Palliative and Supportive Care

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Quality of life among relatives of patients with amyotrophic lateral sclerosis: A prospective and longitudinal study

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Original Article

Cite this article: Larsson BJ, Ozanne A, Nordin K, Nygren I (2022). Quality of life among relatives of patients with amyotrophic lateral sclerosis: A prospective and longitudinal study. *Palliative and Supportive Care* **20**, 203–211. https://doi.org/10.1017/S1478951521000778

Received: 24 October 2020
Revised: 11 May 2021
Accepted: 30 May 2021

Key words:
ALS; Anxiety; Depression; Quality of life; Relatives

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Abstract

Objective. Relatives are often central in caring for patients with amyotrophic lateral sclerosis (ALS), involving considerable physical, emotional, and social challenges. The aim of this study was to describe individual quality of life (iQoL) among relatives of patients with ALS, from diagnosis through disease progression.

Method. A total of 31 relatives were included. Data collection was performed at five time points: 1–3 months after their relatives had been diagnosed with ALS and every 6 months for 2 years. Quality of life was determined using the Schedule of Evaluation of Individual Quality of Life — Direct Weighting (SEIQoL-DW), emotional distress with the Hospital Anxiety and Depression Scale (HADS), and the illness severity of the patients was determined with the Amyotrophic Lateral Sclerosis Functional Rating Scale (ALS FRS-R).

Results. The SEIQoL-DW involves participants nominating the important life areas. The most nominated areas were family, friends, health, and leisure. Although most relatives had overall good and stable iQoL, several had scores indicating poor iQoL on some occasions during the disease trajectory. The relatives' iQoL correlated with emotional well-being and the patient's physical function at different time points.

Significant of result. Social relations, emotional well-being, and rapid decline in the patient's physical function influence the relatives' iQoL. Measuring both emotional well-being and iQoL, with a focus on the relatives' own descriptions of perceived iQoL and those factors contributing to their iQoL during the disease trajectory may improve the possibility of identifying and supporting those relatives with poor iQoL.

Introduction

Amyotrophic lateral sclerosis (ALS) is a neurodegenerative disorder affecting the motor neurons controlling the voluntary muscles, leading to the gradual development of muscle weakness and atrophy (Mitchell and Borasio, 2007). Relatives have an important role in caring for a patient with ALS, which involves considerable physical, emotional, and social challenges. Relatives often dedicate many hours every day to help the patient with daily activities (Krivickas et al., 1997; Aoun et al., 2012). The burden of care increases for the relatives during disease progression; moreover, due to symptoms and progression of the disease, the relatives' lives become focused around their home and the patient, leading to a restricted social life and decreased activities (Trail et al., 2003; Hughes et al., 2005).

Quality of life (QoL) may be difficult to define, but for most individuals, QoL is associated with life satisfaction and well-being. Quality of life is subjective and multidimensional; moreover, different individuals value different aspects of life, and the meaning of QoL means different things for different individuals (Carr et al., 2001). The World Health Organization's (WHO) definition of QoL is "an individual's perception of their position in life in the context of the culture and value systems in which they live, and in relation to their goals, expectations, standards and concerns" [The World Health Organization Quality of Life assessment (WHOQOL): position paper from the World Health Organization, 1995]. Despite the severity of the disease, several studies have shown that most patients with ALS have a relatively good QoL (Neudert et al., 2001; Trail et al., 2003; Chio et al., 2004; Fegg et al., 2005; Nygren and Askmark, 2006; Gauthier et al., 2007; Roach et al., 2009; Jakobsson Larsson et al., 2017). When comparing QoL in patients with ALS and their relatives, it has been found that relatives estimated their QoL as poorer than the patients (Bromberg and Forshew, 2002; Olsson et al., 2010), while other studies found no difference between how patients and their relatives rated their QoL (Trail et al., 2003; Lo Coco et al., 2005; Gauthier et al., 2007).

Few longitudinal and prospective studies have been conducted with relatives of patients with ALS with the perspective to describe individual QoL (iQoL; Gauthier et al., 2007; Roach et al., 2009; Olsson et al., 2010), and none, to our knowledge, with relatives of newly diagnosed patients with ALS and over time. This present study aims to describe iQoL from diagnosis and throughout disease progression among relatives of patients with ALS and to evaluate if iQoL correlates with the patient's physical function or the relatives' own psychological well-being.

Methods

This was a prospective, longitudinal, and descriptive study.

Relatives of patients with probable or definite ALS, according to the El-Escorial WFN revised criteria (Brooks et al., 2000), who were treated by the ALS teams at three separate hospitals in Sweden, were asked to participate in the study. Inclusion criteria were being older than 18 years of age and understanding and being able to express themselves in the Swedish language. This is part of a longitudinal study with 36 patients who had been newly diagnosed with ALS (Jakobsson Larsson et al., 2017); three of the patients could not identify a relative to participate in the study. A total of 33 relatives were included at the start of the study.

Data collection was performed using questionnaires and a semi-structured interview instrument. The data collection started 1–3 months after the patient was diagnosed with ALS and continued for a period of 2 years with the purpose of evaluating iQoL over time. After having given written informed consent, the relatives answered the questionnaires and the semi-structured interview were performed by the first author (BJL) during home visits or by telephone. The patient's physical function was evaluated at the same time point as the interviews.

