

Attitudes of caregivers towards prolonging and shortening life in advanced stages of amyotrophic lateral sclerosis

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Folia Neuropathol 2023; 61 (4): 349-359

DOI: <https://doi.org/10.5114/fn.2023.130444>

Abstract

Introduction: Inevitable disease progression in amyotrophic lateral sclerosis (ALS) forces patients and their caregivers (CGs) to reflect on end-of-life treatment. The CGs are often heavily burdened with their role of surrogate decision-makers. The aim of the study was to analyze attitudes of CGs and presumable attitudes of ALS patients from the CGs' perspective towards palliative care in advanced disease stages.

Material and methods: One hundred and sixty four CGs from Germany and Poland were interviewed regarding their own preferences and patients' ideational attitudes towards life-prolonging (invasive and non-invasive ventilation, tube feeding) and life-shortening methods (termination of measures, active measures if permitted by law). The data were correlated with patient- and CG-related factors: demographic and clinical data, care commitment, depression and quality of life (QoL).

Results: The CGs were mostly female spouses of ALS patients, with secondary/higher education. Nearly 70% (81% in Poland, 57% in Germany; $p = 0.0001$) reported positive attitudes towards life-prolonging methods, which positively correlated with religiousness and negatively with patients' age. Approximately 40% of CGs (25% and 51% respectively; $p = 0.001$) reported positive attitudes towards life-shortening methods. It positively correlated with time since diagnosis and negatively with the CG's QoL, religiosity and religious/spiritual faith as factors that significantly influenced end-of-life decisions. There was a strongly positive correlation between CGs' positive attitudes towards life-shortening methods and presumed positive patients' attitudes assessed by their CGs ($p < 0.000001$).

Conclusions: Although attitudes towards treatment differed between countries, the CGs of ALS patients were generally positive towards life-prolonging treatment. A greater acceptance of life-shortening methods in the case of longer disease duration and poorer QoL may indicate worse coping with disease progression and weaker adaptation mechanisms in CGs compared to those previously reported in ALS patients. A close resemblance of the CGs' answers to probable patients' attitudes reported by the CGs indicates that many GCs might actually express their own culturally shaped attitudes towards end-of-life methods. In light of earlier-reported discrepancies between presumed opinions of the CGs and of patients themselves, a greater focus should be placed on thorough discussions on future treatment options with ALS patients in the presence of their CGs, to stay in line with the patient's authentic will.

Key words: palliative, depression, quality of life, caregivers, amyotrophic lateral sclerosis, end of life, life-sustaining methods, life-shortening methods, shared decision-making.

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Introduction

Due to a lack of efficient neuroprotective treatment, the therapy of amyotrophic lateral sclerosis (ALS) mainly focuses on symptomatic and palliative care. Although muscle weakness constitutes the main burden for the patients, there are two major risks of the disease: malnutrition and respiratory insufficiency. Up to 80% of ALS patients develop bulbar syndrome [45], which may severely affect food intake. As malnutrition worsens the disease prognosis [27], patients require a high-calorie diet [31], fluid thickeners, appropriate nutritional supplies followed/accompanied by tube feeding [45]. Introducing gastrostomy reduces the risk of weight loss, dehydration, fear of choking and social isolation associated with prolonged time of the meals and their aesthetics [42]. Despite obvious clinical benefit, many patients are reluctant to undergo a surgical procedure [30]. Concomitantly, during the disease progression, the involvement of respiratory and paraspinal muscles leads to respiratory insufficiency. An increase in the partial pressure of carbon dioxide in blood causes fatigue and drowsiness, followed by orthopnea and dyspnea at rest [17]. Respiratory failure requires the use of assisted breathing techniques, such as non-invasive (NIV) and invasive ventilation (IV) [46]. NIV is generally more acceptable due to its non-invasive and intermittent character, relatively good tolerability and ease of use [17]. Invasive ventilation, which is usually introduced due to progression of respiratory insufficiency or sialorrhea [50], is generally associated with good acceptance by patients, a reduction of a feeling of suffocation and easier mucus removal. On the other hand, it causes communication difficulties and a need for 24 h-home care, which burdens the caregivers (CGs) [4,28,47]. To facilitate the choice of optimal treatment when the disease advances, ALS patients and their CGs should be informed about life-prolonging treatments in advance. The information is advised to be given at the time of diagnosis and periodically afterwards [39]. Time of discussions may also be determined by country-specific legal regulations, including advanced directives. Starting a discussion concerning end-of-life treatment might be difficult for physicians with regard to the right timing of appropriate information according to patients' needs, which may thus be determined not only by physicians' training, but also by professional and life experience [7]. Also not all the patients are eager to undertake such discussions, halted at times by their avoidant type of coping strategies with the fatal disease [8,19,21], which are known to be protective to secure QoL for a certain amount of time [38]. Patients' ultimate choices may thus be influenced by legal regulations (advance directives, power of attorney

