Abstract: 1202

Longitudinal but not circumferential proximal aorta strain predicts aortic root dilation rate and major cardiovascular events in Marfan syndrome

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BACKGROUND:
Marfan syndrome (MFS) is a hereditary connective tissue disorder caused by mutation in the FBN1 gene. Typical cardiovascular complications of MFS is dilation of the aortic root, which has been related to an increase in the risk of dissection. Current guidelines suggest a close follow-up of aortic diameter, and preventive aortic root replacement when diameter reaches 50 mm or 45 mm in the presence of fast diameter progression. However, aortic diameter has some limitations and other imaging biomarkers are needed to improve risk stratification of rapid progressive dilation and complications.

PURPOSE:
The aim of the present study was to investigate the capacity of proximal aorta longitudinal strain, circumferential strain and distensibility to predict aortic root dilation rate and occurrence of major cardiovascular events in Marfan patients.

METHODS:
Eighty seven Marfan patients without previous aortic dissection, cardiac/aortic surgery and moderate/severe aortic regurgitation were prospectively included in a multicenter protocol of clinical and imaging follow-up. At baseline, CMR was performed and proximal aorta longitudinal and circumferential strain and distensibility were computed.

RESULTS:
During a mean follow-up duration was 81.6 ± 17 months, mean dilation rate was 0.65 ± 0.67 mm/year, z-score growth rate was 0.07 ± 0.13 1/year, 11 patients underwent elective aortic root replacement, and 2 experienced type A aortic dissection. Once corrected for clinical and demographic characteristics and aortic root diameter, proximal aorta longitudinal strain but not AAo 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 major cardiovascular events (p=0.018, p=0.064 and p=0.205, respectively).

CONCLUSIONS:
Longitudinal strain of the proximal aorta is an independent predictor of aortic root dilation rate and major cardiovascular events in Marfan syndrome patients beyond aortic root diameter, clinical risk factors and demographic characteristics.
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