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Original articles

Dysarthria and quality of life in patients with amyotrophic lateral sclerosis

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ABSTRACT

Purpose: to analyze the impact of dysarthria on the quality of life in patients with amyotrophic lateral sclerosis.

Methods: the study consisted of 32 subjects, divided into two groups (control group and study group) who underwent an initial interview for background information, followed by an evaluation based on the Dysarthria Assessment Protocol and completion of quality of life questionnaire "Living with Dysarthria - (LwD)". Exploratory data analysis was collected through mean, median, SD, minimum and maximum measures. A comparison was performed between the studied groups and a correlation was carried out between scores. The significance level adopted was 5%.

Results: according to the findings, all sub-items analyzed by the dysarthria assessment protocol were statically significant ($p < 0.001$) when comparing the groups. Regarding quality of life, a moderate positive correlation ($p = 0.0008$; Spearman's coefficient $= 0.75202$) was observed between the total score of the two protocols used, indicating that the higher the degree of dysarthria, the worse the Quality of Life (QOL) of the subject, according to the parameters evaluated.

Conclusion: dysarthria affects all speech parameters herein assessed, in varying degrees, negatively impacting communication and quality of life.

Keywords: Amyotrophic Lateral Sclerosis; Dysarthria; Quality of Life; Voice; Rehabilitation of Speech and Language Disorders

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INTRODUCTION

Amyotrophic lateral sclerosis (ALS) belongs to the group of motor neuron disease and is characterized by the degenerative progression of upper motor (UMN) and lower motor neurons (LMN), usually associated with bulbar and pyramidal involvement [1].

ALS is classified as sporadic (90 to 95% of the diagnosed cases), of unknown etiology, and familial. It is believed that there are several factors that may contribute to the onset of the disease, such as excito-toxicity by neurotransmitter glutamate, accumulation of neurofilaments, deficiency Neurotrophic factors, immunity changes, physical trauma, persistent viral infections, as well as chemical and physical environmental factors. Familial onset of ALS occurs in 5 to 10% of the occurrences, with an autosomal dominant inheritance pattern, mutations of enzymes or mutations of chromosomes that contribute to the onset of the disease, such as the mutation in the gene encoding the enzyme superoxide Dismutase 1 (SOD1) [2]. The most frequent clinical sign is muscle weakness, which occurs in approximately 60% of the patients. It affects the upper limbs, lower limbs, and, at some point in the disease, speech, swallowing, chewing and breathing [3].

The dysarthria which characterizes the disease, presents as slow, weak, and imprecise articulation. The disease also presents with incoordination of the stomatognathic system, compromising the respiratory, phonatory, resonant and articulatory systems.

With the progress of the disease, communication difficulties increase while sentences become simpler and shorter, culminating, in advanced phases, to answering questions through the use of keywords or "yes / no" answers"[4]. These changes, together with the loss of functional independence caused by ALS, lead to an extremely discouraging situation for the individual [5]. Studies reaffirm that quality of life is largely affected by the presence of dysarthria caused by various underlying diseases [6,7].

The decline in functional capacity coupled with the relatively preserved cognitive capacity observed in subjects with ALS are considered the primary problems to be addressed. As such, these individuals are often unable to take pleasure in living, thus experience a decline in their quality of life [8].

Thus, it is important to understand the negative impact that speech disorders have on subjects with ALS. In turn, this may help improve the therapeutic process and alleviate suffering. All this highlights the uniqueness of this study, in light of the shortage of this topic in the literature, in particular, the absence of a link between the impact of dysarthria to quality of life. In recent years, some studies have focused on the quality of life of subjects with ALS; however, despite the protocols used were related to communication, the aspects specifically related to dysarthria were not discussed in detail as they were in this article.

Considering that the subjects affected with ALS will present with speech alterations as described above, the deterioration of this function will gradually interfere with their communication ability. Communication disability, in its advanced stages, may lead to isolation from social interaction with other people, directly affecting quality of life.

