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Complexity of needs in amyotrophic lateral sclerosis (ALS) patients using the ENP-E scale in the north-eastern region of Spain

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Abstract

Objectives. This study aimed to explore the clinical characteristics of amyotrophic lateral sclerosis (ALS) patients in Spain's north-eastern region, their inclusion in chronic care programmes, and their psychosocial and spiritual needs (PSNs).

Methods. A longitudinal descriptive study in adult patients with ALS. We analyzed clinical variables and participation in chronicity and PSNs assessment using the tool Psychosocial and Spiritual Needs Evaluation scale in end-of-life patients (ENP-E scale).

Results. 81 patients (average age 65.6 $\pm$ 11.7) were studied. At the study's outset, 29.7% employed non-invasive ventilation (NIV), increasing to 51.9% by its conclusion. Initial percutaneous endoscopic gastrostomy (PEG) utilization was 14.8%, rising to 35.85%. Chronic care programme participation was as follows: home care (24.7% initially, 50.6% end), palliative care (16% initially, 40.7% end), case management (13.6% initially, 50.6% end), and advance care planning registration (6.2% initially, 35.8% end). At study start, 47.8% of patients ($n = 46$) showed moderate-to-severe complexity in PSNs assessment using the ENP-E scale, without showing differences in age, sex, and time of evolution; whereas, on the evolutionary analysis, it was 75% ($n = 24$). A higher evolutionary complexity was observed in males <60 and >70 years, with no PEG and evolution of ALS of <2 and ≥ 5 years, and not included in chronicity programmes. When assessing concerns, physical pain and family aspects stand out in all measurements. Forty-eight percent of patients at study start and 71% at end of study showed external signs of emotional distress.

Significance of results. Most ALS patients showed a high degree of complexity and were not integrated in chronicity programmes. A "care path" is proposed to integrate ALS patients in these programmes and systematically assess their needs.

Introduction

Amyotrophic lateral sclerosis (ALS) is a neurodegenerative disorder that leads to progressive loss of motor neurons. Although the onset of the disease varies, with some patients experiencing spinal or limb weakness and others showing bulbar involvement, all forms lead to increasing physical dependence (Al-Chalabi and Hardiman 2013; Hardiman et al. 2011). Patients usually succumb to respiratory failure within 3 years of symptom onset (Brown and Al-Chalabi 2016).

Currently, there is no healing treatment, so patients need palliative care and management of their symptoms, which sometimes include the use of percutaneous endoscopic gastrostomy (PEG) and invasive/non-invasive mechanical ventilation systems (IV/NIV) (Bensimon et al. 1994; Hobson and McDermott 2016; Miller et al. 2012).

The incidence in European populations is between 1.8 and 3.3 cases per 100,000 inhabitants/year over 18 years old, being slightly higher in men than in women (Marin et al. 2017).

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Worldwide prevalence is unknown, but in Catalonia (Spain), it is between 5.4 and 7.8 patients/100,000 inhabitants over 18 years old (Castro-Rodríguez et al. 2021; Pradas et al. 2013).

Care management of these patients in the hospital setting is based on the care model used by multidisciplinary units. Their goal is to improve patients' survival and quality of life through the collaboration of professionals from various fields, such as neurophysiology, pneumology, palliative care, specialized nursing, physical therapy, occupational therapy, nutrition and dietetics, speech therapy, and social work (Hobson and McDermott 2016).

Their participation in care management, in the primary care (PC) setting, working in synergy with the same goals, allows ALS patients to be integrated in the area of chronicity as a care model. Different professionals participate in this model, such as the reference team (doctor, nurse and social worker) that facilitates patient integration in the home care programme (ATDOM, for its Spanish acronym) (Grupo ATDOM, 2003), the teams from the public healthcare network's palliative home care programme (PADES, for its Spanish acronym) (Busquets 2001), and the case manager (CM), who addresses all situations of complexity, dependence and frailty in a holistic manner and also acts as a link between the different care levels (Catalonia Health Plan 2016-2020 2023; Garcés and Ródenas 2015). Patients integrated in the PC area of chronicity present with one or more diseases of long evolution, which professionals perceive as having a complex clinical management. For this reason, they are identified and registered as complex chronic patients (CCPs) or included in the advanced chronicity care model (MACA, for its Spanish acronym) (Amblàs-Novellas et al. 2016; Department of Health, 2021; Gómez-Batiste et al. 2013). The model emphasizes person-centered care, prompting identified patients to engage in Advance Care Planning (ACP), which relies on patients' understanding of their illness, values, experiences, and specific choices (Lasmarías et al. 2016; Limón et al. 2018).

In recent years, a tool that assesses emotional, social and spiritual needs in end-of-life situations has been developed and validated. This tool is the Psychosocial and Spiritual Needs Evaluation (ENP-E, for its Spanish acronym) scale in end-of-life patients (Figure 2). This scale detects the complexity of patients' psychosocial and spiritual needs (PSNs), concerns, and external signs of emotional distress. Including the ENP-E scale in these patients' management facilitates to systematically evaluate their PSNs and allows us to monitor them over time, improving the implementation of personalized interventions with high sensitivity and specificity (Mateo-Ortega et al. 2019).

This study aims to know the clinical characteristics of ALS patients, their integration in chronicity programmes, and their PSNs in the north-eastern area of Spain.

Methods

Study design and patients

This was a longitudinal descriptive study carried out in the north-eastern area of Spain, close to Barcelona, with over 1.3 million inhabitants. The study period is between March 1, 2017 and February 28, 2019.

Data were sourced from computerized medical histories (eCAP, from its Spanish acronym) and the records from the Hospital Multidisciplinary Unit of Bellvitge, Hospitalet de Llobregat, Barcelona.

