Perspectives and a Systematic Scoping Review on Longitudinal Profiles of Posterior Cortical Atrophy Syndrome

Kate Abe-Ridgway BS, Victoria Pelak MD, Asher Mahmood MS. Departments of Neurology and Ophthalmology, University of Colorado, Aurora CO.

University of Colorado Anschutz Medical Campus

Background

- Posterior Cortical Atrophy (PCA) is a neurodegenerative syndrome characterized by prominent visual dysfunction (alexia, visual space and object perception dysfunction, apraxia, homonymous visual field defects, and elements of Balint and Gerstmann syndromes) and progressive dementia
- PCA is considered a "visual variant" of Alzheimer's Disease (AD)
- Despite its recognition as a distinct clinical entity, there is a significant lack of comprehensive longitudinal data on PCA
- The lack of longitudinal data and validated outcome measures specific to PCA has historically excluded these patients from clinical trials for potential dementia treatments
- Understanding the longitudinal profiles of PCA is crucial for informing patients and families about prognosis, guiding appropriate clinical care, and developing outcome measures for inclusion in treatment trials.

Methods

- A search was conducted through PubMed databases for English-language articles published between 1976 and August 8, 2022, using the terms (longitudinal or natural history or follow-up) and (posterior cortical atrophy)
- A single reviewer screened titles and abstracts, and then full-text articles, for studies that included at least two PCA participants and reported longitudinal measures
- For included studies, we extracted data on sample size, PCA diagnostic criteria, baseline symptom duration, length of follow-up, and longitudinal measures and outcomes

Results

- We identified 13 longitudinal studies of PCA, with only two having over 30 participants
- PCA shows faster progression and greater severity of cognitive/functional decline and brain atrophy in posterior regions compared to non-posterior areas, distinguishing it from typical Alzheimer's disease (AD)
- Longitudinal studies using MRI show fastest atrophy in the occipital and parietal lobes in PCA, the areas responsible for visual processing and spatial awareness
- Longitudinal cognitive measures that show distinct trajectories in PCA are primarily those assessing higher-order visual processing, which tend to decline rapidly. Conversely, working memory skills appear to decline at a slower rate in PCA
- Limited survival data from two trials estimates a median survival of 8-10 years after PCA diagnosis

Figure 1 – study identification flow diagram

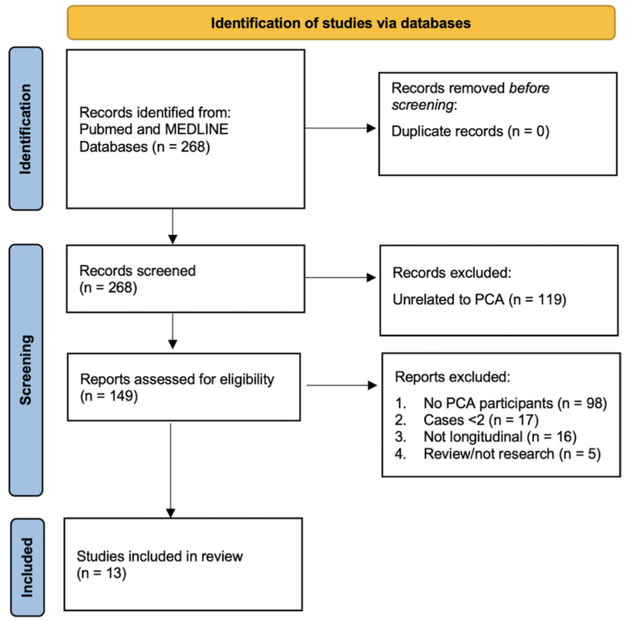
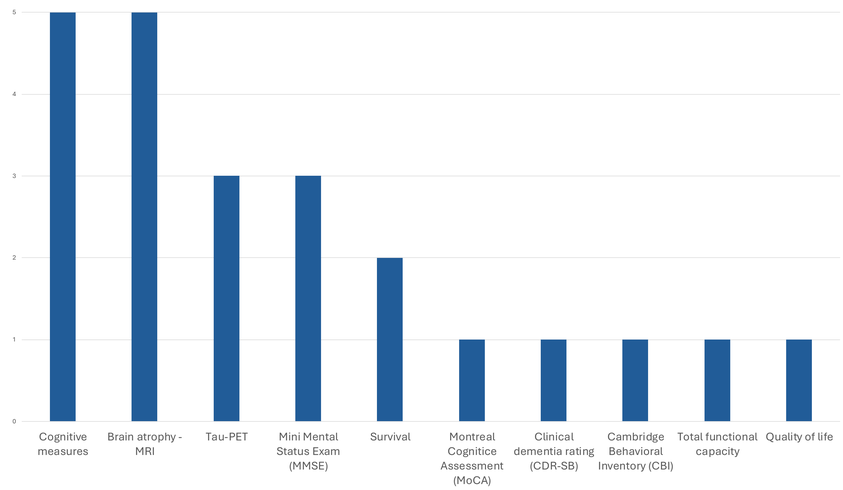


Figure 2 – Longitudinal measures tracked in PCA studies



Conclusions

- This review demonstrates large gaps in our understanding of the clinical progression of PCA, including expected biomarker progression, cognitive function, and survival
- Understanding PCA's longitudinal progression is vital for accurate prognosis for patients and their families, enabling tailored symptomatic treatments throughout the syndrome.
- Identifying distinct PCA cognitive trajectories and longitudinal biomarker changes is essential for developing PCA-specific outcome measures, which are needed to include PCA patients in treatment trials.

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