Circulation: Cardiovascular Imaging

EDITORIAL

New Kid on the Block: Double-Peak Strain Pattern in Arrhythmic Mitral Valve Prolapse

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Mitral valve prolapse (MVP) is the most common valvular heart disease that affects 1% to 3% of the general population. Most patients are asymptomatic with good long-term prognosis. Arrhythmic MVP, defined as MVP in the presence of frequent or complex ventricular arrhythmia (VA), is a rare feature (<1% per year) among MVP patients.[1] However, it is overrepresented (up to 7%) in patients with otherwise unexplained sudden cardiac death on autopsy series.[2]

See Article by Nagata and Bertrand et al

A typical arrhythmic MVP phenotype would be that of a young female patient with inferior T-wave inversion on ECG, bileaflet prolapsed, and mitral annular disjunction (MAD) on echocardiogram. Other high-risk echocardiographic features that have been suggested include, but are not limited to, the Pickelhaube sign (lateral annular velocity spike of ≥16 cm/s), systolic curling of the posterior wall, marked billowing excursion, mechanical dispersion, and postsystolic shortening.[3] The presence of fibrosis on cardiac magnetic resonance (CMR), either in the form of replacement fibrosis on late gadolinium sequences or interstitial fibrosis on T1 mapping, and extracellular volume measurement has also been shown to predict malignant arrhythmias.[4] Note that not all the aforementioned high-risk imaging parameters are included in the risk stratification framework of the recently published European Heart Rhythm Association expert consensus statement.[1] In asymptomatic patients with MVP, high-risk imaging parameters that warrant further investigations are limited to MAD, redundant mitral leaflets, enlarged left atrium, left ventricular ejection fraction ≤50%, and the presence of late gadolinium enhancement (LGE).[1]

The interaction between the morphology and function of MVP has long been a subject of interest. A common theme that runs through the morphofunctional characteristics of arrhythmic MVP, for example, MAD, systolic curling, and the Pickelhaube sign is a hypercontractile basal inferolateral segment that is subjected to mechanical stretch. Speckle-tracking echocardiography is well suited at assessing myocardial deformation in response to the vectors generated by various components of the mitral valve apparatus during the cardiac cycle.

In this issue of *Circulation: Cardiovascular Imaging*, Nagata and Bertrand et al[5] reported a novel double-peak pattern (DPP) on speckle-tracking echocardiography–derived longitudinal strain curves in patients with MVP. As the name implies, DPP is characterized by 2 distinct peaks. The first peak denotes normal systolic shortening in early systole. This is followed by paradoxical systolic expansion of the mitral annulus in late systole, tugging the basal posterior segment and causing it to lengthen in late systole. The second peak represents postsystolic shortening, as the stretched basal posterior segment recoils to release the tension that was built up earlier. In this study cohort of 113 patients with MVP, DPP was reported in slightly less than half, 47% (n=53) of the study population, mostly in those with fibrosis on CMR (n=35). DPP was also more frequently found in those with superior leaflet displacement, MAD, systolic curling, and papillary muscle displacement.

These findings, specifically the postsystolic shortening, concur with that reported by Huttin et al in an earlier

Key Words: Editorials ■ fibrosis ■ mitral valve prolapse ■ prognosis ■ prolapse

The opinions expressed in this article are not necessarily those of the editors or of the American Heart Association.

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study. In the study by Huttin et al[6] that included patients with MVP and normal LV ejection fraction, pathological early systolic shortening and postsystolic deformation was observed. The present article by Nagata et al offers further insights into the underlying mechanisms of the strain pattern and provides outcome correlation.

