ScienceDirect

Neuroscience & Biobehavioral Reviews
Volume 111, April 2020, Pages 1-11

Quality of life and functional independence in amyotrophic lateral sclerosis: A systematic review

Jéssica Paloma Rosa Silva [a c], José Bomfim Santiago Júnior [c], Elizabete Lima dos Santos [c], Fernanda Oliveira de Carvalho [a b], Iandra Maria Pinheiro de França Costa [a], Deise Maria Furtado de Mendonça [c]

Show more

https://doi.org/10.1016/j.neubiorev.2019.12.032
Get rights and content

Abstract

Amyotrophic Lateral Sclerosis (ALS) leads to functional capacity decline, generating great impact in quality of life. Quality of life is directly related to physical integrity and functional independence. This systematic review aimed to analyze treatment protocols and their outcomes from clinical trials with focus on ALS rehabilitation that evaluated the effects on quality of life and functional independence from their intervention process. A literature search was conducted through MEDLINE-PubMed, Science Direct, Web of Science and Scopus databases. A total of 3630 articles were identified. Eleven studies met the inclusion criteria. They focused on different aspects of quality of life or functional independence, which are: respiratory care, mental health, communication skills and exercises. Use of bipap and inspiratory muscle training, anxiety and depression, communication devices implementation and exercises safety and tolerability were considered as key points. However, the drastic evolution of the disease is a limiting factor to the perception of quality of life improvement by patients. Further studies should be done to validate the benefits on patients' quality of life.

Introduction

Amyotrophic Lateral Sclerosis (ALS) is a neurodegenerative disease characterized by the degeneration and death of upper and lower motor neurons (Saberi et al., 2015; Hardiman et al., 2017). In most cases, patients evolve to death on average within 3–5 years after the onset of symptoms (Bendotti et al., 2016; Mehta et al., 2017). The disease does not have a defined etiology, but studies point to the possibility that the disease is multifactorial, and some environmental and genetic factors are mentioned (Bendotti et al., 2016). The disease does not have laboratory or imaging biomarkers for diagnosis. The diagnosis of ALS is made by means of clinical manifestations and complementary tests such as electroneuromiography (ENMG) (Brooks, 1994; Makki and Benatar, 2007). There is no cure for ALS and Riluzole is so far the only approved drug used in this population, however it only prolongs the patient's survival time in about 3 months (Miller et al., 2012; Hardiman et al., 2017; Schultz, 2018). Recently, Edaravone, a new drug resource with Riluzole-like action, has been approved for intravenous treatment, but there are still few descriptions of its efficacy in the literature (Rothstein, 2017; Dash et al., 2018; Okada et al., 2018; Schultz, 2018).

ALS symptoms are quite heterogeneous, with the most common symptoms being muscle weakness, spasticity, atrophy and fasciculations (Bandeira et al., 2010; Hardiman et al., 2017). The relentless decline of functional capacity with relative cognition and

sensitivity preservation is perceived in patients with ALS. It is possible to assume that patients with a progressive and disabling disease such as ALS are unable to enjoy living and invariably experience a decrease in their quality of life (Mello et al., 2009; Felgoise et al., 2010). Thus, it is of fundamental importance an appropriate multidisciplinary diagnosis, care and therapies with the purpose of providing greater functionality and consequent improvement in the quality of life of these patients (Felgoise et al., 2010; Morris, 2015).

ALS is part of a group of diseases that have a negative effect on muscle control, which has a dramatic adverse effect on the function and well-being of those with this diagnosis. The disease produces significant impacts in different spheres, such as economic, social, emotional and family life, affecting aspects of the functionality and quality of life of patients (Bandeira et al., 2010; Silva et al., 2014).

