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Associations between illness cognitions and health-related quality of life in the first year after diagnosis of amyotrophic lateral sclerosis

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ABSTRACT

Objective: To describe illness cognitions among patients with amyotrophic lateral sclerosis (ALS), to study cross-sectional associations between illness cognitions and health-related quality of life (HRQoL) and to study the predictive value of illness cognitions measured shortly after the diagnosis for HRQoL at follow-up.
Methods: Prospective longitudinal design. We administered Self-report questionnaires at study onset (n = 72) and follow-up (n = 48). Median follow-up period was 10.0 months. At baseline median ALS Functional Rating Scale-Revised was 43, median time since onset of symptoms was 13.6 months, 79% of patients presented with spinal onset. Illness cognitions Helplessness, Acceptance and Disease Benefits were measured with the Illness Cognitions Questionnaire (ICQ) and HRQoL with the ALS Assessment Questionnaire (ALSAQ-40). Correlational and regression analyses were used.
Results: Patients experienced more Helplessness at follow-up. We found no significant changes in Acceptance or Disease Benefits at follow-up. In cross-sectional analyses, Helplessness was independently related to worse HRQoL at baseline (β = 0.44; p = .001) and Acceptance and Disease Benefits were independently related to worse HRQoL at follow-up (β = −0.17, p = .045) and (β = −0.186, p = .03 respectively). Longitudinal analyses showed that, adjusted for disease severity at baseline, Helplessness at baseline was a predictor of worse HRQoL at follow-up (β = 0.43; p = .006). None of the illness cognitions were a significant predictor of HRQoL with adjustment for baseline HRQoL.
Conclusion: Helplessness was independently associated with HRQoL in the cross-sectional and longitudinal analyses. These results can help us identify patients shortly after diagnosis who might benefit from psychological interventions.

1. Introduction

Amyotrophic Lateral Sclerosis (ALS) is a fatal progressive neurodegenerative disorder. Despite extensive research, there is currently no curative treatment available. Daily care focuses on symptom management and preserving Health-Related Quality of Life (HRQoL) [1]. There is an increasing awareness that psychological and behavioral determinants are associated with HRQoL among patients with ALS [1,2].

The concept of illness cognitions and related concepts such as appraisals, illness beliefs, or illness perceptions refer to the way people think about and perceive their disease [3–5]. The importance of this is increasingly being recognised across a broad range of conditions, including stroke [6], cancer [7–10], Huntington [11], Parkinson's disease [12], multiple sclerosis [13], spinal cord injury [14] and muscle disease [15]. One previous study on illness cognitions among ALS patients described two clusters of ALS patients according to their illness representations: adaptors and non-adaptors [16]. The two groups were characterized by different forms of thinking about and perceiving their disease, with impact on their level of health-related quality of life. Additionally, research among other diagnostic groups has suggested

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that different illness beliefs may be prominent at different disease stages [17]. However, no longitudinal studies among ALS patients have been performed on this subject, and, therefore, we do not have insight in how illness cognitions relate to QoL among patients with ALS during the progression of their disease. For daily practice, having insight in patients at risk of developing a lower QoL shortly after diagnosis, could be helpful in delivering personalized care.

The aims of our study are (1) to describe positive and negative illness cognitions in ALS patients using a validated questionnaire, (2) to study cross-sectional associations between illness cognitions and HRQoL, and (3) to study the longitudinal associations between illness cognitions measured shortly after the diagnosis of ALS with HRQoL at follow-up. Knowledge about illness cognitions and HRQoL could help us identify patients who may benefit from interventions.

2. Patients and methods

2.1. Patients

This study used data collected in a multicentre trial (FACTS-2-ALS). The methods have been published elsewhere [18]. Recruitment took place between 2009 and 2015. The Medical Ethics Committees from all participating centres approved the study protocol and informed consent was obtained from all patients.

Inclusion criteria were: age between 18 and 80 years; life-expectancy of more than 1 year; predicted forced vital capacity of at least 80%; diagnosed with probable or definite ALS [19], at least one month post-diagnosis and able to walk and cycle. Data for the current study were collected at inclusion (T0) and follow-up (after 10 months; T1). Relevant exclusion criteria were: cognitive impairment (whether or not related to ALS, preventing the intervention from being completed) and psychiatric disorder, both assessed using the Cumulative Illness Rating Scale [20]. Patients could be included for 2 interventions or Usual Care (control group).

