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Paradoxical restricted motion in diastole associated to mitral valve prolapse/dystrophy: a frequent finding

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Background: Filamin-A mitral valve prolapse/dystrophy (FLNA-MVP) phenotype associates moderate MVP and a paradoxical restricted motion in diastole.

Purpose: We aimed to assess the association of MVP with restricted motion in diastole in MVP patients (restricted MVP).

Methods: We prospectively enrolled 433 MVP probands (57 ± 16 years). Patients underwent a clinical examination and a comprehensive echocardiographic analysis of mitral valve apparatus.

Results Among the 433 probands, 27 (6.2%, 95% CI 3.9-8.5) had both a MVP and a doming aspect in diastole. Patients with restricted MVP exhibited shorter posterior chordae tendinae (24.8 ± 6.3 vs 27.2 ± 5.9 mm, $P = 0.037$), and a shorter distance between papillary muscle (PM) tips and mitral annulus (anterior PM: $P = 0.0001$; posterior PM: $P = 0.009$). Anterior mitral valve leaflet was lengthened (15.5 ± 2.4 vs 14.3 ± 2.6 mm/m², $P = 0.018$), but leaflet thickness, leaflet prolapse, and mitral valve annulus did not differ between the 2 groups. Bicuspid aortic valve was more frequent in patients with restricted phenotype (14.8 vs 2.9%, $P < 0.05$). Familial recurrence of restricted MVP was identified even in the absence of Filamin-A mutation.

Conclusion: Restricted MVP is a quite frequent finding in MVP patients and is associated with PM tips location closer to mitral annulus. Restricted MVP can be regarded as a third type of MVP beside myxomatous Barlow disease and fibro-elastic deficiency MVP.