California Pacific Medical Center
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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:

Mean age: 59
Mean FVC: 73%
Mean education: 15.64 yrs
Average ALSFRS-R: 32
Mean disease duration: 2.7 yrs

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)

Correlations with Cognitive Score of ALS-CBS™

Cognitive score & FVC:	-0.08	ns
Cognitive score & ALSFRS-R:	0.04	ns
Cognitive score & education:	**0.38**	
Cognitive score & age:	-0.11	ns
Cognitive score & symptom duration	0.08	ns

Suggested Cognitive Cutoff Scores	Total: 20
Normal Scores	≥16
Suspected Impairment (2-3 SD below mean)	12-15
Probable Impairment (>3 SD below mean)	≤ 11
ALS-FTD High Suspicion (10 SD below mean)	≤ 5

Figure 1: Current Version of Cognitive Section of ALS-CBS™

Figure 2: Current Version of Behavioral Section of ALS-CBS™

RESULTS:

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)

Correlations with Behavioral Score of ALS-CBS™

Behavioral score & FVC:	0.20	ns
Behavioral score & ALSFRS-R:	0.29	ns
Behavioral score & cognitive score:	**0.37**	
Behavioral score & age	-0.04	ns
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 included:

- Decreased emotional responsiveness
- Withdrawal without sadness
- Confusion or distraction
- Decreased awareness/denial of problems and changes

Suggested Behavioral Score Cutoffs	Total: 45
Normal Scores	≥ 35
Suspected Impairment (2-3 SD below mean)	28-34
Probable Impairment (>3 SD below mean)	< 28
ALS-FTD High Suspicion (6 SD below mean)	≤ 21

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.