To evaluate iQoL, the Schedule of Evaluation of Individual Quality of Life — Direct Weighting (SEIQoL-DW) was used. This is a semi-structured interview instrument to evaluate a person's iQoL by letting the respondents nominate the five most important areas (cues) of their life at present, determine the level of functioning for each cue on a visual analogue scale from worst possible (0) to best possible (100). Finally, the respondent determined the relative importance of each of these cues using a pie chart with sections, where he or she could adjust the size to reflect the relative percentage importance of the different cues. By multiplying the cue level by the cue weight for each cue and summing the values for each of the five cues, a SEIQoL-DW index score is calculated. This index can range from 0 (lowest QoL) to 100 (highest QoL) (Hickey et al., 1996). In the present study, a modified version of the SEIQoL-DW was used (Wettergren et al., 2003). The respondents were asked to nominate the five most important areas, both good and bad, for their overall quality of life. The definition of each area (cue) given by the respondents was documented directly on a cue definition record form. Thereafter, each area was rated on a 7-point scale regarding how well it functioned: "As bad as it could possibly be" (scored 1) and "As good as it could possibly be" (scored 7). In the original version of SEIQoL-DW, a direct weighting procedure is conducted to determine the importance of each cue. Earlier studies have found that the weighting procedure does not have an impact on the total index (Wettergren et al., 2005, 2008). Therefore, the overall individual QoL score (SEIQoL index) was calculated by taking the sum of the ratings divided by the number of nominated areas. Finally, the relatives self-rated how they experienced their overall QoL based on the areas they nominated themselves for and how they rated each area (SR-QoL). By describing and documenting each cue nominated by the respondent, a better understanding and a clinical application will be achieved.

The Hospital Anxiety and Depression Scale (HADS) was used to evaluate the relatives' emotional well-being. This questionnaire consists of two subscales: 7 items for anxiety (HADSa) and 7 items for depression (HADSd). The scale measures the presence and severity of symptoms of anxiety and depression in the past week. Two cut-off scores have been suggested, 8–10 = doubtful cases and ≥11 = cases (Zigmond and Snaith, 1983).

The patient's physical function was measured using the revised Amyotrophic Lateral Sclerosis Functional Rating Scale (ALS FRS-R). This functional rating scale consists of 12 questions to assess the patients' levels of functioning: bulbar function, fine- and gross motor tasks and respiratory function. Each function is graded from 0 (total lost) to 4 (normal function); a lower score indicates more disability (Cedarbaum et al., 1999).

Analysis

Descriptive statistics were used to describe the demographic and clinical characteristics of the participants. For each time point, the results are based on the subgroup for patients who have survived up to that time point. The answers to the questions in the SEIQoL-DW were written down during the interview by the first author (BJL). The written answers were grouped into areas based upon the domain "the cue primarily belonged to" by two of the authors (BJL, AO). The content in each area was described qualitatively. A mixed model, with time as a continuous fixed variable with the corresponding 95% confidence interval, was used for the development of the SEIQoL-DW index and the other scales over time. Spearman's rho was used to analyze the correlation between the SEIQoL-DW, HADS, and ALS FRS-R. SPSS, version 17 (Chicago, IL, USA) was used for statistical analysis. The significance level was $p \leq 0.01$ since the p-value was not adjusted for the number of tests. All tests were two-tailed. The number of dropouts for reason other than death of patients is small ($n = 2$) and thus taking into account dropouts due to other reasons was deemed not necessary.

Results

A total of 33 relatives were included, 24 women and 9 men. Of those, 2 relatives were missed at time point 1 (TP1) but were included at time point 2 (TP2). The sample was divided into five groups, depending on time in the study; group 1 includes relatives that only participated at the first assessment (i.e., 1–3 months after diagnosis), and group 5 includes only relatives that participated at all five assessments. Two relatives dropped out on their own initiative due to psychological distress, and the other dropouts were due to the death of patients. The characteristics of the group are shown in Table 1.

Areas of importance for the iQoL

In total, 528 cues were nominated by the relatives. These cues were grouped into 14 areas based on the cue label and the relatives' description of each cue. Thirty relatives nominated five cues at the different assessments, one relative nominated only one cue at the second assessment, another relative nominated four cues at the third assessment and yet another relative nominated three cues at all five assessments. The most nominated area of importance was "family" at all five time points (Table 2). This area included different family members and was

https://doi.org/10.1017/S1478951521000778 Published online by Cambridge University Press

Table 1. Socio-demographic characteristics

Characteristics upon entry to the study	Relatives ($n = 33$)	Group 1 ($n = 3$)	Group 2 ($n = 7$)	Group 3 ($n = 9$)	Group 4 ($n = 4$)	Group 5 ($n = 10$)
Male (n)	9	0	2	3	1	3
Age mean (SD)	58.1 (18)	–	36.5 (7.8)	74.3 (7.0)	63.0 (–)	54.7 (18.2)
Range	31–81	–	31–42	67–81	63–63	35–71
Female (n)	24	3	5	6	3	7
Age mean (SD)	60.4 (13.9)	67.7 (20.0)	58.2 (15.7)	65.2 (12.5)	58.3 (6.1)	55.7 (14.7)
Range	33–87	47–87	43–78	42–76	53–65	33–68
Education (n)						
Mandatory	10	0	2	5	1	2
High school	10	1	3	1	2	3
University	13	2	2	3	1	5
Relation to patient (n)						
Spouse	25	2	4	7	4	8
Children	6	1	3	0	0	2
Parent	1	0	0	1	0	0
Sibling	1	0	0	1	0	0
Employment status (n)						
Full-time	11	1	3	1	2	4
Part-time	4	0	1	1	1	1
Unemployed	1	0	1	0	0	0
Retired	17	2	2	7	1	5