The ENP-E tool was chosen because it has been validated in our setting to identify the complexity of patients' PSNs, their concerns and outward signs of emotional distress and allows the implementation of personalized interventions with high sensitivity and specificity (Mateo-Ortega et al. 2019).

Inclusion criteria were patients that were alive and residing in the area, with an ALS diagnosis recorded in both study settings at the beginning of the period. Patients with unconfirmed diagnosis, not residing in the region under study and/or hospitalized in health care centers were excluded from the study.

The ENP-E survey was always administered by the same research team member, who had received prior training from the team that designed the survey. This training included detailed instructions on how to administer the survey and interpret patient responses. Every patient received an individual briefing about the survey's purpose, and their families were similarly informed to ensure understanding and support.

Variables

Clinical variables were sex, age ($<$60, 60–69, and $\geq$70 years), initial form of the disease (spinal, bulbar, or other), use of PEG, NIV, or IV by patients, and time of evolution of the disease from diagnostic confirmation of ALS until study start ($<$2, 2–4,and $\geq$5 years). These age categories and time of disease progression were chosen considering that the risk of developing ALS in Europe peaks at 65 years of age. Survival is highly variable, but respiratory failure usually results in death typically 2–4 years after onset (Brown and Al-Chalabi 2016).

Variables associated with integration in chronicity care programmes were patient identification as CCPs or inclusion in MACA on eCAP, patients registered in ATDOM, PADES and CM programmes, and patients with an ACP registration.

Variables associated with PSNs were those included in the psychosocial and spiritual needs evaluation (ENP-E) scale in end-of-life patients (Figure 1).

The ENP-E scale consists of three sections (A, B, and C) with different measuring methods:

Section A: complexity

It consists of 13 questions with a score of 0–5 points for the answers to each of them.

A global score of $\geq$28 points indicate moderate-to-severe complexity in PSNs and recommends specialized intervention (Figure 1).

It contains five key questions. A score of $\geq$4 points in any one of them also warns of a psychosocial need and recommends specialized intervention (Figure 1).

Section B: concerns

It analyses patients' concerns in the financial, family, emotional, spiritual, physical, or other aspects.

A score of $\geq$4 points in any of them is considered an affirmative answer, and it is recommended to assess the need for a specific approach and evolutionary follow-up (Figure 1).

Section C: external signs of emotional distress

The detection of any external sign of emotional distress becomes a dichotomous variable (YES/NO). If the answer is YES, specialized intervention is also recommended (Figure 1).

Initial and evolutionary assessments of PSNs have been performed through personalized interviews at the Multidisciplinary Unit of the hospital of reference using the ENP-E scale. Initial

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PSYCHOSOCIAL CARE TOOLS

2 Psychosocial and spiritual needs evaluation (ENP-E) scale in end-of-life patients

A) ANALYSIS OF PSYCHOSOCIAL NEEDS (Questions for the patient)

Below, we will ask you a series of questions about your feelings, yourthoughts, and what activities you have been doing recently:

	QUESTIONS	Score
1	**What is your state of mind?** ☐ Not explored[0] ☐ Very bad[5] ☐ Bad[4] ☐ OK[3] ☐ Good[2] ☐ Very good[1]	
2	**Are you sad?** ☐ Not explored[0] ☐ Not at all[1] ☐ A little[2] ☐ Neither a lot nor a little[3] ☐ Quite a bit[4] ☐ A lot[5]	
3	**Are you nervous?** ☐ Not explored[0] ☐ Not at all[1] ☐ A little[2] ☐ Neither a lot nor a little[3] ☐ Quite a bit[4] ☐ A lot[5]	
4	**How are you handling your situation?** ☐ Not explored[0] ☐ Very badly[5] ☐ Badly[4] ☐ OK[3] ☐ Well[2] ☐ Very well[1]	
5	**Do you speak openly with your family about your disease?** ☐ Not explored[0] ☐ Not at all[5] ☐ A little[4] ☐ Neither a lot nor a little[3] ☐ Quite a bit[2] ☐ A lot[1]	
6	**Do you believe that you need to know more about your disease?** ☐ Not explored[0] ☐ No [1] ☐ A little more[2] ☐ Indifferent [3] ☐ Quite a bit more[4] ☐ A lot more[5]	
7a	**Do you feel cared for/supported by your family members?** ☐ Not explored[0] ☐ Not at all[5] ☐ A little[4] ☐ Neither a lot nor a little[3] ☐ Quite a bit[2] ☐ A lot[1]	
7b	**Do you feel cared for/supported by your friends?** ☐ Not explored[0] ☐ Not at all[5] ☐ A little[4] ☐ Neither a lot nor a little[3] ☐ Quite a bit[2] ☐ A lot[1]	
8	**In general, do you feel satisfied with what you have done during your life?** ☐ Not explored[0] ☐ Not at all[5] ☐ A little[4] ☐ Neither a lot nor a little[3] ☐ Quite a bit[2] ☐ A lot[1]	
9	**Do you feel at peace? (no regrets or feelings of guilt)** ☐ Not explored[0] ☐ Not at all[5] ☐ A little[4] ☐ Neither a lot nor a little[3] ☐ Quite a bit[2] ☐ A lot[1]	
10	**Do you believe that your life has meaning?** ☐ Not explored[0] ☐ No[5] ☐ A little[4] ☐ Neither a lot nor a little[3] ☐ Quite a bit[2] ☐ A lot[1]	
11	**Do your beliefs and values help you in this situation?** ☐ Not explored[0] ☐ Not at all[5] ☐ A little[4] ☐ Neither a lot nor a little[3] ☐ Quite a bit[2] ☐ A lot[1]	
12	**How would you assess your overall well-being?** ☐ Not explored[0] ☐ Very bad[5] ☐ Bad[4] ☐ OK[3] ☐ Good[2] ☐ Very good[1]	
	Results – Section A	
	If the patient was able to answer every question from Section A (pages 1-12), indicate the total score after adding up the points corresponding to all the answers.	
	Did the patient score ≥4 in one or more of the key questions (1, 2, 4, 10, 12)?	☐ No ☐ Yes

B) ANALYSIS OF CONCERNS (Questions for the patient)

Taking into account everything we have talked about, How concerned are you about the following aspects?

