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CHARACTERISTICS OF FAMILY MEMBERS' CARE IN NURSING CARE OF PATIENTS WITH AMYOTROPHIC LATERAL SCLEROSIS

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Annotation. Amyotrophic lateral sclerosis is a rapidly progressive and fatal neurodegenerative disease. The relative number of patients throughout the population with this disease is small. There is no effective way to treat the disease or to halt its progression, therefore only symptomatic treatment is available and continuous nursing care of relatives is essential. Around 50% of the patients die within 30 months of the onset of symptoms, often as a result of respiratory failure, and around 10% may survive for more than a decade. Amyotrophic lateral sclerosis is characterised by degeneration of the upper and lower motor neurons and frontotemporal systems. Although the cause of the disease is largely unknown, 10-20% of the cases have a certain genetic aetiology.

Keywords: amyotrophic lateral sclerosis, neurodegenerative disease, nursing care

INTRODUCTION

Neurodegenerative diseases affect around one billion people worldwide, and the root causes of these diseases are unclear. Neurodegenerative diseases can occur due to a combination of genomic, epigenomic, metabolic and environmental factors. In the context of progressive neurodegenerative processes and neuroinflammation, no therapy or treatment can slow down or stop the progression of these diseases. Amyotrophic lateral sclerosis, multiple sclerosis, Parkinson's, Alzheimer's and Huntington's diseases can severely affect patient's daily life (Ruffini et al., 2020). Neurodegenerative diseases are chronic diseases that damage nerve cells in the brain and cause memory loss, behavioural, speech and mobility disorders and have similar cellular and molecular development mechanisms. According to Geevasinga et al. (2016), oxidative stress is considered to be one of the most important pathophysiological mechanisms underlying neurodegenerative diseases.

One of the most rapidly progressive neurodegenerative diseases is amyotrophic lateral sclerosis (hereinafter referred to as ALS) (Benatar et al., 2019). The name of the disease refers to the changes that occur in a human body when it strikes. "Amyotrophic" refers to the atrophy of a person's muscles; "lateral sclerosis" refers to the presence of sclerotic changes in the lateral parts of the spinal cord. ALS is a neurological disease that affects the neurons involved in the transmission of nerve impulses to muscles (motor neurons) and is classified under motor neurone disease group.

Different number of ALS cases is observed in various countries around the world. The age of people with ALS also varies between countries. The average age of patients at the time of diagnosis varied between 54 and 66 years. This high prevalence is thought to be due to the higher proportion of older people (Longinetti & Fang, 2019).

The aim of the study is to assess the characteristics of family members' care in nursing care of patients with amyotrophic lateral sclerosis.

The object is the characteristics of family members' care in nursing care of patients.

The objectives of the study are as follows:

1. To analyse the factors of amyotrophic lateral sclerosis onset in amyotrophic lateral sclerosis patients and the basis for its management based on scientific literature.
2. To identify the challenges experienced by family members in nursing care of patients with amyotrophic lateral sclerosis.
3. To identify the major nursing care factors in the care of patients with amyotrophic lateral sclerosis.

Research methods: 1. Analysis of scientific literature. 2. Qualitative research: unstructured interviews. 3. Descriptive analysis of the data collected.

THE CONCEPT OF NEURODEGENERATIVE DISEASE: LITERATURE OVEREVIEW

ALS is a rare and progressive neurodegenerative disease that affects nerve cells in the brain and spine. ALS is a fatal neurodegenerative disorder that affects adults and is characterised by degeneration of both upper motor neurons in the primary motor cortex and lower motor neurons in the brainstem and spinal cord. Initially, symptoms of ALS manifest in muscle atrophy and weakness. This is often followed by the development of progressive paralysis of the voluntary muscles and eventually of the respiratory muscles. Corcia et al. (2017) observed that the initial onset of ALS usually develops in the limbs, masticatory and swallowing muscles, spreading into other areas of the body, and resulting in dysfunction of respiratory muscles. The majority (more than 90%) of ALS cases are of the sporadic form, while a small proportion (around 5-10%) are of the familial form. The onset of the disease is usually manifested by weakness in the limbs, called spinal onset, or difficulty with speech or swallowing, called bulbar onset. Other forms of ALS may also be observed in patients, including mixed spinal and bulbar form, chest onset, thoracic or dementia symptoms, respiratory

symptoms, thoracic or respiratory symptoms, or cognitive change (Longinetti & Fang, 2019). Scientific progress in genetic research has led to the identification of genes contributing to the pathogenesis of this disease. However, there is currently no effective treatment for patients with ALS.

