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ORIGINAL COMMUNICATION

Experience matters: neurologists' perspectives on ALS patients' well-being

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Abstract Despite the fatal outcome and progressive loss of physical functioning in amyotrophic lateral sclerosis (ALS), many patients maintain contentment in life. It has been shown that non-professionals tend to underestimate the well-being of patients with ALS, but professionals' perspective is yet to be studied. In total, 105 neurologists with varying degrees of experience with ALS were included in an anonymous survey. They were asked to estimate the quality of life and depressiveness of ALS patients with artificial ventilation and nutrition. Physicians' estimations were compared with previously reported subjective ratings of ALS patients with life-prolonging measures. Neurologists with significant experience on ALS and palliative care were able to accurately estimate depressiveness and quality of life of ALS patients with life-prolonging measures. Less experienced neurologists' estimation differed more from patients' reports. Of all life-prolonging measures neurologists regarded invasive ventilation as the measure associated with lowest quality of life and highest depressiveness of the patients. Experienced neurologists as well as neurologists with experience in palliative care are able to better empathize with patients with a fatal illness such as ALS and support important decision processes.

Keywords Amyotrophic lateral sclerosis (ALS) · Depression · Quality of life · Life-prolonging measures · Physician

Introduction

In amyotrophic lateral sclerosis (ALS), the employment of non-invasive (NIV) or invasive (IV) artificial ventilation and/or nutrition via a feeding tube (percutaneous endoscopic gastrostomy, PEG) may prolong patients' survival and improve their quality of life [1–3]. In practice, decisions regarding therapeutic options are not solely made by the patient, but are influenced by the social environment [4, 5]. Whether the influence acts pro or con for certain measures strongly depends on the person's image of well-being of patients living with such measures [6].

Neurologists who have medical expertise and are acquainted with life-prolonging measures have a crucial role in the patient's decision-making process [7]. However, similar to non-professionals, the physician's appraisal of therapeutic options might be framed according to his/her own attitudes [8] and perception of the patient's psychological well-being [6, 9, 10]. This, in turn, may influence the way physicians discuss the available therapeutic options with the patient [6, 11].

Despite the fatal outcome and progressive loss of physical functioning in ALS [12], many patients maintain contentment in life [13]. It has been shown that non-professionals have a negatively biased image of the well-being of patients with ALS [14, 15]. Even close relatives tend to underestimate ALS patients' quality of life and overestimate their depressiveness [16, 17]. It is not yet clear how medical professionals reflect the well-being of the patients with ALS.

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Health professionals are important sources to drive improvement in ALS care [18], but there is a need for quantitative measures to further support this. Therefore, our aim was to study neurologists' perception of the impact of therapeutic interventions on ALS patients' psychological well-being in relation to their professional expertise and to compare it with patients' own reports.

Subjects and methods

Overall, $n = 813$ German neurologists were considered. $N = 449$ neurologists received a paper version of the questionnaire by mail and $n = 364$ a link by e-mail the online version of the questionnaire, generated with the Survey Monkey online survey tool (http://www.surveymonkey.com). Responses were collected between July 2014 and May 2016 from the neurologists of all specialized ALS clinics in Germany and registered neurologists in Southern Germany (states of Saarland, Rhineland-Palatinate, Baden-Wuerttemberg and Bavaria).

In total, $n = 114$ physicians returned the questionnaire (response rate 18% for the paper questionnaires and 11% for the online survey), including $n = 9$ who returned it without answers, claiming to have no experience with ALS. Thus, $n = 105$ neurologists' responses were included in the final analysis (Fig. 1; Table 1).

Out of $n = 813$ neurologists considered, $n = 100$ ($n = 51$ by mail, $n = 49$ by e-mail) were not reached. Out of $n = 114$ who returned the questionnaires, $n = 78$ received it by mail and $n = 36$ online. $N = 9$ returned the questionnaire unanswered due to lack of experience with ALS patients. Reports of $n = 105$ ($n = 30$ online, $n = 75$ regular) neurologists were included in the study.

