

Impact of Cognitive Impairment from Frontotemporal Spectrum Disorder on Quality-of-Life Outcomes in Amyotrophic Lateral Sclerosis

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Background

- Of those with ALS, up to 50% will experience cognitive impairment and 25% will be diagnosed with frontotemporal dementia (FTD)¹
- ALS is theorized to be on a spectrum with FTD, known as ALS-frontotemporal spectrum disorder (ALS-FTSD)¹
- It is generally understood that patients with ALS-FTSD have a poorer prognosis with shortened lifespan²
 - The effects on patient quality of life and caregiver burden have yet to be assessed

Questions

- How does cognitive impairment in patients with ALS affect the following:
 - Patient quality-of-life (QOL)
 - Patient depressive symptoms
 - Caregiver burden and QOL
 - End-of-life preferences
 - Opinion of the California End-of-life Options Act (EOLOA) for both patients and caregivers

Methods

Recruitment & Criteria

- Single-center cross-sectional study: patients from the UCSF ALS Clinic
- Recruited between August – November 2023 through direct recruitment in clinic or via EHR message (MyChart)
- Criteria: UCSF patient, diagnosed with ALS, caregiving needs of at least 20 hours/week, no FTD diagnosis

Screenings & Surveys

ALSFRS-R, ALS-CBS (Cognitive Behavioral Screen), McGill Quality of Life Scale – Revised, Geriatric Mood (Depression) Scale, Caregiver Burden Scale, UCSF End-of-Life Survey (for patients and caregivers)

Statistical Methods

Continuous variables were summarized using means (SDs) and medians (IQRs) as appropriate. Categorical variables were summarized with counts and percentages. Comparisons between those with and without cognitive impairments were made using t-tests for normally distributed variables and the Wilcoxon rank sum test for continuous variables that were not normally distributed. Categorical variables were compared between the two groups using chi-square tests, or Fisher's exact tests when appropriate.

Preliminary Results

48% (14/29) of patients tested positive for cognitive impairment (ALS-FTSD) using the ALS-CBS.

Table 1: Demographics and disease features of all 29 patients and p-values when compared between the ALS and ALD-FTSD groups.

Demographics	% (n/total)	p-value
Female identifying	45% (13/29)	0.58
Race – White	80% (23/29)	0.33
Ethnicity – Hispanic	7% (2/29)	0.14
Bachelor's degree or higher	76% (22/29)	0.19
Medicare/Medicaid (public) insurance	69% (20/29)	0.38
Working full- or part-time	31% (9/29)	0.31
Income >\$200,000 USD	24% (7/29)	0.75
(continued)	Median (IQR)	p-value
Age (years)	65 (59-73)	0.13
Time from ALS symptom onset (months)	39 (23-48)	0.27
ALSFRS-R at time of survey (out of 48)	32 (25-36)	0.11

Table 2: Symptoms of depression and preferences for the EOLOA in patients with ALS and ALS-FTSD.

	ALS % (n)	ALS-FTSD % (n)	p-value
Self-reported symptoms of depression	33% (5/15)	47% (7/15)	0.36
Aware of the EOLOA	(8/14)	(8/14)	1.00
Interested in the EOLOA	(7/14)	(5/14)	0.46

Figure 1: Patient QOL between those with ALS and ALS-FTSD based on a scale of increasing QOL from 1-10