The two interventions comprised of cognitive behavioral therapy (CBT) or aerobic exercise therapy (AET). For CBT, an additional inclusion criterium comprised of a Hospital Anxiety and Depression score (HADS) [21] above 8 points. Patients in the control group were not made aware of the possibility of the AET or CBT intervention to avoid a bias relating to negative feelings concerning not participating in the treatment arm.

2.2. Measurements

Demographic variables (age, gender), time since onset of first symptoms and site of first symptoms were collected at inclusion. All measurements at follow up were collected in the same way as the first time at T0. Disease severity was assessed using the revised ALS Functional Rating Scale-Revised (ALSFRS-R) [22]. The ALSFRS-R, a valid, reliable and sensitive instrument includes 12 items structured on a 5-point scale (0 = unable, 4 = normal). The items assess limb, bulbar and respiratory function.

Forced Vital capacity (FVC) as a determinant of lung-capacity was measured with a spirometer (MicroRPM; PT Medical, Leek, The Netherlands) and the score was expressed as a percentage of the predicted score based on the patient's gender, weight, race and height. In case of insufficient lip closure a face mask was used. Each participant made 2 attempts and the maximum score was recorded.

Illness cognitions were measured using the Illness Cognitions Questionnaire (ICQ) [3,23]. This questionnaire consists of 18 items (three 6-item scales), with a 4-point response scale ranging from ‘not at all’ to ‘completely’. The three subscales reflect different illness cognitions: Helplessness as a way of emphasizing the aversive meaning of the disease, Acceptance as a way to diminish the aversive meaning and Disease Benefits as a way of attributing positive meaning to a disease. Scale scores are calculated by summing up the item scores and range from 0 to 24. Higher scores indicate greater presence of the illness cognition in question. The three-factor structure [23] and the clinical usefulness have been studied and supported by various groups [13,14]. In sum, the ICQ showed a strong internal consistency, reliability, and good predictive and construct validity. Intercorrelations between the scales were moderate, which revealed their content validity.

HRQoL was assessed using the Dutch version of the ALS Assessment Questionnaire (ALSAQ-40) [24]. The ALSAQ-40 is a disease-specific questionnaire with 40 questions, each with a 5-point response scale. Domains are mobility, independence in mobility and self-care, eating and drinking, communication, emotional functioning. The total score has a range from 0 to 100, with higher scores indicating poorer health status. Validity and reliability of the ALSAQ-40 are reported to be good [24,25].

2.3. Statistical analyses

Descriptive statistics were used to describe characteristics of the study population, ICQ and ALSAQ-40 scores at baseline and at follow-up. At follow-up, it was assessed whether there were differences in the baseline scores of those who continued to participate and those who dropped out. Wilcoxon Signed Rank tests were performed to examine changes in ALSFRS-R, FVC, ALSAQ-40 and ICQ scores between onset and follow-up. Effect sizes were calculated using the formula $r = Z/\sqrt{N}$. Spearman's rank correlation coefficients were computed to assess cross-sectional associations between potential determinants and ALSAQ-40 scores at T0 and at T1. To study the possible correlation between the illness cognition domains and the rate of disease progression evaluated by the difference between ALSFRS-R score at baseline and at follow-up (Δ ALSFRS-R). This allowed us to understand how the level of disease progression may influence the illness cognitions in ALS patients. Using Cohen's rule of thumb, a correlation of 0.10 was considered ‘weak’, of 0.30 ‘moderate’ and of 0.50 ‘strong’ [26,27]. Hierarchical linear regression was used to study the associations between illness cognitions and ALSAQ-40 scores, controlling for disease severity or HRQoL. Because of the restricted sample size, only determinants that showed a p-value < .05 in the correlation analysis (ALSFRS-R and FVC), were entered into the regression models. Variables were entered in the following order: step 1: Illness cognitions; step 2: disease severity variables, and demographics; Step 3: To study the impact of participating in AET or CBT, two dummy variables reflecting participating in either AET or CBT were added to the regression analysis.

Hierarchical linear regression analyses were performed to study the predictive value of illness cognitions at baseline, corrected for CBT or AET intervention, on HRQoL at follow-up, while controlling first for disease severity at baseline and second for HRQoL at baseline.

Residual analyses were performed and multi-collinearity was tested to search for violations of the assumptions underlying multiple regression. For all questionnaires, up to 25% of missing values were permitted. These were replaced by the mean of the missing values of the same scale.

SPSS version 24 for Windows was used for all statistical analyses.

