

Coping with amyotrophic lateral sclerosis: an integrative view

Tamara Matuz, Niels Birbaumer, Martin Hautzinger, Andrea Kübler

▶ To cite this version:

Tamara Matuz, Niels Birbaumer, Martin Hautzinger, Andrea Kübler. Coping with amyotrophic lateral sclerosis: an integrative view. Journal of Neurology, Neurosurgery and Psychiatry, 2010, 81 (8), pp.893. 10.1136/jnnp.2009.201285. hal-00557441

HAL Id: hal-00557441

https://hal.science/hal-00557441

Submitted on 19 Jan 2011

HAL is a multi-disciplinary open access archive for the deposit and dissemination of scientific research documents, whether they are published or not. The documents may come from teaching and research institutions in France or abroad, or from public or private research centers.

L'archive ouverte pluridisciplinaire **HAL**, est destinée au dépôt et à la diffusion de documents scientifiques de niveau recherche, publiés ou non, émanant des établissements d'enseignement et de recherche français ou étrangers, des laboratoires publics ou privés.

TITLE PAGE

Coping with amyotrophic lateral sclerosis: an integrative view

Tamara Matuz Dr. ¹, Niels Birbaumer Prof. Dr. ^{1,2}, Martin Hautzinger Prof. Dr. ³, Andrea Kübler Prof. Dr. ^{1,4}

¹ Institute of Medical Psychology and Behavioral Neurobiology, Eberhard-Karls-University Tübingen, Germany

² Istituto di Ricovero e Cura a Carattere Scientifico (IRCCS), Ospedale San Camillo, Venezia, Lido, Italy

³ Department of Psychology, Eberhard-Karls-University Tübingen, Germany

⁴ Department of Psychology I, Biological Psychology, Clinical Psychology, and Psychotherapy, University of Würzburg, Germany

Correspondence to: Tamara Matuz

Address: Eberhard-Karls-University Tübingen, Institute of Medical Psychology and Behavioral Neurobiology, Gartenstr. 29, 72074 Tübingen, Germany. Telephone number: +49 (0)7071 2975997; Fax number: +49 (0)7071 295956.

Email address: tamara.matuz@medizin.uni-tuebingen.de

Email address of the co-authors: niels.birbaumer@uni-tuebingen.de; martin.hautzinger@uni-tuebingen.de; martin.hautzinger@uni-tuebingen.d

Word count for the paper 3193 and for the abstract 217.

Number of tables: 5; Number of figures: 2; Number of references: 27

Search Terms: 1. psychosocial adjustment; 2. ALS; 3. coping; 4. depression; 5. quality of life

ABSTRACT

Objectives - To identify predictors of psychosocial adjustment to motoneurone disease.

Methods - A total of 27 individuals, with a confirmed diagnosis of amyotrophic lateral sclerosis (ALS), participated in the study. The ALS functional rating scale mean score indicated a high physical impairment of the sample. Months since diagnosis varied between 4 und 129 (*Mdn* 36). Adjustment outcomes were severity of depressive symptoms and individual quality of life (QoL). Predictors included: social support, cognitive appraisal, coping strategies, and illness parameters.

Results - Multiple regression analysis revealed that approximately 60% of the variance of depression and QoL were accounted for by social support, coping strategies and cognitive appraisal. The degree of physical impairment did not explain any variance of the adjustment outcomes. The best predictors for the severity of depressive symptoms were perceived social support and appraisal of coping potential (internal locus of control) and for individual QoL perceived social support.

Conclusions - The focus on medical issues in treatment of ALS is not sufficient. A palliative approach to ALS must equally imply advice with regards to adequate coping strategies, provide the adequate amount of disease and support related information at any one time, and encourage patients to seek social support. Sufficient medication and psychotherapy has to be provided for those patients who show depressive symptoms or disorder.

List of acronyms: ALS = amyotrophic lateral sclerosis; MS = multiple sclerosis; QoL = quality of life; BDI = Beck's Depression Inventory; SEIQoL- DW = The Schedule for the Evaluation of Individual Quality of Life Direct Weighting; BSSS = Berlin Social Support Scales; MNDCS = Motor Neuron Disease Coping Scale;

INTRODUCTION

People with fatal diseases such as HIV, cancer, multiple sclerosis (MS) or amyotrophic lateral sclerosis (ALS) are permanently challenged by disease progression. The impact of symptoms, in ALS and MS particularly the increasing physical impairment due to motor paralyses, and the lack of a curative treatment require adaptive mechanisms. In the context of chronic illness and physical disability the person-environment equilibrium is disrupted and for its restoration patients have to adjust their internal needs to the new external demands. This process has been referred to as the *psychosocial adjustment to chronic illness and disabilities* [1].

