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Left ventricular systolic dysfunction in Marfan syndrome is related to aortic distensibility

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Background: The cardiovascular involvement in Marfan syndrome (MS) is characterized mainly by dilatation and reduced distensibility of the ascending aorta. Left ventricular (LV) dysfunction has been detected in MS and a primary cardiomyopathy has been suggested but the pathophysiology is unclear. The purpose of this study was to evaluate the LV function in patients with MS and to assess the relationship with the aortic distensibility, as possible underlying mechanism.

Methods: We studied 53 patients with MS (27.0 ± 11 years, 29 men) without valvular or hypertensive heart disease and a control group of 26 healthy subjects (29 ± 9 years, 17 men). All underwent echocardiography and cardiac magnetic resonance (CMR) and the following indexes were analyzed: a) Echo: mitral flow velocities E/A ratio and deceleration of mitral E; E' velocity obtained from the medial and lateral mitral ring; E/E' ratio b) CMR: LV volumes indexed to BSA and ejection fraction (EF); global longitudinal strain (GLS, Circle cvI42); maximal size and distensibility of proximal ascending aorta. Distensibility was calculated from maximal (systolic) and minimum (diastolic) area of maximal proximal ascending aorta (cross-sectional images from SSFP sequence) using the formula=(maximum area-minimum area)/[minimum area x (systolic BP-diastolic BP)].

Results: 17 from 53 patients with MS had increased LV end-diastolic volume, 14 decreased EF (<50%) and 22 decreased GLS. Compared with the control group, patients with MS showed increased LV end-diastolic volume (85.0 ± 5.2vs 80.1 ± 5.3ml/m2, p = 0.04), lower GLS (17.1 ± 1.9 vs 18.9 ± 2.2, p = 0.01) lower LVEF (53.0 ± 7.1% vs 56.0 ± 5.4%, p = 0.03) and higher E/E' value (9.2 ± 1.3 vs. 6.2 ± 0.8, p = 0.01) as well as larger dimensions of proximal aorta (39 ± 2.2cm vs 32 ± 0.8,p = 0.010) and lower aortic distensibility (3.4 ± 1.9 vs. 4.4 ± 1.7 dynes cm-1, p = 0.03). In the group of patients, a correlation was found between the aortic distensibility and the LVEF (r = 0.47, p = 0.045) and GLS (r = 0.65, p = 0.02), but not with the other variables. Aortic distensibility was an independent predictor of LV dysfunction using a cutoff of 50% for LVEF and 19% for GLS.

Conclusion: In our population of patients with MS, we found changes in the LV function, which were related with aortic distensibility, possibly sharing a common pathway. The prognostic significance of these findings is under evaluation.