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*Paradoxical restricted motion in diastole is a frequent finding in mitral valve prolapse/dystrophy patients*

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

Purpose: We aim to assess the association of mitral valve prolapse to restricted motion in diastole in MVP patients (restricted MVP).

Methods: We prospectively enrolled 475 MVP probands (64±13 years) and controls relatives. Patients underwent a clinical examination and a comprehensive echocardiographic analysis of mitral valve apparatus.

Results: Among 475 consecutive probands, 48 (10.1%, 95% CI 7.7-13.3) had both a MVP and a doming aspect in diastole. Patients with restricted MVP exhibited shorted chordae tendinae, and a shorter distance between papillary muscle tip and mitral annulus. Compared with controls, mitral valve leaflets were lengthened, thickened and mitral valve annulus was enlarged. The prevalence of polyvalvular disease and bicuspid aortic valve was not increased in restricted MVP patients compared with conventional MVP. Familial form 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 unique features of the MV apparatus. Restricted MVP can be regarded as a third type of MVP beside myxomatous Barlow disease and fibro-elastic deficiency MVP.