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# FUNCTIONALITY AND QUALITY OF LIFE IN PATIENTS WITH AMYOTROPHIC LATERAL SCLEROSIS: A CROSS-SECTIONAL STUDY



original paper
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**PRISCILA GOMES DE JESUS OLIVEIRA, ROBERTA DA SILVA ARAMAKI, ALEX EDUARDO DA SILVA, GUSTAVO JOSÉ LUVIZUTTO, LUCIANE APARECIDA PASCUCCI SANDE DE SOUZA**

Federal University of Triângulo Mineiro, Uberaba, Brazil

ABSTRACT

**Purpose.** To identify factors constraining functionality and life quality in amyotrophic lateral sclerosis (ALS).

**Methods.** Cross-sectional study in 6 ALS patients aged 51.16 ± 8.70 years, with disease duration of 3.16 ± 2.63 years. Amyotrophic Lateral Sclerosis Severity Scale (ALSSS) and Amyotrophic Lateral Sclerosis Functional Rating Scale (ALSFRS-R/BR) served to evaluate functionality; Amyotrophic Lateral Sclerosis Assessment Questionnaire (ALSAQ-40) analysed life quality. Descriptive statistics was applied to clinical characteristics. The correlation of clinical profile and disease duration with the degree of functionality and life quality was verified by Spearman's correlation, with values considered significant if $p < 0.05$ and $r > 0.8$.

**Results.** There was a positive correlation of age with ALSFRS ($r = 0.852$), disease duration with ALSAQ-40/physical mobility ($r = 0.805$), ALSFRS-R with ALSSS ($r = 0.972$), physical mobility with food ($r = 0.881$), and communication with emotional status ($r = 0.947$). Negative correlations were observed of age with mobility ($r = –0.805$) and emotional status ($r = –0.876$), disease duration with ALSFRS-R ($r = –0.858$) and ALSSS ($r = –0.930$), ALSFRS-R with physical mobility ($r = –0.978$) and food ($r = –0.855$), ALSSS with physical mobility ($r = –0.910$) and daily life activity ($r = –0.874$).

**Conclusions.** Longer disease was associated with lower functionality and poor life quality. Better functionality indicated lower severity and higher life quality in physical mobility and food. Higher severity was bound with poor life quality in physical mobility and daily life activity. We also observed correlations of physical mobility with food, and communication with emotional status.

**Key words:** amyotrophic lateral sclerosis, quality of life, questionnaires

## Introduction

Amyotrophic lateral sclerosis (ALS) is a disease that affects lower motor neurons, located in the brainstem and anterior region of the spinal cord, and upper motor neurons, located in the motor area of the central nervous system [1–3]. Age is an important factor related to ALS development: the disease occurs mainly in patients aged 55–75 years [4]. In Brazil, it is estimated that the prevalence ranges from 0.9 to 5 per 100,000 inhabitants [5]. The average survival of patients after the onset of symptoms is 3–5 years [6]. ALS causes motor deficit and decreases functionality. The classic form of ALS is characterized by insidious onset, asymmetrical muscle weakness, fasciculation, hyperreflexia, and limb and tongue atrophy [7, 8].

There are currently new forms of ALS therapies, and the traditional assessment of the disease would not be sufficient to characterize some important changes during the evolution; the evaluation process would be facilitated by specific scales [9]. The comprehension of clinical and epidemiological factors in ALS patients correlated with functional impairment and quality of life is important to improve prognosis and reduce complications in these patients [10, 11].

*Correspondence address:* Gustavo José Luvizutto, Department of Applied Physical Therapy, Institute of Health Sciences, Federal University of Triângulo Mineiro, Casa Terra – Rua Capitão Domingos, 309. Abadia. Uberaba, MG, 38025-180, Brazil, e-mail: gustavo.luvizutto@uftm.edu.br

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ALS has no cure, and the patients are treated in tertiary reference centres in many countries. Although multidisciplinary treatment prolongs survival in ALS, the effect on quality of life is not known. ALS evaluation is a relevant topic to guide and identify significant and realistic goals for treatment [4]. Thus, the care of these patients by a multidisciplinary team (physicians, physiotherapists, nurses, psychologists, speech therapists, nutritionists, and social workers) trained with specific evaluation tools application facilitates the strategies and monitoring of ALS progression [12–14].