2.4. Results

A total of 72 patients were included in the FACTS-ALS trial and 48 patients completed all questionnaires at both baseline and follow-up. Median follow up period was 10.0 months, mean follow up period was 10.1 months (SD 0.57, range 9–12 months). Of these 48 patients, 6 were allocated to the CBT intervention, 16 to the AET intervention (11 of whom completed the module) and 26 to the usual care group. The most frequent reason for dropping out of the trial was death or because they experienced participation as too burdensome. Table 1 presents patient characteristics and scores on the primary outcome measures. No significant differences (p < .05) at base line were found between patients who participated at follow-up and those who dropped out of the study.

Table 1
Patients' characteristics at baseline (T0) and follow up (T1).

	T0 all patients (n = 72)	T0 patients who completed T1 (n = 48)	T1 (n = 48)	Difference at T0 between participants and dropouts at T1,p
Age in years *mean (SD)*	59.9 (10.6)	60.3 (9.4)	60.5 (9.4)	0.91
Sex, male *n* (%)	50 (69.4)	31 (64.6)	31 (64.6)	0.21
Time since onset in months *Mdn, (IQR)*	12.0 (8–21)	13.6 (9–23)	24.0 (20 − 32)	0.38
Time since diagnosis in months *Mdn (IQR)*	3.3 (2–5)	3.3 (2–5)	13.0 (12–15)	0.20
Spinal onset n (%)	53 (73.6)	38 (79.2)	38 (79.2)	0.13
ALSFRS-R *Mdn, (IQR)*	43.0 (40–45)	43.0 (40–46)	34 (26–39)	0.11
Severe (≤27)	1 (1.4%)	1 (2.1%)	13 (27.1%)	
Moderate (28–37)	6 (8.3%)	3 (6.3%)	21 (43.8%)	
Mild (≥38)	65 (90.3%)	44 (91.7%)	14 (29.2%)	
FVC% *Mdn (IQR)*	94.0 (82.2–104)	97 (85–104)	74 (66.3–82.8)	0.19
ALSAQ[a] *Mdn (IQR)*	26.9 (17.2–35.6)	23.1 (15.6–35.6)	40.9 (26.4–53.8)	0.09
Nr of patients in AET intervention		6		
Nr of patients in CBT intervention		16		

ALSFRS-R, revised ALS Functional Rating Scale; FVC, Forced vital capacity; ALSAQ, ALS Assessment Questionnaire.
[a] Higher ALSAQ scores indicate lower health related quality of life.

Table 2
Illness cognition scores at baseline and follow-up and change in Illness cognition scores between baseline and follow-up.

ICQ	T0 (n = 71)		T0 (n = 48)		T1 (n = 48)		Effect size, r
	Median (IQR)	Mean (SD)	Median (IQR)	Mean (SD)	Median (IQR)	Mean (SD)	
Helplessness	12.0 (10–16)	13.2 (4.4)	12.0 (10–16)	12.8 (4.5)	15.0 (13–19)	15.7 (4.1)	0.54*
Acceptance	15.0 (12–17)	15.9 (3.8)	15.0 (13–18)	15.3 (3.8)	16.0 (13 − 20)	16.3 (4.4)	0.23
Disease benefits	13.0 (10–15)	13.0 (3.7)	13.0 (10–15)	12.9 (3.7)	13.0 (10–16)	13.4 (3.9)	0.09

ICQ, Illness cognition questionnaire; T0 start trial, T1 10.6 months later.
IQR, interquartile range; Wilcoxon signed Rank effect size, $r = Z/\sqrt{N}$.
* $p < 0.05$

Table 2, distributions of the ICQ scores. Helplessness scores increased significantly between baseline and follow-up, but no significant changes in Acceptance or Perceived Benefit scores were seen.

Table 3 presents the item scores of the ICQ over time. All item scores of the Helplessness domain increased over time. Overall Acceptance scores appeared to be high compared to scores of the Helplessness domain.

Table 3
ICQ item scores of Helplessness, Acceptance and Disease Benefits. % of participants scoring Yes on this items.