Interestingly, this study has shown that DPP is an independent predictor of VAs, regardless of the presence of fibrosis. In fact, the effect of fibrosis on VAs was rendered statistically insignificant after adjusting for DPP (model 7 in Table 3 and Table S4B) or after adjusting for both DPP and history of VA (model 9 in Table 3 and Table S4B). Whether DPP in itself is arrhythmogenic still requires further validation in larger, prospective studies. However, this echoes previous observation that the pathogenesis of arrhythmias in some MVP patients is independent of replacement fibrosis detected by CMR as LGE. Studies have shown that arrhythmias can occur in patients without the evidence of LGE on CMR, for example, those with focal basal posterior hypertrophy or ballerina-foot pattern.[7] VAs in the absence of LGE might be attributed to interstitial fibrosis or other disease processes, for example, inflammation or ischemia.[8–10] Late gadolinium sequence strength lies in its ability to detect replacement fibrosis. However, it lacks sensitivity at detecting interstitial fibrosis compared with parametric mapping techniques, for example, T1 and extracellular volume. Interstitial fibrosis on parametric mapping has been associated with increased risk of cardiac arrest in the absence of LGE.[4] Study using fluorine 18-labeled fluorodeoxyglucose positron emission tomography imaging in patients with MVP and ventricular ectopy showed metabolic activity, a surrogate for myocardial inflammation, or ischemia, which suggests ongoing occult disease activities beyond replacement fibrosis.[11]

In the context of arrhythmia prediction, DPP adds to the expanding list of risk markers in patients with MVP. However, it is unclear whether DPP provides incremental prognostic values beyond the current established risk markers or whether DPP still has a role when all things are considered. Further, larger longitudinal outcome studies are required to better define the role of DPP in arrhythmic