The present study aims to analyze the impact of dysarthria on the quality of life of patients with amyotrophic lateral sclerosis, based on the following questions: Is there a dysarthria that subjects with ALS have in common, that interferes with their quality of life? Is there a direct relationship between the worsening of dysarthria and quality of life? Does the time of diagnosis interfere with the perception of quality of life related to dysarthria in ALS?

METHODS

This is a comparative cross-sectional study, and the procedures performed were approved by the Ethics and Research Committee (CEP) of the State University of Campinas, number CAAE 43795515.1.0000.5404.

Selection of Subjects

To conduct the analysis, two groups were formed, called the study group (SG) and the control group (CG). The aim of the CG was to ensure that the findings of the assessment of quality of life and dysarthria of SG were characteristics of ALS.

The sample consisted of 32 participants, 16 in the SG and 16 in the CG. This sample was collected in twelve-months, during a specialized service in the care of patients with neurological disorders.

Inclusion criteria for the SG were individuals who had previous diagnosis of sporadic ALS and had signed a free informed consent (TCLE). Anarchic subjects were excluded, because they would have not have had the ability to respond adequately to the protocols used, thus making it impossible to analyze some parameters suggested, mainly by the dysarthria protocol.

The CG was matched to the SG according to age and sex, and was made up of participants without a

history/complaint of neurological alterations and/or speech disorders due to some underlying pathology, and subjects undergoing voice or speech therapy were excluded.

Procedures

This study used two instruments for data collection and evaluation: a) The Protocol of Evaluation of Dysarthria [9] and b) The questionnaire on quality of life "Living with Dysarthria" [10].

The data collection took place in a quiet room, without the presence of noise that could have compromised the quality of the recording for later analysis of the data, according to the consensus among the research team. The speech and voice samples were obtained with the following devices: A mobile device (Motorola Moto X 2014 XT1097 32GB) with a 5.2-inch screen with an AMOLED panel of 1920 × 1080 pixels, a competent Snapdragon 801 with a quad-core processor of 2.5 GHz and GPU video card Adreno 330, and a 13 megapixel camera with flash.

Voice parameters were recorded in audio by means of sustained phonation, with the participants seated, with feet comfortably supported on the floor and with the mobile device at a distance of approximately 15 cm from the midline of the mouth. For audio recording purposes, the sampling rate used was 44100 Hz. During video recordings, which were carried out to test the parameters of respiration, resonance, articulation and prosody, the subjects remained in the same position and the distance of the mobile device was manipulated by the researcher to allow visualization of the trunk and the head of the participant, in order to favor the adequate completion of the dysarthria evaluation protocol.

The protocol, as adapted to Portuguese[9], included five parameters: a) respiration, b) phonation, c) resonance, d) articulation and e) prosody, which were evaluated by means of specific tests for further analysis, as follows:

- Respiration: Counting breath cycles (expiration / inspiration) for one minute and at rest; Maximum phonation times with emission of sustained vowels (/ a /; / i /); s/z ratio; Words by expiration through counting (40-1).
- Phonation: Sustained phonation of the vowels /a/ and /i/.
- Resonance: Velar movement by means of the alternating emission of /a/ and /ã/; use of the Altmann millimeter mirror during the emission of words with nasal and oral sounds; and movement of the pharyngeal wall during continuous emission of / ka,ka /.
- Articulation: Lip movement (alternating /i/, /u/, and /pa/); Tongue movement with /ka/ /ta/ emission at increasing speed; Spontaneous description of figures; and monosyllabic reading.
- Prosody: Reading sentences (affirmative, interrogative and exclamatory).

With the exception of the "respiration" parameter that used objective data such as number of respiratory cycles and number of words per expiration, the other parameters utilized auditory-perceptual and visual parameters assessed by the evaluators, who must determine the degree of change from 0 to 6 (Score 0 equals no change and score 6 at the highest possible rate of change). The final score of the protocol was computed by the sum of each section, which provided the degree of dysarthria, so that the final score from 0 to 10 would determine mild dysarthria, from 11 to 20 moderate dysarthria and from 21 to 30 severe dysarthria.

