



Quality of life of patients with Amyotrophic Lateral Sclerosis

Qualidade de vida de pacientes com Esclerose Lateral Amiotrófica

Sue Christine Siqueira¹, Priscila Valverde de Oliveira Vitorino², Cejane Oliveira Martins Prudente², Tainara Sardeiro de Santana¹, Gleydson Ferreira de Melo¹

Objective: to analyze the scientific evidence that evaluated the quality of life of patients with Amyotrophic Lateral Sclerosis through ALSAQ-40/ALSAQ-5. **Methods:** this is an integrative review carried out with eight articles in the database, using controlled keywords. **Results:** the most impaired domains of quality of life were physical mobility and activities of daily living. Patients with bulbar Amyotrophic Lateral Sclerosis, communication and feeding were more impaired. Problems with health care and lack of social support to the caregiver negatively impacted the patient's quality of life. Emotional stress was correlated with communication, physical mobility and disease progression in patients with breathing and artificial feeding, but it is not directly related to the advances of physical deterioration, but rather how the patient processes the experiences. **Conclusion:** patients with Amyotrophic Lateral Sclerosis have a poorer quality of life regarding the motor aspect, and lack of caregiver support worsens the patient's quality of life.

Descriptors: Quality of Life; Amyotrophic Lateral Sclerosis; Nursing.

Objetivo: analisar as evidências científicas que avaliaram a qualidade de vida de pacientes com Esclerose Lateral Amiotrófica por meio do ALSAQ-40/ALSAQ-5. **Métodos:** revisão integrativa realizada com oito artigos em base de dados, utilizando-se descritores controlados. **Resultados:** os domínios mais prejudicados da qualidade de vida foram mobilidade física e atividades de vida diária. Os pacientes com Esclerose Lateral Amiotrófica bulbar, comunicação e alimentação foram mais prejudicados. Problemas com a assistência à saúde e a falta de apoio social ao cuidador impactaram negativamente a qualidade de vida do paciente. Estresse emocional correlacionou com a comunicação, mobilidade física e progressão da doença em pacientes com respiração e alimentação artificial, mas não está diretamente relacionado aos avanços da deterioração física e sim como o paciente processa as experiências. **Conclusão:** pacientes com Esclerose Lateral Amiotrófica têm pior qualidade de vida referente ao aspecto motor; e a falta de suporte ao cuidador piora a qualidade de vida do paciente.

Descritores: Qualidade de Vida; Esclerose Amiotrófica Lateral; Enfermagem.

¹Faculdade Estácio de Sá de Goiás. Goiânia, GO, Brazil.

²Pontifícia Universidade Católica de Goiás. Goiânia, GO, Brazil.

Corresponding author: Sue Christine Siqueira

Rua 67-A, Quadra 140, nº216, Setor Norte Ferroviário - CEP: 74.063-320. Goiânia, GO, Brazil. E-mail: sue.siqueira@estacio.br

Introduction

Amyotrophic Lateral Sclerosis is a progressive neurodegenerative disease with involvement in upper and lower motor neurons, without cognitive and sensory impairment. It is associated with the death of the patient in a time of three to four years, usually due to respiratory failure. The disease affects the fibers along the corticospinal tract, which transmit impulses that control voluntary movements. These patients have progressive muscle weakness with a reserved prognosis, absent or diminished deep reflexes, fasciculations and spasticity⁽¹⁻³⁾.

The causes are multifactorial, such genetic factors (5 to 10.0%), excitotoxicity, mitochondrial damage, protein aggregation, apoptosis, immune system, muscle activity, smoking and occupation⁽⁴⁾. Its incidence is 1.4 to 1.6 per 100 thousand inhabitants⁽⁵⁾.

The progression of weakness, loss of physical independence, lack of a cure, fears about death and daily concerns of patients with a serious illness such as Amyotrophic Lateral Sclerosis influence the quality of life⁽⁶⁾.

The quality of life is the individual's perception of their position in life, in the context of the culture and value system in which they live and to their goals, expectations, standards and concerns⁽⁷⁾.

The quality of life is the satisfaction in living. Based on this concept, it is considered necessary for society in general to know the impact of Amyotrophic Lateral Sclerosis on the biopsychosocial aspect of the patient's life, since only public policies regarding the care of these patients can be developed⁽⁸⁾.

The quality of life can be measured by generic or disease-specific instruments. The ALS Amyotrophic Lateral Sclerosis Assessment Questionnaire (ALSAQ-40) and its brief form (ALSAQ-5) were developed as specific health status instruments, of a well-established standard in Amyotrophic Lateral Sclerosis since they were designed by deeply explanatory findings interviews of patients with Amyotrophic Lateral Sclerosis whose scores range from zero to one hundred.