There is a general agreement that despite the continuously increasing physical impairment

ALS patients adjust effectively to their illness and are often perceived as particularly positive people. These observations are supported by a number of studies investigating different adjustment variables, e.g. quality of life, depression or anxiety, social support (for an elaborated review see McLeod & Clarke, 2007 [2]) as well as by few studies exploring the neural correlates of emotional processing in ALS [3, 4].

Behavioral and emotional changes in motor neuron disease (MND), when present, have been shown to be partially in line with the spectrum of symptoms that characterize the behavioral variant of frontotemporal dementia (bvFTD) (e.g. apathy, self-centredness, blunting of emotions)[5]. Findings of imaging studies on emotional processing in ALS could, however, not confirm a causal relationship between the altered emotional responses in ALS (e.g. more positive valence and decreased excitability to extreme emotional stimuli) and frontal lobe dysfunction [3].

Although there are several studies investigating different psychosocial aspects of MND/ALS, to our knowledge, no effort has been made to integrate the different determinants of psychosocial adjustment into a model of coping with ALS. To have a model of psychosocial adjustment to ALS would be of value to guide intervention and therapy when patients fail to

cope with the disease leading to depression, poor quality of life, and the wish for a hastened death [6]. The current study aimed at providing such a model. As we assumed that living with ALS constitutes a stressful event in life, we based our approach on a stress-coping model (e.g. Lazarus and Folkman [7]) adapted to chronic diseases [8, 9]. According to this model, psychosocial adjustment to illness is determined by a complex interaction between four variables: *illness parameters*, *cognitive appraisal*, *coping resources* and *coping strategies*. Possible outcome measures of psychosocial adjustment are the affective state and quality of life (QoL) [10].

The objective of the present study was firstly, to examine the utility of Lazarus and Folkman's stress-coping model, i.e. how much of the variance in psychosocial adjustment could be explained by the model in a sample of people with ALS. Secondly, we aimed at determining to what extend the model could predict successful psychological adjustment to ALS.

METHODS

Participants and procedure

The study was approved by the Ethical Committee of the Medical Faculty, University of Tübingen. The inclusion criterion for patients was a neurologist's diagnosis of ALS. Patients with diagnosed frontotemporal dementia, alcoholism and poor knowledge of the German language were excluded. Twenty seven ALS patients (mean age \pm SD: 55.3 \pm 11.1; 35-73; 12 women) were included. Fifteen patients were recruited from the ALS outpatient clinic of the Department of Neurology, University of Ulm, 6 from the Institute of Medical Psychology and Behavioural Neurobiology, University of Tübingen and 6 through an appeal for participation in the study in a biannual magazine published by the German Society for Muscular Diseases. All patients were visited at their homes by a clinical psychologist (TM), signed informed

consent and were interviewed. Patients were interviewed alone, except those (N=2) who used eye movement for communication and needed occasional communicative support by caregivers or family members in the case the interviewer could not decode/understand the message. According to the interviewer, the presence of the third person did not influence the respondent's answers. However, an eventual bias can not be totally excluded based on this assessment.

Measures

Background data. Patients' demographic data, including age, sex, marital status, level of education, and living arrangement and communication abilities were obtained with a semi-structured interview.

Illness parameters. Three illness related variables were assessed: duration of illness (month since diagnosis), dependence on life sustaining treatment (ventilation and nutrition) and physical disability (ALS Function Rating Scale [11] with scores range from 0 = complete paralysis to 40 = normal physical functioning).

Appraisal components. Patients' primary and secondary appraisals were assessed with four face-valid items designed to measure motivational relevance, motivational congruence, problem focused and emotion focused coping potential [12]. Patients rated each item on a 9-point Likert scale (1 referred to *not at all* and 9 to *extremely*). The larger the difference between the two items of primary appraisal "motivational relevance" and "motivational congruence" the more patients feel threatened by the disease.

Social support. The Berlin Social Support Scales (BSSS) [13] was developed based on a multidimensional approach and includes 5 subscales measuring both cognitive and behavioral aspects of social support: perceived available social support, actually received social support, need for support, search for support, and protective buffering. Patients rated their agreement with the statements from each of the scales on a 4-point Likert scale ranging from strongly disagree (1) to somewhat disagree (2), somewhat agree (3) and strongly agree (4).

Coping strategies. Coping strategies can be classified in four categories: problemmanagement, problem-appraisal, emotion-management and emotional-avoidance [14]. Problem-management coping strategies describe active attempts to manage the situation (e.g., seek for information) whereas problem-appraisal coping involves attempts to avoid a direct and active confrontation with the problem by either reappraisals of the stressfulness of the situation (e.g., positive thinking) or by behavioral distraction (e.g., positive acting). Emotionmanagement strategies can be conceptualized as efforts to gain knowledge, to understand and to express emotions engendered by the situation whereas emotional avoidance represents the active attempts to avoid emotions induced by the stressful event [14, 15]. Coping strategies were assessed with the Motor Neuron Disease Coping Scale (MNDCS) [16]. Eighteen items were assigned to 6 subscales. Patients expressed the extent in which they relied on the coping strategies in the last month using a 6-point Likert scale. We integrated the 6 subscales of this measure in the classification of coping strategies described above, as follows: information seeking and support scales as problem management strategies; positive thinking and positive action scales as problem appraisal strategies; independence scale as emotion management coping and avoidance as emotional avoidance strategy. One score for each type of coping was obtained by generating the mean score of the grouped scales.

