Journal of Statistics on Health Decision

ORIGINAL ARTICLE

Contribution to the translation and reliability analysis of the Self-Administered Amyotrophic Lateral Sclerosis Functional Rating Scale - Revised, for European Portuguese

Maria da Assunção Matos[1,2], Isabel Costa[1,2], Raquel Portinha[3], Beatriz Afonso[4], Katia Barbosa[1], Carla Gouveia[5], Pedro Sá-Couto[6]

[1] School of Health Sciences (ESSUA), University of Aveiro, Aveiro, Portugal
[2] Center for Health Technology and Services Research (CINTESIS@RISE), University of Aveiro, Aveiro, Portugal.
[3] Multiclínica, Moimenta da Beira, Portugal;
[4] Agrupamento de Escolas de Nelas, Nelas, Portugal;
[5] Centro Hospitalar Tondela-Viseu, E.P.E., Viseu, Portugal;
[6] Center for Research and Development in Mathematics and Applications (CIDMA), Department of Mathematics, University of Aveiro (DMAT), Aveiro, Portugal.

ABSTRACT

Objective: To contribute to the translation of the self-administered ALSFRS-R - European Portuguese version (ALSFRS-R: EP), and to analyse its reliability to enable its use by ALS patients in Portugal.

Methods: The study was developed in 3 phases. Phase 1: Translation and cultural adaptation of the Self-Administered ALFRS-R; Phase 2: Content validation by study participants; Phase 3: Exploratory factor analysis (EFA), analysis of its internal consistency and test-retest reliability.

Results: According to a panel of experts (N=6), a CVI of 100% were obtained for all Self-Administered ALSFRS-R: EP items. A sample of 18 people with ALS (13 male) fulfilled the test and evaluated its items regarding its clarity, comprehension, difficulty, and relevance, obtaining values varying from 8.6 and 8.9, 8.7 and 8.9, 8.5 and 8.8, and 8.5 and 8.9, respectively. The EFA revealed three factors representing the following domains: (1) bulbar function; (2) fine and gross motor function; and (3) respiratory function. The instrument total score and its subscales presented good internal consistency (Cronbach's α: ranging from 0.72 to 0.92 in the test; from 0.70 to 0.95 in the retest) and good to excellent test-retest reliability (Kendall tau: ranging from 0.58 to 0.99). Considering the scale total score for the test, no statistical differences were observed between females and males nor between bulbar ALS and medullar ALS. The correlation between the total score and age showed to be significant and negative (-0.53).

Conclusions: The Self-Administered ALSFRS-R: EP version was successfully translated, validated, and presented good to excellent reliability results, with similar EFA structure to other studies. It will allow its use by European Portuguese ALS patients, enabling their health professionals to monitor the disease progression at home.

Keywords:
Amyotrophic Lateral Sclerosis; Self-administered ALSFRS-R; Functionality; Quality of life.

Corresponding author:
Maria da Assunção Coelho de Matos,
School of Health Sciences (ESSUA), Edifício 30, Agras do Crasto - Campus Universitário de Santiago, 3810-193 Aveiro, Portugal
maria.matos@ua.pt

Support statement:
PSC work was partially supported by National Funds through FCT - Fundação para a Ciência e a Tecnologia), through CIDMA - Center for Research and Development in Mathematics and Applications, within project UIDB/04106/2020 and UIDP/04106/2020.

Conflict of interest:
The authors certify that they have no affiliations with or involvement in any organization or entity with any financial interest or non-financial interest in the subject matter or materials discussed in this manuscript.

Received date: 04.MAY.2022
Accepted date: 15.APR.2023
First published: 28.APR.2023

Introduction

Amyotrophic Lateral Sclerosis (ALS), also known as motor neurone disease or Lou Gehrig's disease, is a rare, fatal, progressive neurodegenerative disease. The main signs and symptoms presented by each patient depend on whether the superior (SMN) or the inferior motor neurons (IMN) are involved as well as the areas innervated by the bulbar nerve fibbers. In addition to motor impairment, cognitive, behavioural, and emotional changes are also reported [1, 2, 3].

Despite all these changes, ALS patients keep their sensory, intestinal, urinary, and sexual functions intact, as well as their awareness, realizing the progression of the disease [4].

ALS is more frequent in males and in ages between 50 and 75 years [5]. The lifetime risk of developing ALS is estimated at 1:350 for men and 1:500 for women [6]. However, according to Pimentel and Ferro [7] it can also be present in young people between 20 and 30 years of age. The incidence of ALS is 2:3 individuals per 100 000 inhabitants in the European population and the prevalence is 3:5 individuals per 100 000 inhabitants, per year [4, 5].

