Amyotrophic Lateral Sclerosis in Argentina: an approach to the realities of people with ALS and their care environments in the year 2023 *

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Keywords
Caregivers; people diagnosed with Amyotrophic Lateral Sclerosis; survey; care work; infrequent illnesses; health coverage; caregiver burden syndrome

Abstract

Introduction. This article explores and analyzes the daily experiences and realities of the universe of people diagnosed with Amyotrophic Lateral Sclerosis (ALS) and their care settings in Argentina in 2023. Methodology. The results of a self-administered online survey conducted from May to July 2023 on people with ALS and primary caregivers are presented. The study included the participation of 111 people: 56 diagnosed with ALS and 55 primary caregivers. **Results and discussion**. 71.7% of the people diagnosed with ALS surveyed considered their household income to be insufficient to meet their expenses. Reaching the correct diagnosis takes in most cases more than a year. The primary caregivers surveyed carry heavy caregiving workloads that, in most cases, exceed 8 hours per day. In addition, they show higher levels of exhaustion, discouragement, nervousness, and discomfort than people with ALS who participated in the study. Those who assume the role of primary caregivers are mostly women, which coincides with the evidence provided by the field of caregiving, which points to the close relationship between gender and caregiving. It is evident the pressing situation that people with ALS and their caregivers go through both economically and in terms of physical and emotional health. **Conclusions**. Based on the study's findings, a set of considerations and recommendations are offered to inform the design of policies and actions aimed at improving the situation of people with ALS and those who assume the role of primary caregivers. It also identifies possible future lines of research that would broaden and deepen the findings presented here.

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- Author 1: Supervision and writing of the original draft.
- Author 2: Acquisition of funds.
- Author 3: Visualisation.
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Esclerosis Lateral Amiotrófica en Argentina: una aproximación a las realidades de las personas con ELA y sus entornos de cuidado en el año 2023

Resumen

Introducción. El objetivo del presente artículo es explorar y analizar las experiencias y realidades cotidianas del universo de las personas con diagnóstico de Esclerosis Lateral Amiotrófica (ELA) y sus entornos de cuidado en Argentina en 2023. **Metodología**. Se presentan los resultados de una encuesta autoadministrada online realizada en los meses de mayo a julio de 2023 a personas con ELA y a cuidadores/as principales. El estudió contó con la participación de 111 personas: 56 con diagnóstico de ELA y 55 cuidadores/as principales. **Resultados y discusión.** El 71,7% de las personas con diagnóstico de ELA encuestadas considera insuficientes los ingresos de su hogar para afrontar sus gastos. Arribar al diagnóstico correcto demora en la mayoría de los casos más de un año. Los/as cuidadores/as principales encuestados/as soportan grandes cargas de trabajo de cuidados que, en la mayoría de los casos, supera las 8 horas diarias. Además, muestran mayores niveles de agotamiento, desánimo, nerviosismo y malestar que las personas con ELA que participaron del estudio. Quienes asumen el rol de cuidadores/as principales son, en su mayoría, mujeres, lo cual coincide con la evidencia aportada por el campo de los cuidados, en donde se señala la estrecha relación que existe entre el género y el trabajo de cuidados. Se evidencia la situación acuciante que las personas con ELA y sus cuidadores/as atraviesan tanto en el plano económico como en el de la salud física y emocional. **Conclusiones**. Con base en los hallazgos del estudio, se ofrecen un conjunto de consideraciones y recomendaciones destinadas a informar el diseño de políticas y acciones dirigidas a mejorar la situación de las personas con ELA y de quienes asumen el rol de cuidadores/as principales. Asimismo, se identifican posibles futuras líneas de investigación que permitirían ampliar y profundizar los hallazgos aquí expuestos.

Palabras clave
Cuidadores; cuidadoras; personas con Esclerosis Lateral Amiotrófica; encuesta; trabajo de cuidados; enfermedades poco frecuentes; cobertura de salud; síndrome del cuidador quemado

Esclerose Lateral Amiotrófica na Argentina: uma abordagem das realidades das pessoas com ELA e seus ambientes de atendimento no ano de 2023

Resumo

Introdução. O objetivo deste artigo é explorar e analisar as experiências e realidades cotidianas do universo de pessoas diagnosticadas com Esclerose Lateral Amiotrófica (ELA) e seus ambientes de atendimento na Argentina em 2023. Metodologia. São apresentados os resultados de uma pesquisa on-line autoadministrada realizada nos meses de maio a julho de 2023 com pessoas com ELA e cuidadores primários. O estudo envolveu 111 participantes: 56 pessoas diagnosticadas com ELA e 55 cuidadores primários. **Resultados e discussão**. 71,7% das pessoas diagnosticadas com ELA pesquisadas consideraram que sua renda familiar era insuficiente para cobrir suas despesas. A obtenção do diagnóstico correto leva, na maioria dos casos, mais de um ano. Os cuidadores primários pesquisados têm cargas de trabalho pesadas de cuidados que, no a maioria dos casos,, excedem 8 horas por dia. Além disso, eles apresentam níveis mais altos de exaustão, desânimo, nervosismo e desconforto do que as pessoas com ELA que participaram do estudo. Aqueles que assumem o papel de cuidadores primários são, em sua maioria, mulheres, o que é consistente com as evidências do campo de cuidados que apontam para a estreita relação entre gênero e cuidados. Está claro que as pessoas com ELA e seus cuidadores estão em uma situação muito difícil, tanto financeiramente quanto em termos de saúde física e emocional. **Conclusões**. Com base nos resultados do estudo, é oferecido um conjunto de considerações e recomendações para informar a elaboração de políticas e ações destinadas a melhorar a situação das pessoas com ELA e daquelas que assumem o papel de cuidadores primários. O estudo também identifica possíveis linhas de pesquisa futuras que poderiam ampliar e aprofundar os resultados aqui apresentados.

Palavras-chave
Cuidadores; pessoas com Esclerose Lateral Amiotrófica; pesquisa; trabalho de assistência; doenças raras; cobertura de saúde; síndrome do cuidador exausto

1. Introduction

Amyotrophic Lateral Sclerosis (ALS) is a progressive neurodegenerative disease that affects the voluntary motor system. It is considered a "rare disease" (RD) or "infrequent disease" (EPOF or EPF), which commonly leads to social and often professional ignorance about it (Gnavi, 2020).

According to data presented by the Fundación Esteban Bullrich (2021), worldwide, it is estimated that the incidence of this disease (new cases per year) is around 1 or 2 per 100,000 people and that its prevalence (number of people with the disease simultaneously) is approximately 5 cases per 100,000 people. It makes it the third most prevalent neurodegenerative disease after Alzheimer's and Parkinson's disease.

At present, there are no precise figures on the number of people diagnosed with ALS in Argentina (Fundación Esteban Bullrich, 2021), but there are different estimates that yield dissimilar results. Thus, for example, a retrospective epidemiological study conducted in the Autonomous City of Buenos Aires (CABA) yielded results aligned with world averages: an incidence of 1.04 per 100,000 p/year and a prevalence of 3.25 per 100,000 p/year (Pérez et al., 2017). However, other research conducted in specialized centers located in the same city yields a higher incidence. Thus, a study conducted at the Ramos Mejía Hospital in the period 2001-2008 reported an incidence of 6 per 100,000 inhabitants (Bettini et al., 2011). On the other hand, a 7-year retrospective study conducted at the Italian Hospital (HIBA) revealed an incidence of 3.17 per 100,000 inhabitants and a prevalence of 8.86 per 100,000 people (Bettini et al., 2011). Although there has been an initiative at the national level to carry out a registry of people with ALS, it has not been completed due to the scarcity of registered cases. Some of the difficulties encountered for this have been the heterogeneity of the health system and the lack of articulation between the National Government and the provinces, among others (Fundación Esteban Bullrich, 2021).

Argentina has a vast regulatory framework that enshrines and protects the right to health of its inhabitants. Its healthcare system is characterized by three coexisting subsystems: (i) the public system - regulated by Ley No. 23,661 of 1988, which created the National Health Insurance System; (ii) the Social Security system - regulated by Ley No. 23,660 of the same year; and the private system - which refers to prepaid medical insurance companies regulated by Ley No. 26,682 of 2011. The minimum medical assistance benefits that both social security and prepaid health insurance companies must cover on a mandatory basis and independently of the health plan are set forth in the Compulsory Medical Program (Decreto 492/1995).

