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RESEARCH ARTICLE (ORIGINAL)

Burden of informal caregivers of people with amyotrophic lateral sclerosis

Sobrecarga dos cuidadores informais de pessoas com esclerose lateral amiotrófica
Sobrecarga de los cuidadores informales de personas con esclerosis amiotrófica lateral

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Abstract
Background: Amyotrophic lateral sclerosis (ALS) is an incurable and disabling progressive neurodegenerative disease, where the caregiver plays a key role in meeting the patient's needs.
Objective: To analyze the association between caregivers' sociodemographic and professional characteristics and their burden and between the sociodemographic and clinical characteristics of individuals with ALS and caregivers' burden.
Methodology: Descriptive-correlational study. Sample of 30 patients and 30 caregivers. The Informal Caregiver Burden Assessment Questionnaire was used. Parametric and non-parametric tests were used for data analysis.
Results: The sample of patients had a mean age of 66.4 ± 11.17 years and homogeneous gender distribution. The majority of caregivers were women, spouses, with moderate levels of burden (56.7%), less affected by "reactions to demands" and more affected by "efficacy and control mechanisms" (mean 25.67 ± 21.76 and mean 66.39 ± 21.50). The caregiver's sociodemographic characteristics, the patient's functional status, the number of care hours, and the use of non-invasive ventilation were associated with burden levels (*sig* < 0.05).
Conclusion: Caregivers have moderate levels of burden, highlighting the importance of proximity care.

Keywords: amyotrophic lateral sclerosis; caregivers; burden; quality of life

Resumo
Enquadramento: A esclerose lateral amiotrófica é uma doença neurodegenerativa, progressiva, incapacitante e sem cura, onde o cuidador assume um papel fundamental na resposta às necessidades.
Objetivo: Analisar a associação entre caraterísticas sociodemográficas e profissionais do cuidador e sobrecarga; e entre caraterísticas sociodemográficas e clínicas da pessoa com ELA e a sobrecarga do cuidador.
Metodologia: Estudo descritivo-correlacional. Amostra de disponíveis com 30 doentes e 30 cuidadores. Como instrumento utilizou-se o Questionário de Avaliação da Sobrecarga do Cuidador Informal. Análise estatística com testes paramétricos e não paramétricos.
Resultados: Doentes com média de 66,4 ± 11,17 anos e distribuição homogénea quanto ao sexo. Cuidadores maioritariamente cônjuges, mulheres e com sobrecarga moderada (56,7%), sendo menos afetada a "reações a exigências" e mais afetada os "mecanismos de eficácia e de controlo" (média 25,67 ± 21,76 e média 66,39 ± 21,50). Caraterísticas sociodemográficas do cuidador, funcionalidade do doente, tempo de cuidados e ventilação não invasiva, relacionam-se com os níveis de sobrecarga (*sig* < 0,05).
Conclusão: Os cuidadores apresentam níveis moderados de sobrecarga, tornando importante cuidados de proximidade.

Palavras-chave: esclerose lateral amiotrófica; cuidadores; sobrecarga; qualidade de vida

Resumen
Marco contextual: La esclerosis amiotrófica lateral es una enfermedad neurodegenerativa, progresiva, incapacitante e incurable, donde el cuidador informal asume un papel fundamental en la respuesta a las necesidades.
Objetivos: Analizar la asociación entre características sociodemográficas y profesionales del cuidador y la sobrecarga; y entre las características sociodemográficas y clínicas de la persona con ELA y la sobrecarga del cuidador.
Metodología: Estudio descriptivo-correlacional. Muestra de disponible con 30 pacientes y 30 cuidadores. Para la evaluación de la sobrecarga, se utilizó el Cuestionario de evaluación de la sobrecarga del cuidador informal. Análisis estadístico con pruebas paramétricas e no paramétricas.
Resultados: Pacientes con una media de 66,4 ± 11,17 años y distribución homogénea por sexo. Cuidadores mayoritariamente cónyuges, mujeres y con sobrecarga moderada (56,7%), siendo menos afectada por "reacciones a demandas" y más afectada por "mecanismos de eficiencia y control" (media 25,67 ± 21,76 y media 66,39 ± 21,50). Las características sociodemográficas del cuidador, la funcionalidad del paciente, la duración del cuidado y la ventilación no invasiva están asociados con los niveles de sobrecarga (*sig* < 0,05).
Conclusión: Los cuidadores tienen niveles de sobrecarga moderados, por lo que son importantes los cuidados de proximidad.

