

Hemodynamic forces and myocardial deformation using cine MRI in Marfan syndrome

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Introduction. Cardiovascular assessment of Marfan syndrome (MS) patients has normally focused on the aortic root and vascular manifestations due to the high risk of aortic dissection.

Although primary myocardial impairment has long been suspected, the evidence has been controversial. Advanced in CMR may support the early detection of cardiac dysfunction. Beyond left ventricle ejection fraction (EF) and myocardial strain (S), a new parameter is emerging, the hemodynamic forces (HF) exchanged between the blood flowing in the heart and the myocardium. The application of these techniques to MS could be useful in demonstrating the presence of primary myocardial impairment.

Aim. The aim of this study is to explore myocardial function in MS through the evaluation in cine CMR of EF, S and cardiac HF exchanged between the blood and the myocardium and compare these data with those of a control group (C).

Methods. We retrospectively analysed CMR cine images of MS (diagnosed according revised Ghent criteria) without valvular disease or previous cardio surgery, and C, in standard long-axis projections, to define endocardial borders for subsequent quantification of left ventricular volumes, EF, longitudinal, circumferential and radial S, apex-to-base and lateral-to-septum HF (expressed in mN and as a percentage of gravity acceleration). The analysis were performed on Medical Imaging Systems (QStrain version 1.3.0.79; MEDIS) (Figure 1).

Results. 108 MS and 44 C had a good quality study, suitable for MEDIS analysis. The mean age was 33 ± 13 ys in MS, 35 ± 12 ys in C; 39% were male in MS, 50% in C.

The results of left ventricular function were: EF $63 \pm 7\%$ in MS vs $66 \pm 5\%$ in C group, $p = .008$, global longitudinal S $-24.5 \pm 4.1\%$ in MS vs $-26.2 \pm 4.1\%$ in C, $p = .014$; global circumferential S $-30.6 \pm 6.3\%$ in MS vs $-33.8 \pm 4.4\%$ in C, $p = .002$; radial S $64.5 \pm 16.2\%$ in MS vs $72.7 \pm 15.9\%$ in C, $p = .005$; apex-to-base HF $13.2 \pm 4.7\%$ in MS vs $17.8 \pm 7.6\%$ in C, $p = .000$; lateral-to-septum $2.6 \pm 1.3\%$ in MS vs $3.1 \pm 1.4\%$ in C, $p = .048$.

Moreover, 4.6% MS patients had mid reduced EF (40-50%); 9.2% had global longitudinal S reduction (cut off -19.3%); 7.4% had global circumferential S reduction (cut-off -21.7%).

Conclusion. These data provide support for the existence of a cardiomyopathy in MS. In our opinion, the term "primary cardiomyopathy" is not appropriate to describe this condition: patients with MS have changes in aortic stiffness and probably in cardiac afterload. The HF data are the most interesting of this study, both in the validation of this new parameter and in early detection a cardiomyopathy in MS

Moreover, the reduction of global circumferential S, as well as global longitudinal S, in MS patients may help provide new elements to characterize the MS cardiomyopathy: sure enough, in literature, circumferential strain abnormalities are related to afterload increase. HF analysis is really a new challenge of cardiac imaging, as sensitive markers of subtle systolic dysfunction.

Abstract Figure. Figure 1. Analysis exemple.