Quality of life is defined through the individuals' understanding of their lives and the context in which they are inserted, associated with their goals, expectations, standards and concerns broadly related to their physical and psychological conditions, independence level, social relations and personal beliefs (The WHOQOL Group, 1995) (Fig. 1). This concept is directly related to functional independence, which is characterized by the functional activities that patients point as fundamental for the maintenance of their physical and psychological well-being (Orient-López et al., 2006). All of which is carried out congruently through the integration of cognitive, behavioral, sensory and motor resources (Cesário et al., 2006; Mcnaughton et al., 2005). Considering the non-existence of a cure for ALS, patients need a specific assistance and care based on their clinical condition, accompanying every evolutionary manifestation of the disease, in a continuous scheme of therapeutic readjustment (Majmudar et al., 2014; Soofi et al., 2018).

In the rehabilitation process, the primary goal is to help patients to stay functionally active in a safe and effective way Hobson and McDermott, 2016. The process should positively impact their quality of life, although the main characteristic of the disease is the reduction of mobility with a consequent decline in daily life activity and instrumental activities of daily life. This decline interferes with the subject's self-care ability, which consists of the individual's ability to take care of themselves, providing benefits to their health and well-being and is directly linked to changes in the process of illness (Silva et al., 2014;)Hobson and McDermott, 2016.

During rehabilitation, the multidisciplinary team provides the patient with a comprehensive and individual practice (Majmudar et al., 2014). Preserving, maintaining, developing or restoring the integrity of systems or functions, as well as the maintenance of functionality and quality of life are the purposes of the rehabilitation process (Honorato and Martins, 2008). In order to do so, it is necessary to use specific evaluation instruments for the variables to ascertain the actual functional stage of the patient, as well as to understand the impact of this decline on quality of life and functional independence. Based on that, it is possible to prepare an individual, accessible and effective therapeutic plan (Cappellato et al., 2015).

The present study is based on a systematic literature review that aims to analyze clinical trials that evaluated treatment effects of rehabilitation programs, in its multidisciplinary concept, in functional independence and quality of life of patients. It was included only clinical trials that used validated instruments of measurement in their intervention process.

Section snippets

Material and methods

This study was conducted following the Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) statement and supplemented by guidance from the Cochrane Collaboration Handbook. A protocol of this systematic review was designed a priori and was registered in the PROSPERO database (registration number CRD42019115729).…

Study selection

The process followed for article selection is presented in Fig. 2. A total of 3630 articles were identified: 2045 articles on Science Direct, 468 on SCOPUS, 304 on WEB OF SCIENCE and 813 on PubMed. After deleting duplicate articles, we proceeded with the reading of 2166 titles and abstracts. 175 articles were selected for full reading. After the assessment of the articles not shown in full, the duplicates, the letters to the reader, the case studies, the articles that were not in English,…

Summary of findings

Despite the complexity of the disease for patient and family, few studies are found in the literature containing evaluation of quality of life and functionality of patients with ALS. The disease has a major impact on patients and families. The disease drastically changes the

course of life of all people involved, directly or indirectly (Majmudar et al., 2014). Gradually, decreasing mobility leads to difficulty in carrying out activities of daily living, and sometimes breathing, communication,…

Conclusion

Until now ALS has no cure and its treatment is bascically symptomatic. Some therapies slow down the progression of the disease and, consequently, increase survival. ALS patients have many needs, since the disease greatly compromises motor skills, communication and, sometimes, nourishment and breathing. Invariably, the condition conducts to enormous decrease in functional independence and quality of life. Quality of life is related to the patient's individual perception about himself/herself in…

Acknowledgements

This research was supported by Pró-reitoria de Pesquisa da Universidade Federal de Sergipe. (POSGRAP/UFS).…

References (56)

K. Sato *et al.*
Seven amyotrophic lateral sclerosis patients diagnosed only after development of respiratory failure
J. Clin. Neurosci. (2014)

P. Sanjuán-López *et al.*
Amyotrophic lateral sclerosis: impact of pulmonary follow-up and mechanical ventilation on survival. A study of 114 cases
Arch. Bronconeumol. (2014)