Palabras clave: esclerosis amiotrófica lateral; cuidadores; sobrecarga; calidad de vida

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Introduction

Amyotrophic lateral sclerosis (ALS) is a rare, rapidly progressive neurodegenerative disease of the central nervous system. It is characterized by degeneration of motor neurons in the cerebral cortex, brainstem, and spinal cord, leading to denervation of nerve cells and atrophy of the corresponding muscle fibers (Brown, 2015).
ALS affects over 70,000 people worldwide, and survival rates range from 3 to 5 years. For Brown (2015), the incidence is 1-3/100,000 individuals, and the prevalence is 3-5/100,000 individuals. Maragakis and Galvez-Jimenez (2018) report slightly higher prevalence rates (2.7-7.4/100,000 individuals). Its incidence increases with age, reaching a peak at 74 years. Prevalence is slightly higher among White men. Even though the average survival rate is 3-5 years, some individuals live more than a decade (Maragakis & Galvez-Jimenez, 2018). Although there are no epidemiological studies in Portugal, the Portuguese Association of ALS estimates that around 800 people in Portugal have ALS (Carvalho, 2016).
Studies report that these people remain at home until an advanced stage of the disease, under the care of caregivers (usually the spouse), and experience increasing levels of suffering with an impact on health (de Wit et al., 2018; Galvin et al., 2016; Pinho & Gonçalves, 2016). Thus, as this is a progressive disabling disease affecting the lives of patients and caregivers, nurses play an important role in training the caregiver to perform self-care tasks and develop strategies to cope with the levels of burden.
This study aimed to analyze the association between caregivers' sociodemographic and professional characteristics and their burden and between the sociodemographic and clinical characteristics of individuals with ALS and caregiver's burden.

Background

The classic presentation of ALS is that of progressive motor deterioration causing upper and lower neuron signs and symptoms. Most patients present with initial muscle weakness in the upper or lower limbs (spinal onset); in 25% of patients, it may affect first the muscles innervated by the bulb (bulbar onset); and in a small number of patients (about 1-2%), it may affect the thoracic and respiratory muscles (Fearon et al., 2015).
Changes in the respiratory pattern usually occur in the later stages of the disease. The weakening of the respiratory muscles and the axial muscles of the trunk triggers symptoms of hypoventilation that cause fatigue, dyspnea, tachypnea, and functional impotence.
This disease has major physical, psychological, and social implications for patients, families, and caregivers. The terms informal caregiver and family caregiver refer to unpaid individuals who provide care. These individuals may be primary or secondary caregivers. The primary caregiver provides most of the care and has more responsibilities (Bruletti et al., 2015). According to de la Cuesta-Benjumea (2011), the family remains the primary source of help preferred by care-dependent people. Martins (2006) refers that the choice of caregivers is not arbitrary and tends to fall on a female member. According to Almeida et al. (2017), the role of women as caregivers is historically and culturally evidenced. However, due to the socio-economic development over the last century, women have entered the job market, and other family members have assumed the role of primary caregivers.
The caregiving routine is often exhausting and can last for a prolonged period of time, leading to caregiver burden. According to Braithwaite (1992), as cited by Martins (2006), caregiver burden is a distress arising from dealing with caregivers' physical dependence and mental incapacity, corresponding to the subjective perception of threats to the caregiver's physiological, social, and psychological needs. In contrast, de Wit et al. (2018) define caregiver burden as the impact on the emotional health, physical health, social life, and the financial status of the caregiver as a result of adopting this role. For Lage (2007), family demands in terms of time, energy, emotional commitment, and resources grow as limitations increase.
ALS causes a rapid decline in physical functioning, requiring physical and emotional adjustments from both patients and caregivers. As the disease progresses, caregivers' stress, worries, and burden increase, with restrictions on their time availability, energy for leisure activities, and time to fulfill their own needs (de Wit et al., 2018). For Pagnini et al. (2010), together with emotional strain, care in ALS requires much physical effort, particularly during the advanced stages of the disease, in which the decline in mobility and the need for mechanical ventilation influence the caregiver's burden. Pinho and Gonçalves (2016) mention that caregivers tend to neglect their own health status due to the amount of time they spend caring for people with ALS. Therefore, health professionals should focus their attention on both people with ALS and their caregivers.