Table 2. Frequencies and mean scores for indication level for each area at the five time points for the total group

	TP1[a] ($n = 31$)		TP2[b] ($n = 30$)		TP3[c] ($n = 23$)		TP4[d] ($n = 14$)		TP5[e] ($n = 10$)	
	%	CL	%	CL	%	CL	%	CL	%	CL
Family	81	5.3	77	5.0	87	4.9	93	5.4	90	5.5
Friends	48	5.4	47	5.3	52	4.4	57	5.1	50	5.4
Own health	48	3.8	47	3.9	56	4.3	50	4.7	50	4.7
Others' health	35	4.0	47	4.3	39	4.4	21	5.0	30	3.7
Own time	19	4.3	17	3.4	30	3.4	36	3.0	30	3.3
Support			3	6.0	4	1.0	28	3.7	40	4.5
Leisure	42	3.6	40	4.8	39	4.4	57	5.4	60	5.1
Hobbies	35	5.0	20	4.4	43	4.5	43	4.5	10	4.0
Social activities	22	3.7	13	5.4	22	4.0	14	1.7	30	3.0
Living	32	4.8	27	4.9					10	2.0
Work	35	5.5	40	5.1	22	6.0	28	5.5	40	6.0
Finances	19	5.5	20	5.2	9	7.0	7	6.0	10	5.0
Church	10	4.7	7	5.5	4	3.0				
Pets	6	6.0	10	5.7	4	6.0	7	7.0	10	6.0

% = percentage of relatives nominating the different areas (each relative could choose more than one area). CL = cue level, how each area is functioning for the relative at present time, range from 1 (as bad as it could possibly be) to 7 (as good as it could possibly be).
[a]1–3 months after diagnosis.
[b]After 6 months.
[c]After 12 months.
[d]After 18 months.
[e]After 24 months.

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Table 3. Descriptions of the different areas nominated by the relatives as being important for their individual quality of life

Areas	Description of the areas
Family	This area includes spouses, children, grandchildren, siblings, and parents. Concerns relations and support. To have a good and worthy life together and to make sure that the sick relative received a good care and that he/she could be cared for at home. It also includes descriptions of love and happiness as well as the burden of caring including limitations in spending time with other family members and worries of not being able to care for their sick relative.
Friends	Includes friends and neighbours. Friends that understand, to ventilate/talk with and to get an opinion from another perspective. Friends were described as important for both practical and psychological support, but also as a possibility to relax and divert their mind from the "misery of their situation". Friends gave them energy and were of great importance especially when the children lived far away.
Health	Includes own physical and psychological health. *Own physical health* was described as recover from own illness, being healthy and having the physical strength so that one could support others. It also included regular sleep and being physically well. *Own psychological health* includes stress, worries, hope, and being able to feel secure that it will work for the spouse when he/she got out.
Others' health	Includes worries about both physical health and emotional distress among spouses, children, grandchildren, siblings, and parents. Hope that the ALS disease would not worsen, and the sick spouse felt that his/her existence was meaningful.
Own time	The relatives described a feeling of always being needed and being available, feelings of being locked up, and never being able to finish activities without interruption due to the relative needing help. They wanted to be able to decide over their own time, needed time for relaxation and to think about their situation. These moments of having their own time gave them the possibility to divert their thoughts and to collect strength.
Support	Includes both physical and psychological support; contact with social workers, help with the care from health professionals or personal assistance.
Leisure	Includes areas such as sports/outdoor activities like own training/exercise, being out in the nature/forest and gardening. These physical activities had a positive influence on their physical well-being, gave energy, helped them to clear their thoughts, ease the stress, and help them to relax.
Hobbies	Was described as more passive activities, for example, watching TV, reading, painting a picture, writing poetry, and solving crosswords. Both watching TV and reading could be about taking part in things that happen in the world and/or to get excitement. The hobby gave them relaxation and helped them to divert their thoughts.
Social activities	Includes areas such as travel, theater, seeing a movie, or restaurant visits with others. It was about joy and pleasure with others.
Living	Includes both the home, the environment, and housework. Living close to nature and having beautiful surroundings was described as important. The home was described as both a security and a burden. To do housework such as cooking and baking was described as joy for some and for others it was a new experience due to the spouse's illness.
Work	Includes both the relations with colleagues and customers, the work itself as well as being unemployed. The work was described as the normality in their ordinary life; it was the thing that was still the same as before the spouse got ill. Work was viewed as therapeutic, something to occupy their thoughts. The work suited them and gave them an opportunity to think of other things. Some relatives described that they had to wind up the company due to the disease.
Finances	Includes both descriptions of having a satisfying economic situation, but also financial worries about the future due to the sick spouse's inability to work.
Church	Here, the relatives mentioned the church, God, and the community. The church gave support, and the faith in God gave a firm ground to stand on. It also included the social dimension of meeting one's friends at church.
Pets	The dog was described as something to care for and being a companion, helping the relatives to get out. Concerns were expressed about not being able to care for the dog.

described as "relations," getting "help from other family members" but also "providing support to other family members" (Table 3). Other areas that were frequently nominated were friends, health, leisure, and hobbies. The cue level showed that most of the nominated areas were functioning well at different time points (Table 2).