Thus, the present study aimed to characterize patients with ALS by means of an evaluation of the functionality and quality of life by using specific scales and to correlate these scales with age and the disease duration and severity.

## Material and methods

### Study design, setting, and participants

A cross-sectional, descriptive study was performed between June and December 2017. We included individuals of both sexes, living in the city of Uberaba, Minas Gerais, Brazil, with a documented or possible diagnosis of ALS. Patients with motor deficits resulting from other associated conditions and with cognitive impairment were excluded.

### Variables

a) Clinical and demographic variables: the general information on the participants were age, sex, and clinical data such as onset of symptoms and evolution of the disease.

b) Functionality: functionality was assessed by using the Amyotrophic Lateral Sclerosis Functional Rating Scale – Revised (ALSFRS-R/BR) and Amyotrophic Lateral Sclerosis Severity Scale (ALSSS) in the Portuguese version.

c) Quality of life: this was evaluated with the Amyotrophic Lateral Sclerosis Assessment Questionnaire (ALSAQ-40).

### Measurement

a) ALSFRS-R/BR: the scale involves 12 items, each scored 0–4, with the total score ranging from 0 to 48; 48 indicates normal functionality and 0 stands for serious impairment [15].

b) ALSSS: the scale is composed of 4 dimensions: lower extremity, upper extremity, speech, and swallowing; each dimension can be scored from 10 to 1. The score can vary between 4 (bad function) and 40 (function within normality) [16].

c) ALSAQ-40: the questionnaire includes 40 questions, divided into the following dimensions: daily life activity and independence (10 items); physical aspects (10 items); food (3 items); communication (7 items); and emotional aspects (10 items). Each item on the scale has the response options of 'never,' 'rarely,' 'sometimes,' 'often,' and 'always'; the higher the score, which can vary from 0 to 100, the worse the quality of life [17].

### Statistical methods

Descriptive statistics (mean and standard deviation) was applied to the data of the clinical, sociodemographic, topographical, and evolution characteristics of the patients. The correlation of the clinical profile, topographic data, and period of disease evolution with the degree of functionality and quality of life was verified by Spearman's correlation, with values considered significant at $p < 0.05$ and $r > 0.8$.

### Ethical approval

The research related to human use has complied with all the relevant national regulations and institutional policies, has followed the tenets of the Declaration of Helsinki, and has been approved by the Federal University of Triângulo Mineiro Institutional Review Board.

### Informed consent

Informed consent has been obtained from all patients or caregivers included in this study.

## Results

Overall, 18 individuals (11 men and 7 women) were identified with the diagnosis of ALS, and 6 (4 men and 2 women) met the inclusion criteria. The main characteristics of the patients are shown in Table 1. The mean age was 51.16 (± 8.70) years. Subjects 3 and 4 were diagnosed with bulbar ALS prior to the present study, while the others were diagnosed with typical ALS.

The results analysed by Spearman's correlation are presented in Table 2. There was a positive correlation of age with ALSFRS-R ($r = 0.852$) and disease duration with ALSAQ-40/physical mobility ($r = 0.805$), ALSFRS-R with ALSSS ($r = 0.972$), ALSAQ-40/physical mobility with ALSAQ-40/food ($r = 0.881$), ALSAQ-40/communication with ALSAQ-40/emotional ($r = 0.947$). However, negative correlations were observed for age

Table 1. Characteristics of the study subjects

| Subject, disease form | Gender | Age (years) | Disease duration (years) | Initial symptom |
|---|---|---|---|---|
| 1 Classic/defined | Male | 43 | 8 | Decreased upper limb strength |
| 2 Classic/defined | Male | 44 | 4 | Decreased upper limb strength |
| 3 Bulbar/defined | Female | 44 | 1 | Difficulty in speaking |
| 4 Bulbar/defined | Female | 54 | 2 | Difficulty in speaking |
| 5 Classic/defined | Male | 59 | 3 | Decreased upper limb strength |
| 6 Flail arm/defined | Male | 63 | 1 | Decreased upper limb strength |