Research question

Is the burden of caregivers of people with ALS associated with the caregivers' sociodemographic and professional characteristics and/or with the sociodemographic and clinical characteristics of people with ALS?

Methodology

A descriptive-correlational, observational, and cross-sectional study was conducted with a sample of 43 individuals with ALS and their caregivers being treated in hospitals of the district of Braga. The following inclusion criteria were applied: people with confirmed medical diagnosis of ALS; people at home under the care of an informal caregiver; pairs of informal caregivers and people with ALS who were willing to participate in the study. Thus, a sample consisting of 30 people with ALS and their caregivers was selected.
Individuals with ALS had a homogeneous gender distri-

bution (50% of women). Their ages ranged from 43 to 85 years, with a mean of 66.4±11.17 years and a median of 68.5 years. The most represented age groups were 40-64 years and 65-79 years (43.3% each), with 13.1% of individuals over the age of 80. Concerning marital status, married/cohabiting couples were predominant (83.3%). The education level ranged from no schooling (6.7%) to higher education (10.0%), with a predominance of the 1st cycle of basic education (50.0%). As for the professional situation, none of the individuals was currently working, and 46.7% were retired due to disability.

The majority of caregivers were women (76.7%) aged 40 to 88 years, with a mean of 60.5±13.63 years and a median of 60.73 years. The most represented age group was 40-64 years (70%), and 16.3% of caregivers were aged 80 years or over. There was a predominance of married/cohabiting couples (86.7%) and of the 1st cycle of basic education (36.7%). Concerning the professional situation, 40.0% were professionally active. Concerning the relationship with the patient, 63.3% were spouses, followed by children (23.3%), parents (10.0%), and siblings (3.3%).

The Amyotrophic Lateral Sclerosis Functional Rating Scale-Revised (ALSFRS-R; Guedes et al., 2010) was used to assess the functional status of people with ALS. It consists of 12 items grouped into four domains. The questions are rated on a Likert-type scale from 0 (*unable*) to 4 (*normal ability*), with higher scores indicating a higher level of functioning. Mioshi et al. (2012) categorize into four stages of severity: mild (37-48), moderate (25-36), severe (13-24), and very severe (0-12). The Cronbach's alpha value of the scale was 0.895.

The Amyotrophic Lateral Sclerosis Assessment Questionnaire (ALSAQ-40; Pavan et al., 2010) was used to assess the quality of life (QoL) of individuals with ALS. It consists of 40 statements rated on a Likert-type scale ranging from *never* (0) to *always* (4). They are grouped into five dimensions, with higher scores indicating worse QoL (Bandeira et al., 2010). The Cronbach's alpha value of the scale was 0.919.

The World Health Organization Quality of Life - BREF (WHOQOL-BREF; Vaz Serra et al., 2006) was used to assess caregivers' QoL. It consists of 26 questions rated on a 5-point Likert-type scale and addresses four domains. Higher scores correspond to better QoL. The Cronbach's alpha value of the scale was 0.838.

The Informal Caregiver Burden Assessment Questionnaire (*Questionário de Avaliação da Sobrecarga do Cuidador Informal*, QASCI) by Martins et al. (2003) was used to assess caregiver burden. It consists of 32 items rated on a Likert-type scale ranging from 1 to 5. The items are divided into seven dimensions: Emotional Burden (EB); Impact on Personal Life of the Caregiver (IPL); Financial Burden (FB); Reactions to Demands (RD); Mechanisms of Efficacy and Control (MEC); Family Support (FSupp) and Satisfaction with Role and with the Relative (SRR; Martins et al. 2004). Higher scores indicate a higher burden. The Cronbach's alpha value of the scale was 0.911. According to the authors' cut-off points, burden can be low (0-25), moderate (26-50), high (51-75), or extreme (> 76).

Internal consistency in all scales ranged from very good to good.

