

Lavoisier Leite Neto¹ 
Marcondes Cavalcante França Júnior² 
Regina Yu Shon Chun¹ Speech intelligibility in people with
Amyotrophic Lateral Sclerosis (ALS)*Inteligibilidade de fala em pessoas com
Esclerose Lateral Amiotrófica (ELA)*

Keywords

Speech Intelligibility
Dysarthria
Speech Disorders
Speech Production Measurement
Amyotrophic Lateral Sclerosis

Descritores

Inteligibilidade de Fala
Disartria
Distúrbios da Fala
Medida da Produção da Fala
Esclerose Amiotrófica Lateral

Correspondence address:

Lavoisier Leite Neto
Programa de Pós-graduação Saúde,
Interdisciplinaridade e Reabilitação,
Faculdade de Ciências Médicas,
Universidade Estadual de Campinas –
UNICAMP
Rua Dona Antônia de Queirós, 239,
Ap 147, Consolação, São Paulo (SP),
Brasil, CEP: 01307-012.
E-mail: lavoisier.leite@hotmail.com

Received: August 21, 2019

Accepted: February 06, 2020

ABSTRACT

Purpose: To evaluate speech intelligibility and dysarthria, correlated to the functional assessment of Amyotrophic Lateral Sclerosis (ALS). **Methods:** Quantitative-descriptive study approved by REC under No. CAAE 62912416.4.0000.5404, comprised of 19 individuals with sporadic or familiar ALS. Data were collected using the Dysarthria Protocol and the Revised Amyotrophic Lateral Sclerosis Functional Rating Scale (ALSFRS-Re). We used visual analogue scale (VAS) to assess speech intelligibility and summary measures; and Spearman's coefficients of correlation for the instruments with significance level of 5%. **Results:** Speech intelligibility is compromised (41.37±39.73) in varied degrees with positive correlation with the general degree of dysarthria ($p < .0001$), and with all the analyzed speech parameters, indicating impact on the speech deterioration of the studied group. There is negative correlation between speech intelligibility and the results of the bulbar sections – speech and deglutition ($p = 0.0166$), arm – activities with the upper limb ($p = 0.0064$) and leg – activities with the lower limb ($p = 0.0391$). Breathing ($p = 0.0178$), phonation ($p = 0.0334$) and resonance ($p = 0.0053$) parameters showed a negative correlation with the item “speech” of the ALSFRS-Re. **Conclusion:** Results show impaired speech intelligibility and dysarthria, and evidence breathing, phonation and resonance as important markers of the disease progression. A thorough and early evaluation of the oral motor production allows for a better management of alterations in ALS.

RESUMO

Objetivos: Avaliar a inteligibilidade de fala e disartria, correlacionando com a avaliação funcional da ELA. **Método:** Estudo quantitativo-descriptivo aprovado pelo CEP, sob nº CAAE 62912416.4.0000.5404, constituído por 19 pessoas com ELA, esporádica ou familiar. Para coleta, aplicou-se o Protocolo de Disartria e Escala de Avaliação Funcional da Esclerose Lateral Amiotrófica (ALSFRS-Re). Para análise, foi utilizada escala visual analógica para inteligibilidade de fala e medidas de resumo e correlação dos instrumentos pelo Coeficiente de Spearman com nível de significância de 5%. **Resultados:** A inteligibilidade de fala está comprometida (41,37±39,73) em graus variados com correlação positiva com o grau geral de disartria ($p < .0001$) e com todos os parâmetros de fala analisados, indicando impacto na deterioração da fala do grupo estudado. Há correlação negativa entre inteligibilidade de fala e resultados das seções bulbar – fala e deglutição ($p = 0,0166$), braço – atividades com membro superior ($p = 0,0064$) e perna – atividades com membro inferior ($p = 0,0391$). Os parâmetros de respiração ($p = 0,0178$), fonação ($p = 0,0334$) e ressonância ($p = 0,0053$) apresentaram correlação negativa com o item “fala” do ALSFRS-Re. **Conclusão:** Os achados mostram prejuízo da inteligibilidade de fala e disartria e evidenciam respiração, fonação e ressonância como importantes marcadores da progressão da doença. Uma avaliação criteriosa e precoce da produção motora oral permite melhor gerenciamento das alterações na ELA.

Study conducted at Programa de Pós-graduação Saúde, Interdisciplinaridade e Reabilitação, Faculdade de Ciências Médicas, Universidade Estadual de Campinas – UNICAMP - Campinas (SP), Brasil.

¹ Programa de Pós-graduação Saúde, Interdisciplinaridade e Reabilitação, Faculdade de Ciências Médicas, Universidade Estadual de Campinas – UNICAMP - Campinas (SP), Brasil.

² Departamento de Neurologia, Faculdade de Ciências Médicas, Universidade Estadual de Campinas – UNICAMP - Campinas (SP), Brasil.

Financial support: Coordenação de Aperfeiçoamento de Pessoal de Nível Superior – CAPES (process n. 01-P-1734/2016, process n. 01-P-3376/2017, process n. 01-P-1604/2018, process n. 02-P-4631/2019).

Conflict of interests: nothing to declare.



This is an Open Access article distributed under the terms of the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly cited.

INTRODUCTION

Amyotrophic Lateral Sclerosis (ALS) is a heterogeneous neurodegenerative disease, which presents motor, cognitive, and behavioral changes. Recent pathophysiological studies show genetic, environmental, and lifestyle causes as the main causes of the disease⁽¹⁾.

According to the literature, 90% of cases appear sporadically and approximately 10% are of a family nature. Although there are no specific disease biomarkers reported in the literature, previous research reported a relationship with findings of ALS-related gene mutations and reduced dismutation activity of the superoxide dismutase enzyme SOD1⁽²⁾. Also, a recent study indicated that skeletal muscle microRNAs (miRNAs), MiR-424, and miR-206 are potential prognostic markers for ALS⁽³⁾. No significant environmental factors are reported; however, smoking and lean body structure can be some aggravating factors⁽²⁾.