The greater the score, the more impaired will be the quality of life of the patient⁽⁹⁾. ALSAQ-40 is a specific measure that assesses the particularities and concerns of patients with Amyotrophic Lateral Sclerosis, addressing the affected and impaired domains, covering important physical and emotional areas, which in a generic instrument would not be possible.

With this review, health professionals will be able to direct their activities to the mentioned aspects regarding the patient's perception of their quality of life.

Given the above, this article aimed to analyze the scientific evidence that evaluated the quality of life of patients with Amyotrophic Lateral Sclerosis by ALSAQ-40/ALSAQ-5.

Methods

This is an integrative review of the literature on the quality of life of patients with Amyotrophic Lateral Sclerosis. The following steps were taken: identification of the theme and definition of the problem; Establishment of criteria for inclusion of studies; Definition of the information to be extracted from the selected studies; Evaluation of studies; Interpretation of results and presentation of the review⁽¹⁰⁾.

The question was formulated to guide the review: what evidence does the quality of life of patients with Amyotrophic Lateral Sclerosis using the specific instrument of quality of life ALSAQ-40/ALSAQ-5?

The keywords were: quality of life and Amyotrophic lateral sclerosis. The search for articles was carried out from September 2014 to June 2016, in the Virtual Health Library (VHL), where the databases of Latin American and Caribbean Literature in Health Sciences (LILACS), International Literature in Health Sciences (MEDLINE), Cochrane Library and Scientific Electronic Library Online (SciELO), Spanish Bibliographical Index of Health Sciences (IBECS) and the United States National Library of Medicine (PubMed) were researched.

The inclusion criteria of the publications were:

a) occurring between 2004 and 2015; B) written in Portuguese or English; C) using the specific quality of life measurement instrument, ALSAQ-40 and/or ALSAQ-5. The exclusion criteria were: a) articles repeated in the search; B) articles of literature review, thesis or dissertation; C) qualitative studies; D) articles validating the ALSAQ-40 and ALSAQ-5 instruments.

A total of 828 articles were found, 386 of them were from the VHL and 442 from PubMed. After reading the titles and abstracts of the articles, they were accepted or rejected. The complete articles and after reading and translating the articles were searched, nine were selected. Eight completed the sample, six of MEDLINE and two of PubMed.

The evidence was classified into seven levels. Level I: systematic review or meta-analysis of randomized controlled trials; Level II: well-delineated randomized controlled clinical trials; Level III: well-delineated clinical trial studies without randomization; Level IV: well-delineated (non-experimental) cohort and case-control studies; Level V: studies of systematic review of descriptive and qualitative studies; Level VI: evidence from a single descriptive or qualitative study; And level VII: evidence from the opinion of authorities and/or expert committee reports⁽¹¹⁾.

The data were analyzed critically, seeking explanations for the results found by the authors. The main results were discussed, comparing the different

studies and identifying conclusions. The data referring to the year of publication, study design, and sample, results, and level of evidence are presented schematically in two figures for better visualization.

Results

All articles that were part of the sample are international and have been published in journals in the areas of neurology, medicine, psychology and specific journals of Amyotrophic Lateral Sclerosis. Seven of the eight articles are of European origin and one of Asian origin. Regarding the year of publication, it was heterogeneous, although they prevailed in 2010 and 2014 with two publications each. Three studies are cross-sectional⁽¹²⁻¹⁴⁾, three clinical trials⁽¹⁵⁻¹⁷⁾ and two longitudinal⁽¹⁸⁻¹⁹⁾.

Three studies aimed to describe or relate aspects that influence the quality of life of patients with Amyotrophic Lateral Sclerosis^(12,14,19), two studies used the ALSAQ-40 instrument as a comparative form of measurement^(13,18) and three studies used the ALSAQ-40 to verify the effect of an intervention⁽¹⁵⁻¹⁷⁾. As for the level of evidence, an article presented level II; two articles were level III, and five articles were level VI. Thus, the eight papers were selected for analysis and presented according to the study design, as shown in Figures 1 and 2.

