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Proximal aorta longitudinal but not circumferential strain predicts aortic events and aortic root dilation rate in marfan syndrome patients

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Background: The most common cardiovascular complications in Marfan syndrome (MFS) are aortic root dilation and type A aortic dissections. Elective aortic root surgery is indicated when maximum aortic diameter is larger than a defined threshold or in the case of fast-progressing dilation. However, maximum aortic diameter is limited for the prediction of aortic events. Indeed, a large international registry of acute aortic syndromes reported that as much as 40% of aortic dissections happen with maximum aortic diameter lower than 50 mm. Consequently, there is a need for new, non-invasive biomarkers to improve the prediction of aortic complications.

Purpose: The aim of the present study was to assess if proximal aorta circumferential and longitudinal strain and ascending aorta distensibility were associated with progressive aortic dilation and incidence of aortic events in Marfan syndrome patients.

Methods: Eighty seven Marfan syndrome patients free from previous cardiac/aortic surgery or dissection, were prospectively included in a multicenter follow-up. Patients were diagnosed by original Ghent criteria. Proximal aorta longitudinal and circumferential strain and distensibility were computed from baseline cine CMR images by means of feature-tracking. The

predictive capacity of each stiffness biomarkers was separately tested with multivariable linear regression analysis (aortic growth) and with Cox logistic regression analysis (aortic events), both corrected for clinical and demographic variables, including baseline maximum aortic diameter.

Results: During a follow-up of 81.6±17 months, mean diameter growth-rate was 0.65±0.67 mm/year and z-score growth rate was 0.07±0.13 / year. Elective aortic root replacement was performed in 11 patients while two patients presented type A aortic dissection. Baseline proximal aorta longitudinal strain was independently related to diameter growth-rate (p=0.001), z-score growth-rate (p=0.018) and aortic events (p=0.018). Conversely, neither circumferential strain nor distensibility were independent predictors of diameter growth-rate (p=0.385 and p=0.381, respectively), z-score growth-rate (p=0.515 and p=0.484, respectively) and aortic events (p=0.064 and p=0.205, respectively).

Conclusions: Proximal aorta longitudinal strain predicts aortic root dilation and major aortic events in Marfan syndrome patients beyond aortic root diameter and clinical and demographic characteristics.