Depressive symptomatology. Severity of depressive symptoms was assessed with Beck's Depression Inventory [17]. Values below 11 indicate no, values between 11 and 18 mild to moderate, and values above 18 clinically relevant severity of depressive symptoms.

Individual quality of life. The schedule for the Evaluation of Individual Quality of Life-Direct Weighting (SEIQoL-DW) [18] allows the patients to propose five areas of their live which they consider most relevant for their QoL. They then indicate the relative importance of each area and then the degree of satisfaction with each of the areas. The resulting SEIQoL index (see results) may range from 0 (worst possible QoL) to 100 (best possible QoL).

Statistical analysis

Normal distribution of the data was tested with Kolmogorov-Smirnov Test. We report the median (*Mdn*) when data were not normally distributed. With continuous data correlational analyses were performed to examine whether the dependent and independent variables varied as a function of the background data. Mann-Whitney *U*-test and Kruskal-Wallis *H*-test were applied to categorical data with the same purpose. Nonparametric tests were used with regard to the small sample size and when data were not normally distributed. To compare two variables within the same participants paired samples t-test or *Z* Wilcoxon signed-rank test were used. For the comparison of more than two conditions in the same participants, ANOVA or Friedman's ANOVA was computed. To evaluate the relative impact of the predictor variables on the measures of psychosocial adjustment, two multiple regression analyses were conducted separately for depression and QoL scores.

Each predictor was assessed through multiple subscales and thus the predictors-sample size ratio was inadequate for regression modelling (the predictors exceeded the sample size). To

reduce the number of predictors multiple linear regressions (stepwise) were first performed with the subscales of each perdictors variable separately. Those subscales that showed an adjusted (Bonferroni corrected at p< 0.01) significant contribution to the prediction of the dependent variable were included in the final regression. Analysis of the model assumptions (i.e., independence of the residuals, absence of the outliers, multicollinearity, linearity and homoscedasticity) was performed. We used SPSS 15.0 for all statistical analyses.

RESULTS

Background data and adjustment outcomes

Background and disease related data are listed in Table 1.

Insert Table 1 about here.

The group's BDI mean score was 11.8 (SD \pm 6.9, range 1-29) and 5 patients (18.5%) had clinically relevant depressive symptoms. Mean score of individual QoL was 67.8 (SD \pm 15.8, range 32-91). Age correlated significantly positive with QoL (r = 0.49, p < 0.01, Pearson's correlation), but not with depression (r = 0.09, p = 0.7, Pearson's correlation). Depression and QoL did not differ between sexes (depression: U = 71, p = 0.3; QoL: U = 57.5, p = 0.1; Mann -Whitney U-tests) and as a function of education (depression: U = 68, p = 0.2; QoL: U = 56, p = 0.09; Mann-Whitney U-tests). Married patients and patients living in partnership reported significantly higher QoL as compared to patients living without a partner (U = 15.5, p = 0.01; Mann -Whitney U-tests). There was no difference between these groups regarding depression scores (U = 36.5, p = 0.2; Mann -Whitney U-tests).

Predictors of psychosocial adjustment: descriptive and comparative analysis

Mean, median, standard deviation and range for all predictor variables are listed in Table 2. With a mean score of 17.4 ALS-Functional Rating Scale (ALS-FRS) values indicated high physical impairment of the patient sample. Correlation analysis revealed that neither severity of depressive symptoms nor QoL correlated significantly with time since diagnosis and the degree of physical impairment. To compare patients with and without life-sustaining treatment Mann-Whitney *U*-tests were conducted and proved non-significant. For all coefficients and probabilities see Table 3.

Scores of received and perceived social support indicated a high availability of social support in this sample (Table 2). Median of primary appraisals evidenced that patients rated their illness situation as being not threatening (Table 2). Significant mean differences were found between the appraisals of problem-focused coping potential and emotion-focused coping potential ($t_{(26)} = -4.5$, p < 0.001, Paired sample t test), meaning that patients believed to deal better with the situation emotionally than by active attempts to change it. However, patients rated both coping potentials as being high, indicating that they experienced a high propensity to cope well with their disease.

Insert Table 2 and Table 3 about here

Friedman's ANOVA revealed significant differences between the frequencies of used coping strategies ($\chi^2_{(3)} = 45.2$, p < 0.0001 Friedman's ANOVA). Wilcoxon signed-rank tests were used for pair-wise comparisons (Bonferroni corrected) and revealed that emotional avoidance was less frequently used then all the other coping strategies (all p < .001). Moreover, patients relied less frequently on problem-management and problem-appraisal strategies than on

emotion-management strategies (Z = -2.7, p < 0.008; Z = -2.6, p < 0.008; Wilcoxon signed rank tests).