N = 48	T0 (%)	T1 (%)
Helplessness		
1. Because of my illness, I miss the things I like to do most	41.7	65.9
2. My illness controls my life	50.0	59.6
3. My illness makes me feel useless at times	10.5	27.7
4. My illness prevents me from doing what I would really like to do	41.7	70.2
5. My illness limits me in everything that is important to me	29.2	46.8
6. My illness frequently makes me feel helpless	16.6	36.2
Acceptance		
7. I can handle the problems related to my illness	75.0	76.6
8. I have learned to live with my illness	47.9	66.0
9. I have learned to accept the limitations imposed by my illness	37.5	55.3
10. I can accept my illness well	50.0	59.5
11. I think I can handle the problems related to my illness, even if the illness gets worse	39.6	54.3
12. I can cope effectively with my illness	62.5	61.7
Perceived benefits		
13. Dealing with my illness has made me a stronger person	20.8	34.0
14. I have learned a great deal from my illness	18.8	38.3
15. My illness has made life more precious to me	50.1	34.0
16. Looking back, I can see that my illness has also brought about some positive changes in my life	14.6	26.0
17. My illness has helped me realize what's important in life	50.0	45.7
18. My illness has taught me to enjoy the moment more	60.5	68.1

Table 4 displays the Spearman Correlations between Illness cognitions questionnaire (ICQ) with demographic and disease characteristics and quality of life (ALSAQ), at T0 and T1.

At follow-up, more Helplessness was strongly related to less Acceptance and moderately related to less Disease Benefits and more Acceptance was moderately related to Disease Benefits. More Helplessness was strongly related to higher ALSAQ-40 scores, both at baseline and follow-up. The relationship between functioning and HRQoL scores was stronger at follow-up compared to baseline. There is a significant correlation between Δ ALSFRS-R and outcome measure ALSAQ and ICQ-Helplessness.

Table 5 summarizes the results of the cross-sectional regression analyses at baseline and follow-up. At baseline, Helplessness was the only ICQ-subscale independently associated with HRQoL, explaining 38% of the ALSAQ-40 score. After adding the other variables, Helplessness was still independently associated with HRQoL (total explained variance 53%). At follow-up, Helplessness was the only ICQ subscale independently associated with HRQoL, explaining 40% of the variance. After adding disease severity and controlling for AET or CBT, Acceptance and Disease Benefit and disease severity (ALSFRS-R) were significantly associated with HRQoL (R^2 = 0.41), explaining 81% of the variance in HRQoL at follow up.

Table 6 summarizes results of the longitudinal analyses. A total of 48% of the variance in HRQoL at follow-up was explained by HRQoL at baseline. Illness cognitions at baseline were not significantly associated with HRQoL at follow-up, when adjusted for baseline HRQoL. When entering ALSFRS-R (baseline) and ICQ scales (baseline) together in the model, 27% of the variance in HRQoL at follow-up was explained by Helplessness scores at baseline.

This model did not change after controlling for CBT or AET.

Table 4
Spearman correlations between Illness cognitions questionnaire (ICQ) with demographic and disease characteristics and quality of life (ALSAQ), at T0 and T1.

	Cross-sectional T0 (n = 72)				Cross-sectional T1 (n = 48)			
	ALSAQ T0	Helplessness	Acceptance	Disease benefits	ALSAQ T1	Helplessness	Acceptance	Disease Benefits
Gender, (m)[a]	0.09	−0.22	−0.34[a]	−0.11	0.11	0.16	−0.25	−0.14
Age	0.05	0.10	−0.25*	−0.09	−0.16	0.18	−0.14	0.06
ALSFRS-R	−0.63**	−0.48**	−0.02	0.09	−0.86**	−0.43**	0.12	0.15
Δ ALSFRS-R					0.81**	0.46**	−0.17	−0.11
FVC	−0.27*	−0.08	0.07	−0.10	−0.52**	−0.31*	0.18	0.16
ICQ-H	0.64**	1.00	−0.10	−0.13	0.51**	1.00	−0.47**	−0.37**
ICQ-A	−0.08	−0.10	1.00	0.18	−0.34*	−0.47**	1.00	0.35*
ICQ-DB	0.07	−0.13	0.18	1.00	−0.32*	−0.37**	0.35*	1.00

T0 start trial, T1 10 months. ALSFRS-R, revised ALS Functional Rating Scale; Δ ALSFRS-R, difference T1-T0 in ALSFRS-R score; FVC, Forced vital capacity. ICQ, Illness cognitions questionnaire; ICQ-H, Illness cognitions questionnaire Helplessness; ICQ-A, Illness cognitions questionnaire-Acceptance; ICQ-DB, Illness cognitions questionnaire-Disease Benefits; Δ ALSFRS-R, difference T1- T0 ALSFRS-R scores.

* $p < .05$.

** $P < .01$.

[a] Point-biserial correlation.

