surveillance.

In the case presented by Kachhwaha et al,[7] a 42-year-old woman with a longstanding diagnosis of bileaflet MVP and a family history of the condition experienced an out-of-hospital sudden cardiac arrest due to ventricular fibrillation. Despite prior evaluations, including a stress echocardiogram in 2020 showing mild MVP and MR, and ambulatory monitoring in 2021 revealing premature ventricular contractions (PVCs) without sustained arrhythmias, she collapsed without warning. Postarrest multimodality imaging—transthoracic echocardiography (TTE), cardiac magnetic resonance (CMR), and coronary computed tomography angiography—uncovered posterior mitral annular disjunction (MAD) measuring 17 mm, focal late gadolinium enhancement (LGE) at the posterior mitral insertion site, moderate MR, and marked left ventricular (LV) dilation (LV end-diastolic volume index: 150 mL/m^2). Management included implantable cardioverter-defibrillator (ICD) placement for secondary prevention, with deferral of mitral valve repair given the absence of severe MR or heart failure symptoms. Follow-up showed clinical stability without recurrent events.

Several key lessons emerge from this case:

First, the patient was exceptionally fortunate to survive a prolonged cardiac arrest (approximately 30 minutes of resuscitation with 6 defibrillations), highlighting the unpredictable and potentially fatal nature of AMVP.[8]

Second, earlier rhythm monitoring could have identified escalating arrhythmic risk: Indeed, the 2022 European Heart Rhythm Association (EHRA) consensus document proposes a two-tiered risk-stratification model: The phenotypic assessment (eg, bileaflet MVP, posterior MAD, electrocardiographic repolarization abnormalities, LGE, LV remodeling) evaluates the risk of developing AMVP and thus the frequency and duration of rhythm monitoring. The arrhythmic burden evaluation (eg, nonsustained ventricular tachycardia, frequent PVCs, unexplained syncope or presyncope) identifies arrhythmias that

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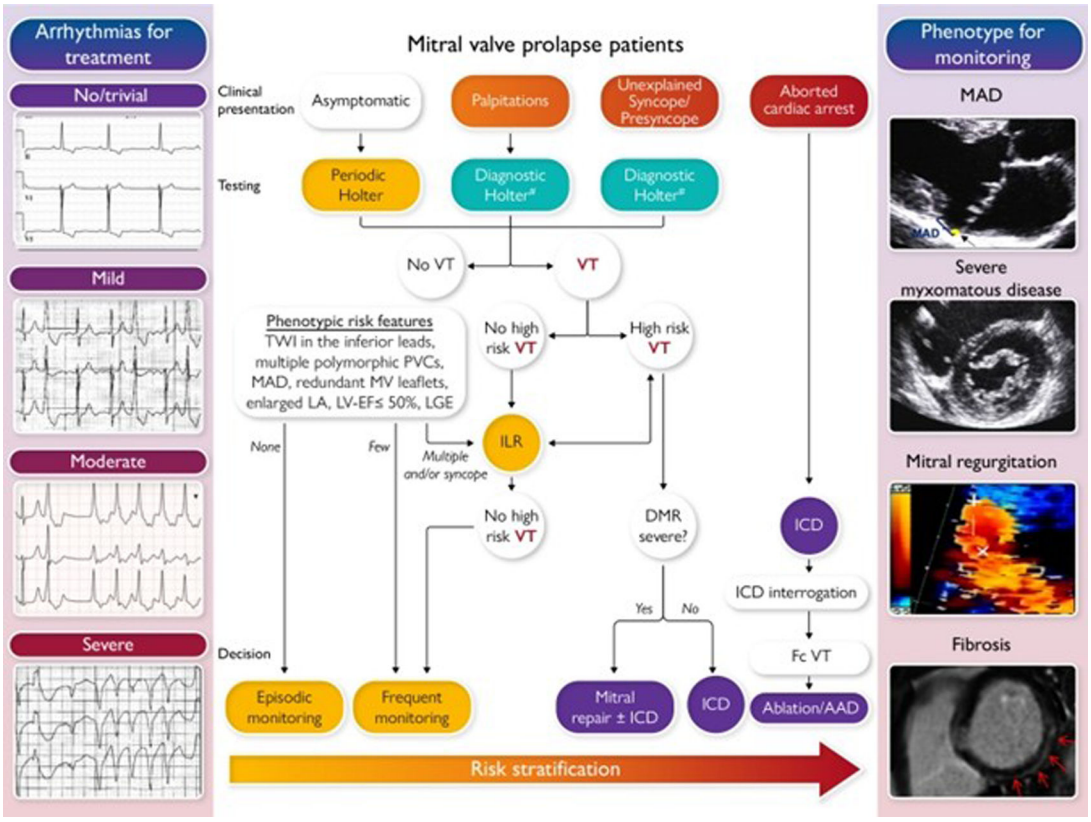
FIGURE 1 Proposed Risk Stratification Algorithm for Arrhythmic Mitral Valve Prolapse, Integrating Phenotypic Markers and Arrhythmic Assessment to Guide Surveillance and Intervention