Descriptive statistics were used for data analysis. Associations and differences were analyzed using the assumption of normality. Parametric tests (Pearson's test and Student's *t*-test) were used in the case of normal distribution (Shapiro-Wilk test). For ordinal qualitative variables or in the case of non-normal distribution, Spearman's correlation coefficient and the Mann-Whitney *U* test were used. For nominal variables, the chi-square test of independence was used. The SPSS® 26.0 for Windows was used for data analysis, and the level of significance was set at 5%.

The study received a favorable opinion from the ethics committees of the hospitals where it was carried out. Participation in the study was voluntary, and prior informed consent was obtained. To ensure compliance with internationally recognized standards, the purpose of the study was described in the informed consent form and clarified when the questionnaires were applied.

Results

The results show that 66.7% of patients had bulbar onset, 26.7% had spinal onset, and 6.6% had respiratory onset. Most patients reported that their symptoms had begun 2 to 5 years earlier and 5 to 10 years earlier (33.3% each), 16.7% of patients reported onset 2 years earlier or less, and the remaining patients reported onset more than 10 years earlier.

As for functional assessment, 33.3% had a very severe degree of functional impairment, 30% had a severe and moderate degree, and 6.7% had a mild degree.

Concerning the dimensions of functional assessment, the respiratory function had the highest scores, with a mean of 7.62 ± 2.90 and a median close to the mean (7.5), while the gross motor function had the lowest scores, with a mean of 2.87 ± 3.46 and a median of 1.5, indicating high variability and asymmetry (Table 1).

Table 1

Descriptive Statistics of Functional Assessment (ALSFRS-R; n = 30)

	min-max	Mean±Deviation	Median
Bulbar	0-12	5.83 ± 4.20	5.00
Fine Motor	0-10	3.27 ± 3.89	1.00
Gross Motor	0-12	2.87 ± 3.46	1.50
Respiratory Function	2-12	7.62 ± 2.90	7.50
TOTAL	1-39	19.20 ± 11.54	16.50

QoL was assessed based on the valid answers of 28 participants: 35.7% of respondents always had difficulties, that is, worse QoL, 32.1% had difficulties very often, 25.0% had difficulties quite often, and 7.1% rarely had difficulties. There was no case of no difficulty. The best perception of QoL was found in Eating and Drinking (mean 55.95 ± 38.08; median 58.33), and the worst scores were found in Physical Mobility (82.00 ± 19.54; median 86.25; Table 2).

Table 2

Descriptive Statistics of Quality of Life (ALSAQ-40; n = 28)

	min-max	Mean±Deviation	Median
Physical Mobility	35-100	82.00 ± 19.54	86.25
Activities of Daily Living	12.50-100	72.32 ± 29.27	81.25
Eating and Drinking	0-100	55.95 ± 38.08	58.33
Communication	0-100	69.13 ± 36.72	87.50
Emotional functioning	7.50-100	56.25 ± 25.02	57.50
TOTAL	30-100	68.49 ± 19.45	68.75

Concerning the caregivers' burden, the lowest scores were found in the RD dimension, with a mean of 25.67 ± 21.76, followed by FB (mean 29.17 ± 36.00) and EB (mean 41.25 ± 22.72), with the IPL showing intermediate scores. The highest burden was found in the MEC (mean 66.39 ± 21.50) and the FSupp dimensions (mean 66.25 ± 31.51; Table 3). Moderate levels of burden were more prevalent (56.7%), followed by high (20.0%) and low levels (23.3%). No cases of extreme burden were found.

Table 3

Descriptive Statistics of the Informal Caregiver's Burden (n = 30) – QASCI

	min-max	Mean±Deviation	Median
Emotional Burden (EB)	0-87.50	41.25 ± 22.72	43.75
Implications for Personal Life (IPL)	6.82-100	51.44 ± 24.12	44.32
Financial Burden (FB)	0-100	29.17 ± 36.01	6.25
Reactions to Demands (RD)	0-100	25.67 ± 21.76	22.50
Mechanisms of Efficacy and Control (MEC)	16.67-100	66.39 ± 21.50	66.67
Family Support (FSupp)	0-100	66.25 ± 31.51	75
QASCI Total	6.97-74.31	35.81 ± 16.28	30.83

Concerning the caregivers' QoL (Table 4), perceived QoL was higher in the Psychological domain (62.36 ± 16.36; median 60.42) and the Physical domain (57.5 ± 18.36; median 55.36). The Environmental domain had a mean of 56.77 ± 12.284 and a median of 59.38, and the Social Relationships domain had the lowest scores (mean 51.67 ± 20.11; median 50.00).