Table 5
Linear regression analysis of the effects of illness cognitions on health related QoL (ALSAQ Sum score) at T0 and T1.

	Cross-sectional T0 (n = 72)		Cross-sectional T1 (n = 48)	
	Step 1, β	Step 2, β	Step 1, β	Step 2, β
ICQ-Helplessness	*0.62	0.44*	0.56*	0.13
ICQ-Acceptance	NA	NA	−0.08	0.17*
ICQ-Disease Benefits	NA	NA	−0.04	0.19*
ALSFRS-R	NA	−0.40*	NA	−0.66*
FVC	NA	−0.20*	NA	−0.16
Dummy CBT			−0.04	−0.08
Dummy AET			0.07	0.09
ΔR^2	0.38	0.15	0.40	0.41
Explained variance		53%		81%

T0 start trial, T1 10 months. β, standardized coefficient. NA, not added. ALSAQ, ALS Assessment Questionnaire; ICQ, Illness cognitions questionnaire; ALSFRS-R, revised ALS Functional Rating Scale; FVC, Forced vital capacity; Yes/no CBT, participation CBT intervention yes/no; Yes/no AET, participation AET intervention yes/no.

* $p < .05$.

Table 6
Predictive linear regression analysis with ICQ subscales and ALSFRS-R or ALSAQ at T0 as possible correlates of ALSAQ at T1.

T0	Step 1	Step 2 with ALSAQ T0	Step 2 with ALSFRS-R T0
	β	β	β
ICQ-Helplessness	0.44*	0.07	0.43*
ICQ-Acceptance	0.03	0.02	0.02
ICQ-Disease Benefits	−0.16	−0.20	−0.17
ALSAQ-T0	NA	0.59*	NA
ALSFRS-R T0	NA	NA	−0.03
ΔR-square	0.27	0.22	0.01
Total R-square	0.27	0.48	0.27

Dependent variable: ALSAQ-T1. β, standardized coefficient. NA, not added in analysis. T0 start trial, T1 10 month; NA, not added in the analysis.

* $p < .05$.

3. Discussion

There is an increasing awareness that psychological factors are associated with HRQoL among patients with ALS. The results of this study showed a significant increase of Helplessness, but no significant changes in Acceptance or Disease Benefits between baseline and follow-up. Despite this, at follow up Acceptance and Disease Benefits measured at follow up were independently related to HRQoL. Helplessness was further independently related to HRQoL at baseline and Helplessness measured at baseline was an independent predictor of HRQoL at follow-up.

The Helplessness score at baseline was equal to scores among patients with Rheumatoid arthritis (RA) and lower compared to scores among breast cancer patients and patients with Multiple Sclerosis (MS), in a latter phase of their disease. [3,10,13]. Baseline Acceptance and Disease benefits scores were lower (= worse) compared to scores among patients with RA, MS and after stroke [3,6,13]. At follow-up Helplessness score were higher (=worse) than the scores found among stroke patients and patients with spinal cord injury [6,14]. Our patients experienced physical deterioration, which is usually not the case among stroke patients and patients with spinal cord injury which can explain the higher scores. Acceptance and Disease Benefits scores at follow-up were lower (=worse) than those found among spinal cord injury patients and stroke patients in a longitudinal study. Again, this could be associated with the physical deterioration our patients experienced. Compared to these patients, ALS patients reported more change in illness cognitions.

The association between the ICQ-helplessness scores and ALSAQ-40 changed over time. Corrected for disease severity, higher Helplessness scores at baseline were associated with lower HRQoL at follow-up. This result implies that we may have found a way to select a subgroup of patients shortly after diagnosis who might need extra attention in daily care. This group might benefit from a psychological intervention, such as described in studies among patients with muscle disorders (including ALS patients) [28–33]. To target helplessness specifically as an unfavourable cognition individual, daily care should focus on 1: physical aspects of helplessness due to physical limitations and ongoing deterioration by providing personalized care, just in time (assistive devices just in time, adequate symptom management and shared decision making during multidisciplinary care). 2: on the feelings of helplessness due to loss of control.

Despite the fact that Acceptance and Disease benefit scores did not increase significantly, these scores were associated with HRQoL at follow up. At that moment patients have had more experience with the impact of the disease. As stated by Evers, Acceptance can be regarded a way to diminish the aversive meaning of disease and Disease Benefits as a way of attributing positive meaning to a disease. This explains the association with HRQoL and gives ground for psychological interventions based on ACT.