Data reduction and selection of predictors

Regression analyses were conducted separately for depression and QoL with the subscales (aspects) of each stress-coping model predictor to (1) investigate the relationships between the aspects of the predictor variables and adjustment outcome and (2) to reduce and select predictors for the final regression analysis. The coefficients are presented in Table 4.

Illness characteristics

Regression analysis revealed that neither severity of depressive symptoms nor QoL could be significantly predicted by the time since diagnosis and the degree of physical impairment (Table 4) and thus these parameters were not included in the final regression.

Social Support

The subscales on *Received social support* and *Perceived social support* of BSSS were included in the regression analysis because these subscales are geared toward assessment of social support as a coping *resource*. The other scales of the instrument were related to coping behaviour and thus, were not included in the final regression to avoid overlap with the coping strategy subscales. Results showed that higher perceived social support could significantly predict lower depression and higher quality of life (Table 4). Thus the scale on *Perceived social support* was selected as a predictor for the final regression.

Coping Strategies

Higher scores on the scale *Independence* significantly predicted lower severity of depressive symptoms (Table 4). Higher QoL scores were significantly predicted by *problem-management* and *avoidant type of strategies*. The *emotion-management strategy* (independence), for depression, and the *problem-management* and *avoidant type of strategies*, for QoL, were included in the final regression analyses.

Cognitive appraisal

The results of the regression analysis showed that higher appraisals of both emotion and problem focused coping potential, which can be interpreted as higher internal locus of control, predicted lower severity of depressive symptoms (Table 4). The variance of the QoL scores could not be significantly accounted for by any of the appraisals scales.

Insert Table 4 about here

Regression analysis

A multiple regression analysis was performed on each of the two dependent variables. For depression the block of predictors included perceived social support, appraisal of coping potential, and emotion management coping (independence scale of the MNDCS). For QoL we included perceived social support, problem-management coping strategies (information seeking and support) and avoidance. Since no hierarchical assumptions about these predictors were made, all of them were entered in one step using the method *Enter*. A summary of the regression analyses is presented in Table 5. Assumptions (see methods) were tested and met. The regression analysis showed that 61% of the variance of severity of depressive symptoms was accounted for by all the predictor variables taken together. The Beta values (Table 5)

Hence, perceived social support and appraisal of coping potential individually accounted for a significant percentage in the variance in severity of depressive symptoms. Higher perceived

indicated the independent contribution of each predictor to the total explained variance.

social support ($t_{(27)}$ = -2.28, p < 0.05) and more confidence in their own coping potential ($t_{(27)}$

= -2.49, p < 0.05) predicted lower depressed mood. None of the coping strategies predicted

the severity of depressive symptoms.

Selected predictors explained 56% of the variance in QoL ($F_{(3,23)} = 9.87$, p < 0.001). Beta values indicated that higher perceived social support ($t_{(27)} = 3.54$, p < 0.01) predicted higher QoL. Patients who searched more frequently for information and support ($t_{(27)} = 2.5$, p < 0.05) and showed more avoidant behaviour ($t_{(27)} = 3.11$, p < 0.05) also reported higher QoL. However, these relationships did not reach significance at the corrected p value. The results of

Insert Table 5 and Figure 1 about here

the final regression analysis are depicted in Figure 1.

DISCUSSION

Perceived social support, appraisal of coping potential and independence all together explained 61% of the variance of severity of depressive symptoms. Perceived social support, seeking for support and information and avoidance accounted for 56% of the variance in QoL. The degree of physical impairment and the time since diagnosis did not explain any variance in the adjustment outcomes. This result is of utmost importance as it corroborates that more severe functional impairment does not inevitably lead to poorer quality of life and depressive symptoms.

Earlier studies on psychosocial aspects in ALS report similar outcomes but mostly as a result of correlative analyses [19-22]. In contrast, our study revealed the predictive power of social

support, coping appraisal, and coping strategies. Only few studies aimed at predicting psychological outcomes in ALS. The outcomes referred to psychological distress (depressive symptoms and anxiety), self-esteem [23, 24] and hopelessness [25]. We are aware of only one study trying to predict the individual QoL among ALS Patients [26]. Results of these studies are difficult to compare since they investigated different sets of predictors, such as religiosity, demographics, self-perceived quality of social support, present marital intimacy, which were not selected on the basis of a model to cope with disease. However, the quality of social support was evident in predicting both QoL and depressive mood, suggesting that received quality of care and the extent of how patients evaluate their social network as being supportive are of utmost importance for patients' affective state and QoL [23, 26, 27]. Our findings serve to further qualify this statement, by demonstrating that perceived social support was the strongest predictor of QoL and severity of depressive symptoms. These results are in line with the stress and coping theory, which was originally proposed by Lazarus & Folkman [7] and has been repeatedly adapted for different chronic illnesses [28-30]. These models posit that appraisal determines the level of stress experienced. Higher perceived social support is presumed to interact with one's appraisal of the stressful situation to reduce the effects of stress and promote well-being. The loss of or difficulty in communication and mobility, which may lead to limited social interaction are examples of the impairment imposed by ALS/MND that could decrease perceived social support. Comprehensive care of patients with ALS/MND should therefore include measures that focus on maintenance of social network by providing novel communication devices and proper assistive technology. This would allow the patients to preserve their active role in the family and society.

