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Are there nursing questionnaires for neurological deterioration in Amyotrophic Lateral Sclerosis?

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Abstract

Objective: to identify the existence of validated measurement scales that help us predict the existence of neurological damage of the respiratory and digestive origin. Methods: a bibliographic review was carried out in December 2019 in the main databases. Results: arti-cles are included that describe instruments for the early detection of neurological change and quality of life in patients with Amyotrophic Lateral Sclerosis. 15 articles are included that provide us with information on the eight most important and validated questionnaires to assess the progression of amyotrophic lateral sclerosis (EuroQoL, SF-12, SPB, ALSFR, ALSFRS-R, NPS, DMN, ALSAQ40). Conclusions: Despite the constant work to improve the quality of life and dignity of patients and their families, there is currently no validated question-naire that predicts the existence of respiratory and digestive neurological risk.
Key-words: Amyotrophic lateral sclerosis. Questionnaires. Digestive. Respiratory. Neurological.

¿Existen cuestionarios de enfermería para predecir el deterioro neurológico en la Esclerosis Lateral Amiotrófica?

Resumen

Objetivo principal: identificar la existencia de escalas de medición validadas que nos ayuden a predecir la existencia de deterioro neurológico del origen respiratorio y digestivo. Metodología: se realizó una revisión bibliográfica en diciembre de 2019 en las principales bases de datos. Resultados principales: se incluyeron artículos que describen instrumentos para la detección precoz del deterioro neurológico y la calidad de vida en pacientes con Esclerosis Lateral Amiotrófica. Se incluyeron 15 artículos que nos brindaron información sobre los ocho cuestionarios más importantes disponibles y validados para evaluar la progresión de la esclerosis lateral amiotrófica (EuroQoL, SF-12, SPB, ALSFR, ALS-FRS-R, NPS, DMN, ALSAQ40). Conclusión principal: a pesar del trabajo constante para mejorar la calidad de vida y la dignidad de los pacientes y sus familias, actualmente no existe un cuestionario validado que prediga la existencia de deterioro neurológico respiratorio y digestivo.
Palabras clave: Esclerosis lateral amiotrófica. Cuestionarios. Digestivo. Respiratorio. Neurológico.

Introduction

With the Orpha code 803, amyotrophic lateral sclerosis (ALS) is defined by the Carlos III Institute of Health as a "neurodegenerative disease characterized by progressive muscular paralysis that reflects a degeneration of motor neurons in the primary motor cortex, corticospinal tract, brain stem, and spinal cord".[1]

If we go back in time to the beginning of the history of this disease in 1848, we could affirm that François Amilçar Aran was the first to speak of it, publishing the description of a pathology "not yet described" characterized by "muscular atrophy progressive". However, the first description of a bulbar form of amyotrophic lateral sclerosis (ALS) is due to Charles Bell and his work on sensory and motor functions as two independent systems, describing in his study the case of a woman who remains quadriplegic, but without suffering impairment of sensitivity, memory, or other intellectual functions. This statement confirmed this neuromuscular alteration at autopsy.[2]

If the first to approach what ALS would be was François Aran, Jean-Martin Charcot was the first to describe it as such between 1865 and 1874, as a result of progressive degeneration of motor neurons, with an idiopathic origin, which generated weakness and generalized, progressive and rapid muscular atrophy, which frequently end in death. He identified this first pathological picture as "primary lateral sclerosis". In 1869, he published with Joffroy two cases that he called "amyotrophic lateral sclerosis" specifying its manifestations, as we know it today.[2,3]

According to various studies, ALS currently has an incidence of around 1 / 50,000 annually, with a prevalence of 1 / 20,000 on average. The average age of onset is around 60 years, and although the male / female ratio was previously

higher, the trend is to equalize, still finding a slight male predominance (1.3: 1).[1]

Since the diagnosis of ALS, the average duration of the disease ranges from two to 5 years, with respiratory failure being the main cause of death in these patients. However, the changes introduced in healthcare in recent years have significantly improved survival.[1,3,4]

Normally, the patient does not have the capacity to adapt or improve; therefore, it is very important to take care not only of the patient but also of his family, who will be the main providers of informal care, considering them a single work unit, within order to help you live to the end of your life. That is why, as nursing professionals, we must not miss the caregiver's health. For a person to be able to provide quality care, they have to start by taking care of themselves. If the caregiver is not taken into account, the care provided to the patient will be of poorer quality. Quite often, the main caregivers are usually seen as an annex of the care receiver, the patient, while they should be seen from the perspective that they have a different perception and needs than the patient, so we should never forget their health. Therefore, with our intervention within the interdisciplinary team, we ensure an improvement in the care that the informal caregiver provides, increasing patient safety, and therefore the quality of life of both, thus also improving communication and information between them and family satisfaction since the caregiver does not reach excessive saturation. In addition to the emotional impact that the whole disease process implies for both, implies, in most cases, a change in the habitual lifestyle, both in the fieldwork as economic (specific care, treatment expensive, transport), affecting to a great extent, as we have commented on the quality of family life.[5-8]