Helplessness at T1 was significantly correlated with disease severity (ALSFRS-R) and change in disease severity (ΔALSFRS-R scores). This association between higher Helplessness scores and disease progression was also found in patients with multiple sclerosis [13]. There is a wide

variety in disease progression and survival among patients with ALS [34]. Future studies including larger samples could compare the course of illness cognitions between subgroups with different survival prognosis. In our population the correlation between Helplessness and disease severity increased over time, which may be explained by greater physical deterioration at follow-up. However, the questions in the Helplessness scale are not all oriented at physical functioning. Patients apparently experience an overall feeling of Helplessness due to deterioration. As the variety in Helplessness is strongly correlated to HRQoL, it is important to monitor patients frequently. In our study, 22 patients participated in an intervention of the FACTS-2-ALS trial (CBT or AET). We evaluated the impact of these patients who participated in an intervention, on our results. This has not lead to different conclusions, and therefore we included the data of these patients in our calculations.

Based on theories about post-traumatic growth and response shift and results from other studies [2,8,35,36] we expected, but did not find an increase of Acceptance and Disease Benefits scores between baseline and follow-up. Posttraumatic growth is defined as a collection of positive changes following a traumatic event which stimulates the individual to re-evaluate his/her worldview. Posttraumatic growth has interfaces with another phenomenon called 'response shift'. The response shift theoretical model [36] posits that a health state change (catalyst) causes an individual to utilize cognitive, behavioral, and emotion-focused coping strategies (mechanisms). Baring these phenomenon in mind, we expected more acceptance and disease benefits in time. Qualitative research has suggested that different illness beliefs may be prominent at different disease stages [16]. Regarding the ICQ item scores, from onset, 50% of the patients score on the acceptance items. Over time, a higher percentage of patients score helplessness, simultaneously. One could conclude that these patients have a realistic insight in the consequences of their disease. Additionally, in accordance with the Theory of Waldron about psychological adaptation to terminal illness, there might be a shift in focus of determinants of QoL, physical functioning to psychological and spiritual domains [37].

This is the first study with a longitudinal focus on illness cognitions in relation to quality of life among ALS patients. Following patients over time has given us more insight into the development of cognitions like Helplessness, Acceptance and Disease Benefits and their associations with change in HRQoL over time.

However, interpretation of our results must take account of the following limitations.

First, patients included in the FACTS-2-ALS trial needed to be able to participate in physical exercise, and therefore the less impaired patients were selected. At diagnosis, there are patients who have already severe physical limitations. Patients with a very progressive disease course are probably not included in this study. However, we do not have insight in the amount of people who were not eligible to participate. Second, the impact of cognitive and/or behavioral changes in the frontotemporal spectrum for example the phenomenon of anosognosia, due to ALS, were not studied, but we would expect a negative association of frontotemporal behavioral changes with adaptative psychological processes. Third, we did not include psychological factors such as resilience or coping in our study; these are factors described among e.g. cancer patients as influencing the adaptation process [38]. Fourth, because of the limited sample size, we were able to add only a limited amount of variables in the regression analysis.

In conclusion, Helplessness was independently associated with HRQoL in the cross-sectional and longitudinal analyses. In daily care, we strive to provide personalized care with the aim to optimize QoL despite physical limitations.The results of this study can help us identify patients with ALS who might benefit from possible psychological interventions e.g. acceptance and commitment therapy (ACT) or mindfulness [32,33,39]. As several authors are indicating that psychological interventions are promising, we should be studying their efficacy.

