The diagnostic, therapeutic and assistance pathway for amyotrophic lateral sclerosis in a north-eastern Italian region: satisfaction of patients and their caregivers

Francesca Palese[1] | Gian Luigi Gigli[2,3] | Paolo Manganotti[4] | Paolo Passadore[5] | Michele Rana[6] | Lorenzo Verriello[7]

[1]Medical Area Department, University of Udine, Udine, Italy

[2]Clinical Neurology Unit, Department of Neurosciences, Santa Maria della Misericordia University Hospital, ASUFC, Udine, Italy

[3]Department of Mathematics, Informatics and Physics, University of Udine, Udine, Italy

[4]Clinical Neurology Unit, Department of Medical, Surgical, and Health Sciences, University Hospital of Trieste, ASUGI, Trieste, Italy

[5]Neurology Unit, Santa Maria degli Angeli Hospital, ASFO, Pordenone, Italy

[6]Neurology Unit, Hospital of Gorizia, ASUGI, Gorizia, Italy

[7]Neurology Unit, Department of Neurosciences, Santa Maria della Misericordia University Hospital, ASUFC, Udine, Italy

Correspondence
Francesca Palese, Institute of Hygiene and Clinical Epidemiology, Medical Area Department, Via Colugna, 50, 33100 Udine, Italy.
Email: francesca.palese@uniud.it

Abstract

In order to evaluate the users' satisfaction degree for the diagnostic, therapeutic and assistance services for amyotrophic lateral sclerosis (ALS) in the Italian region Friuli-Venezia Giulia (FVG), a self-compiled anonymous multiple-choice questionnaire was administered to ALS patients and their caregivers. The questionnaire explored 41 different issues covering the following areas: (a) access to diagnostic pathway and communication among patients, families and health professionals; (b) quality of disease monitoring and effectiveness of interventions aimed at mitigating ALS symptoms; (c) easiness of access to assistive devices (e.g. wheelchair, ankle-foot-orthosis) and home assistance; (d) patient' choices sharing and health professionals empathy. The same issues were proposed both to patients and carers, appropriately adapting the questions, during the period between June and December 2019. The answers were categorised according to criticality level. Median with interquartile range of the numeric variables and percentages of the categorical variables and of the answers to questions were calculated. The mean percentage of satisfied users was 72.8%, considering all the areas. Pain treatment and easiness of access to ambulance transport were the most positive aspects (95.7% and 92.5% of satisfied respondents, respectively), while information about possible enrolment in clinical trials and about possible registration to the regional ALS association were the most critical issues (30.9% and 43.4% of satisfied users). Although the satisfaction level of ALS patients and their caregivers for the services provided resulted generally good, there were some areas that have to be improved. For this purpose, enhancement of multidisciplinary collaboration, sharing of points of view from users and different practitioners and rising awareness among healthcare professionals through clinical audits could be useful. Further research is needed to identify a wider range of users' unexplored unmet needs.

KEYWORDS

amyotrophic lateral sclerosis, caregivers, healthcare, patients, satisfaction, services, survey

What is known about this topic and what this paper adds?

- Health and social workers have low probabilities of coming in contact with patients affected by amyotrophic lateral sclerosis (ALS), a rare disease, and therefore, it is very difficult for them to have proper knowledge about their needs.

- As ALS patients' and their caregivers' benefit from a wide range of health and assistance services, it is important to evaluate their satisfaction degree with the services offered.
- We found that the mean percentage of satisfied users was 72.8%, although some areas have to be improved.
- Information about possible enrolment in clinical trials and about possible registration to the local ALS association appeared as critical areas.

1 | INTRODUCTION

Amyotrophic lateral sclerosis (ALS) represents the most common form of motor neuron disease in adulthood. It is a degenerative disease whose incidence rates in Europe vary from 1.92 to 2.35 cases per 100,000 person-years (Marin et al., 2017). Due to its rarity, health and social workers have low probabilities of coming in contact with this kind of patients. Indicatively, it is estimated that a general practitioner is likely to encounter only 1–2 cases of ALS in his or her working life (Levvy, 2000). Therefore, it is very difficult for health and social professionals to have proper knowledge about the needs of this particular group of patients, differently to other ones, suffering from more common diseases (Hughes et al., 2005; van Teijlingen et al., 2001).

The progression of this illness results in a long clinical course of disease characterised by progressive degrees of disability. This implies that ALS patients will need assistance with daily living activities, mobility, medical and nursing care (Krivickas et al., 1997). Consequently, people living with ALS (that includes not only patients but also their carers, who endure the bulky burden of the daily management of their affected loved ones) come in contact with a wide range of health and assistance services.

In the Italian region Friuli-Venezia Giulia (FVG) a consistent part of health and assistance services are provided by the National Health System, which is a public institution and offers universal and free of charge healthcare assistance to all citizens. Moreover, in the last years, there has been made an effort to create a homogenous and standardised level of care for ALS patients, organised on the basis of evidence based medicine. In particular, a team of experts (comprising neurologists, pneumologists and physiotherapists) dedicated to ALS multidisciplinary management was created.

In order to test the degree of satisfaction with the diagnostic, therapeutic and assistance services offered by FVG to ALS patients and their families, a survey was carried on.