13a	**Financial** ☐ Not explored[0] ☐ Not at all[1] ☐ A little[2] ☐ Neither a lot nor a little[3] ☐ Quite a bit[4] ☐ A lot[5]	
13b	**Family** ☐ Not explored[0] ☐ Not at all[1] ☐ A little[2] ☐ Neither a lot nor a little[3] ☐ Quite a bit[4] ☐ A lot[5]	
13c	**Emotional** ☐ Not explored[0] ☐ Not at all[1] ☐ A little[2] ☐ Neither a lot nor a little[3] ☐ Quite a bit[4] ☐ A lot[5]	
13d	**Spiritual** ☐ Not explored[0] ☐ Not at all[1] ☐ A little[2] ☐ Neither a lot nor a little[3] ☐ Quite a bit[4] ☐ A lot[5]	
13e	**Physical pain** ☐ Not explored[0] ☐ Not at all[1] ☐ A little[2] ☐ Neither a lot nor a little[3] ☐ Quite a bit[4] ☐ A lot[5]	
13f	*Is there anything else that you consider important that concerns you? How much of a concern is it?*	☐ No other concerns ☐ Yes, specify: .....
	☐ Not explored[0] ☐ Not at all[1] ☐ A little[2] ☐ Neither a lot nor a little[3] ☐ Quite a bit[4] ☐ A lot[5]	
	Results – Section B Has the patient expressed any concerns?	☐ No ☐ Yes

C) EXTERNAL SIGNS OF EMOTIONAL DISTRESS (ESD)

(to be completed by the professional)

Rate the extent to which the following external signs of emotional distress are evident (items 14a–14f) where 0 = "Not present at all" and 10 = "Totally present."

14a	Facial expression and behaviour (visible signs of sadness, fear, anger, nervousness, etc.)	(0-10):
14b	Isolation (mutism, request to lower the blinds, refusal to accept visits, lackof distractions, lack of communication, etc.)	(0-10):
14c	Demand for company/attention (constant complaints, etc.)	(0-10):
14d	Nighttime behavioural disturbances (insomnia, nightmares, unjustified requests for rescue medication, complaints, etc.)	(0-10):
14e	Other signs, specify:	(0-10):
14f	Provide an overall assessment of the intensity of emotional distress in this patient	(0-10):
	Results – Section C Does this patient show any external signs of emotional distress?	☐ No ☐ Yes

RESULTS AND DIAGNOSES OF THE ENP-E SCALE

SECTION A – Questions for the patient		
Questions 1-12 Total score (sum)	☐ Unable to exploreall the questions.	It is recommended to examine any unanswered questions in the following interventions and re-evaluate the patient when all questions have been answered.
	☐ <26. No complex needs detected.	An evolutionary follow-up is recommended.
	☐ ≥26 y <28. Borderline cases.	It is recommended to examine patients' needs in greater depth, taking into account their answers to the key questions, as well as their concerns and externalsigns of emotional distress to assess the need of care by a specialized team.
	☐ ≥28 and ≤30. Moderate warning indicators of complex needs.	A specialized intervention assessment is recommended.
	☐ >30 and ≤33. High warning indicatorsof complex needs.	A specialized intervention assessment is recommended.
	☐ >33. Severe warning indicators of complex needs.	A specialized intervention assessment is recommended.
Key questions: 1, 2, 4, 10, 12 Did the patient score ≥4 in one or more of these questions individually? (not the sum)	☐ No. No complex needs detected.	Follow the indications described for the ENP-E total score.
	☐ Yes, the patient scored ≥4 in a key question. Complex needs detected.	A specialized intervention assessment is recommended.
SECTION B – Questions for the patient		
Question 13 Does the patient have any concerns?	☐ No. No complex needs detected.	Follow the indications described for the ENP-E total score.
	☐ Yes.	It is recommended to use specific approach to address the detectedconcerns.
SECTION C - Professional evaluation		
Question 14 Are any external signs of emotional distress evident in this patient?	☐ No. No complex needs detected.	Follow the indications described for the ENP-E total score.
	☐ Yes. Complex needs detected.	A specialized intervention assessment is recommended.

Interpretation of results and practical considerations of the ENP-E scale

- Who should use this tool? Psychosocial professionals (psychologists and/or social workers) who know the patient and can start the PSICPAL programme with the ENP-E.
- The total score on the scale can range from **13 to 65points.**

  A score ≥28 means a needs complexity ranging from moderate to severe, so a **specialized intervention assessment is recommended**.

  Scores <28: This does not mean that the patient has no psychosocial and/or spiritual needs; it rather indicates a needs complexity ranging from no complexity at allto borderline cases:

  - ✓ **Scores <26: No complex needs** detected. ***An evolutionary follow-up is recommended.***
  - ✓ **Scores ≥26 and ≤28. Borderline cases:** ***It is recommended to examine patients' needs in greater depth****, taking into account their answers to the key questions, as well as their concerns and external signs of emotional distress to assess the need of care by a specialized team.*

- Analysis of concerns (Section B).

  Even though they are not part of the total score, they must be taken into account for a specific analysis. **Concerns** refer to the specific factor(s) that may be influencing patients' emotional distress and, therefore, theirpsychosocial needs. Once the patient's concerns have been identified, the need to use a specific approach to address them and perform an evolutionary follow-up should be assessed.