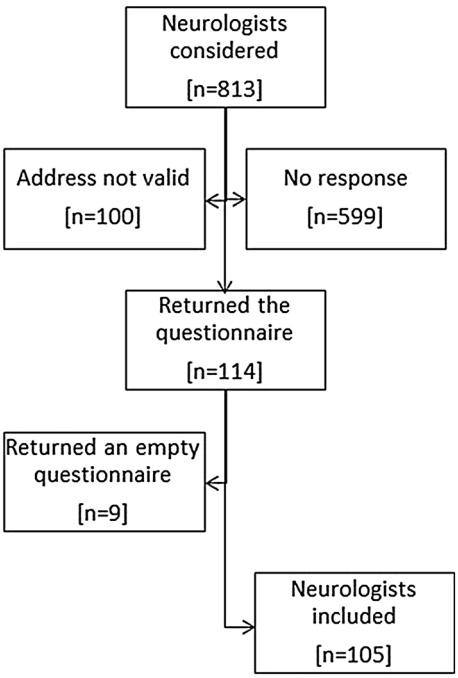

Fig. 1 Neurologists being selected in the study

Table 1 Demographics of the neurologists ($n = 105$) included in the study

Variable		Percentage
Age (mean ± SD)	50.3 ± 12.0	
Gender	68 male	65
	31 female	30
	6 no information	5
Experience with ALS (years)	<1	13
	1–3	61
	4–10	14
	>10	12
ALS patients seen per month	<1	6
	1–5	15
	5–10	14
	>10	65
Completed palliative care training	12 with	11
	93 without	89

SD standard deviation

The 29-item questionnaire was to be filled out anonymously which took about 10 min time. Questions included in the final analysis encompassed items addressing the experience of the physician with ALS and estimation of the level of quality of life and depressiveness of ALS patients with NIV, IV and PEG (additional data are given in Online Resource 1). Because of the need to maintain anonymity, no reminder was possible. The study was approved by the Ethical Committee of the University of Ulm (19/12) and was therefore performed in accordance with the ethical standards laid down in the 1964 Declaration of Helsinki and its later amendments. To maintain anonymity, no written consent was requested. All participating physicians were provided with an informative briefing.

Clinical experience

The experience of the neurologist was defined as the average number of ALS patients seen per month (with four choices from less than one to more than ten patients) multiplied by the average number of years of experience with ALS patients (with four choices from less than 1 year to more than 10 years). Experience was classified as either low (average number of ALS patients seen in total < median, $n = 32$), moderate (average number of ALS patients seen in total = median, $n = 51$) and high experience on ALS (average number of ALS patients seen in

total > median, $n = 22$). Additionally, physicians were asked whether they had completed specific palliative care training according to the German Medical Association (Bundesärztekammer).

Well-being

Neurologists were requested to estimate the psychological well-being (depressiveness and quality of life) of ALS patients with NIV, IV and PEG. Depressiveness was assessed on a Likert scale ranging from 0 to 10 and quality of life was assessed according to an adapted version of the anamnestic comparative self-assessment (ACSA) [19] on a Likert scale ranging from −5 to 5.

Additionally, the data of $N = 52$ patients diagnosed with probable or definite ALS according to the revised El Escorial criteria [20] were included in the study (Table 2). These patients having NIV, IV and/or PEG had been interviewed in a previous study on their subjective well-being [21]. Neurologists' reports on the quality of life and depressiveness of ALS patients with the aforementioned life-prolonging measures were compared with the reports of ALS patients' own reports published earlier [21]. Patients' depression was measured with the "Allgemeine Depressionskala" (ADSK) [22], the German version of the Center for Epidemiologic Studies Depression Scale, CES-D, (range: 0–60; threshold >16). To compare the depressiveness scores estimated by the neurologists with patients' reports, neurologists' estimation of patients' depressiveness on a scale of 0–10 was adjusted to the range of the ADSK (0–60) that was used for the patients' subjective rating for depression. Patients' quality of life was assessed with the anamnestic comparative self-assessment (ACSA [19], ranging from −5 for as bad as possible to +5 for as good as possible).

