

POSTER PRESENTATION

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Aortic arch stiffness in Fabry disease

Zoubir M Bensalah^{1*}, Cédric Collin³, Alban Redheuil², Pierre Boutouyrie³, Dominique Germain⁴, Elie Mousseaux²

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Background

Background: Aortic thoracic remodelling has been recently described in FD, however no data was available concerning AAS in this rare genetic disease.

Aim of this study was to assess aortic arch stiffness (AAS) parameters in male patients with Fabry disease (FD) using cardiovascular magnetic resonance imaging (CMR).

Methods

Twenty nine males with FD matched with 58 controls for age underwent CMR using cine and phase contrast velocity sequences.

Thoracic aortic diameter; local (distensibility, β -index stiffness), global (pulse wave velocity) stiffness parameters of the aortic arch and cardiac proprieties were assessed by CMR.

Results

Aortic arch PWV was significantly increased in FD patients (6.5 ± 3.1 vs 5.0 ± 1.5 m/s, $p < 0.01$)

Compared to control subjects, patients with FD had also markedly decreased distensibility (2.73 ± 1.14 vs 3.45 ± 1.13 10^{-2} kPa $^{-1}$, $p < 0.01$) and increased stiffness index beta ($9.4 \pm 6.7 \cdot 10^{-2}$ vs $5.9 \pm 2.7 \cdot 10^{-2}$, $p < 0.001$) in the ascending aorta.

Descending aortic stiffness parameters were also impaired with a trend for decreased distensibility (2.26 ± 1.15 vs 3.15 ± 1.0 10^{-2} kPa $^{-1}$, $P = 0.06$) and significant increased for β -index stiffness ($8.5 \pm 3.9 \cdot 10^{-2}$ vs $2.9 \pm 0.9 \cdot 10^{-2}$, $p < 0.0001$).

Conclusions

FD patients exhibited impairment of both local and global aortic arch stiffness parameters.

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Author details

¹Radiology, HOSPITAL AMBROISE PARE, Boulogne-Billancourt, France.

²Cardiovascular Radiology, Hospital George Pompidou, Paris, France.

³Department of Pharmacology, Hospital George Pompidou, Paris, France.

⁴Genetic, Hospital Raymond Poincaré, Garches, France.

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¹Radiology, HOSPITAL AMBROISE PARE, Boulogne-Billancourt, France
Full list of author information is available at the end of the article