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EDITORIAL

From an Anatomic Curiosity to a Treatable Arrhythmic Syndrome

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Note

See related article, Montana et al. 2025;3(2): pages 22-28.

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The case series by Montana and colleagues reframes arrhythmogenic mitral valve prolapse (MVP) and mitral annular disjunction (MAD) as clinically actionable entities rather than incidental imaging findings or postmortem diagnoses. Although MVP is common and usually benign, this work reinforces that a distinct subset of patients carries a disproportionate risk of malignant ventricular arrhythmias and sudden cardiac death. By presenting four cases across a wide clinical spectrum, the authors illustrate how arrhythmogenic MVP represents a continuum of mechanical stress, myocardial remodeling, and electrical instability rather than a binary condition.[1]

A central contribution of this article is its emphasis on multimodality imaging—particularly cardiac magnetic resonance imaging (CMR)—as the cornerstone of risk recognition. Several cases demonstrate how transthoracic echocardiography may underestimate or miss high-risk phenotypes, while CMR reveals bileaflet prolapse, MAD, and late gadolinium enhancement (LGE) in the papillary muscles or inferobasal myocardium. These findings provide a mechanistic link between leaflet prolapse, repetitive mechanical traction, myocardial fibrosis, and ventricular arrhythmogenesis. The discussion of echocardiographic markers such as the Pickelhaube sign further highlights how careful imaging interpretation can uncover dynamic clues to arrhythmic risk.

The series also challenges the traditional reliance on mitral regurgitation severity as the primary determinant of prognosis. Montana et al. show that malignant arrhythmias may occur even in patients with mild or moderate regurgitation, particularly when additional risk modifiers—such as MAD, ventricular ectopy, family history of sudden cardiac death, or LGE—are present. This reinforces the need for a layered approach to risk stratification that integrates clinical history, electrocardiography, ambulatory rhythm monitoring, and advanced imaging, rather than a single dominant metric.

Equally important is the paper's treatment of intervention as more than symptomatic management. Mitral valve surgery is presented not only as a hemodynamic solution but also as a potential means of reducing ongoing mechanical stress and

progressive myocardial fibrosis. Similarly, catheter ablation is framed as targeted therapy for scar-mediated ventricular tachycardia when arrhythmias persist despite structural correction. The proposed clinical algorithm, aligned with contemporary European consensus guidance, offers a pragmatic framework while acknowledging gaps in evidence and variability in resource availability.

Overall, this article advances the field by shifting arrhythmogenic MVP from a retrospective diagnosis to a prospective, treatable syndrome. It highlights how thoughtful integration of imaging, electrophysiology, and clinical judgment can identify patients at risk before catastrophic events occur. While unanswered questions remain regarding optimal screening thresholds and timing of intervention, the work by Montana et al. makes a compelling case that MVP should no longer be viewed as uniformly benign and that vigilance, when guided by pathophysiology, can meaningfully alter outcomes.

References

1- Montana P; Goodrich R; Rodriguez-Lozano PF; Morsy M; Prousi GS. Arrhythmogenic Mitral Valve Prolapse and Mitral Annular Disjunction: From Observation to Intervention. J Arrhythm Electrophysiol 2025;3(2):22-28

Conflict of Interests

None

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