

Title: Assessment of Cognitive and Behavioral Changes in Amyotrophic Lateral Sclerosis Related Frontotemporal Spectrum Disorder (ALS-FTSD): Call for a Global Normative Project.

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Cognitive and behavioral impairments in amyotrophic lateral sclerosis (ALS) have gained recognition as important facets of the frontotemporal neuropathology. ALS-Frontotemporal Spectrum Disorder (ALS-FTSD) has been identified in up to 75% of patients with ALS, with approximately 15-41% meeting diagnostic criteria of Frontotemporal Dementia (FTD). The presence of ALS-FTSD has been linked to shorter survival and reduced quality of life, two primary outcomes measures in drug treatment and development.

While we know that ALS and FTD may have a shared neuropathology and even arise from common genetic mutations, the majority of ALS research has been conducted exclusively with homogeneous groups. As we move towards genetic therapies for these diseases, expanding our understanding of ALS-FTSD with racially, culturally, and linguistically diverse populations will be essential.

The Edinburgh Cognitive and Behavioural ALS Screen (ECAS) and the ALS Cognitive Behavioral Screen (ALS-CBS) are two commonly used measures in ALS-FTSD research. While both have shown demonstrated strong sensitivity and specificity when applied to patient samples that reflect the original normative groups, their use with diverse populations remains limited. While both measures have undergone “transcultural normative projects,” review of resulting normative standards reveal a wide range of variability with significantly different psychometric properties (Table1).

The ECAS and ALS-CBS are promising assessment tools. While translation and validation efforts have been beneficial, they largely reflect isolated efforts. A global normative project is needed to facilitate collaborative pooling of data. Additionally, linguistic, cultural, and educational variables that influence test performance should be quantified and statistically controlled to improve generalizability.

Biography

Dr. Christin Bermudez Andrews completed her PhD at Nova South Eastern University. She is an Assistant Professor of Clinical Neuropsychology at the University of Miami Miller School of Medicine Department of Neurology. Dr. Bermudez specializes neurodegenerative disorders, motor neuron disease, and in linguistically and culturally informed cognitive assessments.

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Table 1.0 ECAS Normative Projects

SampleSize	Executive_Mean	Executive_SD	Langauge_Mean	Language_SD	Fluency_Mean	Fluency_SD	ALS_Specific_Mean	ALS_Specific_SD	Memory_Mean	Memory_SD	Visuospatial_Mean	Visuospatial_SD	ALS_NonSpecific_Mean	ALS_NonSpecific_SD	Total_Mean	Total_SD	Language	Age_Mean	Age_SD	Education_Mean	Education_SD
40.00	40.48	3.54	27.63	0.70	19.85	2.50	87.95	4.98	18.68	2.73	10.00	12.00	30.53	2.96	118.48	6.64	English	59.20	12.57	12.28	2.52
18.00	38.94	4.93	26.72	1.53	19.11	2.59	84.78	7.06	18.06	3.26	11.67	0.49	29.72	3.12	114.50	8.99	English	62.40	8.32	13.50	3.61
26.00	40.42	5.73	27.46	0.81	19.85	2.19	87.73	7.30	18.81	2.97	11.85	0.46	30.65	3.16	118.38	8.77	English	62.40	8.32	13.50	3.61
17.00	37.12	7.66	25.12	3.48	18.94	2.36	81.18	11.79	16.24	5.12	11.35	1.06	27.59	5.97	108.76	15.93	English	62.40	8.32	13.50	3.61
21.00	37.81	6.90	27.29	1.31	20.38	1.75	85.48	8.63	19.29	2.83	11.86	0.65	31.14	3.07	116.62	10.54	English	62.40	8.32	13.50	3.61
57.00	38.98	5.59	27.14	1.87	19.61	2.71	85.65	8.56	18.46	3.43	11.68	0.74	30.14	3.73	115.79	11.26	English	62.32	13.36	14.87	3.19
57.00	39.82	5.37	26.74	2.09	19.82	2.43	86.39	8.55	18.51	3.13	11.39	1.00	29.89	3.66	116.28	11.38	English	62.32	13.36	14.87	3.19
51.00	39.75	5.68	26.76	1.89	19.69	4.22	86.20	9.49	18.88	2.89	11.53	0.70	30.41	3.11	116.61	11.87	English	62.32	13.36	14.87	3.19
40.00	35.57	8.07	25.25	2.50	17.48	5.02	78.36	12.91	14.29	3.62	11.74	0.53	26.04	3.65	104.65	15.61	Spanish	55.40	#NULL!	19.08	#NULL!
277.00	33.78	8.52	24.25	3.80	18.82	5.91	76.86	15.97	15.03	4.29	11.06	1.36	26.09	4.93	102.95	19.54	Italian	55.25	13.15	10.88	4.97
160.00	37.10	5.50	26.90	1.60	18.80	4.00	82.80	8.50	16.20	3.80	11.70	0.70	28.00	4.00	110.70	10.80	German	60.00	12.60	13.90	3.10
84.00	37.53	6.26	21.71	3.06	19.44	3.98	78.68	9.85	15.60	3.89	11.96	0.24	27.56	3.93	106.24	12.16	Chinese	54.83	10.98	11.51	3.36