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Palliative Care in Amyotrophic Lateral Sclerosis: A View of Comprehensive Care

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Abstract— *Objective: Discuss through a literature review about the need for palliative care for patients with amyotrophic lateral sclerosis (ALS). Method: An integrative literature review was adopted, since it contributes to the process of systematization and analysis of results, aiming at the understanding of a certain topic, based on other independent studies. This is a study with a qualitative approach, to identify productions on the topic of palliative care in patients with ALS. Results: The involvement of palliative care clinics composed of doctors, nurses, physiotherapists, speech therapists, psychologists and nutritionists promote programmed and continuous actions that can increase survival and improve comfort at the end of life in ALS. The integration of palliative care into health systems benefits the user by increasing access to this care and can also strengthen health systems, as it results in benefits that spread and improve the quality of care. Final Considerations: The care of ALS patients is complex, however, as the disease burden increases, access to these specialist clinics can become difficult or impossible for the patient. Understanding the current multidisciplinary team paradigm to include collaborations with local providers, including palliative care physicians and allied healthcare providers, can ensure that patient needs are met throughout the course of illness, especially during terminal illness.*

I. INTRODUCTION

Amyotrophic lateral sclerosis (ALS) is a degenerative neuronal disease with a highly predictable clinical course, progressive and relentless in nature. Due to its clinical characteristics, ALS represents a paradigm for palliative care in neurological diseases, so that it should start at the moment or soon after diagnosis.[1]

ALS also affects the muscles used for breathing, speech, and swallowing, which can cause physical discomfort and emotional pain. Most people with the disease worry about how this loss will make them dependent on others for even the most common tasks, such as bathing, dressing or eating.[2]

Care is intensified when symptoms gradually increase, associated with disease progression. End-of-life care for all disorders is associated with increased use of health services. Palliative services are associated with fewer hospital and intensive care unit (ICU) visits and more cost-effective care.[3]

Palliative care (PC) consists of assistance, promoted by a multidisciplinary team, which aims to improve the quality of life of the patient and their family members of a disease that threatens the continuity of their life, through the prevention and relief of suffering, early identification, relentless assessment and treatment of pain.[4]

The objective of this research was to discuss, through a literature review, about the need for palliative care for patients with ALS.

II. METHOD

This is a study with a qualitative approach, to identify productions on the topic of palliative care in patients with ALS. An integrative literature review was adopted, since it contributes to the process of systematization and analysis of results, aiming at the understanding of a certain topic, based on other independent studies.

The integrative literature review proposes the establishment of well-defined criteria for data collection, analysis and presentation of results, from the beginning of the study, based on a previously elaborated and validated research protocol. [5] The focus in the elaboration of the research was to understand the findings on palliative care for patients with ALS, and to highlight which care would be important, with a focus on hospital care.

For that, the six steps indicated for the constitution of the integrative literature review were adopted: 1) selection of the research question; 2) definition of inclusion criteria for studies and sample selection; 3) representation of the selected studies in table format, considering all common characteristics; 4) critical analysis of the findings, identifying differences and conflicts; 5) interpretation of results and 6) clearly reporting the evidence found.

The study identification and selection strategy was the search for publications indexed in the Medical Literature and Retrivial System on Line (MEDLINE), Latin American and Caribbean Health Sciences (LILACS) and Scientific Electronic Library Online (SciELO) databases, in April 2021.

The following criteria were adopted for the selection of articles: all article categories (original, literature review, reflection, update, experience report, etc.); articles with abstracts and full texts available for analysis; those published in Portuguese and English, between 2017 and 2021, and articles that contained in their titles and/or abstracts the following descriptors in health sciences (DeCS): Amyotrophic Lateral Sclerosis; Palliative care; Comprehensive care. The resource used in the research was the expression “exact term”, associated with specific descriptors.

The exclusion criterion for the articles was: studies that did not meet the aforementioned inclusion criteria. In the search in the databases, 263 articles were found talking about Amyotrophic Lateral Sclerosis, related to palliative care were only 32, of which each abstract/article was carefully read, highlighting those that responded to the objective proposed by this study, the in order to organize the findings.

