FULL-LENGTH ARTICLE NPub.org/x4ey2i

ALS-specific cognitive and behavior changes associated with advancing disease stage in ALS

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Study objective

To elucidate the relationship between disease stage in amyotrophic lateral sclerosis (ALS) and ALS-specific cognitive and behavioral changes.

Summary results

ALS-specific cognitive deficits and behavioral impairments are more common in more advanced ALS stages.

What is known and what this paper adds

Cognitive deficits and behavioral impairments commonly occur in ALS, but past longitudinal studies have been confounded by factors such as small sample sizes. This study examined cognitive deficits and behavioral impairments across different ALS stages in a large cross-sectional patient sample.

Participants and setting

This study recruited 161 patients (67.1% male; mean age, 61.39 ± 11.58 years) who met the El Escorial diagnostic criteria for possible, probable, or definite ALS from centers in Edinburgh, Dublin, and London between July 2014 and July 2016. This study also recruited 80 demographically matched healthy controls (60% male; mean age, 60.83 ± 13.23 years) without any family history of ALS from Dublin and Edinburgh.

Design, size, and duration

Demographic and clinical data were collected through semi-structured interviews. The patients were sorted into disease stage groups based on the King's Clinical Staging System. Each participant's cognitive performance was assessed with the Edinburgh Cognitive and Behavioral ALS Screen (ECAS). The ECAS includes a caregiver interview that was used to detect behavioral impairments.

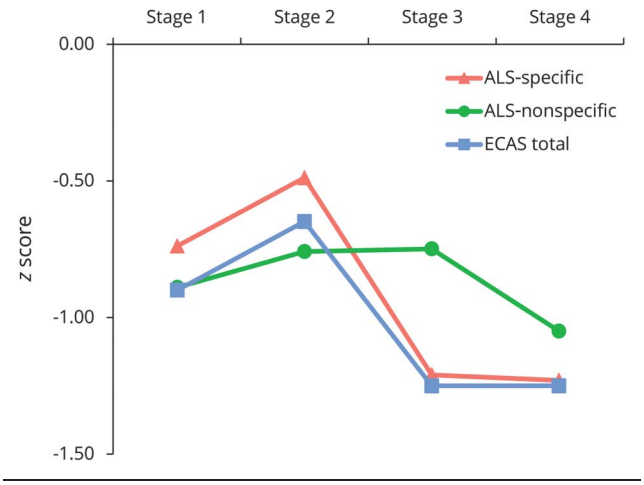
Primary outcome measures

The primary outcomes were differences in cognitive or behavioral ECAS results between ALS stages.

Main results and the role of chance

More advanced ALS stages were associated with greater cognitive dysfunction in the ECAS ALS-specific score of executive, language, and letter fluency domains ($p = 0.022$) but not in the ALS nonspecific score of memory and visuospatial functioning. More advanced ALS stages were associated with greater behavioral impairment burdens ($p < 0.001$).

Figure Cognitive performance across ALS stages



Bias, confounding, and other reasons for caution

This study's cross-sectional design limited its ability to elucidate longitudinal changes in ALS pathology. Patients with severe cognitive deficits or behavioral impairments might have been underrepresented.

Generalizability to other populations

This study's large sample size and multisite nature favor the generalizability of the results.

Study funding/potential competing interests

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A draft of the short-form article was written by M. Dalefield, a writer with Editage, a division of Cactus Communications. The authors of the full-length article and the journal editors edited and approved the final version.

