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Prognostic value of proximal aorta longitudinal strain for aortic events and dilation in Marfan syndrome patients

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BACKGROUND

Aortic root dilation and type A aortic dissection are the most common cardiovascular complications of Marfan syndrome (MFS) patients. In clinical practice, MFS patients are closely followed up for aortic root dilation. Current guidelines support the use of aortic diameter to indicate preventive aortic root surgery, which is indicated if diameter reaches a threshold or in case of fast-progressing dilation. However, the predictive capacity of diameter is limited, given that a large part of acute aortic events happen in aorta with a diameter lower than the suggested threshold. New non-invasive biomarkers to improve risk stratification are thus needed.

PURPOSE

We aimed to evaluate whether proximal aorta circumferential and longitudinal strain and ascending aorta distensibility predict aortic root diameter growth and incidence of aortic events in Marfan patients.

METHODS

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

RESULTS

During a follow-up of 81.6 ± 17 months, mean diameter and z-score growth-rates were 0.65 ± 0.67 mm/year and 0.07 ± 0.13 1/year, respectively. Two patients presented type A aortic dissection while 11 required elective aortic root replacement. Proximal aorta longitudinal strain but not circumferential strain and distensibility were independent predictors of diameter growth-rate ($p = 0.001$, $p = 0.385$ and $p = 0.381$, respectively), z-score growth-rate ($p = 0.018$, $p = 0.515$ and $p = 0.484$, respectively) and aortic events ($p = 0.018$, $p = 0.064$ and $p = 0.205$, respectively), in multivariable analyses corrected for demographic and clinical characteristics, including baseline aortic root diameter.

CONCLUSIONS

In Marfan syndrome patients, proximal aorta longitudinal strain independently predicts aortic root dilation and major aortic events beyond aortic root diameter and clinical risk factors