Following the inclusion criteria, 16 studies were selected for analysis, which are referenced in this text. A bibliometric analysis was carried out to characterize the selected studies. Subsequently, the concepts addressed in each article and of interest to the researchers were extracted.

III. RESULTS AND DISCUSSION

In 2014, the World Health Organization (WHO) published the Global Atlas of Palliative Care, where it presented the results of its survey on the development of palliative care around the world. Brazil received a 3 A rating, which means that palliative care is limited in terms of networking, with funding sources heavily dependent on donations; limited availability of morphine; and a small number of palliative care services compared to the size of the population.[6]

According to The Economist (2017) who evaluated the quality of death in 80 countries. Assessed availability of access to opioids, existence of specific public health policies on palliative care and access to palliative care in health services. Brazil ranked 42nd, a ranking worse than Chile (27th), Costa Rica (29th), Panama (31st), Argentina (32nd), Uruguay (39th), South Africa (34th), Uganda (35th), Mongolia (28th) or Malaysia (38th). [7]

In 2017 the Lancet Commission on Access to Opioids and Palliative Care published results of their worldwide survey, the results were frightening. It has been shown that Brazil suffers from a shortage of opioids. The amount of opioids prescribed in the country is insufficient to provide adequate pain relief that seriously ill patients living in the country have. The common conclusion among all these studies is the evidence that in Brazil it is common for people with serious illnesses to often live with uncontrolled pain and without receiving adequate palliative care. [8]

ALS is accompanied by progressive muscle weakness, accompanied by muscle atrophy, fasciculations, muscle cramps, and slowness of movement with muscle stiffness.[9] The onset of muscle weakness in ALS is usually focal and typically spreads to adjacent body regions. This pattern is compatible with the spread of disease pathology within the motor system, with neuroanatomical spread within the spinal cord and motor cortex segments.[1]

Motor neurons run from the brain to the spinal cord and from the spinal cord to muscles throughout the body, the progressive degeneration of motor neurons in ALS eventually leads to their death. When motor neurons die, the brain's ability to initiate and control muscle movement is lost, with voluntary muscle action progressively affected, people may lose the ability to speak, eat, move, and breathe. [10]

The response to their needs is usually delayed and largely based on institutional palliative care focused on cancer. There is a great need to identify these patients and integrate an early palliative approach according to their individual needs in all settings, as suggested by the WHO.[11]

The involvement of palliative care clinics composed of doctors, nurses, physiotherapists, speech therapists, psychologists and nutritionists promote programmed and continuous actions that can increase survival and improve comfort at the end of life in ALS. The integration of PC into health systems benefits the user by increasing access to PC and can also strengthen health systems, by strengthening and improving the quality of care. [12]

Patients with ALS have a some advantages in the decentralized PC service of hospitals should be highlighted: early dehospitalization; cost reduction; training family members with programs that facilitate alternative non-verbal communication; management of total or partial oral supply or suspension of food, maintaining oral pleasure; possibility of an interdisciplinary team trained to monitor the clinical condition of patients; respiratory comfort with adequate ventilation patterns; monitoring of nutritional and metabolic status.[13]

In some patients, muscle weakness is preceded by a period when fasciculations, muscle cramps, or slight weight loss have been observed. The progressive and relentless nature of amyotrophic lateral sclerosis (ALS) culminating in respiratory failure makes end-of-life care planning and implementation an integral component of caring for ALS patients.[13]

Traditional models of palliative care do not sufficiently address the specific needs of patients and families living with a neurological diagnosis. In addition, palliative physicians who are extensively trained in internal medicine may be less comfortable with neurological diseases than with cancer or other medical diagnoses.[13]

Patients with limiting neurological diseases usually have a long and variable disease progression, punctuated by cognitive impairment, behavioral and communication problems, in addition to motor symptoms.[9] This trajectory differs from the sharp decline seen in many cancer patients. Palliative care services are increasingly recognizing the needs of cancer-free patients, particularly in rapidly progressing neurological conditions such as motor neuron diseases, however this growth is gradual.[11]

