Letter by Sheppard et al Regarding Article, “Arrhythmic Mitral Valve Prolapse and Sudden Cardiac Death”

To the Editor:

We read with great interest the excellent article by Basso et al1 on arrhythmic mitral valve prolapse (MVP) and sudden cardiac death in Circulation. The investigators reported a relatively high prevalence of MVP in individuals experiencing sudden death. Such deaths were more common in young women and were associated with myocardial fibrosis in the papillary muscles and the inferobasal wall of the left ventricle. The data were supported by clinical cases of patients with mild MVP and complex ventricular arrhythmias that underwent contrast-enhanced cardiac magnetic resonance. Most patients exhibited late gadolinium enhancement within the papillary muscles, in the adjacent left ventricular wall, and within the inferobasal left ventricular segment beneath the posterior mitral valve leaflet. Some patients also revealed focal endocardial late gadolinium enhancement in the same area, featuring a fibrous plaque. The authors hypothesized that the myocardial fibrosis may be the result of increased tension on the chordae and transmission of this force to the mitral support apparatus.1 Equally important may be the contact lesions that result from the redundant valve prolapsing into the atrium in systole and snapping back against the ventricular myocardium during diastole as highlighted in an accompanying editorial by Noseworthy and Asirvatham.2

We have found very similar pathological findings in cases in our sudden cardiac death cohort that were subject to detailed postmortem examination of the heart.3 Of 3680 cases, we detected 62 cases with isolated MVP (1.7%). Our cohort did not show a female preponderance; the female to male ratio was ≈ 1 (29 females:33 males). We also noted that the majority (74%) died at rest or during sleep. Affected individuals in our series showed predominantly bileaflet mitral valve involvement with an increased heart weight. As reported by Basso et al, approximately half our cohort had an in vivo diagnosis of MVP and arrhythmias. We identified left ventricular fibrosis in 46 (74%) of our cases, involving 1 or both papillary muscles and the adjacent left ventricular wall. Fibrosis was predominantly localized within the posteromedial papillary muscle and the inner wall of the adjacent posteroinferior wall. Moreover, the anterolateral papillary muscle and the adjacent anterolateral wall of the left ventricle were frequently involved. Consistent with the findings of Basso et al, the fibrosis was of the replacement and interstitial type, with subendocardial and midmural distribution, involving the trabeculae, but never transmural. Our autopsy series support the findings of Basso et al and suggest that, in the subset of patients with MVP with high-risk clinical markers and especially complex ventricular arrhythmias, contrast-enhanced cardiac magnetic resonance may play an extremely important role in the management and risk stratification of these patients. Finally, Basso et al reported that MVP made up 7% of the cardiac pathology in victims of SCD, in comparison with just 1.7% in our cohort. We suspect that the higher percentage of MVP in the Italian series reflects the efficacy of their state-sponsored ECG program in excluding other major pathologies such as cardiomyopathies and ion channel diseases.4

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Disclosures
None.

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References
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