Table 4

Descriptive statistics of the QoL of caregivers of people with ALS (n=30) – WHOQOL-BREF

	min-max	Mean±Deviation	Median
Domain 1 – Physical health	10.91-96.43	57.5 ± 18.36	55.36
Domain 2 – Psychological	4.17-87.50	62.36 ± 16.36	60.42
Domain 3 - Social Relationships	16.67-83.33	51.67 ± 20.11	50.00
Domain 4 – Environment	28.13-81.25	56.77 ± 12.284	59.38
Overall QoL	0.00-87.50	49.17 ± 18.26	50.00

The analysis of the association between caregivers' burden and their sociodemographic characteristics revealed a moderate positive correlation (r = 0.480; *sig* = 0.007) between age and FSupp. Concerning the degree of kinship, when comparing spouses and other family members, significant differences were found in the FB and FSupo dimensions (Z = 2.028; *sig* = 0.043 and Z = 2.203; *sig* = 0.028, respectively), with spouses showing higher mean ranks than other family members (*FB* 17.82 vs. 11.50; *FSupp* 18.13 vs. 10.95). With regard to the professional situation, when comparing active and non-active individuals, significant differences were found in the RD and FSupp dimensions (Z = -2.134; *sig* = 0.033 and Z = -2.037; *sig* = 0.042, respectively), with non-active individuals showing higher scores than active individuals (mean ranks of 18.28 vs. 11.33 and 18.11 vs. 11.58, respectively).

The analysis of the association between the caregiver's burden and perceived QoL showed significant negative correlations between FB and the Physical, Psychological, Social Relationships, Environmental, and Overall QoL domains (r = -0.441, *sig* = 0.015; r = -0.528, *sig* = 0.003; *rs* = 0.487, *sig* = 0.006; r = -0.446, *sig* = 0.013; r = -0.570, *sig* = 0.001). Statistically significant negative correlations were also found between the IPL dimension and the Physical, Psychological, Social Relationships, Environmental, and Overall QoL domains (r = -0.633, *sig* = 0.000; r = -0.528, *sig* = 0.003; *rs* = -0.460, *sig* = 0.011; r = -0.656, *sig* = 0.000; r = -0.700, *sig* = 0.000, respectively). A significant correlation was found between the FB dimension and the Social Relationships dimension of QoL (r = -0.390; *sig* = 0.033). Significant positive correlations were found between the MEC dimension and the Physical, Psychological, Social Relationships, Environmental, and Overall QoL domains (r = 0.447, *sig* = 0.013; r = 0.374, *sig* = 0.041; *rs* = 0.726, *sig* = 0.000; r = 0.364, *sig* = 0.048; r = 0.364, *sig* = 0.048, r = 0.365, *sig* = 0.047, respectively). Concerning the total QASCI score, statistically significant negative correlations were observed with the Physical, Psychological, Social Relationships, Environmental, and Overall QoL domains (*rs* = -0.548, *sig* = 0.002; *rs* = -0.467, *sig* = 0.009; *rs* = -0.717; *sig* = 0.000; *rs* = -0.552, *sig* = 0.002; *rs* = -0.551, *sig* = 0.002, respectively).

As for the association between the caregiver's burden and the functional status of the person with ALS (total score), moderate negative correlations were found in the EB and IPL dimensions (r = -0.402; *sig* = 0.028 and r = -0.436; *sig* = 0.016, respectively). Regarding the number of care hours, positive correlations were found between the IPL and RD dimensions (*rs* = 0.443; *sig* = 0.014 and *rs* = 0.370; *sig* = 0.044, respectively). As for the presence of non-invasive ventilation (NIV), differences were found in the FB (Z = -1.971, *sig* = 0.049), where the caregivers of people with NIV had higher scores than those without NIV (mean scores 16.98 vs. 9.58). Differences were also found in the total QASCI score (t = -2.194; *df* = 16.91; *sig* = 0.042), where caregivers of people with NIV had higher mean scores than those without NIV (37.97 ± 17.17 vs 27.21 ± 8.40).