The disease presents several symptoms including muscle weakness as the most noticeable characteristic, affecting all muscles in the body, including the respiratory muscles. Also, 30% of the population has a bulbar onset of symptoms, commonly associated with a worse prognosis, showing, gradual changes in speech production and swallowing since the diagnosis more intensely, and more than 80% of the cases show the same symptoms during its progression, even those that reach anarthria^(1,4-6).

Frequent speech motor changes in ALS, such as dysarthria, are characterized by impaired oral production due to impaired neuromuscular control, slower, weak, and/or inaccurate communication, impaired breathing, phonation, resonance, articulation, and prosody, with wide a variability of symptoms⁽⁷⁾. As dysarthria progress, they compromise the functionality of communication in day-to-day activities, social participation, and negatively interfere with the person's quality of life^(6,8).

In ALS, dysarthria is generally characterized by decreased strength and velopharyngeal movement, larynx, lips, tongue, and mandible; articulatory imprecision; decreased speech rate; hypernasality and hoarseness; tension, breathiness and harshness and monotony^(4,9).

Speech intelligibility is related to how much the message is understood by the interlocutor, i.e., the ease of understanding the content in a conversation. Therefore, it must be perceived from the point of view of the speaker and his communication partner⁽¹⁰⁾. In ALS, the decrease in speech rate shows the beginning of the decline in this parameter⁽⁵⁾.

Longitudinal studies assessing the progression of dysarthria in ALS are scarce. However, the literature reports that in approximately one and a half years there is an evolution from mild to severe and anarthria can occur 17 months after the beginning of the first speech changes⁽⁶⁾.

The speech changes experienced by the person with ALS, and the precipitation of the loss of this function, makes communication vulnerable because it compromises the speech⁽¹¹⁾. In this situation, the use of Supplementary and/or Alternative Communication (SAC) is configured as a communication strategy with the family, the health team, and close friends, facilitating socialization, autonomy, and decision making⁽¹²⁾. The communicative vulnerability situation experienced by the

person with ALS, especially after the loss of communication, can lead to emotional problems and/or anxiety, frustration, fear, and sadness^(8,12), and can be considered as one of the worst aspects of the disease⁽⁶⁾.

Therefore, we emphasize the importance of speech care in maintaining the quality of life of the person with ALS, especially in a context with progressive limitations during the course of the disease, motivating the search for specific markers that allow the early diagnosis of bulbar change and enable to treat the aggravation of motor speech production, from the initial stages, when the first symptoms appear.

Thus, this article aims to assess speech intelligibility and dysarthria, correlated with the functional assessment of ALS, based on the following guiding questions: Which speech components interfere most intensely in their intelligibility and may be possible markers of impaired motor oral production? Are dysarthria and impaired speech intelligibility related to the severity of the disease?

METHODS

This is a quantitative and descriptive study, approved by the Ethics and Research Committee (ERC), under No. CAAE 62912416.4.0000.5404.

Sample constitution

The study included 19 participants diagnosed with ALS and seen at a Neuromuscular Diseases Clinic. Fifteen were sporadic cases, and 4 were familial cases. The following inclusion criteria were considered: confirmed diagnosis of Amyotrophic Lateral Sclerosis, sporadic or familial, according to the revised criteria El Escorial; heterogeneous demographic characteristics and varied stages of the disease; both genders; and age greater than or equal to 18 years old. The exclusion criteria included: people with advanced ALS stage, having concomitant unrelated neurological conditions; inability to respond adequately to the instruments used in the research, even with the help of the researcher and use of tracheostomy and/or nasogastric tube.

Data collection procedures

Participants were contacted during medical care at the Neuromuscular Diseases Outpatient Clinic, with periodic follow-up with a pre-scheduled appointment, at times previously established by the sector and convenient, without causing any damage in the follow-up.

For data collection, the researcher sent the participant to a reserved room, with a minimum of external noise, so as not to interfere with the data collection procedures, where the following research instruments were applied:

- a) Initial interview to collect data related to gender, age, years of education, and length of illness of each interviewee;
- b) Dysarthria evaluation through the "Dysarthria Evaluation Protocol"⁽¹³⁾ (Annex 1), which includes the analysis of the following components: breathing, phonation, resonance,

articulation, and prosody. The Protocol was translated and adapted into Portuguese, and despite not being validated, it was sensitive for assessing the speech of the individuals with ALS, according to a previous study⁽⁷⁾. The instrument required some adaptations for its use in this study due to the reality of the studied population and available material so that the analysis of the five parameters was as follows:

- Breathing: counting the number of breathing cycles for one minute; sustained emission of the phonemes / a /, / i /, / s /, / z /; and the word count by expiration;
- Phonation: analysis performed through the sustained emission of vowels, words, phrases, and spontaneous speech;
- Resonance: emission of phonemes / a / e / ã / alternately, repetition of the syllable / ka / and reproduction of oral and nasal words and phrases;
- Articulation: spontaneous and forced emission of the phonemes / i /, / u / and syllable / pa /; spontaneous repetition in increasing speed of the syllables / ka /, / ta / and diadocokinesia (Pataka, Badaga, Fasacha); and naming figures. When the evaluated participant did not know or remember the name of the figure presented, it was disregarded. Analysis of articulatory precision and speech intelligibility was performed;
- Prosody: Spontaneous speech and reading of affirmative, interrogative, and exclamatory phrases. If the participant did not know how to read, the sentences were read by the evaluator, without an intonation mark corresponding to each type of sentence.

- c) Application of the revised version of the Functional Assessment of Amyotrophic Lateral Sclerosis (ALSFRS-Re)⁽¹⁴⁾ (Annex 2). The ALSFRS-R is a functional inventory specific to ALS, validated for the Portuguese language and composed of 12 items: speech, salivation, swallowing, writing, cutting food and handling utensils, dressing and hygiene, turning in bed, walking, climbing stairs, dyspnea, orthopnea and respiratory failure⁽¹⁴⁾. Each item has five possible answers, ranging from 0 to 4, with zero indicating severe impairment and 4 indicating no change. The sum of the items determines the total score. The protocol is divided into 4 sections of the body: bulbar, arms, chest, and legs, with the score of these sections calculated from 0.0 to 1.0. The closer to 1.0, the lower the commitment.