On the other hand, some of the **questions oritems differentiate themselves from the others. These key questions are:**

A1: "What is your state of mind?"

A2: "Are you sad?"

A4: How are you handling your situation?

A10: "Do you believe that your life has meaning?"

A12: "How would you assess your overall well-being?"

If the patient **scores ≥4** in any of these questions, this **would be a warning of** a psychosocial need.

- Presence of external signs of emotional distress (ESD).

  Patients with psychosocial needs present with more external signs of emotional distress, and, therefore, attention must be paid to these behavioural and communicative elements. If the presence of ESDs is detected, the recommendation will be to perform a specialized assessment of the patient.

- This should be carried outby a psychologist or a social workerin the context of a psychosocial interview with the patient, finding the most appropriate time to do so, without interfering with the natural, patient-led communication process and, preferably, gradually introducing the patient in as many interventions asnecessary.
- To identify these psychosocial and spiritual needs, professionals must have basic training in this field of care allowing them, on the one hand, to provide patients with immediate care and, on the other, to properly refer them to specific specialists in a timely manner.

Figure 1. The ENP-E scale.

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assessments of PSNs were performed in 46 of the 81 patients included (Figure 2).

In 24 of these patients, the evolutionary study of the PSNs using the ENP-E scale was usually carried out every 3 months (occasionally 6 months in some of them), coinciding with the visits made in the hospital unit during the study period (Figure 2).

Ethical aspects and confidentiality of data

All study patients signed an informed consent. The study was approved by the Clinical Research Ethics Committee of the Primary Care Research Institute (IDIAP, for its Spanish acronym) Jordi Gol of Barcelona (reference no. P17/005). Research good practice standards of the Helsinki Declaration were observed, as well as the Fortaleza (Brazil) amendment of 2013.

Confidentiality of all personal and research data was ensured in accordance with the applicable European legislation (European regulation 2016/679 of 27 April) on the protection of physical persons with regard to the processing of personal data and on the free movement of such data, and with the Spanish Organic Law 3/2018, of 5 December, on the protection of personal data and the guarantee of digital rights.

Statistical analysis

Quantitative variables were estimated using measures of central tendency (mean, median) and dispersion (standard deviation, percentiles), and qualitative variables were described as absolute frequencies and percentages.

Contingency tables were used to describe the potential relationship between categorical variables using Pearson's chi-squared test or Fisher's exact test, when appropriate. The means of quantitative variables were compared using Student's t-test and an ANOVA test to check the normality of data.

To analyze potential increases in the use of resources from the different health settings between the beginning and the end of study, the McNemar test was used.

To know the evolution of PSNs of patients, 1–4 interviews were carried out using the ENP-E scale. These were analyzed using relevant paired data tests (Friedman Test). Cochran's test was used to compare percentages.

The statistical analysis of data was performed using the SPSS software for Windows, version 11.5. All differences with $p < 0.05$ were considered statistically significant.

Results

The study included 81 cases registered as live ALS patients on March 1, 2017 (Figure 2).

Regarding clinical characteristics, a mean age of 65 years ($\pm$11.7) was observed at study start, with no differences between sexes. The most common initial form of the disease in both sexes was the spinal form. At study start, 14.8% were PEG users and 29.7% used NIV. During follow-up, an increase in the use

