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?

Conclusions

1. AoR growth rate 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.

Results

Table 1: Females were older and had a higher BSA at baseline. AoR diameter, however, did not differ between both groups

	Male (n=29)		Female (n=15)		p-value
Age baseline (yr)	7.59 (4.27)		10.60 (3.38)		**0.022**
Current age (yr)	16.64 (5.95)		16.93 (4.10)		0.867
Mean follow-up (yr)	8.25 (3.74)		6.61 (3.75)		0.202
Height (cm)	139.53 (31.40)		166.38 (19.35)		**0.010**
Weight (cm)	32.32 (18.07)		45.23 (13.75)		**0.037**
BSA (m^2)	1.08 (0.41)		1.36 (0.39)		**0.031**
Fullfill Ghent-2 N(%)	22 (75.9)		12 (80)		0.693
AoR diameter (mm)	29.62 (5.98)		29.73 (3.15)		0.935
AoR z-score	3.62 (1.88)		2.83 (0.96)		0.070
ST-junction (mm)	23.22 (5.28)		23.31 (2.39)		0.944
Aortic regurgitation N(%)	No Trace/mild	27 (93.1) 2 (6.9)	No Trace/mild	15 (100) 0	0.533
MVP (all forms) N(%)	No Yes	15 (51.7) 14 (48.3)	No Yes	4 (26.7) 12 (73.3)	0.167
Mitral regurgitation N(%)	No Trace/mild Moderate	21 (72.4) 7 (24.1) 1 (3.4)	No Trace/mild Moderate	8 (53.4) 6 (34) 1 (6.7)	0.194

Figure 1: AoR growth rate was higher in males than in females with MFS

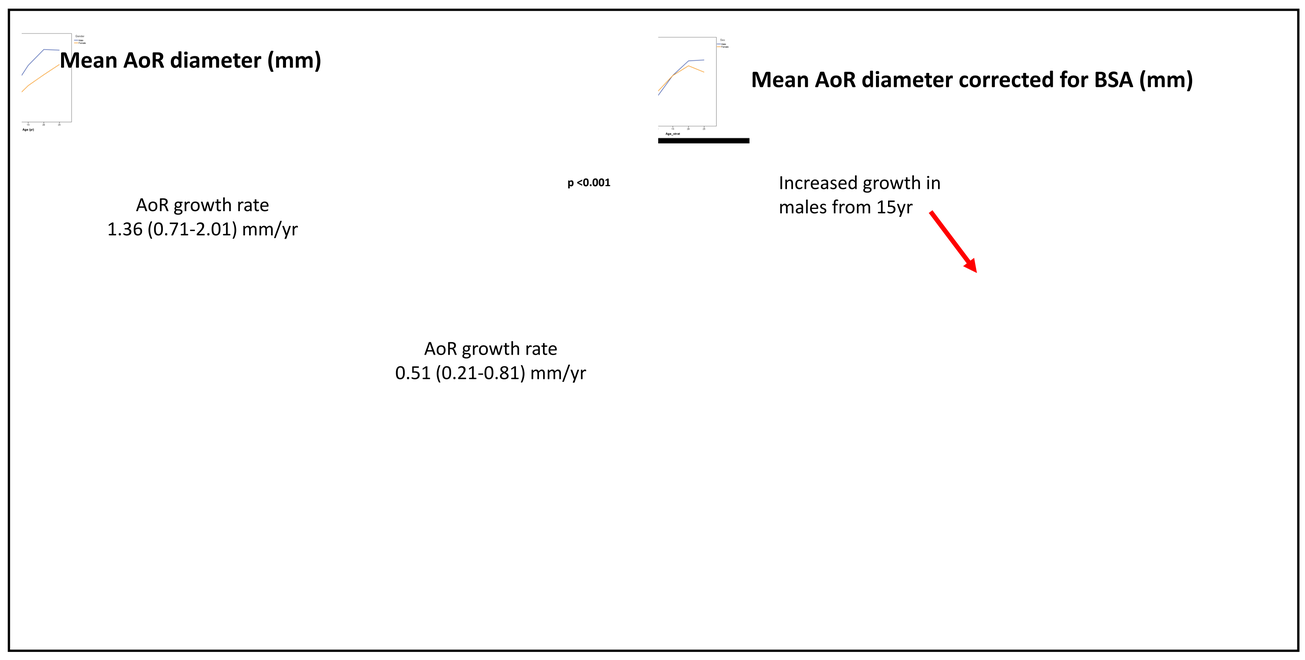


Figure 2: A z-score at baseline of 4,19 or more can predict AoR replacement during childhood accurately