Individual QoL over time

The mean score for the SEIQoL index and the SR-QoL index indicated that most relatives had a relatively good iQoL at the different assessments (Figure 1). However, when looking at each individual, the result showed that 19 relatives had scored <4 on SEIQoL and/or SR-QoL at any of the assessments, which indicated a poor iQoL. Areas that contributed to low QoL were: "family" (i.e., change relations), "own health," "others" health" (i.e., the ALS disease), and "lack of own time" (data not shown). Even though the SEIQoL index for the total group of relatives did not change over time ($p = 0.570$), the mean score for both SEIQoL-DW and SR-QoL indicated a decline in iQoL in group 4 from TP2 to TP4 (Figure 1).

Relatives' emotional well-being and patient's physical function over time

The results showed that several relatives had scores within the cut-off score for doubtful cases or cases on the HADSa (anxiety) and that some relatives had the symptoms of depression (Figure 2). When looking at each individual, the result showed that relatives in group 4 had more symptoms of anxiety and depression compared with the other groups, with score ≥10 on both HADSa and HADSd at TP3 and TP4, respectively. The ALS FRS-R mean score for the patients show an increase in function disabilities over time and that patients of relatives in group 4 had a more rapid decline in physical function between TP3 and TP4 compared with the other groups (Figure 1).

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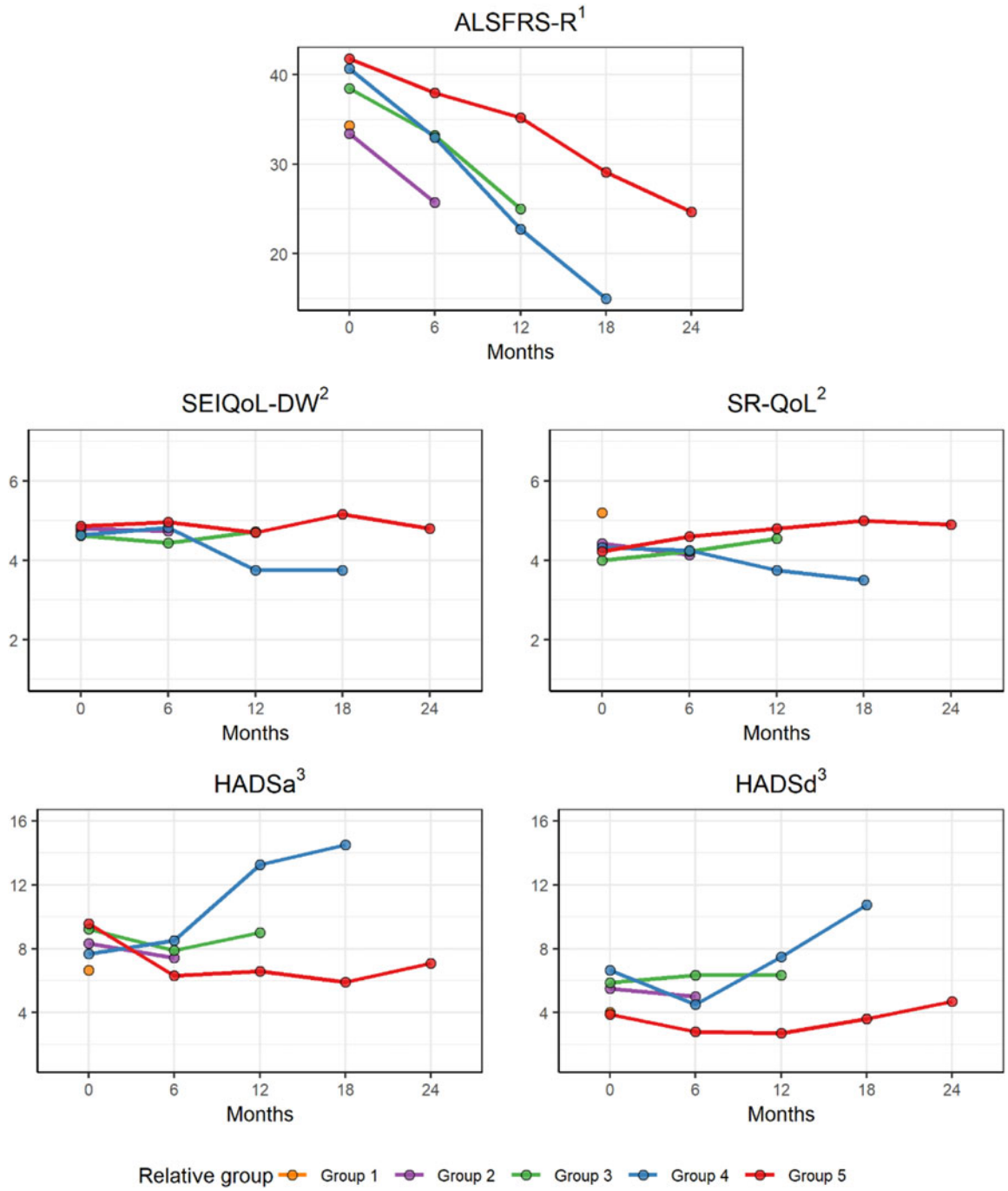

Fig. 1. Mean score for QoL, emotional well-being, and the patient's physical function in the different relative group during the disease progression. [1]range 0–48, a lower score indicates more disabilities, [2]range 0–7, higher score represents a better QoL, [3]range from 0 (no distress) to 21 (maximum distress).

