

**ALS Specific Quality of Life- Short Form (ALSSQOL-SF): A Brief, Reliable and Valid
Version of the ALSSQOL-R**

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ABSTRACT

Introduction: The ALS Specific Quality of Life instrument and its revised version (ALSSQOL, ALSSQOL-R) have strong psychometric properties, and have demonstrated research and clinical utility. The current study aimed to develop a short form (ALSSQOL-SF) suitable for limited clinic time and patient stamina. *Methods:* The ALSSQOL-SF was created using Item Response Theory and confirmatory factor analysis on 389 patients. A cross-validation sample of 162 patients assessed convergent, divergent, and construct validity of the ALSSQOL-SF compared to psychosocial and physical functioning measures. *Results:* The ALSSQOL-SF consisted of 20 items. Compared to the ALSSQOL-R, optimal precision was retained, and completion time was reduced from 15-25 minutes to 2-4 minutes. Psychometric properties for the ALSSQOL-SF and its subscales were strong. *Discussion:* The ALSSQOL-SF is a disease-specific global QOL instrument that has a short administration time suitable for clinical use, and provides clinically useful, valid information about persons with ALS.

Keywords: Amyotrophic lateral sclerosis, ALS, Quality of life, Psychometrics, ALSSQOL, assessment,

INTRODUCTION

Quality of life (QOL) is a broad term with many operational definitions. Health-related QOL, or health status, is most commonly used to describe an individual's perception of their well-being secondary to their physical and mental functioning^{1,2}. However, QOL is often conceived more broadly as being impacted by many factors other than the physical or mental health domains, such as family and friends, finances, religion and spirituality, and existential concerns³. When defined in this manner, overall QOL in patients with amyotrophic lateral sclerosis (ALS) does not correlate with physical function and appears to be maintained over time as physical function declines⁴⁻¹⁰. Baseline and ongoing assessments provide insight into factors that impact QOL, and that may guide multidisciplinary interventions for ALS patients. One instrument that captures this global self-perception of QOL is the 59-item ALS Specific Quality of Life Instrument (ALSSQOL). This questionnaire, and its shorter, revised version, the ALSSQOL-R, have been validated in multicenter studies¹¹⁻¹³. The ALSSQOL-R is a 50 item (46 scored) instrument that measures overall QOL with 6 subscales (Negative Emotion, Interaction with People and the Environment, Intimacy, Religiosity, Physical Symptoms, and Bulbar Function) and an Average Total QOL score. Every subscale consists of several items, each of which is rated by the patient on a 0 to 10 point Likert-type scale (0 represents least desirable; 10 is most desirable). Average Total QOL score is the average of all items, and ranges from 0 (worst QOL) to 10 (best QOL).

Completion of the ALSSQOL-R by persons with ALS takes approximately 15-25 minutes by interview. Fatigue may make an assessment of this length highly taxing or burdensome for some

patients, particularly in view of research showing that patients already cite the length of visits as a negative feature of multidisciplinary ALS clinic¹⁴. In addition, it may be challenging for busy ALS clinic personnel to find time to administer the questionnaire. In view of the utility of the ALSSQOL-R and these limiting factors, we sought to develop and validate a shorter version, the ALSSQOL-Short Form (ALSSQOL-SF).

METHODS

For all studies and phases, Institutional Review Board (IRB) approval was obtained from all participating institutions, and all participants provided informed consent.

Phase 1: Creation of the ALSSQOL-SF

Patients: Phase 1 was a cross-sectional study using data from a validation study of the ALSSQOL-R, collected between 2005 and 2008¹³. All participants had definite, probable, probable laboratory-supported, or possible ALS¹⁵, were 18 years of age or older, and fluent in English at the 6th grade level or higher. Patients were excluded if a physician or psychologist at the center judged them to have cognitive impairment sufficient to preclude the granting of informed consent and participation in the study.

