

Symptom-staging for primary progressive aphasia syndromes

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State-of-the-art: The primary progressive aphasia (PPAs) are complex language-led neurodegenerative diseases comprising three main variants: semantic (svPPA), nonfluent/agrammatic (nfvPPA), and logopenic (lvPPA). No clinically-defined staging system describing symptom progression in each main PPA variant separately currently exists. Here we propose a new symptom-led staging system for major PPA syndromes, based on detailed online surveys of patients' primary caregivers, covering four broad symptom domains (communication, general cognition, behaviour and wellbeing, and physical neurological).

Methodology: We solicited initial feedback on the proposed symptoms and stages from 118 caregiver members of PPA Support Groups in the UK. From another 110 UK and Australian PPA caregivers, we then collected qualitative and quantitative feedback to refine the proposed stages, for each PPA subtype separately. Symptoms were retained if they were rated as 'present' by more than 50% of caregiver respondents representing a PPA subtype, and moved to an earlier/later stage based on respondent majority consensus.

Results: A six-stage framework was adopted for each variant, ranging from 1 (Very mild) to 6 (Profound). Fifty-four symptoms were included across the six stages for svPPA; 41 for nfvPPA; and 46 for lvPPA. Earlier stage symptoms were typically specific to each variant, with convergence of symptoms across variants in later stages. Using framework analysis of qualitative responses, we identified five major themes and 15 subthemes covering respondents' experiences of PPA, and suggestions for staging implementation.

Conclusions: These newly-defined stages for the three main PPA subtypes promise to facilitate diagnosis, prognosis, care, and treatment planning for people living with PPA.

Conflicts of interest

N/A