Correlations between iQoL, emotional well-being, and the patient's physical function

Both the SEIQoL index and the SR-QoL index correlated negatively with emotional well-being at several time points and positively with the ALS FRS-R at TP4 (Figure 3). There was also a positive correlation between the SEIQoL index and the SR-QoL index ($r_s = 0.77$).

Discussion

This present study aims to describe iQoL from diagnosis through 2 years of disease progression among relatives of patients with ALS and to evaluate if iQoL correlated with the patient's physical function or the relatives' own emotional well-being. The most nominated QoL areas were: "family" at all time points, but also "friends," "health" (both own health and others' health), and "leisure" were areas of importance for the relatives' iQoL, which is in accordance with previous studies (Bromberg and Forshew, 2002; Lo Coco et al., 2005; Felgoise et al., 2009; Olsson et al., 2010). The results from the semi-structured interview on the descriptions of the areas show the burden that the ALS disease has on the relatives' life due to the increased physical and emotional demands, but also due to limited time for oneself.

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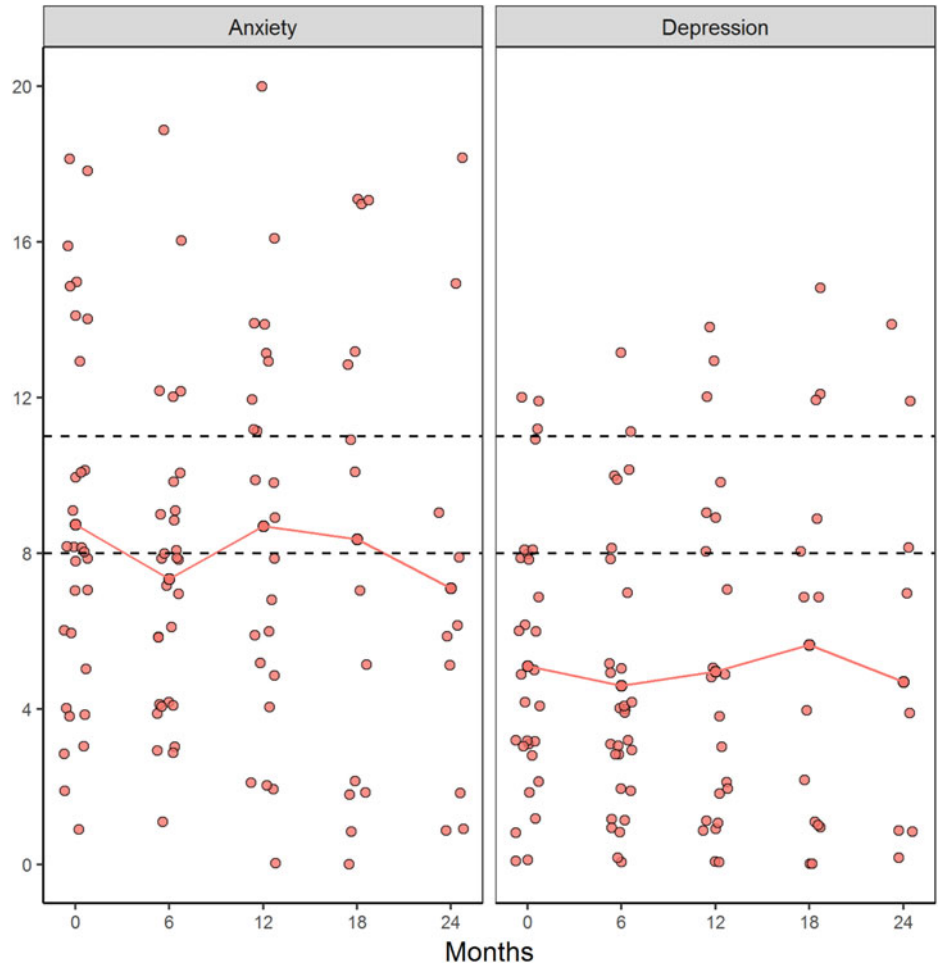

Fig. 2. Doubtful cases (score 8–10) and cases (score ≥11) on HADS two subscales at different time points.

The relatives described worries, stress, the burden of caring, and a need to have time for relaxation and to think about their situation. Although the cue level for each area showed that most areas functioned well, there were variations for some areas between different assessments. The results show that areas such as "own time" and "social activities" had poorer functioning compared with the other areas. To some extent, these areas include a need to be able to decide over one's own time and time outside the home, which most often is limited for relatives involved in the care of patients with ALS. Earlier studies have shown that relatives often rate their QoL as relatively good, despite the impact of the disease on their social life (Galvin et al., 2016) and spending several hours per day to care for the patient (Murphy et al., 2009). When looking at the mean score, the results showed that most relatives rated their iQoL as being relatively good; however, when looking at each participant, relatives in group 4 had a worse iQoL compared with the other groups. The individual descriptions for the cues showed that these relatives with poor iQoL had nominated cues that had a negative impact on their social life (i.e., lack of own time, restrictions in leisure activities, and changed family relations).

