Behavioral and Cognitive Dysfunction in Amyotrophic Lateral Sclerosis
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INTRODUCTION
About 49% of amyotrophic lateral sclerosis (ALS) patients exhibit at least mild cognitive dysfunction, and 15% meet criteria for frontotemporal dementia (FTD). Other evidence supporting the association between ALS and FTD includes pathologic, imaging, and genetics investigations.

In FTD, frontal-lobe mediated behavioral dysfunction is commonly found. This type of dysfunction, in the form of apathetic behavior, has been suggested in ALS. However, the extent to which there is behavioral dysfunction in ALS and its relationship to cognitive impairment requires further elucidation.

METHODS
- Behavioral Status
- Frontal Systems Behavior Scale (FrSBe)
- Family members rated patients on behavioral change from before- to after-onset of ALS
  - Measure behavioral dysfunction (Total Score) by combining subscores for Apathy, Disinhibition and Executive Dysfunction
  - Elevated scores on the FrSBe indicated increased behavioral digressions
  - T-score ≥ 65 shows clinical behavioral impairment
- Cognitive Status
- Comprehensive neuropsychological testing
- Cluster analysis using Block Design, Logical Memory II, RAVLT-Delay, VSAT-time and errors, Stroop color-word, Trails B-time, FAS and Animal Fluency found cognitive subgroups

RESULTS

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%), Caucasian (78.7%), and male (64.6%)

Table 1. Participant Demographics (N=225)

<table>
<thead>
<tr>
<th>Trait</th>
<th>Mean (SD)</th>
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<tbody>
<tr>
<td>Age</td>
<td>57.65 (13.98)</td>
</tr>
<tr>
<td>Education</td>
<td>14.05 (3.05)</td>
</tr>
<tr>
<td>FSIQ</td>
<td>100.67 (16.34)</td>
</tr>
<tr>
<td>VIQ</td>
<td>99.88 (13.12)</td>
</tr>
<tr>
<td>PIQ</td>
<td>101.71 (17.41)</td>
</tr>
</tbody>
</table>

Behavior impairment was found in a quarter of ALS patients
Apathetic behavior was the most frequent; however, disinhibition and executive dysfunction were also common.

Cluster analysis revealed 3 cognitive groups: Cognitively Intact, Mild Executive Impairment, and Moderate Impairment suggesting cognitive impairment exists on a continuum
Frequency of behavioral and cognitive impairment was not affected by site of onset
However, dysarthria and weak upper extremity function were not controlled for
Further studies should assess this question
Age did not impact frequency of cognitive impairment
Age was significantly older in patients with behavioral impairment vs those without behavioral impairment
Poor performance on Animals predicts poor behavioral performance

CONCLUSIONS
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