In recent years, there are randomized controlled clinical trials that have shown that long-term mechanical ventilation improves survival and quality of life in these patients. As the disease progresses, symptoms appear such as difficulty clearing respiratory secretions, dyspnoea on exertion, sleep pattern disturbance, fatigue, night awakenings, irritability, daytime sleepiness, difficulty concentrating, impaired cognition, altering your quality of although we have tests that analyse lung function such as vital capacity, oximetry, arterial blood gas, polysomnography, total lung capacity, and inspiratory capacity, lying vital capacity, inspiratory flow, which may indicate signs of poor prognosis for the introduction of tracheostomy, need for non-invasive ventilation, use of mechanical ventilation, we lack tools such as scales or questionnaires that allow us to predict this non-invasive respiratory and digestive neurological deterioration.[9]

The aim of our study was to identify the existence of measurement scales validated help us predict the existence of neurological impairment respiratory and digestive order to be able to intervene at an early stage with the aim to improve the quality of life of these patients.

Methodology

A bibliographic search that ends on December 14, 2019, was performed in the international databases Pubmed, CINHAL, Cochrane, and the national CUIDEN. The DECS terms for the bibliographic search were those shown in Table 1.

The search strategy used was the one shown in Table 2.

After reading the summaries, those articles that addressed all the points marked in our objective were included, selecting 24 articles published between the years 2000 and 2019, of which only 15 were included in our review as they were completely in line with the content of our search (see table 3). Table 4 collects in a flow diagram the information of the methodology through the different phases of the systematic review.

Table 1. *DECS search terms*

Spanish	**English**
Encuestas y Cuestionarios	Surveys and Questionnaires
Cuestionario de Salud del Paciente	Patient Health Questionnaire
Esclerosis Amiotrófica Lateral	Amyotrophic Lateral Sclerosis
Técnicas de Diagnóstico Neurológico	Neurological Diagnostic Techniques
Examen Neurológico	Neurologic Examination
Técnicas de Diagnóstico del Sistema Respiratorio	Diagnostic Techniques, Respiratory System
Sistema Respiratorio	Respiratory System
Insuficiencia Respiratoria	Respiratory Insufficiency
Técnicas de Diagnóstico del Sistema Digestivo	Digestive System Diagnostic Techniques
Sistema Digestivo	Digestive System
Anomalías del Sistema Digestivo	Digestive System Abnormalities