We further found that appraisal of emotion- and problem -focused coping potential contributed significantly to the prediction of the severity of depressive symptoms in ALS, illustrating that higher internal locus of control predicts lower severity of depressive symptoms. Cognitive appraisals might have weaker effect on QoL because they rather

influence emotion and subsequent coping behaviour [12]. QoL is a more complex psychological construct which cannot be reduced to emotional dimensions alone. QoL was better predicted by coping strategies. Searching for information and support as well as avoidance were relevant predictors of QoL such that more support and information seeking behaviour and more emotional avoidance predicted higher QoL. These results suggest that asking for instrumental and emotional support (e.g., emotional intimacy and comfort; maintaining mobility with walking sticks / wheelchair) and receiving more information about the illness condition lead to higher QoL.

Although it is commonly assumed that 'confrontation is good and avoidance is bad' [31, 32] several studies indicated that the effects of approach and avoidance coping may vary as a function of the type of problem [33] or of the stages of the illness [34]. Studies of coping among cancer and HIV positive patients showed that denial (avoidant type of coping) may reduce negative emotional reactions in the early stages but may have maladaptive consequences if it leads to the postponement of medical care [34]. The degenerative course of ALS exposes patients to short and long term stressors which require different strategies depending on the specific stressful aspect of the illness with which they are coping at any one time. Our results suggest, that the combination of confronting and avoiding coping strategies might be beneficial for ALS patients, because more frequent search for information and for support may on the one hand help them to initiate actions which are necessary to ensure optimal future care (e.g., clarify support by the health insurance). On the other hand emotional avoidance through minimizing the importance of the diagnosis or directing attention away from the negative information and consequences that may occur successively as their illness progresses may prevent patients from psychological distress and despair. However, as the disease progresses, avoidance is no longer an adaptive strategy as it prevents patients from taking appropriate measure to cope further with the illness.

Among the sociodemographic variables age and marital status showed a significant relationship with QoL. Older and married ALS patients reported higher individual QoL than younger patients and patients living alone. These results are consistent with findings from studies of other diseases that lead to severe physical impairment (e.g. multiple sclerosis, spinal cord injury, cancer) [35, 36] and studies on healthy subjects [37]. During their life span older people most likely get more experienced in handling stressful situations and thus develop a higher resistance to psychological stress [38] leading to higher scores in QoL as compared with younger people [39].

The main limitations of the present study are the small sample size and the high collinearity between some predictor variables which did not allow us to include all the subscales in the regression analysis. Nevertheless, we succeeded in defining predictors of psychosocial adjustment (see Figure 1). Taking into consideration at least one dimension of each stress coping predictor from Lazarus and Folkman's model we could show that not illness related characteristics but the psychosocial variables, first and foremost social support, best predicted psychosocial adaptation to ALS.

In summary, the present study contributes to a better understanding of psychosocial adjustment to ALS as it elucidates predictive relationships between the model variables: illness related parameters, coping strategies, cognitive appraisals and social support and the outcome variables depression and quality of life. However, more theory driven and longitudinal research is necessary to clarify which factors have the most powerful influence on psychosocial adjustment. Such an approach would allow us to suggest and test interventions for those patients who poorly cope with the disease.

ACKNOWLEDGMENTS

We thank patients and caregivers for their readiness to participate in the study. We are grateful to Profs. Christoph Braun and Boris Kotchoubey for their support in statistical analysis of the data and to Adrian Furdea for designing and making the figures.

COMPETING INTERESTS

All authors declare that the answers to the questions on your competing interest form are all 'No' and therefore have nothing to declare.

Signatures
Tamara Matuz
Dr. Andrea Kuebler Andrea Edillo
Prof. Dr. Niels Birbaumer
Prof. Dr. Martin Hautzinger
- 1

FUNDING

This study was supported by the Deutsche Forschungsgemeinschaft (DFG) through the Research Training Group "Bioethics" at the Interdepartmental Centre for Ethics in the Sciences and Humanities (Interfakultären Zentrum für Ethik in den Wissenschaften, IZEW) of the University of Tübingen. Dr. Matuz received research support in form of a PhD scholarship for three years.