Three speech-language pathologists, with experience in attending patients with neurological disorders were invited to perform the voice and speech analysis according to the adopted protocol. Their role was to judge the parameters presented in the evaluations to ensure the fairness of the trials. Each one of the three judges received individual training by the researchers on how to perform the measurements, emphasizing the subjective nature of the auditory-perceptual and visual evaluation. The researchers prepared a mock test evaluation with the professionals in order to exemplify the evaluation process and guarantee its orientation.

Following the data collection, to assess the quality of life related specifically to dysarthria, the principal investigator applied the self-assessment questionnaire called "Living with Dysarthria" translated and culturally adapted into Brazilian Portuguese [9]. The purpose of this questionnaire was to verify how the participants perceived themselves and their speech difficulties and how they adjusted themselves to different situations. The questionnaire consisted of 50 statements divided into 10 distinct sections and the total score of the questionnaire was performed by summing the score of each statement from all sections. The minimum score was 50 points, which suggested little impact of dysarthria on quality of life and the maximum score was 300

points, which indicated a high impact of dysarthria on the quality of life of the subject.

Data Analysis

Data from the present study were tabulated and statistically analyzed using the software SAS System for Windows (Statistical Analysis System), version 9.4.

Exploratory statistical data were obtained by mean, standard deviation, and minimum, median and maximum for sample characterization of the studied groups (age, sex, time of disease diagnosis) and dependent variables (auditory-perceptual analysis parameters).

From the results obtained in the auditory-perceptual evaluation, an inter-rater reliability analysis was performed, using the Kappa Fleiss Concordance Index. After the analysis, the arithmetic mean of each analyzed parameter was extracted.

A comparison between the groups was performed using the Mann-Whitney, Chi-Square and Fisher's exact tests. The level of significance was 5%.

For the data analysis of the protocol "Living with Dysarthria", each item that presented with "strongly agree" and "totally agree" (5 and 6) were extracted in all the sections present in the protocol. The mean and standard deviation were then obtained for the ten sections of the questionnaire and for the total score.

RESULTS

The sample characteristics were similar between the groups, except for the variable "study time", where it was possible to observe a higher level of education of the CG. As such, the groups were not homogeneous; however the difference of this variable did not interfere with the overall findings of the research (Table 1).

Table 1. Sample characterization according to gender, age, and disease duration (study group)

Variables		Groups		Value of p
		SG (n=16)	CG (n=16)	
Age	Mean	58.8	57.4	0.72*
	Median	62	60	
	Minimum-Maximum	30-79	32-75	
	SD	14.4	12.6	
Sex	Female	9	9	1.00**
	Male	7	7	
School time (years)	Mean	7.4	11.5	0.042*
	Media	4.5	11	
	Minimum-Maximum	3-15	2-23	
	SD	4.5	5.6	
Disease duration (years)	Mean	4.4	-	
	Median	3.5		
	Minimum-Maximum	1-12	-	
	SD	3.8	-	

(*) Mann Whitney Test; (**) Chi-Square test
Legend:
SD - Standard Deviation
SG – Study Group/CG – Control Group

Among the subjects who participated in the research, for each group, nine (56.3%) were female and seven (43.8%) were male.

The mean age for the SG was 58.8 ± 14.4 years. Among the 16 participants in this group, nine were ≥ 60 years of age, while the other seven were between the age range of 30 to 59 years. There was no statistical difference (p = 0.72) between the ages of the participants of the SG and CG, since the latter presented a mean age of 57.4 ± 12.6 years.

In regards to the time of the disease, the mean of the participants was 4.4 ± 3.8 years of diagnosis. A greater variation of diagnosis time was observed with nine individuals presenting between 1 and 5 years, five individuals between 6 and 9 years, and only two with ≥ 10 years.

The inter-rater reliability analysis of the dysarthria evaluation was performed using the Kappa Fleis test and showed a moderate degree of reliability for the parameters respiration, resonance, articulation and prosody, and a weak reliability for phonation.