It has been observed that the majority of ALS patients die of respiratory failure, usually within 3-5 years of the onset of signs and symptoms (Longinetti &Fang, 2019). Recent studies also emphasise that longer survival is associated with male gender, younger age at the onset of disease and diagnosis, higher body mass index and weight gain following diagnosis. Tracheostomy is another factor prolonging the patient's life. It leads to a significant increase in survival time of patients. Benjaminsen et al. (2018) demonstrated in their study that the median survival from the symptom onset to death can be longer than five years if tracheostomy was performed on ALS patients.

SYMPTOMS OF AMYOTROPHIC LATERAL SCLEROSIS AND THE DEMAND FOR NURSING CARE IN PATIENTS WITH AMYOTROPHIC LATERAL SCLEROSIS

ALS is a neurodegenerative disease characterised by progressive muscle paralysis. Benatar et al. (2022) note that men are slightly more likely to suffer from this disease than women. The highest incidence of the disease is observed between 58 and 63 years of life, and only about 5% of cases are diagnosed before the age of 30 years. The concept of ALS identifies two phases of the disease: asymptomatic and symptomatic. The asymptomatic (clinically silent) phase begins at the onset of the disease, but it is not identifiable at that time. ALS is a progressive neurodegenerative disease involving both upper and lower motor neurons, leading to paralysis and eventually death. Symptomatic treatments, such as suppression of salivation, relief of muscle cramps and relief of spasticity and pain, still play an important role in improving the quality of life (Xu et al., 2021). Benatar et al. (2022) report that in some patients, the clinical onset of ALS is preceded by a prodromal phase characterised by non-specific symptoms (e.g. muscle cramps, reduced exercise tolerance), signs (e.g, fasciculation, isolated loss of ankle reflexes, diffuse hyperreflexia), abnormalities (e.g. positive sharp waves in the muscles of one limb or in the thoracic paraspinal muscles) in the absence of progressive muscle weakness. As a sign of progressive disease, patients may develop limb spasms that affect hand dexterity, balance and gait. Early symptoms of ALS may involve muscle twitching, cramps, stiffness or weakness, slurred speech and/or difficulty in chewing or swallowing. As the disease progresses, people may develop weakness and eventually become dependent on a wheelchair.

Nursing care has always been focused on preserving health, comfort and providing care. Nursing care addresses a wide range of patient needs, from the most complex procedures to the simple act of gently taking a patient's hand and giving him or her reassurance. The main purpose of nursing care is to meet physical, spiritual, emotional and social needs of patients. Therefore, nursing care is about caring for others. Chronic progressive neurological diseases require integrated palliative care. Advance planning of care is an integral part of this process (Seeber et al., 2019). The severity of illness and uncertainty about the timing of disability and the evolving burden of care cause significant stress not only for the affected individuals but also for their families. Upon hearing a diagnosis of ALS, the lives of everyone around patients change and health care providers themselves are faced with the enormous task of caring for these individuals affected by ALS (Bromberg et al., 2011). Family members of patients with ALS are very much involved in the treatment process from the earliest stages of the disease and take on a huge share of the responsibility in helping patients.

ANALYSIS OF THE STUDY RESULTS

In order to achieve the aim and objectives of the study, a qualitative research method, unstructured interviews, was selected. This method allows for a better assessment of a person's experiences and emotions. The qualitative research was aimed at describing and understanding how different people involved in the study understand the phenomenon under investigation, what meanings they give to the phenomenon, and disclose the characteristics of nursing care of a severely ill person. As the survival time of people with ALS is limited, interviews were conducted with relatives of patients who were with their loved one during their illness. All respondents were consulted in advance of the scheduled interview. Out of 10 contacted people, only 5 agreed to take part in the study. The study was carried out with the consent to use the information obtained for scientific purposes, ensuring the anonymity of the respondents and confidentiality of the information, respecting the ethical principles (goodwill, privacy, voluntary nature, respect and fairness) and not violating the participants' rights.

The sample of the study consists of 5 respondents: 3 female and 2 male. The age of respodents participating in the study ranged from 45 to 55 years old, with an average age of 48 years. All respondents had a higher education degree. The respondents had been providing nursing care as follows: two for 1 year, two for 5 years and one for 7 years. The age of the patients was between 49 and 79 years, with an average age of 63 years.

The analysis of the study is linked to the knowledge of the most important factors in the situation. The questions and challenges raised relate to the search for meaning and a new approach to certain situations.

The interview was structured into 4 categories and 7 sub-categories.