Year	Design/Sample	Conclusion	Level
2010 ⁽¹²⁾	Cross-sectional/44 patients	The physical mobility domain was the most negatively impacted in the quality of life, followed by the daily life activity domain and the feeding domain had the least influence on the quality of life of the patient. The physical mobility domain was negatively correlated with low education.	VI
2012 ⁽¹³⁾	Cross-sectional/404 patients and 404 caregivers. Two groups: those who reported one or no problem with health and social services and those who reported two or more problems.	The physical mobility domain was the most impaired, followed by the Daily Life Activities. The food was the one that had the least negative impact. The patient's age and time since diagnosis did not have significant differences between the two groups. Four domains had significant differences between the two groups: physical mobility, daily life activities, feeding and emotional function.	VI
2014 ⁽¹⁴⁾	Cross-sectional/121 patients	The physical mobility domain was the most affected, followed by activities of daily living, communication, emotional function and the feeding domain. The more impacting feelings of the emotional function domain were: concern about the future, feeling of lack of freedom, concern about being a burden, and hopelessness with the future. This domain was impacted using measures of prolonged life.	VI

Figure 1 - Description of Cross-sectional Design research

Year	Design/Sample	Conclusion	Level
2010 ⁽¹⁵⁾	Randomized double-blind, cross-over/27 patients	The use of Tetrahydrocannabinol (THC) had no effect on the measures of quality of life.	III
2014 ⁽¹⁶⁾	Randomized controlled clinical trial/71 patients in the intervention group and 61 patients in the control group	The domain measured emotional function did not change over time. There was no significant difference in the values of the instrument between the intervention group and the control group.	II
2015 ⁽¹⁷⁾	Controlled multicenter clinical trial/103 in the intervention group and 97 in the control group.	There were no differences between groups in assessing the quality of life.	III
2008 ⁽¹⁸⁾	Longitudinal/24 patients	In patients with moderate Amyotrophic Lateral Sclerosis, a significant correlation was observed between the level of Chromogranin A (CgA) salivary and the Emotional Function domain score of the instrument.	VI
2013 ⁽¹⁹⁾	Longitudinal/110 patients	Spinal onset patients: mobility domain as the most impaired. Patients with bulbar onset: feeding and communication domains. In the domain of the emotional function: concerns about the future, being a burden, lack of freedom and hopelessness were the most impaired. In the second evaluation, there was a correlation between all domains and the domain emotional function, prevailing communication. In the third assessment, communication was the most impaired and the least were physical mobility and feeding.	VI

Figure 2 - Description of Longitudinal Design and Clinical Trials

Discussion

The analysis of the articles that were part of this study showed that the physical mobility domains and activities of daily living were the ones that most negatively impacted the quality of life of patients with Amyotrophic Lateral Sclerosis⁽¹²⁻¹⁴⁾ and Spinal Amyotrophic Lateral Sclerosis⁽¹⁹⁾. In the patients with Amyotrophic Lateral Sclerosis of bulbar onset, the domains feeding and communication were the most impaired, while the difficulties in the physical mobility affected the quality of life of these patients to a lesser extent, which is justified because the physical disabilities of the patients with Bulbar Amyotrophic Lateral Sclerosis and Spinal Amyotrophic Lateral Sclerosis are at different times during the course of the disease⁽¹⁸⁻¹⁹⁾.

Overall, nutrition⁽¹³⁻¹⁴⁾ and emotional function^(12,19) were the domains that had the least negative impact on quality of life.

The deficits found in the domains evaluated are related to the characteristics of Amyotrophic Lateral Sclerosis, as a degenerative disease of the whole motor neuron system, including limbs, bulb resulting in progressive paralysis of skeletal muscles, including speech, swallowing and respiratory muscles^(12-14,18-19). However, as the physical deterioration of the patient with Amyotrophic Lateral Sclerosis increases, there is a loss of independence, inability to communicate and demoralization. However, even though there is a greater emotional challenge, this domain remains stable⁽¹⁹⁻²⁰⁾.

With the diagnosis and evolution of the disease,

there is a negative impact on the patient's life and a change in routine. Amyotrophic Lateral Sclerosis causes progressive loss of autonomy and a greater need for assistance with activities of daily living, restructuring in eating, difficulties in communication, and impairment of the emotional state^(12-16,18-20).

Although the impairment is not significant in the quality of life of the patient with Amyotrophic Lateral Sclerosis, the emotional function is a domain studied by researchers, and it is a high priority for clinical care since there is evidence that emotional stress in patients with Amyotrophic Lateral Sclerosis is associated with an increased risk of mortality. Some feelings in this domain, such as being a burden, loss of freedom and hopelessness were the most affecting the quality of life of the patients, and hopelessness was the feeling that persisted during the evolution of Amyotrophic Lateral Sclerosis⁽¹⁹⁾. The feelings that affected the quality of life to a lesser degree were the anger of the disease and depression^(14,19).

