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Marfan syndrome (MS): other cardiovascular manifestations rather than aortic root dilatation.

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Introduction:
Although aortic root (AoR) dilatation and mitral valve prolapse (MVP) are the most frequent cardiac manifestations of Marfan syndrome (MS), these patients can also develop ventricular arrhythmias and dilated cardiomyopathy. Our aim is to describe the incidence of these findings in our population and see if there is any correlation among them.

Materials:
Retrospective study of 57 Marfan paediatric patients (under 18) followed up in our ICC clinic from 2000-2018.

Results:
Median age of 10 (IQR 8,15); 56% females. 86% with FBN1 mutation (24,6% de novo). 75,4% on medical treatment (29,8% on losartan, 29,8% on BB and 15,8% on both). 17,5% symptomatic, 4 of them had arrhythmias: 2 with supraventricular tachycardia and another 2 with non-sustained ventricular tachycardia (NSVT). No late gadolinium enhancement was found in any of MRIs performed. There were no sudden deaths.

Echocardiographic findings: 56,1% had AoR dilatation [median 31mm (IQR 27,36)]; 73,7% had mitral regurgitation (MR) (17,5% significant degree) and 52,6% had evidence of MVP. 19,3% of the patients showed LV dilatation, all with normal systolic and diastolic function [median LVEF on MM 64,5% (IQR 61,70%), median S wave on TDI 10cm/seg (IQR 8.3,10)]. There was a significant correlation between the presence of MVP and LV dilatation (p=0.005) but LV dilatation seemed not to be related to the presence of a significant MR (p=0.25). None had significant aortic regurgitation.

Electrocardiographic data: 21% of the patients had baseline repolarization abnormalities mainly in form of inverted/flattened T waves in inferior/lateral leads, which was correlated to the presence of LV dilatation (p=0.03) and MVP (p=0.03). 59,6% had other ECG abnormalities mainly mild intraventricular conduction delay and LVH (15,8%). The presence of arrhythmias didn´t show significant correlation to LV dilatation (p=0.16), MVP (p=0.61) nor repolarization abnormalities (p=0.33).

Conclusion:
MS can be associated to many other cardiac comorbidities rather than AoR dilatation. The only factor associated to LV dilatation in our series was the presence of MVP. Patients with a dilated LV didn't show ventricular dysfunction. Although no cases of sudden arrhythmic death, 2 patients had runs of NSVT. Ventricular arrhythmias did not correlate with MVP nor LV dilatation.