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Non-motor markers improve survival prediction in Progressive Supranuclear Palsy

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State of the art
The pathology of Progressive Supranuclear Palsy (PSP) causes Richardson's syndrome (RS) and variant clinical phenotypes, with differential cognitive, behavioural and motor deficits. Survival is 3-4 years from diagnosis. The PSP Rating Scale (PSPRS) is prognostically informative, but the impact of cognitive/behavioural change on survival is less clear. Here we test univariate and multivariate models of survival to determine the best clinically-applicable prediction of survival.

Methodology
The MDS 2017 criteria were used to phenotype patients at the Cambridge Centre for Parkinson-plus (UK). Univariate and multivariate logistic regression models assessed the relationship between survival and clinical variables (PSPRS, MMSE, Addenbrooke's Cognitive Examination, Cambridge Behavioural Inventory, CBI, alone or in combination).

Results
335 people (male=56%, age 71.4±7.2 years) were identified with possible, probable or definite PSP (RS: n=233, male=53%, age 71.5±7.3years; or variant-PSP: n=102, male=64%, age 71.2±6.8 years). RS and variant groups had similar disease severity at baseline assessment (PSPRS: 35.6±14 vs 34.6±15.8, p=0.6) and survival (RS 6.1±2.7 years, variant-PSP 6.8±3.5 years, p=0.2). For 3-year mortality, PSPRS was the most reliable single predictor (AUC=0.68). Age, sex and PSPRS improved the model (AUC=0.71) but over all models Akaike's Information Criterion identified the best model for RS to include PSPRS, CBI and MMSE (AUC=0.79, p=0.01). CBI and MMSE also improved the model for variant-PSP (PSPRS, CBI and MMSE AUC=0.89 vs 0.73, p=0.01).

Conclusion
Inclusion of cognitive and behavioural measures improves the prediction of mortality in PSP. Such non-motor features may improve stratification and design of clinical trials of PSP disease-modifying treatments.

Conflicts of interest

N/A