given to the CG, withdrawing life-sustaining therapies on request and assisted dying), individual attitudes towards life-prolonging/shortening methods, and personal circumstances (medical, social, financial, cultural etc.) [9,49,54]. ALS patients usually discuss treatment options with their family members/caregivers. In case of disease progression, the latter might need to take surrogate decisions on end-of-life treatments. Therefore the CGs' personal perspective may be crucial in the decision-making process. The aim of the study was to analyze the caregivers' attitudes towards life-prolonging and life-shortening methods in the advanced stage of ALS, as well as their determinants. It was also considered important to find out the opinion of the CGs on presumable attitudes of ALS patients towards decisions of prolonging/shortening life. The study was performed in two neighboring European countries, Germany and Poland, with different legal regulations. The main differences concern advanced directives and the possibility of treatment withdrawal. The first applies in Germany, but not in Poland. As for the latter, although active suicide assistance remains illegal in both countries, the withdrawal of artificial life support is allowed in Germany, while it is considered an assisted suicide in Poland [5,41].

Material and methods

We recruited 80 Polish and 84 German informal CGs of ALS patients followed at the Department of Neurology, Medical University of Warsaw and University of Ulm. The study was conducted prospectively using a questionnaire designed within the JPND Needs in ALS project (www.NEEDSinALS.com). The inclusion criteria were: taking care of patients with clinically possible, probable or definite ALS according to the El Escorial criteria [11] and disease duration of > 3 months. The exclusion criteria included clinical diagnosis of dementia, additional neurological/psychiatric disorders or locked-in state.

The CGs were asked a series of questions concerning their own attitudes towards the use of life-prolonging methods (NIV, IV, percutaneous gastrostomy – PEG) for the patient in case it was medically justified, and life-shortening methods (if permitted by law) at the advanced stage of the disease (Table I). They were also asked about presumed attitudes of ALS patients they took care of to life-prolonging and life-shortening methods (Table I). The CG- and patient-dependent factors which could potentially influence their attitude were also recorded (Tables II, III). The CG-dependent factors included: age, sex, years of education, relation to the patient, living in a shared household with the patient, time spent on functional care (assistance in walking, changing position, hygiene, meals) and emotional sup-

Table I. Survey questions considering the caregiver's opinion on the use of life-extension and life-shortening methods

Survey questions	Answer
1. Would you like your relative with amyotrophic lateral sclerosis to use life-prolonging measures?	a) definitely not b) probably not c) probably yes d) definitely yes
2. If it were permitted by law, would you like your relative with amyotrophic lateral sclerosis to use life-shortening measures (active or passive euthanasia or switching off the life-sustaining measures)?	a) definitely not b) probably not c) probably yes d) definitely yes
3. What do you think is the patient's attitude towards life-shortening methods (possibility of disconnecting life-support equipment, active euthanasia)?	a) negative b) mostly negative c) mostly positive d) positive
4. Have you ever searched for information on euthanasia/active dying assistance?	a) no b) yes
5. Can your religious/spiritual/faith influence your possible decisions concerning extending or shortening your life in a serious disease?	a) no, it cannot b) it may have some effect c) it has a significant impact

Table II. Characteristics of the sample of patients with amyotrophic lateral sclerosis

Patients	Entire group N = 164	Polish group n = 80	German group n = 84	p-value	
Gender F : M (%)	41.46 : 58.54	41.25 : 58.75	41.66 : 58.34	0.95	
Age (years), mean \pm SD	59.26 \pm 12.58	59.11 \pm 11.59	59.41 \pm 13.45	0.88	
Time since onset (months), mean \pm SD	29.5 \pm 21.6	27.8 \pm 14.44	31.1 \pm 26.45	0.33	
Time since diagnosis (months), mean \pm SD	13.65 \pm 12.42	12.05 \pm 8.53	14.4 \pm 14.8	0.4	
ALSFERS-R (0-48), mean \pm SD	34.07 \pm 7.53	34.36 \pm 6.12	33.8 \pm 8.5	0.65	
MIs (%)					
	NIV	12.8	2.5	22.6	0.001
	IV	0.6	0.0	1.2	–
	PEG	6.7	5.0	8.33	0.05
Patient's children, %					
	0	18.6	8.3	27.4	0.02
	1	17.9	6.7	19.0	
	2	52.6	62.5	44.0	
	≥ 3	10.9	12.5	9.6	
Children in the same household					
	Yes	71.9	62.9	78.6	0.2
	No	28.1	37.1	21.4	
Partner in the same household					
	Yes	81.2	84.3	78.6	0.33
	No	18.8	15.7	21.4	

