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Prognostic impact of quantification of late gadolinium enhancement in cardiac magnetic resonance on mortality and morbidity in hypertrophic cardiomyopathy with systolic dysfunction

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Purpose: A subset of patients with hypertrophic cardiomyopathy (HCM) develop systolic dysfunction so-called end-stage phase. Reduced left ventricular (LV) ejection fraction (EF) reflects poor prognosis. Late gadolinium enhancement (LGE) in cardiac magnetic resonance (CMR) detecting myocardial fibrosis is associated with major adverse cardiac events (MACE) in general HCM population. Inverse relationship is present between LGE and LVEF in HCM, however, which of LGE or LVEF predicts mortality and morbidity more precisely in HCM with systolic dysfunction remains unclear.

Methods: We assessed the extent of LGE with a threshold of 6 SD expressed as a percentage of the total LV mass (%LGE) in 47 consecutive HCM patients with systolic dysfunction defined as LVEF <50% (average 34±13%) who underwent CMR (36 males, mean age 59±14 years) and followed them over 4.5±1.6 years. The primary composite end point was MACE such as all-cause death, lethal arrhythmia, LV assist device implantation, heart transplantation or stroke due to cardiac emboli. Two separate secondary end points were defined, heart failure (HF) end point including unplanned heart failure hospitalization (UPHFH) and HF death and arrhythmic end point including lethal arrhythmia and sudden cardiac death.

Results: LGE was detected in all patients with an extent of 30±15% of LV mass. During the follow up, 19 of 47 patients developed primary end point of MACE. On the other hand, 23 of 47 patients had secondary HF end point and 11 of 47 patients reached secondary arrhythmic end point. Multivariable analysis revealed %LGE as the only independent predictor of both primary and secondary HF end points. These risks increased as %LGE increased (hazard ratio [HR]: 2.89 /10%, 95% confidence interval [CI] = 1.39 to 7.82, p=0.003 and HR: 3.48 / 10%, 95% CI = 1.62 to 10.0, p<0.001, respectively). LVEF was inversely related to %LGE (r = -0.46; p<0.001) and was a predictor of both primary and secondary HF end points in univariate analysis, but not an independent predictor in multivariate analysis. The number of recurrent UPHFH per patient was related to not LVEF but %LGE (r=0.49; p<0.02). Neither %LGE nor EF was a predictor of secondary arrhythmic end point in univariate analysis.

Conclusion: %LGE is the only independent predictor of MACE and HF end point and reflects mortality and morbidity more precisely than LVEF in HCM with systolic dysfunction. Quantification of LGE is useful for predicting MACE and HF events and can contribute to further risk stratification in HCM with systolic dysfunction.