



THE RELATIONSHIP BETWEEN FRONTAL BEHAVIORAL AND COGNITIVE CHANGES IN ALS

Alicia Salamone, Diane M. Mosnik, PhD, Mariana Witgert PhD, Major Bradshaw PhD, and Paul E. Schulz, MD

BAYLOR COLLEGE OF MEDICINE—Houston, TX USA

INTRODUCTION

It is estimated that 51% of amyotrophic lateral sclerosis (ALS) patients exhibit at least a mild cognitive dysfunction throughout the course of their disease, with 15% of these patients meeting criteria for fronto-temporal dementia (FTD)¹. However, the frontal behavioral changes that may be associated with ALS have not been well described. Clinical observation suggests that patients with ALS tend to be particularly "nice", and they also may exhibit behavioral changes similar to those seen in FTD patients including apathy, disinhibition and/or executive dysfunction. These changes can affect patients' daily activities and increase caregiver's burden and distress².

Preliminary research by our group (in preparation) has revealed that 24.4% of ALS patients experience some form of behavioral impairment, with 31.1% showing changes in apathy, 16.9% showing changes in disinhibition, and 19.6% showing impairment in executive function in a cohort of 225 sALS patients. In this study, we sought to examine further the behavioral aberrations seen in ALS.

SUBJECTS

- Recruited from the Baylor College of Medicine MDA/ALS Outpatient Clinic
- Diagnosis of probable/definite ALS (El Escorial criteria)
- A majority of subjects were right handed (91.6%) and Caucasian (78.7%). 64.6% of subjects were male.

Table 1. Participant Demographics (N=225)

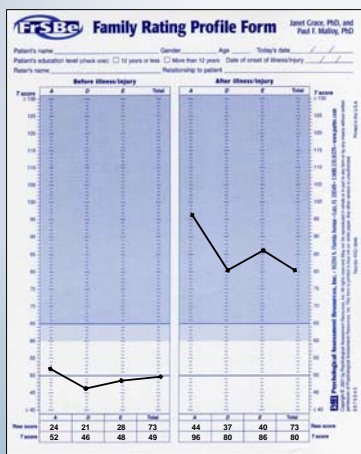
	Mean (SD)
Age	57.65 (13.98)
Education	14.05 (3.05)
FSIQ	100.67 (16.34)
VIQ	99.88 (13.12)
PIQ	101.71 (17.41)

METHODS

- Neuropsychological testing
- Family members rated patients on behavioral change from before- to after-illness
 - Frontal Systems Behavior Scale (FrSBe)³
 - Total Score
 - Apathy, Disinhibition, Executive Dysfunction Subscores
 - Increased points on the FrSBe indicated increased behavioral digressions

FrSBE Ratings (Figure 1)

- Ratings measured by T-Score
- T-score ≥ 65 indicates impaired behavior



REFERENCES

- Ringholz GM, et al. Neurology. 2005 Aug 23; 65(4):586-90.
- Hecht MJ, et al. Palliat Med. 2003 Jun; 17(4):327-33.
- Grace J & Malloy PF. Professional manual. 2001

RESULTS

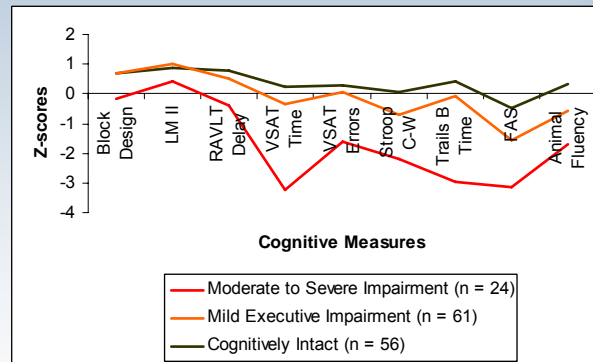


Figure 2. Cognitive Clusters. Cluster analysis of neuropsychological results found a three group solution (n = 141): Cognitively Intact (n = 56), Mild Executive Impairment (n = 61), and Moderate to Severe Impairment (n = 24). Z-scores for cognitive measures are displayed for each group.

Behavioral	Cognitive Impairment (n = 56)	Mild Executive Impairment (n = 61)	Cognitively Intact (n = 24)
Total	58.71 \pm 18.22	54.25 \pm 15.68	50.50 \pm 15.54
Apathy	64.42 \pm 17.65	57.43 \pm 16.12	52.30 \pm 16.04
Disinhibition	53.42 \pm 17.96	50.52 \pm 14.76	50.00 \pm 13.38
Executive Dysfunction	55.50 \pm 17.48	53.07 \pm 13.83	49.27 \pm 11.49

Table 2. Behavioral Impairments in Cognitive Clusters. FrSBe Total Score and Apathy, Disinhibition, and Executive Dysfunction Subscores \pm SD for Cognitive Groups.

FrSBe Scale	Mean T-Score \pm SD	Significance
Total (Before)	50.57 \pm 13.19	p < 0.01
Total (After)	57.62 \pm 16.66	
Apathy (Before)	48.21 \pm 11.69	p < 0.01
Apathy (After)	60.03 \pm 17.49	
Disinhibition (Before)	51.10 \pm 15.02	p < 0.05
Disinhibition (After)	53.62 \pm 15.96	
Executive Dysfunction (Before)	52.54 \pm 11.43	p < 0.01
Executive Dysfunction (After)	56.28 \pm 14.28	

Table 3. Behavioral Change in ALS.

FrSBe Total Score and Apathy, Disinhibition, and Executive Dysfunction Subscores were analyzed before illness and after illness onset using paired-sample T-tests (n = 39).

CONCLUSIONS

- Cluster analysis revealed 3 cognitive groups: Cognitively Intact, Mild Executive Impairment, and Moderate-Severe Impairment.
- A general trend emerged in which the highest FrSBe scores were found in the Moderate-Severe Impairment group and the lowest scores in the Cognitively Intact group, suggesting an interaction between cognitive impairment and behavioral dysfunction.
- Statistically significant changes in behavior were found from before to after-illness onset on the FrSBe Total Score and the three Subscores, with the greatest change noted on the Apathy scale.
- Caregivers should receive education regarding the effects and consequences of behavioral changes, such as apathy, executive dysfunction, and disinhibition, in their loved ones. Educating patients and caregivers will help them be able to cope with this disease and plan appropriately.
- Future studies will determine whether the identification of behavioral problems has any prognostic value in predicting cognitive decline.