

MP3-10

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

Muiño-Mosquera L., De Groot K., Vandekerckhove K., Panzer J., De Wilde H., De Wolf D., De Backer J.

Ghent University Hospital, Ghent, Belgium

Background:

Aortic root (AoR) dilation and subsequent dissection are the most life threatening complications of Marfan syndrome (MFS). Known determinants of aortic dissection include the diameter and growth rate. Knowledge of risk factors favouring rapid AoR growth is important to tailor treatment.

Data regarding gender differences in children with MFS are scarce. Our aim was to study whether AoR growth in this group differs depending on the gender.

Methods:

We retrospectively reviewed the echocardiography and clinical data of our cohort of MFS patients younger than 25yrs. MFS was diagnosed after familial screening in 50% or due to clinical features of MFS in the rest. All patients had an underlying FBN1 gene mutation. First digital echocardiography available was considered as baseline. Last echocardiography at follow-up was used to calculate AoR growth rate.

Results:

Forty MFS patients were included in this study. Twenty-seven (69,2%) were males. Mean age and mean AoR diameter at baseline were $8,41 \pm 3,9$ yrs and $29,5 \pm 4,82$ mm (z-score $3,29 \pm 1,95$) respectively.

During a mean follow up time of $4,63 \pm 2,95$ yrs no aortic dissection occurred. Eight patients (20%) underwent elective AoR replacement at a mean age of $15 \pm 3,7$ yrs. All these patients were males. One patient died of arrhythmia a few months after elective AoR replacement during orthopaedic surgery. Mean AoR growth in this period was $1,1 \pm 0,68$ mm/yr. There was a significant difference between males and females ($1,36 \pm 0,65$ versus $0,51 \pm 0,30$ mm/yr; $p < 0,001$) and between operated males and non-operated males ($2,02 \pm 0,53$ vs $1,09 \pm 0,49$ mm/yr; $p < 0,001$). Operated males also had significant higher AoR z-score at baseline in comparison to non-operated males ($5,35 \pm 1,43$ vs $2,68 \pm 1,47$; $p < 0,001$).

Conclusion:

In this study males with MFS had higher AoR growth rate and higher incidence of elective AoR replacement than females with MFS. This supports the argument that there is a gender difference. The group of males undergoing elective AoR replacement had significantly higher AoR z-score at baseline and more rapid AoR growth during follow-up. These results suggest that there is a group of MFS patients showing rapid AoR growth from a very young age. Further study is necessary to elucidate why this happens.