In this way, emotional stress is directly related to the characteristics of the disease, the low life expectancy, not having a significant treatment, being incurable and having a rapid progression. These characteristics lead the patient with the progression of Amyotrophic Lateral Sclerosis to have negative feelings due to the loss of freedom and independence and the need to use supportive measures such as respirator and feeding. Although impairing quality of life in the physical mobility domain, these deficits do not negatively impact the emotional function domain of the patient with Amyotrophic Lateral Sclerosis^(14,18).

Depression is expected in most patients with Amyotrophic Lateral Sclerosis, as the disease's characteristics are confusing with somatic symptoms such as weight loss and fatigue. However, it is rare to find this feeling in patients, which is called depressive mood⁽²¹⁾.

There is a concern in assessing and analyzing the most impacts on the emotional function of the patient with Amyotrophic Lateral Sclerosis to subsidize an adequate planning of psychotherapeutic strategies. It is important to emphasize that the concern with the impact of physical disabilities on the emotional state of the patient encourages the studies about this aspect, most confirming that there is a weakness in the emotional function, but that it is not directly related to the physical aspects^(12-14,18).

Patients with Amyotrophic Lateral Sclerosis use medications to treat psychological symptoms, but the effects on the quality of life of these patients are limited, suggesting a greater emphasis on alternative therapies to minimize emotional stress, since it does not depend exclusively on physical damage caused throughout life, but rather as the individual processes in his/her mind and interprets lived experiences. That is, physical dysfunction alone is not the main contributor to emotional distress⁽¹⁸⁻¹⁹⁾.

The quality of life of the patient with Amyotrophic Lateral Sclerosis in a study carried out in Poland with 44 patients was not affected by sociodemographic factors such as age and gender and clinical factors as type of treatment. The low educational level has a negative impact on the physical mobility domain⁽¹²⁾. The domains of physical mobility, daily life activities, feeding and emotional function of patients with Amyotrophic Lateral Sclerosis obtained significant differences among the group of caregivers who reported having one or no problem with the provision of health care services and social support than the group who claimed to have two or more problems. In this same study, the patient's age and time since diagnosis did not have significant differences between the two groups. This is valid because the reduction of the quality of life of the patient is related to the number of problems reported by the caregivers and the hours

spent in the care provided⁽¹³⁾.

There were no changes in the result of ALSAQ-40 in 27 patients in a clinical trial comparing medication intervention and placebo^(15,17). In another case management study with 132 patients (71 in the intervention group and 61 in the control group), did not differ between the intervention group and the control group, besides not having changed over time, with the evolution of the disease⁽¹⁶⁾. Amyotrophic Lateral Sclerosis has its characteristics, and few interventions significantly change the course of the disease because it is a progressive neurodegenerative disease that affects motor neurons⁽²²⁻²³⁾.

The two studies that had an intervention⁽¹⁵⁻¹⁶⁾ showed that there were no significant differences in ALSAQ-40 values, mainly in the emotional function domain. Although the study authors have not discussed this result, it may be because the disease has a peculiar feature, that cognitive function remains intact in most cases. The patient is aware of the whole evolution of the disease, and the emotional aspects are individual because it depends on how each one processes their experience.

No articles of systematic review or meta-analysis of the quality of life of patients with Amyotrophic Lateral Sclerosis were found. We believe that due to the small number of randomized clinical trials published on the theme to the different methodologies adopted in the existing ones and to the low level of evidence of most of the studies found (level VI), there is a need to deepen the research on the topic.

The small sample of this review, using different methods to evaluate the quality of life in the studies found and low level of evidence in most publications were limitations.

Conclusion

This study identified factors that negatively impacted the quality of life of patients with Amyotrophic Lateral Sclerosis through the specific quality of life instrument for patients with Amyotrophic Lateral Sclerosis. The physical mobility domains and activities of daily living have impaired the quality of life. However, the feeding and communication domains were more impaired in patients with bulbar amyotrophic lateral sclerosis. Variables such as age, gender, and type of treatment had no relation to quality of life. Problems with health care and lack of social support to the caregiver are factors that negatively correlated with the patient's quality of life.

The emotional function domain was identified as being a priority in the life of the patient with amyotrophic lateral sclerosis, and it was more impacted by the difficulties found in communication and physical mobility. Being a burden, loss of freedom and hopelessness are feelings that have most affected the patient's emotional function, and anger and depression were the least impacts in the first year of the illness. With the progression of the disease, hopelessness persisted, and depression was less pronounced.

Collaborations

Siqueira SC, Vitorino PVO and Prudente CMO contributed to the project design, data collection, data analysis and interpretation, conception, article writing, critical review and final approval of the version to be published. Santana TS and Melo GF contributed to the data collection.

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