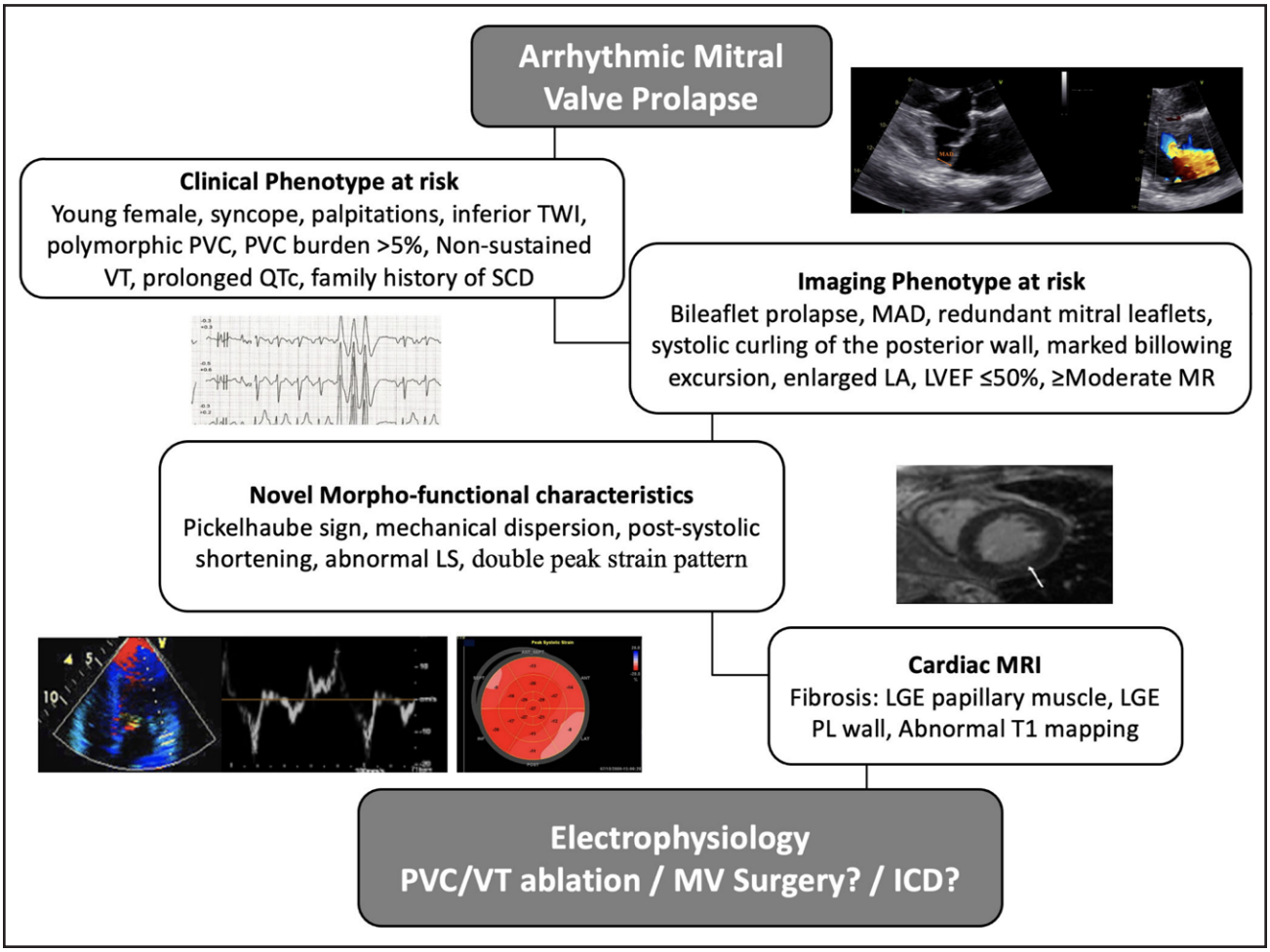

Figure. Features associated with arrhythmic mitral valve prolapse.
ICD indicates implantable cardioverter-defibrillator; LA, left atrium; LGE, late gadolinium enhancement; LS, longitudinal strain; LVEF, left ventricular ejection fraction; MAD, mitral annular disjunction; MR, mitral regurgitation; MRI, magnetic resonance imaging; MV, mitral valve; PL, posterior-lateral; PVC, premature ventricular contraction; SCD, sudden cardiac death; TWI, T-wave inversion; and VT, ventricular tachycardia.

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MVP risk stratification. Also, more data are required to make sense of the relationship between DPP and other imaging markers that are also a result of a hypercontractile inferolateral segment and whether they are indeed different angles that describe the same phenomenon.

Nagata et al also reported decreased regional peak systolic longitudinal strain in the basal inferior and posterior segments, both in MVP patients with and without LGE. This contrasts previous study that reported supranormal peak systolic strain in the posterolateral trident.[3] Nagata et al added that the areas of decreased regional longitudinal strain mirrored the distribution of fibrosis on CMR, thus the decreased longitudinal strain in patients with LGE. However, it does not explain the decreased regional longitudinal strain in patients without fibrosis. On the other hand, the supranormal peak systolic strain parallels the Pickelhaube sign, both the result of a hypercontractile basal posterior segment. Further studies are required to reconcile these contradictory findings.

Risk stratification in arrhythmic MVP is challenging. A multitude of parameters, ranging from imaging to ECG to genetics to electrophysiologic study, have been associated with arrhythmias, although none conclusively and most limited by the size of the studies or the lack of longitudinal follow-up. Compared with hypertrophic cardiomyopathy that has similar annual incidence of sudden cardiac death, risk stratification in arrhythmic MVP is still in its infancy. The European Heart Rhythm Association expert consensus statement is the start of a long journey that calls for constant review of emerging literature and iterative refinement of the recommendations. There are still many gray areas, from screening to risk stratification to management, that could benefit from further guidance and clarification. One area would be the role of surgery in arrhythmic MVP. In patients with severe mitral regurgitation, the decision for surgery is relatively straightforward. However, in patients with less than severe mitral regurgitation, whether arrhythmic MVP lowers the threshold for surgery is unclear. Small studies have suggested that mitral valve repair may reduce the burden of malignant arrhythmias. However, surgery has not been shown to decrease the risk of sudden cardiac death among patients with MVP.[1,2]

Another area that requires further guidance is the role of multimodality imaging in patients with suspected arrhythmic MVP. All patients in this study underwent CMR as part of the study entry criteria. Thus, the present study was not poised to answer the perennial question of who among the MVP patients should undergo a CMR study. The European Heart Rhythm Association expert consensus suggests that CMR may be useful in patients with MVP who have at least 1 phenotypic risk feature, namely palpitations, inferior lead T-wave inversion, polymorphic premature ventricular complexes, MAD, redundant mitral leaflets, enlarged left atrium, or ejection fraction ≤50%.[1]

Arrhythmic MVP is a complex disease entity that has been much talked about but not fully understood or at least not just yet. Collaboration between the imagers, electrophysiologists, pathologists, and the surgeons is needed for us to stand the best chance at resolving the many uncertainties surrounding the most common valve disease in the community—the MVP (Figure).

ARTICLE INFORMATION

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Disclosures

None.

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