Currently, the diagnosis of ALS is made based on clinical aspects (e.g., history and progression of the disease) and on evidence of impairment of the IMN and SMN, detected through tests such as electromyography, and based on the exclusion of other diseases [8, 2, 3, 9].

Since ALS is a fatal disease with no cure so far [10], the intervention involves the multidisciplinary monitoring of the functional status of the individual, since the diagnosis is confirmed [11, 12]. This multidisciplinary approach has been shown to prolong survival and improve quality of life of ALS patients [6].

The revised Amyotrophic Lateral Sclerosis Functional Rating Scale (ALSFRS-R) was created several years after the ALSFRS [13] due to the acknowledgment that respiratory function (and its deterioration) is a critical prognostic factor. Thus, the original 10-item scale was expanded to the current 12-items. The ALSFRS and its revised version are the most widely applied rating scales for measuring both severity and disease progress overtime in ALS patients and is one of the most frequently used scales in the neurological and functional assessment of these patients [14, 15, 16]. The ALSFRS-R was shown to be a more reliable predictor of prognosis compared to the original ALSFRS [14].

In order to reduce the burden on patients with ALS derived from frequent visits to the trial centres as well as for neurologists to benefit from the ability to monitor global function in patients at home, Montes et al [17] developed the Self-Administered version of the ALSFRS-R. The Self-Administered ALSFRS-R showed excellent reliability (intraclass correlation = 0.93, 95% CI: 0.88 to 0.96) and similar sensitivity to change over time as the standard evaluator-administered ALSFRS-R [17]. The aim of this study is to translate and validate to European Portuguese (EP) language the Self-Administered ALSFRS-R [17] and to analyse its reliability, to enable its use by European Portuguese ALS patients.

Materials and Methods

Evaluation tools

The Self-Administered ALSFRS-R: EP version was used (phase 1 of the study). This is a self-administered questionnaire with 12 items, organized in 4 dimensions: bulbar (Items 1, 2 e 3), fine motor (items 4, 5 e 6), gross motor (items 7, 8 e 9) and respiratory function (items 10, 11 e 12). Each of the items can be classified in a Likert scale with a score of 0 to 4, thus allowing obtaining a final total score, resulting from the sum of all the obtained items, quote that enables the quantification of the participants' level of functioning. It can be completed either by the patient or caregiver.

Ethical procedures

The author of the original version of the scale was contacted and a formal authorization to use it in this study was obtained. The study was approved by the ethical committee of the Centro Hospitalar Tondela - Viseu, Portugal, where the study was developed. All participants signed an informed consent form. After being selected, participants were randomly numbered to keep their anonymity.

Phases of the study/Statistical analyses

Phase 1: Translation and cultural adaptation of the Self-Administered ALSFRS-R

Translation of the Self-Administered ALSFRS-R into EP was carried out using the method suggested by Beaton et al. [18]. Two independent bilingual translators (one Speech and Language Therapist (SLT) and a professional translator) obtained the first two EP versions of the English scale (versions 1 and 2). The translations were reconciled into a single translation (version 3) by the authors. This reconciled version was then back translated into English by a third independent bilingual translator.

A panel of experts (N=6) constituted by 1 SLT, 1 Physiotherapist (PT), 3 Occupational Therapists (OT), and a Physician (P) with professional experience in ALS field evaluated the final translation (version 3). The following parameters were analysed: equivalence in word meaning; equivalence in idioms and colloquial expressions; equivalence in the target cultural context; equivalence between the concept and the experiences of the target culture. Comments and modification suggestions was possible. After incorporation, a final version of ALSFRS-R: EP (version 4) and a second evaluation by the same experts was obtained.

The content validity index (CVI), which "measures the proportion or percentage of experts agreeing on certain aspects of the instrument and its items" [19] was calculated. Once the panel consists of six or more experts, the CVI average should not be less than 78% [18]. This method employs a Likert scale with a score of 1 to 4. To assess relevance/representativeness, the answers were 1 = "no equivalence", 2 = "poor equivalence", 3 = "quite equivalence", 4 = "much equivalence" [19]. This index is calculated by the proportions of a sum of agreement of the items that were scored with "3" or "4" by the experts. Items scored with "1" or "2" should be reviewed or deleted.