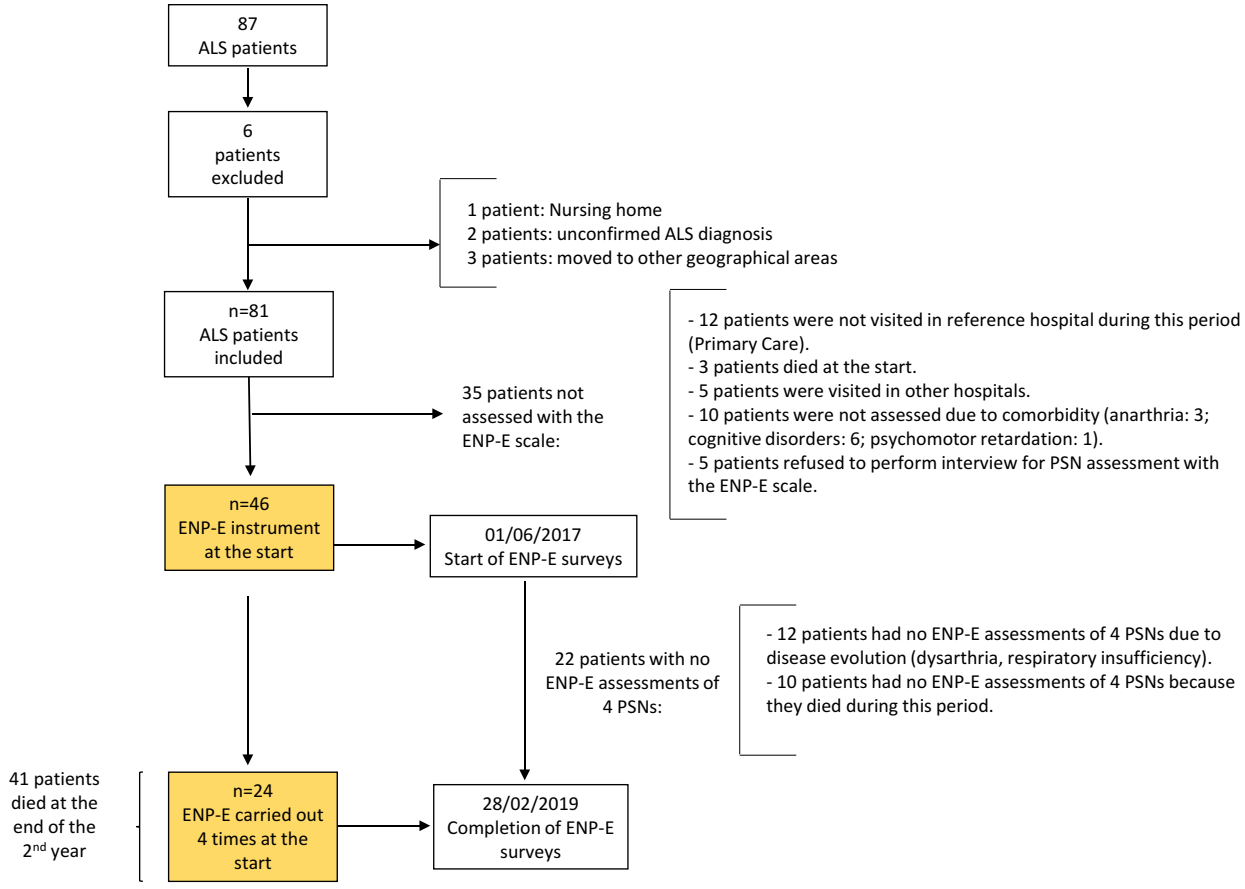

Figure 2. Study diagram.

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Table 1. Clinical characteristics and integration in chronicity care programmes of patients with ALS at the beginning and the end of the study (2 years)

Patients $n = 81$	**Age (years) Mean**	**SD**	**%**	***p*-value**
Male, $n = 40$	66.4	12.1	49.4	0.131[a]
Female, $n = 41$	64.8	11.3	50.6	

Initial clinical forms	**Spinal form**		**Bulbar form**		**Other**		
	n	**%**	***n***	**%**	***n***	**%**	***p*-value**
Male	31	77.5	6	15.0	3	7.5	0.064[b]
Female	25	61.0	15	36.6	1	2.4	

Use of PEG and NIV devices	**Initial phase**		**Follow-up period**		**Increase in device use: *p*-value**
	n	**%**	***n***	**%**	
PEG users	12	14.8	29	35.8	<0.001[c]
NIV users	24	29.7	42	51.9	<0.001[c]

Care resources from the area of chronicity	**Initial phase**		**Follow-up period**		**Increase in resource use: *p*-value**
	n	**%**	***n***	**%**	
CCP/MACA	11	13.6	50	61.7	<0.001[c]
ATDOM	20	24.7	41	50.6	<0.001[c]
PADES	13	16.0	33	40.7	<0.001[c]
CM	11	13.6	41	50.6	<0.001[c]

ACP registration on eCAP	**Initial phase**		**Follow-up period**		**Increase in ACP registrations: *p*-value**
	n	**%**	***n***	**%**	
Registered ACP	5	6.2	29	35.8	<0.001[c]

n, number of cases with this registration in their medical history; SD, standard deviation; males: 40 patients; females: 41 patients; %, percentage; Initial phase: 03/01/2017; Follow-up period: 03/01/2017 to 02/28/2019.
PEG = percutaneous endoscopic gastrostomy; NIV = non-invasive ventilation; CPP/MACA = complex chronic patient/advanced chronicity care model; ATDOM = home care; CM = case management; PADES = home care programme by teams specialized in palliative care; ACP = advance care planning.
[a]Student's *t*-test.
[b]Chi-squared test.
[c]McNemar test.

of PEG and NIV of up to 35.8% and 51.9%, respectively, was observed (Table 1). No patients used IV during this period.

Regarding the integration of patients in chronicity care programmes at study start, 13.6% were identified as CCPs/included in the MACA programme, 24.7% were in the ATDOM programme, 16% in the PADES home care programme, and 13.6% in CM. At the end of the follow-up period, these numbers had significantly increased to 61.7%, 50.6%, 40.7%, and 50.6%, respectively. Regarding ACP registration on eCAP, 6.2% had registered at study start, reaching 35.8% at the end of the study (Table 1).

One limitation of this study is the sample size at the outset (81 patients). Baseline psychosocial needs (PSNs) were assessed using the ENP-E scale in 46 patients at the beginning of the study (Figure 2). Furthermore, a longitudinal follow-up was carried out for 24 of these patients, making it possible to evaluate the evolution of PSNs at four time points in relation to variables such as age, sex, duration of the disease, usage of NIV, and PEG. Additionally, the identification in the chronic care programme and the utilization of resources such as the ATDOM, PADES, and GdC programmes were assessed (Figure 2).

The initial assessment of PSNs using the ENP-E scale of the 46 patients analyzed has been broken down in three sections (Table 2).

Section A: Complexity. A mean group score of 28.8 points is observed: 22 (47.8%) patients had a score of ≥28 points and 16 (34.8%) scored ≥4 points in any of the key questions. No significant differences were observed in complexity results regarding the following variables: distribution by sex ($p < 0.365$), age ($p < 0.655$), and time of evolution of the disease ($p < 0.360$) (Table 2).

Section B: Concerns. A higher score in concerns associated with physical pain and family aspects is observed ($p = 0.001$) (Table 2).

Section C: External signs of emotional distress. It is observed that 22 (47.8%) patients show external signs of emotional distress (Table 2).