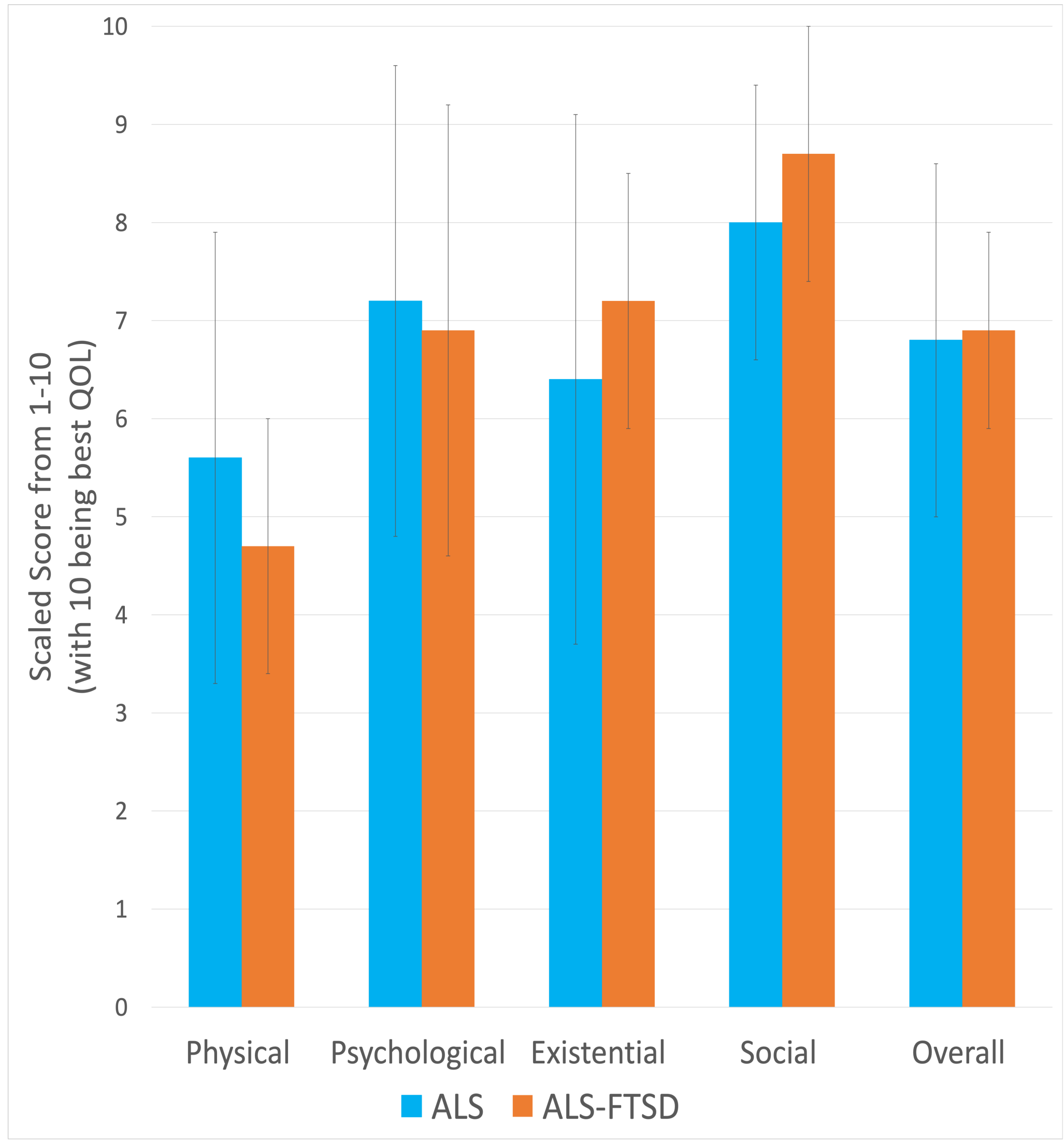


Figure 2: Select QOL features in caregivers/loved ones of those with ALS and ALS-FTSD scaled on a frequency of 1) never, 2) seldom, 3) sometimes, and 4) often.