Item Response Theory Methodology: Item Response Theory (IRT) analyses were conducted to inform ways of shortening the ALSSQOL-R with minimal loss of information¹⁶. IRT is a test design, analysis, and scoring approach that utilizes statistical modeling to distinguish between the characteristics of the person from the characteristics of the items to which they responded¹⁶. IRT is useful in test

construction by using item level information to identify items that best suit the purpose for a given measure. In an IRT context, information is equivalent to the concept of reliability in classical test theory, except that it can vary. In IRT the precision of an item or measure depends on the location of the individual on the trait scale whereas reliability makes an assumption that error is constant. Using IRT to build a measure involves defining the ideal information function and then selecting items that best approximate that shape¹⁷. In designing a short form, the ideal information function is the information function for the original form. Thus, IRT can be effective in creating a short form that provides similar information across the trait scale.

To provide guidance about which items should be removed from the ALSSQOL-R to create the ALSSQOL-SF, item parameters for each of the 6 subscales were estimated with the Modified Graded Response Model (M-GRM)¹⁸, using Parscale¹⁹. The slope parameter (α) is an indicator for discrimination power, the extent to which the item can distinguish between individuals with low and high trait levels whereas the location parameter (β) indicates the targeted trait value²⁰. Items were selected based on (i) large α parameters, (ii) relevance to the construct the item was intended to measure, and (iii) contribution to the shape of the information function for the particular subscale.

Confirmatory Factor Analysis: A confirmatory factor analysis (CFA) was conducted with Amos 21²¹ to test the fit of the short form to the original factor structure in order to substantiate construct validity. Using the retained items from the IRT analysis, the hypothesized model was fit to the data using maximum likelihood estimation. Model data fit was analyzed using the comparative fit index (CFI²²), the root mean square error of approximation (RMSEA²³), and the standard root mean square

residual (SRMR). The literature suggests CFI > .95, RMSEA < .06, and SRMR < .08 for good fit and CFI > .90, RMSEA < .10, and SRMR < .10 for acceptable fit^{22, 24, 25}.

Phase 2: Cross-Validation Sample

Once the ALSSQOL-SF was created, a prospective study of ALS patients attending 10 multidisciplinary clinics was conducted to test its validity. Inclusion and exclusion criteria were identical to those in Phase 1. Study measures were completed onsite via interviews with patients by ALS clinic personnel who were part of the site study team. Patient enrollment and data collection were performed from January through December 2014.

Participant demographics and descriptive information were recorded, including the ALS Functional Rating Scale-Revised (ALSFRS-R).²⁶ After the completion of the ALSSQOL-SF, participants completed the following measures to evaluate the construct, convergent, and discriminant (divergent) validity of the overall ALSSQOL-SF and its subscales. These measures were selected to replicate previous methodology used for the development of the ALSSQOL-R, and were determined to be reliable and valid^{11,27-29}: The McGill Quality of Life Single Item Score (MQOL-SIS)³⁰, modified to assess QOL over the past 7, rather than 2 days, appears on the ALSSQOL-SF as the first question, though scored separately. This measure was used for testing of validity of the ALSSQOL-SF's assessment of global QOL. The World Health Organization Quality of Life Instrument-Brief version (WHOQOL-BREF)³¹ domain scores (Psychological, Social Relationships, Environment, Physical Health) were compared to the ALSSQOL-SF Average Total Score or to subscales as follows: Psychological domain was compared with the Negative Emotion subscales; Social Relationships domain

was compared with the Interaction with People and the Environment and Intimacy subscales; No specific comparison measure of intimacy was used, because most measures require information from both partners; Environment domain was compared with Interaction with People and the Environment subscale; and Physical Health domain was compared with Physical Symptoms subscale. The Idler Index of Religiosity³² (IIR), used to evaluate the ALSSQOL-SF Religiosity subscale, has two subscales - Public Religiousness and Private Religiousness. The Satisfaction with Life Scale³³ (SWLS) was included to evaluate the ALSSQOL-SF Interaction with People and Their Environment subscale. The Center for Epidemiological Studies- Depressed Mood Scale³⁴ (CES-D) was compared to the Negative Emotion subscale. The Brief Symptom Inventory-18³⁵ (BSI) also was compared to the Negative Emotion subscale.