S. Saberi *et al.*
Neuropathology of amyotrophic lateral sclerosis and its variants
Neurol. Clin. (2015)

J.D. Rothstein
Edaravone: a new drug approved for ALS
Cell. (2017)

M. Okada *et al.*
Long-term effects of edaravone on survival of patients with amyotrophic lateral sclerosis
eNeurologicalSci. (2018)

H.K. Mcnaughton *et al.*
Functional measures across neurologic discase states: analysis of factores in common
Archives Physical Medicine and Rehabilitation, Reston. (2005)

T. Makkonen *et al.*
Oral motor functions, speech and communication before a definitive diagnosis of amyotrophic lateral sclerosis
J. Commun. Disord. (2016)

C. Jenkinson *et al.*
The amyotrophic lateral sclerosis assessment questionnaire (ALSAQ-40): tests of data quality, score reliability and response rate in a survey of patients
J. Neurol. Sci. (2000)

B.R. Brooks
El Escorial World Federation of Neurology criteria for the diagnosis of amyotrophic lateral sclerosis. Subcommittee on Motor Neuron Diseases/Amyotrophic Lateral Sclerosis of the World Federation of Neurology Research Group on Neuromuscular Diseases and the El Escorial “Clinical limits of amyotrophic lateral sclerosis” workshop contributors
J. Neurol. Sci. (1994)

F.M. Bandeira *et al.*
Evaluation of the quality of life of patients with Amyotrophic Lateral Sclerosis (ALS) in Brasília
Rev. Neurocienc. (2010)

View more references

Cited by (16)

Efficient machine learning approach for volunteer eye-blink detection in real-time using webcam
2022, Expert Systems with Applications

> *Citation Excerpt :*
> …This set of symptoms characterizes, among other diseases, Amyotrophic Lateral Sclerosis (ALS) (Barbalho et al., 2021; Kiernan et al., 2011; van Es et al., 2017) whose patients are potential users of this work. The quality of life of patients affected by ALS is directly related to their communication capacity (Fernandes et al., 2021; Rosa Silva et al., 2020; Rousseau et al., 2015). Considering that ALS patients are able to blink even in advanced stages of the disease and the interest in enhancing their communication capacity, this work presents a low-cost and real-time eye-blink detector system to be used as an activation and control tool for Alternative Communication Systems (ACS)….

Show abstract

Effectiveness of Nonpharmacological Interventions in the Field of Ventilation: An Umbrella Review
2023, International Journal of Environmental Research and Public Health

Phase angle is related to physical function and quality of life in preoperative patients with lumbar spinal stenosis
2022, ResearchSquare

Muscular Response in ALS Patients during Maximal Bilateral Isometric Work of the Biceps Brachii until Fatigue
2022, Life

International depiction of the cost of functional independence limitations among older adults living in the community: a systematic review and cost-of-impairment study
2022, BMC Geriatrics

Phase angle is related to physical function and quality of life in preoperative patients with lumbar spinal stenosis
2022, medRxiv

View all citing articles on Scopus

Recommended articles (6)

Research article

Disentangling the molecular mechanisms of multiple sclerosis: The contribution of twin studies
Neuroscience & Biobehavioral Reviews, Volume 111, 2020, pp. 194-198

Show abstract

Research article

Odors: from chemical structures to gaseous plumes
Neuroscience & Biobehavioral Reviews, Volume 111, 2020, pp. 19-29

Show abstract

Research article

ALS: Management Problems

Neurologic Clinics, Volume 38, Issue 3, 2020, pp. 565-575

Research article

Pain in amyotrophic lateral sclerosis

The Lancet Neurology, Volume 16, Issue 2, 2017, pp. 144-157

Show abstract

Research article

Nonmotor Symptoms in Amyotrophic Lateral Sclerosis: A Systematic Review

International Review of Neurobiology, Volume 134, 2017, pp. 1409-1441

Show abstract

Research article

Care management in amyotrophic lateral sclerosis

Revue Neurologique, Volume 173, Issue 5, 2017, pp. 288-299

Show abstract

View full text