Speech samples were collected using a Canon Powershot SX530 HS Digital Camera with 3-inch LCD, 16.0 megapixel 1/2.3-inch CMOS sensor, 50x optical zoom (24-1200mm), Canon DIGIC 4 Image processor, shoots in Full HD 1920 x 1080, NTSC/PAL video output (dedicated [female] connector with unified type of digital, audio and video) mini-HDMI connector; stereo audio output (dedicated [female] connector with unified digital, audio and video type), as well as mini-tripod for the camera for better stabilization and adjustment of the images.

The video recorded the participants in a sitting position, at a distance that allowed them to view their torso and head for further analysis of the images.

Two judges analyzed the data collected from the Dysarthria Protocol and speech intelligibility to guarantee the impartiality of the judgment of the samples. They were speech therapists with experience in neurological disorders, both of whom received prior individualized training by one of the researchers responsible for the protocol applied to increase the reliability of the analysis.

One of the researchers carried out the application of the Functional Evaluation Scale through direct questioning to the participant about the items contained in the ALSFRS-Re.

Due to the severity of the speech symptoms, some individuals needed the help of the researcher and/or a family member or companion to answer both protocols. For that, when possible, we accepted gestures with head movements indicative of “yes” and “no”, the use of figures, writing, and typing on the cell phone, when the participants were already using some of these strategies. None of them used supplementary and/or alternative communication devices.

Data analysis form

The literature described several methods for evaluating this parameter, divided into two groups: identification and graduation, in which the visual analog scale was used by the judges, considered a viable method, easy to perform, and widely used in scientific circles⁽¹⁰⁾.

The visual analog scale consists of a horizontal line with limits that determine its extremes. In this case, from 0 to 100, with zero indicating greater impairment and one hundred indicating no change in speech intelligibility. The trial was carried out through the recording of spontaneous speech samples and the evidence contained in the Dysarthria Protocol.

The data were tabulated and analyzed, statistically, using the software The SAS System for Windows (Statistical Analysis System), version 9.4.

Exploratory data analysis was performed using summary measures (mean, standard deviation, minimum, median, maximum, frequency, and percentage). The agreement between the judges was analyzed using the Intraclass Correlation Coefficient. Spearman's Coefficient analyzed the correlation between the instruments and the instruments with age and disease duration. The level of significance adopted was 5%.

RESULTS

Table 1 shows the summary measures related to the characterization of the participants.

The average age of the participants is 58.42 ± 13.27 , lower than the average of symptoms onset found in the literature, which is in the sixth decade of life^(1,15,16). Nine participants (47.36%) were under 60 years old, a factor that can influence the prognosis and functional decline of individuals with the progression of the disease⁽¹⁵⁾.

Table 1. Characterization of participants regarding age, gender, time of study, and time of diagnosis

	Variables	Participants
Age	Mean	58.42
	Median	61.00
	Minimum-Maximum	31-79
	Standard Deviation	13.27
Gender	Female	8 (42.11%)
	Male	11 (57.89%)
Time of study (years)	Mean	9.95
	Median	11.00
	Minimum-Maximum	2-15
	Standard Deviation	4.25
Time of diagnosis (years)	Mean	4
	Median	1-9
	Minimum-Maximum	4
	Standard Deviation	2.91

Epidemiological studies show little variability of the disease concerning age; however, there is a higher prevalence in males⁽¹⁷⁾, an aspect that corroborates our findings.

The average study participation time was 9.95 ± 4.25 years, with eleven of the participants having a level of education between high school and higher education.

The average time of diagnosis was 4 ± 2.91 years; however, most of the participants had up to 5 years and, only five had between 6 and 9 years.

According to the intra-judge agreement test, regarding the parameters of breathing, phonation, resonance, articulation, prosody, general degree of dysarthria, and speech intelligibility, there was a high or very high agreement, between the judges, for the analyzed items, which makes the analysis more reliable.

Table 2 summarizes the measures of the study sample related to the parameters evaluated (breathing, phonation, resonance, articulation, prosody), general degree of dysarthria, and speech intelligibility.

The average of the general degree of dysarthria of the participants was 12.4 ± 10.7 , indicating moderate impairment but with great variation in results among people with ALS (0.5-30.0).

The average of the participants in the speech intelligibility was 41.37 ± 39.73 , showing an important impairment of this variable. However, these results have no significant correlation with time of disease onset and age, and participants with shorter time of diagnosis and younger, also showed changes in speech, as seen in Figure 1.

Table 3 shows a comparison between speech intelligibility and parameters of breathing, phonation, resonance, articulation, prosody, and general degree of dysarthria.

There is a positive correlation between the parameters analyzed, showing that all the speech components studied have a significant influence on intelligibility. In the analysis of dysarthria, all the items evaluated indicate mild to moderate impairment, allowing the inference that, even in the initial

Table 2. Summary measures according to the parameters of breathing, phonation, resonance, articulation, prosody, general degree of dysarthria, and speech intelligibility

Variables	Sample (n=19)*	
Breathing	Mean	2.9
	Median	2.0
	Minimum-Maximum	0.5-6.0
	Standard Deviation	2.0
Phonation	Mean	2.7
	Median	2.0
	Minimum-Maximum	0.0-6.0
	Standard Deviation	2.2
Resonance	Mean	2.2
	Median	1.0
	Minimum-Maximum	0.0-6.0
	Standard Deviation	2.3
Articulation	Mean	2.4
	Median	1.5
	Minimum-Maximum	0.0-6.0
	Standard Deviation	2.4
Prosody	Mean	2.0
	Median	0.5
	Minimum-Maximum	0.0-6.0
	Standard Deviation	2.3
General Degree	Mean	12.4
	Median	7.5
	Minimum-Maximum	0.5-30.0
	Standard Deviation	10.7
Intelligibility	Mean	41.37
	Median	20.5
	Minimum-Maximum	2-100
	Standard Deviation	39.73

Caption: (*) the average of the two judges was used; n = sample number

stages and/or with mild alteration of oral production, there is impaired speech intelligibility.