All 46 patients were stratified by sex, three age groups (<60, 60–69 and ≥70 years) with 21.7% (10 patients), 41.3% (19 patients), and 37% (17 patients), respectively, and time of evolution of

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Table 2. Baseline assessment of PSNs using the ENP-E scale (Sections A, B, and C) in 46 ALS patients

	Group score		Patients with a score ≥28 points		Patients with a score ≥4 points in one or more answers to key questions	
Assessment Section A Range 13–65 in the ENP-E scale	**Mean**	**SD**	***n***	**%**	***n***	**%**
	28.83	7.94	22	47.8	16	34.8

	Analysis of concerns financial		Analysis of family concerns		Analysis of emotional concerns		Analysis of spiritual concerns		Analysis of concerns physical pain		
Assessment Section B Range 0–5 in the ENP-E scale	**Mean**	**SD**	**Mean**	**SD**	**Mean**	**SD**	**Mean**	**SD**	**Mean**	**SD**	***p*-value**
	2.46	1.2	4.11	1.8	2.98	1.32	1.65	0.67	4.30	1.05	0.001[a]

	Presence of external signs of emotional distress	
Assessment Section C YES/NO value in the ENP-E scale	**YES**	**%**
	22	47.8

ENP-E = Psychosocial and Spiritual Needs Evaluation scale in end-of-life patients; SD = standard deviation; *n* = number of cases with this registration; % = percentage; concerns range = 0–5 points.
[a]ANOVA test.

the disease (<2, 2–4, ≥5 years) with 43.4% (20 patients), 28.3% (13 patients) and 28.3% (13 patients), respectively. No significant differences were observed in complexity score results regarding the following variables: distribution by sex ($p < 0.365$), age groups ($p < 0.655$), and time of evolution of the sssdisease ($p < 0.360$).

Results from the evolutionary assessment of PSNs at the four interviews carried out with the 24 patients analyzed (Figure 1) during the study period are shown in Table 3.

In Section A (Complexity), a linear progression in the complexity score throughout the four measurements was observed, with a significant increase in the results of the group score, percentage of patients with a score ≥28 points, and percentage of patients with a score of ≥4 points in one or more answers to key questions.

In Section B (Concerns), it was observed that results with higher scores associated with physical pain and family aspects remain the same.

In Section C (External signs of emotional distress), a significant increase in the percentage of patients showing external signs of emotional distress during this period was observed.

Concerning the score of Section A (Complexity) of the ENP-E related with sex, a linear progression in all four assessments was observed, with significant differences in males (Table 4).

For the three age groups (Table 4), results show a linear progression in all four assessments, with significant differences between the groups <60 years and ≥70 years (Table 4).

Regarding score results associated with time of evolution of ALS, a linear progression in the four assessments of all three groups was observed, with significant differences between patients with a time of evolution <2 years and ≥5 years (Table 4).

As for the use of PEG and NIV devices (YES/NO) (Table 4), a linear progression of the complexity score in the four assessments of all four groups was observed, with a linear trend in the first three assessments. In turn, there were significant differences between the group of patients that did not use PEG, the group of NIV users and the group of patients that did not use NIV. No significant differences were observed in the group of PEG users.

When examining the complexity score results of Section A related with patient identification as CCP/inclusion in the MACA programme and integration in the ATDOM, PADES, and CM programmes, a linear progression in the four assessments of all groups was observed, with significant differences in the score in the group of patients not identified as CCP/included in MACA in the area of chronicity and the three groups of patients not integrated in the ATDOM, PADES, or CM programmes (Table 5).

Discussion

This study analyses relevant aspects about the clinical characteristics, the integration in chronicity care programmes and the PSNs assessed using the ENP-E scale of ALS patients in the North-eastern area of Spain, which are little known and have not been fully analyzed to date.

In the group of patients under study, results show that clinical characteristics such as age, distribution by sex, and initial clinical forms of the disease are similar to results published in developed countries (Al-Chalabi and Hardiman 2013; Hardiman et al. 2011) regarding a mean age over 65 years old and the prevalence of the initial spinal form in older patients, with the exception of a slight non-significant prevalence in women regarding distribution by sex.

The use of PEG feeding tubes and NIV increased significantly during the study, aligning with current trends emphasizing their benefits on survival and quality of life (Hobson and McDermott 2016); it also favors integration of ALS patients in chronicity programmes (Castro-Rodríguez et al. 2021).

At the end of the period analyzed, this study showed that more than a third of ALS patients were not integrated in chronicity

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Table 3. Evolutionary analysis of PSNs using all four assessments of the ENP-E scale (Sections A, B, and C) in 24 patients with ALS during a period of 2 years

	Group score				Patients with a total score of ≥28 points			Patients with a score ≥4 points in one or more answers to key questions		
Assessment Section A Range 13–65 in the ENP-E scale	**Median**	**P25**	**P75**	***p*-value**	***n***	**%**	***p*-value**	***n***	**%**	***p*-value**
First	26.50	21.50	33.00	0.001[a]	11	45.8	0.003[b]	9	37.5	0.012[b]
Second	30.50	23.00	33.00		15	62.5		8	33.3	
Third	31.00	24.00	36.00		16	66.7		12	50.0	
Fourth	30.50	27.25	36.00		18	75.0		14	58.3	

	Analysis of concerns financial			Analysis of family concerns			Analysis of emotional concerns			Analysis of spiritual concerns			Analysis of concerns physical pain		
Assessment Section B Range 0–5 in the ENP-E scale	**Median**	**P25-P75**	***p*-value**	**Median**	**P25-P75**	***p*-value**	**Median**	**P25-P75**	***p*-value**	**Median**	**P25-P75**	***p*-value**	**Median**	**P25-P75**	***p*-value**
First	2.00	2.00 4.00	0.632[c]	4.00	4.00 5.00	0.370[c]	3.00	2.00 4.00	0.707[c]	2.00	1.00 2.00	0.260[c]	5.00	4.00 5.00	0.321[c]
Second	2.00	2.00 4.00		4.00	4.00 5.00		2.50	2.00 4.00		1.50	1.00 2.00		5.00	4.00 5.00	
Third	2.00	2.00 4.00		4.00	4.00 5.00		3.00	2.00 4.00		2.00	1.00 2.00		5.00	4.00 5.00	
Fourth	2.00	1.25 4.00		4.00	4.00 4.75		3.00	2.00 4.00		2.00	1.00 3.00		4.00	4.00 5.00	

	Presence of external signs of emotional distress		
Assessment Section C YES/NO value in the ENP-E scale	**Yes**	**%**	***p*-value**
First	12	50.0	0.012[b]
Second	14	58.3	
Third	16	66.6	
Fourth	17	70.8	

ENP-E = Psychosocial and Spiritual Needs Evaluation scale in end-of-life patients.
[a]ANOVA test.
[b]Cochran's *Q* test.
[c]Friedman's test.