ALSFERS-R – ALS Functional Rating Scale–revised version, MIs – medical interventions, NIV – non-invasive ventilation, IV – invasive ventilation, PEG – percutaneous gastrostomy. *Statistically significant correlations in bold.

port, CG's personal involvement in patient support associations, former experience in caring for chronically ill people, suffering from chronic diseases themselves, sleeping problems, seeking information on euthanasia/

assisted dying. The CGs filled in questionnaires on the QoL (Anamnestic Comparative Self Assessment – ACSA [10] and Quality of Life in Life-Threatening Illness-Family Career Version – QOLLI-F [14]), religiosity

Table III. Demographic characteristics of the caregivers of patients with amyotrophic lateral sclerosis

CGs	Entire group N = 164	Polish group n = 80	German group n = 84	p-value
Gender (%)				0.05
Male	32.3	25	39.3	
Female	67.7	75	60.7	
Age (years) (%)				0.38
Mean ±SD	52.40 ±14.59	51.36 ±14.70	53.39 ±14.43	
Median	53	53.5	53	
< 30	7.2	9.0	6.0	
30-39	12.0	17.8	7.2	
40-49	22.3	17.8	27.6	
50-59	24.2	26.4	24.8	
60-69	22.7	14.4	18.2	
> 70	14.4	10.2	14.4	
Education (years)				0.85
Mean ±SD	14.40 ±2.78	14.36 ±2.58	14.45 ±2.97	
Median	14	15	14	
Religion (%)				0.0001
Catholic	59.8	91.3	29.8	
Protestants	20.1	0.0	39.3	
Atheists	18.9	8.8	28.6	
Other	1.2	0.0	2.4	
IIR (4-17)				0.0001
Mean ±SD	6.82 ±4.933	11.10 ±3.634	2.75 ±0.890	
Median	6	12	5	
Attending services (2-10)				0.0001
Mean ±SD	4.07 ±2.530	5.88 ±2.166	2.36 ±1.411	
Median	3	6	2	
Degree of belief (2-7)				0.0001
Mean ±SD	3.96 ±1.784	5.23 ±1.599	2.75 ±0.890	
Median	4	6	3	
CG's family status (%)				0.54
Spouse/partner	65.9	61.3	70.2	
Children	22.0	25.0	19.0	
Parents	3.7	5.0	2.4	
Siblings	3.7	2.5	4.8	
Others	4.9	6.3	3.6	
ECAS-BS (0-10)				0.1
Mean ±SD	0.55 ±0.99	0.63 ±1.02	0.48 ±0.97	

CGs – caregivers, IIR – Idler Index of Religiosity, ECAS-BS – Edinburgh Cognitive and Behavioural ALS Screen-Behavioural Score; *Statistically significant correlations in bold.

(Idler Index of Religiosity – IIR) [26] and a question on a possible influence of religious/spiritual faith on their attitudes towards life-prolonging and life-shortening methods (“May your religion/spiritual faith influence your potential decisions on prolonging or shortening life in a severe disease?”). The data were analyzed in

the context of patient-related factors of the corresponding ALS patients. They included functional status (ALS Functional Rating Scale-Revised – ALSFRS-R [13]), behavioral status (CG survey component of the Edinburgh Cognitive and Behavioral ALS Screen – ECAS-BS [1]), disease duration, time from diagnosis, and the time

period in which the patient received information about the life-prolonging and life-shortening methods. The study was approved by the institutional Ethical Committees (KB/138/2013, LU 336/13-2 BI 195/54-2 and 01GM1103A). Each participant signed written informed consent prior to participation in the study.

Statistical analyses

The analysis of data was performed using SPSS version 18, with a p -value 0.05 considered statistically significant. Normal distribution was tested with the Kolmogorov-Smirnov test. Descriptive statistics were used to analyze the participant's demographics and distribution of sociodemographic variables, clinical findings, CG and family details collected using the structured interview schedule. Comparisons were assessed using Student's t -test, while the correlation was determined with Pearson's test. Spearman rank order was used for some correlation tests.