2 | MATERIALS AND METHODS

2.1 | Setting and study population

The survey was conducted in FVG, a north-eastern Italian region of about 1,200,000 inhabitants and an estimated prevalence of ALS of 8.0 cases per 100,000 inhabitants (Palese et al., 2018). During the period between June and December 2019, a self-compiled anonymous questionnaire was administered to ALS patients and their caregivers. Patients were eligible to the survey if ALS diagnosis was established at least 6 months before and if they were resident in FVG. The list of potential candidates to survey was compiled considering the patients periodically examined by the neurologists of FVG hospitals and the ones registered at the regional section of the non-profit Italian ALS Association (AISLA). The ethical approval was granted by the FVG Ethical Committee.

2.2 | Data collection

White and grey literature (Muraro & Coccia, 2015; van Teijlingen et al., 2001) and the local organisation peculiarities were considered in order to design the appropriate questions comprising the wide range of health and assistance services offered to manage ALS cases. The questionnaire was composed of 41 multiple choice questions addressing four main thematic areas: (a) access to diagnostic pathway and communication among patients, families and health professionals; (b) quality of disease monitoring and effectiveness of interventions aimed at mitigating ALS symptoms; (c) easiness of access to assistive devices (e.g. wheelchair, ankle-foot-orthosis) and home assistance; (d) patient' choices sharing and health professionals empathy. The same issues were proposed both to patients and carers, appropriately adapting the questions. Basic demographic and illness duration data were collected, too.

The questionnaires were administered in the hospital by the reference neurologist (in the event of a periodic examination) or at home by the AISLA volunteers (in case of particularly complicated patients, difficult to transport). The patients and caregivers compiled the questionnaires autonomously or, if necessary, with the help of a carer. The questionnaire completing occurred at the moment of the administration or later at home. If the latter, the respondents returned the completed form in occasion of the following neurological examination or visit by AISLA volunteers.

2.3 | Statistical analysis

To facilitate the interpretation, the answers were categorised according to the criticality level (Muraro & Coccia, 2015):

Example 1: according to you, are the suggestions you receive about how to face daily life difficulties useful?

- Yes, always.
- Almost always.
- Only sometimes.

§ Almost never.
§ Never.
* I have never needed them.
* Not answered question / Not pertinent question.

Example 2: have you ever have been briefed on the clinical trials you could be enrolled in?

◦ Yes.
§ No.
◦ No criticalities.
§ Presence of criticalities.
* Data not usable for analyses.

Median with interquartile range (IQR) of the numeric variables and percentages of the categorical variables and of the responses to the questions were calculated. Results presented in the following section account only for answers that were given, as some questions were left without any answer. We explored a possible relation between basic demographic and clinical characteristics (patients vs. caregivers, age, sex, educational level, duration of disease) and lack of answer to questions by means of chi-square test and Wilcoxon–Mann–Whitney test.

Data were analysed using SAS© software version 9.4 (SAS).

3 | FINDINGS

3.1 | Base demographic characteristics and ALS duration

Of about 70 patients and the same number of caregivers who were expected to be potentially engaged on the basis of a pilot evaluation, 42 patients and 41 caregivers were effectively enrolled in the study. Their characteristics are summarised in Table 1. It was possible to administer the questionnaire only to the patients who were regularly visited in one of the four regional Neurology departments during the study period and/or were members of the AISLA. No one of them refused to participate in the survey.

The remaining estimated 30 patients resulted impossible to contact for three reasons:

1. they were in a later stage of disease. As a consequence, they did not undergo neurologic examinations as they were treated exclusively in a home-care context;

   or

2. they were regularly visited in a Neurologic department situated out of the FVG region.

   and

3. they were not registered to the AISLA association, so the AISLA volunteers could not administer the questionnaire.

TABLE 1 Characteristics of the respondents

Characteristics	Patients (*N*, %)	Caregivers (*N*, %)
Total	42 (50.6)	41 (49.4)
Sex		
Male	23 (54.8)	14 (34.2)
Female	18 (42.9)	21 (51.2)
No response	1 (2.4)	6 (14.6)
Age (years)		
Median (IQR)	65.5 (55.8–73.8)	57.5 (51.8–65.3)
No response	2 (4.8)	5 (12.2)
Employment		
Occupied	2 (4.8)	17 (41.5)
Unoccupied or pensioner	37 (88.1)	16 (39.0)
Student	1 (2.4)	0 (0.0)
No response	2 (4.8)	8 (19.5)
Educational level		
Elementary school diploma	7 (16.7)	4 (9.8)
Middle license	13 (31.0)	8 (19.5)
High school diploma	18 (42.9)	16 (39.0)
Degree	2 (4.8)	3 (7.3)
No response	2 (4.8)	10 (24.4)
Duration of illness (years)		
Median (IQR)	4.0 (2.0–6.0)	4.0 (2.0–6.0)
No response	2 (4.8)	7 (19.4)

3.2 | General considerations

The number of persons who did not answer the base demographic or clinical questions ranged from 5 to 11, while it ranged from 0 to 8 considering the questions related to satisfaction level. Therefore, a percentage of answers to each question ranging from 0% to 9.6% remained unanswered.