Several studies have shown that patients often rated their QoL as good and that it did not change despite the disease progression (Nygren and Askmark, 2006; Gauthier et al., 2007; Jakobsson Larsson et al., 2017). These ratings and the stability of QoL over time may be explained by the response shift theory; the patient reappraised values, internal standards, and conceptualiation of QoL during the disease trajectory (Albrecht and Devlieger, 1999; Schwartz et al., 2007). This revaluation may help them to adjust to their situation, which may explain that most relatives rated their QoL as relatively good despite the situation. When looking at how relatives have estimated the various cues, it seems that most areas that are important also work well, but maybe it is because they are working well that they also become important and not vice versa. The individual description of the cues by relatives with a poor iQoL indicated that they had difficulties in doing this revaluation, leading to difficulties in adjusting to their situation and worse iQoL.

The ALS disease is related to both physical and psychological stress for the relatives during the disease trajectory. Earlier studies have shown that caregiver burden affects the emotional well-being of the caregivers, and that it is not uncommon with symptoms of anxiety (Burke et al., 2015; Galvin et al., 2016). Our results showed that several relatives had scores within the cut-off score for doubtful cases to cases at the HADS and that emotional well-being correlated with both the SEIQoL index and the SR-OoL index. This correlation between emotional well-being and QoL has been found in earlier studies (Murphy et al., 2009; O'Connor and McCabe, 2011; Burke et al., 2015). When looking at the mean score for iQoL index and HADS in group 4 one year after diagnosis, these relatives had more symptoms of anxiety/depression and a poorer iQoL compared with the other groups in the late stage of the disease. Whether it is the emotional well-being that affects the iQoL or if it is the other way around is unclear.

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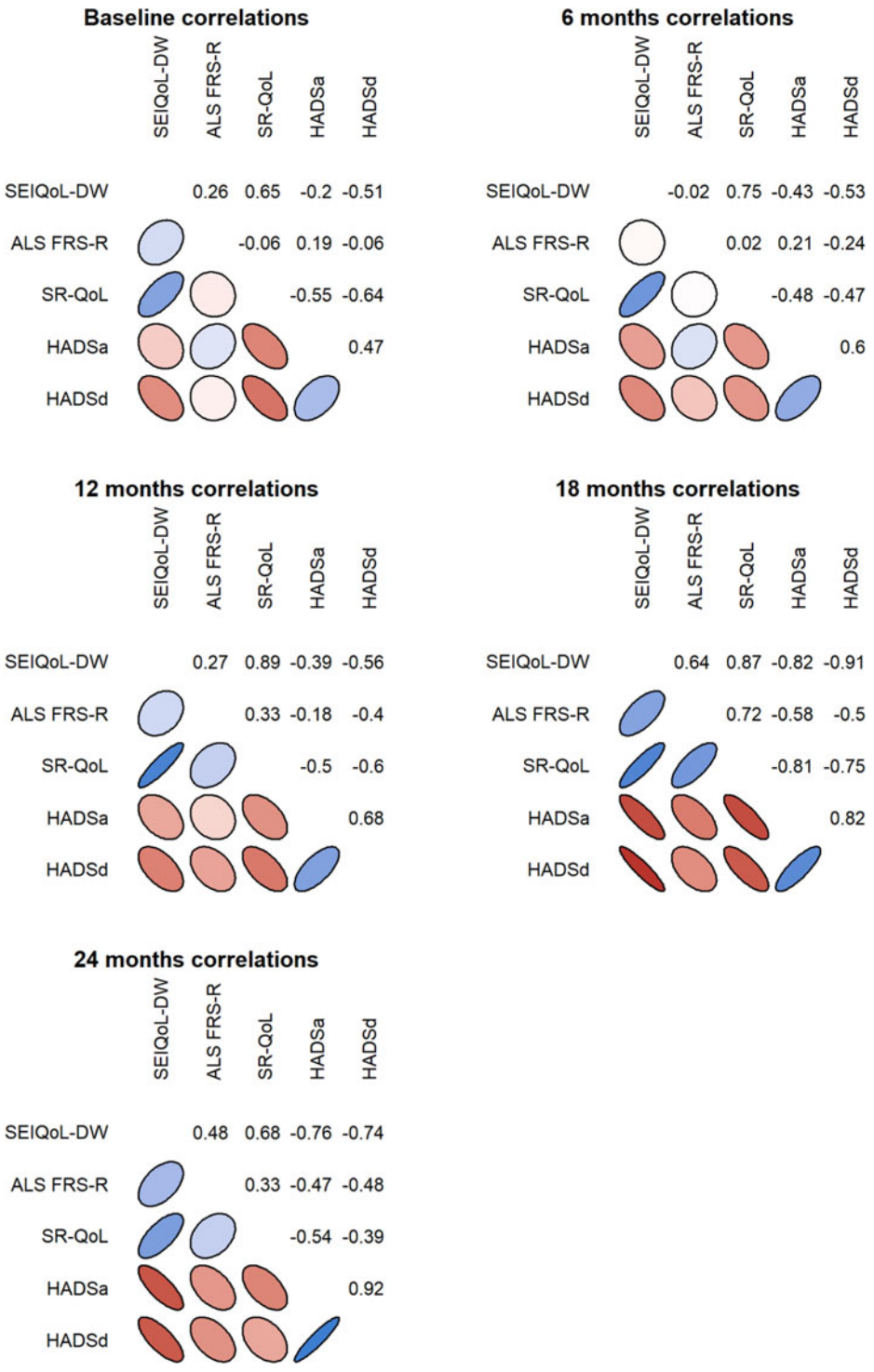

Fig. 3. Correlation between the relatives' QoL, emotional well-being, and the patient's physical function during the disease progression. The shapes color scale range from dark red (strong negative correlation), white (no correlation) and to dark blue (strong positive correlation).The shape of the ellipse illustrates the strength of the correlation where a more narrow ellipse indicates a stronger relationship.