Results

Patients from both countries had a similar frequency of IV use, while the use of PEG and NIV was more frequent among German patients. There were no differences in the functional status or disease duration between groups (Table II). The majority of CGs were patients' partners or spouses (65.9%), 67.7% females, mean age of 53.39 ± 14.43 years and mean years of education of 14.45 ± 2.97 years (Table III). The characteristics of the study group were previously described [37].

Attitude of caregivers towards life-prolonging methods. Overall

The majority of CGs (69.5%) had a positive attitude towards life-prolonging methods. The above attitude positively correlated with CGs' religiosity ($\tau = 0.219$, $p = 0.001$) and negatively with patients' age ($\tau = -0.135$,

Table IV. Attitude of Polish and German caregivers to life-prolonging and life-shortening medical interventions

	Entire group $N = 164$	Polish group $n = 80$	German group $n = 84$	p -value
<i>Would you like your relative with ALS to use life-prolonging measures?</i>				
No or probably not (%)	30.5	17.5	42.9	0.0001
Probably yes and yes (%)	69.5	82.5	57.1	
<i>If it were permitted by law, would you like your relative with ALS to use life-shortening measures (active or passive euthanasia or switching off the life-sustaining measures)?</i>				
No or probably not (%)	61.3	73.8	48.8	0.001
Probably yes or yes (%)	38.7	26.2	51.2	

ALS – amyotrophic lateral sclerosis. *Statistically significant correlations in bold.

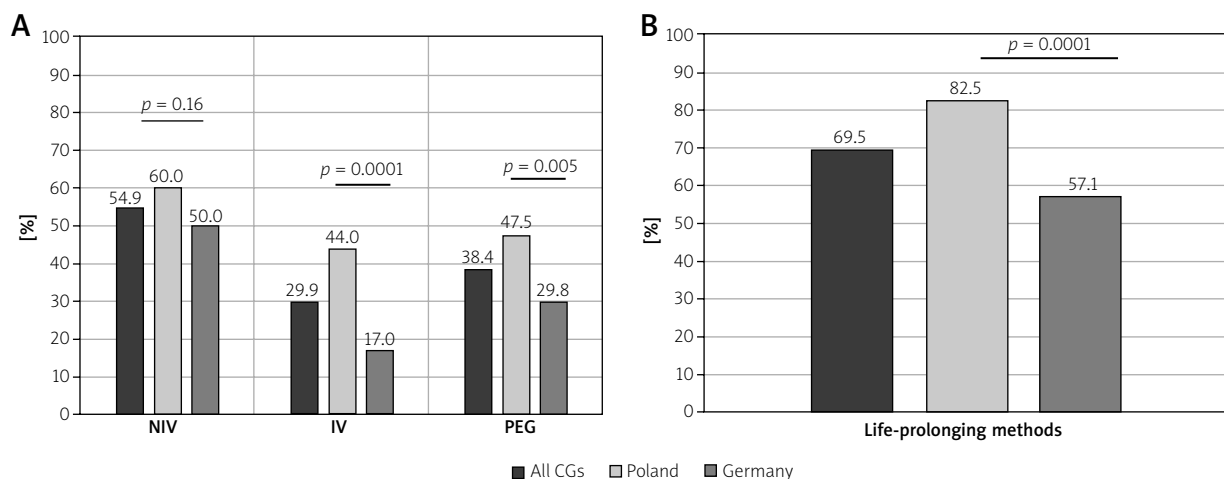


Fig. 1. Caregivers' opinion on the patient's attitude to the use of medical interventions in the advanced stage of the disease (A) and attitude of Polish and German caregivers to life-prolonging medical interventions (B).

$p = 0.038$) (Table IV). The CGs' response concerning a significant impact of the religious/spiritual faith on end-of-life decisions correlated with a positive attitude towards life extension methods ($\tau = 0.162, p = 0.029$). There was no correlation with other CG-related factors: gender, relationship, education, suffering from chronic diseases, presence of sleep problems, experience in the care of chronically ill people, involvement in associations, QoL (QOLLI-F and ACSA), time spent on functional and emotional care; or patient-related factors: ECAS-B, ALSFRS-R, living in a shared household with the patient and children, disease duration or time from the diagnosis and the time of receiving information concerning life-prolonging methods. When asked more specific questions considering the decision the patient has probably taken considering the use of NIV, IV or PEG in future, the responses were significantly lower compared to CGs' preferences and ranged from 29.9% (IV) through 38.4% (PEG) to 54.9% (NIV) (Fig. 1).