All dysarthria parameters analyzed showed differences between the groups (p <0.001). The overall score averaged 9.9 ± 4.3 for SG, indicating a slight degree of impairment, however, ranging from mild to moderate degree of dysarthria (2.8-17.6 points) (Table 2).

Table 2. Description of dysarthria in terms of respiration, phonation, resonance, articulation and prosody parameters of the study group and the control group

Variables		Groups		
		SG (n=16)	CG (n=16)	Value of p*
Respiration	Mean	3.6	0.9	<0.001
	Median	3.7	1	
	Min-Max	1.0-6.0	0.0-3.0	
	Standard Deviation	1.3	0.8	
Phonation	Mean	4.1	1.4	<0.001
	Median	4.2	1.2	
	Min-Max	2.0-5.7	0.3-4.7	
	Standard Deviation	1	1.1	
Resonance	Mean	3	0.1	<0.001
	Median	3	0	
	Min-Max	0.3-6.0	0.0-0.7	
	Standard Deviation	2.2	0.2	
Articulation	Mean	3.3	0.1	<0.001
	Median	3.7	0	
	Min-Max	0.3-6.0	0.0-1.3	
	Standard Deviation	2	0.3	
Prosody	Mean	2.5	0.3	<0.001
	Median	2.3	0	
	Min-Max	0.3-0.6	0.0-1.3	
	Standard Deviation	1.9	0.4	
Total Score	Mean	9.9	1.7	<0.001
	Median	10.2	1.2	
	Min-Max	2.8-17.6	0.4-5.8	
	Standard Deviation	4.3	1.4	

(*) Mann Whitney Test
Legend:
Min- Minimum/Max- Maximum
SG – Study Group/CG – Control Group

In the quality of life analysis related to dysarthria, four out of ten sections of the questionnaire presented with a mean percentage equal to or greater than 50% of individuals who indicated affirmatively "mostly true" and "definitely true" (5-6) on questions such as: "How do you perceive changes and the possibility of changing your way of speaking?" (76.8%). Participants reported being aware of the possibility of worsening in their speech and reported creating strategies when they were not understood, including to rest a little during conversation and even avoid talking. "Communication problems related mainly to speech" (65%), in which subjects with ALS perceived themselves with slower and more drawn speech, having to repeat the words for others to understand. "My communication difficulties hamper my chances of ..." (57.5%), in which the difficulties presented restricted their social life. Finally "Effects on emotion" (55%), showing that the difficulties caused by the disease produced an effect on their self-image.

Section 10 had the highest score obtained, with an average of 4.8 ± 1.8, followed by section 1 with a mean of 4.7 ± 1.6, and section 7 with a mean of 4.4 ± 1.8 (Table 3).

Table 3. Comparison of means and standard deviation of the ten sections and total score of the "living with dysarthria" protocol

	SG	CG
1. Communication problems related primarily to speech	4.7±1.6	1.7±1.3
2. Communication problems related primarily to language/cognition	2.3±1.8	2.2±1.6
3. Communication problems related primarily to fatigue	1.8±2.1	1.8±1.3
4. Effects of emotions	3.9±2.2	1.9±1.5
5. Effects of different persons	3.9±0.8	1.3±0.8
6. Effects of different situations	3.9±2.0	1.5±1.1
7. My difficulties in communicating affect my possibilities to …	4.4±1.8	1.3±0.9
8. What do you think contributes to the changes in the way you communicate?	3.6±2.2	1.7±1.4
9. How altered is my communication?	2.9±2.0	1.4±1.0
10. How do you perceive changes and the possibility to alter your way of speaking?	4.8±1.8	1.6±1.3
Total Score	189±49.6	81.2±30.3

Legend:
SG – Study Group/CG – Control Group

The Spearman coefficient was used to investigate the correlation between dysarthria and time of diagnosis with quality of life (QOL). It showed a moderate positive correlation between the total dysarthria score and the total quality of life score in the SG ($p = 0.0008$; Spearman coefficient = 0.75202). However, there was no correlation between diagnostic time and QOL ($p = 0.84$, Spearman coefficient = -0.05469) (Figure 1). The best result for the group studied was that the greater the dysarthria, the greater the impact on the quality of life of the individual with ALS.