Along with these regulations that seek to regulate the functioning of the health system, it is also possible to point out others that seek to protect and guarantee more directly the rights of patients, taking into account their particular state of health. Among them, it is worth mentioning Ley No. 26,529 of 2009 on Patients' Rights in their Relationship with Health Professionals and Institutions. This law establishes a series of patients' rights, including the right to decide to receive (or not) the necessary health information related to their health, which is defined in article 3 as: "information that, in a clear, sufficient and adequate manner to the patient's capacity of understanding, informs about their state of health, the studies and treatments that need to be performed and the foreseeable evolution, risks, complications or sequelae of the same". More recently, also closely linked to the requirements arising from ALS, Ley No. 27,678 on Palliative Care was passed in 2022, which "aims to ensure patients' access to comprehensive palliative care services in their different modalities, in the public, private and social security spheres, and the accompaniment of their families in accordance with these provisions" (art. 1). To this end, it establishes the formation of interdisciplinary and multidisciplinary work teams, the implementation of actions to promote access to essential drugs and training in palliative care for health personnel.

Even so, ALS does not seem to have entered the agenda of the national legislature yet since, to date, no national laws have been passed that directly address issues related to this disease -such as its diagnosis, treatment, and research- (Fundación Esteban Bullrich, 2021). Even so, ALS does not seem to have entered the agenda of the national legislature yet since, to date, no national laws have been passed that directly address issues related to this disease -such as its diagnosis, treatment, and research- (Fundación Esteban Bullrich, 2021). Exceptionally, only one of the 23 provinces of the country, Corrientes, has had a specific law on ALS since 2011 ("Ley N° 6.072"), which, however, is still awaiting its regulation. Faced with this situation, those who suffer from this disease are only protected through regulations that involve them in a less specific, more general, and indirect way. Thus, Ley No. 26,689 on Rare Diseases -among which ALS is included-, enacted in 2011, promotes comprehensive health care for those who suffer from this type of disease and, among other issues, establishes the obligation for social security and prepaid health insurance companies to guarantee coverage. Even so, several of the articles

that make up this legislation have not been regulated to date, including the one that assigns to the State the responsibility of guaranteeing comprehensive health care for those who do not have coverage by prepaid health insurance companies or social security companies -art. 7- (Fundación Esteban Bullrich, 2021). Furthermore, only 18 of the 24 jurisdictions that make up the national territory have adhered to the law above, which means that 6 of them have not yet done so. Complementarily, there is another regulation that reaches a subgroup of those who suffer from this disease, namely, those patients who are able to apply for the Single Certificate of Disability -also known as CUD- (Ley 22.431). Thus, those who obtain a CUD can, through the different laws related to disability, access tax benefits and pensions, as well as receive the medical and welfare benefits required for their treatment (Fundación Esteban Bullrich, 2021).

This article presents some of the main results obtained in quantitative research conducted in 2023 in Argentina, whose purpose has been to produce knowledge about this problem from a sociological and interdisciplinary perspective by surveying people diagnosed with ALS and primary caregivers. More specifically, the objective was to explore and analyze the main dimensions of their living conditions.

In this way, we have sought to contribute to the production of knowledge on a comparatively underexplored topic at the national and international level, as well as to generate evidence that contributes to making visible the realities of those who suffer from this rare disease and their care environments. In doing so, the aim is to provide empirical evidence that can be retrieved and used by different institutions -especially state agencies and third-sector organizations- to inform the design of new regulations, policies, and actions aimed at improving the situation of people diagnosed with ALS and those who act as their main caregivers.

The article is structured in a total of 4 sections and conclusions. After this introduction, in the section entitled Background Review, a brief compilation of research background related to the topic studied is offered. At the same time, the conceptual tools used in the analysis of the findings obtained have been recovered. Section 3, Methodology, details the research activities carried out within the framework of the study, as well as the ethical considerations adopted. Next, in the section Results and Discussion, the main results of the study are presented, described, and analyzed, using theoretical tools that contribute to their interpretation and establishing a dialogue with the results of previous studies. Finally, the Conclusions offer a set of recommendations or considerations for the design of policies and actions aimed at accompanying those who suffer from this disease and their care environments. It also identifies possible future lines of research that will contribute to deepening and broadening the findings of this study.

2. Background review

Amyotrophic lateral sclerosis (ALS) is a neurodegenerative disease with sudden onset and rapid progression, leading to complex disabilities with fatal consequences (Cipolletta and Amicucci, 2015). To date, not only the causes of this disease have not been identified, but also there is still no cure. Consequently, the progression of this disease is inevitable, and the therapeutic objective, in the absence of a satisfactory cure, focuses on preventing or delaying its progression by procuring the best possible quality of life for the patient and his family (Gómez, Ballester and Cantus, 2020). In other words, since its inception, treatment has been based on palliative symptom control measures (Ando, Cousins and Young, 2019; Camacho, Esteban y Paradas, 2014). It is extremely costly, which is why it is considered a disease that generates catastrophic expenses (Fundación Esteban Bullrich, 2021).

ALS is one of the hardest diseases emotionally for patients, their families, and caregivers (Ballester, García Berenguer and Lloret Irles, 2022). The low life expectancy of people with ALS entails a high level of suffering for the sufferer, as well as for their environment, since it requires an increasing demand for care as it continues to develop (Dumancela Mina, 2012). In this sense, studies linked to the disease have two main foci of study. On the one hand, those who focus on people with ALS and, on the other hand, those who focus on the patient's environment, either family members and/or those who assume the role of caregivers.

In relation to the first group, the first line of work that focuses on the diagnostic process has been identified. Richards, Morren and Pioro (2020) provide a review of most of these studies related to the diagnostic delay in the ALS patient population. These authors show that the shortest interval - calculated from the median - from symptom onset to diagnosis was 9.1 months, as reported in a national study of ALS in France. In contrast, the longest time to diagnosis has been 27 months, as reported in a U.S. study between 2008 and 2009. A study conducted in Spain also points in the same direction,

evidencing that the mean diagnostic delay was 13.1 months (median 11.7) (Vázquez-Costa et al., 2021). Most studies reported a delay of 10 to 16 months to reach the diagnosis of ALS.

In summary, the diagnostic delay seems to be independent of the country and the health system and has not changed significantly in the last 20 years despite the efforts made (Vázquez-Costa et al, 2021). The diagnostic delay of ALS seems to be determined by disease-specific factors and independent of the health system (Vázquez-Costa et al, 2021). Although the disease progresses rapidly, early diagnosis can be a major challenge due to its insidious onset, clinical heterogeneity, and lack of clear diagnostic markers (Vázquez-Costa et al, 2021). Accurate diagnosis of a patient presenting with suspected ALS is important. However, early diagnosis of ALS is often difficult, resulting in a diagnostic delay of approximately one year, which results in approximately one-third of the overall survival time (Vázquez-Costa et al, 2021). Mistaking ALS for another, often more benign disease can result in an inaccurate prognosis and an inadequate treatment program (Iwasaki, Ikeda and Kinoshita, 2001). Some of the barriers that have been identified in diagnosis include multiple referrals to specialists, unnecessary tests and procedures/surgeries, and misdiagnosis (Richards et al., 2020;Vázquez-Costa et al, 2021).

Overall, studies agree that these delays could be reduced with increased awareness and clinical education about ALS directed toward both the general population and primary care providers and all physicians who evaluate these patients prior to making a definitive diagnosis (Richards et al., 2020;Vázquez-Costa et al, 2021).

Complementarily, within the first group of studies, which focuses on ALS patients, is a second strand that focuses on patient emotional issues. As a fatal and rapidly progressive disease, ALS has devastating effects on the mental health of people with ALS (Smith, Crossley, Greenberg, Wilder and Carroll, 2000).). As the disease progresses, patients face physical disabilities, and gradually, the incidence of psychological disorders such as depression increases (Dutta, Thammisetty, Boutej, Bareil and Julien, 2020). Evidence shows that ALS patients are more susceptible to being depressed and feeling disappointed (Beswick et al., 2021), as well as developing a range of emotional problems, especially anxiety, fear, and depression (Unglik et al., 2018). Physical disability and impending death increase the likelihood of hopelessness and the desire for medical assistance for suicide (Paganoni et al., 2017).

Depression is recognized as one of the most important complications of ALS (Chiò et al., 2019). An extensive review of existing studies linked to depression in people with ALS can be found in Heidari et al. (2021). According to various studies, its prevalence in ALS patients varies from 10% to 45% (Prado et al., 2017; De Marchi et al., 2019). ALikewise, one study has shown that male patients have more symptoms of depression than female patients and that anxiety and depression are more common in older patients at the onset of the disease (Wicks et al., 2007).

There is a large number of investigations that focus on quality of life (QoL), which is highlighted as a very relevant issue in people with ALS (Ilse et al., 2015). Considering that ALS is a disease that, to date, has no cure, improving the quality of life of patients is the main focus of treatment (Ando et al., 2019). It is noteworthy that, in the different studies, there is no consensus on the indicators that should be used to measure it, which is rooted in the fact that there are multiple definitions of quality of life (Ilse et al., 2015; Ando, et al., 2019).

