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503 SPECKLE-TRACKING ECHOCARDIOGRAPHY FOR ARRHYTHMIC RISK ASSESSMENT IN HYPERTROPHIC AND FABRY CARDIOMYOPATHY

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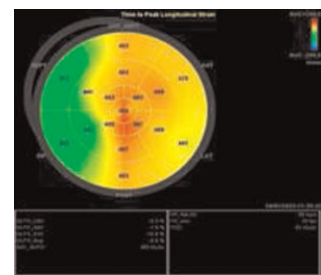
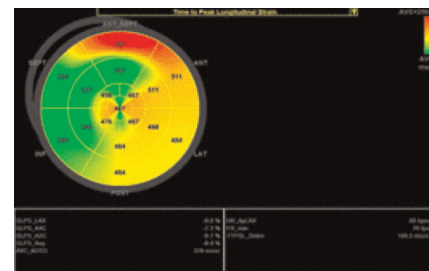
Background: Hypertrophic Cardiomyopathy (HCM) is burdened by sudden cardiac death (SCD) risk of 0.9%/year, and is the most common cause of SCD in young adults. It is an autosomal dominant inherited disease caused by mutations in cardiac sarcomere genes, but the hypertrophic phenotype can also be an expression of cardiac involvement in multiorgan metabolic storage diseases, such as Anderson-Fabry disease (AFD). Mechanical Dispersion (MD) by Speckle-Tracking Echocardiography (STE) has recently emerged as an additional arrhythmic risk marker.

Purpose: Aim of the study was to evaluate LV systolic and diastolic function, global longitudinal strain (GLS) and MD by STE and analyze their association with ventricular arrhythmias in patients with HCM and AFD.

Methods: We included in our analysis 36 patients with HCM, 54 with AFD, of which 10 with left ventricular hypertrophy (AFD-LVH) and 44 without (AFD-N), and 27 healthy subjects. We performed a comprehensive basic echocardiographic study and analyzed GLS and MD (post-processing through EchoPAC 2.02). We also evaluated ventricular arrhythmias (V-AR), including ventricular fibrillation and sustained and non-sustained ventricular tachycardia, by Holter ECG, and the data obtained by cardiac magnetic resonance (CMR) in hypertrophic patients. Data were analyzed by unpaired Student t-test or chi-square/Fisher's exact test as appropriate, and binary logistic regression (SPSS Statistics ver.26).

Results: Diastolic function was impaired in HCM and AFD-LVH patients compared to control and AFD without LVH. GLS was significantly lower in the V-AR group compared to patients without V-AR (9.7 ± 2.9 vs 14.1 ± 4 , $P=0.007$), MD was significantly higher in the V-AR group (111 ± 47 vs 68.1 ± 16 , $P=0.03$). We found a significant association between ventricular arrhythmias and GLS ($P=0.005$) and between ventricular arrhythmias and MD ($P<0.001$). We found also a significant association of late gadolinium enhancement at CMR with GLS ($P=0.005$) and MD ($P=0.03$).

Conclusions: GLS and MD are useful additional indices in the evaluation of patients with HCM or AFD, also in presence of preserved LV ejection fraction, and promising prognostic predictors to identify patients at high risk for ventricular arrhythmias



Figures: MD in an HCM patient (on the left) and in an AFD-LVH patient (on the right).