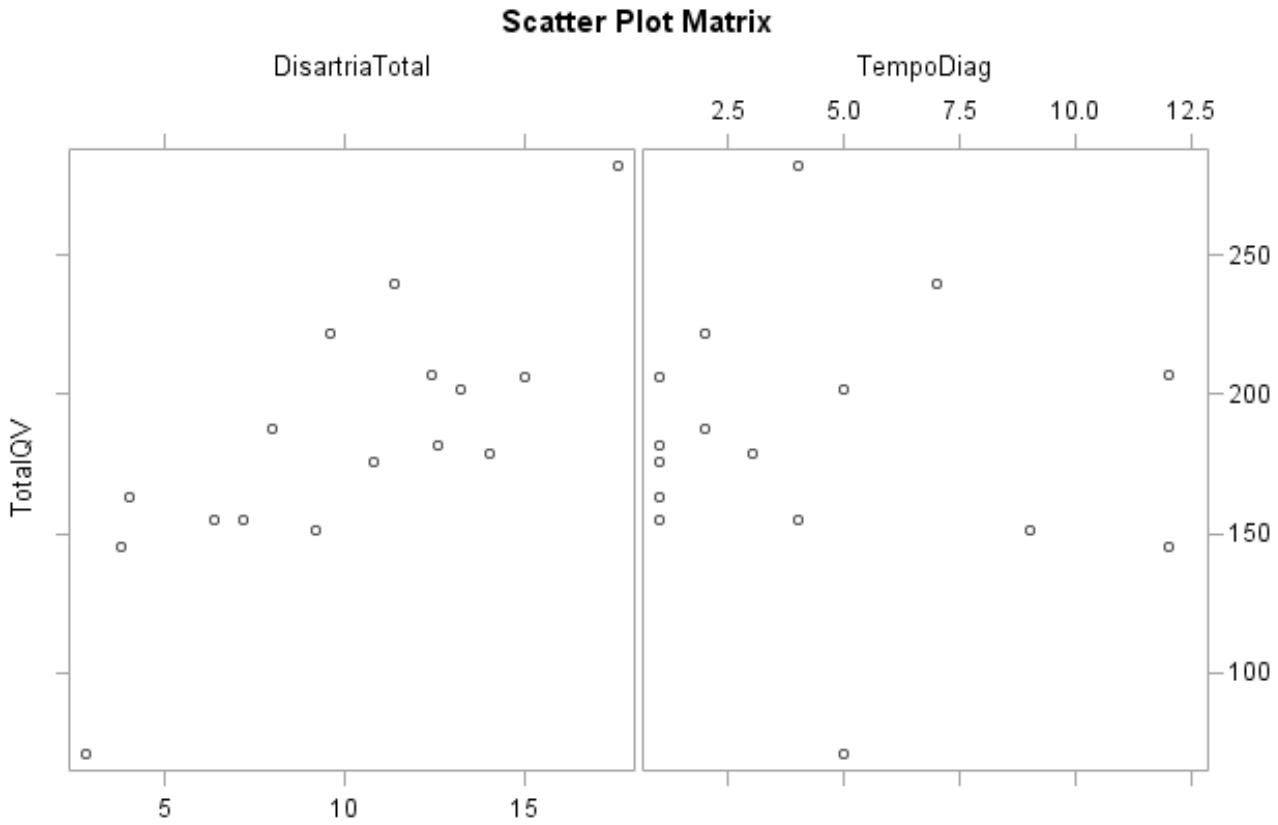

Figure 1. Spearman correlation between dysarthria and time of diagnosis with quality of life in the study group

DISCUSSION

This study presented different results from those described in the literature in regards to gender, since studies reported a higher incidence of the disease in men than in women [11,12]. However the results in regards to age and duration of illness were similar and correlated with Epidemiological studies which reported the onset of diagnosis being within the 5th decade of life [13].