Quality of life measurements should capture several individual factors, including physical, psychological, social, and spiritual aspects. In general, quality of life (QoL) is determined by health-related factors but also by other factors that include employment status, as well as religious activities and/or with family and friends (Ilse et al., 2015). Standard indicators of quality of life are wealth, employment, residence, physical and mental health, education, recreation and free time, and social belonging (Martínez-Campo et al., 2017). Many ALS studies focus specifically on health-related quality of life (HRQOL), which focuses on the physical and mental aspects of quality of life that affect health and disease (Ilse et al., 2015; Sánchez-López, Perestelo-Pérez, Ramos-Pérez, López-Bastida and Serrano-Aguilar, 2014). HRQOL is a global and multidimensional concept that refers to both objective conditions and subjective components, and these include all relevant and health-related aspects of a patient's life. It reflects how individuals perceive and react to their health status and other health-related factors, including perceptions of physical and mental health and their correlates (functional status, social support, and socioeconomic status) (Sánchez-López et al., 2014).

Various investigations have sought to delve into the factors that determine the quality of life of people with ALS. The relationship between disease severity and quality of life in ALS patients has been a matter of debate (Ilse et al., 2015).). There is evidence that, although HRQoL in ALS patients decreases during the disease, overall quality of life seems to

remain at a stable level, even in patients with advanced ALS (Cupp et al., 2011; Goldstein, Atkins y Leigh, 2002; Neudert, Wasner and Borasio, 2004). It reflects the "paradox of well-being," which is well-known in studies related to the quality of life (Ilse et al., 2015). However, other studies do show that both the clinical status and quality of life deteriorate in patients with ALS as the disease progresses (Martínez- Campo et al., 2017).

The social context does seem to be correlated with quality of life: several studies have highlighted the importance of social support for quality of life in patients with ALS (Goldstein et al., 2002; Chiò et al., 2004; McLeod and Clarke, 2007; Matuz, Birbaumer, Hautzinger and Kübler, 2010; Ganzini, Johnston and Hoffman, 1999). Asimismo, hay estudios que evidencian que el bienestar psicológico determina su calidad de vida (Neudert et al., 2004).

In this context, for example, Nelson et al. (2003) spoint out that ALS patients who had a better quality of life were younger, had a shorter duration of the disease, and experienced less disease severity as well as more adequate finances and less stress due to the characteristics of the disease.

The second group of studies, which focuses on the patient's environment, whether on family members and/or caregivers, mostly focuses on quality of life, mental health, and the needs of the environment. Caregivers are key figures for ALS patients, both in terms of care and the physical and emotional support they provide, and they play a key role in decision-making related to the disease (Galvin et al., 2018).

Throughout the disease, in a short period, patients go through a situation of loss or decrease in their self-sufficiency, which results in the inability to carry out daily life activities without support from third parties. As ALS progresses, patients require assistance to move, eat, dress, and nursing care. In its more advanced stages, this disease requires the presence and care of health professionals who, together with the family and/or emotional environment, play a very important role in the course of this disease (Gnavi, 2020).

The family is usually the main provider of this care (Galvin et al., 2018), with the patient's partner normally assuming the role of the primary caregiver (Cipolletta y Amicucci, 2015; Gómez et al., 2020), which impacts both the number of hours they dedicate to caring for the sick person and the emotional consequences and implications (Gómez et al., 2020). ESometimes, this person spends more than 11 hours a day with the patient (Pagnini, 2013). In this context, there are more and more studies that aim to understand the consequences that all this entails for the so-called primary caregivers. The primary caregiver is understood to be the person who plays the role of caring for and attending to the needs of the sick person. Different studies carried out agree that those who occupy this role are usually female (Paz-Rodríguez, Andrade-Palos y Llanos-Del Pilar, 2005; Perdomo-Romero y Ramírez-Perdomo, 2017).

Studies that focus on the needs of caregivers highlight different relevant aspects. On the one hand, Galvin et al. (2018) underline the importance of having external support and assistance. The need for these services to be provided flexibly is emphasized so that they adapt to the situation at different stages of the disease. These authors also point out the need for caregivers to have someone to talk to and to listen to them, as well as to have time away from care and the opportunity to pursue their interests (Galvin et al., 2018).

Ballester et al. (2022) review the works that focus on the psychosocial needs of people with ALS and their caregivers. In relation to the latter, in addition to the factors mentioned above, there is also the need for financial support - since the disease causes a loss of employment and income and an increase in expenses - as well as needs related to the health system, such as reduction of referral and diagnosis times, coordination between specialists and caregivers, facilitating access to the system and empathy on the part of medical staff.

In addition to needs, various studies focus on well-being and, especially, on the burden of caregivers (Aoun et al., 2013; Pinho and Goncalves, 2016; Bergin y Mockford, 2016; De Wit et al., 2018). Knowing which factors are associated with the caregiver's burden is essential to designing strategies that allow supporting those who occupy this role (De Wit et al., 2018). An extensive review of the published literature regarding which patient and caregiver factors are associated with the burden of informal caregivers of ALS patients can be found in De Wit et al. (2018).

As mentioned above, the progressive nature of the disease accentuates the patient's dependence on the primary caregiver. However, conclusions drawn from the evidence on the relationship between the caregiver's psychological state and the

patient's disability are controversial (Cipolletta and Amicucci, 2015). Some studies have shown that caregiver burden and depression are positively associated with physical disability in ALS patients (Pagnini et al., 2010) In contrast, other studies show that caregivers with lower levels of quality of life are not always those who have to care for people with greater physical or psychological problems (Lo Coco et al., 2005). Rabkin, Albert, Rowland, and Mitsumoto (2009) also found no clear association between the degree of patient disability and caregiver depression or perceived burden. In any case, most of the studies carried out to date agree on the existence of what some authors have agreed to call the "burnout syndrome" (Gómez et al., 2020; Martínez Pizarro, 2020). As Gómez et al. (2020) suggest, people who take on the care of a person with a pathology of this type often experience serious physical and mental health problems over time. Thus, they must face, on the one hand, the stress generated by the act of caring itself and also the deterioration caused by assisting for a long period. All of this usually leads to physical, psychological, and even social or economic problems. The results of these studies underline the need to offer care and support to each individual in the family, as they show that both patients and family members are affected by a decrease in their quality of life and by anxiety (Olsson Ozanne, Strang and Persson, 2011).

3. Methodology

The article presents the results of quantitative research carried out in the months between May and July -inclusive- of 2023, which was possible to carry out because it was one of the winning projects of the "Public Contest for Social Research on ALS" carried out by the Ministry of Science, Technology, and Innovation of the Nation (Argentina) and the Esteban Bullrich Foundation.

It consisted of a self-administered online survey, which could be answered by cell phone or computer in no more than 25 minutes. Two different and complementary questionnaires were designed by the subuniverse: one for people with ALS and another for primary caregivers, all of whom will be residing in Argentina in 2023. People were contacted by email, mainly through a contact list provided by the Esteban Bullrich Foundation. In addition, the project was disseminated through social networks, mainly Facebook, Instagram, and LinkedIn. All persons who expressed interest in participating were sent an initial email with information about the study and then another with two individual links to access and answer the questionnaires. Both the email and the survey interface included information about the characteristics of the research, its objectives, how the data would be used, and the institutions involved in order to guarantee the conditions for informed consent. The approximate duration of the survey was also specified, and the voluntary, anonymous, and confidential nature of participation was clarified, as well as the guarantee of safeguarding the information collected. In addition, they were given contact information for the research team so that participants could contact them if they wished. All this information was included at the beginning of the online survey, which could only be answered if this informed consent was accepted.

The study included the participation of a total of 111 people: 56 answered the survey for people diagnosed with ALS, and 55 answered the survey for primary caregivers. Among them, 36 can be considered a dyad, which allows the information collected to be linked since the survey was answered by both the person with ALS and their respective primary caregiver. The inclusion criteria were that they were people with ALS or primary caregivers of a person with this diagnosis - the latter understood as those people who dedicate more time to accompanying them in daily tasks - and that they were residing at the time of the study in one of the 23 provinces of Argentina. The data obtained were processed using the SPSS statistical software, and the figures were prepared using Excel.

Specifically, the 56 people diagnosed with ALS who responded to the survey have an equal composition in terms of gender: 47.3% identify as women and 52.7% as men. The average age is 57 years. The educational level of the people with ALS surveyed is high: 18.2% report having completed university, and 34.5% have incomplete university or completed tertiary education. The remaining 47.2% is distributed between 23.6% who report having completed secondary school or incomplete tertiary education and 23.6% who have incomplete secondary school as their highest educational level. Regarding the province of residence, most of the people with ALS surveyed are concentrated in the Province of Buenos Aires (60%): 16.4% in CABA and 43.6% in Buenos Aires. Secondly, 9.1% (5 cases) reside in Córdoba and 7.3% (4 cases) in Santa Fe. The remaining 1.8% live in other provinces -3.6% of people who reside in Catamarca (1), Chaco (1), Chubut (1), Formosa (2), La Rioja (1), Misiones (1), Neuquén (2), Río Negro (2), San Juan (1) and Santiago del Estero (1)-.