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References

[1] Z. Simmons, B.A. Bremer, R.A. Robbins, S.M. Walsh, S. Fisher, Quality of life in ALS depends on factors other than strength and physical function, Neurology 55 (2000) 388–392.
[2] A.C. Van Groenestijn, E.T. Kruitwagen-van Reenen, J.M.A. Visser-Meily, L.H. van den Berg, C.D. Schröder, Associations between psychological factors and health-related quality of life and global quality of life in patients with ALS: a systematic review, Health Qual. Life Outcomes 14 (2016) 107.
[3] A.W. Evers, F.W. Kraaimaat, W. van Lankveld, P.J. Jongen, J.W. Jacobs, J.W. Bijlsma, Beyond unfavourable thinking: the illness cognition questionnaire for chronic diseases, J. Consult. Clin. Psychol. 69 (2001) 1026–1036.
[4] H. Leventhal, M. Diefenbach, E.A. Levental, Illness cognitions: using common sense to understand treatment adherence and affect cognition interactions, Cogn. Ther. Res. 16 (1992) 143–163.
[5] M.S. Hagger, S. Orbell, A meta-analytic review of the common-sense model of illness representations, Psychol. Health 18 (2003) 141–184.
[6] M. van ML, H. van CM, M.W.M. Post, K. de PLM, J.M.A. Visser-Meily, Life satisfaction post stroke: the role of illness cognitions, J. Psychosom. Res. (2015) 137–142.
[7] J.C. Coyne, H. Tennen, Positive psychology in cancer care: bad science, exaggerated claims, and unproven medicine, Ann. Behav. Med. 39 (2010) 16–26.
[8] N.K. Mc Corry, M. Dempster, J. Quinn, A. Hogg, J. Newell, M. Moore, et al., Illness perception clusters at diagnosis predict psychological distress among women with breast cancer at 6 months post diagnosis, Psycho-Oncology 22 (2013) 692–698.
[9] H. Rozema, T. Vőllink, L. Lechner, The role of illness representations in coping and health of patients treated for breast cancer, PsychoOncology 18 (2009) 849–857.
[10] J. Han, J.-E. Liu, H. Qiu, Z.-H. Nie, Y.-L. Su, Illness cognitions and the associated socio-demographic and clinical factors in Chinese women with breast cancer, Eur. J. Oncol. Nurs. (2018) 33–39.
[11] A.A. Kaptein, D.I. Helder, M. Scharloo, et al., Illness perceptions and coping explain well-being in patients with Huntington's disease, Psychol. Health 21 (2006) 431–446.
[12] D. Evans, P. Norman, Illness representations, coping and psychological adjustment to Parkinson's disease, Psychol. Health 24 (2009) 1181–1196.
[13] P. van der Werf, A. Evers, P.J.H. Jongen, G. Bleijenberg, The role of helplessness as mediator between neurological disability, emotional instability, experienced fatigue and depression in patients with multiple sclerosis, Mult. Scler. 9 (2003) 89–94.
[14] C.M.C. Van Leeuwen, Y. Edelaar-Peters, C. Peter, A.M. Stiggelbout, M.W.M. Post, Psychological factors and mental health in persons with spinal cord injury: an exploration of change or stability, J. Rehabil. Med. 47 (2015) 531–537.
[15] C.D. Graham, J. Weinman, R. Sadjadi, T. Chalder, R. Petty, M.G. Hanna, et al., A multicentre postal survey investigating the contribution of illness perceptions, coping and optimism to quality of life and mood in adults with muscle disease, Clin. Rehabil. 28 (2014) 508–519.
[16] M. Miglioretti, L. Mazzini, G.D. Oggioni, L. Testa, F. Monaco, Illness perceptions, mood and health-related quality of life in patients with amyotrophic lateral sclerosis, J. Psychosom. Res. 65 (2008) 603–609.