Difference between Poland and Germany

A significantly higher proportion of the CGs from Poland (82.5%) reported that they would like the patient to undergo life-prolonging interventions as compared to the German CGs (57.1%, $p = 0.0001$) (Table IV). In the opinion of Polish but not German CGs, their religious/spiritual faith could influence their decisions concerning extending or shortening life in advanced disease stages ($\chi^2 = 23,547, p = 0.0001$). Forty percent of Polish CGs rated the above influence as significant and 21.3% as moderate, as compared to 8.3% and 25.0% of the German CGs, respectively. There was no correlation between the rating of the potential influ-

ence and the CGs' attitude towards particular methods extending or shortening life. Among factors that differed between the two countries and which could potentially influence the CGs' attitude was the time of receiving information about life-prolonging methods. Although both groups obtained information mainly on their own request (36.3% and 37.3% in Poland and Germany, respectively), in the German group 37.3% of patients received information at the time of diagnosis. In the Polish group, the second most frequent moment of obtaining information was approximately 6 months before it was medically indicated (26.3%) (Fig. 2). The results did not correlate with the CGs' attitude towards life-prolonging methods in any of the populations.

Attitude of caregivers towards life-shortening methods. Overall

Over 1/3 of CGs (38.7%) would like the ALS patients to use life-shortening methods if they were legally allowed (Table IV). The attitude towards the use of life-shortening methods positively correlated with time since diagnosis (Kendall's tau correlation; $\tau = 0.131, p = 0.046$) and negatively with CGs' QOLLI-F ($\tau = -0.140, p = 0.03$). CGs' religiosity ($\tau = -0.161, p = 0.018$) and CGs' religious/spiritual faith were factors that were significantly associated with end-of-life decisions ($\tau = -0.244, p = 0.001$). CG's attitude did not depend on CG-related factors: age, gender, relationship, education, suffering from chronic diseases, presence of sleep problems, experience in the care of chronically ill people, involvement in associations, QoL (ACSA), time spent on functional and emotional care; or patient-related factors: ECAS-B, ALSFRS-R, living in

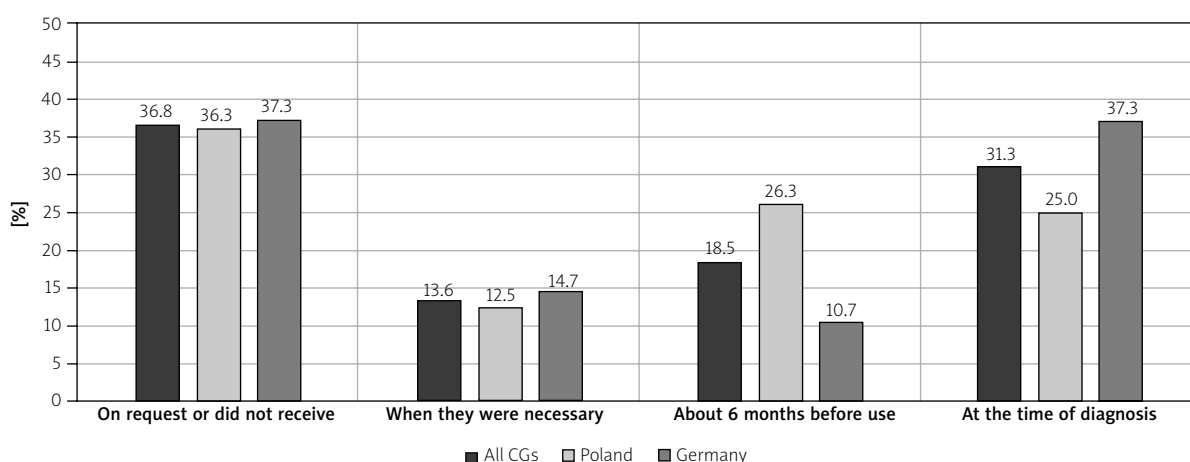


Fig. 2. Time of informing patients with amyotrophic lateral sclerosis about life-extending methods in Poland, Germany and in both countries in general.

a shared household with the patient and children, disease duration and the time of receiving information concerning life-shortening methods (all $r < 0.13$ and $p > 0.057$).

Difference between Poland and Germany

If the life-shortening methods were legally allowed, 26.2% of Polish and 51.2% of German CGs would like the patient to use them ($\chi^2 = 10.711$, $p = 0.001$) (Table IV). Positive attitudes towards the use of life-shortening methods negatively correlated with years of education in Poland ($\tau = -0.237$, $p = 0.016$), and with the number of the patient's offspring in Germany ($\tau = 0.211$, $p < 0.04$).

