Detection of cardiomyopathy in children with Marfan Syndrome with 2D strain echocardiography

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Introduction: Studies have found cardiomyopathy in adult patients with marfan syndrome (MS) with systolic dysfunction. Sophisticated techniques such as 2D strain echocardiography or "speckle tracking imaging" (STI) have been reported to detect early cardiac dysfunction and to be more accurately. The aim of our study was to evaluate the validity of STI for the study of myocardial function in children with MS, and to assess the interest for cardiomyopathy detection in this population.

Methods and Results: Echocardiographic parameters of left ventricular systolic function were compared between two populations in standard 2D, STI and cardiac MRI. We included 39 MS (clinically confirmed from the 2010 modified criteria of Ghent or genetically with mutations on the FBN1 gene) and 41 healthy children, aged 4 to 18 years old respectively on average 12.07± 4.81 and 11.82 ± 3.64. They were paired on the body surface.The values of global strain longitudinal peak (SLG) of MS were compared with values of ejection fraction of the left ventricle (LVEF) assessed by cardiac MRI (gold standard). Relations between the two types of mutations (PTC and inframe) in MS and altered strain were studied. Impaired SLG was significant for 7 segments : basal inferoseptal (p = 0.0001), mid anterolateral (p = 0.0083), basal anterolateral (p <0.0001), basal inferior (p = 0.0057), apical lateral (p = 0.0084), basal anterior (p <0.0001) and basal inferolateral (p = 0.0103). This segmental impairment predominated the basal level with apex-to-base gradient. Patients with more altered SLG appeared to have a greater dilatation of the ascending aorta (p = 0.03).

Conclusion: This study is the largest pediatric cohort who compare 2D strain echocardiography and MRI in children with MS. The interest for primary or secondary cardiomyopathy detection remains to be validated and consolidated with larger studies and could justify not only to focus on the aorta.