[17] C.S. Hurt, C.L. Julien, R.G. Brown, Measuring illness beliefs in neurodegenerative disease: why we need to be specific, J. Health Psychol. 20 (1) (2015 Jan) 69–79.
[18] A.C. Van Groenestijn, I.G. van de Port, C.D. Schröder, M.W.M. Post, L.H. van den Berg, E. Lindeman, et al., Effects of aerobic exercise therapy and cognitive behavioral therapy on functioning and quality of life in amyotrophic lateral sclerosis: protocol of the FACTS-2-ALS trial, BMC Neurol. 11 (2011) 70.
[19] B.R. Brooks, R.G. Miller, M. Swash, T.L. Munsat, El Escorial revisited: revised criteria for the diagnosis of amyotrophic lateral sclerosis, Amyotroph. Lateral Scler. Other Motor Neuron Dis. 1 (2000) 293–299.
[20] B.S. Linn, M.W. Linn, L. Gurel, Cumulative illness rating scale, J. Am. Geriatr. Soc. 16 (1968) 622–626.
[21] A.S. Zigmond, R.P. Snaith, The hospital anxiety and feelings of depression scale. The hospital anxiety and feelings of depression scale, Acta Psychiatr. Scand. 67 (1983) 361–370.
[22] J.M. Cedarbaum, N. Stambler, E. Malta, C. Fuller, D. Hilt, B. Thurmond, et al., The ALSFRS-R: a revised ALS functioning rating scale that incorporates assessments of respiratory function. BDNF ALS Study Group (Phase III), J. Neurol. Sci. 169 (1999) 13–21.
[23] E. Lauwerier, G. Crombez, S. Van Damme, L. Goubert, D. Vogelaers, A.W.M. Evers, The construct validity of the illness cognition questionnaire: the robustness of the three-factor structure across patients with chronic pain and chronic fatigue, Int. J. Behav. Med. 17 (2010) 90–96.
[24] M. Maessen, M.W.M. Post, R. Maillé, E. Lindeman, R. Mooij, J.H. Veldink, et al., Validity of the Dutch version of the amyotrophic lateral sclerosis assessment questionnaire, ALSAQ-40, ALSAQ-5, Amyotroph. Lateral Scler. 8 (2007) 96–100.
[25] C. Jenkinson, R. Fitzpatrick, C. Brennan, M. Swash, Evidence for the validity and reliability of the ALS assessment questionnaire: the ALSAQ-40, Amyotroph. Lateral Scler. Other Motor Neuron Dis. 1 (1999) 33–40.
[26] J. Cohen, Statistical Power Analysis for the Behavioural Sciences, Academy Press, New York, 1988.
[27] A. Field, Discovering Statistics Using IBM SPSS Statistics, Fourth edition, Sage Publications, Thousand Oaks, California, 2013.
[28] C.D. Graham, Z. Simmons, S.R. Stuart, M.R. Rose, The potential of psychological interventions to improve quality of life and mood in muscle disorders, Muscle Nerve 52 (2015) 131–136.
[29] R.L. Gould, M.C. Coulson, R.G. Brown, L.H. Goldstein, A. Al-Chalabi, R.J. Howard, Psychotherapy and pharmacotherapy interventions to reduce distress or improve well-being in people with amyotrophic lateral sclerosis: a systematic review, Amyotroph. Lateral Scler. Frontotemporal Degener. 16 (2015) 293–302.
[30] M.C.F. Oberstadt, P. Esser, J. Classen, A. Mehnert, Alleviation of psychological distress and the improvement of quality of life in patients with amyotrophic lateral sclerosis: adaptation of a short-term psychotherapeutic intervention, Front. Neurol. 16 (9) (2018) 1–6.
[31] T.J. Connerty, V. Knott, Promoting positive change in the face of adversity: experiences of cancer and post-traumatic growth, Eur. J. Cancer Care 22 (2013) 334–344.
[32] F. Pagnini, A. Marconi, A. Tagliaferri, G.M. Manzoni, R. Gatto, V. Fabiani, et al., Meditation training for people with amyotrophic lateral sclerosis: a randomized clinical trial, Eur. J. Neurol. 24 (2017) 578–586.
[33] F. Pagnini, D. Phillips, C. Bosma, A. Reece, E. Langer, Mindfulness, physical impairment and psychological well-being in people with amyotrophic lateral sclerosis, Psychol. Health 30 (2015) 503–517.
[34] H.-J. Westeneng, T.P.A. Debray, A.E. Visser, R.P.A. van Eijk, J.P.K. Rooney, A. Calvo, et al., Prognosis for patients with amyotrophic lateral sclerosis: development and validation of a personalised prediction model, Lancet Neurol. 17 (2018) 423–433.
[35] R.G. Tedeschi, L.G. Calhoun, Posttraumatic growth: conceptual foundations and empirical evidence, Psychol. Inq. 15 (2004) 1–18.
[36] C.E. Schwartz, E.M. Andresen, M.A. Nosek, G.L. Krahn, the RRTC Expert Panel on Health Status Measurement, Response shift theory: important implications for measuring, quality of life in people with disability, Arch. Phys. Med. Rehabil. 88 (2007) 529–536.
[37] D. Waldron, C.A. O'Boyle, M. Kearney, M. Moriarty, D. Carney, Quality-of-life measurement in advanced cancer: assessing the individual, J. Clin. Oncol. 17 (1999) 3603–3611.
[38] D.M.J. Walsh, T.G. Morrison, R.J. Conway, E. Rogers, F.J. Sullivan, A. Groarke, A model to predict psychological and health-related adjustment in men with prostate cancer; the role of post traumatic growth, physical post traumatic growth, resilience and mindfulness, Front. Psychol. 9 (2018) 136.
[39] K.R. Weeks, R.L. Gould, C. Mcdermott, J. Lynch, L.H. Goldstein, C.D. Graham, et al., Needs and preferences for psychological interventions of people with motor neuron disease, Amyotroph. Lateral Scler. Frontotemporal Degener. 20 (7–8) (2019) 521–531.