In the opinion of 30.0% of Polish and 53.0% of German CGs, the patients had a positive or rather positive attitude towards life-shortening methods (withdrawal from life support and/or active euthanasia) ($\chi^2 = 8.872$, $p = 0.003$). The results closely mirrored the CGs' own responses; positive attitude of CGs strongly correlated with CGs' opinion on presumed positive attitude of the patient they would take care of ($R = 0.6$, $p < 0.000001$; Spearman's rho) (Fig. 3).

Also the time of receiving information about life-shortening methods differed between the two countries. Almost 64.8% of patients in Germany and 90.0% in Poland (78.1% overall) received information on life-shortening methods at their own request ($\chi^2 = 18.882$, $p = 0.0001$). A significant group of 18.3%

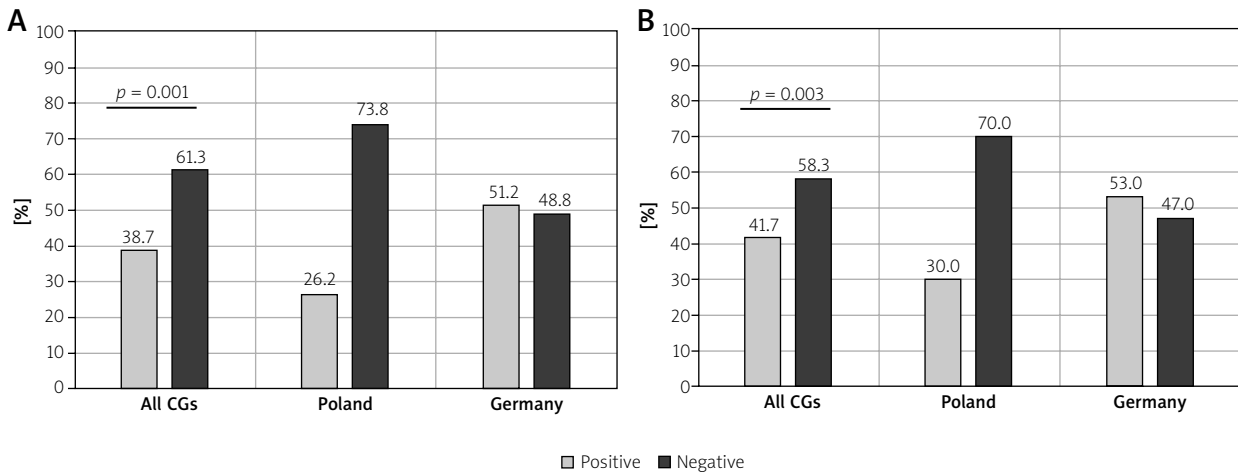


Fig. 3. The attitude of caregivers (A) and the ideational attitude of patients with amyotrophic lateral sclerosis (B) to life-shortening methods at an advanced stage of the disease in Poland and Germany.

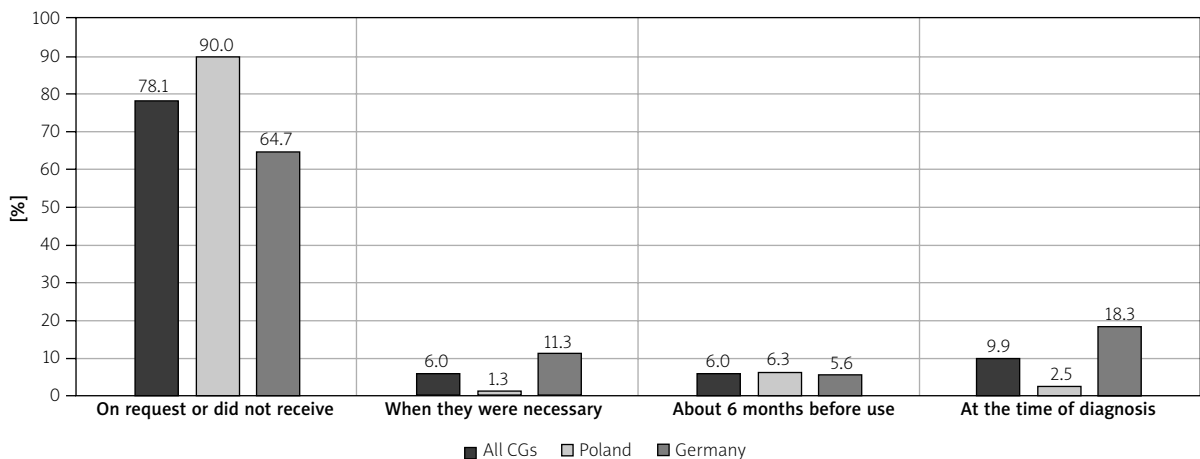


Fig. 4. Time of informing patients with amyotrophic lateral sclerosis about life-shortening methods in Poland, Germany and in both countries in general.

of German and only 2.5% of Polish patients received the information at the time of diagnosis (9.9% overall) (Fig. 4). The results did not correlate with CGs' attitude towards life-prolonging methods in either population.