On the other hand, regarding the 55 people who are primary caregivers of people with ALS and reside in Argentina in 2023 who responded to the survey, it is important to highlight the marked presence of women: 81.5% identify as women,

and 18.5% as men. The age distribution shows that caregivers are younger than people with ALS. While the vast majority of people with ALS (77.8%) are 50 years old or older, this percentage is reduced to 51% in the case of caregivers. Among them, there is a greater presence of younger age groups: 27.5% are between 40 and 49 years old, 15.7% are between 30 and 39 years old, and 5.9% are under 30 years old.

Likewise, the educational level of the caregivers surveyed is high, even higher than that of the patients. 37% reported having completed university education, and 27.8% have completed tertiary education or incomplete university education, while these percentages are 18.2% and 34.5% -respectively- in the case of people with ALS. The remaining 35.2% is distributed between 27.8% of caregivers who report having completed secondary education or incomplete tertiary education as the highest educational level achieved and a small proportion who have even incomplete secondary education (7.4%).
Below are the most outstanding findings of the survey, both on people with ALS and on those who assume the role of primary caregivers of patients with this disease.

4. Results and discussion

The results obtained from the survey show the complex reality that people with ALS in Argentina must face, as well as the major disruption they experience in their daily lives. In this sense, on the one hand, the negative economic impact that going through this disease has on household income is noteworthy, which most of the people surveyed consider insufficient to cover their expenses, including ALS treatments. The majority of people with ALS surveyed (62.3%) said that the income their household currently receives is insufficient to cover their expenses, including ALS treatments, even though they consider that these were sufficient before the first symptoms of the disease. In contrast, 28.3% consider that the income received by the household is and was sufficient to cover expenses, and 9.4% stated that the household income has always been insufficient, both before the disease and at present (Figure 1).

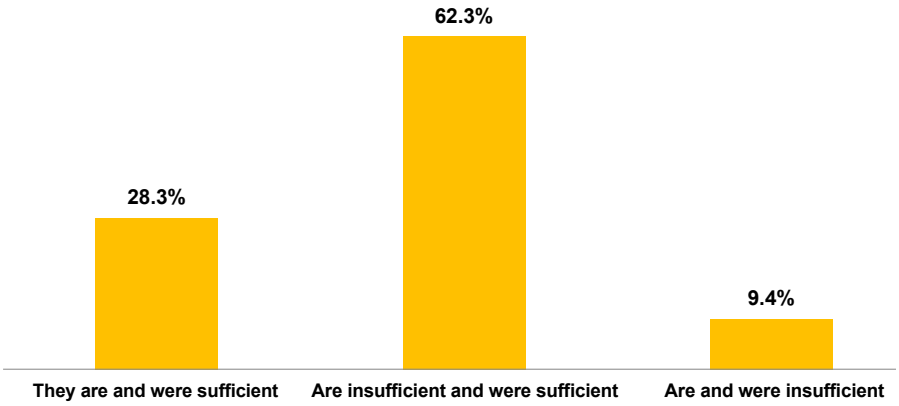

Figure 1. Perception of the sufficiency of household income before and after the first symptoms of ALS (%)
Source: Prepared by the authors based on survey data.

In parallel, the majority of people diagnosed with ALS surveyed (74.1%, equivalent to 40 cases) stated that they were not working at the time of this study. It is due, in most cases, to the symptoms and consequences of the disease: among those who stated that they were not working, more than half (59%) indicated that they had had to stop working due to the symptoms and consequences of ALS.

Even so, a considerable percentage, 25.9% (representing 14 cases), indicated that they were working. Regarding the occupational status of those who make up this last group of people who are currently working, 64.3% (9 cases) stated that they were salaried employees, 28.6% (4 cases) stated that they worked for their own company, business, or activity and only 7.1% (1 case) stated that they were an employer. In any case, almost all of them indicated that they had to make changes in their work due to ALS - only one person stated the opposite. Predominantly, the respondents in this sub-universe indicated that they had to reduce their working hours (42.9%), make changes in their daily work tasks/activities (35.7%), and/or start teleworking (28.6%). To a lesser extent, they also indicated that they had to introduce breaks throughout the working day (14.3%) and/or make or request adaptations in their workspaces and/or furniture (14.3%).

In line with the pressing economic situation faced by many of the households of the people with ALS surveyed, the results obtained in the survey show that the majority of them, 62.5%, do not receive any pension or financial aid that contributes to alleviating their pressing economic reality. However, 37.5% do so: 32.1% indicated that they are beneficiaries of a pension or state-type financial aid, and 3.6% (2 cases) indicated that they receive financial help from friends and/or family. Only 1.8% (1 case) of the respondents belonging to this sub-universe receive help from both sources.

In this context, there is also a significant percentage of people with ALS who declare that they pay for the expenses associated with the disease themselves, even though they have health coverage. The majority of the people with ALS surveyed (91.1%) have health coverage: 33.9% (19 people) have social security, another 33.9% have prepaid or private insurance, 17.9% (10) have PAMI, and 5.4% (3) have a state or federal program or plan. 8.9% (5 cases) do not have health coverage (Figure 2).

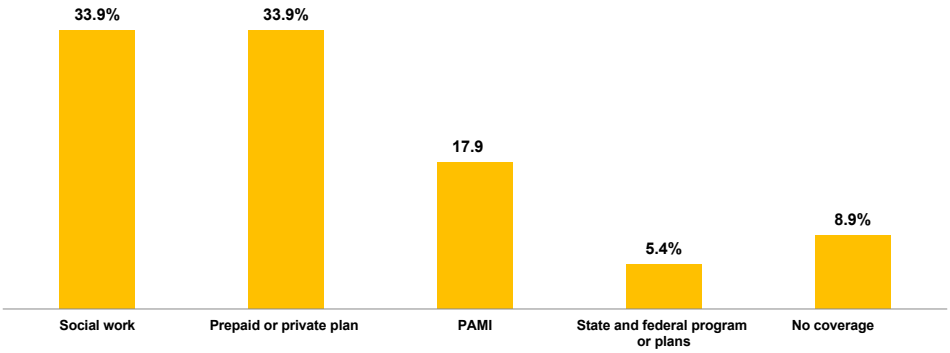

Figure 2. Type of health coverage of people with ALS surveyed (%)
Source: Prepared by the authors based on survey data.

Even so, the fact that most people with ALS have health insurance does not guarantee coverage of all expenses associated with the disease. Although more than half of the people surveyed (63.6%, 35 people) indicate that the main way of covering the expenses associated with the disease is through their social security, prepaid or private plan (Figure 3), at the same time, a considerable percentage of people pay for them privately. Specifically, 14.5% of the respondents (8 people) stated that they do so mainly through financial resources provided by friends and/or family and 12.7% (7) with financial resources from home (Figure 3). EThis results in 27.2% (15) using out-of-pocket expenses to cover expenses associated with the disease.

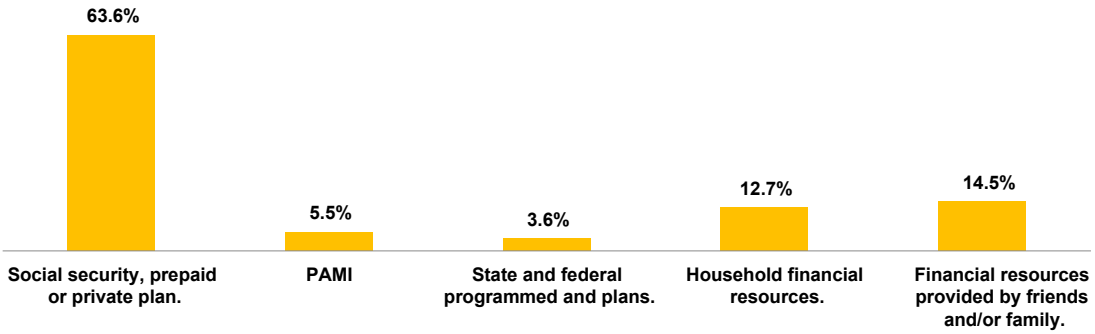

Figure 3. Main way of covering expenses associated with the disease (%)
Source: Prepared by the authors based on survey data

The comparison between the type of health coverage of the people with ALS surveyed and the main form of coverage of the expenses associated with the disease allows us to verify that a portion of the people with ALS declare that they pay for the expenses associated with the disease on their own, even if they have health coverage. There are 22.2% of people (4 cases) who have social security and, in any case, they use household economic resources as the main form of coverage for expenses, as well as 33.1% (1 case) of the people who are covered by some state or federal program or plan. Likewise, 50% (5 cases) of the people who have PAMI indicate that they cover the expenses from household economic resources

or resources provided by family and/or friends. Those who have prepaid or private plans are the only ones who, in their entirety, report that said health coverage is the main way in which they pay for the expenses associated with ALS.

