
Format: Abstract


Development and Validation of a New Risk Prediction Score for Life-Threatening Ventricular Tachyarrhythmias in Laminopathies.


Author information

Abstract

BACKGROUND: An accurate estimation of the risk of life-threatening (LT) ventricular tachyarrhythmia (VTA) in patients with LMNA mutations is crucial to select candidates for implantable cardioverter-defibrillator implantation.

METHODS: We included 839 adult patients with LMNA mutations, including 660 from a French nationwide registry in the development sample, and 179 from other countries, referred to 5 tertiary centers for cardiomyopathies, in the validation sample. LTVTA was defined as (1) sudden cardiac death or (2) implantable cardioverter defibrillator-treated or hemodynamically unstable VTA. The prognostic model was derived using the Fine-Gray regression model. The net reclassification was
compared with current clinical practice guidelines. The results are presented as means (SD) or medians [interquartile range].

**RESULTS:** We included 444 patients, 40.6 (14.1) years of age, in the derivation sample and 145 patients, 38.2 (15.0) years, in the validation sample, of whom 86 (19.3%) and 34 (23.4%) experienced LTVTA over 3.6 [1.0-7.2] and 5.1 [2.0-9.3] years of follow-up, respectively. Predictors of LTVTA in the derivation sample were: male sex, nonmissense LMNA mutation, first degree and higher atrioventricular block, nonsustained ventricular tachycardia, and left ventricular ejection fraction (https://lmna-risk-vta.fr). In the derivation sample, C-index (95% CI) of the model was 0.776 (0.711-0.842), and the calibration slope 0.827. In the external validation sample, the C-index was 0.800 (0.642-0.959), and the calibration slope was 1.082 (95% CI, 0.643-1.522). A 5-year estimated risk threshold ≥7% predicted 96.2% of LTVTA and net reclassified 28.8% of patients with LTVTA in comparison with the guidelines-based approach.

**CONCLUSIONS:** In comparison with the current standard of care, this risk prediction model for LTVTA in laminopathies significantly facilitated the choice of candidates for implantable cardioverter defibrillators.

**CLINICAL TRIAL REGISTRATION:** URL: https://www.clinicaltrials.gov. Unique identifier: NCT03058185.

**KEYWORDS:** death, sudden; defibrillators, implantable; tachycardia, ventricular

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