Statistics

Statistical analyses were performed using the Statistical Package for Social Sciences (SPSS, IBM, version 21.0). Mean and standard deviations or alternatively medians are given. The normality of the data was tested with the Kolmogorov–Smirnov test and either parametric or non-parametric statistical tests were applied accordingly. For between group comparisons (patients vs. neurologists; neurologists with palliative care training vs. neurologists without palliative care training vs. patients; neurologists with high experience vs. patients; neurologists with low experience vs. patients) non-parametric Mann–Whitney U tests were conducted. Pearson correlation was conducted for the association of estimation of patients' well-being (the depressiveness and quality of life) and total number of ALS patients seen on average and Spearman correlation for the association of estimation of patients' well-being and years of experience or engagement in ALS research. Linear regression analysis was applied for association between the average number of patients seen per month and estimation of the patients' depressiveness and quality of life. For the analysis of neurologists' reports, a threshold of $p < 0.05$ was adopted for statistical significance. To minimize possible false interpretations owing to small patient groups, a conservative threshold of $p < 0.01$ was chosen for statistical significance for comparisons between neurologists' reports and patients' subjective ratings.

Results

Overall, neurologists (with varying degrees of experience) estimated higher depressiveness scores for patients with any life-prolonging measure (with NIV: $U = 488.5$,

Table 2 Demographics of the ALS patients from Lulé et al. [21]

Variable	Patients with NIV ($n = 29$) mean ± SD	Patients with IV ($n = 6$) mean ± SD	Patients with PEG ($n = 17$) mean ± SD
Age	58.07 ± 12.09	51.50 ± 7.50	53.53 ± 13.43
Gender	8 female	3 female	7 female
	21 male	3 male	10 male
ALS-FRS-R	24.59 ± 10.09	10.50 ± 9.81	15.06 ± 11.48
Time since onset [months]	49 ± 41	104 ± 67	66 ± 53
Progression [loss of ALS-FRS scores per month]	0.9 ± 0.9	0.5 ± 0.3	0.7 ± 0.4
Site of onset	21 spinal	4 spinal	9 spinal
	7 bulbar	2 bulbar	8 bulbar
	1 n/a		

ALS-FRS-R ALS functional rating scale revised form [23]. *NIV* non-invasive ventilation, *IV* invasive ventilation, *PEG* percutaneous endoscopic gastrostomy, *SD* standard deviation

$p < 0.001$; with IV: $U = 39$, $p < 0.001$; with PEG: $U = 380.5$, $p < 0.001$) and lower quality of life scores for patients with IV ($U = 69$, $p = 0.002$), but not with NIV ($U = 1372$, $p > 0.01$) or PEG ($U = 713$, $p > 0.01$) than what the patients had reported previously [21].

However, neurologists with high experience estimated quality of life close to patients' subjective estimation for all measures as no statistically significant difference was seen between them (NIV: $U = 236$, $p > 0.01$; IV: $U = 39$, $p > 0.01$; PEG: $U = 149.5$, $p > 0.01$). Similarly, they estimated depression with PEG ($U = 109$, $p > 0.01$) close to the patients' subjective estimation. High experienced neurologists estimated depressiveness significantly higher for the patients with NIV ($U = 152$, $p = 0.001$) and IV ($U = 10$, $p = 0.002$). In contrast to this, less experienced neurologists' estimation on depressiveness was higher than the reports of the patients with all measures (NIV: $U = 106.5$, $p < 0.001$; IV: $U = 7$, $p < 0.001$; PEG: $U = 99.5$, $p = 0.001$). Neurologists with low experience also estimated quality of life lower for the patients with IV ($U = 12.5$, $p = 0.001$), but not for the patients with NIV ($U = 421.5$, $p > 0.01$) and PEG ($U = 189.5$, $p > 0.01$) (Fig. 2).