When comparing the sections of the ALSFRS-Re with the parameters of the Dysarthria Protocol of the studied group, we observed a negative correlation between the general degree and speech intelligibility with bulbar sections, arms, and legs, as shown in Table 4. Speech impairment advanced with the worsening of the severity of the assessed functions, which make up these sections. Such an interpretation is justified since while in the ALSFRS-Re, the level of commitment is lower with a higher score, in the Dysarthria Protocol the opposite happens, there is a greater commitment with a lower score.

When comparing the severity of speech with the parameters of dysarthria and intelligibility of the participants, we observed a negative correlation between the progress of functional impairment with the items breathing, phonation, resonance, and intelligibility, indicating that these have a greater impact on the deterioration of speech. Thus, as there is a worsening of these components, communication becomes more impaired, which does not happen with articulation and prosody, which probably interferes, later, as shown in Table 5.

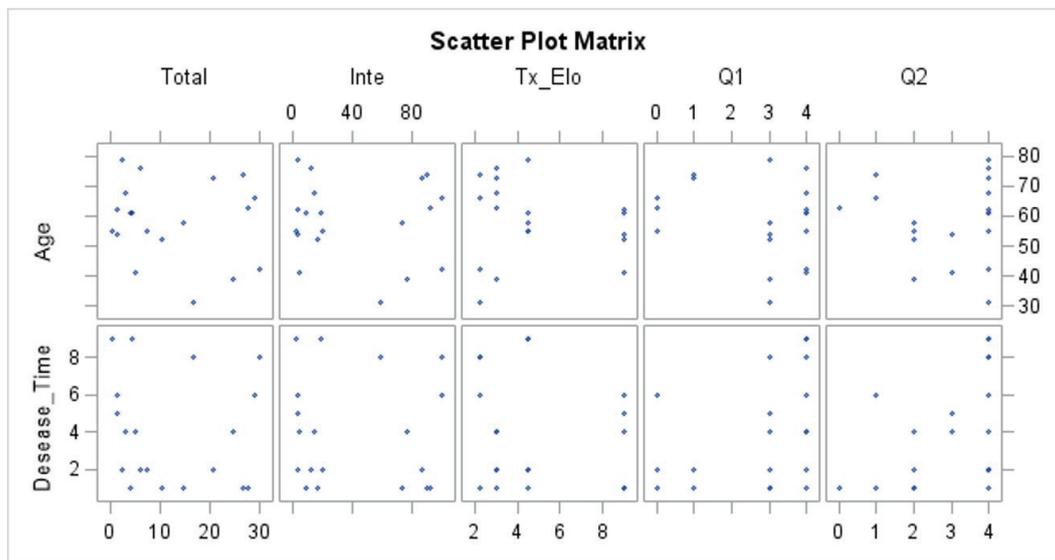


Figure 1. Correlation of age and disease duration with speech intelligibility (*)
Subtittle: Inte – Speech intelligibility; (*) Spearman's coefficient

Table 3. Comparison between speech intelligibility with breathing, phonation, resonance, articulation, prosody, and general degree of dysarthria

Variables	Breathing	Phonation	Resonance	Artic	Pros	Total
Intel	0.83314	0.82192	0.89138	0.94748	0.86010	0.96047
	<.0001	<.0001	<.0001	<.0001	<.0001	<.0001

Caption: Artic – Articulation; Pros – Prosody; Total – General Degree; Intel – Intelligibility

Table 4. Correlation between the results of the Dysarthria Protocol and the sections of the ALSFRS-Re

sections/ variables	Breathing	Phonation	Resonance	Artic	Pros	Total	Intel
B	-0.53599	-0.52612	-0.62119	-0.45129	-0.28546	-0.56519	-0.54179
	0.018	0.0207	0.0045	0.0524	0.2362	0.0117	0.0166
A	-0.60748	-0.66058	-0.60627	-0.60496	-0.41614	-0.67065	-0.60204
	0.0058	0.0021	0.0059	0.0061	0.0764	0.0017	0.0064
C	-0.28236	-0.30125	-0.47654	-0.44825	-0.03047	-0.30959	-0.30708
	0.2415	0.1541	0.0391	0.0543	0.9015	0.1971	0.201
L	-0.5105	-0.34019	-0.47654	-0.44825	-0.25961	-0.46435	-0.47653
	0.0255	0.1541	0.0391	0.0543	0.2831	0.0452	0.0391

Caption: Artic – Articulation; Pros – Prosody; Total – General Degree; Intel – Intelligibility; B: Bulbar; A: Arms; C: Chest; L: Leg

Table 5. Comparison between speech severity, breathing, phonation, articulation, prosody, and speech intelligibility

Issues	Breathing	Phonation	Resonance	Artic	Pros	Total
1. Speech	-0.53691 (0.0178)	-0.48956 (0.0334)	-0.61266 (0.0053)	-0.45322 (0.0513)	-0.26617 (0.2707)	-0.54823 (0.0151)

Caption: Artic – Articulation; Pros – Prosody; Total – General Degree; Intel – Intelligibility

DISCUSSION

Speech unintelligibility is one of the main manifestations in people with acquired speech impairments and, although there is no consensus in the literature about methods for evaluating this parameter, it suffers interference from several factors in its judgment, such as speech task used, stimulus offered, mode of presentation of the samples to be evaluated, type of response in the identification of stimuli, analysis in transcription, the gender of the listener and familiarity with the speaker⁽¹⁰⁾. According to the same authors, the visual analog scale that is a strategy used

in this study is a grading method that enables the analysis of this parameter, which, together with the Dysarthria Protocol, proved to be sensitive in previous research⁽⁷⁾. Also, the Functional Evaluation Scale (ALSFRS-Re) of established scientific use obtained the research data.

The results show that speech intelligibility is compromised (41.37 ± 39.73) in people with ALS studied and this aspect is influenced by all the components analyzed (breathing, phonation, resonance, articulation, and prosody).

Such findings corroborate previous studies that state that with the progression of ALS, there is a loss of speech intelligibility,

which evidences a gradual loss of control and muscle strength of phonoarticulatory organs, reduction, and increase in the duration of articulatory movements⁽⁶⁾, decrease progressive speech rates and an increase in the number and duration of pauses in speech⁽¹⁸⁾. Also, other authors reinforce the negative impact of these changes for the impoverishment of language, which makes the discourse increasingly laborious and reduced^(7,19), distancing the individual from social life⁽¹²⁾.