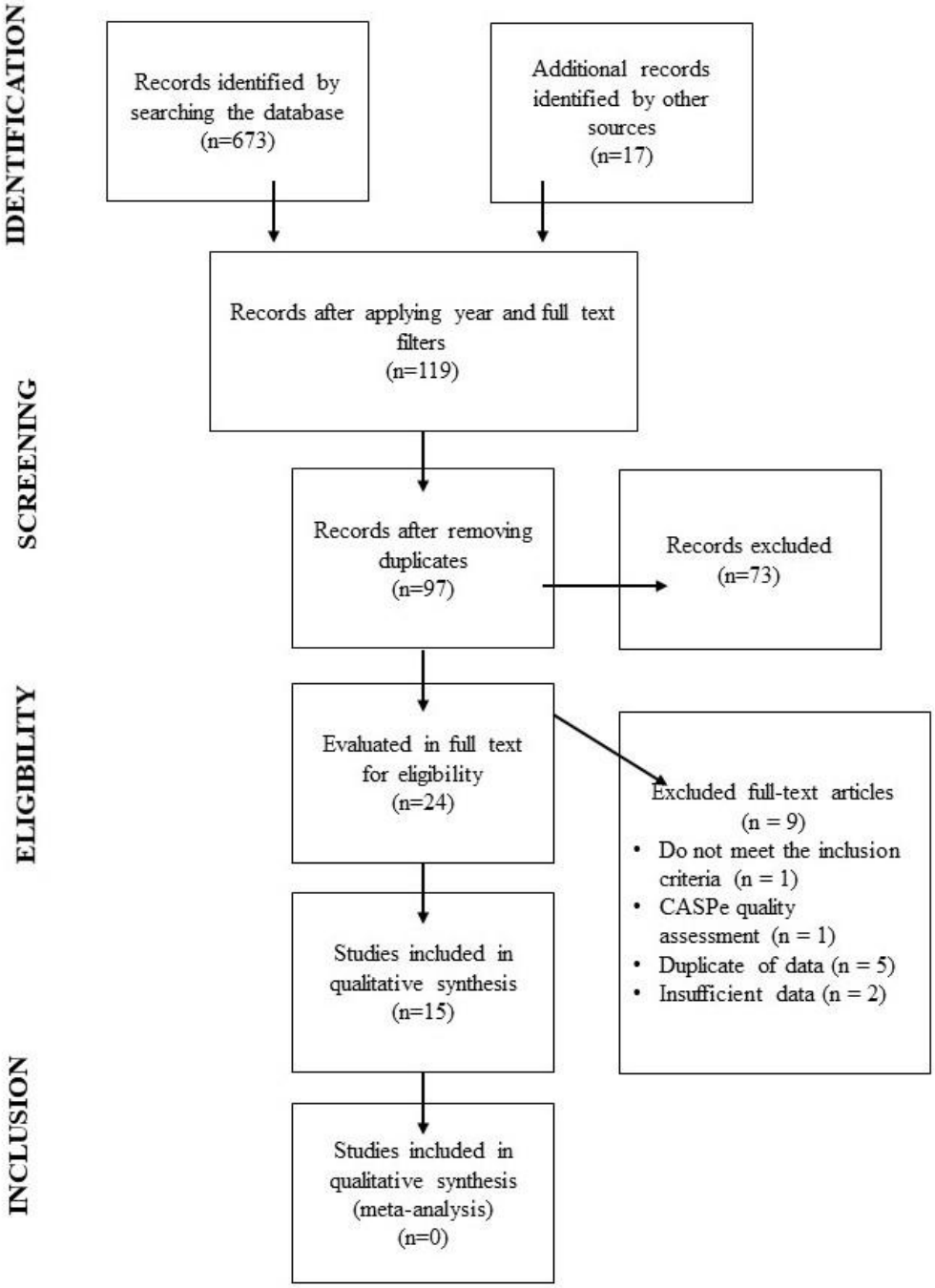
Table 2. *Search strategy*

Database	Strategy		Filter	
Pubmed	(((Amyotrophic Lateral Sclerosis) AND Questionnaires) AND Respiratory System) AND Digestive System	8		
	((((amyotrophic lateral sclerosis) AND questionnaires) AND ((respiratory system) OR (digestive system)))	47	10 year, free full text	11: 3
CUIDEN	("ALS")AND("questionnaires")	354	0	0
	("ALS")AND(("questionnaires")AND(("RESPI")OR("DIGES")))	10	0	0
Cinhal	(amyotrophic lateral sclerosis) AND questionnaires	194	2000-2019, full text	40: 15
Cochrane	"amyotrophic lateral sclerosis" in Title Abstract Keyword AND "questionnaire"	60	2000-2019	60:8

Table 3. *Studies included in the systematic review. Type of study, and level of evidence*

Authors	Year	Type of study	Level of evidence	Incorporation
Johnson et al	2017	Original article	2	YES
Felgoise et al	2018	Original article	2	YES
Geng et al	2016	Original article	2	YES
Vitacca et al	2013	Original article	2	YES
Rutkove	2015	Bibliographic review	3	YES
Molina et al	2013	Original article	2	NO
Hwang et al	2016	Original article	2	NO
Callagher et al	2009	Original article	2	NO
Grehl et al	2010	Original article	2	YES
Correia et al	2014	Original article	2	NO
Siciliano et al	2019	Original article	2	YES
Moore et al	2018	Original article	2	YES
Martínez-Campo et al	2017	Bibliographic review	3	YES
Santos et al	2016	Original article	2	NO
Ciotti et al	2018	Original article	2	YES
Creemers et al	2017	Bibliographic review	3	NO
Lavernhe et al	2017	Bibliographic review	3	NO
Malik et al	2014	Study of cases	5	NO
McGeachan et al	2016	Bibliographic review	3	NO
Montes et al	2006	Original article	2	YES
Jenkinson et al	2000	Original article	2	YES
Neudert et al	2001	Original article	2	YES
Ortiz Corredor et al	2011	Original article	2	YES
Rooney et al	2016	Bibliographic review	3	YES

Table 4. *Information flow diagram through the different phases of the systematic review*



Results

Due to its rapid evolution, and respiratory failure as the most common cause of death within the first 3 years from diagnosis, the main focus of care is on palliative care for the patient with ALS, with the aim of improving the quality of life of both the patient and their family, due to the great psychological impact that this entails for both, taking on greater importance in the approach to improving the quality of life of the patients with Amyotrophic Lateral Sclerosis. However, the concept of quality of life is multifactorial and, therefore, difficult to define.[10-11]