Phase 2: Content validation by study participants

A group of patients with ALS were invited to participate. Inclusion factors were defined (being diagnosed with ALS, doing their physical therapy, occupational therapy and/or speech therapy treatments in

the Rehabilitation Department of a central hospital in Portugal - Centro Hospitalar Tondela -Viseu). The patients who were involved (N=18) fulfilled the Self-Administered ALSFRS-R: EP final version (version 4) as well as a questionnaire organized in order to evaluate all the items according to the following parameters: clarity, comprehension, difficulty and relevance. A visual scale with possible values between 0.0 and 9.0 was used. Making suggestions of modification was also a possibility.

Phase 3: Exploratory factor analysis, internal consistency and test-retest reliability

The exploratory factor analysis (EFA) was applied to determine the number of the dimensions that underlie the scale, by indicating the number of factors within a set of items and to determine which items are linked to which factor. The EFA was employed using principal components method with a Varimax rotation. Kaiser-Meyer-Olkin (KMO) and Bartlett's Test of Sphericity were considered to determine the adequacy of the sample adequacy. The number of factors to extract was established based on the scree plot and with eigenvalues greater than 1. The criterion considered to identify the number of items per factor was having loadings superior than 0.4. [19]

The internal consistency of the scale was analysed through the Cronbach's alpha value. Values between 0.7 and 0.8 are considered acceptable, between 0.8 and 0.9 classified as good, and superior to 0.9 are considered excellent [20].

The questionnaire was first applied (test) and after a period of approximately one week, it was re-applied (retest) to the same group of patients to study its test-retest reliability. The test-retest reliability was measured using the Kendall tau correlation coefficient to consider the concordant and discordant pairs for each item and for the total score. To test the temporal stability of the results between test-retest, a paired t-test was conducted. Identical conclusions were obtained with the non-parametric Wilcoxon sign rank test (not presented).

Statistical analysis

All collected data were analysed using the Statistical Package for the Social Sciences (SPSS)-version 24. Descriptive statistics are presented as mean (M) and standard deviation (SD) or in percentage (%) depending on the nature of study variable. The Chi-square test or Fisher Exact test was used to test associations between qualitative variables; the Mann-Whitney test in order to determine differences between independent groups, and the Spearman Rank test for the correlation analysis between quantitative variables. The significance value used was 5%. A simple linear regression analysis was conducted between the variables scale total score, age and TSD. The Normal distribution of the residuals was checked by visual inspection of the PP plot.

Results

Phase 1: Translation and cultural adaptation of the Self-Administered ALFRS-R

The panel of experts (N=6) evaluated the Self-Administered ALSFRS-R: EP version in two different moments. After the first evaluation, all the items are properly validated, with CVI values of 100%, except for the item 1, which has a value of 83.3% (see table 1).

The experts, however, have provided some suggestions for improving the EP version content making it more easily understood by the target population. In the first question, the word "discourse" was replaced by "speaking"; in the third question, "swallowing" was replaced by "deglutition" and "suffocation" replaced by "choking"; in the fourth question, "cuff or brace" was replaced by "assistive products"; and in all questions where the term "feed tube" arose, it was replaced by "nasogastric tube or PEG". The heading was also placed on all pages of the questionnaire, as well as the phrase "Compared to the time before ALS symptoms". The scale was formulated in the third person as it can be completed not just by ALS patients but also by their caregivers. A final evaluation was performed and a CVI of 100% were obtained for all items (version 4).

Phase 2: Content validation by study participants

Sample characterization

The sample population comprised 18 subjects, 13 (72.2%) males and 5 (27.8%) females (Table 2) with an age range from 45 to 77 years. All female patients suffer from bulbar ALS. Three male patients suffer from bulbar ALS and 10 from medullar ALS. Time passed from the symptom onset to diagnosis (TSD) range from 0 to 5 years. A statistical association between ALS and gender was found (p=0.001). No statistical mean differences were observed in age and TSD between females and males (p>0.05).

Table 1 - Results of Experts panel first evaluation of EP (version 3)

Item	SLT1	PT1	OT1	OT2	OT3	PH1	CVI (%)
1. Speech	3	4	4	4	4	2	83.3
2. Salivation	3	4	4	4	4	3	100
3. Swallowing	3	3	3	4	4	4	100
4. Handwriting	4	4	3	4	4	4	100
5A. Cutting food and handling utensils (patients without gastrostomy)	3	4	4	4	4	3	100
5B. Cutting food and handling utensils (patients with gastrostomy)	3	4	4	4	4	3	100
6. Dressing and hygiene	4	4	4	4	4	3	100
7. Turning in bed and adjusting bed clothes	4	4	4	4	4	4	100
8. Walking	4	4	3	4	4	3	100
9. Climbing stairs	4	4	4	4	4	4	100
10. Dyspnoea	4	3	4	4	4	4	100
11. Orthopnoea	4	4	4	4	4	4	100
12. Respiratory insufficiency	4	4	4	4	4	4	100