We explored possible relations between basic demographic and clinical characteristics and lack of answer to questions, but we found no statistically significant results. We cannot exclude that this could be related to the limited number of the sample, considering also that 14 interviewees (4 patients and 10 caregivers) did not give an answer to one or more questions about basic demographic and clinical characteristics.

Globally, both ALS patients and their caregivers expressed a good level of satisfaction for the services provided: the mean percentage of the users that considered the issues as not critical was 72.8%, calculating the mean value for all the areas. The level of agreement among patients and carers varied, differing the percentage of satisfied users from 0.07% to 18.3% (mean value: 3.8%) according to the different questions.

Patients showed a lower satisfaction level than their caregivers for 10 items out of 41, whereas they expressed a higher satisfaction level

for the remaining 31 items. Anyway, the differences between the two groups were lower than 10 percentage points in most cases. We found a higher difference for only 6 items (see Figures 1–4). Note that, except for one item ('time appropriateness of assistive devices provision'), patients always expressed more satisfaction than their carers.

3.3 | Access to diagnostic pathway

The survey revealed that more than 75% of the respondents had no difficulty in accessing a first neurologic examination (81.0% of patients and 75.7% of caregivers) or diagnostic instrumental examinations (90.5% and 80.6%, respectively; see Figure 1). On the converse, of the respective 20.5% and 14.0% of respondents who found it difficult to book either type of examinations, two of them (2.7%, one patient and one caregiver) declared that the wait time needed in a public healthcare context was so long that they had to refer to private healthcare.

With regards to follow-up examinations, a quarter of patients experienced difficulties in neurologic and physiatric follow-up appointment booking, while <10% in the case of pneumological check-up.

3.4 | Communication and information flow between patient and neurologist

Respondents were asked if they were satisfied by the way the neurologist communicated ALS diagnosis and if he/she fulfilled their information needs about the illness and its course. Responses were positive for more than 78% of the interviewees (77.5% and 78.1% of patients and 82.4% and 78.4% of their carers, respectively) who gave an answer, as displayed in Figure 1. Differently, only a minority of respondents were given information about possible enrolment in clinical trials (31.7% of patients and 30.0% of their carers) and possible registration to AISLA association (41.5% and 45.2%, respectively). The opportunity of obtaining psychological support was envisaged for 69.1% of patients and 60.0% of their carers.

With regards to the understandability of explanations the neurologist gave to patients and caregivers about their ALS-related queries, even 90.2% of respondents (95.2% of patients and 85.0% of caregivers) declared themselves satisfied. In contrast, only two-thirds of the interviewees (69.1% and 69.3%, respectively) refer that the treating neurologist informed them about the risks and benefits of the treatments (drugs, ambulatory and surgical interventions) proposed.

3.5 | Exemption certification and drugs supply

The neurologists appeared available to provide ALS exemption certification (that allows to obtain health services related to illness for free; 87.5% of patients and 94.4% of their carers) and hospital pharmacy delivered the drugs needed for the first cycle of therapy in the large majority of cases (78.1% and 78.4%, respectively).

3.6 | Effectiveness of interventions against anxiety and depression, pain and stiffness

The survey revealed that less than one-third and one-quarter of patients, respectively, suffered from anxiety or depression and pain, which brought them to get medical help. On the contrary, about 70% of patients suffered from severe muscle stiffness.

While all the cases of pain were treated, about 20% of patients did not receive medical treatment for anxiety or depression and muscular stiffness (see Figure 2).

Respondents were asked to assess the degree of treatment effectiveness using a score ranging from 0 (no benefit) to 10 (maximum benefit). Median value of benefit obtained for these non-motor symptoms was evaluated at 6.5 (IQR: 2.5–8.8) for depression (patients: 6.5, IQR: 6.3; caregivers: 5.0, IQR: 3.0), 5.0 (IQR: 5.0–7.0) for pain (patients: 5.0, IQR: 2.0; caregivers: 6.0, IQR: 2.0) and 6.0 (IQR: 4.5–8.0) for stiffness (patients: 6.0, IQR: 3.5; caregivers: 6.0, IQR: 3.0).

3.7 | Information flow among healthcare providers

One-third of the respondents (31.0% of patients and 43.9% of caregivers) reported that they received different opinions from different healthcare providers about the same issue and one-quarter of the interviewees (16.7% and 35.0%, respectively) noticed errors during information flow among healthcare professionals.

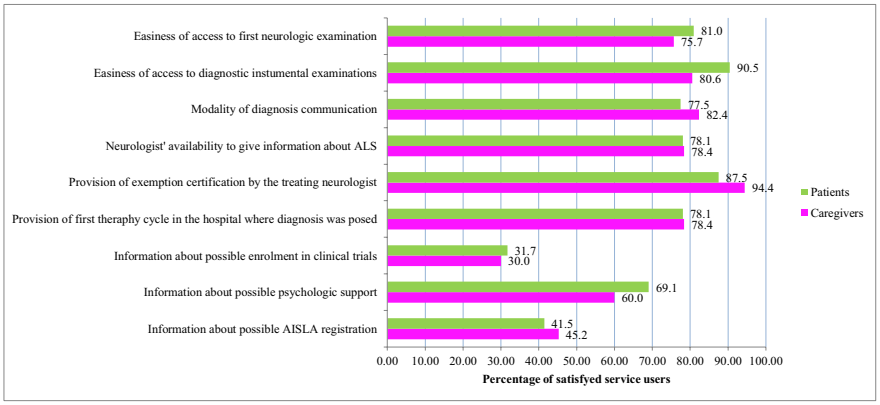

FIGURE 1 Satisfaction degree for access to diagnostic pathway and for communication among patients, families and health professionals [Colour figure can be viewed at wileyonlinelibrary.com]