Category 1: **Experiences in the care of patients with amyotrophic lateral sclerosis.** Family members face a variety of problems related to nursing care. They have different experiences, positive and negative emotions and gain new experience. Uncertainty, fear, physical and emotional fatigue are common symptoms, experienced not only by

the patient but also by the entire family. Caregivers suffer from psychological stress in the form of depression, anxiety and sleep disorders. Psychological support for patients and family members is essential.

Category 2: **Interventions to promote independence.** Caregivers say that the disease is slowly taking away the patient's capacity to function, but the mind remains lucid and all skills are lost. Therefore, assistance from a variety of specialists is required: physiotherapists, occupational therapists, speech therapists and dieticians. A properly adapted home environment is essential for comfortable nursing care and better quality of life of the patient. However, with the rapid progression of the disease, it is often impossible to continuously change the environment according to the needs of the patient. A special feeding table, a bathroom bench, a toilet chair and many other items are required, as well as armrests for comfortable and safe access at home. If the person being cared for walks, mobility aids such as a walker, a cane must be provided, and the layout of the room where the person lives must be adapted to allow safe movement. If the patient is bedridden, a hydraulic bed is required, and the position of the patient must be changed to prevent pressure sores, he or she must be fed, washed, dressed, hygiene products must be changed, and the patient must be given or administered medication at the right time.

Category 3: **Contribution of the family to supporting the patient with amyotrophic lateral sclerosis**. The help and support of close relatives is an important factor. It covers sharing information or experiences, sharing tasks, supporting each other psychologically, spending meaningful time with the patient. The family provides security, support and care. The most important step is regular health monitoring.

Category 4: **Self-awareness**. Information can be sought from information dissemination means, most of them in English, however there is no specific information on how to increase self-care for patients and family members caring for them (self-awareness means personal freedom, the pursuit for improvement, the pursuit of the individual's potential and desires). Mental health and self-awareness are key factors determining the effectiveness of nursing care. Initial information should always be provided by a doctor. All caregivers, whether a nurse or a family member, experience stress to a lesser or greater extent. Engaging in positive activities, sports, work that a person loves, can prevent burnout. There are also meditations and prayer, as well as inner dialogues and conversations with psychologists (see Figure 1).

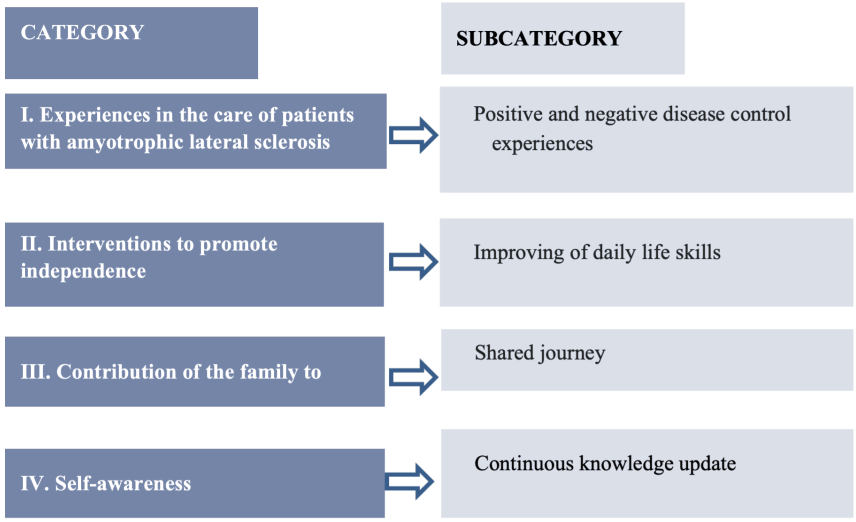

Figure 1. **Instrument for the Analysis of Characteristics of Nursing Care Provided by Family Members**

The key need is to adapt quickly to the patient's changing deterioration in condition, as social needs and skills disappear. Challenges, which are divided into three groups and are essential for meeting the needs of the patients, are outlined below:

Domestic (household) challenges are faced in the provision of suitable accommodation for the comfortable care of a seriously ill patient, special equipment, and a comfortable vehicle for transporting the patient.

Physical challenges include heavy strain (caring for a sick person, having a formal employment), and physical exhaustion.

Psychological challenges include uncertainty, fear, anxiety, sleep disturbances, emotional exhaustion, and depression (see Figure 2).

