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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) whereas conventional echocardiographic parameters showed normal systolic function. The aim of our study was to evaluate the validity of the 2D strain echocardiography or "speckle tracking imaging" (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 the two populations (39 MS and 41 healthy children) in standard 2D, STI and cardiac MRI. 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) and altered strain were studied. Impaired SLG was significant for 7 segments. 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.

Conclusion:
This is the largest pediatric cohort who compare STI and MRI in children with SM.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.