Of all life-prolonging measures, neurologists regarded IV as the measure associated with higher depression and lower quality of life than NIV and PEG. Neurologists estimated depressiveness higher (NIV vs. IV: $U = 3350.5$, $p < 0.001$; PEG vs. IV: $U = 3527.5$, $p < 0.001$) and quality of life lower (NIV vs. IV: $U = 2508.5$, $p < 0.001$; PEG vs. IV: $U = 2531.5$, $p < 0.001$) for patients with IV than for patients with NIV and PEG. Quality of life ($U = 5027$, $p > 0.05$) and depressiveness ($U = 5093.5$, $p > 0.05$) of patients with NIV and PEG were estimated in a similar range.

Negative Pearson correlation was seen between the experience of the neurologist (patients seen in total) and his/her estimation of depressiveness of patients with NIV ($r = -0.217$, $p = 0.028$) and with IV ($r = -0.241$, $p = 0.017$) but not with PEG ($r = -0.088$, $p > 0.05$), and positive correlation between experience of neurologists (patients seen in total) and estimation of quality of life of patients with IV ($r = 0.263$, $p = 0.009$) but not with NIV ($r = 0.167$, $p > 0.05$) or PEG ($r = 0.185$, $p > 0.05$).

Linear regression analysis revealed that the more ALS patients were seen by a neurologist monthly, the lower did the neurologist estimate the depressiveness of patients (with NIV: $R^2 = 0.051$, $\beta = -0.226$, $p = 0.022$; with IV: $R^2 = 0.109$, $\beta = -0.33$, $p = 0.001$; with PEG: $R^2 = 0.051$, $\beta = -0.225$, $p = 0.023$) and the higher did he/she estimate the quality of life of the patients (with NIV: $R^2 = 0.053$, $\beta = 0.229$, $p = 0.021$; with IV: $R^2 = 0.057$, $\beta = 0.239$, $p = 0.018$; with PEG: $R^2 = 0.113$, $\beta = 0.337$, $p = 0.001$) with any life-prolonging measure.

Neurologists with completed palliative care training estimated higher quality of life for patients with NIV ($U = 244.5$, $p = 0.006$) and PEG ($U = 243$, $p = 0.005$) but not with IV ($U = 259.5$, $p > 0.01$) than the neurologists without palliative care training. Neurologists with and without palliative care training did not significantly differ in their estimation of depressiveness (NIV: $U = 310.5$, $p > 0.01$; IV: $U = 301.5$, $p > 0.01$; PEG: $U = 434$, $p > 0.01$). When comparing patients [21] and the neurologists with palliative care training, no statistically significant difference was seen in rating of depressiveness (NIV: $U = 138.5$, $p > 0.01$; IV: $U = 16$, $p > 0.01$; PEG: $U = 69$, $p > 0.01$) or quality of life (NIV: $U = 100.5$, $p > 0.01$; IV: $U = 10.5$, $p > 0.01$; PEG: $U = 73$, $p > 0.01$). However, neurologists without palliative care training differed in the estimation of quality of life with patients with IV ($U = 58.5$, $p = 0.001$) but not with NIV ($U = 1271.5$, $p > 0.01$) or PEG ($U = 350$, $p > 0.01$) and in the estimation of depressiveness with patients with all measures (NIV: $U = 350$, $p < 0.001$; IV: $U = 23$, $p < 0.001$; PEG: $U = 311.5$, $p < 0.001$) (Fig. 3).

Additionally, Spearman correlation revealed that the more actively neurologists participated in ALS research, the lower did they estimate patients' depressiveness with all measures (NIV: $r = -0.271$, $p = 0.006$; IV: $r = -0.253$, $p = 0.012$; PEG: $r = -0.330$, $p = 0.001$) and the higher the quality of life for the patients with PEG ($r = 0.237$, $p = 0.017$). Neurologists' engagement in ALS research was not correlated with their estimation of quality of life of the patients with NIV ($r = 0.085$, $p > 0.05$) or IV ($r = 0.061$, $p > 0.05$).