Table 2. Values of Spearman's correlation between the variables of age, disease duration, ALSFRS-R, ALSSS, and ALSAQ-40

| Variables | | Age | Disease duration | ALSFRS-R | ALSSS | ALSAQ-40 | | | | |
|---|---|---|---|---|---|---|---|---|---|---|
| | | | | | | Physical mobility | Food | DLA | Communi-cation | Emotional |
| Age | (rho) | 1 | | | | | | | | |
| | (*p*) | | | | | | | | | |
| Disease duration | (rho) | –0.523 | 1 | | | | | | | |
| | (*p*) | 0.435 | | | | | | | | |
| ALSFRS-R | (rho) | 0.852* | –0.858* | 1 | | | | | | |
| | (*p*) | 0.028* | 0.029* | | | | | | | |
| ALSSS | (rho) | 0.797 | –0.930* | 0.972* | 1 | | | | | |
| | (*p*) | 0.010 | 0.007* | 0.001* | | | | | | |
| ALSAQ-40/ physical mobility | (rho) | –0.805* | 0.805* | –0.978* | –0.910* | 1 | | | | |
| | (*p*) | 0.002* | 0.049* | 0.001* | 0.012* | | | | | |
| ALSAQ-40/food | (rho) | –0.670 | 0.716 | –0.855* | –0.787 | 0.881* | 1 | | | |
| | (*p*) | 0.135 | 0.110 | 0.030* | 0.063 | 0.020* | | | | |
| ALSAQ-40/DLA | (rho) | –0.746 | 0.778 | –0.767 | –0.874* | 0.633 | 0.503 | 1 | | |
| | (*p*) | 0.100 | 0.068 | 0.075 | 0.023* | 0.177 | 0.309 | | | |
| ALSAQ-40/ communication | (rho) | –0.713 | 0.440 | –0.549 | –0.592 | 0.398 | 0.414 | 0.679 | 1 | |
| | (*p*) | 0.155 | 0.383 | 0.259 | 0.216 | 0.435 | 0.415 | 0.138 | | |
| ALSAQ-40/ emotional | (rho) | –0.876* | 0.455 | –0.696 | –0.678 | 0.593 | 0.599 | 0.678 | 0.947* | 1 |
| | (*p*) | 0.001* | 0.364 | 0.125 | 0.139 | 0.215 | 0.209 | 0.139 | 0.004* | |

ALSFRS-R – Amyotrophic Lateral Sclerosis Functional Rating Scale, ALSSS – Amyotrophic Lateral Sclerosis Severity Scale, ALSAQ-40 – Amyotrophic Lateral Sclerosis Assessment Questionnaire, DLA – daily life activity, rho – Spearman's correlation coefficient, *p* – correlation significance
* statistically significant values were $p < 0.05$ and $r > 0.8$

Table 3. Descriptive statistics of the scales results

| Variable | Minimum | Maximum | Mean (± *SD*) |
|---|---|---|---|
| Disease duration | 1 | 8 | 3.16 (± 2.63) |
| ALSFRS-R/BR | 15 | 48 | 33.5 (± 12.84) |
| ALSSS | 7 | 39 | 26.5 (± 11.36) |
| ALSAQ-40/physical mobility | 0 | 100 | 42.5 (± 46.44) |
| ALSAQ-40/food | 18 | 100 | 63.6 (± 35) |
| ALSAQ-40/DLA | 0 | 100 | 35.3 (± 35) |
| ALSAQ-40/communication | 0 | 100 | 72.0 (± 37) |
| ALSAQ-40/emotional | 0 | 75 | 53.0 (± 27) |

ALSFRS-R/BR – Amyotrophic Lateral Sclerosis Functional Rating Scale, ALSSS – Amyotrophic Lateral Sclerosis Severity Scale, ALSAQ-40 – Amyotrophic Lateral Sclerosis Assessment Questionnaire, DLA – daily life activity, *SD* – standard deviation

with ALSAQ-40/physical mobility ($r$ = –0.805) and ALSAQ/emotional ($r$ = –0.876), disease duration with ALSFRS-R ($r$ = –0.858) and ALSSS ($r$ = –0.930), ALSFRS-R with ALSAQ-40/physical mobility ($r$ = –0.978) and ALSAQ-40/food ($r$ = –0.855), ALSSS with ALSAQ-40/physical mobility ($r$ = –0.910) and ALSAQ-40/daily life activity ($r$ = –0.874).