We observed no correlation between intelligibility and disease duration or age in the population studied here. Such findings allow us to infer that, even in the early stages, people with ALS may present significant impairment of speech motor function, which compromises their contact with other people, even family members, and close friends, and limits their autonomy as indicated in the literature^(7,12).

The results found here differ from other studies^(4,15) that indicate that the loss of speech intelligibility happens in more advanced stages of the disease. These authors highlight that the age of onset of symptoms interferes with the prognosis and decline of bulbar and respiratory function, although the mechanism is not clearly understood. It is important to consider that some known prognostic factors such as cognitive impairment and nutritional status were not considered in the studies, which can be a limiting factor, despite ratifying the genetic and pathophysiological heterogeneity of ALS. Thus, other authors reinforce the importance of the time of disease onset, as being one of the most significant factors that contribute to the deterioration of speech functions; however, they indicate that neither age nor gender is significant in the decline in these functions⁽⁶⁾.

On the other hand, the findings of this research indicate a positive correlation between speech intelligibility and the parameters of dysarthria analyzed, corroborating the findings in the literature that, as dysarthric changes worsen, speech becomes increasingly unintelligible⁽⁵⁾.

In the results of the Dysarthria Protocol, we observed a moderate level of general changes in the studied group, a result similar to the previous study⁽⁷⁾, with breathing (2.9 ± 2.0), phonation ($2.7 \pm 2, 2$), and articulation (2.4 ± 2.4) the most compromised variables among the analyzed speech parameters.

Our findings related to breathing showed the presence of mild to moderate impairment, similar to the literature that indicates the presence of impairment of this function in ALS, affecting the inspiratory, expiratory, and bulbar muscles and can generate oxygen desaturation due to hypercapnia, cough and excessive saliva aspiration. In advanced stages of the disease, it is also possible to observe symptoms of respiratory muscle weakness, such as dyspnea on exertion, fatigue, morning headache, or sleep disorders, culminating in nocturnal hypoventilation, most of the times with the need to use mechanical ventilation⁽²⁰⁾.

Our results also show a positive correlation between the severity of speech and the breathing parameter, showing that the increase in the impairment of respiratory function harms communication. This finding converges with another study that approaches that the general respiratory state can affect the speech performance of patients with ALS and complicate the articulatory execution⁽⁴⁾.

As in the breathing parameter, the phonation has the presence of mild to moderate impairment and a positive correlation between it and the speech severity in the analyzed individuals, reinforcing that these changes significantly interfere with communication. This finding is similar to other studies, which add that phonoarticulatory impairment is one of the first bulbar symptoms to appear, and often goes along with vocal aspects of breathiness, hoarseness, tension, decreased intensity, and hypernasality^(7,21). Also, the same authors reiterate that the assessment of the presence of these vocal changes is important and can contribute to the differential diagnosis of the disease, providing subsidies to its evolution.

As previously mentioned, we identified changes in the articulation parameter in this research, which varied from mild to moderate and a positive correlation with speech intelligibility, showing that as the articulation movements worsen, speech becomes less intelligible. This finding converges with the literature that states that, in ALS, the ability to produce such movements accurately is impaired, as the decline in speech motor function occurs⁽²²⁾, making it unintelligible once these changes progress^(18,22,23). Other authors explain that the early diagnosis of joint damage is essential in detecting bulbar involvement and must be considered⁽²⁴⁾.

Although the articulation influenced intelligibility, there was no correlation with speech severity. Thus, despite the presence of the impairment, it is not frequently perceived and/or diagnosed.

The other parameters of dysarthria analyzed in this study were resonance and prosody and they were altered and showed a positive correlation with speech intelligibility, despite showing less impact on it. Such findings are consistent with those found in the literature, which states that the disease progresses with decreased strength and velopharyngeal movement, producing hypernasality⁽⁸⁾ and monotonous voice, with few melodic variations⁽⁷⁾.

In this research, when comparing the sections of the Functional Assessment Scale (ALSFRS-Re) with the intelligibility and general degree of dysarthria, we observe negative correlation between the bulbar, arm, and leg sections, showing that the worsening of speech accompanies the loss of motor function. The results indicate that the ALSFRS-Re scale is a sensitive measure to assess speech functionality since the beginning of the appearance of bulbar symptoms, unlike another study⁽⁶⁾.

Also, the findings of ALSFRS-Re indicate the need to use SAC. With the impairment of intelligibility and the possibility of total speech loss, the need to use supplementary and/or alternative communication is essential, from the initial stages^(12,25,26). In a previous study, 60% of the participants needed to use this strategy to maintain the functionality of the communication⁽⁶⁾.

Also, the results show that the parameters of breathing, phonation, and resonance seem to have a greater impact on the severity of speech in the perception of the studied group. Thus, such components may constitute important markers of loss of speech intelligibility in ALS. Therefore, an accurate speech evaluation by a specialist professional such as a speech therapist from the first diagnostic consultations can favor the perception of the first bulbous symptoms, becoming a fundamental aspect for the differential diagnosis, and to guide clinical and therapeutic

follow-up and indicate alternative forms of communication such as SAC.

Finally, the results reaffirm that the presence of a multidisciplinary and interdisciplinary team, composed of neurologists, psychologists, nutritionists, pulmonologists, physiotherapists, speech therapists, and specialized nurses is essential in the management of changes and assistance to the person with ALS to prolong survival, reduce hospitalization time and costs and improve the quality of life of this population group, as discussed by authors in the area⁽¹⁾.

CONCLUSION

The findings indicate a positive correlation between speech intelligibility and dysarthric changes in the population studied, which negatively influence the communication functionality of the person with ALS. The results show that the parameters of breathing, phonation, and resonance have a greater impact on speech production, showing that they can constitute important markers for speech unintelligibility.

Thus, the findings reaffirm the importance of a careful assessment of speech by a specialized professional such as the speech therapist, from the initial diagnosis of the disease. A proper care team will enable the continuous management of the changes in the oral motor production, even when there is still no significant impairment of speech intelligibility, and representing a relevant contribution to the differential diagnosis of ALS, since it may indicate bulbar involvement. Early diagnosis and specialized follow-up may favor the promotion of other forms of communication as necessary to minimize the negative impact on communication and quality of life of these people.