On the other hand, as part of the most relevant results, it is worth mentioning that more than half of the people with ALS surveyed mentioned that obtaining the correct diagnosis took more than a year. This finding verifies that Argentina has levels of delays similar to those indicated by previous studies (Richards et al., 2020), lwhich seems to indicate that these are independent of the country and the health system (Vázquez-Costa et al, 2021). More than half of the respondents (57.1%, 32 people) took more than a year to obtain the correct diagnosis if the first symptoms of the disease were considered as a starting point. 21.4% (12) took between 7 months to a year, and 17.9% (10) waited 3 to 6 months. Only 3.6% (2) obtained it in less than three months (Figure 4).

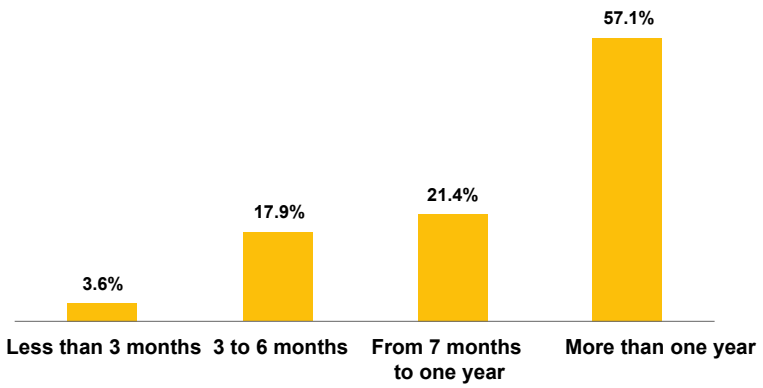

Figure 4. Time elapsed from the first symptoms of ALS to reaching the correct diagnosis of the disease (%)
Source: Prepared by the authors based on survey data**.**

The results suggest that, indeed, the delay in diagnosis of ALS seems to be determined by factors specific to the disease (Vázquez-Costa et al., 2021) and independent of the type of health coverage available. Only 20% (1 case) of those without coverage indicate that their diagnosis took more than a year, while the majority of them (80%, 4 cases) indicate a shorter time. On the other hand, among those who have prepaid or private insurance, there is a higher proportion of people who indicate a delay in diagnosis of more than a year (47.4%, 9 cases). The same occurs among those who have PAMI or social security: 60% (6 cases) and 68.4% (13 cases), respectively, indicate that their diagnosis took more than a year. Although it is important to highlight that the small number of cases does not allow for conclusive statements to be made in this regard, the results seem to confirm that early diagnosis is a great challenge due to issues inherent to the disease, including its clinical heterogeneity and the lack of clear diagnostic markers (Vázquez-Costa et al., 2021).

The time taken by the health system is one of the major obstacles that people with ALS have to face: 71.4% of the people surveyed believe that the administrative-bureaucratic time taken by the health system hindered, delayed, or hindered effective access to the elements required for the treatment of the disease, much or quite a lot, compared to 28.6% who believe the opposite, that is, that the time taken by the health system delayed access very little or not at all.

Even so, the evaluations that people with ALS have of health personnel are markedly favorable. The majority of the people surveyed positively evaluated the amount of information that was provided to them by health personnel about the disease: 41.1% stated that they received a lot of the information they needed or wanted to know about ALS, and 19.6% said that they were given all of it. Even so, a considerable percentage, 37.5%, believe that they were given little, and 1.8% that they were given none of the information about the disease that they needed or wanted to know. Similarly, people predominantly agree with the positive evaluation of the treatment received by health personnel: 80.4% consider it good or very good. A small percentage evaluate it negatively; 8.9% consider it bad or very bad, while 10.7% consider it average (Figure 5).

The gradual loss of autonomy that ALS brings makes caregiving a central dimension of the study. In this regard, it is highlighted that, according to what was stated by the respondents, the main caregivers are almost entirely family members.

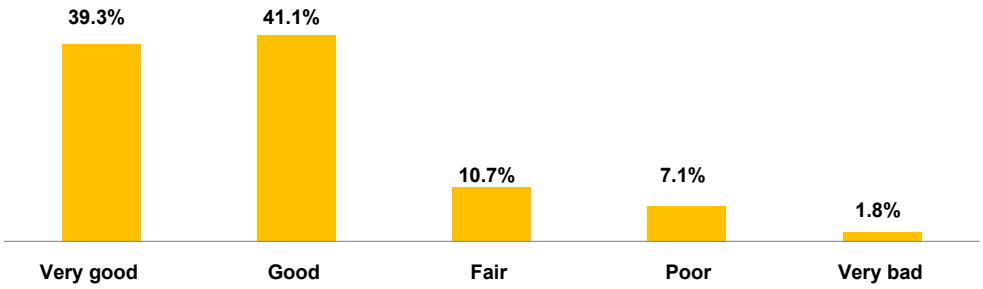

Figure 5. Evaluation of the treatment received by health personnel (%)
Source: Prepared by the authors based on survey data.

They are mainly couples: women in 44.4% of cases and men in 20.4%. These results are in line with those of previous studies carried out in other contexts, which show that it is normally the patient's partner who assumes the role of the main caregiver (Cipolletta and Amicucci, 2015; Gómez et al., 2020). They are followed by daughters in 14.8% of cases and sons, although in a markedly lower percentage in 3.7% of cases. In 1.9% of cases, the main caregiver is the mother of people with ALS. Outside of family ties, in 13% of cases, a paid person is the one who dedicates the most time to accompanying the people with ALS surveyed, and in 1.9% of cases, it is friends or acquaintances who do so (Figure 6).

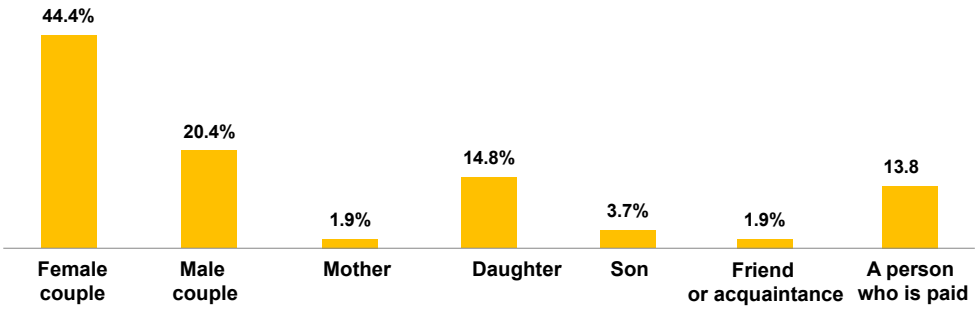

Figure 6. Person who assumes the role of primary caregiver for people with a diagnosis
Source: Prepared by the authors based on survey data.

It is interesting to note that the vast majority of the men with ALS surveyed (82%) indicate their female partner as their main caregiver. In contrast, in the case of the women diagnosed with ALS surveyed, the main caregivers are divided between their male partner (36%, 9 cases), their female daughter (28%, 7 cases), and a person who is paid (28%, 7 cases). Even among women, only 8% (2 cases) indicate their male son as their main caregiver. These data seem to suggest the central role of women as the main responsible for providing well-being and care to other people, as indicated in the literature both in relation to people with ALS and other populations (Paz-Rodríguez et al., 2005; Zibecchi, 2014; Perdomo-Romero y Ramírez-Perdomo, 2017). In summary, the results of this study show that 85.2% of the people who provide care are family members, and among them, 71.7% are women (partner, mother, or daughter).

This situation becomes more complex if we take into account that the majority of the primary caregivers surveyed spend more than 8 hours a day assisting the people with ALS under their care in different tasks and activities: more than half (56.6%) spend more than 8 hours a day on this task, with 45.3% spending more than 12 hours, far exceeding a working day for the market (Figure 7). The number of hours that primary caregivers spend caring for the sick person has been highlighted in previous literature, highlighting the implications that this has on both the physical and emotional health of caregiver (Pagnini, 2013; Gómez et al., 2020).

The high daily workload that the people surveyed dedicate to caring for the person with ALS is surely associated with the cohabitation situation: the majority (63%) state that they live with the person with ALS every day of the week. However, there is also a notable proportion of people who do not live with the person (27.8%) and a small portion who do so only some days (9.3%) (Figure 8).