The analysis of the association between the informal caregiver's burden and the perception of QoL of individuals with ALS showed a positive correlation between the FSupp dimension and the Eating and Drinking domain (r = 0.404; *sig* = 0.033). The Emotional functioning domain showed positive correlations with the EB and IPL dimensions (p = 0.500; *sig* = 0.007 and p = 0.413; *sig* = 0.029, respectively. Negative correlations were also found between the Emotional functioning domain and the MEC dimension (r = -0.469; *sig* = 0.012).

Discussion

ALS is a disabling disease that is more common in men, adults, and older adults. In this study, people with ALS have a homogeneous gender distribution, which is not observed in more recent studies that found a slightly higher prevalence in men (Maragakis & Galvez-Jimenez, 2018). They are predominantly adults and older adults (66.4 ± 11.17 years), and none of the persons with ALS are professionally active. As for marital status, the majority of them are married/cohabiting. The education level ranged from no schooling to higher education, with a higher frequency of the 1st cycle of basic education.

The majority of caregivers were women (76.7%), which is in line with similar studies (Almeida et al., 2017; Martins, 2006). Of the various roles women play, the role of caregiver is historically and culturally evidenced. For a long time, women have been responsible for providing care to family members with acute or chronic illnesses (Martins, 2006). They are all adults, with a significant percentage of the very old. Demographic aging and the increased incidence and prevalence of chronic or disabling diseases are key factors for the increase in the number of

cases of care-dependent persons in family contexts, and, consequently, the aging of the caregivers themselves. Older spouses are thus one of the greatest guarantees of support in old age (Lage, 2007). As for marital status, most of them are married or cohabiting. Their academic qualifications range from being able to read/write to having a Master's degree, with a prevalence of the 1st cycle of education. As for the professional situation, 40.0% are professionally active. Concerning kinship relationships with the patient, it is mostly the spouse who assumes the role of informal caregiver, which can also be assumed by children, parents, and siblings.

Concerning the clinical characteristics, most individuals had bulbar onset ALS, followed by spinal onset, and a small percentage of individuals had respiratory onset, contrary to what has been observed in other studies (Fearon et al., 2015). Half of the participants reported onset of symptoms less than 5 years earlier. However, 16.7% had been diagnosed more than 10 years earlier, which is in line with Maragakis and Galvez-Jimenez (2018), who reported that the average survival rate is 3-5 years but some people with ALS live more than a decade as a result of intense and careful assistance.

The results of the functional assessment of people with ALS indicate that most of them have a very severe degree of dependence and that the gross motor function was the most affected domain. For caregivers, this means that they have to perform the care for the patient in many physical activities, leading to a higher burden. The respiratory function was the dimension with the best functional scores, which can be explained by the timely use of technical aids such as NIV and mechanically assisted coughing.

The majority of caregivers had moderate levels of burden (56.7%). The results show that they have the lowest levels of burden in the RD dimension (25.67 ± 21.76), that is, the caregivers perceive that they meet the demands arising from their role. On the other hand, they had the highest levels of burden in the MEC dimension (66.39 ± 21.50). The demands inherent to prolonged and increasingly demanding caregiving activities may lead to progressive strain among the caregivers, their inability to use coping strategies, and, consequently, higher burden.

As for the caregiver's sociodemographic characteristics associated with burden, age was positively correlated with the FSupp dimension: the older the caregiver, the higher the level of burden in this dimension. Knowing that FSupp refers to the family's recognition and support in the face of events caused by the family member's disease and adaptation (Martins, 2006), we advance the hypothesis that the physical and cognitive decline inherent to the aging process leads to a higher perception of burden. Their own condition as older people may imply a higher perception of frailty and that they themselves need to be cared for. With regard to the degree of kinship, spouses have a higher burden than other family members in the FB and FSupp dimensions. Spouses are naturally expected to provide support and assistance at various levels in their life together and not in a life of dependence. This fact may explain the spouses' perception of a higher burden in these dimensions. Concerning the professional situation, the non-active caregivers had a higher burden than the active caregivers in the RD and FSupp dimensions. Despite the burden that the active caregivers may feel, the differentiation of roles may moderate this effect. The possibility of performing different tasks and contacting with other people may relativize the role of caregiver, leading to a lower perception of the impact of this role.