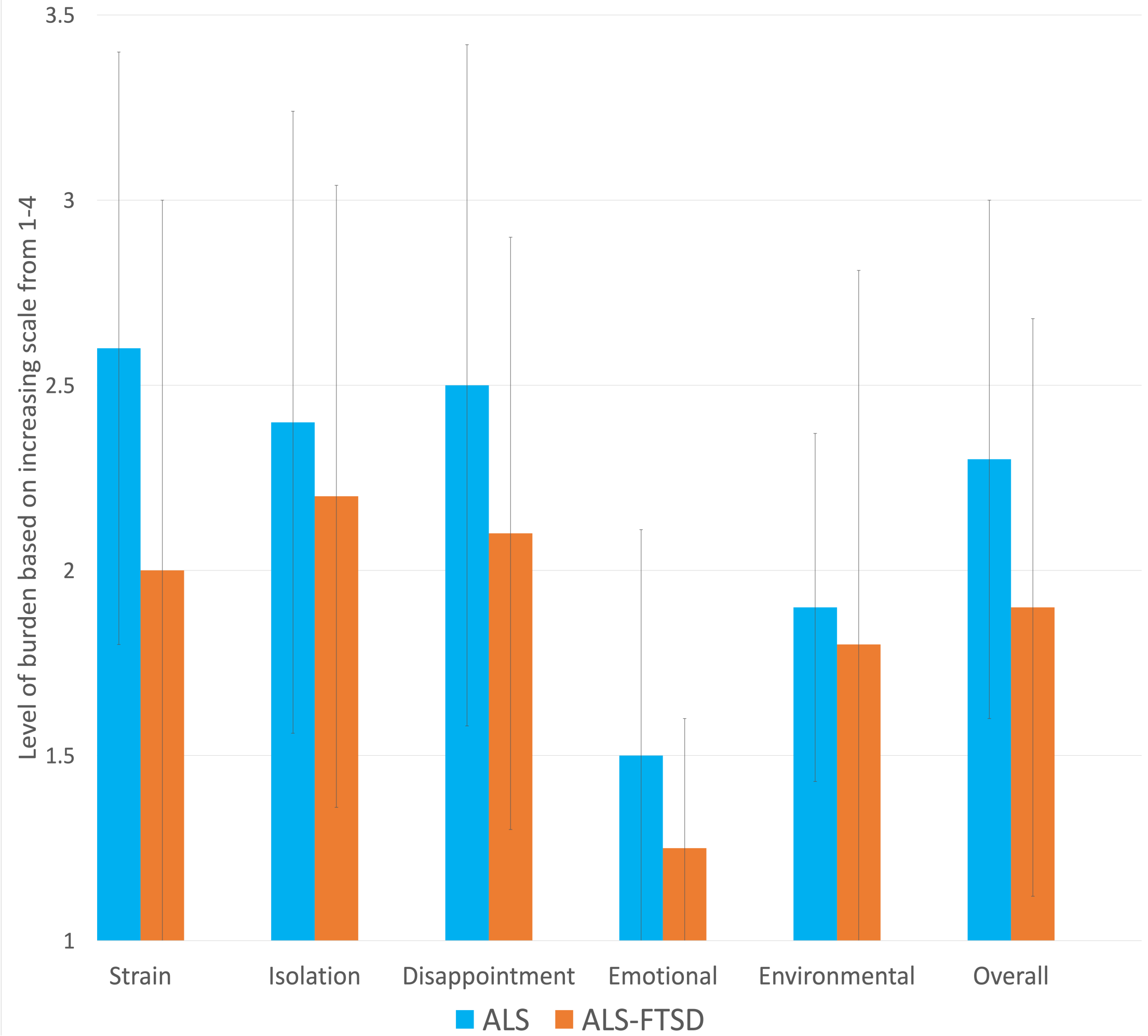


Table 3: Select statements and common sentiments from caregivers on how ALS has affected their QOL

On caring for their loved one...	"I fight a lot of feelings of inadequacy. I worry about the future symptoms and what I'll need to do - it gives me nightmares."
On resources for ALS patients and their families...	"There is very little help for everyone who has all the insurance and some income...We did everything we were supposed to do, followed all the rules, and left out to dry."
On how their own health is affected....	"I think I'm dealing with depression in a way I haven't before. It's like a train wreck waiting to happen. We spent our 4th anniversary in the clinic finding out he had ALS."

Discussion

- There is little difference in self-reported QOL, depressive symptoms, and EOLOA preferences between patients with ALS and ALS-FTSD
- Though there were not statistically significant differences, caregivers had lower mean burden in all categories when the patient had ALS-FTSD
 - Loved ones' comments reflected "frustration" with patient mood and stress/anxiety towards their illness
- Common sentiments from caregivers included:
 - Concern for future ALS symptoms, lack of resources for ALS patients, and personal health troubles
- More data is needed to understand if there are significant differences in caregiver burden and how they can be better supported

References

1. Strong MJ, Abrahams S, Goldstein LH et al. Amyotrophic lateral sclerosis - frontotemporal spectrum disorder (ALS-FTSD): Revised diagnostic criteria. Amyotroph Lateral Scler Frontotemporal Degener. 2017; 18(3-4):153-174.
2. Cividini C, Basaia S, Spinelli EG et al. Amyotrophic Lateral Sclerosis – Frontotemporal Dementia: Shared and Divergent Neural Correlates Across the Clinical Spectrum. Neurology. 2022; 98(4):e402–e415.

Future Directions

Limitations

- Small sample size: cancellations due to illness and ALS symptoms – we will be interviewing more people in the coming months
- Largely white, upper middle class sample
- Not all surveys were made specifically for the ALS population
- Surveys could be limited by privacy if the person is not alone

Streamlined data collection

- Complete CBS and ALSFRS-R in clinic or 15-minute video call
- Send other surveys via email to be completed on their own time
- Use ALS-specific surveys on depression and other QOL measures

Broader outreach

- Expansion across multiple sites to diversify population
- Working with the ALS caregiver Facebook group, which many of the participants are part of and referenced