Speech and voice parameters analyzed demonstrated differences between SG and CG subjects. All items related to respiration, phonation, resonance, articulation and prosody of the SG changed, ensuring that the dysarthria findings were characteristics of the ALS group studied.

Findings of the SG's respiration parameters suggested a moderate degree of alteration (4.1 ± 1). These findings corroborated the literature on respiratory failure [14], indicating that it is possible to observe changes in ventilation control in individuals with ALS. These changes indicated signs of respiratory muscle fatigue, changes in the mechanical properties of the respiratory system, changes in gas exchange, especially at night, and dysfunction of the upper respiratory tract, causing the patient in advanced stages to require the use of mechanical ventilation in order to improve comfort and quality of life, and increase survival rate [15].

The effects of respiratory changes are evident in their communication abilities, as it interfered in their fluency, making speech more laborious and impairing intelligibility. According to the literature, respiration is directly linked to phonation, as it compromises the necessary connection between the aerodynamic and myoelastic mechanisms, leading the individual to present an uncoordinated system [16].

In regards to phonation, the findings showed a moderate degree of impairment in the SG (3 ± 2.2). Impairments included hoarseness, breathiness, roughness and vocal instability as the most frequently observed alterations in this study. Some authors have reported that vocal deterioration is common in individuals with ALS, and it is an important clinical feature in the diagnostic process of the disease as it signals bulbar involvement [15]. The main vocal symptom in ALS is hoarseness [17]; however, other symptoms can also be observed, such as: tension, breathiness, roughness, hypernasality, low frequency, low intensity, monotonous voice [18] and tremor [19].

In the voice analysis, performed by the evaluators, there was no consensus between the level of involvement found. Thus, it is important to note that the evaluators' experience is critical in order to minimize the subjectivity regarding the correct definition of the subject's vocal type. Moreover, auditory-perceptual analysis is sensitive to the quality of the recording, the presence of varying levels of environmental noise, and to the subjects' diversity participating in the research, in this case, especially in relation to age and degree of the disease that showed great variability. It is important to emphasize that this parameter was the one that presented with the highest score in the CG. Although the participants of this group did not present with any initial vocal complaints, the hypothesis about the level of alteration perceived by the judges may be due to the effects that several factors has overtime on the voice, such as eating, smoking, alcoholism, abusive vocal practices or even aging itself, which may cause physiological or functional changes to the vocal mechanism.

The findings on resonance showed a moderate degree of alteration among the SG participants (3 ± 2.2), characterized by the presence of hypernasality. Despite resonance changes not being frequently studied in the literature, research corroborated the findings of this study and reported that hypernasality [20] is present in ALS due to impairment of the velopharyngeal muscles and is acoustically characterized by decreased projection and reduction of vocal intensity, Which also commonly interferes with speech intelligibility [21].

Articulation is often affected in individuals with ALS and is part of the overall dysarthria found from the early stages as a result of the disease, which significantly compromises their speech intelligibility [13,16]. In most cases, the articulation changes happen due to deficiency of the orofacial muscles and atrophy of the tongue and lips [22]. The findings of this study indicated a moderate degree of change to this parameter (3.3 ± 2), which reaffirmed what is found in the scientific literature.

Prosody is related to three parameters: fundamental frequency, which is the physical correlate corresponding to the pitch; duration, which is the rhythm of speech (including speech rate and articulation); and intensity, related to the vocal energy used by the speaker [23]. The findings on prosody suggested a degree of mild to moderate impairment in the SG (2.5 ± 1.9). In subjects with ALS, it is possible to observe slowed rate of speech, which occurs due to decreased muscle tone and strength, reduced amplitude of articulatory movements and, eventually, tremor of the orofacial structures [14,16]. Another rationale is related to the vocal findings of reduced intensity and monotonous voice [18], which hinder the melodic variations of speech. Previous research using the same protocol for the evaluation of dysarthria, focusing on Parkinson's disease (PD), showed that prosody is a parameter that has been significantly altered since the initial phases, negatively influencing the QoL of subjects with PD [6].