The mean score for ALS FRS-R showed that the patients of relatives in group 4 were severely impaired between TP3 and TP4, which probably increased the burden for these relatives. Even if previous studies have shown that the patient's physical function does not seem to affect relatives' QoL (Lo Coco et al., 2005; Felgoise et al., 2009; Burke et al., 2017), we found a correlation between iQoL and ALS FRS-R at time point 4. This result indicates that a rapid decline in physical function among the patients had a negative impact on the relatives' iQoL. Perhaps the prospective design, with measurements from diagnosis through disease trajectory, could explain the difference between the present study and previous studies.

The strength of this study is the prospective and longitudinal design, with relatives of newly diagnosed patients with ALS, and that there were few dropouts. Felgoise et al. (2009) emphasized that caution must be taken when using SEIQoL-DW of groups, which the results of the present study show. When looking at the mean score of the group, we may have missed those who suffer from poor iQoL, who are the ones that need to be in focus. Felgoise et al. (2009) also suggested that the SEIQoL-DW might

https://doi.org/10.1017/S1478951521000778 Published online by Cambridge University Press

be measuring happiness rather than QoL. The findings in the present study showed a high agreement between the SEIQoL index score and the SR-QoL index score, indicating that the SEIQoL-DW is a useful instrument for assessing iQoL.

The limitation of this study is the small sample size. Another limitation is that we did not use the weighting procedure to estimate the relative importance of the cues. The reason for this was the need to be able to collect data by a telephone interview if we failed to collect data in accordance with home visits. Even though studies have shown that the weighting procedure does not have an impact on the total QoL index (Wettergren et al., 2005, 2008), it may provide valuable information that can be useful in clinical setting when evaluating how to best support the individuals. The participants were asked to nominate areas of importance for their iQoL, both those functioning well or poorly. The results showed that most of the areas nominated were functioning well, but the present study highlights the importance of the individual descriptions of the different cues for a better understanding of how to best support the relatives to maintain a good iQoL.

Even though the present study provided information on areas of importance for relatives' iQoL, more knowledge is needed on specific kinds of support that relatives need and how to best provide this support on an individual basis. Relatives are often central in the care for patients with ALS and dedicate many hours to help and support the patient with daily activities (Krivickas et al., 1997), focusing on the sick relative (i.e., ALS patient) and seldom on their own needs (Larsson et al., 2015). Individual and qualitatively measurements of QoL can be used to identify factors that contribute to the experience of good or bad QoL among relatives of patients with ALS.

In conclusion, our study has shown that social relations with family and friends, but also health and leisure are important for iQoL, with both negative and positive impact. Even though most relatives rated their iQoL as relatively good and stable, the result indicates that a rapid decline in the patients' physical function and emotional distress have a negative impact on iQoL among relatives of patients with ALS. To be able to support and help relatives in their situation, health professionals need to measure both emotional well-being and iQoL, from diagnosis and throughout the disease trajectory, with the focus on the relatives' own descriptions of perceived iQoL and those factors contributing to their iQoL. The evaluation will give the relative and health professionals insight into areas of importance and how they are functioning. This information can help health professionals to provide individual support with the aim to help relatives to obtain a good iQoL and to manage their situation.

Acknowledgments. We thank the relatives of ALS patients who participated in the study. We would also like to thank the Statisticians Marcus Thuresson and Fabian Söderdahl for their support with the statistical analysis.

Funding. The research was funded by Uppsala University, Uppsala University Hospital, Ulla-Carin Lindqvist ALS Research Foundation, and Norrbacka-Eugenia Foundation.

Conflict of interest. None.

References

Albrecht GL and Devlieger PJ (1999) The disability paradox: High quality of life against all odds. *Social Science & Medicine* **48**(8), 977–988.

Aoun SM, Connors SL, Priddis L, *et al.* (2012) Motor Neurone Disease family carers' experiences of caring, palliative care and bereavement: An exploratory qualitative study. *Palliative Medicine* **26**(6), 842–850.

Bromberg MB and Forshew DA (2002) Comparison of instruments addressing quality of life in patients with ALS and their caregivers. *Neurology* **58**(2), 320–322.

Brooks BR, Miller RG, Swash M, *et al.* (2000) El Escorial revisited: Revised criteria for the diagnosis of amyotrophic lateral sclerosis. *Amyotrophic Lateral Sclerosis and Other Motor Neuron Disorders* **1**(5), 293–299.

Burke T, Elamin M, Galvin M, *et al.* (2015) Caregiver burden in amyotrophic lateral sclerosis: A cross-sectional investigation of predictors. *Journal of Neurology* **262**(6), 1526–1532.

Burke T, Galvin M, Pinto-Grau M, *et al.* (2017) Caregivers of patients with amyotrophic lateral sclerosis: Investigating quality of life, caregiver burden, service engagement, and patient survival. *Journal of Neurology* **264**(5), 898–904.

Carr AJ, Gibson B and Robinson PG (2001) Measuring quality of life: Is quality of life determined by expectations or experience? *BMJ* **322**(7296), 1240–1243.

Cedarbaum JM, Stambler N, Malta E, *et al.* (1999) The ALSFRS-R: A revised ALS functional rating scale that incorporates assessments of respiratory function. BDNF ALS Study Group (Phase III). *Journal of the Neurological Sciences* **169**(1–2), 13–21.