Table 4. PSN evaluation using four assessments of the ENP-E scale (Section A) in 24 patients with ALS during a period of 2 years

	First assessment		Second assessment		Third assessment		Fourth assessment		
Distribution by:	Median	P25–P75	Median	P25–P75	Median	P25–P75	Median	P25–P75	***p*-value**
Both sexes									
Males $n = 12$ (50%)	26.00	21.00–32.75	30.50	23.25–33.00	31.00	24.00–33.75	31.00	28.00–34.75	0.018[a]
Females $n = 12$ (50%)	27.50	23.25–34.75	30.00	22.25–34.00	30.50	23.50–41.25	29.50	25.50–42.50	0.078[a]
Three age groups									
<60 years $n = 5$ (20.8%)	24.00	21.00–32.00	32.00	22.00–33.00	33.00	25.50–39.50	33.00	27.00–40.00	0.022[a]
60–69 years $n = 11$ (45.9%)	27.00	20.00–33.00	32.00	20.00–33.00	31.00	20.00–32.00	30.00	24.00–35.00	0.411[a]
≥70 years $n = 8$ (33.3%)	27.00	24.50–34.75	28.50	25.00–34.00	31.00	25.25–42.75	30.00	28.25–42.75	0.004[a]
Time of evolution in years									
<2 years $n = 12$ (50 %)	25.50	21.00–34.75	26.50	21.50–34.00	29.00	23.25–38.25	29.00	25.50–41.25	0.006[a]
2–4 years $n = 6$ (25 %)	23.50	20.25–33.00	27.50	20.75–33.00	27.50	19.75–31.50	29.50	23.00–33.25	0.675[a]
≥5 years $n = 6$ (25 %)	30.00	26.75–35.00	32.00	28.75–33.50	34.00	31.25–37.75	33.50	29.75–39.50	0.046[a]
Based on the use of PEG and NIV devices									
PEG users $n = 9$ (37.5 %)	27.00	25.00–38.50	32.00	23.50–34.50	32.00	23.50–36.00	31.00	27.50–37.00	0.157[a]
Non-users of PEG $n = 15$ (62.5 %)	25.00	21.00–33.00	28.00	21.00–33.00	30.00	24.00–36.00	29.00	27.00–36.00	0.005[a]
NIV users $n = 6$ (25 %)	26.00	23.50–32.50	28.00	23.50–32.50	30.00	24.00–34.00	29.00	25.50–35.00	0.047[a]
Non-users of NIV $n = 18$ (75 %)	30.00	21.00–34.00	32.00	21.00–34.00	32.00	23.00–36.00	33.00	28.00–38.00	0.041[a]

ENP-E = Psychosocial and Spiritual Needs Evaluation scale in end-of-life patients; P25–P75 = 25th percentile–75th percentile; PEG = percutaneous endoscopic gastrostomy; NIV = non-invasive ventilation.
[a]Friedman's test.

Table 5. Relationship between the evolutions of all four complexity scores taken from Section A of the ENP-E scale in the different resources of the area of chronicity

	First assessment		Second assessment		Third assessment		Fourth assessment		
Distribution by:	Median	P25–P75	Median	P25–P75	Median	P25–P75	Median	P25–P75	*p* value
Identification as CCP/inclusion in MACA									
Identified $n = 12$ (50%)	26.50	24.00–32.75	28.50	23.00–32.75	28.00	23.25–35.00	29.50	24.25–35.75	0.082[a]
Not identified $n = 12$ (50%)	27.00	21.00–33.75	30.50	22.75–33.75	31.50	28.25–38.25	31.00	28.25–42.25	0.006[a]
ATDOM CARE programme									
Integrated $n = 8$ (33.3%)	26.50	23.50–32.75	28.00	22.20–32.75	27.50	21.00–35.00	29–50	24.00–35.50	0.029[a]
Not integrated $n = 16$ (66.6%)	27.00	21.00–33.75	30.50	23.50–33.75	31.50	25.75–38.25	31.00	28.00–41.75	0.004[a]
PADES CARE programme									
Integrated $n = 6$ (25%)	25.50	22.00–31.25	24.50	21.75–32.75	24.50	22.25–33.00	28.00	24.00–32.00	0.299[a]
Not integrated $n = 18$ (75%)	29.00	21.00–33.25	32.00	22.75–33.25	31.50	26.75–36.75	32.00	28.00–37.75	0.005[a]
CM CARE programme									
Integrated $n = 12$ (50%)	26.50	24.00–32.75	28.50	23.00–32.75	28.00	23.25–35.00	29.50	24.25–35.75	0.082[a]
Not integrated $n = 12$ (50%)	27.00	21.00–33.75	30.50	22.75–33.75	31.50	28.25–38.25	31.00	28.25–42.25	0.006[a]

CCP/MACA, identification as complex chronic patient/inclusion in the Advanced chronicity care model; *n*, number of patients; P25–P75, 25th percentile–75th percentile; ATDOM, home care programme; PADES, home care programme by teams specialized in palliative care; CM, case management.
[a]Friedman's test.

programmes. This may mean that a significant number of them stopped having access to the comprehensive care model that provided a multidimensional vision of ALS patients and their families (Gómez-Batiste et al. 2010). This multidimensional vision also entails an assessment of healthcare needs, including access to professional support, such as social workers, who implement effective

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intervention plans to organize patients' care, the involvement and cohesion of the family, and improve the setting, whether by adapting the patient's home or their social surroundings (Department of Health 2021). In this sense, we propose to create a territorial "care path" for ALS patients to improve their integration in chronicity programmes. This "care path" must be understood as an agreement between professionals and organizations from different care settings, operating within the same region and that care for the same population that has a specific chronic health issue, to efficiently apply good practice criteria, optimize care circuits, and plan a response to predictable scenarios (Department of Health 2021).