There is a study that analysed how suffering from a neurological disease affected the quality of life, through a survey, concluding that it worsened considerably compared to the general population and highlighting the lack of support and difficulty in accessing services that these patients manifested. [11]

Another study also analyzed quality of life, this time, from a rehabilitation centre. The scale used was generic and not specific for motor neuron disease, resulting in an increase in quality of life in those patients who go to rehabilitation more times, without agreement on the reason for this increase.[12]

There is another study in which they identified the lack of consideration of the cognitive-emotional domain of these patients, beginning to address it with their study. For this, they used the Parkinson's Hoehn and Yahr scales, The Schwab and England (S & E) Activities of Daily Living (ADL) Scale, and Unified Parkinson's Disease Rating Scale,

also evaluating depression, with a great impact on the quality of life and physical deterioration. This study determined that motor deficiencies should not only be improved to improve the quality of life in these and patients and that this improvement when recognized and treated.[11,12]

Other authors assessed the quality of life in ALS patients using the McGill questionnaire for quality of life, the sickness Impact Profile ALS-19 questionnaire, and the Idler index of religiosity (measures the high correlation between internal and external support and quality of life), concluding the need to go beyond the physical to assess the quality of life of patients, and highlighting the great importance of external support.[11]

Another study tells us about the instruments available to assess the quality of life in these patients. The ALSSQOL scale and its short version ALSSQOL-R measure global quality of life with 6 subscales (negative emotion, interaction with people and the environment, intimacy, religiosity, physical symptoms, and bulbar function, as well as an average quality score of total life), and are measured with a Likert-type scale of 0-10 (0 less desirable and 10 is the most desirable). In this study, difficulties are shown in passing the questionnaires in their long version, due to the appearance of fatigue and the short time by the professional who carried out the questionnaires. Regarding the short version, it presented a better reception, maintaining the objective of being a valid and specific quality of life instrument for ALS and enhancing the areas that patients indicated as most important for improving quality of life, such as the planning of professionals' interventions with the patient and the comprehensive care of the multidisciplinary team.[13]

As the disease progresses, patients with ALS become weak, experiencing pain and anxiety as symptoms deteriorate. The goal for healthcare professionals and caregivers is to slow down this progression and make its impact less significant since they should know that a disease progression can significantly affect the resignation of these patients and relatives. It is then when everything possible must be offered to provide appropriate psychological support while preserving the dignity of the patient at all times.[12,14]

In addition, this progression of the disease implies an increase in the responsibility of the caregiver to whom, in addition to the physical demands, they are often attributed with important decision-making, which, adding daily physical and emotional support, can lead to depression, which their quality of life decreases. Some authors consider caregiver overload as the appearance of feelings of burden (self-perceived) that are postulated as a multidimensional construction, which arises from feelings of dependency and frustration in the care recipient, leading to feeling guilty and responsible for the caregiver's burden, manifesting as pictures of tension and anxiety and affecting personal relationships and their quality of life. That is why it is important to value the quality of life as a family nucleus that includes both the patient with ALS and their caregiver, evaluating the relationship between the two. Today there is no questionnaire that specifically evaluates these parameters. Tests like SIP / ALS – 19 and the ALS Assessment Questionnaire are patient-centered. An instrument that values the quality of life as the core is needed.[9,11,12]

There is another study of 30 patient-caregiver couples, using the Short Form Health Survey Questionnaire (SF-12), the abbreviated version of SF-36, with 12 questions to measure the respondent's functional health and well-being, with 8 domains: general health, psychic functioning, physical role, body pain, mental domains, vitality, social functioning, and emotional and mental health. They were passed 2 times, once to assess their quality of life, and once to assess the perspective of the caregivers; and in the case of caregivers as well, one for their own perspective and one for that of the patient. After analyzing the results obtained in this study, the importance of health professionals emphasizing communication between the patient and their caregivers was highlighted, in the face of emotional well-being, including spirituality, social and mental well-being.[11]

Other authors conducted a study that identified an increased burden on caregivers of ALS patients using the Self-Perceived Overload Scale (SPB), and therefore the perceived quality of life in ALS patients decreased. This overload tended to be greater in female patients, with more knowledge of the disease and severe respiratory problems, and on the caregiver side, severe motor dysfunction and increased cost of care were the factors that increased the burden. Other studies indicated that weight loss, oropharyngeal dysphagia and pain are other symptoms that had a negative impact on the quality of life of the ALS patients.[12]

