Aortic root growth in children with Marfan syndrome: evidence for gender differences

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Background

- The most known and life threatening cardiovascular complication of MFS is aortic dilation and subsequent dissection.
- Adult male MFS patients have more aortic events than females.¹
- Scarce data on gender differences in the paediatric MFS population are available.

Known risk factors of AoR dissection in MFS

- Family history of AoR dissection
- AoR diameter >50mm
- Rapid AoR growth (>2mm/y)

Aim

1. Does aortic root growth differ between genders during childhood and adolescence?
2. Are there any factors which can identify children at risk for rapid aortic growth or aortic surgery?

Results

In our cohort of children with MFS, 8 underwent elective AoR replacement.

Mean age: 15 (11.3)
Height: 157.2 (14.9)
Weight: 67.8 (11.7)
BSA: 1.53 (0.13)
AoR z-score: -0.84 (1.3)

Only males, no females

Conclusion 1.

Aortic root growth is higher in males than in females with MFS. Differences in BSA seems responsible for this differential growth until 15yr.

2. In our cohort of children with MFS, 8 underwent elective AoR replacement, all of whom were males.

3. Males undergoing AoR replacement have higher AoR z-score at baseline and higher AoR growth rate during FU.

4. Z-score>4.19 at baseline is a good predictor of AoR replacement during childhood. This group might benefit from a more aggressive medical treatment.

Materials and Methods

- Inclusion: Our cohort of MFS patients ≤ 25yr with FBN1 mutation
- Retrospective analysis
- Collection of clinical data
- Review of yearly echocardiography

Abbreviations:
MFS: Marfan syndrome; MVP: mitral valve prolapse; AoR: aortic root replacement; Pos: positive, FN: false negative, Sensitivity: 100%, Specificity: 83.9%

References:
¹ Groth KA et al. Clin Res Cardiol 2017 Feb; 106 (2) 105-112.
² Campens LA et al. Am J Cardiol 2014 Sep; 114 (6) 914-920.

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