Quality of life analysis revealed a correlation between dysarthria and quality of life of subjects with ALS, that is, with the increase in severity of the dysarthria, there was worsening of the QOL related to dysarthria. This finding confirms the initial hypothesis of this research and alludes to the importance of early diagnosis in order to mitigate the effects of the disease in communication. On the other hand, there was no correlation between quality of life and time of diagnosis. The absence of correlation shows that a shorter disease time may have a great impact on QOL related to dysarthria, or vice-versa, a longer time of diagnosis may have less of an impact on QOL. This contradicted an earlier study that pointed out a correlation between these two variables, based on the rationale that in the advanced stages of ALS individuals present with respiratory compromises, making activities of daily life (ADLs) and communication difficult [24]. Perhaps this finding is justified by the fact that the present study evaluated only the communication domain, as other aspects such as mobility and ADL have greater interference in quality of life [25].

The results of the "Living with Dysarthria" questionnaire indicated self-image difficulties in the SG. These difficulties lead individuals with ALS to social isolation as they are typically active participators in the social environment. A previous study [16] showed that in ALS, social withdrawal, together with communication difficulty, profoundly affects the patient's feelings of hope, self-esteem and dignity, as he/she is not understood often by their listeners, requiring constant repetition of words or phrases or in most situations choosing not to talk. Another study suggested that the difficulties of the listener to understand the speech of the subject with ALS and the unavoidable repetitions, decrease the communication rate of these individuals [7]. These difficulties have been also described in other neurodegenerative diseases such as Parkinson's disease [6].

The individual's perception of their own illness and communication difficulties were frequently self-reported by the participants in this study, as in the statements: "I worry about my speech difficulties" and "I think my

speech can change". These statements were marked with a high agreement rate by 87.5% of the participants.

When the two groups were compared, the CG presented with lower scores in the assessment of quality of life, suggesting that their speech difficulties, if any, did not have a significant impact on their QOL.

In light of the above, it is possible to conclude that ALS patients' dysarthria interferes with their communication, compromising their productivity, reducing social interaction and negatively influencing their QOL. Therefore, it is extremely important to understand the impact of these changes on quality of life, as it allows health professionals to better assist with therapeutic choices based on the correlation between objective findings of the evaluation and patients' self-perception about their communication. This will allow the patient to continue to be part of their social environment and will also allow him/her to participate in the decision-making process, planning and intervention plans together with the medical and support staff, and treatment services [25].

Study Limitation

Limitations of this study include the use of a mobile device, adopted for its practicality and possibility of simultaneous recording of audio and video samples. However, although this resource has the potential to be an important tool in the clinical practice, its use in scientific studies is still small and needs further research, in order to assess the possible effects on speech and speech analysis.

The use of translated protocols, but not validated for Brazilian Portuguese, can also be considered a limitation to the study. However, the use of the two protocols has allowed for the collection of data about dysarthria and its impact on the quality of life in individuals with ALS that would have not been possible without them.

Another important point to consider was the poor reliability of the phonation parameter in the protocol for the evaluation of dysarthria after application of the Kappa Fleiss Concordance Index. The different amount of professional voice experience among the three judges who participated can also be considered a limitation to the study because it may have corroborated the low correlation in this parameter, for an evaluation that is essentially subjective to the judges' expertise.

CONCLUSION

This study demonstrated a moderate positive correlation between QOL and dysarthria in ALS, revealing that the higher the severity of dysarthria, the more negative was its impact on the quality of life of the individuals with the disease. In addition, all speech parameters evaluated: respiration, phonation, resonance, articulation and prosody were altered in this group to varying degrees and contributed to the deterioration in communication.

It is evident that issues related to speech disorders, in different situations of daily life, compromise the functionality of communication and restrict social participation, even with close family and friends, which further worsens their quality of life.

Thus, it is important to evaluate and monitor dysarthria as well as aspects of quality of life, in order to improve clinical practice and provide the best treatment options for individuals with ALS, and to help them with their social interaction needs in their advanced stages.

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