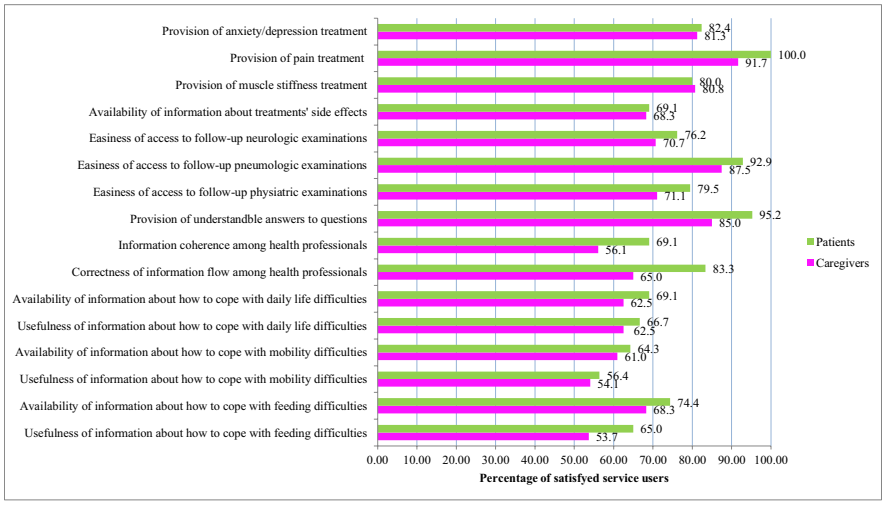

FIGURE 2 Satisfaction degree for disease monitoring and interventions effectiveness [Colour figure can be viewed at wileyonlinelibrary.com]

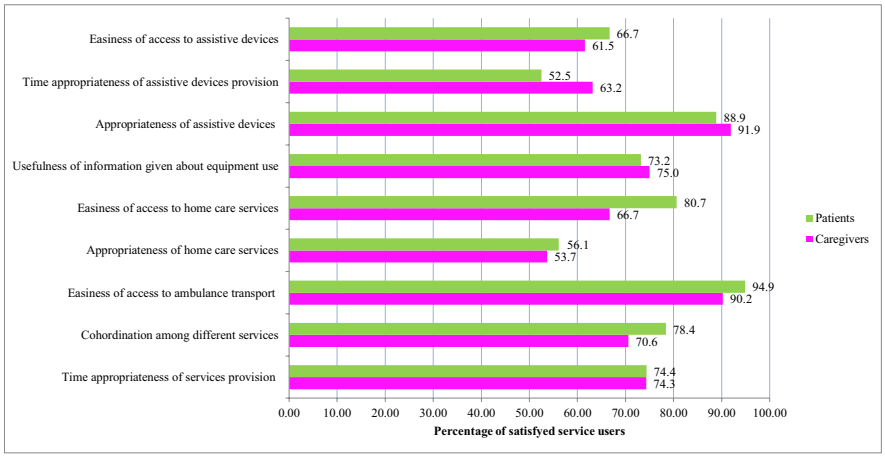

FIGURE 3 Satisfaction degree for easiness of access to assistive devices and home care services [Colour figure can be viewed at wileyonlinelibrary.com]

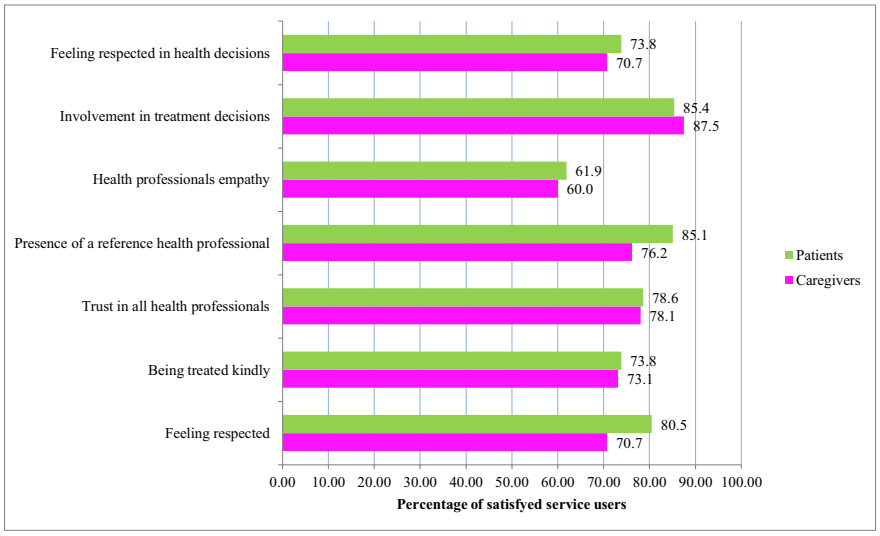

FIGURE 4 Satisfaction degree for patient choices sharing and health professionals empathy [Colour figure can be viewed at wileyonlinelibrary.com]

3.8 | Usefulness of information about daily life strategies

A percentage of respondents ranging from 55.3% (56.4% of patients and 54.1% of caregivers) to 71.3% (74.4% and 68.3%, respectively) appreciated the suggestions they received about how to face difficulties in everyday life, in movement and in feeding.