SLT: Speech language therapist; PT: Physical therapist; OT: Occupational therapist; P: Physician

Table 2 - Sociodemographic data of the participants

	Total N=18	Gender		Statistical results
		Female N=5 (27.8%)	Male N=13 (72.2%)	
ALS (N (%))				
Bulbar	8 (44.5)	5 (62.5)	3 (37.5)	Fisher=0.0065; p=0.007
Medullar	10 (55.5)	0 (0)	10 (100)	
Age (years, M ± SD)	65.6 ± 8.4	61 ± 5.3	67 ± 9.0	U=16.0; p=0.103
TSD (years, M ± SD)	1.5 ± 1.2	1.2 ± 0.8	1.6 ± 1.3	U=27.0; p=0.562

ALS: Amyotrophic lateral sclerosis: TSD: Time from symptom onset to diagnosis

Self- Administered ALSFRS-R: EP version evaluation

All items of the EP version (version 4) were evaluated from each participant's perspective regarding the following parameters: clarity, comprehension, difficulty, and relevance (Table 3). Overall, it can be observed that the average of the scores attributed to the items is between 8.6 and 8.9 in relation to the clarity parameter, between 8.7 and 8.9 for comprehension, between 8.5 and 8.8 in difficulty and between 8.5 and 8.9 in the relevance parameter. In addition, standard deviation values are relatively low on all items.

Table 3 - Evaluation of the items of the Self-Administered ALSFRS-R: EP version (version 4) for the domains: clarity, comprehension, difficulty, and relevance.

Item	Clarity		Comprehension		Dificulty		Relevance	
	M	SD	M	SD	M	SD	M	SD
1. Speech	8.7	0.5	8.9	0.2	8.8	0.2	8.9	0.2
2. Salivation	8.8	0.3	8.8	0.5	8.7	0.7	8.8	0.4
3. Swallowing	8.8	0.3	8.8	0.4	8.8	0.3	8.8	0.4
4. Handwriting	8.9	0.3	8.8	0.4	8.8	0.4	8.8	0.3
5. Cutting food and handling utensils	8.6	1.1	8.8	0.4	8.5	1.1	8.8	0.4
6. Dressing and hygiene	8.7	0.5	8.8	0.4	8.5	1.1	8.8	0.3
7. Turning in bed and adjusting bed clothes	8.8	0.3	8.8	0.4	8.8	0.4	8.8	0.4
8. Walking	8.8	0.4	8.8	0.4	8.8	0.4	8.8	0.3
9. Climbing stairs	8.7	0.6	8.7	0.4	8.8	0.4	8.5	0.9
10. Dyspnoea	8.8	0.3	8.7	0.4	8.8	0.4	8.8	0.4
11. Orthopnoea	8.6	1.1	8.8	0.3	8.8	0.3	8.8	0.3
12. Respiratory insufficiency	8.8	0.4	8.8	0.4	8.8	0.4	8.8	

Phase 3: Exploratory factor analysis, internal consistency and test -retest reliability

The EFA results presents a 3-factor structure for the test and retest, confirmed by the scree plot (results not presented) and by the eigenvalues greater than 1 (Table 4). For both situations, the Bartlett test is significant, the total variance explained are 77.92% and 78.20%, respectively, but the KMO for the test is somewhat lower than 0.5. The distribution of the items throughout the factors is also similar for both cases. The Cronbach's α coefficient values show high internal consistency values for the achieved solution. The factor 1 corresponds to the dimensions of fine and gross motor together; factor 2 corresponds to the respir-

atory dimension and factor 3 corresponds to the bulbar function dimension. However, the original Self-Administered ALSFRS-R scale presents a 4-factor structure, having separate factors for the fine motor and for the gross motor (Table 5). Nevertheless, the distribution of the items throughout the factors are identical in both cases (see Tables 4 and 5).

