Background: Although hypertrophic cardiomyopathy (HCM) with preserved systolic function (PSF) defined as ejection fraction (EF) > 50% reveals favorable clinical course, HCM with systolic dysfunction (SD) defined as EF < 50% reflects poor prognosis. Presence of marked fibrosis in left ventricle (LV), detected by cardiac magnetic resonance (CMR) as late gadolinium enhancement (LGE), is a main factor responsible for SD and is associated with major adverse cardiac events (MACE). Both of LVEF and LGE have prognostic importance, however, which predicts prognosis more precisely in HCM with PSF or SD remains unresolved.

Methods: We assessed baseline clinical and CMR parameters including LVEF and the extent of LGE with a threshold of 6 standard deviation expressed as a percentage of the total LV mass (%LGE) in 106 consecutive HCM patients (69 males, mean age 56 ± 18 years) including 56 PSF and 50 SD and followed them over 1364 ± 543 days. The composite endpoint was MACE such as all-cause death, lethal arrhythmia, LV assist device implantation, heart transplantation or stroke due to cardiac emboli.

Results: LVEF was inversely related to %LGE in total of 106 HCM (r = -0.63; p < 0.0001). HCM with PSF showed higher EF (61±5% versus 34±12%, p < 0.0001) and lower %LGE (12±11% versus 30±15%, p < 0.0001) than HCM with SD. During the follow up, 17 of 50 HCM with SD and 1 of 56 HCM with PSF developed MACE (34% versus 2%, p < 0.0001). In total of 106 HCM, multivariable analysis revealed LVEF as the only independent predictor of MACE. The risk decreased as EF increased (hazard ratio [HR]: 0.92 / %, 95 % confidence interval [CI] = 0.84 to 0.99, p = 0.028). Interestingly, in 50 HCM with SD, %LGE was the only independent predictor of MACE although EF was not an independent predictor. The risk increased as %LGE increased (HR: 1.08 / %, 95 % CI = 1.01 to 1.16, p = 0.018). In 56 HCM with PSF, no predictor of MACE existed due to low frequency of MACE.

Conclusion: These results demonstrated that two-staged classification by LVEF followed by %LGE when LVEF < 50% is useful for risk stratification in HCM. We suggest that, in addition to screening HCM with SD, quantification of LGE is useful for predicting MACE in HCM with SD.