In this sense, the results confirm that in comprehensive care for people with AL, the participation of a multidisciplinary and interdisciplinary team is essential, prolonging survival, reducing hospitalization time and costs, and improving the quality of life of this population group.

The results reiterated the applicability of the Dysarthria Protocol for the analysis of speech components in ALS as found in previous research. The ALSFRS-Re is also a sensitive scale to analyze speech disorders from the initial stages, enabling to guide clinical and therapeutic follow-up in this area.

The findings of the ALSFRS-Re scale also showed a negative correlation between the bulbar, arm, and leg sections with speech intelligibility and dysarthria, showing the impairment of these variables accompanies the deterioration of speech motor function.

Therefore, the study shows that the loss of speech intelligibility in ALS is inevitable, and attention at this aspect is essential to maintain communication, autonomy, decision making, and quality of life for this population group, in favor of assistance to multi and interdisciplinary, integral and humanized health. For this reason, the results of this research can generate safety in clinical practice and are configured as allies to sensitize health professionals to develop strategies for better acceptance of the use of Supplementary and/or Alternative Communication, between people with ALS and their families and/or caregivers.

REFERENCES

1. Hardiman O, Al-Chalabi A, Chio A, Corr EM, Logroscino G, Robberecht W, et al. Amyotrophic lateral sclerosis. *Nat Rev Dis Primers*. 2017;3(1):17071. <http://dx.doi.org/10.1038/nrdp.2017.71>. PMID:28980624.
2. Chiò A, Battistini S, Calvo A, Caponnetto C, Conforti FL, Corbo M, et al. Genetic counselling in ALS: facts, uncertainties and clinical suggestions. *J Neurol Neurosurg Psychiatry*. 2014;85(5):478-85. <http://dx.doi.org/10.1136/jnnp-2013-305546>. PMID:23833266.
3. de Andrade HM, de Albuquerque M, Avansini SH, de S Rocha C, Dogini DB, Nucci A, et al. MicroRNAs-424 and 206 are potential prognostic markers in spinal onset amyotrophic lateral sclerosis. *J Neurol Sci*. 2016;368:19-24. <http://dx.doi.org/10.1016/j.jns.2016.06.046>. PMID:27538595.
4. Rong P, Yunusova Y, Wang J, Green JR. Predicting early bulbar decline in amyotrophic lateral sclerosis: a speech subsystem approach. *Behav Neurol*. 2015;2015:183027. <http://dx.doi.org/10.1155/2015/183027>. PMID:26136624.
5. Rong P, Yunusova Y, Wang J, Zinman L, Pattee GL, Berry JD, et al. Predicting speech intelligibility decline in amyotrophic lateral sclerosis based on the deterioration of individual speech subsystems. *PLoS One*. 2016;11(5):e0154971. <http://dx.doi.org/10.1371/journal.pone.0154971>. PMID:27148967.
6. Makkonen T, Ruottinen H, Puuto R, Helminen M, Palmio J. Speech deterioration in amyotrophic lateral sclerosis (ALS) after manifestation of bulbar symptoms. *Int J Lang Commun Disord*. 2018;53(2):385-92. <http://dx.doi.org/10.1111/1460-6984.12357>. PMID:29159848.
7. Leite L No, Constantini AC. Dysarthria and quality of life in patients with amyotrophic lateral sclerosis. *Rev CEFAC*. 2017;19(5):664-73. <http://dx.doi.org/10.1590/1982-021620171954017>.
8. Makkonen T, Korpjaakko-Huuhka AM, Ruottinen H, Puuto R, Hollo K, Ylinen A, et al. Oral motor functions, speech and communication before a definitive diagnosis of amyotrophic lateral sclerosis. *J Commun Disord*. 2016;61:97-105. <http://dx.doi.org/10.1016/j.jcomdis.2016.04.002>. PMID:27110704.
9. da Costa Franceschini A, Mourão LF. Dysarthria and dysphagia in Amyotrophic Lateral Sclerosis with spinal onset: A study of quality of life related to swallowing. *NeuroRehabilitation*. 2015;36(1):127-34. <http://dx.doi.org/10.3233/NRE-141200>. PMID:25547774.
10. Barreto SS, Ortiz KZ. Medidas de inteligibilidade nos distúrbios da fala: revisão crítica da literature. *Pró-Fono*. 2008;20(3):201-6. <http://dx.doi.org/10.1590/S0104-56872008000300011>. PMID:18852969.
11. Chun RYS, Leite L No, Zaquero VF, Mais ALW, Farias LP. Comunicação vulnerável em casos de alta complexidade: perspectiva de atuação e pesquisa fonoaudiológica em hospital-escola. In: Silva RM, Bezerra IC, Brasil CCP, Moura ERF, organizadores. *Estudos qualitativos: enfoques teóricos e técnicas de coleta de informações*. Sobral: Edições UVA, 2018. pp. 81-96.
12. Leite L No, Constantini AC, Chun RYS. Communication vulnerable in patients with Amyotrophic Lateral Sclerosis: a systematic review. *NeuroRehabilitation*. 2017;40(4):561-8. <http://dx.doi.org/10.3233/NRE-171443>. PMID:28222570.
13. Fracassi AS, Gatto AR, Weber S, Spadotto AA, Ribeiro PW, Schelp AO. Adaptação para a língua Portuguesa e aplicação de protocolo de avaliação das disartrias de origem central em pacientes com Doença de Parkinson. *Rev CEFAC*. 2011;13(6):1056-65. <http://dx.doi.org/10.1590/S1516-18462011005000030>.
14. Cedarbaum JM, Stambler N, Malta E, Fuller C, Hilt D, Thurmond B, et al. The ALSFRS-R: a revised ALS functional rating scale that incorporates assessments of respiratory function. BDNF ALS Study Group (Phase III). *J Neurol Sci*. 1999;169(1-2):13-21. [http://dx.doi.org/10.1016/S0022-510X\(99\)00210-5](http://dx.doi.org/10.1016/S0022-510X(99)00210-5). PMID:10540002.
15. Yokoi D, Aotsuta N, Watanabe H, Nakamura R, Hirakawa A, Ito M, et al. Age of onset differentially influences the progression of regional dysfunction in sporadic amyotrophic lateral sclerosis. *J Neurol*. 2016;263(6):1129-36. <http://dx.doi.org/10.1007/s00415-016-8109-0>. PMID:27083563.
16. Chiò A, Logroscino G, Traynor BJ, Collins J, Simeone JC, Goldstein LA, et al. Global epidemiology of amyotrophic lateral sclerosis: a systematic