Table 4 - Exploratory factor analysis for the Self-Administered ALSFRS-R: EP version (version 4)

	Test (KMO=0.469; $\chi^2(66)=197.9$; p<0.001)			Retest (KMO=0.601; $\chi^2(66)=182.7$; p<0.001)		
	Factor 1	**Factor 2**	**Factor 3**	**Factor 1**	**Factor 2**	**Factor 3**
1. Speech			0.880			0.890
2. Salivation			0.803			0.755
3. Swallowing			0.666			0.672
4. Handwriting	0.898			0.946		
5. Cutting food and handling utensils	0.777			0.741		
6. Dressing and hygiene	0.962			0.953		
7. Turning in bed and adjusting bed clothes	0.834			0.940		
8. Walking	0.907			0.927		
9. Climbing stairs	0.793			0.839		
10. Dyspnoea		0.600			0.547	0.451
11. Orthopnoea		0.859			0.880	
12. Respiratory insufficiency		0.875			0.916	
Eigenvalues	5.614	2.350	1.387	6.625	1.139	2.620
Variance explained (%)	46.78	19.58	11.55	46.87	9.49	21.83
Cronbach's α	0.948	0.754	0.720	0.957	0.753	0.706

From Table 5, it is possible to verify that the obtained Cronbach's α coefficient values are similar for the test and retest situations, varying between 0.72 and 0.92 in the test and between 0.70 and 0.95 in the retest. The Cronbach's α values are classified as good or excellent, with the exception of subscale bulbar function (only acceptable). The reliability results presented by the Kendall tau are very high except for item 2 (salivation), which presents a moderate correlation. All the results are significant. Finally, no statistical differences were noticed between the test and retest situations, showing temporal stability between the two measures. Of notice, the total score presented almost excellent internal consistency and a very high correlation measure.

Table 5 - Internal consistency and test-retest reliability results (n=17) for Self-Administered ALSFRS-R: EP version (version 4)

Subscale			**Statistical results**	
Nr. Item	**Test**	**Retest**	**Difference** (paired t-test)	**Correlation** (Kendall tau)
Bulbar function (Cronbach's α)		0.70		
1. Speech (M±SD)	1.9±1.3	1.9±1.3	t(17)= 0.6;p=0.579	0.882*
2. Salivation (M±SD)	2.5±1.4	2.4±1.5	t(17)= 0.4;p= 0.707	0.575*
3. Swallowing (M±SD)	2.8±1.2	2.8±1.3	t(17)= 1.0;p=0.331	0.990*
Fine motor (Cronbach's α)	0.90	0.91		
4. Handwriting (M±SD)	2.2±1.4	2.1±1.4	t(17)= 1.4;p=0.187	0.881*
5. Cutting food and handling utensils (M±SD)	1.7±1.6	1.7±1.4	t(17)= 0.0;p=1.000	0.857*
6. Dressing and hygiene (M±SD)	1.5±1.7	1.6±1.7	t(17)=-1.5;p=0.163	0.948*
Gross motor (Cronbach's α)	0.92	0.95		
7. Turning in bed and adjusting bed clothes (M±SD)	1.9±1.7	2.1±1.6	t(17)=-0.9;p=0.381	0.844*
8. Walking (M±SD)	2.0±1.8	1.9±1.8	t(17)= 0.6; p=0.579	0.908*
9. Climbing stairs (M±SD)	1.6±1.7	1.4±1.7	t(17)= 1.0; p=0.331	0.942*
Respiratory function (Cronbach's α)	0.75	0.74		
10. Dyspnoea (M±SD)	2.6±1.7	2.7±1.6	t(17)=-0.8;p=0.430	0.824*
11. Orthopnoea (M±SD)	1.7±1.9	1.4±1.9	t(17)= 1.4; p=0.172	0.929*
12. Respiratory insufficiency (M±SD)	2.3±1.2	2.6±1.1	t(17)=-1.5;p=0.163	0.901*
Total score (Cronbach's α)	0.88	0.87		
Total score (M±SD)	24.7±12.4	24.5±11.9	t(17)=-0.7;p=0.508	0.902*

*p<0.001

Considering the scale total score for the test, no statistical differences were observed between females and males (33.0±15.5 vs 21.5±9.9, U=16.5, p=0.114) nor between bulbar ALS and medullar ALS (30.8±12.7 vs 19.9±10.3, U=21.5, p=0.100). The correlation between the total score and age showed to be significant and negative (r=-0.53, p=0.024; Total Score=73.91(p=0.03)-0.75(p=0.031)*Age, R^2=0.26, see figure 1) but not with time from symptom onset to diagnosis (TSD, r=0.16, p=0.536; Total Score=22.62 (p<0.001)+1.40 (p=0.607)*TSD, R^2=0.02). Similar results were observed for the retest situation.

