A prognostic model for overall survival in sporadic Creutzfeldt-Jakob disease

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Background: Sporadic Creutzfeldt-Jakob disease (sCJD) is the world's most common invariably fatal human prion disease with an incidence rate of 1–2 cases per million and year. Disease duration averages 5–6 months from diagnosis to death, but ranges from weeks to several years.
Aims: To develop an individual prognostic prediction model based on cerebrospinal fluid (CSF) biomarkers and other proposed disease survival modifiers, which are easily obtainable in routine settings at the time of diagnosis.
Methods: Probable or definite sCJD cases from a German surveillance study (1993–2017) were included. The prognostic accuracy to predict overall survival after sCJD diagnosis was measured by the *c* statistic of a model derived from a multivariable Cox proportional hazard regression.
Results: Complete information about age, sex, codon 129 genotype, presence of 14-3-3 in the CSF, and CSF tau concentrations was available for 1,226 out of 2,908 sCJD cases. The median age at diagnosis was 66 years (range 19–89 years). The male-to-female ratio was 1:1. A Cox proportional hazard model containing age, sex, genotype, CSF tau and the interaction terms age × tau, sex × tau, and sex × genotype was selected as the model with the highest c statistic (0.686, 95% CI 0.665–0.707) using cross-validation. This model was well calibrated. A score chart was derived to predict 6-month survival and median survival time (Figure 1).
Conclusions: We developed an individual prediction model with moderate to good accuracy. The score chart developed in this study serves as a hands-on prediction tool for clinical practice, allowing better planning of care after sCJD diagnosis and easier identification of potential participants for future treatment trials.

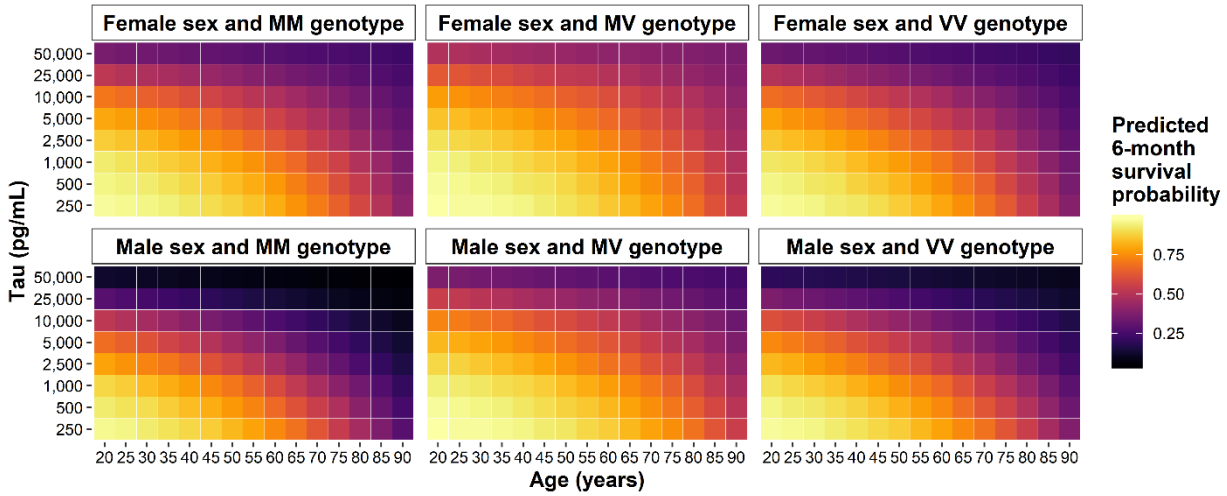

Figure 1: Score chart for predicting 6-month survival probability of sCJD patients

Keywords
Cerebrospinal fluid, neurodegeneration, prognosis, sporadic Creutzfeldt-Jakob disease, tau