Discussion

In the current study, experienced neurologists accurately estimated psychological well-being in ALS. In terms of rating depressiveness and quality of life of patients with NIV, IV or PEG, neurologists with high experience (high number of ALS patients seen in total) mostly did not differ from the subjective rating of ALS patients with life-prolonging measures. Similarly, the more neurologists were engaged in ALS research, the closer they estimated patients' well-being. Therefore, our results suggest that with increasing patient contacts besides increased general medical expertise, physicians also refine the ability to better recognize patients' affective state and empathize with their life with certain therapeutic measures.

It has been suggested that palliative care training may improve the knowledge, communication, confidence and symptom management of the physician [24] and that earlier exposure to death [25] and experience on palliative care may decrease physician's anxiety [26] and negative

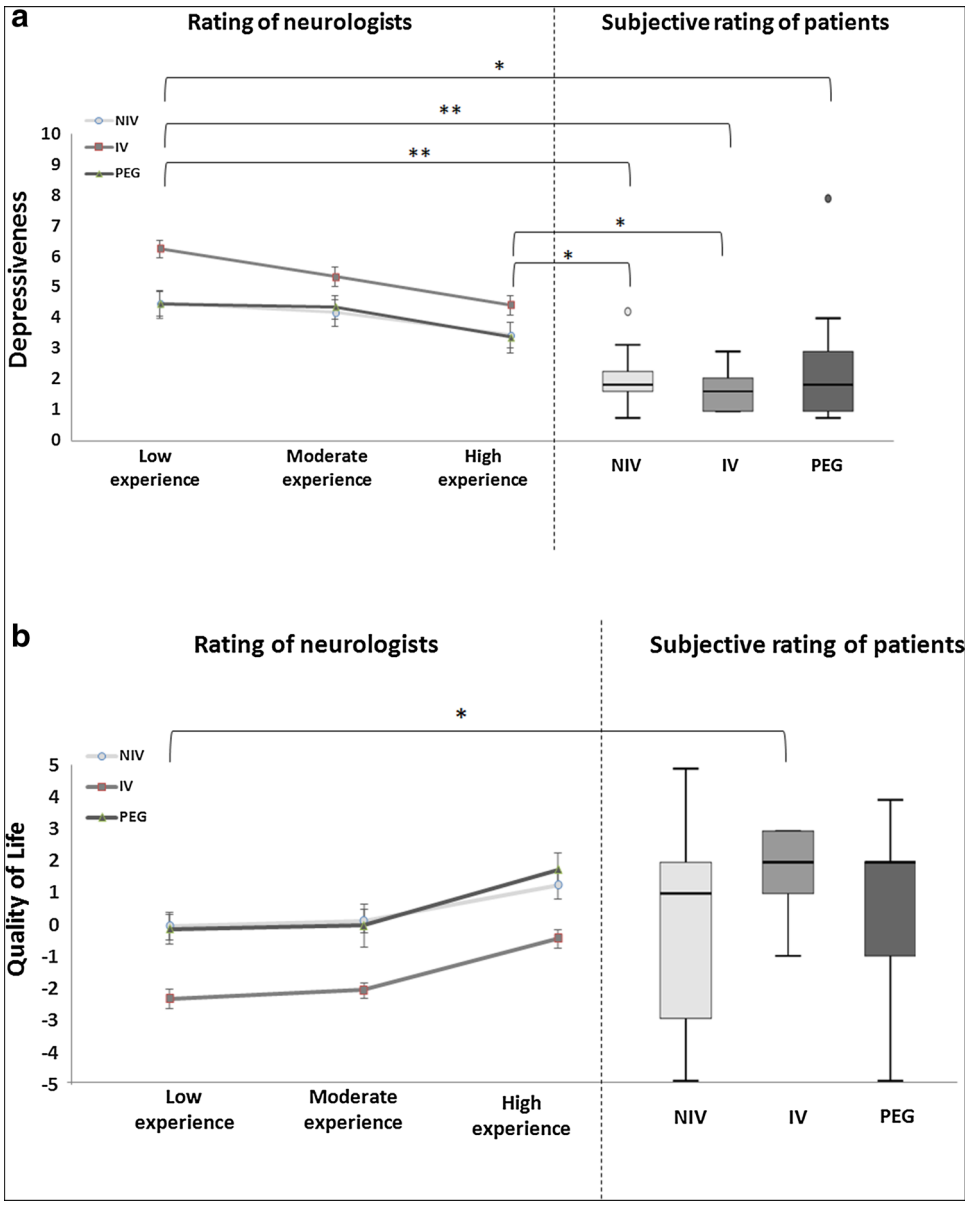

Fig. 2 Rating of depressiveness (**a**) and the quality of life (**b**) of ALS patients by neurologists with varying degrees of experience compared with patients' own reports Depressiveness was estimated on a Likert scale ranging from 0 to 10 and the quality of life on a Likert scale ranging from −5 to 5. The line charts (left figure in a and b) show change in neurologists' estimations of patients' well-being in relation to their experience. Box plots (right figure of a and b) show patients' [21] subjective rating on their depressiveness and quality of life. Medians, first and third quartiles, range and the outliers are shown. * indicates statistical significance with $p < 0.01$ and ** with $p < 0.001$ between patients and neurologists (with either low or high experience) in Mann–Whitney U test. Low experience: average number of ALS patients seen by the neurologist < median; moderate experience: average number of ALS patients seen by neurologists = median; high experience: average number of ALS patients seen by neurologists > median. NIV = non-invasive ventilation, IV = invasive ventilation, PEG = percutaneous endoscopic gastrostomy

attitudes toward living and dying with fatal illness [27]. Similarly, our results show that neurologists with completed palliative care training rated lower depressiveness and higher quality of life for ALS patients with life-prolonging measures, thus being close to the subjective rating of patients. Therefore, also our results support the importance of training and knowledge on palliative care when treating patients with fatal illnesses.

More difference was seen between rating of less experienced neurologists and subjective rating of patients. The depressiveness of patients with life-prolonging measures was rated higher and the quality or life lower by the neurologists with low experience in ALS. Also, rating of neurologists without completed palliative care training mostly differed from the patients. Discrepancy between the rating of patients and neurologists was present especially for the well-being of patients with IV, which neurologists also overall rated more negatively than PEG and NIV. Similarly, it has been shown that IV might be associated with negative attitudes among some physicians [28, 29]. Additionally, IV is a less desired option also among many ALS patients and caregivers who do not have experience with it [21, 30–32].

There might be some reasons given to the rather negative associations related to IV. Besides the facts that IV is an invasive measure [28] and might increase caregiver burden [29, 33] as well as dependency of the patient [34], rareness of IV might contribute to the associations related to the measure. Physicians see relatively rarely patients with IV, as it is not as commonly employed as NIV or PEG