One of the tools included in chronicity programmes and PC in general is to promote ACP creation and registration (Limón et al. 2018). This study shows that only one third of ALS patients have an ACP registered by the professionals who care for them in their electronic medical records. During the interviews, we found that no such ACP records in any other format. For this reason, it is important to promote actions to identify the barriers that make it difficult, or that do not favor ACP creation and registration, whether they are related with patients, professionals or organizations (Lasmarías et al. 2016), to then increase and improve ACP registration in ALS patients' medical histories.

One of the tools included in chronicity programmes and PC in general is to promote the creation and registration of PCAs (Limón et al. 2018). This study shows that only one third of ALS patients have an ACP recorded in their electronic medical records to identify barriers that hinder or do not favor the creation and recording of ACP, whether related to patients, professionals or organizations (Lasmarías et al. 2016), in order to then increase and improve the recording of ACP in the clinical records of ALS patients.

PSNs evaluation using the ENP-E scale is a novel perspective in the evolutionary analysis of ALS patients, which examines little-known factors such as the levels of complexity assessed by clinicians, but also includes aspects that are typical of patients, such as concerns and potential signs of emotional distress.

Results show that almost half of patients under study present with moderate-to-severe levels of PSNs complexity at the beginning of the study, which progressively and significantly increase until reaching 75% of patients at the end of the study.

Regarding concerns detected through Section B of the ENP-E scale, those particularly associated with family aspects and physical pain stand out. Regarding the presence of external signs of emotional distress detected in Section C of the ENP-E scale, it is worth noting that, at the end of the study period, three out of four patients showed external signs of emotional distress, which is consistent with the impact of the disease concerning dependence and the fateful prognosis (Brown and Al-Chalabi 2016).

The presence of these signs may seem logical, but it might help that professionals and relatives identify them to seek the necessary support and resources.

The results obtained are unequivocal warning indicators of a psychosocial need and are consistent with the opinions of the authors of the ENP-E scale that it is necessary to systematically perform an assessment and an evolutionary follow-up, and address PSNs using the ENP-E scale (Mateo-Ortega et al. 2019) during the care process of all patients in an advanced stage of a disease, to improve the quality of care in ALS patients.

Regarding PSNs complexity scores of the assessments grouped by sex, age and time of evolution of the disease at the beginning of the study, no differences in complexity have been observed. However, in the evolutionary analysis of PSNs, there is a significant increase in overall complexity scores of male patients. Regarding patients' age, there is a higher complexity in younger and older patients.

In terms of disease evolution time, an increase in patients with a time of less than 2 years and equal to or more than 5 years of evolution has been observed. It is difficult to find an explanation for these variations with our current knowledge of PSNs (Mateo-Ortega et al. 2019). Some factors may include the small number of cases analyzed, as well as other cultural, financial and social factors that were not the object of this study. More extensive studies are needed to compare these results.

Regarding the evolution in PSNs complexity scores associated with the use or not of feeding and breathing devices, such as PEG and NIV, a progression in group scores is also observed, with the exception of PEG users, where differences were not significant. Causes of this can be attributed to the low number of cases included in each group, although, in the group of PEG users, this could also be explained by the improvement in nutritional and life quality aspects perceived, an opinion shared with other authors (Katzberg and Benatar 2011).

Patients not integrated into chronic care programmes showed a greater increase in PSN complexity scores than those integrated (Boult et al. 2011; Lynn and Adamson 2003). Although sample size could affect the statistical results, these scores suggest that palliative measures by these teams could reduce perceived complexity. The evaluation of the ENP-E scale underscores the need to include all ALS patients in chronicity programmes and to involve professionals such as psychologists and social workers to address the specific NPS of ALS patients (Amblàs-Novellas et al. 2016; Department of Health 2021).

This study has some strengths and limitations. Among the main strengths of this study are the selection of the ENP-E scale, a validated tool in our specific context, and the exhaustive inclusion of all patients identified within a public health service covering a wide geographical area. In addition, it has provided us with the opportunity to explore initial and progressive PSNs which, to our knowledge, have not been previously published in the existing literature.

A limitation of our study lies in the relatively small sample size. This limitation is a common feature of most published studies in this field (Barker and Rose 1992). At the beginning of the study, we were only able to assess baseline PSNs using the ENP-E scale in 46 patients (Figure 2). However, we did not correlate the results of these 46 patients with other variables.

On the other hand, it was possible to perform a longitudinal follow-up in a subset of 24 of these patients. This follow-up allowed us to assess the progression of PSNs at four different time intervals, taking into account several variables (Figure 2). Despite its limited size, this longitudinal follow-up provides valuable information on the evolution of PSNs in ALS patients.

One conclusion that we have reached is that we were able to detect a high number of ALS patients that are not integrated in chronicity programmes or using their care resources; one way to improve this integration could be to create a specific "care path" for ALS patients in the region. Another relevant aspect is that ALS patients under study showed high levels of PSNs complexity, which can serve as a warning in the care of these patients. These data support the proposal of exploring PSNs using the ENP-E scale and the specific approach of PSNs in all patients with ALS (Mateo-Ortega et al. 2019).

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Results and conclusions of this study must be interpreted as hypothesis generators, which will have to be compared in more extensive and complex studies over a longer period of time and a wider geographical area.

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