COPYRIGHT LICENCE STATEMENT

The corresponding author, Tamara Matuz, grants on behalf of all authors, an exclusive licence on a worldwide basis to the BMJ Publishing Group Ltd, and its Licensees to permit this article (if accepted) to be published in BMJ editions and any other BMJPGL products and to exploit all subsidiary rights, as set out in their licence.

Signature

Thatus

REFERENCES

- [1] Livneh H, Antonak RF. Psychosocial adaptation to chronic illness and disability. Gaithersburg: MD: Aspen 1997.
- [2] McLeod JE, Clarke DM. A review of psychosocial aspects of motor neurone disease. Journal of the neurological sciences. 2007 Jul 15;258(1-2):4-10.
- [3] Lule D, Kurt A, Jurgens R, Kassubek J, Diekmann V, Kraft E, et al. Emotional responding in amyotrophic lateral sclerosis. Journal of neurology. 2005 Dec;252(12):1517-24.
- [4] Lule D, Diekmann V, Anders S, Kassubek J, Kubler A, Ludolph AC, et al. Brain responses to emotional stimuli in patients with amyotrophic lateral sclerosis (ALS). Journal of neurology. 2007 Apr;254(4):519-27.
- [5] Lillo P, Hodges JR. Frontotemporal dementia and motor neurone disease: overlapping clinic-pathological disorders. J Clin Neurosci. 2009 Sep;16(9):1131-5.
- [6] Kurt A, Nijboer F, Matuz T, Kubler A. Depression and anxiety in individuals with amyotrophic lateral sclerosis: epidemiology and management. CNS Drugs. 2007;21(4):279-91.
- [7] Lazarus RS, Folkman S. Stress, appraisal and coping. New York: Springer 1984.
- [8] Pakenham KI. Adjustment to multiple sclerosis: application of a stress and coping model. Health Psychol. 1999 Jul;18(4):383-92.
- [9] Pakenham KI, Dadds MR, Terry DJ. Relationships between adjustment to HIV and both social support and coping. J Consult Clin Psychol. 1994 Dec;62(6):1194-203.
- [10] Anderson NB. Encyclopedia of health and behavior. California: Sage publications 2004.
- [11] Cedarbaum JM, Stambler N, Malta E, Fuller C, Hilt D, Thurmond B, et al. 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. 1999 Oct 31;169((1-2)):13-21.

- [12] Smith CA, Lazarus RS. Appraisal components, core relational themes and the emotions. Cognition and emotion. 1993;7(3/4):233-69.
- [13] Schulz U, Schwarzer R. Soziale Unterstützung bei der Krankheitsbewältigung. Die Berliner Social Support Skalen (BSSS). Diagnostica. 2003;49(2):73 82.
- [14] Terry DJ, Hynes GJ. Adjustment to a low-control situation: Reexamining the role of coping responses. Journal of personality and social psychology. 1998;74(4):1078-92.
- [15] Osowiecki DM, Compas BE. A prospective study of coping, percieved control and psychological adaption to breast cancer. Cognitive therapy and research. 1999;23(2):169-80.
- [16] Lee JN, Rigby SA, Burchardt F, Thornton EW, Dougan C, Young CA. Quality of life issues in motor neurone disease: the development and validation of a coping strategies questionnaire, the MND Coping Scale. Journal of the neurological sciences. 2001 Oct 15;191(1-2):79-85.
- [17] M. Hautzinger MB, H. Worrall, F. Keller. Das Beck Depressions-Inventar (BDI). 2 Auflag ed. Bern: Huber 1995.
- [18] Hickey AM, Bury G, O'Boyle CA, Bradley F, O'Kelly FD, Shannon W. A new short form individual quality of life measure (SEIQoL-DW): application in a cohort of individuals with HIV/AIDS. BMJ (Clinical research ed). 1996 Jul 6;313(7048):29-33.
- [19] Kübler A, Winter S, Ludolph AC, Hautzinger M, Birbaumer N. Severity of depressive symptoms and quality of life in patients with amyotrophic lateral sclerosis. Neurorehabil Neural Repair. 2005 Sep;19(3):182-93.
- [20] Hammer EM, Hacker S, Hautzinger M, Meyer TD, Kubler A. Validity of the ALS-Depression-Inventory (ADI-12)- A new screening instrument for depressive disorders in patients with amyotrophic lateral sclerosis. J Affect Disord. 2008 Feb 7.
- [21] Rabkin JG, Albert SM, Del Bene ML, O'Sullivan I, Tider T, Rowland LP, et al.

 Prevalence of depressive disorders and change over time in late-stage ALS. Neurology. 2005

 Jul 12;65(1):62-7.