Chio A, Gauthier A, Montuschi A, *et al.* (2004) A cross sectional study on determinants of quality of life in ALS. *Journal of Neurology, Neurosurgery, and Psychiatry* **75**(11), 1597–1601.

Fegg MJ, Wasner M, Neudert C, *et al.* (2005) Personal values and individual quality of life in palliative care patients. *Journal of Pain and Symptom Management* **30**(2), 154–159.

Felgoise SH, Stewart JL, Bremer BA, *et al.* (2009) The SEIQoL-DW for assessing quality of life in ALS: Strengths and limitations. *Amyotrophic Lateral Sclerosis* **10**(5–6), 456–462.

Galvin M, Corr B, Madden C, *et al.* (2016) Caregiving in ALS - A mixed methods approach to the study of burden. *BMC Palliative Care* **15**(1), 81.

Gauthier A, Vignola A, Calvo A, *et al.* (2007) A longitudinal study on quality of life and depression in ALS patient-caregiver couples. *Neurology* **68**(12), 923–926.

Hickey AM, Bury G, O'Boyle CA, *et al.* (1996) A new short form individual quality of life measure (SEIQoL-DW): Application in a cohort of individuals with HIV/AIDS. *BMJ* **313**(7048), 29–33.

Hughes RA, Sinha A, Higginson I, *et al.* (2005) Living with motor neurone disease: Lives, experiences of services and suggestions for change. *Health & Social Care in the Community* **13**(1), 64–74.

Jakobsson Larsson B, Ozanne AG, Nordin K, *et al.* (2017) A prospective study of quality of life in amyotrophic lateral sclerosis patients. *Acta Neurologica Scandinavica* **136**(6), 631–638.

Krivickas LS, Shockley L and Mitsumoto H (1997) Home care of patients with amyotrophic lateral sclerosis (ALS). *Journal of the Neurological Sciences* **152**(Suppl. 1), S82–S89.

Larsson BJ, Frojd C, Nordin K, *et al.* (2015) Relatives of patients with amyotrophic lateral sclerosis: Their experience of care and support. *Palliative & Supportive Care* **13**(6), 1569–1577.

Lo Coco G, Lo Coco D, Cicero V, *et al.* (2005) Individual and health-related quality of life assessment in amyotrophic lateral sclerosis patients and their caregivers. *Journal of the Neurological Sciences* **238**(1–2), 11–17.

Mitchell JD and Borasio GD (2007) Amyotrophic lateral sclerosis. *Lancet* **369** (9578), 2031–2041.

Murphy V, Felgoise SH, Walsh SM, *et al.* (2009) Problem solving skills predict quality of life and psychological morbidity in ALS caregivers. *Amyotrophic Lateral Sclerosis* **10**(3), 147–153.

Neudert C, Wasner M and Borasio GD (2001) Patients' assessment of quality of life instruments: A randomised study of SIP, SF-36 and SEIQoL-DW in patients with amyotrophic lateral sclerosis. *Journal of the Neurological Sciences* **191**(1–2), 103–109.

Nygren I and Askmark H (2006) Self-reported quality of life in amyotrophic lateral sclerosis. *Journal of Palliative Medicine* **9**(2), 304–308.

O'Connor EJ and McCabe MP (2011) Predictors of quality of life in carers for people with a progressive neurological illness: A longitudinal study. *Quality of Life Research* **20**(5), 703–711.

Olsson AG, Markhede I, Strang S, *et al.* (2010) Differences in quality of life modalities give rise to needs of individual support in patients with ALS and their next of kin. *Palliative & Supportive Care* **8**(1), 75–82.

https://doi.org/10.1017/S1478951521000778 Published online by Cambridge University Press

Roach AR, Averill AJ, Segerstrom SC, *et al.* (2009) The dynamics of quality of life in ALS patients and caregivers. *Annals of Behavioral Medicine* **37**(2), 197–206.

Schwartz CE, Andresen EM, Nosek MA, *et al.* (2007) Response shift theory: Important implications for measuring quality of life in people with disability. *Archives of Physical Medicine and Rehabilitation* **88**(4), 529–536.

Trail M, Nelson ND, Van JN, *et al.* (2003) A study comparing patients with amyotrophic lateral sclerosis and their caregivers on measures of quality of life, depression, and their attitudes toward treatment options. *Journal of the Neurological Sciences* **209**(1–2), 79–85.

Wettergren L, Bjorkholm M, Axdorph U, *et al.* (2003) Individual quality of life in long-term survivors of Hodgkin's lymphoma – A comparative study. *Quality of Life Research* **12**(5), 545–554.

Wettergren L, Bjorkholm M and Langius-Eklof A (2005) Validation of an extended version of the SEIQoL-DW in a cohort of Hodgkin lymphoma' survivors. *Quality of Life Research* **14**(10), 2329–2333.

Wettergren L, Sprangers M, Bjorkholm M, *et al.* (2008) Quality of life before and one year following stem cell transplantation using an individualized and a standardized instrument. *Psycho-Oncology* **17**(4), 338–346.

The World Health Organization Quality of Life Assessment (WHOQOL): Position paper from the World Health Organization (1995) *Social Science & Medicine* **41**(10), 1403–1409.

Zigmond AS and Snaith RP (1983) The hospital anxiety and depression scale. *Acta Psychiatrica Scandinavica* **67**(6), 361–370.

https://doi.org/10.1017/S1478951521000778 Published online by Cambridge University Press