In Table 3, we can observe the minimum and maximum values obtained in the scales.

## Discussion

In our study, it was observed that older patients presented better functionality and quality of life, and increased disease duration was associated with poor quality of life in the physical mobility domain. Correlations between ALSFRS and ALSSS were also revealed, and better functionality was associated with lower ALS severity. A low score in physical mobility was correlated with the food domain, and communication with the emotional status domain. We also noted that increased disease duration was related to lower scores in the functionality scales. Poor functionality was associated with poor quality of life in the physical mobility and food domains, and higher severity with poor quality of life in the physical mobility and daily life activity domains.

The present study demonstrated a high correlation of older age with better functionality and quality of life, and of longer disease duration with poor functional scales results and quality of life. In older individuals, the time between the onset of symptoms and diagnosis was shorter, thus a lower effect of ALS on the motor system was observed. ALS progression was correlated in another study with lower ALSFRS-R scores [18]. With the disease progression, there is an increase in motor neuron degeneration and motor function loss, which leads to difficulties in performing daily life activities, impairing functionality and quality of life.

Higher ALS severity was associated with poor functionality, as well as poor physical mobility and daily life activity results in ALSAQ-40. Other studies reported that greater severity of the disease occurred in more advanced stages, and motor impairment decreased the ability to perform daily life activities. The scientific literature identifies 3 stages of functional impairment during ALS progression: in the initial stage, the individuals independently perform daily life activities; the intermediate stage is characterized as a semi-dependent phase, and the patients need help to perform some tasks; in the final stage, the individuals need total assistance to perform daily life activities [19–22].

A low score in physical mobility was correlated with the food domain in our study. The deficits found in these domains can be explained by the fact that ALS is a degenerative disease of the whole motor neuron system, including progressive paralysis of skeletal muscles, with an impact on speech, swallowing, and respiratory muscles [23]. ALS causes a progressive loss of autonomy and need for assistance during daily life activities, mainly during eating. We also identified a significant correlation between communication and emotional state. The emotional function is an important domain studied by other authors, and it constitutes a high priority for clinical care since there is evidence of emotional stress in ALS individuals [24, 25]. Physical deterioration during ALS progression is associated with a loss of independence, inability to communicate, and a considerable emotional challenge [26, 27].

The physical mobility and food domains of ALSAQ-40 presented a significant correlation with functionality. The physical mobility domain was the most impaired in ALS patients in other studies, and showed correlation with quality of life, mainly in the food domain. There is an increase in the metabolic demand during ALS progressions, and weight loss has multifactorial causes, including oropharyngeal dysphagia, secondary loss of appetite, and loss of autonomy [28, 29].

The limitations of the study are related to the sample size and the absence of forced vital capacity data, which could not be analysed on account of the great heterogeneity of topographies and lack of the information in the medical charts. However, the purpose of the present study was to identify factors that negatively impacted on the functionality and quality of life in ALS patients. There is a need to improve the evaluation tools because of the complications during ALS progression, which refer to strength, tone, trophism, respiratory function, and depression, as well as to analyse the impact of the environment context.

## Conclusions

Our results revealed that an increase in disease duration was associated with lower functionality and poor quality of life, and better functionality with less severity and higher quality of life in the physical mobility and food domains. Higher ALS severity was associated with poor quality of life in the physical mobility and daily life activity domains. We also observed correlations between physical mobility and food, and between communication and emotional status domains in ALS patients.

## Disclosure statement

No author has any financial interest or received any financial benefit from this research.

## Conflict of interest

The authors state no conflict of interest.

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