3.9 | Access to assistive equipment and home assistance

Even if 40% of respondents considered as critical the access to assistive devices (33.3% of patients and 38.5% of caregivers) and their provision time appropriateness (47.5% and 36.8%, respectively), more than 90% of them considered the granted supportive devices as suited for their needs (88.9% and 91.9%) and 74% (73.2% and 75.0%) of them appreciated the information they received about their correct use (Figure 3).

Even if only one-quarter of the people answered they experienced difficulties in obtaining home care services (19.4% and 33.3%), one-half of the users (43.9% and 46.3%) considered them as unappropriated for their needs.

About 75% of the respondents (78.4% and 70.6%) considered the devices and home assistance services they used as well-coordinated and 92% of them (94.9% and 90.2%) did not face difficulties in obtaining ambulance transport.

3.10 | Patients' choices sharing and empathy

About 75% of patients and caregivers expressed satisfaction for issues regarding the relational aspects experienced with health professionals, ranging from 61% of respondents (61.9% of patients and 60.0% of their carers) that perceived empathy from healthcare providers to 86.4% (85.4% and 87.5%, respectively) that felt involved in decisions related to their health problems (as showed in Figure 4).

4 | DISCUSSION

The present study recruited a region-based sample of 83 persons and investigated a wide range of topics, differently from other studies, that deepened more specific themes in smaller samples (Brown, 2003; Hocking et al., 2006; Hughes et al., 2005; McCabe et al., 2008; O'Brien et al., 2011; Silverstein et al., 1991). Our data were collected in a quantitative way, while the majority of the available literature is derived from interviews and has a descriptive nature (Bolmsjö & Hermerén, 2001; Brown, 2003; Foley et al., 2012; Hocking et al., 2006; Hughes et al., 2005; McCabe et al., 2008).

Our findings revealed that the satisfaction degree of ALS patients and their caregivers for the health and assistance services provided in FVG was generally good, even if some critical areas are still perceived as not satisfying by patients and/or caregivers.

We decided to enrol both patients and caregivers because of the fundamental role in ALS management of the latter. As a matter of fact, carers are almost always in charge of quite all the management of ALS patients, even in the last and severe stages, while healthcare professionals' interventions are usually limited in time (Robinson & Hunter, 1988). Despite this, the needs of ALS carers are often underestimated by health and social care professionals (Levvy, 2000).

4.1 | Access to diagnostic pathway and communication

The moment of diagnosis communication represents a fundamental step for building the following relationship between patients and their families and physician. Most of our interviewees believed that diagnosis was communicated by the neurologist leisurely and spending all the time needed, in line with a survey and an audit carried out at an Italian and an English ALS specialised centres (Callagher et al., 2009; Chiò et al., 2008), which found that even though service users experienced negative feelings about their diagnosis, most of them were satisfied with the way physicians communicated the bad news (Foley et al., 2012).

People newly diagnosed with ALS and their carers often require practical information about the illness (Hughes et al., 2005) and receiving the proper information, especially in the first phase of the illness, appears as particularly important. This is because patients and families obtaining incomplete and inappropriate information from healthcare providers will probably find (through internet, books, leaflets or media) autonomously abundant information, not always correct and suitable for the specific patient, thus generating possible confusion. With regards to this issue, it appeared substantially uncritical in our respondents' opinion, in contrast to other studies, revealing that users expected more information about specific aspects of the disease than that which was provided to them (Hughes et al., 2005; Silverstein et al., 1991; Wicks & Frost, 2008).

The important role played by patients' associations in supporting ALS patients and their families were highlighted in a New Zealander study (Hocking et al., 2006), where the Australian ALS Association resulted as particularly appreciated for its responsiveness and capacity to accompany during the illness journey. Nevertheless, only less than half of respondents were informed by the treating neurologist of the possibility to register to AISLA, which offers relational, psychological as well as material (equipment and transport) support. A possible explanation may be that physicians could neglect the importance of associations, maybe because they are more focused on merely medical and treating aspects.

Only a minority of users declared they were notified about the possible enrolment in clinical trials. This is a particularly critical aspect, as this delayed or even missing information could reflect in a loss of opportunities to benefit from potentially effective new drugs.

As a fact, therapeutic trials usually recruit patients with relatively mild disability (Chiò, 1999) and, therefore, in an early stage of the disease.