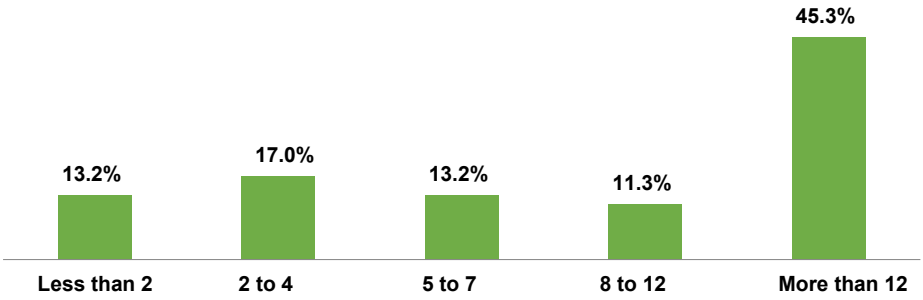

Figure 7. Daily hours that the surveyed caregivers spend caring for the person with ALS (%).
Source: Prepared by the authors based on survey data.

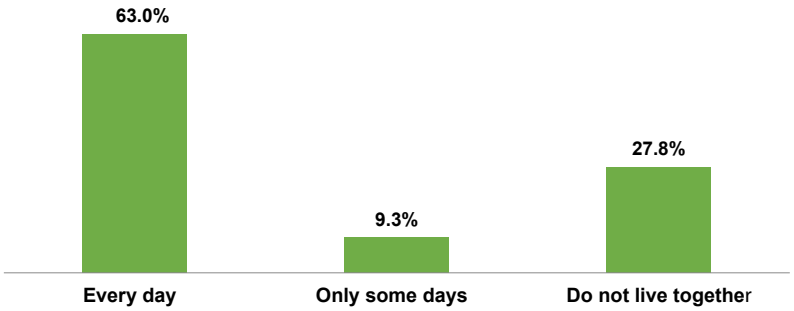

Figure 8. Caregivers surveyed according to whether they live (or not) with the person with ALS (%)
Source: Prepared by the authors based on survey data.

It is also noteworthy that the vast majority of caregivers (86.5%) have been doing this task for at least a year (Figure 9). Although it is not possible to know whether the daily hours dedicated to care have been constant over this time, the data suggest a situation of heavy care workload for the caregiver.

Finally, the results show the multiple and, in some cases, drastic changes that caregivers experience in their daily lives after assuming this role; transformations that are characterized by a strong reduction in the time that they can devote to their work, their personal life, and their recreational and leisure activities. In line with this, caregivers register negative impacts on their physical and emotional health, which is expressed in the higher levels of exhaustion that they manifest compared to those declared by the patients. This finding is in line with the majority of studies that have been carried out to date linked to caregivers, which agree on the existence of what some authors agreed to call the "burnout syndrome" (Gómez et al., 2020; Martínez Pizarro, 2020).

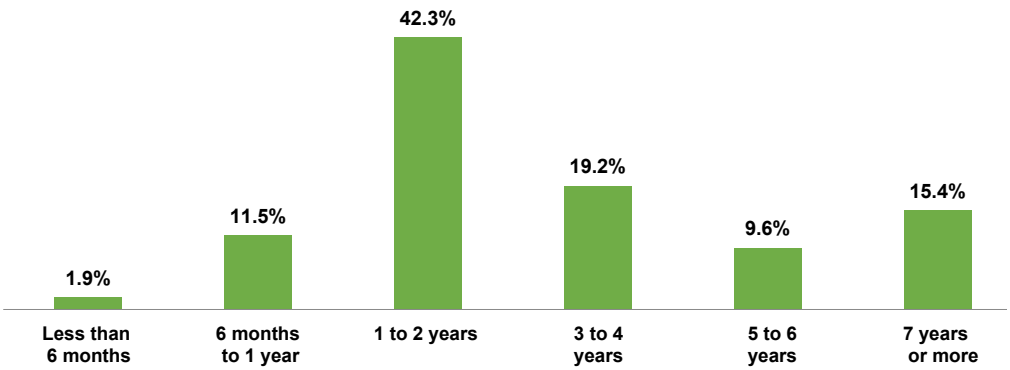

Figure 9. Time that the caregivers surveyed have been caring for the person with ALS (%)
Source: Prepared by the authors based on survey data.

In this context, it is not surprising that 66% of the caregivers surveyed have indicated that, since they have been caring for the person with ALS, they have felt especially stressed, anxious, and/or depressed, this being the main change noted (Figure 10). In second place are those who indicate that they have less time for themselves (49.1%) and then those who indicate that they have less time for leisure activities (45.3%). These results are part of the line of studies that focus on the needs of caregivers (Ballester et al., 2022), among which the need to have time away from care and the opportunity to pursue their interests stands out (Galvin et al., 2018). Only 5.7% (3 people) have indicated that they have not experienced any change.

For all the reasons previously mentioned, it is not surprising that 41.2% and 47.2% of the surveyed caregivers report a high level of nervousness and exhaustion (Figure 11). In general terms, it is observed that the surveyed caregivers show higher levels of exhaustion, discouragement, nervousness, and discomfort than people with ALS.

These results show that ALS is an emotionally difficult disease for both patients and caregivers (Ballester et al., 2022). Several studies focus on this field and explore, on the one hand, the differences in the psychological and emotional state and quality of life of ALS patients according to age, gender, social support, and degree of disease progression and/or physical disability (Ganzini et al., 1999; Goldstein et al., 2002; Chiò et al., 2004; Wicks et al., 2007; McLeod y Clarke, 2007; Matuz et al., 2010; Dutta et al., 2020). This study sought to explore these questions. In relation to age, the results of the survey suggest that younger people with ALS (under 50) are more likely to show a high level of discouragement, nervousness, and malaise (Figure 12).

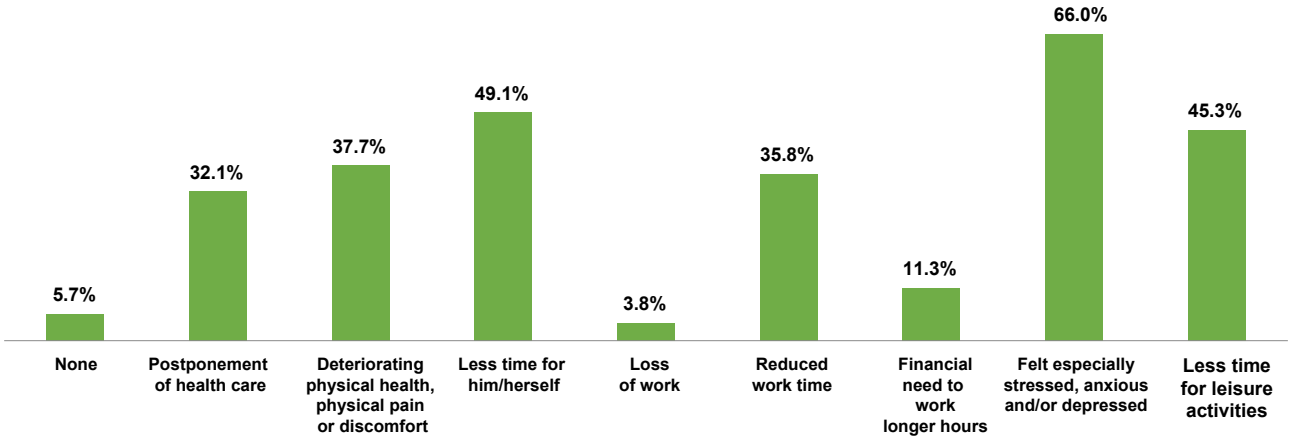

Figure 10. Main changes experienced by the caregivers surveyed - since they have been caring for the person with ALS - (%). Multiple choice questions.
Source: Prepared by the authors based on survey data.

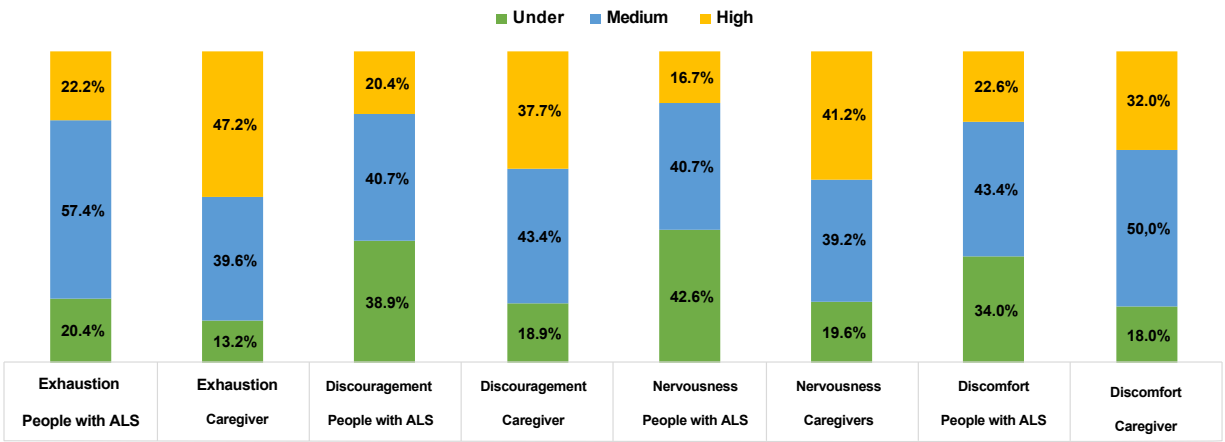

Figure 11. Level of exhaustion, discouragement, nervousness, and discomfort of people with ALS and the surveyed caregivers at the time of the survey -2023- (%)
Source: Prepared by the authors based on survey data.