Differences between neurology and other patients include symptom profiles, psychosocial issues, caregiver needs, and effects on spiritual well-being. As examples, patients with motor neuron disease experience more demoralization, hopelessness, and suicidal ideation than patients with metastatic cancer, patients with brain cancer have distinct symptom profiles, including more cognitive problems, seizures, and communication deficits than patients with brain cancer living with other types of cancer; and Huntington's disease patients have distinct social work needs as a result of combined behavioral, psychiatric, movement, and cognitive issues.[14]

Physical and cognitive impairments associated with neurological diseases also contribute to the feeling of being “useless” or a “burden” and may contribute to higher rates of demoralization.[22] Neurological diseases are associated with caregiver suffering and reduced quality of life related to care, well-being, depression and demoralization.[15]

The concept of adequate neuropalliative care is defined by Sutton (2008) as “a holistic approach to the care of neurological patients with significant disabilities, complex needs and a potentially shortened life expectancy. It is patient-centered and involves diagnosing clinical problems at all stages, rehabilitation to maintain function, coordination of care, and adequate palliation to alleviate symptoms.”[16]

For neurological patients, the most appropriate forms seem to be the so-called “early” palliative care and the “dynamic model” of palliative care.[13] Early palliative care does not exclude the simultaneity of curative care, the intensity of palliative care services gradually increasing as the disease progresses.[17]

In the dynamic model, specialized palliative care services are provided based on trigger points, that is, in intermittent periods according to the needs of patients. [18] The objective of palliative care at all stages of the disease should be to achieve the ideal quality of life for patients and their families.[19]

On the other hand, specialized palliative care must be provided in the form of complex interventions that positively impact the quality of life of patients and their families, reduce the burden of physical symptoms and have a positive effect on psychosocial and spiritual issues, individual preference for care, and the quality of death and dying.[20] The analysis of patients' subjective perception of the impact of the disease and treatment on activities of daily living, self-care capacity, emotional experiences, social relationships and level of anxiety, tension and depression is very important for the evaluation of the quality of care provided to patients with ALS.[21]

Based on a systematic review and meta-analysis, the PC intervention is associated with improved quality of life for patients with life-limiting disease and an attenuated symptom burden. However, they found no connection between palliative care and survival. Few studies have been published analyzing the positive effects of neuropalliative care on the quality of life of ALS patients and their family caregivers, or on satisfaction with the care provided.[22]

Several studies have indicated an improvement in quality of life and a reduction in pain, dyspnea, sleep disturbances and intestinal symptoms, after the application of palliative care in patients with amyotrophic lateral sclerosis, multiple sclerosis, Parkinson's disease, neurodegenerative diseases and, in addition, a reduced burden for family caregivers.[20]

A randomized trial confirmed the financial efficiency of early palliative care in patients with multiple sclerosis.[23] Additional research to focus are recommend on the effect of complex, specialized palliative care interventions on all aspects of the quality of life of ALS patients.[24]

IV. FINAL CONSIDERATIONS

Palliative care are practices aimed at the final period of life of patients outside of therapeutic cure procedures, such practices seek to control the physical and psychological signs and symptoms of the advanced stage of ALS, in addition to improving the quality of life.

The care of ALS patients is complex, however, as the disease burden increases, access to these specialist clinics can become difficult or impossible for the patient. Extending the current multidisciplinary team paradigm to include collaborations with local providers, including local palliative care physicians and allied healthcare providers, can ensure that patient needs are met throughout the course of illness, especially during terminal illness.

Much can be done to mitigate the disease burden of ALS patients through the use of palliative interventions. There is considerable evidence that PC intervention improves the quality of life of patients and caregivers. Although new treatments for ALS are being developed, they are not curative and only offer the potential to slow its progression.

Therefore, both the multidisciplinary palliative care team and the neurology team are essential to provide a high standard of care and allow the maintenance of quality of life.

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