Adapted with permission from the European Heart Rhythm Association (EHRA) consensus statement[5] and Essayagh et al.[9]
AAD = antiarrhythmic drug therapy; DMR = degenerative mitral regurgitation; ICD = implantable cardioverter-defibrillator; ILR = implantable loop recorder; LA = left atrium; LV-EF = left ventricular ejection fraction; MAD = mitral annular disjunction; MV = mitral valve; PVC = premature ventricular contraction; TWI = T-wave inversion; VT = ventricular tachycardia.

may warrant immediate treatment and, if not, the requirements for repeated monitoring.[5] In our recent review,[9] we outlined a similar tiered approach integrating TTE for valve morphology and MAD, CMR for fibrosis and remodeling, and extended ambulatory monitoring (Figure 1). This patient, with prior syncope, PVCs, and inferior T-wave inversions on electrocardiography, was only captured at the algorithm extreme right—at the SCD stage—but she may have been pre-emptively diagnosed by an earlier repeated monitoring in view of the many phenotypic features suggestive of AMVP, emphasizing the need for routine rhythm surveillance in all MVP patients, with frequency of repeats determined by symptoms, initial arrhythmia burden, and presence of phenotypic AMVP traits.

Third, the case exemplifies the critical phenotypical evaluation. This patient exhibited the full triad of AMVP risk features: MVP with redundant leaflets and extensive posterior MAD, electrocardiographic repolarization abnormalities, and CMR-detected focal LGE indicative of myocardial fibrosis.[10,11] MAD, even after mitral surgery, is independently associated with the development of subsequent arrhythmias.[12] Fibrosis on CMR, particularly in the perimitral region and even in the absence of severe MR or reduced ejection fraction, supports a substrate-trigger model, where MAD-induced mechanical stress fosters fibrosis (substrate), and prolapse dynamics precipitate arrhythmias (trigger).[11] LV dilation is frequently noted in AMVP but can be underestimated by TTE, suggesting a more widespread use of CMR to detect subclinical remodeling.[5]

Fourth, MAD characterization remains uneasy,[12] requiring the precise location of the mitral annulus position throughout the cardiac cycle, demonstrating

posterolateral myocardium separation from annulus in systole, with myocardium return under the annulus in diastole. This case seems to show a persistent separation of myocardium from annulus in diastole in certain views, while in others myocardium and annulus appear reunited. This potential systolo-diastolic MAD feature, suggested by some authors,[13] remains in doubt, suggesting that MAD development and extent warrant further investigation to refine diagnostic criteria.

Fifth, regarding arrhythmia detection/treatment, progression from PVCs and syncope to ventricular fibrillation aligns with the electrical instability of AMVPs.[5] Management remains empirical, using ICD for secondary prevention after cardiac arrest, but with more uncertainty for primary prevention.[14] Detection of ventricular tachycardia, severe with fast rates, multifocal, or associated with syncope/pre-syncope (not the phenotype), raises the indication of primary prevention of SCD by ICD implantation, but the exact criteria for defining severe arrhythmias in the context of AMVP are poorly defined.[9] Emerging data suggest that early mitral repair in AMVP with MAD may reduce arrhythmic burden, possibly by alleviating mechanical stress and even in patients without severe MR, but survival benefit remains unproven.[12]

In conclusion, this case illustrates the risk of SCD with MVP, the necessity of regular rhythm monitoring in patients with MVP, and the importance of multimodality imaging in uncovering the AMVP phenotype. By integrating historical insights with modern risk stratification, clinicians can shift from reactive to proactive management, potentially preventing SCD in this under-recognized subset.

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