Significant negative correlations were also found between the level of burden and the QoL domains perceived by the caregiver. Caregivers with higher levels of burden have a worse perception of QoL, and the strongest correlations were found in the IPL, MEC, and Total QASCI dimensions ($r > 0.7$). Caregivers with higher levels of burden have more mental health changes that translate into fatigue and worse perceived QoL (Galvin et al., 2016; Pagnini et al., 2010).

The patient's clinical characteristics also impact the caregiver's perceived burden. The degree of dependence, the number of care hours, and the need for NIV influence the level of burden, and it should be considered that these characteristics define the type, amount, and quality of care. Therefore, they have a strong influence on perceived burden. The degree of dependence influences EB and IPL dimensions, and the more dependent the patient is, the higher the level of burden. This association is justified because the person with ALS who has a poor functional status is more dependent and demanding of the caregiver. The caregiver has to perform more and more complex tasks (e.g., mobility changes, need for respiratory and feeding support, or communication changes) that imply spending more time delivering care and a higher level of physical and emotional burden (Bruletti et al., 2015; Pinho & Gonçalves, 2016). The number of care hours was associated with the IPL and RD dimensions, indicating that the more care hours, the higher the burden. As described by de Wit et al. (2018), as the disease progresses, both in terms of the worsening of symptoms and dependence, caregivers have less time to fulfill their own needs and less energy to perform leisure activities. As for the RD dimension, caregivers may feel that patients ask for their help more often than necessary. As for the presence of NIV, the caregivers of people with NIV have a higher level of burden in the FB dimension. The disease itself involves a financial effort from the caregiver. Unemployment, sick leave, or even early retirement can have a significant economic impact on the lives of these families. On the other hand, this perception may worsen with NIV, which is one of the most adverse first measures that may contribute to the caregiver's perception that dependence is worsening.

Concerning the QoL of people with ALS, most of them always have difficulties. According to Bandeira (2010), worse QoL was found in Mobility and better QoL in Eating and Drinking. It should be noted that the Emotional functioning domain is not the one with the worst scores (56.25 ± 25.02), which can be explained by the different coping skills, but also by the clinical assistance, the existing support network, and the fact that patients remain at home.

The perceived QoL of people with ALS influences the level

of burden felt by the caregivers. The worse the perceived QoL of people with ALS in the Emotional functioning domain, the higher the level of burden felt by the caregivers in the EB and IPL dimensions. A similar situation was found in the Eating and Drinking domain of QoL, where the burden in the FSupp dimension is higher.
The limitations of this study were its sample size and sampling technique, which do not allow generalizing the results, and the restriction of the study to a specific region. Future studies in this area should include a larger and multicenter sample.

Conclusion

ALS is a rare, rapidly progressive degenerative disease of the central nervous system. In Portugal, it is estimated that 800 people have ALS. As the disease progresses, patients require a high number of care services, specialized clinical monitoring, specialized units for long-term hospitalization and caregiver's rest, timely and adapted technical aids given the rapid progression of the disease, faster and more specific responses from physical medicine and rehabilitation services, permanent caregiver for activities of daily living, and increased support to families at various levels, among others.
Understanding the factors related to QoL and burden is becoming increasingly important. Society must know the biopsychosocial impact of the disease on the lives of patients and caregivers in order to allow and encourage the development of public policies aimed at these people.
The results indicate that the sample of patients with ALS consists mostly of non-active adults with bulbar onset. The profile of caregiver corresponds to an adult family member, usually a spouse, with basic education, and a higher prevalence of women. The family is the first and most requested caregiver in care organization. The results reveal a moderate level of burden, indicating the importance of the support networks' response in reducing this burden, given that the caregivers are the key pillars for these patients to stay at home. Older caregivers, spouses, and non-active caregivers reported higher levels of burden, especially in the family dimension. The caregiver's perceived QoL is negatively influenced by burden.
The levels of burden are also higher in caregivers of more dependent patients who require more care hours and NIV. This study concluded that the worsening of the functional status and the need for complex care influence the levels of burden and perceived QoL.
Some recommendations emerged from this study to improve caregiver support: empowerment of caregivers through information and health education; relief of caregiver burden with a proactive response from professionals; timely assessment of caregiver needs due to the rapid progression of the disease; and flexible and timely allocation of resources.

Author contributions

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