

DOES GLOBAL LONGITUDINAL STRAIN PREDICT EARLY SYSTOLIC DYSFUNCTION IN PEDIATRIC PATIENTS WITH MARFAN SYNDROME?

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INTRODUCTION

Marfan syndrome (MFS) is an inherited connective tissue disorder caused by mutations in FBN1 gene, which encodes for the extracellular matrix protein Kbrillin-1. The clinical spectrum of MFS includes cardiovascular, ocular and skeletal involvement and diagnosis often requires both the identification of clinical signs and molecular analysis of FBN1 gene, according to Ghent revised criteria. Heart and large-vessel involvement is the major cause of death in MFS: aortic root aneurysms and aortic dissection represent the most severe and life-threatening clinical events in MFS, nevertheless mitral and tricuspid valve prolapse, late-occurrence aortic valve dysfunction and dilated cardiomyopathy have been described. Although an increased prevalence of left ventricular systolic dysfunction (LVSD) has been reported in MFS, predictive factors of LVSD in patients without evidence of valvular heart disease have not been addressed in MFS.

METHODS

The aim of this study is to assess whether myocardial deformation indexes, including global longitudinal strain (GLS), circumferential and radial strain are impaired earlier and at greater extent in patients with MFS, compared with age-adjusted control population. From an initial screening population of 37 patients with genetically confirmed MFS, we enrolled 20 patients (aged 4-16) with clinically and genetically-confirmed MFS, according to revised Ghent criteria and 20 healthy controls. Patients older than 16, presence of moderate/severe valvular heart disease and/or other causes of left ventricular dysfunction were considered exclusion criteria for the population of study. A complete echocardiographic and clinical assessment was performed, Wilcoxon-Mann-Whitney 2-sample rank sum and chi-square test were used when appropriated.

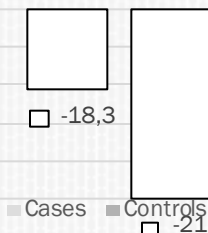
RESULTS

Results: Compared with controls, patients with MFS showed a significant reduction in global longitudinal strain (GLS $-18,3$ vs -21 , $p<0.01$) and strain rate $0.99\% \pm 0.19\%$ vs $1.07\% \pm 0.18\%$, $p=0.03$) whereas no significant differences were found in left ventricular ejection fraction (EF $64 \pm 4\%$ vs $67\% \pm 3\%$, $p=0.07$) radial (mean $43,07$ vs $47,88$ $p=0.3$) and circumferential ($-21,06$ vs $-22,08\%$, $p=0,8$) strain. Among adult patients with MFS the prognostic role of GLS as early indicator of LVSD has been clearly demonstrated.

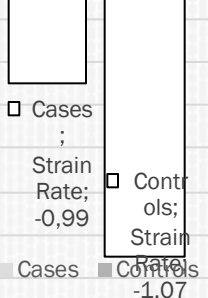
CONCLUSIONS

Although larger pre-specified studies are needed, this study shows that pediatric patients with MFS have lower GLS compared with controls, therefore it is possible that longitudinal strain could represent an early index of LVSD in this clinical setting.

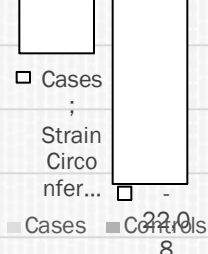
Longitudinal Strain



Strain Rate



Circumferential Strain



Radial Strain

