Prognostic role of the progression of late gadolinium enhancement in hypertrophic cardiomyopathy

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Background: Late Gadolinium Enhancement (LGE) extent $\geq 15\%$ of left ventricular (LV) mass is a widely accepted prognostic risk factor in hypertrophic cardiomyopathy (HCM). However, LGE extent increases over time and the clinical role of the progression of LGE (LGE-rate) was not prospectively evaluated in previous studies.

Purpose: We evaluated the clinical and prognostic role of LGE-rate in HCM.

Methods: 105 patients with HCM underwent cardiac-MR at baseline (CMR-I) and after $\geq 2$ years of follow-up (CMR-II). LGE extent was measured in both the examinations (LGE was defined using threshold validated method of $\geq 6$ SD exceeding the mean of normal myocardium (1)).

LGE-rate was defined as the ratio between the increase of LGE extent (g) and the time interval (months) between CMR-I and CMR-II. A combined endpoint of sudden cardiac death (SCD), resuscitated cardiac arrest, appropriate ICD intervention and sustained ventricular tachycardia was used (hard events).

Results: The percentage of patients with LGE extent $\geq 15\%$ increased from 9$\%$ to 20$\%$ at CMR-II (p=0.03), compared to CMR-I. During a median follow-up of 52 months, 25 hard events were recorded. The presence of LGE $\geq 15\%$ at CMR-II allowed a significant reclassification of the risk of patients than at LGE $\geq 15\%$ at CMR-I (NRI 0.21, p=0.046). At maximally selected rank statistical analysis the optimal prognostic cut-point for LGE-rate was $>0.07$ g/months. At Kaplan-Meier curve analysis patients with LGE-rate $>0.07$ had significant worse prognosis than those without (p<0.0001) (figure 1). LGE-rate $>0.07$ allowed a significant reclassification of the risk compared to LGE $\geq 15\%$ at CMR-I and at CMR-II (NRI 0.49, p=0.003) (figure 2). At Cox regression multivariable analysis, LGE-rate $>0.07$ was the best independent predictor of hard events.

Conclusions: LGE-rate is heterogenous among patients with HCM and a follow-up CMR should be performed after 2 years to reclassify the risk of SCD of those patients. High LGE-rate should be considered as a novel prognostic risk factor in HCM.

Figure 1
Figure 2