Preliminary Validation of the ALS Cognitive Behavioral Screen (ALS-CBS)™

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BACKGROUND: Research has shown that 48.5% of patients demonstrate cognitive decline at the time of ALS diagnosis and 15% meet criteria for Frontotemporal Dementia (FTD) (Wheaton et al., 2007, In Press). However, full neuropsychologic testing for cognitive and behavioral changes is often unavailable or difficult to complete given the patients' significant physical symptoms. Thus, a brief screening measure to identify ALS patients who may benefit from comprehensive neuropsychological assessment is warranted.

OBJECTIVE: The ALS Cognitive Behavioral Screen (ALS-CBS) was created at the Forbes Norris ALS Research Center with the goal of identifying ALS patients who have cognitive and/or behavioral changes suggestive of FTD. This measure is composed of 10 items that examine changes in frontal lobe functioning, specifically attention, working memory, ocular function and verbal fluency. The screen also consists of an 8-item caregiver-rated behavioral change questionnaire. The goal of this study is to provide an introduction to and preliminary validation of the ALS-CBS.

METHODS
Participants: The ALS-CBS was administered to 24 ALS patients from the ALSA Clinic at Baylor College of Medicine with confirmed diagnoses based on cognitive and behavioral data. Patients did not differ on age, education, gender, or site of onset between the clinical diagnosis groups.

	Unimpaired (n=9)	Subtle Cognitive Changes (n=3)	Behaviorally Impaired (n=7)	FTD (n=5)
Age	52.8 (13.3)	50.5 (12.0)	59.0 (9.90)	50.0 (4.36)
Education	14.3 (1.63)	16.0 (2.83)	13.3 (4.51)	14.0 (4.0)
Gender (M/F)	4/5	2/1	5/2	4/1
Site of Onset Bulbar/Limb/Missing	5/4/0	1/1/1	1/3/4	2/3/0

RESULTS
Cognitive Component: Significant differences were found between diagnostic classifications on the cognitive component of the screen (p=0.02).
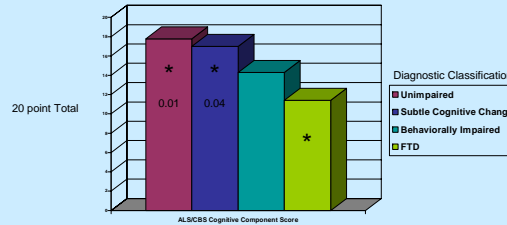
ALS patients diagnosed with FTD scored significantly lower than the unimpaired patients (p<0.01) and those with subtle cognitive changes (p=0.04); the FTD patients did not differ from those patients who were only behaviorally impaired (p=0.17).

A subset of 11 patients received both the cognitive component of the screen and a neuropsychological battery. The unimpaired and FTD patients did not differ on demographic variables. A K-means cluster analysis resulted in 91% correct classification by the screen.
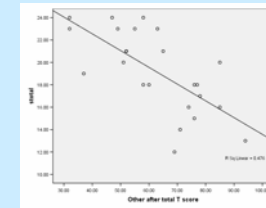
	Unimpaired (n=9)	Impaired (n=2)
Age	53.7 (11.4)	49.0 (5.66)
Education	14.9 (2.20)	14.0 (5.65)
Gender (M/F)	6/3	1/1
Site of Onset Bulbar/Limb/Missing	4/4/1	2/0

Neuropsychological Scores

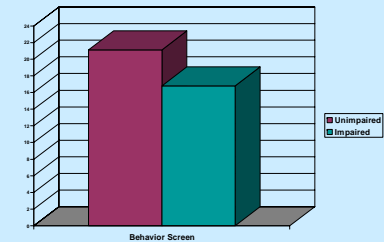|	Unimpaired		Impaired	
Measures	M	SD	M	SD
Basic Orientation	9.89	0.33	9.00	1.41
Reading	53.6	3.41	51.0	0.00
WAIS-III Similarities	23.7	4.82	15.5	7.78
VSAT-Time	73.5	23.0	89.0	0.00
Digit Span	17.3	2.82	13.0	2.83
Trails A*	26.3	9.71	53.5	4.95
FAS*	35.7	9.95	14.5	9.19
Animals*	19.9	3.76	8.50	3.54
RAVLT-Total	52.5	8.59	37.5	9.19
RAVLT-Immediate	10.0	4.28	7.50	0.71
RAVLT-Delay	10.4	3.50	8.00	1.41
BVMT-R-Immediate	21.1	5.33	19.5	0.71
BVMT-R-Delay	8.33	2.12	6.50	2.12
Trails B*	70.4	34.2	194	65.7
WSCT-Cat*	3.89	0.93	1.00	1.41
WCST-Persev	12.9	17.5	29.0	22.6
BAI	16.5	11.9	2.50	0.71
BDI-II*	12.1	3.98	3.50	3.54
Cognitive Screen	16.7	3.28	12.0	2.83

* p<.05

Behavior Component: The behavior component of the screen was examined in 24 ALS patients who had data for both the behavioral screen and the Frontal Systems Behavior Scale Other Rating (FrSBe).

The behavioral screen was significantly correlated with the total FrSBe T score (p=0.01), indicating a strong association between the behavioral screen and the FrSBe.

Behaviorally impaired ALS patients performed worse than their unimpaired counterparts on the behavioral screen (p=0.002). However, the behavioral screen did not accurately classify the patients into their respective categories (67%) based on a K-means cluster analysis.

CONCLUSIONS: Preliminary analyses show that the ALS-CBS cognitive component significantly distinguishes between cognitively impaired and non-impaired ALS patients and the validation of the measure in a small sample appears promising. The differences in written versus verbal responses will be examined in future research.

However, the behavioral component, while significantly correlated to the FrSBe, does not appear to capture ALS patients with mild to moderate behavioral changes. Future research will focus on modifying the behavioral screen and conducting a validation study with a larger sample.