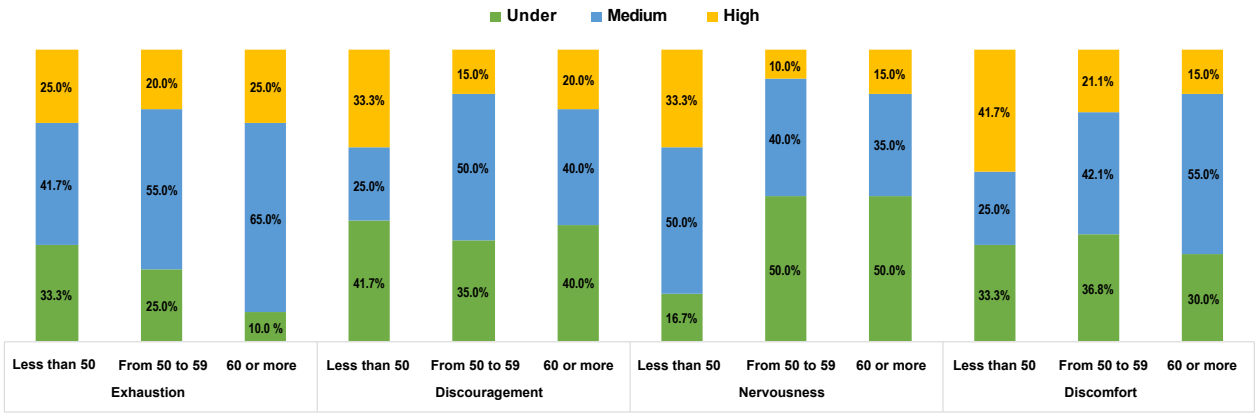

Figure 12. Level of exhaustion, discouragement, nervousness, and malaise of people with ALS by age group (%))
Source: Prepared by the authors based on survey data.

Gender differences are not as pronounced, and no clear pattern can be distinguished. For example, a higher proportion of men with ALS report a high level of discomfort. However, the opposite trend is seen in relation to nervousness: Here, women report a high level in greater proportion (Figure 13).

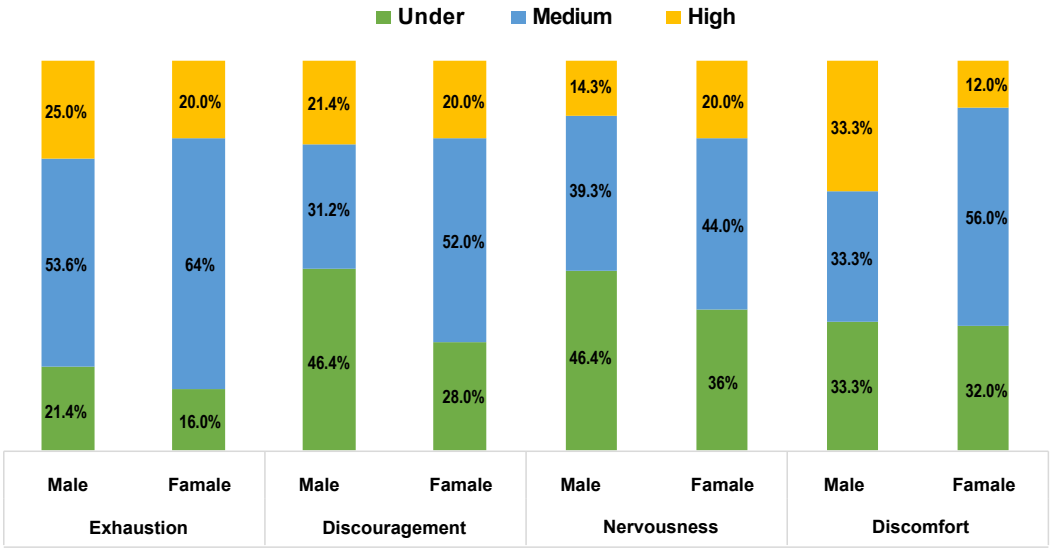

Figure 13. Level of exhaustion, discouragement, nervousness, and discomfort of people with ALS by gender (%)
Source: Prepared by the authors based on survey data.

It is also observed that those with greater physical limitations show a higher proportion of high levels of exhaustion, discouragement, nervousness, and discomfort. Physical limitations are explored from a survey question in relation to the degree to which their physical health has made it difficult for them to eat (including cutting food and others) in the last four weeks. In this sense, for example, it is observed that 30.3% (10 cases) of the people surveyed for whom physical health has made it very or quite difficult to eat have a high level of discouragement, while this level of discouragement is distinguished only in 4.8% (1 case) of the people surveyed who indicated that they had had little or no difficulty (Figure 14).

Finally, in relation to social support, the data suggest that people with ALS who have less support show a higher proportion of high levels of exhaustion, discouragement, nervousness, and discomfort. Social support is explored from a survey question in relation to the degree to which the person has felt supported and/or supported by their friends and close people from their first symptoms until the present. It is observed that 45.5% (5 cases) of the people surveyed who feel little or not at all supported and/or supported have a high level of discomfort, while said level of discomfort is distinguished in

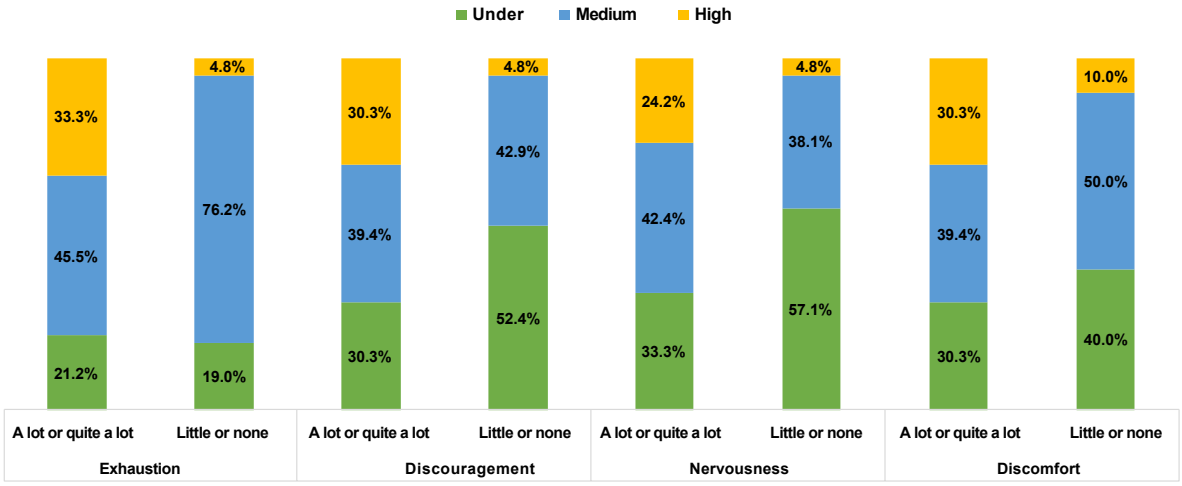

Figure 14. Level of exhaustion, discouragement, nervousness, and discomfort of people with ALS according to the degree to which their physical health has made it difficult for them to eat in the last four weeks (%)
Source: Prepared by the authors based on survey data.

16.7% (7 cases) of the people surveyed who indicate that they feel very or quite supported. A similar trend is distinguished in relation to the levels of exhaustion, discouragement, and nervousness (Figure 15).