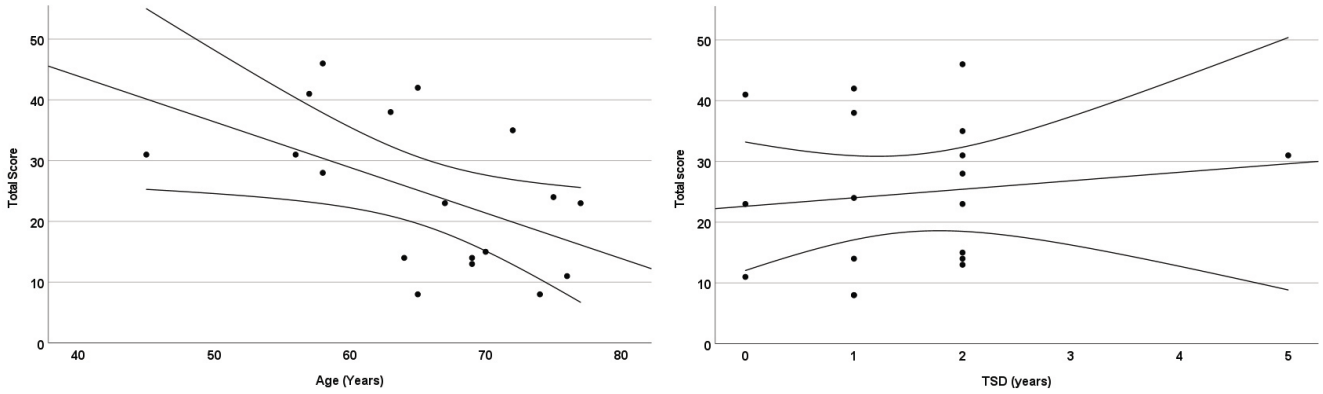

Figure 1 - Scatterplot results for the ALSFRS-R self-questionnaire, EP version (version 4) total score with age and TSD, respectively. The linear regression and the correspondent 95% CI are also presented.

Discussion

The results obtained show that the Self-Administered ALSFRS-R: EP version was successfully translated and validated. Despite the small sample size, the results from EFA, internal consistency and test-retest reliability were sufficiently strong to ensure good psychometric capabilities of the translated scale. Although, in a first moment, some reformulations were suggested by the experts', the final items of the scale were reassessed as being equivalent to the original form, with a CVI of 100% for all its items, guaranteeing its cross-cultural adaptation [17, 20].

Concerning to its content validation by a group of ALS patients (N=18), the participants state that its items are explicit, clear and, therefore, easy to understand and answer. They also have considered that the items are relevant to assess their functioning as well as the disease impact on their quality of life. As the standard deviation values obtained are relatively low in all items, it can then be concluded that the participants are generally satisfied with the scale.

The study sample is composed mainly of male individuals (n=13), with an average age of 67 years. This is similar with the existing literature that claims that ALS appears more frequently in men, in the age groups between 50 and 75 years [4, 5]. According to some authors, this fact can be attributed to female hormones and their neuroprotective effect, which makes men more susceptible to this disease [22]. Most male participants of our study present a medullar ALS, whereas bulbar ALS predominates in the female group, which agrees with what was reported by Wijesejera & Leigh [22] in their study.

It was also observed that the time interval between the onset of the first symptoms and the diagnosis is, on average, 1 year and 5 months. These findings corroborate again the existing literature that indicates that most times there is a period between 13 to 18 months among the onset of symptoms and diagnostic confirmation. This can be because most symptoms appear gradually, but it can also be derived from the fact that establishment of initial diagnosis are often erroneous as a result of the lack of a specific exam for a diagnosis of ALS [23, 6].

From the EFA results, a 3-factor solution was achieved compared to a 4-factor solution of the original scale. The only difference is that the 3-factor solution combines the fine motor factor with gross motor factor. This 3-factor solution with the same combination of factors and items is also suggested in other works, such as Franchignoni et al. [24] for the Italian version e Ohasi et al. [25] for the Japanese version, while a 4-factor solution is presented by Maksymowicz et al. [26] for the Polish version.