- [22] Simmons Z, Bremer BA, Robbins RA, Walsh SM, Fischer S. Quality of life in ALS depends on factors other than strength and physical function. Neurology. 2000 Aug 8;55(3):388-92.
- [23] Goldstein LH, Atkins L, Landau S, Brown RG, Leigh PN. Longitudinal predictors of psychological distress and self-esteem in people with ALS. Neurology. 2006 Nov 14;67(9):1652-8.
- [24] Hogg KE, Goldstein LH, Leigh PN. The psychological impact of motor neurone disease. Psychological medicine. 1994 Aug;24(3):625-32.
- [25] Plahuta JM, McCulloch BJ, Kasarskis EJ, Ross MA, Walter RA, McDonald ER. Amyotrophic lateral sclerosis and hopelessness: psychosocial factors. Social science & medicine (1982). 2002 Dec;55(12):2131-40.
- [26] Chio A, Gauthier A, Montuschi A, Calvo A, Di Vito N, Ghiglione P, et al. A cross sectional study on determinants of quality of life in ALS. Journal of neurology, neurosurgery, and psychiatry. 2004 Nov;75(11):1597-601.
- [27] Hecht M, Hillemacher T, Grasel E, Tigges S, Winterholler M, Heuss D, et al. Subjective experience and coping in ALS. Amyotroph Lateral Scler Other Motor Neuron Disord. 2002 Dec;3(4):225-31.
- [28] Pakenham KI. Adjustment to multiple sclerosis: application of a stress and coping model. Health Psychology. 1999 Jul;18(4):383-92.
- [29] Pakenham KI, Dadds MR, Terry DJ. Relationships between adjustment to HIV and both social support and coping. Journal of Consulting and Clinical Psychology. 1994 Dec;62(6):1194-203.
- [30] Stanton AL, Snider PR. Coping with a breast cancer diagnosis: a prospective study. Health Psychology. 1993 Jan;12(1):16-23.
- [31] Roth S, Cohen LJ. Approach, avoidance, and coping with stress. Am Psychol. 1986 Jul;41(7):813-9.

- [32] Holahan CJ, Moos RH. Life stressors, personal and social resources, and depression: a 4-year structural model. Journal of abnormal psychology. 1991 Feb;100(1):31-8.
- [33] Stanton AL, Danoff-Burg S, Cameron CL, Ellis AP. Coping through emotional approach: problems of conceptualization and confounding. Journal of Personality and Social Psychology. 1994 Feb;66(2):350-62.
- [34] Aldwin CM, Revenson TA. Does coping help? A reexamination of the relation between coping and mental health. Journal of personality and social psychology. 1987 Aug;53(2):337-48.
- [35] Waldron D, O'Boyle CA, Kearney M, Moriarty M, Carney D. Quality-of-life measurement in advanced cancer: assessing the individual. Journal of Clinical Oncology. 1999 Nov;17(11):3603-11.
- [36] Hammell KW. Exploring quality of life following high spinal cord injury: a review and critique. Spinal Cord. 2004 Sep;42(9):491-502.
- [37] Browne JP, O'Boyle CA, McGee HM, Joyce CR, McDonald NJ, O'Malley K, et al. Individual quality of life in the healthy elderly. Quality of Life Research. 1994 Aug;3(4):235-44.
- [38] Kunzmann U. Wisdom Adult development and emotional-motivational dynamics. In: Fernández-Ballesteros, ed. *Geropsychology*. Göttingen: Hogrefe 2007:224-38.
- [39] Lulé D, Häcker S, Ludolph. A, Birbaumer N, Kübler A. Depression und Lebensqualität bei Patienten mit amyotropher Lateralsklerose. Deutsche Ärzteblatt. 2008;105(23):397-403.

Tables

Table 1 Background and disease related data of the patient sample.

Education		Marital		Living		C			
Level		Status		Environment		Sex		Communication *	
Higher	13	Married	21	Own household	25	Male	15	Normal	7
Lower	14	Single	3	Nursing home	2	Female 12		Impaired	15
		Widowed	2					With devices	3
		Divorced	1						
Onset of ALS		Type of ALS		Artificial Ventilation		Invasive Ventilation		PEG	
Bulbar	2	Sporadic	27	Yes	11	Yes	3	Yes	6
Spinal	25	Genetic	0	No	18	No 24		No	21

^{*} Normal communication refers to normal speech process; impaired communication ranged from "detectable speech disturbance" to "communication intelligible with repeating"; with devices refers to loss of useful speech and communication exclusively possible by using assistive technology and/or eye movements.

Table 2 Predictor variables: Mean (M), Median (Mdn), standard deviation (SD), range, maximum possible values and group differences depending on the time since diagnosis.