Another of the available scales was ALSFR, designed as a scoring system for the functional capacity of ALS patients. It consists of 10 questions, rated on a scale of 0 to 4, in all the different domains that could be affected by the disease, including respiratory bulbar function and upper and lower limb function. The original was replaced a few years later by ALSFRS-R when it was recognized that respiratory deterioration needed to be measured. The new scale consists of 12 questions that add up to 48 points and values movement especially of the upper extremities such as writing, feeding, clothing (fine motor), speech, swallowing, salivation (bulbar function), turning, dressing, walking, climbing (gross motor) and dyspnoea, orthopnoea, respiratory failure (respiratory function).[12,15]

There is another study in which they used the ALSFRS-R scale to assess the severity of functional ALS. The conclusion of this study concluded that it is a reliable tool, although the result is quite a challenge for neurologists and nurses around the world when having to decide when and how to give a diagnosis of ALS to patients. However, this questionnaire has a series of limitations, for example, that it is not sensitive to the progression of the disease, and can be affected by emotions as it is a subjective test, as well as by the degree of social support and presence of depression.[12,15]

In addition, the revised ALS functional rating scale (ALSFRSR) predicts the progression of the disease in terms of neurological involvement, as well as the survival time of these patients. Several studies have shown that this scale can be administered by an evaluator, or self-administered, with the same reliability. This scale has been used in ALS clinical trials as a secondary and primary outcome measure so that both neurologists and ALS patients could benefit from the ability to monitor the progression of the disease of these patients from home using the self-administered questionnaire, resulting in this questionnaire being beneficial for

both research and clinical practice in ALS. The study also shows that the self-administered version is better valued than when the healthcare professional administers it.[16]

For the functional classification of ALS patients, this system is easy and quick to apply and it stratifies the severity groups in terms of prognosis. In this way, a study managed to discriminate based on the use ALSFRSR shows five patterns of disease progression, depending on the state of functional deterioration and time of evolution.[17]

In the absence of an established biomarker that allows us to measure disease progression, the decrease in the value obtained on the ALSFRS-R scale has become the measure of progression most used by research clinicians.[18]

Another study showed that ALS patients had a correlation between increased physical dependence and the onset of depression, as well as between impaired speech and anxiety. The inevitable progression of ALS tends to be reflected in a decrease in quality of life. In this study, it is shown that, before the deterioration of physical function; it worsens the quality of life and dignity respectively, compared to healthy controls. This deterioration, in turn, did not seem to be associated with factors such as the onset of the disease, age of onset, or sex. The introduction of technical aids such as non-invasive BPAP ventilation or the placement of a PEG had a significant and negative association with the quality of life of patients with ALS.[14]

As a result of the previous one and to conclude, we identified one of the most important and complete standardized questionnaires used in Spain to measure the subjective well-being of patients with ALS, as well as the progression of the disease. This is the ELA assessment questionnaire (ALSAQ40), the Spanish adaptation of the ALSFR-R and which has five domains (physical mobility, activities of daily living and independence, eating and drink, communication and emotional reactions). The maximum score is 200 points, and the higher the score, the lower the quality of life.[14-19]

Discussion

Today, the prediction of neuromuscular impairment in ALS is very difficult, with changes in neurological impairment occurring from month to month, or even weeks, due to its rapid evolution. Despite the multiple organ involvement, and the multiple tests that could be applied to identify signs of deterioration, early diagnosis continues to be a challenge, sometimes being a problem, choosing which tests to use, for example, two people with the same disease, may have a different presentation and evolution (a person with bulbar involvement, will have a different evolution than someone with predominantly lower limb weakness). To all this, it must be added that some patients do not survive more than a year from diagnosis.[15]

Furthermore, due to its poor prognosis and the associated psychosocial impact, ALS is a neurodegenerative disease that clearly affects the quality of life and dignity of the patient and his family, with a close relationship between clinical deterioration and a worsening in the quality of life, becoming an important goal in ALS management within the multidisciplinary team.[14]

At each stage of the disease, there are specific functional problems that progressively lead to severe disability, which generally leaves a good level of cognitive function. The capacity limitation is gradual and becomes dependent on environmental factors, such as caregivers and technology, which in turn influence their quality of life.[20]

As we have seen, there is no questionnaire that specifically values the quality of life as a family, viewing the patient as a family unit.[11]

Having said this, it should be concluded that despite the constant work to improve the quality of life and dignity of patients and their families, currently there is no validated questionnaire that predicts the existence of respiratory and digestive neurological deterioration, a tool that would be very useful for early intervention in the face of this deterioration, improving the quality of life and dignity of these patients and their families, the main point of approach for this disease.

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