Statistical Analyses

Descriptive statistics were conducted using means and frequencies. Some demographic data was not completed by the participants, and was de-identified before it could be retrieved, resulting in minor variation in sample size for demographic data. The primary analysis of the study used CFA to verify the structure and content of the ALSSQOL-SF. The relationships between subscales of the ALSSQOL-SF and external measures of the same constructs were evaluated using Pearson Product-Moment Correlations. Hierarchical linear regression was used to evaluate the subscales' ability to predict global QOL. All analyses were conducted with SPSS 22.0 statistical software³⁶ and AMOS²¹. Probability values less than 0.05 were considered significant. A minimal sample size of 200 was determined to be sufficient to conduct IRT, and greater than needed to achieve 0.80 power with a medium effect size and

alpha of 0.01 for all remaining analyses. 200 subjects is considered a small but reasonable sample for IRT³⁷; a larger sample would have been more difficult to obtain from this population within the planned timeframe for this study. The most important factor in determining the appropriate sample size for this analysis is dependent on how much information can be extracted from the data, and the standard error for the estimates parameters for the information functions to show that they had similar shapes, and this was accomplished here.

RESULTS

Phase 1: Creation of the ALSSQOL-SF

There were 389 participants (59% male). Ages ranged from 27 to 88 years (mean 61, SD 11). Duration of symptoms at study enrollment averaged 39.7 months (SD 39.2; median 26; range 3 to 232; N=324). Length of time from diagnosis to study enrollment averaged 21.5 months (SD 27.0; median 12.5; range = 0 to 223; N= 362). ALSFRS-R scores (N=385) ranged from 6 to 48 (mean 33.11, SD 8.0).

Supplementary Table 1 presents the estimated α and β parameters for the ALSSQOL-R. Figure 1 presents the final information functions for each subscale. The findings across the 6 graphs indicate that the ALSSQOL-SF has slightly greater imprecision compared to the ALSSQOL-R, though that is expected given that the ALSSQOL-SF is composed of fewer items. Table 1 provides a structural comparison between the original and short form. Twenty items were retained, reducing the length of the ALSSQOL-R by more than 50%. The Physical Symptom subscale was altered the least, with only 1 item removed, given that any further reductions to the original scale would have reduced reliability

below acceptable levels. Negative Emotion and Interaction with People and the Environment subscales were reduced the most.

For the CFA, the hypothesized model is displayed in Figure 2, using the retained items from the IRT analysis. The CFA results found the model fit to be acceptable: $\chi^2_{(164)} = 527.20$, $p < .001$; CFI = .876; RMSEA = .076, SRMR = .078. Table 2 shows the factor loadings by item for the ALSSQOL-SF. All items loaded onto their respective factors as suggested by the model, supporting the same factor structure as the original ALSSQOL-R. Correlations between factors (Table 3) were mostly low, ranging from 0.06 to 0.42, and were similar to results for the ALSSQOL-R.¹³ Table 3 represents the intercorrelations among the factors in the ALSSQOL-SF, and is designed to show that the results of the original Principal Component Analysis with Orthogonal Varimax rotation that was conducted to reduce the ALSSQOL to the ALSSQOL-R¹³, retaining the individuality of the factors, remained intact when the ALSSQOL-R was further reduced to the ALSSQOL-SF. As such, the correlations between subscales are generally low to not significant.

A copy of the current version of the ALSSQOL-SF is presented as supplementary material.