4.2 | Disease monitoring and interventions effectiveness

Given the widespread idea of ALS as a merely motor disease, non-motor symptoms as pain, stiffness and depression could be overlooked by physicians. As a matter of fact, a recent study (Nicholson et al., 2018) reported that 84% of ALS patients suffer from muscle stiffness, 59% from pain and 52% from depression, but only 40%–50% of these disorders obtains medical treatment, even if it is known that these problems heavily affect the patients' and, consequently, caregivers' quality of life. Furthermore, there is evidence that pain could have a prognostic significance, too (Chiò et al., 2017). In contrast to prior findings, our sample showed a larger sensitivity of the physicians about this theme, as the majority of patients received medical treatment for stiffness, pain and anxiety or depression. In this regard, it is important the active evaluation of possible extra-motor symptoms (especially depression) in occasion of follow-up examinations as, according to our experience, ALS patients often do not report them spontaneously.

In our study, the query 'correctness of information flow among health professionals' showed the most important difference (18.3 percentage points) in satisfaction between patients and caregivers, being the latter less satisfied. In our experience, caregivers, more than patients, often complain of problems regarding the information flow between hospital and territorial welfare service health professionals. This sometimes brings difficulties in home care, supply of aids and initiation of practices for the recognition of disability, leading to a greater care burden and time waste for carers.

For the item 'usefulness of information about how to cope with feeding difficulties' the percentage score difference was 11.3, again resulting in the caregivers less satisfied than the patients. This difference could be explained by the fact that ALS patients with dysphagia often tend to underestimate the problem, sometimes up to the advanced stages of the disease, because they do not want to use food thickeners or due to fear of being subjected to PEG. Conversely, caregivers often show greater concern, even when dysphagia is not yet severe, because they have fear their dears could suffocate.

As regards the questions 'information coherence among health professionals' and 'provision of understandable answers to questions', a difference of 13.0% and 10.2% points was found, between patients and caregivers, respectively. The method of communicating ALS diagnosis, its evolution and the necessary treatments take into account the patient's level of education, his/her specific requests at a given time, the willingness or not to address certain aspects related to disease progression (such as, e.g. PEG placement and invasive ventilation via tracheostomy), as well as family background. In the early stages of the disease, when the patients have not yet had time to elaborate the diagnosis, caregivers sometimes wish to obtain detailed information on the progression of the disease, as well as on the duration. In this context, the communication of the various problems related to the progression of ALS and the possible treatments requires a certain graduality and is not always homogeneous among the various professionals involved. Therefore, the answers could be, especially in caregivers' opinion, not clearly explanatory or not completely coherent among the different professionals.

4.3 | Access to assistive equipment and home assistance

As patient's conditions worsen, home care becomes more challenging. In Italy, it is delegated almost entirely to family members, with only some support from public services as regards financing and social professionals' assistance (Cipolletta et al., 2018). The physical and psychological burden of caregivers of ALS patients is overwhelming, and previous literature has shown an increased incidence of depression and psychophysical health deficits in caregivers of patients affected by other degenerative diseases (Kalb, 1995; Olin et al., 1996). In addition, Evans and colleagues (Evans et al., 1991) demonstrated that patients cared for by depressed caregivers receive suboptimal home care. In order to reduce these effects, the primary caregiver burden should be eased as much as possible (Krivickas et al., 1997), for example ensuring periods of relief or enhancing home care services. Given the findings of our survey, the FVG region appears to provide substantially adequate home care services in terms of easiness of access, timing and coordination, even if appropriateness appeared as a critical aspect. A possible explanation for this last point may be the unfamiliarity with ALS of care staff, given the disease rarity.

The treating neurologist may play a fundamental role in correctly informing patients and carers about the home services offered for their needs and who to contact to request them.

A consistent difference in the percentage of satisfaction was found between the two groups of respondents for the item 'easiness of access to home care services'. This could be related to the fact that in case of moderate-severe disability, the patient tends to rely mainly on its caregiver. Therefore, the difficulty in accessing home care has a much greater weight for caregivers than for patients.

As regards the item 'time appropriateness of assistive devices provision', a difference of 10.7% points was found, being, conversely, the patients less satisfied than their carers. This result could be related to the fact that devices (e.g. wheelchairs) allow patients to preserve a certain degree of autonomy. Therefore, patients could be more sensitive than their caregivers to possible delays in devices provision, which may affect greatly their daily life autonomy.

4.4 | Sharing of patients' choices and empathy

With regards to this field, the level of criticality varied greatly according to the specific aspect investigated. Perceived empathy from

healthcare providers appeared as the most problematic aspect, being considered as critical for 39.0% of users. These findings are consistent with an English study (Brown, 2003), that underlined the feeling of inadequate emotional support and a detached professional approach, pointed out by both patients and carers. In this regard, healthcare providers recognised the importance of trying to emotionally adapt to every single patient in order to create a constructive relationship and identified time as a limiting factor. On the contrary, Ray and colleagues (Ray et al., 1986) found that some physicians treating breast cancer patients deliberately avoided forming relationships as they thought that this could invalidate their objectivity and judgement.

The large majority of our sample felt satisfied with their involvement in decisions related to health problems, similarly to an American study (Silverstein et al., 1991), which revealed that 70% of patients wanted to participate in decisions about medical care, whereas the remaining 30% preferred to leave decisions to the physician.

