Utility of scoring system including relative apical sparing pattern for screening cardiac amyloidosis in patients with left ventricular hypertrophy

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Background: Cardiac amyloidosis (CA) is an infiltrative disease mimicking left ventricular hypertrophy (LVH), although its prognosis is poorer than other diseases with LVH. Moreover, because CA is treatable, appropriate screening for CA is an important area of study for clinicians to prevent and treat the disease. Several imaging predictors of CA have been reported so far;. in particular, deformation parameters such as relative apical sparing patterns of longitudinal strain (RASP) may diagnose CA with better precision than conventional parameters. Accordingly, we hypothesized that the inclusion of deformation parameters into the established diagnostic parameters would permit derivation of a risk score for CA screening in patients with LVH. Thus, we aimed to 1) investigate the incremental benefits of deformation parameters over established diagnostic parameters for CA screening in patients with LVH; 2) determine the risk score to screen CA patients with LVH using all of these variables; and 3) externally validate the score.

Methods: We retrospectively studied 295 consecutive non-ischemic patients with LVH who underwent echocardiography as well as the detailed work-up for LVH (biopsy, technetium pyrophosphate scintigraphy (99mTc-PYP) or cardiac magnetic resonance imaging) (median age, 67 years; MWT, 12 mm). CA was diagnosed by biopsy or 99mTc-PYP. The base model consisted of age (≥65 [male], ≥70 [female]), low voltage in electrocardiography, and posterior wall thickness ≥14 mm in reference to previous studies. Continuous echocardiographic variables were binarized by the use of generally accepted external cutoff points to avoid best clinical scenario. Incremental benefits were assessed using receiver operating characteristic curve analysis and area under the curve (AUC) comparison. Multiple logistic regression analysis was performed to determine the risk score. The score was then validated in the external validation sample (N=178, median age, 70 years; MWT, 12 mm).

Results: CA was observed in 54 patients (18%) and of the several echocardiographic parameters studied, only RASP demonstrated a significant incremental benefit for the screening of CA over the base model (Figure A). After multiple logistic regression analysis in the prediction of CA with 4 variables (RASP and basal model components), each was assigned a numeric value based on its relative effect (Figure B). The incidence rate of CA clearly increased as the sum of the risk score increased (Figure C). The score had good discrimination ability, with an AUC of 0.87, a total score of ≥2 with 70% sensitivity and 90% specificity. Similarly, the discrimination ability of the score in the validation cohort was sufficient (AUC = 0.87).

Conclusion: Overall, we determined a simple risk score including RASP to screen CA. This score takes into account 4 common parameters used in daily practice, and therefore, has potential utility in risk stratification and management of patients with LVH.
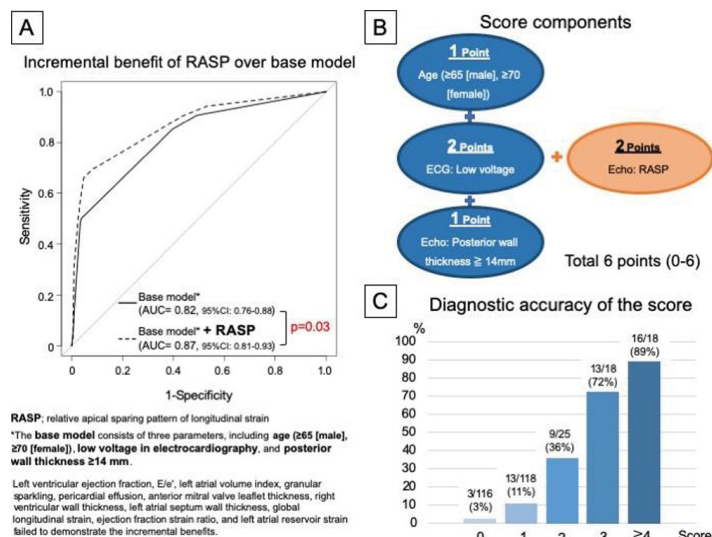
Figure 1