- review of the published literature. *Neuroepidemiology*. 2013;41(2):118-30. <http://dx.doi.org/10.1159/000351153>. PMID:23860588.
17. Prado LG, Bicalho IC, Vidigal-Lopes M, Ferreira CJ, Mageste Barbosa LS, Gomez RS, et al. Amyotrophic lateral sclerosis in Brazil: case series and review of the Brazilian literature. *Amyotroph Lateral Scler Frontotemporal Degener*. 2016;17(3-4):282-8. <http://dx.doi.org/10.3109/21678421.2016.1143011>. PMID:26854959.
 18. Yunusova Y, Graham NL, Shellikeri S, Phuong K, Kulkarni M, Rochon E, et al. Profiling speech and pausing in amyotrophic lateral sclerosis (ALS) and frontotemporal dementia (FTD). *PLoS One*. 2016;11(1):1-18. <http://dx.doi.org/10.1371/journal.pone.0147573>. PMID:26789001.
 19. De Marco M, Merico A, Berta G, Segato N, Citton V, Baglione A, et al. Morphometric correlates of dysarthric deficit in amyotrophic lateral sclerosis. *Amyotroph Lateral Scler Frontotemporal Degener*. 2015;16(7-8):464-72. <http://dx.doi.org/10.3109/21678421.2015.1056191>. PMID:26121168.
 20. Almeida SRM, Silva LBC, Guerreiro CAM, Nucci A. Amyotrophic lateral sclerosis: prospective study on respiratory parameters. *Arq Neuropsiquiatr*. 2010;68(2):258-62. <http://dx.doi.org/10.1590/S0004-282X2010000200020>. PMID:20464296.
 21. Pontes RT, Orsini M, de Freitas MRG, Antonioli R S, Nascimento OJM. Alterações da fonação e deglutição na Esclerose Lateral Amiotrófica: revisão de Literatura. *Rev Neurocienc*. 2010;18(1):69-73. <https://doi.org/10.34024/mc.2010.v18.8505>.
 22. Wang J, Kothalkar PV, Kim M, Yunusova Y, Campbell TF, Heitzman D, et al. Predicting intelligible speaking rate in individuals with amyotrophic lateral sclerosis from a small number of speech acoustic and articulatory samples. *Workshop Speech Lang Process Assist Technol*. 2016;2016:91-7. <http://dx.doi.org/10.21437/SLPAT.2016-16>. PMID:29423454.
 23. Mefferd AS, Pattee GL, Green JR. Speaking rate effects on articulatory pattern consistency in talkers with mild ALS. *Clin Linguist Phon*. 2014;28(11):799-811. <http://dx.doi.org/10.3109/02699206.2014.908239>. PMID:24724615.
 24. Marchand DLP, Bonamigo AW. Atuação fonoaudiológica na voz do idoso: revisão sistemática exploratória de literatura. *Distúrb Comun*. [Internet]. 2015;27(2):309-17. [citado em 2019 Ago 21]. Disponível em: <http://revistas.pucsp.br/dic/article/view/21349>
 25. Caligari M, Godi M, Guglielmetti S, Franchignoni F, Nardone A. Eye tracking communication devices in amyotrophic lateral sclerosis: impact on disability and quality of life. *Amyotroph Lateral Scler Frontotemporal Degener*. 2013;14(7-8):546-52. <http://dx.doi.org/10.3109/21678421.2013.803576>. PMID:23834069.
 26. Ray J. Real-life challenges in using augmentative and alternative communication by persons with amyotrophic lateral sclerosis. *Comm Disord Q*. 2015;36(3):187-92. <http://dx.doi.org/10.1177/1525740114545359>.

Author contributions

LLN participated in the idealization of the study, collection, analysis and interpretation of data and writing of the article; MCFJ participated, as co-advisor, in the idealization of the study, supervision of the data collection and writing of the article; RYSC participated, as a supervisor, in the idealization of the study, analysis, data interpretation and writing of the article.

Annex 1. Brazilian Portuguese version of Dysarthria Evaluation Protocol
PROTOCOLO DE AVALIAÇÃO DAS DISARTRIAS