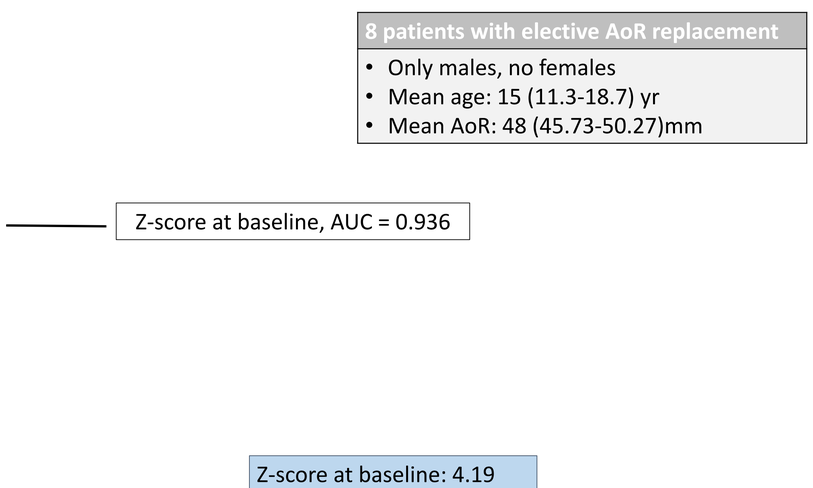


Table 2: only aortic root diameter and z-score at baseline differ significantly between the AoR replacement group and the other groups

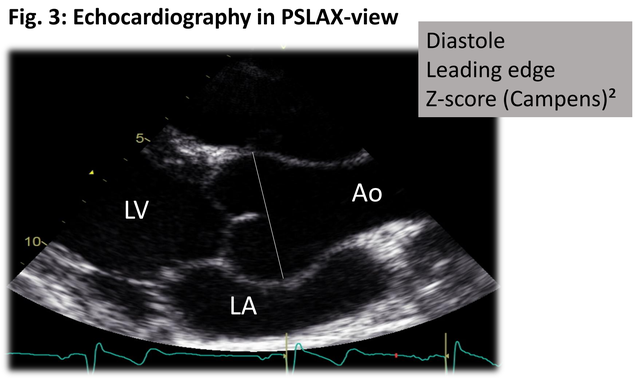
	AoRR (N=8)	Non-operated males (N=20)	Females (N=15)	p-value
Height at baseline (cm)	130.75 (11.98)	143.36 (35.59)	166.38 (19.35)	0.116
Weight at baseline (kg)	23.50 (4.36)	35.57 (20.10)	45.23 (13.75)	0.152
Fullfill Ghent-2 N (%)	8 (100)	14 (65)	12 (80)	0.081
Pos FH N (%)	3 (37.5)	13 (65)	7 (46.7)	0.380
AoR at baseline (mm)	33.88 (3.44)	28.15 (6.11)	29.73 (3.15)	0.026
AoR z-score at baseline	5.52 (1.48)	2.89 (1.51)	2.83 (0.96)	<0.001*
ST-junction at baseline (mm)	27 (25.3-29)	22.25 (19.25-23.75)	23.31 (2.39)	0.104
AoR growth rate (mm/yr)	2.02 (0.53)	1.09 (0.49)	0.55 (0.32)	<0.001^

Post-Hoc analysis (Bonferroni):
*Statistical difference between AoRR and both non-operated males and females (p<0,001). No difference between non-operated males and females
^Statistical difference between the 3 groups (p<0,001)

Materials and Methods

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

Fig. 3: Echocardiography in PSLAX-view



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

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Limitations

- Retrospective character
- Small group of patients
- Variability in the studied group in terms of age, time of follow up and treatment

Abbreviations:
MFS: Marfan syndrome, MVP: mitral valve prolapse, AUC: area under the curve, AoRR: aortic root replacement, Pos: positive, FH: family history, FBN1: fibrillin 1

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