All-cause mortality and cardiovascular death in 52091 patients with hypertrophic cardiomyopathy. A nationwide cohort study

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Background: Patients with hypertrophic cardiomyopathy (HCM) have high risk of death related to cardiovascular (CV) death. Improvements in risk stratification are needed to help identify those HCM patients at higher risk of all-cause death and cardiovascular death.

Methods: This French longitudinal cohort study from the database covering hospital care from 2010 to 2019 analyzed adultshospitalized with isolated HCM. The overall sample of 52,091 patients was randomly partitioned into derivation (n=26,067) and validation (n=26,024) populations. A logistic regression model was used to construct HCM death and CV-death scores in the derivation sample, which were compared to the Charlson index, Frailty index and CHA2DS2VASc scores using c-indexes and calibration analysis.

Results: In 52,091 patients with isolated HCM, 12,676 (24.0%) died during follow-up of 3.0±2.8 years (median 2.3, interquartile range 0.4–5.0). Rate of all-cause death was 8.10%/year (7.96–8.24) and was 2.76%/year (2.68–2.84) for CV death.Independent predictors of CV death in HCM were older age, diabetes mellitus, heart failure, history of pulmonary edema, atrial fibrillation, ventricular tachycardia or fibrillation, ischemic stroke, while smoking and poor nutrition were associated with better survival (all $p<0.05$). In addition to these, male sex, vascular disease, alcohol related diagnoses, kidney disease, lung disease, liver disease anemia and cancer were independent predictors of all-cause death. In the derivation cohort, c-indexes for the HCM death score were 0.720 (0.713–0.727) for all-cause death and 0.695 (0.685–0.705) for CV death. For the HCM CV-death score, c-indexes were 0.679 (0.671–0.686) for all-cause death and 0.723 (0.712–0.733) for CV death. Performances were very similar in the validation cohort. Both scores had good calibrations. Charlson and Frailty indexes however had a better clinical usefulness than the HCM death score and HCM CV-death scores for predicting all-cause death. Decision curve analysis for CV death demonstrated that the HCM CV-death score had the best clinical usefulness of all the tested risk scores.

Conclusion: HCM patients have a high risk of all-cause and CV mortality. Independent predictors of CV-mortality in HCM were used to derive and validate a simple risk prediction model (French HCM CV-mortality score) which performed better than clinical scores, Charlson Index and Frailty Index; showing the best clinical usefulness, with good calibration.