Phase 2: Cross-Validation Sample

There were 162 participants. The majority were male (59.3%; N=96), with a mean age of 60.5 years (SD=10.99, range 26 to 85, N=154). The mean ALSFRS-R score was 31.08 (SD=9.15; range 1 to 48; median 32). Time from symptom onset to data collection averaged 35.6 months (SD = 34, median = 27.5, range 4 to 273 months), and duration of time from diagnosis to study entry averaged 20.8 months (SD =30.2, median = 13, range 0 to 253 months). Completion of the ALSSQOL-SF required 2-4

minutes. 154 patients completed all items on the ALSSQOL-SF, and were included in the CFA without imputation. Series-mean imputation was used for missing items to allow more complete data for additional analyses.

Evaluation of the ALSSQOL-SF: The initial CFA run estimated a negative error variance for item 12 and as a result the model was unable to converge. A negative error variance may have been caused by the small sample size³⁸, or a Heywood case³⁹ since item 12 is an indicator variable for the religiosity factor which is only composed of 2 items⁴⁰. A Heywood case means the factor is possibly not contributing unique variance. However, the negative error variance was not statistically different from 0 ($p > .05$). Thus, the negative value is most likely due to sampling error as a deficiency in the model and can safely be constrained to a non-negative value to allow the model to converge. After constraining the error variance for item 12 to 0, the model converged with a similar fit to the previous CFA: $\chi^2_{(165)} = 349.92$, $p < .001$; CFI = .857; RMSEA = .083, SRMR = .10.

ALSSQOL-SF Scores: The mean score of all ALSSQOL-SF items (Average Total Score) was 6.5 (SD = 1.1, range 3.7 to 9.4). Scores for subscales are provided in Table 4. Range of scores showed maximum use of the scale (minimum 0, maximum 10) for most of the subscales.

Validity Testing: Correlations are reported in Table 5 for the ALSSQOL-SF Average Total and subscale scores as assessments of convergent, divergent, and construct validity. The ALSSQOL-SF Average Total Score and ALSFRS-R showed a significant, moderate correlation. Moderate, positive, significant correlations were found between the ALSSQOL-SF Average Total Score and other measures of global QOL: MQOLSI, WHOQOL subscales, and SWLS. Hierarchical multiple regression

conducted using MQOL-SIS as the dependent variable, and the subscales of the ALSSQOL-SF as the independent variable revealed only three (Negative Emotion, Interaction with People and the Environment, Physical Symptoms) of the six variables were significant predictors of MQOL-SIS, accounting for 2.7%, 9.6%, 7.8%, respectively ($F(3, 153) = 5.083, p = 0.026$), and a total of 44.7% of the variability. Beta weights are presented in Supplementary Table 2, available on-line.

Correlations of the Negative Emotion subscale with the BSI-18 Depression and Anxiety Subscales, CESD, and WHOQOL-Psychological Domain, were moderate to small. Correlations between Interaction with People and the Environment and the WHOQOL Social Relationships Subscale, WHOQOL Environment Subscale, and SWLS were significant, and moderate to small. No direct measure of intimacy was used. However, the ALSSQOL-SF Intimacy subscale and WHOQOL Social Relationships correlated moderately. Strong, positive, and significant relationships were found between the Religiosity subscale and both the Public Religiosity Subscale of the IIR and the total/overall IIR score.

When assessing correlations of the Physical Symptoms subscale, a positive, moderate correlation was found with the total ALSFRS-R score. A closer comparison of subscales of the ALSFRS-R shows significant correlations among all, but most closely with ALSFRS-R Fine Motor and ALSFRS-R Gross Motor Functioning, less so with ALSFRS-R Speech and Respiratory. The relationship between the ALSSQOL-SF Physical Symptoms and the WHOQOL Physical Domain was strong and positive. Bulbar Function demonstrated strong, positive, and significant correlations with the ALSFRS-R Speech subscale. The relationship between this subscale and the ALSFRS-R Respiratory subscale was

significant, but weak. There was not a significant correlation between the ALSSQOL-SF Bulbar Function and the WHOQOL Physical Domain.