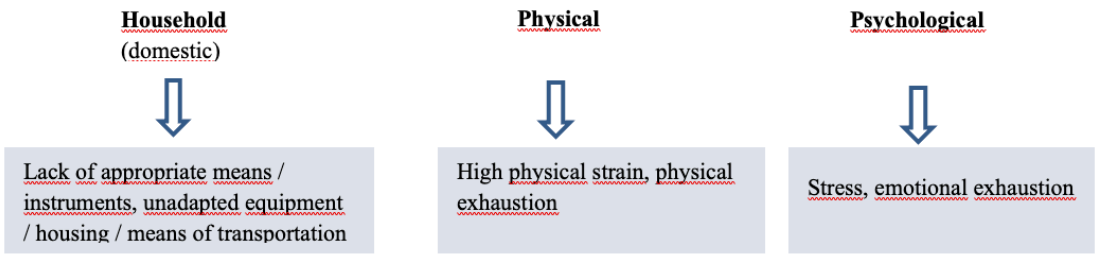

Figure 2. **Challenges Faced by Family Members in Nursing Care of Patients with Amyotrophic Lateral Sclerosis**

The family members help to adapt to the changed living conditions, adapt the environment to meet the patients' needs and try to give meaning to their lives. Adaptation to the disease is determined by personal qualities and the interaction of the support system (family, friends, various medical institutions, caregivers), while psychological adaptation is affected by internal (age, heredity, gender, other illnesses) and external factors (stress, harmful habits, diet, physical fitness) affecting the patient and family members. The most important are as follows:

Multidimensional nursing care support includes the provision of comfortable accommodation adapted to the patient's needs, adaptation of the transportation vehicle, provision of hygiene and nursing care products, and the provision of necessary information about the illness through the support of relatives.

Continuous knowledge update includes access to the necessary information and cooperation with the doctors.

Regular health monitoring means that the patient should never be left alone, his or her condition should be constantly monitored, the necessary tests should be carried out, and the necessary medicines should be taken and/or administered.

Consistent and good quality care includes providing appropriate and comfortable conditions for nursing care, and consistent and high quality care (see Figure 3).

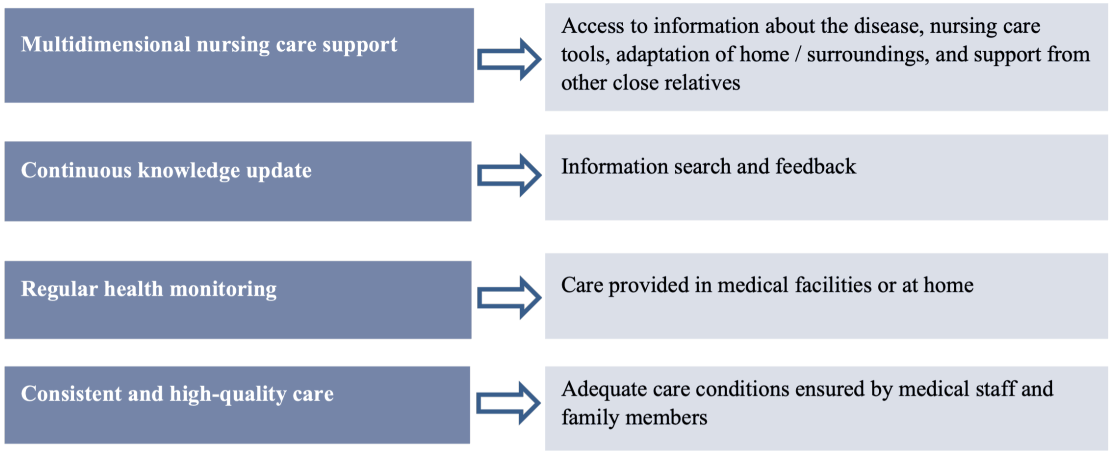

Figure 3. **Key Nursing Care Factors in Nursing Care of Patients with Amyotrophic Lateral Sclerosis**

In summarising all ALS cases, it was revealed that, based on the respondents' accounts, the course of the disease varies for all patients, although the outcome of the disease is unfortunately identical. It is evident from the data obtained that there are differences in the onset and course of the disease, as well as in the medical treatment used. It is probable that the caregivers' emotions and reactions to the disease may also differ.

CONCLUSIONS

• Amyotrophic lateral sclerosis is a rapidly progressive and incurable disease, with oxidative stress and heredity being the most common factors of its onset. There is currently no available cure, and symptomatic and palliative care form the basis for disease management.

• It has been disclosed that the disease causes many inconveniences and a variety of domestic and psychological challenges for the patient and the caregiver, and therefore requires disease management and improvements in daily living skills.

• Nursing care support, continuous knowledge update, regular health monitoring, and consistent and high-quality care are essential to sustain the patient's vital functions as long as possible.

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