1. Respiração		
Velocidade (Ciclos/Minuto) - Normal 12-20 c/min		Normal 12-20 c/min
Tempo Máximo de Fonação	/a/	Normal 10-20 segundos
	/i/	Normal 10-20 segundos
	/s/	Normal 10-20 segundos
	/z/	Normal 10-20 segundos
Relação s/z		Normal 0,7 a 1,3
Palavras por expiração - contagem espontânea de números (40 a 1):		
Respiração: Análise indica comprometimento de grau:	0---1---2---3---4---5---6	(grau 0 ausência; 6 grave)
2. Fonação		
Qualidade Vocal		normal/pastosa/trêmula/rouca/aspera/soprosa
Ataque Vocal		isocrônico/brusco/aspirado
Intensidade Vocal		adequada/alta/baixa
Altura Vocal		adequada/grave/aguda
Varição da Qualidade		estável/instável
Voz: Análise indica comprometimento de grau:	0---1---2---3---4---5---6	(grau 0 ausência; 6 grave)
3. Ressonância		
Movimento velar	/a/; /ã/ alternadamente	adequada/mínima/ausente
Movimentação de parede faríngea	ka ka	adequada/mínima/ausente
Emissão nasal	Mamão x papai / pau x mau; Vovó viu a uva; Papai pediu pipoca; A fita de filó é verde; Amanhã mamãe amassará mamão.	Normal/hipernasal/hiponasal
Ressonância: Análise indica comprometimento de grau:	0---1---2---3---4---5---6	(grau 0 ausência; 6 grave)
4. Articulação		
Movimento de lábio	(i/u e pa) – espontâneos e forçados	normal/alterado
Língua	ka/ta – velocidade crescente	normal/alterado
Mandíbula	Abertura	normal/alterado
conversa espontânea	Desenho para paciente descrever n°:	respiração/articulação/ressonância
Leitura de monossílabos e de frases		
Plosiva	Banco, Tucano, Dedo, Panela	normal/alterado
	Porco, Gato, Batata, Tomate	
Plosivos e nasais, na emissão de palavras e fora	Cama, Caminhão, Balão, Pão, Mão	normal/alterado
Ligações consonantais e Fricativas	Janela, Vaso, Gilete, Vaca, Faca, Lanche, Sapo, Farinha, Chave, Chapéu, Fogão, Gema	normal/alterado
Vogais isoladas e vogais nas palavras	A E I O U, Meia, Pia, Bóia, Baú	normal/alterado
Líquidas	Lápis, Milho, Lua, Olho, Bolo, Ilha	normal/alterado
Encontros consonantais	Prato, Blusa, Flores, Fralda	normal/alterado
Diadococinesias (repetição espontânea – velocidade crescente)	Pataka, Badaga, Fasacha	normal/alterado
Precisão articulatória	0---1---2---3---4---5---6	(grau 0 – ininteligível; 6 - inteligível)
Articulação: Análise indica comprometimento de grau:	0---1---2---3---4---5---6	(grau 0 ausência; 6 grave)
5. Prosódia		
Entonação	Afirmção: É proibido fumar aqui; Interrogação: Você gostaria de comprar bolo ou sorvete?; Exclamação: Maria chegou!	normal/alterado
Velocidade		normal/alterado
Pausas na fala	0---1---2---3---4---5---6	(grau 0 – sem pausas; 6 muitas pausas)
Prosódia: Análise indica comprometimento de grau:	0---1---2---3---4---5---6	(grau 0 ausência; 6 grave)
Total Disartria	Leve	1 a 10 pontos
	Moderada	11 a 20 pontos
	Grave	21 a 30 pontos

Annex 2. Brazilian Portuguese version of Functional Assessment of Amyotrophic Lateral Sclerosis (ALSFRS-Re)

Paciente: _____		Data: ___/___/___	
AMYOTROPHIC LATERAL SCLEROSIS FUNCTIONAL RATING SCALE - REVISED (ALSFRS-R)			
1. Fala		7. Transferência na cama e arrumar roupa de cama	
4	Fala normal	4	Normal
3	Distúrbio de fala perceptível	3	Um pouco lento e desajeitado, mas não precisa de ajuda
2	Inteligível com repetição	2	Pode se transferir sozinho ou arrumar os lençóis, mas com grande dificuldade
1	Fala combinada a comunicação não verbal	1	Capaz de iniciar, mas não se transfere ou arruma o lençol
0	Perda da fala	0	Necessita de ajuda
2. Salivação		8. Andar	
4	Normal	4	Normal
3	Leve excesso de saliva na boca, pode ocorrer sialorréia noturna	3	Dificuldade de deambulação precoce
2	Saliva moderadamente excessiva; pode haver uma mínima sialorréia	2	Caminha com ajuda
1	Marcante excesso de saliva com sialorréia	1	Ausência de deambulação funcional
0	Sialorréia marcante, requer limpeza constante com uso de lenços de papel ou lenços de pano	0	Sem movimento intencional da perna
3. Deglutição		9. Subindo escadas	
4	Hábitos alimentares normais	4	Normal
3	Distúrbios precoces na alimentação - engasgo ocasional	3	Lento
2	Mudanças na consistência da comida	2	Instabilidade moderada ou fadiga
1	Necessita de tubo de alimentação suplementar	1	Precisa de ajuda
0	Ausência de alimentação oral (Alimentação exclusivamente parenteral ou enteral)	0	Não consegue subir
4. Escrita		10. Dispnéia	
4	Normal	4	Nenhuma
3	Lenta ou sinuosa; todas as palavras são legíveis	3	Ocorre quando caminha
2	Nem todas as palavras são legíveis	2	Ocorre em uma ou mais das seguintes atividades: alimentação, banho, vestuário (AVD)
1	Capaz de segurar a caneta, mas incapaz de escrever	1	Ocorre em repouso, dificuldade de respirar tanto sentado quanto deitado
0	Incapaz de segurar a caneta	0	Dificuldade significativa considerando o uso de suporte mecânico respiratório
5a. Cortando alimentos e manuseando utensílios (pacientes sem gastrostomia)		11. Ortopnéia	
	Normal	4	Nenhuma
	Um pouco lento e desajeitado, mas não precisa de ajuda	3	Um pouco de dificuldade ao dormir devido à falta de fôlego, não usa mais de dois travesseiros rotineiramente
	Capaz de cortar a maioria dos alimentos, embora seja lento e desajeitado, precisa de ajuda	2	Precisa de travesseiros extras para poder dormir (mais de dois)
	O alimento tem que ser cortado por alguém, ainda assim é capaz de se alimentar vagarosamente	1	Dorme somente sentado
	Precisa ser alimentado	0	Incapaz de dormir
5b. Cortando alimentos e manuseando utensílios (escala alternativa para pacientes com gastrostomia)		12. Insuficiência respiratória	
4	Normal	4	Nenhuma
3	Desajeitado, mas capaz de executar todas as manipulações independentemente	3	Uso intermitente do BIPAP
2	Precisa de alguma ajuda para fechar ou apertar objetos	2	Uso contínuo do BIPAP durante a noite
1	Da assistência mínima ao cuidador	1	Uso contínuo do BIPAP durante o dia e a noite
0	Incapaz de executar qualquer tarefa	0	Ventilação mecânica invasiva por intubação ou traqueostomia
6. Vestuário e higiene			
4	Função normal		
3	Cuidado pessoal independente e completo com esforço ou eficiência diminuída		
2	Assistência intermitente ou substituí métodos		
1	Precisa de assistência para cuidado pessoal		
0	Dependência total		
		TOTAL:	