DISCUSSION

The ALSSQOL-R was successfully modified to a briefer version, the ALSSQOL-SF, while maintaining the goal of being a valid, disease-specific QOL instrument. The ALSSQOL-SF retains the value of reflecting areas that patients volunteered as most important to their QOL, thus potentially assisting healthcare professionals with intervention planning and comprehensive inter-professional care. The new short form contains less than half of the number of items of previous versions, which shortened time to completion from 15 to 25 minutes to 2 to 4 minutes.

The results of Phase 1 represent a vast improvement of the model fit compared to the original ALSSQOL-R¹³. The improvement in fit may be due to removal of items that measured different facets of the constructs and/or partially measured other characteristics, decreasing the efficiency of the scale. This improvement resulted in information being maximized in similar regions as in the original form, contributing to optimal precision with fewer items.

We chose to retain a 0-10 rating scale on the ALSSQOL-SF for administration/scoring of the measure because it allows for more sensitivity to subtle changes in clinical symptoms for individuals in a treatment setting. It is also consistent with other scales to which patients are accustomed, such as pain scales. The IRT scoring was used for validation of the measure, but is not implemented in regular use of the measure.. It is likely, however, that the correlation between participant scores on both scales would

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be close to a perfect correlation. Thus, even though there may be less precision in someone's category score, it is leading to the same inference and therefore maintaining score validity. Future studies could evaluate a reduction in scale from 0-10 to 0-5 for clinical and research utility.

Negative Emotion and Interaction with People and the Environment subscales originally had the most items compared to the other subscales. They also had the greatest reduction in items, and therefore, clinically, these domains lost the most information. However, given that these were two of the more stable subscales of the ALSSQOL-R, the shortened versions are sufficient for representing and evaluating these constructs.

Phase 2 demonstrated that the ALSSQOL-SF and its subscales have construct, convergent, and divergent (discriminant) validity. The weak to moderate correlations between the ALSQOL-SF Average Total Score and general global measures of QOL suggests that they measure related but not overlapping constructs.

The significant, yet small to moderate correlations between the Negative Emotion subscale and the BSI-18 and CESD suggest either that negative emotion is more than just anxiety or depression for persons with ALS as previously identified¹². The Negative Emotion subscale appears to be appropriate as a brief screening tool, but additional evaluation of depressed mood, anxiety, coping, and adjustment should be performed if scores on this subscale are low.

The Interaction with People and the Environment subscale represents social relationships and existential considerations such as one's appraisal of and reciprocal relationship with the environment. The moderate to small correlations between this subscale and the WHOQOL Social Relationships,

WHOQOL Environment, and SWLS verify the complexity of these concepts, shedding additional light on topics of importance to ALS patients.

The concept of intimacy is not directly assessed in global or general QOL measures. Measuring intimacy in ALS patients and monitoring its change over time can be an important route of intervention and for problem solving ways to bolster QOL in ALS patients, as it can be achieved despite physical dysfunction and decline.

The Religiosity subscale suggests that engaging in religious practices as part of a community may have intrinsic and social benefits that contribute to overall QOL. However, this does not mean that more is better. Rather, the Religiosity subscale may allow for a more comprehensive needs assessment and may give insight as to whether referrals for religious support are warranted if there is a concordant decline in Religiosity and in the overall QOL or the Negative Emotion subscale.

Concurrent validity of the ALSSQOL-SF Physical Function subscale is supported by the strong, positive, and significant relationship between that scale and the WHOQOL Physical Domain, both of which evaluate patients' perceptions of physical functioning. The convergent and construct validity of the ALSSQOL-SF Bulbar Function Subscale was evident via its correlations with the ALSFRS-R Speech Subscale, whereas the lack of correlations with the ALSFRS-R Fine Motor and Gross Motor subscales and the WHOQOL Physical domain demonstrate appropriate divergent/discriminant validity. This supports Bulbar Function and Physical Symptoms remaining as two distinct subscales of the ALSSQOL-SF. Prior research also supports this distinction, as verbal communication was found to be closely related to QOL⁴¹, whereas overall physical function has often been shown to not correlate with

global QOL.

