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


Friedreich Ataxia (FRDA) is due to mitochondrial dysfunction caused by abnormal repetition of GAA expansion:
• First genetic cerebellar ataxia: 1/50000 patients
• Autosomal recessive transmission
• Moderate cardiac hypertrophy: frequent abnormal ECG (80%)
• Cardiopathy is the main cause of death in FRDA before 40 years old (heart failure)
• Cardiological evolution is slow with a progressive decrease in Left Ventricular Ejection Fraction (LVEF)
• Global Longitudinal strain is an effective technique to detect subtiles changes in LV function

OBJECTIVES: To evaluate the prognostic value of global longitudinal strain (GLS) in patient with FRDA as compared to LVEF.

RESULTS (1): Clinical biological and echocardiographic characteristics of the population

a) Clinical data
Male: 51%, mean age = 35,1 ± 12y
Age at diagnostic: 17,2 ± 10,6y
Wheelchair patients: 72% aged 26 ± 10 y
Symptoms (20%): 50% dyspnea, 26% palpitations chest pain 24%

b) ECG data
Abnormal ECG: n=136 (87%)
Negatives T wave :n=126 (80%)

RESULTS (2): EVENTS

a) EVENTS, Follow up : 7.7 ± 4.0 y

b) Factors Associated with All-cause Mortality

GLS (%)
Age (years)
Age at diagnostic (years)
GAA on the shorter allele
LVMI (g/m²)
LVEF (%)

Correlation coefficient: GLS and LVEF: r = 0.31 (0.15 ; 0.45), p=0.0002

CONCLUSION
➢ GLS is a predictor of morbimotality in FRDA
➢ GLS is not superior to LVEF as a prognostic marker
➢ Proposed cut off value of GLS to diagnose FRDA cardiopathy: -15.4%
➢ Prospective studies are mandatory to assess the early predictive value of GLS in FRDA

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