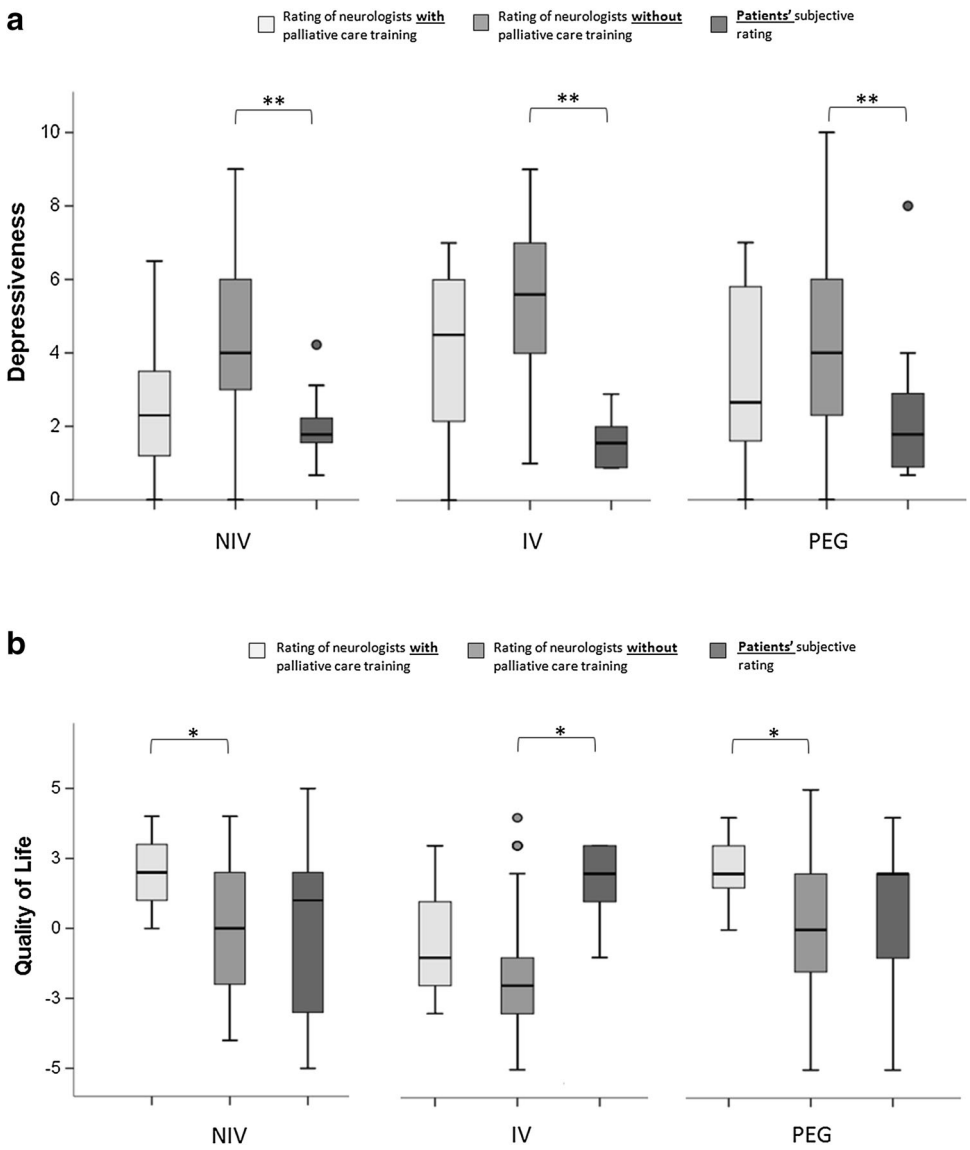

Fig. 3 Estimation of patients' depressiveness (**a**) and the quality of life (**b**) by neurologists with and without completed palliative care training compared to patients' [21] subjective rating Depressiveness was estimated on a Likert scale ranging from 0 to 10 and the quality of life on a Likert scale ranging from −5 to 5. Box plots show the comparison of the neurologists' (with completed palliative care training: $n = 12$; without completed palliative care training: $n = 93$) estimation on patients' well-being as well as patients' [21] own reports. Medians, first and third quartiles, range and the outliers are shown. * Indicates the statistical significance with $p < 0.01$ and ** with $p < 0.001$ in Mann–Whitney U test. NIV = non-invasive ventilation, IV = invasive ventilation, PEG = percutaneous endoscopic gastrostomy

especially in some Western countries [31, 35, 36]. However, again both experience in palliative care and experience in dealing with ALS patients were strongly associated with neurologist's more positive rating of well-being with IV. Thus, experienced neurologists' rating was closer to the subjective rating of patients [21] and earlier findings, showing that patients with IV have rather good quality of life and low depressiveness [37] and do not significantly differ from patients with other life-prolonging measures [21, 33]. Accordingly, it has also been frequently shown that satisfaction in life does not necessarily decline with reduced physical functioning [13, 16, 30, 32, 38].

