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Clinical recognition of patients with myxomatous bileaflet mitral valve prolapse (MBMVP) at risk of sudden cardiac death remains elusive (1). We report our experience using speckle-tracking echocardiography (STE) to identify arrhythmogenic substrate.

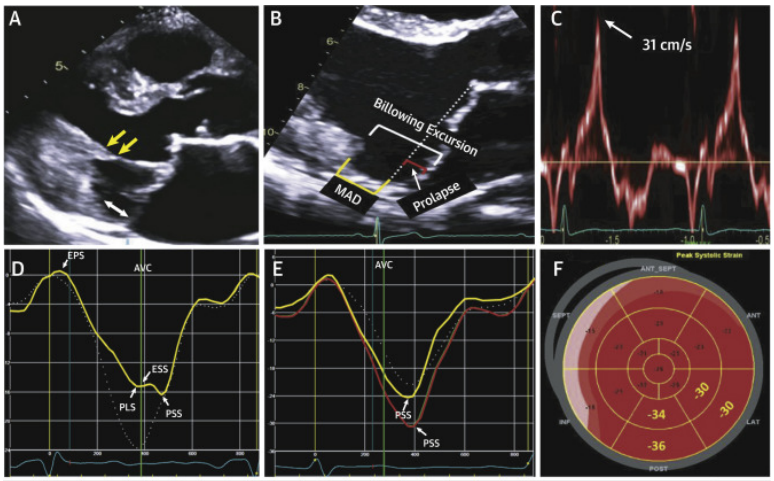
Of 327 patients with MBMVP in our echocardiography database, 54 were referred to the specialized cardiology clinic. Ten patients with severe mitral regurgitation (MR) or flail leaflets were excluded. Baseline demographic and clinical characteristics of 44 patients were collected. The institutional review board approved the study.

Using a GE Vivid E9 or E95 echocardiography machine (GE Healthcare, Waukesha, Wisconsin), the following measurements were recorded: prolapse extent; billowing excursion (BE), defined as maximum displacement of the mitral leaflets from the left ventricular (LV) myocardium; mitral annular disjunction (MAD) (1); and Pickelhaube spike (2). STE assessment of LV deformation was performed off-line from the standard 3 apical views (GE Healthcare EchoPAC) (3). STE parameters measured were early prestretch strain (EPS); peak-systolic strain (PLS); end-systolic strain (ESS); maximum strain in cardiac cycle (PS); postsystolic shortening (PSS) (3); time to peak strain; and mechanical dispersion, defined as standard deviation of the time to peak strain of each segment. Prestretch index (PST) and postsystolic index (PSI) were calculated as: $PST = 100 \cdot EPS/(EPS - ESS)$ and $PSI = 100 \cdot (PS - PLS)/(PS)$. Values of PLS, PSI, and PST were obtained for all 18 echocardiographic segments and averaged for global values (3). LV longitudinal strain bull's-eye plot derived from 2D STE was analyzed for regional strain.

Data are presented as mean ± SD for continuous and frequency/percentage for categorical variables; chi-square or Fisher exact and analysis of variance (ANOVA) or Kruskal-Wallis were used accordingly for comparison. Of 44 patients (mean age 50 ± 15 years; 59% female), 9 had malignant ventricular tachycardia (MVT), 6 nonsustained VT (NSVT), and 29 no VT (NVT). There was no difference in clinical characteristics, standard echocardiographic measurements, severity of MR, leaflet thickness, mitral annular diameters, global PLS, PSI, global and regional PST, time to peak strain, mechanical dispersion, or cardiac magnetic resonance (CMR) features among the groups.

