## Mitral annulus disjunction is associated with adverse outcome in patients with Marfan syndrome and Loeys-Dietz syndrome

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**Background:** Mitral valve prolapse is a common finding in patients with Marfan (MFS) and Loeys-Dietz syndromes (LDS). Mitral annulus disjunction (MAD) is an atrial displacement of the hinge point of the mitral valve that frequently coexists with mitral valve prolapse, but its clinical relevance in connective tissue disorders is unknown.

Purpose: To explore the association between MAD and severity of mitral valve and aortic disease in patients with MFS and LDS.

**Methods:** We included consecutive MFS patients and LDS patients fulfilling established diagnostic criteria. MAD was identified by echocardiography and defined as the distance from the ventricular myocardium to the hinge point of the posterior mitral leaflet (Figure, panel A). Aortic surgery was defined as emergency surgery for aortic dissection or prophylactic aortic surgery for aortic aneurysm. We recorded the need of mitral valve surgery including mitral valve repair or replacement.

**Results:** We included 168 patients of whom 103 (61%) had MFS and 65 (39%) had LDS. We identified MAD in 69 (41%) patients. Aortic surgery was performed in 112 (67%) patients (27 dissections and 85 prophylactic interventions). Patients with MAD were younger at the time of aortic surgery than those without MAD (p log rank = 0.02) (Figure, panel B). Patients needing aortic surgery had greater MAD distance (8 [7-10] mm vs. 7 [6-8] mm, p = 0.04). Mitral valve surgery was performed in 12 (7%) patients, more frequently in patients with MAD than in those without (16% vs. 1%, p < 0.001, p log rank < 0.001) (Figure, panel C).

**Conclusion:** MAD was frequent and detected in 41% of patients with MFS and LDS. MAD was associated with a more severe disease phenotype including aortic surgery at younger age and frequent need for mitral valve surgery. Screening patients with MFS and LDS for MAD may provide prognostic information and may be relevant in planning surgical interventions.