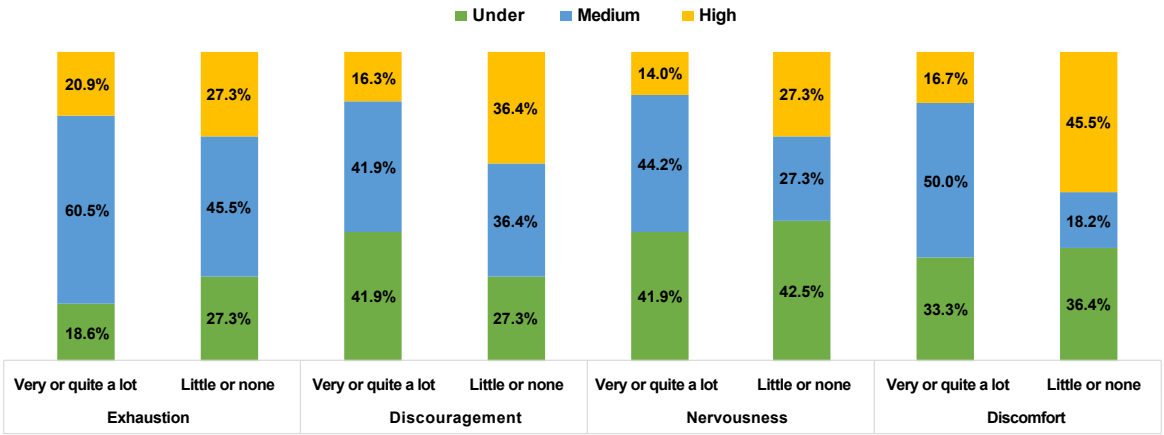

Figure 15. Level of exhaustion, discouragement, nervousness, and discomfort of people with ALS according to whether the person has felt supported and/or supported by their friends and close people (%)
Source: Prepared by the authors based on survey data.

There are, on the other hand, various studies that focus on the differences in the psychological and emotional state and quality of life of caregivers depending on the degree of progression of the disease and/or physical disability of the person with ALS. As mentioned above in the background review, the conclusions drawn from these investigations are controversial (Cipolletta y Amicucci, 2015). The results of the present study show that caregivers of people with ALS who have greater physical limitations show a higher proportion of high levels of exhaustion, discouragement, nervousness, and discomfort in relation to caregivers of people with fewer limitations. In this sense, for example, it is observed that 57.1% (12 people) of caregivers of people with ALS for whom physical health has made eating very or quite difficult have a high level of exhaustion. In comparison, this high level of exhaustion is distinguished in 15.4% (2) of caregivers of people who report having had little or no difficulty. Although these findings should be taken with caution considering the limited number of cases, it is suggested that the caregiver's condition and the patient's disability are associated (Figure 16).

In summary, the results of the survey show the different impacts that both living with ALS and assuming the role of primary caregiver of a person with this disease have on the daily life, health, household economic situation, and emotional well-being of those involved in this dyad.

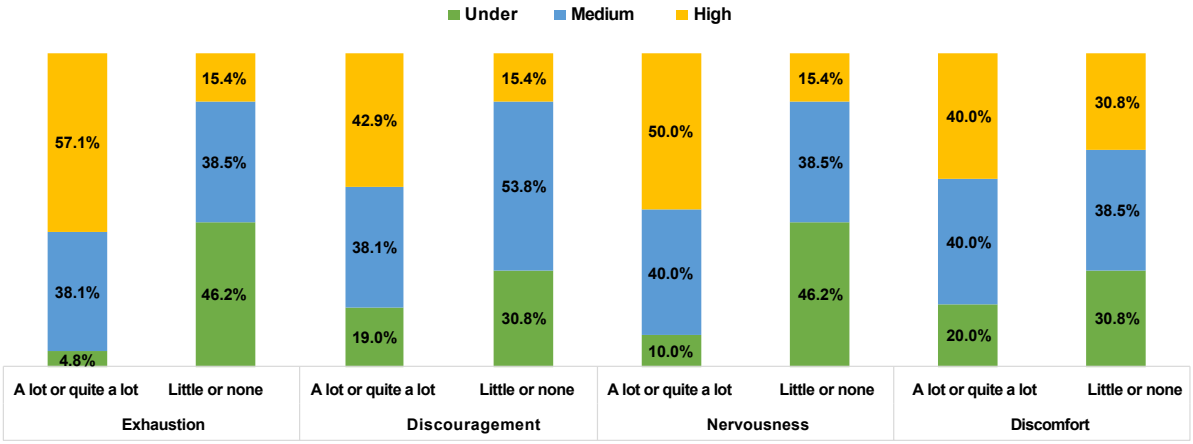

Figure 16. : Level of exhaustion, discouragement, nervousness, and discomfort of the caregiver according to the degree to which the physical health of the person with ALS has made eating difficult in the last four weeks (%)
Source: Prepared by the authors based on survey data**.**

5. Conclusiones

This study contributed, through the production and analysis of quantitative data, to deepen the body of knowledge available on the living conditions of people diagnosed with ALS and those who assume the role of primary caregivers in Argentina in 2023. The results of the survey show the pressing situation that this population is going through both economically and occupationally, as well as in terms of health and emotions as a consequence of the disease. They are in line with the findings of previous studies. It is important to highlight the relevance of the present work, taking into account the existing gaps identified in the background review: there are no studies of this type in Argentina, and even worldwide, there are few investigations that jointly and simultaneously study people with ALS and their caregivers - as is done in the present article.

Even being an exploratory study, the results obtained allow outlining a set of recommendations or considerations that, being evidence-based, can be taken up by different actors and institutions -mainly state agencies and civil society organizations- for the informed design/(re)elaboration of policies and actions aimed at improving the living conditions of people living with this disease, as well as their family and caregiving environment.

In the first place, there is a need to reduce the time required to reach an accurate diagnosis. Doing so could contribute to reducing the levels of anguish and anxiety involved in this process, as well as accelerate the initiation of the palliative care measures required to improve the quality of life of patients and their caregivers. In this regard, it would be beneficial to carry out dissemination and training actions aimed at the early identification of symptoms compatible with ALS, aimed both at the general public and, fundamentally, at health teams -including primary care providers-. Complementarily, it would be desirable that they aim to increase the population's knowledge of the specialized institutions/organizations to which it is possible to turn to obtain reliable information on this disease. In dialogue with the preceding point, there is room for improvement in communication between health personnel and people who receive this diagnosis, fundamentally in terms of providing and obtaining sufficient information on the disease to understand its implications and scope. In line with this, a possible policy line could be aimed at strengthening verbal, nonverbal, and paraverbal communication strategies between medical professionals and patients in order to help guarantee the right of the latter to receive information on the diagnosis, prognosis, and treatment related to their State of health. Third, there is a need to provide financial support to the households of people with ALS in order to ensure that they have sufficient economic resources to cover the costs of the disease and to cope with the impact that this has on the personal and/or family economy due, for example, to the reduction or loss of income from work. As evidenced by this study, and in line with previous research, ALS is a disease with catastrophic expenses that, in turn, usually has a negative impact on the employability of those who suffer from it and their main caregivers, resulting in a dramatic reduction in income that needs to be addressed by the State. Fourthly, in view of the heavy care workloads borne by primary caregivers and the resulting physical and emotional impacts, it is suggested that measures be promoted to facilitate access to and affordable hiring of care professionals specialized in accompanying people

with ALS in order to relieve the family and emotional environment of people with this disease. To this end, the creation of a federal public registry of caregivers specialized in the care of persons with ALS -as well as of other neurodegenerative diseases- could be beneficial for the search and hiring of specialized personnel, facilitating this task. Finally, transversally to the recommendations presented above, it is recommended to adopt a gender-sensitive perspective in all policies and actions designed and implemented focused on this problem in order to avoid -even in unintentional ways- the volume of care work that falls mostly on women is intensified.

This study offered a first approach to the reality of people with ALS and their caregivers in Argentina and, therefore, constitutes an invitation to go deeper into it. It is possible to identify possible lines of research through which to continue investigating this issue. In the first place, the need for a national census of people diagnosed with ALS is highlighted in order to know the universe and its main characteristics. It would be useful both for research in the field of social sciences and in the field of health. Likewise, each of the problems identified in this research could be deepened by carrying out works that focus on each of them. Thus, for example, it would be pertinent to investigate the strategies that people with ALS and their relatives deploy to obtain the material and economic resources needed to cope with the disease and to map the actors involved in them. Similarly, the itinerary that people with ALS must go through until they reach the correct diagnosis could be studied in order to have an approach to the causes that delay this process, among many other possible topics to be explored from a qualitative-quantitative approach that allows to reach a comprehensive understanding of the realities of people with ALS and their caregivers.

Notes

1. It is important to consider that the findings should be taken with caution, due to the limited number of cases, especially with regard to certain groups, as a result of crossing variables.

2. It should be noted that, for these analyses, the sample consists of only 34 caregivers since these are the cases that can be considered a dyad - that is, that allow linking the information collected since the survey has been answered by both the person with ALS and his/her respective primary caregiver - and that, at the same time, do not have missing values in these variables.

Conflict of interest

The authors declare no conflict of interest.

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