ALS-CBS™: A Cognitive Behavioral Screen for Use with ALS Patients

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OBJECTIVE:
To report updated data on the ALS Cognitive Behavioral Screen (ALS-CBS™).

BACKGROUND:
A screening method for detecting cognitive and behavioral impairment in ALS is most helpful in clinical settings where resources for detailed testing are limited.

The ALS-CBS™ is a brief assessment tool, developed at the Forbes Norris ALS Center (FNC), which contains tasks and questions sensitive to frontal lobe dysfunction.

The screen consists of a cognitive section with five domains specific to frontal lobe function, and a behavioral section composed of caregiver-directed questions intended to detect changes since disease onset.

The screen takes less than 5 minutes and can be completed in a routine clinical setting by any member of the care team.

METHODS: Phase I:
• A preliminary version of the ALS-CBS was administered to 150 consecutive patients at three multidisciplinary ALS centers.
• Preliminary validation was performed by two centers (Forbes Norris Center and Baylor University) and presented at the 18th Int’l Symposium last year.
• Based on preliminary results, the ALS-CBS™ was modified into its current version (See Figures 1 and 2)

METHODS: Phase II:
Preliminary validation of the screen was completed in three separate analyses:
• ALS-CBS™ scores of ALS patients (n = 80) were compared to non-ALS controls (n=15)
• 10 ALS patients and 4 ALS-FTD patients completed both the ALS-CBS™ and a complete neuropsychological assessment battery

RESULTS:

Cognitive Results
Normal (non-ALS) controls mean: 18.6 (0.8)
ALS cognitively normal mean: 18.1 (1.3)
ALS cohort (excluding FTD) mean: 15.9 (3.3)
ALS-FTD mean: 2.5 (range 0-5)

Cognitive score & FVC: 0.20   ns
Cognitive score & ALSFRS-R:      0.29   ns
Cognitive score & cognitive score: 0.37
Cognitive score & symptom duration 0.08   ns

Cognitive score & age -0.04   ns
Cognitive score & symptom duration     0.08   ns
Cognitive score & education: 0.38
Correlations with Cognitive Score of ALS-CBS™

Behavioral Results
Normal (non-ALS) controls mean: 44 (1)
ALS behaviorally normal mean: 41 (3.3)
ALS cohort (excluding FTD) mean: 35 (9)
ALS-FTD mean: 21 (range 11-36)

Behavioral score & FVC: 0.30   ns
Behavioral score & ALSFRS-R:      0.29   ns
Behavioral score & cognitive score: 0.37
Behavioral score & symptom duration -0.22 ns
Behavioral score & FrSBe -0.89

This correlation with standardized change scores (Total T scores: current-premorbid) from the Frontal System Behavioral Scale (FrSBe) Family Rating Form suggests that the behavioral screen score may estimate results on a standardized behavioral rating scale.

The most common behavioral changes endorsed by caregivers of ALS-FTD patients include:
• Decreased emotional responsiveness
• Withdrawal without sadness
• Confusion or distraction
• Decreased awareness/denial of problems and changes

METHODS: Phase III:
Preliminary validation of the screen was completed in three separate analyses:
• ALS-CBS™ scores of ALS patients (n = 80) were compared to non-ALS controls (n=15)
• 10 ALS patients and 4 ALS-FTD patients completed both the ALS-CBS™ and a complete neuropsychological assessment battery

CONCLUSIONS:
• This brief screening tool for identifying cognitive and behavioral impairment appears useful and valid in the clinical setting.
• Screen scores should be interpreted with caution in patients with advanced age (>80) or other risk factors such as significant vascular disease, head injury, or major depression. For older patients, a MMSE or other screen should be considered to rule out Alzheimer’s disease or other dementia.
• Cognitive scores are moderately correlated with education, and therefore screens for patients with limited education need to be interpreted with caution.
• This screen is intended to identify patients in need of further assessment and should not be used as a substitute for standardized testing or formal assessment. A diagnosis of FTD or other dementia should not be given solely on the basis of this screen.