Searching for information on life-shortening methods. Overall

A total of 14.0% of CGs reported having looked for information about active life-shortening methods. The search negatively correlated with CGs' religiosity as assessed by the IR scale ($\tau = -0.224$, $p = 0.001$) and positively with CGs' belief that their religious/spiritual faith would not influence the end-of-life decision ($\tau = -0.198$, $p = 0.008$). It did not depend on the CGs' age, gender, relationship with patient, education, QoL (ASCA, QOLTI-F) and patient's functional status, the presence of behavioral impairment, disease duration, time from diagnosis, or living in a shared household with CGs or their children (all $\tau < 0.115$, $p > 0.08$).

Differences between countries

The CGs from Germany searched for information on active life-shortening methods or euthanasia significantly more often compared to the Polish CGs (22.6% vs. 5.0% in Germany and Poland, respectively, $\chi^2 = 10.549$, $p = 0.001$). The search for the above information among the Polish CGs positively correlated with the presence of sleep disorders ($\tau = 0.301$, $p = 0.007$). The search was not related to any other of the CG- or patient-dependent factors.

Discussion

The inevitable disease progression of ALS forces the patient and his/her family to reflect on the end-of-life treatment. The CGs of ALS patients are often heavily burdened with their role and experience severe emotional and physical distress [16,24]. In the past years it was observed that the CG's judgment of the patient's QoL and mood was lower, while the wish for hastened death heavily misjudged – either higher or lower but not in line with the patient's self-rating [6,32]. The opinions of the CGs related to patient's QoL and wish of death were conditioned by the CGs' personal physical and emotional state [32], which could have possibly influenced the surrogate decisions they might have to take in case of the patient being in an unresponsive state (e.g. locked-in state).

We aimed to analyze the attitude of a large group of CGs towards the general use of life-prolonging and life-shortening methods, as well as to identify its determinants. The character of the CG population was similar to that in previous studies [48,55], with the

majority being women in their middle age, most frequently patients' spouses or life partners. A positive attitude of the CGs towards life-prolonging methods in ALS dominated in both countries. A similar attitude towards these methods was previously expressed by ALS patients [33], as well as ALS patients in locked-in syndrome – individuals using life-prolonging methods would decide to use them again if time was turned back and would never consider giving up their use [29]. In our study, positive attitudes towards prolonging life correlated with higher religiosity of the CGs and a subjective rating of the potential influence of their religious/spiritual faith on end-of-life decisions. Since the protection of human life is deeply rooted in most religions, it may influence decisions about medical interventions in both patients and their CGs [35,52]. A negative correlation with the patients' age could potentially be related to a worse general condition, coexistence of additional life burdens not analyzed in this study and/or a general acceptance of the natural course of life and death in advanced age [20]. Interestingly, despite a positive attitude of the CGs towards the future use of life-prolonging methods in general, their view of the actual positive decisions that patients were likely to make regarding the use of either PEG, NIV or IV were significantly lower. It may be assumed that the responses possibly expressed the CG's own attitudes towards these particular interventions, indicated by the similarity of responses as opposed to the general wish for life prolongation of their proxies. The correlation between the direct attitudes of ALS patients towards end-of-life treatment and the patients' ideational attitudes in the opinions of their respective CGs were recently reported in an article by Andersen *et al.* [6].

Caring for a patient who requires nutritional/respiratory support is extremely demanding, especially in the case of PEG and IV due to their invasiveness and possible complications [12,30,40,53]. As previously reported, CGs had fewer concerns about caring for patients using NIV compared to IV [40]. In our study, similarly, the patients' positive ideational decisions on the future use of IV and PEG were less frequent than NIV.