From the application of the test and the retest, it was possible to verify that most of the participants revealed to present alterations essentially in terms of personal care, in the ability to climb stairs and, related to respiratory issues. Consequently, most of the individuals in the study are dependent on their caregiver, unable to climb stairs and experience shortness of breath or difficulty breathing when lying on their backs, therefore needing to use BIPAP (bilevel positive airway pressure) continuously at night. In addition, it was possible to verify that the values of Cronbach's alpha for the same domains are identical, which indicates that their questions are consistent, and that people answered them consciously. As the Cronbach's alpha values ranged between 0.77 and 0.91 in all subscales, we can conclude that the scale has a high internal consistency [23]. The total test-retest reliability result was similar (0.902) to the original study (0.87) [17]. Despite the differences encountered in some items among the two evaluation moments, these were not significant, given that the p-value values are greater than α (0.05) in all of them. Through Kendall tau test, it was also observed that all correlation values are greater than 0.82, except for item 2, which shows a lower but significant correlation (0.575). It is possible to conclude that there is a strong reliability between the test and retest moments.

As conclusion, we can affirm that, although we have used a small sample, the Self-Administered ALS-FRS-R: EP version presents good to excellent reliability values, allowing its use by European Portuguese ALS patients and enabling their health professionals to monitor the disease progression at home. In future studies it would be important to apply it to a larger sample, involving several geographical regions of our country. It also would be interesting to carry out a comparison between the data obtained in the completion of the self-questionnaire by the patient or caregiver and the results of completing the ALSFRS-R, applied by the health professional.

Acknowledgments

We would like to thank all the ALS patients that have participated in our study. Without them, it would not be possible.

