Abstract: P903

Is left ventricular longitudinal strain a good pronostic factor in friedreich ataxia?

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Introduction: Friedreich ataxia (FRDA) is a rare genetic sensory ataxia. The causal mutation is an expanded trinucleotide repeat (GAA) in the frataxin gene. Hypertrophic cardiomyopathy is associated with FRDA and is the major cause of early death before 40 years old. Patients with progressive decline of the left ventricular ejection fraction (LVEF) have the worse prognosis. Speckle tracking echography with 2D longitudinal myocardial strain (GLS) is recognized as a more effective technique than conventional LVEF in detecting subtle changes in LV function.

Purpose: Evaluate the prognostic value of global longitudinal strain (GLS) in patients with FRDA as compared to LVEF.

Methods: From 2003 to 2017 consecutive patients with genetically confirmed FRDA were included. Longitudinal strain analysis was retrospectively performed with Tomtec software. News were obtained until April 2018, no patient was lost during follow-up.

Results: The study included 156 patients (51% male) of 35±12 years (mean ± SD) with an age at disease onset of 17±11 years, age at wheelchair use of 26±10 years, and GAA repeat on the shorter allele of 590±241 pb. The following echocardiographic parameters were studied at baseline: LVEF 64±9%, GLS −19.8±5% (n=141), septal wall thickness (SWT) 11.4±2.5 mm, posterior wall thickness (PWT) 10.4±1.8 mm, LV end diastolic diameter (LVEDD) 44.4±6mm. Correlation between GLS and LVEF was 0.31 (p=0.0002).

After a mean follow-up of 7.7±4.0 years, 17 (11%) patients died and the outcome (cardiac arrhythmia, heart failure, stroke or death) concerned 28 (18%) patients. In univariate analysis (Cox model), factors associated with mortality were: GLS (HR: 1.2; 95% CI 1.10–1.32, p=0.0001), LVEF (HR: 0.88; 95% CI 0.85–0.92, p<0.0001), GAA (HR: 1.28; 95% CI 1.11–1.47, p=0.0008), age at onset (HR: 0.84; 95% CI 0.76–0.94, p=0.002), LVMi (HR: 1.02; 95% CI 1.01–1.04, p=0.0078), SWT (HR: 1.18 95% CI 1.01–1.36, p=0.03) and LVEDD (HR: 1.09; 95% CI 1.00–1.19, p=0.04). In multivariate analysis LVEF was the only independent predictor of long-term mortality (HR: 0.93; 95% CI 0.88; 0.99, p=0.02). Similarly GLS was not an independent predictor of the composite outcome in multivariate analysis.

Conclusion: GLS is a predictor of morbimortality but is not superior to LVEF in FRDA patients. Further prospective studies are mandatory to assess the early predictive value of 2D GLS.