4.5 | Weaknesses of the study

There are several limitations to this study. First, not all respondents completed every question of the survey, maybe due to the relatively high number of questions or, as regards basic demographic data, the fear of being identified. It is also possible that more satisfied or dissatisfied patients and caregivers had increased motivation to express their opinions by completing the survey. We did not find a statistically significant relation between basic demographic and illness duration and lack of answer to questions, but this could be related to the limited number of the sample.

Moreover, a possible bias could be related to the fact that some potentially eligible patients were regularly visited in a Neurologic department situated outside of the FVG region. In some cases, the choice of an extra-regional centre could be the result of a previous bad experience in one of the FVG Neurologic departments. As a consequence, our findings do not take into account of a number of dissatisfied patients and carers, bringing therefore to a possible overestimation of the satisfaction level.

Additionally, self-compiled questionnaire surveys make it difficult to assure that patients or caregivers were responding for themselves, thus affecting the trueness of the answers.

Lastly, this study refers to a relatively small base population, a small geographic area and a local service organisation. Consequently, our findings are probably not generalisable to the whole ALS population.

5 | CONCLUSION

The present study revealed that the satisfaction level of ALS patients and their caregivers for the health and assistance services provided in FVG was generally good, even if some critical areas are still perceived as not satisfying by patients and/or caregivers. This is a particularly significant result, especially considering that they referred to services offered in a public health frame, in contrast to other geographical contexts, where charitable support and private assistance appear as fundamental (Krivickas et al., 1997; van Teijlingen et al., 2001).

There are some areas that need to be improved and, for this purpose, enhancement of multidisciplinary collaboration, sharing of points of view from users and different practitioners and rising awareness among healthcare professionals about ALS users' needs through clinical audits could be useful.

Further research is needed to identify a wider range of users' unexplored unmet needs. Future studies might be conducted with larger samples and by means of interviews or written open-ended questions with the aim of better understanding the expectations and improving the quality of the services provided.

ACKNOWLEDGEMENTS

The authors are grateful to ALS patients, their families and the FVG AISLA volunteers for their precious collaboration.

AUTHOR'S CONTRIBUTION

FP and LV: conception and design. GLG, PM, PP, MR, and LV: acquisition of data. FP: data analysis. FP and LV: data interpretation. FP: writing of original draft and review. GLG, PM, PP, MR, and LV: final approval of the version to be published.

DISCLOSURE STATEMENT

The authors report no conflicts of interest. This study did not receive any financial support.

DATA AVAILABILITY STATEMENT

Data available on request from the authors.

ORCID

Francesca Palese https://orcid.org/0000-0001-7119-4445
Gian Luigi Gigli https://orcid.org/0000-0003-3526-1414
Paolo Manganotti https://orcid.org/0000-0001-5305-6268

REFERENCES

Bolmsjö, I., & Hermerén, G. (2001). Interviews with patients, family, and caregivers in amyotrophic lateral sclerosis: Comparing needs. *Journal of Palliative Care*, *17*, 236–240. https://doi.org/10.1177/082585970101700403

Brown, J. B. (2003). User, carer, and professional experiences of care in motor neurone disease. *Primary Health Care Research & Development*, *4*, 207–217. https://doi.org/10.1191/1463423603pc151oa

Callagher, P., Mitchell, D., Bennett, W., & Addison-Jones, R. (2009). Evaluating a fast-track service for diagnosing MND/ALS against traditional pathways. *British Journal of Neuroscience Nursing*, *5*, 322–325. https://doi.org/10.12968/bjnn.2009.5.7.43241

Chiò, A. (1999). ISIS Survey: An international study on the diagnostic process and its implications in amyotrophic lateral sclerosis. *Journal of Neurology*, *246 Suppl 3*, III1–5. https://doi.org/10.12968/bjnn.2009.5.7.43241

Chiò, A., Montuschi, A., Cammarosano, S., De Mercanti, S., Cavallo, E., Ilardi, A., Ghiglione, P., Mutani, R., & Calvo, A. (2008). ALS patients

and caregivers communication preferences and information seeking behaviour. *European Journal of Neurology*, *15*, 55–60. https://doi.org/10.1111/j.1468-1331.2007.02000.x

Chiò, A., Mora, G., & Lauria, G. (2017). Pain in amyotrophic lateral sclerosis. *The Lancet Neurology*, *16*, 144–157. https://doi.org/10.1016/S1474-4422(16)30358-1

Cipolletta, S., Gammino, G. R., Francescon, P., & Palmieri, A. (2018). Mutual support groups for family caregivers of people with amyotrophic lateral sclerosis in Italy: A pilot study. *Health & Social Care in the Community*, *26*, 556–563. https://doi.org/10.1111/hsc.12558

Evans, R. L., Bishop, D. S., & Haselkorn, J. K. (1991). Factors predicting satisfactory home care after stroke. *Archives of Physical Medicine & Rehabilitation*, *72*, 144–147. https://doi.org/10.5555/uri:pii:000399939190123Z

Foley, G., Timonen, V., & Hardiman, O. (2012). Patients' perceptions of services and preferences for care in amyotrophic lateral sclerosis: A review. *Amyotrophic Lateral Sclerosis*, *13*, 11–24. https://doi.org/10.3109/17482968.2011.607500

Hocking, C., Brott, T., & Paddy, A. (2006). Caring for people with motor neurone disease. *International Journal of Therapy & Rehabilitation*, *13*, 351–355. https://doi.org/10.12968/ijtr.2006.13.8.351

Hughes, R. A., Sinha, A., Higginson, I., Down, K., & Leigh, P. N. (2005). Living with motor neurone disease: Lives, experiences of services and suggestions for change. *Health & Social Care in the Community*, *13*, 64–74. https://doi.org/10.1111/j.1365-2524.2005.00530.x

Kalb, R. C. (1995). *Families affected by multiple sclerosis: Disease impacts and coping strategies*. National Multiple Sclerosis Society.