Finally, it can be speculated whether high empathy on the physician's side for the patient's well-being is even favorable for every medical encounter, as a certain emotional distance might be preferable in a professional relationship. Also, our results show that instead of complete empathy with patients' condition, neurologists, and especially neurologists with significant experience in ALS, are able to empathize with patient's well-being as seen in the accurate estimation of patients' depressiveness and quality of life.

Limitations

First, in such a diverse disease as ALS, only very rough estimations can be made as patients' progression as well as their psychological well-being varies greatly between individuals, which is known especially among experienced neurologists. Ideally, pairwise comparisons allowing the neurologists to evaluate each patient individually and comparing this estimation with patient's subjective estimation would have been done. However, owing to the

anonymity, such a study design was not possible. Moreover, a larger patient cohort would be preferable but due to the rareness of employment of invasive ventilation in Germany [17, 21], it is hard to accomplish. Furthermore, to strengthen the reliability of the conclusions made of the patient–neurologist comparisons, a conservative level of $p < 0.01$ was chosen for statistical significance.

Additionally, a direct comparison of different depression scales filled by the neurologists and patients might be problematic. Patients filled out a proper validated questionnaire with 15 items giving a depression score between 0 and 60, whereas neurologists were only asked to estimate the depressiveness on a visual analog scale between 0 and 10. However, time-consuming questionnaires are not desirable in physician surveys and we believe that comparison and a suggestive conclusion are justifiable through the adjustment of the scales and strict statistical analyses.

Finally, the results of our study may be influenced by a possible selection bias owing to the relatively low response rate which, however, has been reported also earlier [39]. Furthermore, the environment where the patient is met might partly account for the estimation difference between patients and neurologists. In contrast to the caregivers who see the patient at home, physicians meet patients in a clinical setting (in- and outpatient clinics). This should be taken into account as it has been suggested earlier that diverging views on the provision of life-prolonging measures of medical health-care professionals and allied health-care professionals might be partly due to the context where they see ALS patients, either in the clinic or in their home [4]. Similarly, the patient cohort from Lulé et al. [21] used for comparison was not interviewed in the clinic but in the patients' home, where patients might generally be more satisfied with their condition. Therefore, further studies with larger patient groups and reduced biasing environmental factors are needed.

Conclusion

Our results emphasize the importance of neurologist's experience in ALS care, as significant experience was strongly associated with better estimation of patients' well-being. Additionally, not just experience in ALS, but also experience in palliative care and engagement in ALS research, might refine the ability to echo psychological well-being of patients with ALS, thus possibly improving the quality of the therapeutic relationship. However, flawed judgment of the well-being of patients with disability (disability paradox) [15], which was previously reported for non-professionals such as caregivers [17], might also be present in some physicians with low experience in ALS.

As decisions regarding therapies usually have to be backed by family and health professionals [28, 40, 41], it is crucial to consider any possible personal bias in the perception of patient's psychological well-being [28]. Therefore, patients with a rare disease such as ALS would be ideally treated by experienced neurologists, when they might receive both more advanced medical expertise and better understanding of their condition with regard to psychological well-being. This is particularly true for end-of-life decisions for which empathy of the professional environment is mandatory.

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Compliance with ethical standards

Conflicts of interest The authors declare that they have no conflict of interest.

Ethical standard The study was approved by the ethics committees of the University of Ulm and the University of Berlin and has, therefore, been performed in accordance with the ethical standards laid down in the 1964 Declaration of Helsinki and its later amendments. All participants gave informed consent prior to their inclusion in the study.

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