Comparison of the ALSSQOL-SF to the ALSSQOL-R

In evaluating the ALSSQOL-SF's ability to predict variance in the global QOL measure, MQOL-SIS, the hierarchical multiple regressions for each of the two ALSSQOL measures were similar but not the same. Only 2 of the 3 predictors (Negative Emotions, Physical Symptoms) were shared between the two instruments, and ALSSQOL-R's more expansive Negative Emotion Subscale predicted 37% of the global QOL in the study sample compared to 2.7% of the variance being accounted for by the Negative Emotion Subscale of the ALSSQOL-SF. The comparisons of the two versions of the ALSSQOL further validate the factor structure, and reflect the differences in the information provided by each. Comparisons of the two instruments also reveal that the new measure has weaker correlations on subscales with external measures,¹³ which is not surprising, given the reduction in items. This suggests that the short form subscales may be less reliable stand-alone measures of these constructs than the ALSSQOL-R subscales. The two versions of the measure provide flexibility. For instance, clinicians may choose to use the ALSSQOL-R for the initial evaluation of patients, and conduct follow-up assessments with the short form. Another option is to routinely use the short form for screening and follow-ups, and to administer the ALSSQOL-R subscales only when concern is raised in a specific area. Researchers may prefer the ALSSQOL-R, given its provision of more information and sensitivity to subtle changes in broadly assessed domains. The flexibility in choosing the version is supported the similar overall QOL score obtained using the ALSSQOL-SF (6.5), ALSSQOL-R (6.8), and ALSSQOL (7.1)^{11,13}

There were methodological limitations to this study. Patient samples were drawn from individuals attending large ALS referral centers, and so may not be representative of the ALS patient population as a whole, despite a similar age and gender distribution to that of the general ALS population.^{42,43} Data from the two studies was collected at different time periods, and it is possible that patient care or values relating to QOL changed in the interval. The close match of results from ALSSQOL-R and ALSSQOL-SF studies reduces this concern. Our studies collected data through interviews; clinics adopting the ALSSQOL-SF may prefer to use computer-based or hard-copy self-completion of the instrument. Although our previous research showed no difference in results based on administration method⁴⁴, it is possible that clinics may find different results using different administration methods.

Limitations also exist regarding measures selected for assessment of validity. A more recent version (CESD-Revised) had been developed and released in 2004, but few appropriate norm scores were available at the time the current study was being developed. The IIR only has four items, and therefore, less variability, despite evidence for its reliability and validity³². Intimacy measures have been validated on younger, healthy populations^{45,46}, or require comparison of dyad responses, which was not practical for this research. Lastly, the items in the ALSSQOL, and therefore, the ALSSQOL-R and –SF, are distributed among several of the subscales, and therefore, the subscales are defined and compared by the dominant concepts reflected; no existential comparative measure was used.

Conclusions

For purposes of measuring QOL as a holistic construct and understanding factors that affect

patients' reports of QOL, the ALSSQOL-SF appears to be an efficient, brief instrument with good psychometric properties. Clinicians and researchers should consider the advantages and limitations of the ALSSQOL-R versus the ALSSQOL-SF when choosing one or the other for a specific use. Guidelines for scoring and routine clinical use of the ALSSQOL-SF will appear in a forthcoming manual.

ABBREVIATIONS

ALSSQOL: ALS Specific Quality of Life Instrument

ALSSQOL-R: ALS Specific Quality of Life Instrument – Revised

ALSSQOL-SF: ALS Specific Quality of Life Instrument -Short Form

BSI-18: Brief Symptom Inventory-18 item version

CES-D: The Center for Epidemiological Studies- Depressed Mood Scale

CFA: Confirmatory factor analysis

CFI: Comparative fit index

IIR: Idler Index of Religiosity³²

IRT: Item Response Theory

M-GRM: Modified Graded Response Model

MQOL-SIS: The McGill Quality of Life Single Item Score

QOL: Quality of life

RMSEA: Root mean square error of approximation

SRMR: Standard root mean square residual

SWLS: Satisfaction with Life Scale

WHOQOL-BREF: World Health Organization Quality of Life Instrument-Brief version

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Table 1. Structural Comparison of ALS Specific Quality of Life Instrument – Revised and Short Form.