Krivickas, L. S., Shockley, L., & Mitsumoto, H. (1997). Home care of patients with amyotrophic lateral sclerosis. *Journal of the Neurological Sciences*, *152*, 82–89. https://doi.org/10.1016/S0022-510X(97)00251-7

Levvy, G. (2000). The role of the lay associations. In R. H. Brown, V. Meininger, & M. Swash (Eds.), *Amyotrophic lateral sclerosis*. Martin Dunitz.

Marin, B., Boumédiene, F., Logroscino, G., Couratier, P., Babron, M. C., Leutenegger, A. L., Copetti, M., Preux, P. M., & Beghi, E. (2017). Variation in worldwide incidence of amyotrophic lateral sclerosis: A meta-analysis. *Internal Journal of Epidemiology*, *46*, 57–74. https://doi.org/10.1093/ije/dyw061

McCabe, M. P., Roberts, C., & Firth, L. (2008). Satisfaction with services among people with progressive neurological illnesses and their carers in Australia. *Nursing & Health Sciences*, *10*, 209–215. https://doi.org/10.1111/j.1442-2018.2008.00399.x

Muraro, L., & Coccia, J. (2015). *Progetto di indagine sulla qualità percepita da utenti coinvolti nel percorso diagnostico terapeutico assistenziale del paziente con SLA*. Azienda Unità Sanitaria Locale di Bologna, UO Qualità e Accreditamento. Retrieved from http://www.assisla.it/wp-content/uploads/2016/12/RelazioneQPpdtasla.pdf

Nicholson, K., Murphy, A., McDonnell, E., Shapiro, J., Simpson, E., Glass, J., Mitsumoto, H., Forshew, D., Miller, R., & Atassi, N. (2018). Improving symptom management for people with amyotrophic lateral sclerosis. *Muscle & Nerve*, *57*, 20–24. https://doi.org/10.1002/mus.25712

O'Brien, M. R., Whitehead, B., Jack, B. A., & Mitchell, J. D. (2011). From symptom onset to a diagnosis of amyotrophic lateral sclerosis / motor neurone disease; experiences of people with ALS/MND and family caregivers: A qualitative study. *Amyotrophic Lateral Sclerosis*, *12*, 97–104. https://doi.org/10.3109/17482968.2010.546414

Olin, J. T., Schneider, L. S., & Kaser-Boyd, N. (1996). Associating personality pathology with emotional distress in caregivers of patients with Alzheimer's disease. *Journal of Clinical Geropsychology*, *2*, 93–101.

Palese, F., Sartori, A., Verriello, L., Ros, S., Passadore, P., Manganotti, P., Barbone, F., & Pisa, F. E. (2018). Epidemiology of amyotrophic lateral sclerosis in Friuli-Venezia Giulia, North-Eastern Italy, 2002–2014: A retrospective population-based study. *Amyotrophic Lateral Sclerosis & Frontotemporal Degeneration*, *15*, 1–10. https://doi.org/10.1080/21678421.2018.1511732

Ray, C., Fisher, J., & Wisniewski, T. K. M. (1986). Surgeons' attitudes toward breast cancer, its treatment, and their relationship with patients. *Journal of Psychosocial Oncology*, *4*, 33–43. https://doi.org/10.1300/J077v04n01_03

Robinson, I., & Hunter, M. (1988). Motor neurone disease. In R. Fitzpatrick & S. Newman (Eds.), *The experience of illness series*. Routledge.

Silverstein, M. D., Stocking, C. B., Antel, J. P., Beckwith, J., Roos, R. P., & Siegler, M. (1991). Amyotrophic lateral sclerosis and life-sustaining therapy: Patients' desires for information, participation in decision making, and life-sustaining therapy. *Mayo Clinic Proceedings*, *66*, 906–913. https://doi.org/10.1016/S0025-6196(12)61577-8

van Teijlingen, E. R., Friend, E., & Kamal, A. D. (2001). Service use and needs of people with motor neurone disease and their carers in Scotland. *Health & Social Care in the Community*, *9*, 397–403. https://doi.org/10.1046/j.1365-2524.2001.00320.x

Wicks, P., & Frost, J. (2008). ALS patients request more information about cognitive symptoms. *European Journal of Neurology*, *15*, 497–500. https://doi.org/10.1111/j.1468-1331.2008.02107.x

How to cite this article: Palese F, Gigli GL, Manganotti P, Passadore P, Rana M, Verriello L. The diagnostic, therapeutic and assistance pathway for amyotrophic lateral sclerosis in a north-eastern Italian region: Satisfaction of patients and their caregivers. *Health Soc Care Community*. 2022;30:124–132. https://doi.org/10.1111/hsc.13379