CMR of this MBMVP cohort revealed only 1 of 5 in the MVT, 2 of 4 in the NSVT, and 9 of 21 in the NVT group had late gadolinium enhancement (LGE). Mean regional (median; interquartile range [IQR]) PSI in basal and midlateral wall segments was 10.79 ± 13.70 (5.2; 1.7 to 17.1), 1.71 ± 2.68 (0; 0.0 to 4.5), 1.99 ± 2.71 (1.1; 0.0 to 2.6) in the MVT, NSVT, and NVT groups, respectively ($p < 0.01$). Mean (median; IQR) Pickelhaube spike velocity in cm/s, 23.8 ± 11.1 (19; 14 to 30), 15.3 ± 6.1 (12.5; 11 to 21), and 16.3 ± 5.5 (16; 13 to 19) ($p = 0.019$), and mean (median; IQR) MAD in mm, 11.1 ± 1.5 (12; 10 to 12), 13.8 ± 4.4 (13.5; 10 to 17), 7.8 ± 4.3 (8; 5 to 11) ($p = 0.002$), were significantly higher in the MVT and NSVT groups than NVT group, respectively. Although there was no significant difference among the groups in degree of prolapse, mean BE (median; IQR) was significantly higher in the MVT and NSVT groups than NVT group (16.7 ± 2.6 [15; 15 to 19], 18.5 ± 5.1 [18.5; 15 to 20], 14.1 ± 4.2 [14.0; 10 to 16]; $p = 0.03$). All patients had a higher mean regional strain in the posterolateral trident (basal and midposterior and posterolateral wall segments) compared with the basal and midseptal segments (–24 ± 5% vs.

–15 ± 4% p < 0.01). Univariate logistic regression identified potential predictors of VT (lateral wall PSI, BE, and Pickelhaube spike). Empirical optimal cutoff points for these variables for VT were identified through receiver operator characteristics analyses (Liu method) and sensitivity, specificity, and area under the curve determined (Figure 1). Multivariate logistic regression showed lateral wall PSI ≥4.5% and BE to be statistically significant (p < 0.05).



Ventricular Tachycardia	Odds Ratio	Empirical Optimal Cutoff Point:	Sensitivity At Cutoff Point:	Specificity At Cutoff Point:	Area Under ROC Curve Overall (95%CI)	p Value
Pickelhaube Spike cm/s	2.02	19	53	72	0.60	0.36
Billowing Excursion (BE) mm	6.66	14.5	80	62	0.72	0.03
Lateral Segment PSI	5.36	4.5	53	86	0.60	0.04
Lateral PSI >4.5/ BE>14.5	6.75		33	90	0.80	0.04

Figure 1. Arrhythmic Myxomatous Bileaflet Mitral Valve Prolapse Syndrome
(A) Long-axis view (systole) demonstrating myxomatous bileaflet mitral valve prolapse, mitral annular dysfunction (MAD) **(white arrow)**, and tug by the prolapsing leaflet **(yellow arrows)**. **(B)** MAD = 12 mm; BE = 15 mm; Dotted line = mitral annular plane; prolapse = 3 mm. **(C)** Pickelhaube spike. **(D)** Longitudinal deformation curve. **(E) Red and yellow curves** = basal and mid inferolateral wall. **(F)** Heterogenous longitudinal strain bull's eye. **(Table)** VT optimal cutoff points.

Our observations indicate that mean Pickelhaube spike, BE, and basal and midlateral wall PSI tend to be higher in patients with malignant arrhythmogenic MBMVP. The Pickelhaube sign represents a marker of myocardial stretch caused by tugging of prolapsing mitral leaflets in systole on papillary muscles and adjacent myocardium, thus altering myocardial deformation in these segments (PSI). In contradistinction to a previous report of CMR LGE (LV fibrosis) in MBMVP syndrome (1), we found CMR evidence of LGE in only 33% of patients. This may be because fibrosis occurs at a later stage in the disease. Thus, tissue Doppler and STE can identify patients at risk of malignant ventricular arrhythmias in MBMVP earlier, before development of fibrosis**.**

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The authors attest they are in compliance with human studies committees and animal welfare regulations of the authors' institutions and Food and Drug Administration guidelines, including patient consent where appropriate. For more information, visit the JACC: Cardiovascular Imaging author instructions page.