Scale	ALSSQOL-R	ALSSQOL-SF	ALSSQOL-R Reliability	ALSSQOL-SF Reliability
Negative Emotion	13 items (28%)	3 items (15%)	0.91	0.86
Interaction	11 items (24%)	4 items (20%)	0.87	0.80
Intimacy	7 items (15%)	4 items (20%)	0.81	0.82
Religiosity	4 items (9%)	2 items (10%)	0.92	0.89
Physical Symptoms	6 items (13%)	5 items (25%)	0.71	0.70
Bulbar Function	5 items (11%)	2 items (10%)	0.83	0.81

ALSSQOL-R: ALS Specific Quality of Life Instrument – Revised; ALSSQOL-SF – ALS Specific Quality of Life Instrument – Short Form; Interaction: Interaction with People and the Environment.

Reliability was measured using Cronbach's α .

Table 2. Confirmatory Factor Analysis: standardized loadings for the ALS Specific Quality of Life

Instrument – Short Form

Short Form Items	NE	IPE	IN	RE	PS	BF
10. I have been depressed.	0.868					
14. I have felt hopeless.	0.737					
15. I have felt sad.	0.838					
8. The world has been caring and responsive to my needs.		0.737				
9. I have felt supported.		0.840				
11. Relationships have been satisfying.		0.629				
16. I have enjoyed the beauty of my surroundings.		0.592				
17. My desire for emotional intimacy has been strong.			0.787			
18. I have shared emotional intimacy with others.			0.762			
19. My desire for physical intimacy has been strong.			0.702			
20. I have shared physical intimacy with others.			0.674			
12. My religion has been a source of strength/comfort to me.			0.969			
13. I consider myself to have been religious or spiritual.			0.834			
1. Pain				0.468		
2. Fatigue				0.676		
5. My Strength and Ability to Move				0.505		
6. Sleep				0.528		
7. I have felt physically terrible.				0.649		

3. Excessive saliva					0.946	
4. Speaking					0.724	

NE = Negative Emotion; IPE = Interaction with People and the Environment; IN =Intimacy; RE =

Religiosity; PS = Physical Symptoms; BF = Bulbar Function

Table 3. Correlations between factors on the ALS Specific Quality of Life Instrument – Short Form.

	IPE	IN	RE	PS	BF
Negative Emotion	0.33**	0.17**	0.11*	0.42**	0.06
IPE		0.30**	0.30**	0.22**	0.07
Intimacy			0.25**	0.06	0.08
Religiosity				0.06	-0.07
Physical Symptoms					0.11*

NE = Negative Emotion; IPE = Interaction with People and the Environment; IN =Intimacy; RE =

Religiosity; PS = Physical Symptoms; BF = Bulbar Function

* $p < 0.05$, ** $p < 0.001$

Table 4. Normative/Mean scores of quality of life in total patient sample completing the ALS Specific Quality of Life Instrument – Short Form

Measure	Mean (SD)	Range
MQOL-SIS (N=162)	7.0 (2.1)	0-10
ALSSQOL-SF Average Total Score (N=160)	6.5 (1.1)	3.7-9.3
ALSSQOL-SF NE (N=161)	5.3 (1.2)	1.0-7.7
ALSSQOL-SF IPE (N=161)	8.6 (1.4)	3.0-10.0
ALSSQOL-SF IN (N=159)	6.7 (2.3)	0.0-10.0
ALSSQOL-SF RE (N=162)	6.7 (3.5)	0.0-10.0
ALSSQO-LSF PS (N=162)	5.3 (2.3)	0.0-10.0
ALSSQO-LSF BF (N=162)	6.4 (3.2)	0.0-10.0
ALSSQOLSF Total Score (N=160)	130.1 (21.9)	74-187