References

1. Luchesi KF, Silveira IC. Palliative care, amyotrophic sclerosis,and swallowing: a case study. CoDAS. 2018;30(5):1-6. https://doi.org/10.1590/2317-1782/20182017215
2. Chieia M, Oliveira A, Silva H, Gabbai A. Amyotrophic lateral sclerosis: considerations on diagnostic criteria. Arq Neuropsiquiatr. 2011;68(6):837-42. https://doi.org/10.1590/S0004-282X2010000600002
3. Hardiman O, Van Den Berg LH, Kiernan MC. Clinical diagnosis and management of amyotrophic lateral sclerosis. Nat Rev Neurol. 2011; 7(11):639-49. doi: 10.1038/nrneurol.2011.153. https://doi.org/10.1038/nrneurol.2011.153
4. Morim L, Rocha J, Almeida A. Intervenção da Terapêutica da fala na Esclerose Lateral Amiotrófica (ELA). Cad Comun e Ling. 2009;1:119-30.
5. Couratier P, Corcia P, Lautrette G, Nicol M, Preux P-M, Marin B. Epidemiology of Amyotrophic Lateral Sclerosis: A review of literature. Revue neurologique. 2015;2:119-32.
6. Salameh JS, Brown RH Jr, Berry JD. Amyotrophic Lateral Sclerosis: Review. Semin Neurol. 2015; 35(4):469-76. doi: 10.1055/s-0035-1558984. https://doi.org/10.1055/s-0035-1558984
7. Pimentel J, Ferro J. Neurologia - Princípios, diagnóstico e tratamento. LIDEL. 2006.
8. Garcia L, Silva A, Carrete Jr H, Favero F, Fontes S, Moneiro M, et al. Relação entre degeneração do trato córtico-espinhal através de ressonância magnética e escala funcional (ALSFRS) em pacientes com esclerose lateral amiotrófica. Arq Neuropsiquiatr. 2007;65(3-B):869-874. https://doi.org/10.1590/S0004-282X2007000500029
9. Van-Es M, Hardiman O, Chio A, Al-Chalabi A, Pasterkamp R, Veldink J, et al. Amyotrophic lateral sclerosis. Lancet. 2017;390(10107):2084-2098. doi: 10.1016/S0140-6736 (17)31287-4. https://doi.org/10.1016/S0140-6736(17)31287-4
10. Matthew C Kiernan, Steve Vucic, Benjamin C Cheah, Martin R Turner, Andrew Eisen, Orla Hardiman, James R Burrell, Margaret C Zoing, Amyotrophic lateral sclerosis, The Lancet. 2011; 377 (9769): 942-955. https://doi.org/10.1016/S0140-6736(10)61156-7
11. Tabor L, Gaziano J, Watts S, Robison R, Plowman EK. Defining Swallowing-Related Quality of Life Profiles in Individuals with Amyotrophic Lateral Sclerosis. Dysphagia. 2016;31(3):376-82. https://doi.org/10.1007/s00455-015-9686-2
12. Silva L, Mourão L, Silva A, Lima N, Almeida S, Jr M, et al. Amyotrophic Lateral Sclerosis: Combined nutritional, respiratory and functional assessment. Arq Neuropsiquiatr. 2008;66(April):354-9. https://doi.org/10.1590/S0004-282X2008000300014
13. Cedarbaum, JM, Stambler, N. Performance of the amyotrophic lateral sclerosis functional rating scale (ALSFRS) in multicenter clinical trials, J. Neurol. Sci. 1997; 152 (19s1-s9, doi:10.1016/S0022-510X(97)00237-2. https://doi.org/10.1016/S0022-510X(97)00237-2
14. Cedarbaum JM, Stambler N, Maltab E, Fullerb C, Hiltb D, Thurmond B, Nakanishi A, BDNF ALS Study Group (Phase III). The ALSFRS-R: a revised ALS functional rating scale that incorporates assessments of respiratory function. Journal of the Neurological Sciences. 1999; 169:13 - 21. https://doi.org/10.1016/S0022-510X(99)00210-5
15. Campos TS, Rodrigez-Santos F, Esteban J, Vazquea PC, Pardina JSM, Carmona AC. Spanish adaptation of the revised Amyotrophic Lateral Sclerosis Functional Rating Scale (ALSFRS-R). Amyotrophic Lateral Sclerosis.2010; 11: 475-477. https://doi.org/10.3109/17482968.2010.489115
16. Bakker LA, Schröder CD, Tan HHG, et al Development and assessment of the inter-rater and intra-rater reproducibility of a self-administration version of the ALSFRS-R Journal of Neurology, Neurosurgery & Psychiatry 2020;91:75-81. https://doi.org/10.1136/jnnp-2019-321138
17. Montes J, Levy G, Albert S, Kaufmann P, Buchsbaum R, Gordon PH, Mitsumoto H. Development and evaluation of a self-administered version of the ALSFRS-R. Neurology.2006; 67:1294-1296. https://doi.org/10.1212/01.wnl.0000238505.22066.fc
18. Beaton, D. E., Bombardier, C., Guillemin, F., & Ferraz, M. B. (2000). Guidelines for the Process of Cross-Cultural Adaptation of Self-Report Measures. Spine, 25(24), 3186-3191. https://doi.org/10.1097/00007632-200012150-00014
19. Hair, J. F., Black, W. C., Babin, B. J., & Anderson, R. E. (2019). Multivariate data analysis (8th ed.). Cengage
20. Alexandre, N. M. C., & Coluci, M. Z. O. Validade de conteúdo nos processos de construção e adaptação de instrumentos de medidas. Ciência e Saúde Coletiva. 2011. 16(7); 3061-3068. https://doi.org/10.1590/S1413-81232011000800006
21. Cohen J. A coefficient of agreement for nominal scales. Educ Psychol Meas. 1960; 20:37-46. https://doi.org/10.1177/001316446002000104
22. Wijesekera L, Leigh N. Amyotrophic lateral sclerosis. Orphanet J Rare Dis. 2009; 22:1-22. https://doi.org/10.1186/1750-1172-4-3
23. Xerez D. Reabilitação na Esclerose Lateral Amiotrofica: revisão da literatura. Acta Fisiátrica. 2008;15(3):182-8. https://doi.org/10.11606/issn.2317-0190.v15i3a102947
24. Ranchignoni F, Mora G, Giordano A, et al Evidence of multidimensionality in the ALSFRS-R Scale: a critical appraisal on its measurement properties using Rasch analysis. Journal of Neurology, Neurosurgery & Psychiatry 2013; 84:1340-1345. https://doi.org/10.1136/jnnp-2012-304701
25. Ohashi Y, Tashiro K, Itoyama Y, Nakano I, Sobue G, Nakamura S, Sumino S, Yanagisawa N. [Study of functional rating scale for amyotrophic lateral sclerosis: revised ALSFRS(ALSFRS-R) Japanese version]. No To Shinkei. 2001 Apr;53(4):346-55. Japanese.

26. Maksymowicz, S., Kukołowicz, P., Siwek, T. et al. Validation of the revised Amyotrophic Lateral Sclerosis Functional Rating Scale in Poland and its reliability in conditions of the medical experiment. Neurol Sci. 2021: 42, 943-949. https://doi.org/10.1007/s10072-020-04565-5

27. 27. Garcia-Marques T, Marôco J. Qual a fiabilidade do alfa de Cronbach? Questões antigas e soluções modernas? Laboratório Psicol. 2006;4(1):65-90.

28. Souza A, Alexandre N, Guirardello E. Propriedades psicométricas na avaliação de instrumentos: avaliação da confiabilidade e da validade. EpidemiolServSaude. 2017;26(3):649-59. https://doi.org/10.5123/S1679-49742017000300022