Predictors of Psychosocial Adjustment		M/	CD	D	Maximum		
		Mdn	SD	Range	possible		
SS	Illness Charac teristics		Physical Impairment	17.4	9.8	0 -36	40
Illne			Time Since Diagnosis	36		4 -129	-
			Perceived	4		2.1 – 4	4
	ort		Received	3.9		1.9 - 4	4
	Social Support		Need for Support	2.9	0.7	1.2 - 4	4
	Social		Search for Support	2.8	0.6 1.2 - 4		4
			Protective Buffering	2.2	0.6	1.1 - 3.5	4
Cognitive			Primary Appraisal	2.6	3.2	1 - 8	8
		praisais	Appraisal of Coping	5.8	1.7	1 - 9	9
7	Co	Potential	3.0	1.,	1 /	,	
			Problem Management	4.1		2.1 - 5	5
ategies			Problem Appraisal	4.2		2 - 5	5
Coping Strategies	Emotion Management	4.5		3.5 - 5	5		
	Emotional Avoidance		2.3	0.8	0.6 - 4	5	

Table 3 Adjustment outcomes and illness parameters; r Pearson's correlation; τ Kendall's correlation; U Mann-Whitney U-Test

Illn	000	Parameters
		r at attieters

Physical Time Since Artificial Artificial Invasive Impairment Diagnosis Nutrition Ventilation Ventilation r = -0.2U = 73U = 28.5 $\tau = 0.01\,$ U = 61Depression p = 0.3p = 0.9p = 0.9p = 0.4p = 0.5 $\tau = 0.14\,$ U = 57U = 82U = 18r = 0.01QoL p = 0.9p = 0.2p = 0.7p = 0.7p = 0.1

Adjustment Outcomes

Table 4 Summary of regression analyses on the two adjustment variables with the subscales of each stress-coping predictors separately. $\Delta R^2 = R$ squared change; B = b-values; SEB = standard error of the b-values; g-standardized Betas

	Dependent Variables	Depress	ion		Quality of Life			
Predictors		В	SE B	В	В	SE B	ß	
Social Support		_						
	Perceived	-5.5	3.03	54**	14.3	4.4	.44**	
	Received	-2.9	2	24	4.9	6.7	.23	
		$R^2 = .33$	$\Delta R^2 = .28$		$R^2 = .3$	$37, \Delta R^2 =$.32	
		F (2,24)=	6.03, p< 0.0	1	$F_{(2,24)} = 7.2, p < 0.01$			
Cognitive Appraisal								
	Primary Appraisal	24	.42	11	-0.2	1.1	004	
	Appraisal of Coping Potential	-2.4	.77	56**	1.33	2.1	.15	
		$R^2 = .31$, $\Delta R^2 = .26$		$R^2 = .0$	$\Delta R^2 =$.06.	
		F (2,24)=	5.6, p<0.01		F _(2,24) = 0.02, p= 0.8			
Coping Strategies								
	Problem Management	-1.6	1.6	22	9.57	3.9	.44**	
	Problem Appraisal	-2.1	1.3	32	2.67	3.1	.15	
	Emotion Management	-5.1	2.7	53**	-8.74	6.4	26	
	Emotional Avoidance	62	1.4	11	7.76	3.4	.39*	
		$R^2 = .40$	$\Delta R^2 = .29$		$R^2 = .3$	88, $\Delta R^2 =$.28.	
		F (4,22)=	9.8, p<0.01		F (4,22)	= 3.48, p<	0.05	
Illness Parameters								
	Physical Impairment	12	.15	17	.08	.34	0.05	
	Time Since Diagnosis	.02	.05	.07	.06	.11	0.11	
		$R^2 = .04, \ \Delta R^2 = .03. \qquad \qquad R^2 = .01, \ \Delta R^2 = .07.$.07.	
		F _(1,24) =	0.04, p= 0.6		$F_{(1,24)} = 0.1, p = 0.9$			

^{*}p<.05, **p<.01

Table 5 Summary of regression analyses of stress-coping variables predicting psychological adjustment to ALS. $\Delta R^2 = R$ squared change; B = b-values; SEB = standard error of the b values; β standardized Betas.

	Dependent Variables	Depres	Depression		Quality of Life		
Predictors		В	SE B	В	В	SE B	ß
Social Support							
	Perceived Social Support	77	.24	43**	2.01	.57	.49**
Cognitive Appraisal							
	Appraisal of Coping	-1.68	54	43**			
	Potential		.54				
Coping Strategies							
	Problem Management	-3.62	2.15	25	6.22	3.09	.29*
	Emotion Management						
	Emotional Avoidance				6.71	2.72	.34*
		$R^2 = .61, \ \Delta R^2 = .56$		$R^2 = .56, \ \Delta R^2 = .51$			
		F _(3,23) =11.8, p=0.0001			$F_{(3,23)} = 9.8, p = 0.0001$		

^{*}p<.05, **p<.01

Figure captions

Figure 1. The modified model of psychosocial adjustment to ALS on the basis of multiple regression analyses. The psychosocial predictor variables accounted together for a considerable amount of variance in both outcomes of psychosocial adjustment (depression and QoL). This is illustrated by the dashed line. Variables which displayed independent predictive power are denoted by the arrows.

Figure 1