MQOL-SIS = McGill Quality of Life Single Item Scale; ALSSQOL-SF = ALS-Specific Quality of Life-Instrument – Short Form; NE = Negative Emotion; IPE = Interaction with People and the Environment; IN =Intimacy; RE = Religiosity; PS = Physical Symptoms; BF = Bulbar Function

Table 5. Correlations between ALS Specific Quality of Life Instrument-Short Form Subscales and Measures to Test Convergent, Divergent, and Construct Validity (N=162 unless otherwise noted)

Measures	Av Tot Sc	NE	IPE	PS	IN	RE	BF
ALSFRS-R Total	0.40**			0.48**			0.29**
Speech				0.17*			0.79**
Fine Motor				0.37**			0.06
Gross Motor				0.44**			-0.04
Respiratory				0.34**			0.23**
MQOL SIS	0.34**	0.28** N=161	0.28** N=161	0.25**	0.10 N=159	0.09	0.11
WHOQOL- Physical	0.47** N=161			0.60** N=161			0.118 N=161
WHOQOL- Psychological	0.52** N=158	-0.37** N=157			0.27* N=155		
WHOQOL- Social	0.57** N=160		0.49** N=159		0.42** N=157		
WHOQOL- Environment	0.39** N=159		0.37** N=158				
SWLS	0.44** N=161	0.33** N=160	0.37** N=160	0.21* N=161	0.28** N=158	0.16* N=161	0.10 N=161
CESD		-0.38** N=160					
BSI-18 Depression		-0.37** N=159					
BSI-18 Anxiety		-0.29** N=159					
BSI-18 Somatization		-0.13 N=159					
BSI-18 Total		-0.33** N=159					
IIR - Public						0.63** N=151	
IIR - Private						-0.21** N=161	
IIR - Total						0.57** N=151	

Av Tot Sc = ALSSQOL-SF Average Total Score; NE = Negative Emotion; IPE = Interaction with

People and the Environment; IN =Intimacy; RE = Religiosity; PS = Physical Symptoms; BF = Bulbar Function; MQOLSIS – McGill Quality of Life Single Item Scale; WHOQOL – World Health Organization Quality of Life; SWLS = Satisfaction with Life Scale; CESD = Center for Epidemiological Studies Depressed Mood Scale; BSI = Brief Symptom Inventory; IIR = Idler Index of Religiosity. * p <.05, **p<.001.

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FIGURE LEGENDS

Figure 1. Information functions by subscale for the ALS Specific Quality of Life Instrument-Revised (ALSSQOL-R) and its short form, the ALSSQOL-SF. In an Item Response Theory (IRT) context, information is the corollary to reliability and can be expressed as the reciprocal of the standard error conditional for a particular trait level:

$$I(\theta) = \frac{1}{SE(\theta)^2}$$

Thus, more information indicates more precision in measurement. Additionally, trait level refers to an individual's capacity (score) for the particular construct measured by the subscale, with 0 representing the average and higher numbers reflecting more characteristics relative to the data in the sample. The ALSSQOL-SF has slightly greater imprecision compared to the ALSSQOL-R, but the information functions display a consistent pattern between both forms, maximizing accuracy at the same points along the trait scale. Interaction = Interaction with People and the Environment.

Figure 2: Hypothesized second-order model of factorial structure for the ALS Specific Quality of Life Instrument – Short Form (ALSSQOL-SF). The items contributing to each of the 6 subscales of the ALSSQOL-SF are shown. Item numbers correspond to the specific items of the ALSSQOL-SF (see supplementary material for the complete questionnaire). QOL = ALSSQOL-SF; Negative = Negative Emotion; Bulbar = Bulbar Function; Physical = Physical Symptoms; Interaction = Interaction with People and the Environment.