Almost twice as many Germans (51.2%) compared to Polish CGs (26.3%) wanted the patients to use life-shortening methods if permitted by law. The positive CGs' attitude correlated with a lower quality of life, possibly due to a high burden of the disease and accumulation of numerous negative life changes associated with prolonged care. It was also related to lower religiosity and the opinion that spiritual faith would not influence their decisions on prolonging/shortening life. The role of religion and spirituality in coping with end-of-life problems has been widely described [3,22,35]. Likewise, lower religiosity was found to be associated

with a greater feeling of hopelessness and a positive attitude towards life-shortening methods [22]. The acceptance of life-shortening methods by the patients' CGs increased with time from diagnosis. It might indicate that – differently from ALS patients [5] – the CGs do not adapt to the disease during its course. It is an important finding in the context of surrogate decisions which the CGs need to take when the patients are no longer able to express their will. Indeed, 53% of German and 30% of Polish CGs reported that the patients considered the use of life-shortening methods in the advanced stage of the disease, which exactly mirrored the opinions of the CGs themselves (52% and 26%, respectively). The results may indicate that the CGs transferred their own feelings to the presumed decisions of their relatives with ALS. Indeed, the CGs' participation in the decision-making process may increase along with a restriction of patients' autonomy and ability to make decisions [25]. In our previous study, we found that although mean results of the wish for hastened death did not significantly differ between ALS patients and the estimated opinion of their CGs, the individual answers of the CGs did not match those of ALS patients at all but were instead highly over- or underrated [6]. In other studies, the majority of patients had a low desire to hasten death, gradually decreasing over time [33], and patients with locked-in syndrome did not consider shortening their lives [34]. Thus, in view of these and current results, it is advisable to verify the compliance of the views of people taking surrogate decisions with the patient. It may include doctors or psychologists conducting open interviews with the patients at a less advanced disease stage in the presence of their CGs [43] and continuing discussing the matter gradually as the disease progresses.

The German CGs significantly more often searched for information about life-shortening methods. Apart from religiosity [19], no other factors seemed to correlate with the above search. Consistently with other studies, the less religious CGs were more likely to look for information on life-shortening methods [2,3,22,23,35,36,44,51].

The EFNS guidelines and the advanced directives (legally binding in Germany but not in Poland) suggest including a palliative approach in the care plan of ALS patients from the moment of diagnosis [18]. However, in both countries, the ALS patients most often received information about life-prolonging and life-shortening methods only at their own request, regardless of disease duration or time from diagnosis. Moreover, only 1/3 of German patients received information about life-prolonging methods at the time of diagnosis, as required by advanced directives. It was only slightly lower in Poland (25%). Although the moment of receiv-

ing information did not affect the patients' attitudes towards life-prolonging and shortening methods [15], with this approach, in patients who develop frontotemporal dementia in a later course of the disease, the information might be given too late to allow correct judgment. This, in turn, might burden the patients' CGs to an even higher extent.

Conclusions

Choosing end-of-life treatment in ALS is challenging for both patients and their CGs. We found that the CGs have a generally positive attitude towards life-extending methods and a negative attitude towards life-shortening methods. Attitudes towards the future use of life-prolonging methods were significantly higher, while those towards life-shortening methods were lower in Poland compared to Germany. Beside country-specific religiosity differences, a higher acceptance of life-shortening methods by CGs in both countries was associated with a poorer quality of life of CGs and a longer time since diagnosis. It may indicate worse coping with disease progression and weaker adaptation mechanisms in CGs compared to those reported previously in ALS patients [33]. A close resemblance of the CGs' answers to probable patients' attitudes reported by the CGs may indicate that many CGs might have actually expressed their own (culturally shaped) attitudes towards end-of-life methods rather than appropriate proxy decisions. In light of previously reported discrepancies between the presumed opinions of the CGs and of ALS patients themselves [6], a greater focus should be placed on thorough discussions of future treatment options with ALS patients in the presence of their CGs. This attitude might support future surrogate decisions in concordance with the patient's authentic will.

Acknowledgements

We sincerely thank the carers and patients for contributing to this study.

It was an EU Joint Programme – Neurodegenerative Disease Research (JPND) project (“NEEDSInALS” 01ED1405). The project is supported by the following organizations under the aegis of JPND – www.jpnd.eu incl. Germany, Bundesministerium für Bildung und Forschung (BMBF, FKZ), Sweden, Vetenskapsrådet Sverige and Poland, National Center for Research and Development (NCBR). This study was also supported by Deutsche Forschungsgemeinschaft (LU 336 / 13-2 BI 195 / 54-2), BMBF (# 01GM1103A), ERA-NET-E-Rare 3/IV/MAX-OMOD/11/2020, the Deutsches Zentrum für Neurodegenerative Erkrankungen (DZNE) and the Kompetenznetzwerk Präventivmedizin Baden-Württemberg

(K.N.K.B.008.04). The funding sources played no role in the preparation of this manuscript.

Disclosure

The authors report no conflict of interest.

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