

# The ALS Specific Quality of Life-Revised (ALSSQOL-R)

## User's Guide

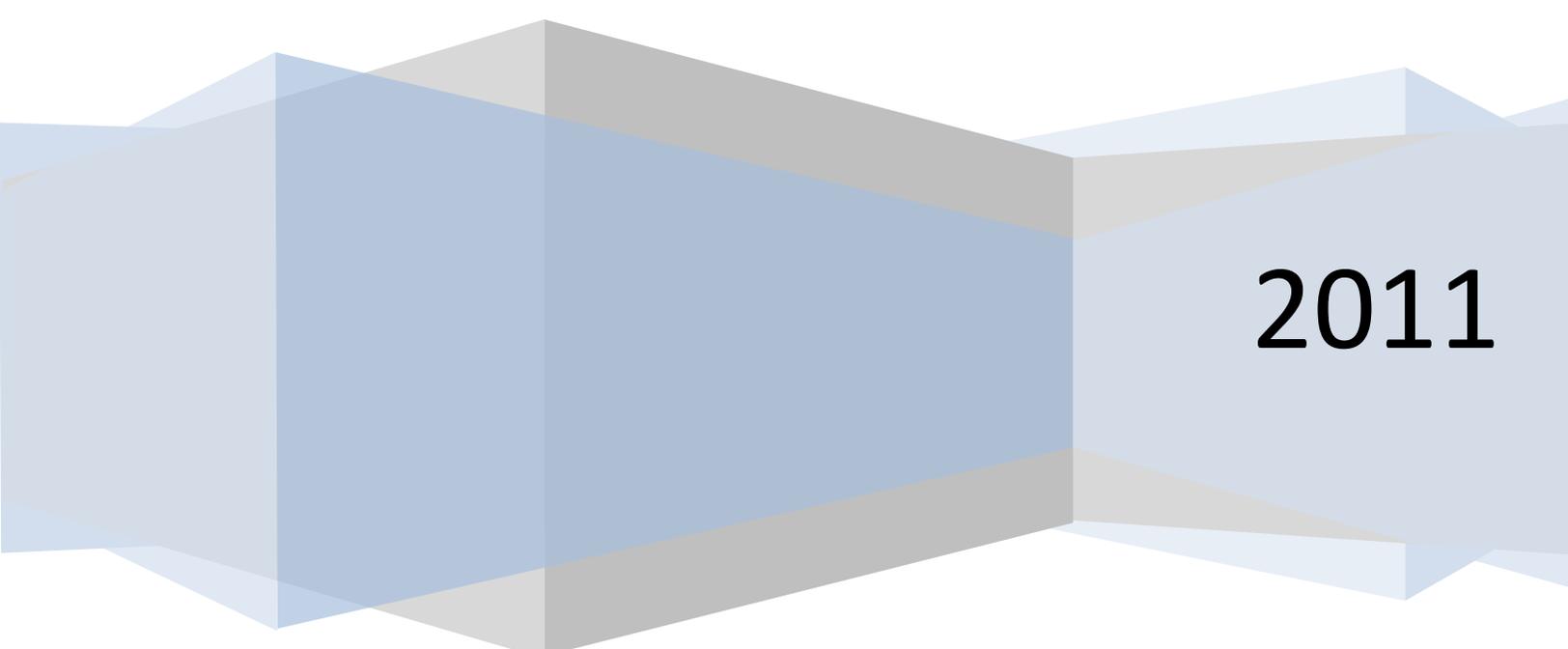
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## Chapter 1: Introduction

The Amyotrophic Lateral Sclerosis Specific Quality of Life Instrument-Revised (ALSSQOL-R) is a 50 item instrument that measures overall quality of life (QOL) and six specific domains for individuals with ALS. Forty-six of the items are used in scoring, to arrive at an average total score and scores for each of the six domains. The ALSSQOL-R can be used in both research and clinical settings, and is useful as a screening tool in the clinical setting where time is a major limiting factor.

### History

Quality of life (QOL) in patients with ALS does not correlate with physical function. Unfortunately, many quality of life (QOL) instruments which have been used to assess individuals with ALS are heavily weighted toward strength and physical function, and therefore fail to capture other important non-health related factors.<sup>1</sup> Existing generic QOL instruments are thought to be unsuitable because they may not assess features unique to ALS, such as the inevitability of death and the rapidly progressive series of functional losses. Using data collected from the McGill Quality of Life Questionnaire (MQOL)<sup>2-5</sup> and from measures of religion and spirituality,<sup>6-9</sup> it was determined that a QOL instrument for ALS should inquire broadly about psychological, support, existential, and spiritual issues, and should have a non-dominant physical domain.<sup>10</sup> We used these principles to develop an ALS-Specific QOL instrument (the ALSSQOL, now revised as the ALSSQOL-R) which would reflect overall self-reported QOL of individuals with ALS.

### Description and Key features of the ALSSQOL-R

The ALSSQOL-R is a 50 item disease-specific questionnaire that is completed by the individual with ALS. Each item of the ALSSQOL-R is rated by the individual using a 0 to 10 point Likert scale, with 0 being the least desirable situation, and 10 being the most desirable. The instrument produces a Single-item QOL score (derived from the McGill Quality of Life Questionnaire – see Chapter 2: Development of the ALSQOL-R), an Average Total QOL score, and 6 domain scores: 1) Negative Emotion; 2) Interaction with People and the Environment; 3) Intimacy; 4) Religiosity; 5) Physical Symptoms; 6) Bulbar Function. The ALSSQOL-R can be completed by most individuals

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with ALS in approximately 15-20 minutes, and can be administered and scored in either paper or computer format.

## Uses

Potential clinical users of the tool are healthcare clinicians caring for individuals with ALS including physicians, nurses, social workers, therapists and counselors. Research users include clinical trial groups and academic researchers in the applied social sciences such as nursing and psychology.

## Clinical Use

The ALSSQOL-R has successfully been implemented in a multidisciplinary ALS clinic. The Penn State Hershey ALS Clinic has paired the ALSSQOL-R with a self-reported ALS Functional Rating Scale-Revised (ALSFRRS-R)<sup>11</sup> as a pre-clinic assessment. Patients complete the assessment in a paper or computer format prior to their scheduled appointment. The ALSSQOL-R scores are shared with the clinic team to guide their assessments and interventions. The ALSSQOL-R can be administered to patients serially over time. Changes in scores can be used to re-assess the patients over time, including their responses to interventions.

## Research Use

The ALSSQOL-R and its original version, the ALSSQOL, have been used or are being used as QOL outcome measures in clinical trial research to examine the effect of experimental treatments on individuals' QOL. Data collected during the development of these instruments has been used to examine various aspects of QOL and ALS.<sup>12-14</sup> Other research efforts are ongoing.

## Cautions of Use

There are many life factors beyond the disease that contribute to QOL such as life events other than ALS, mood state, cultural factors and response biases that may skew ALSSQOL-R scores. The ALSSQOL-R should be considered as one component of a comprehensive diagnostic evaluation. The use of the ALSSQOL-R is not appropriate for individuals with cognitive impairment or with significant psychiatric illness.

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## Appropriate Samples

The ALSSQOL-R has been validated on a national sample of adult men and women from 27 to 88 years of age who are receiving care for their disease in ALS multidisciplinary clinics. It has been used in a broader clinical sample of patients who are in treatment trials or who are not receiving multidisciplinary care. Normative total scores and scores for specific domains can be found in Chapter 6.

## Chapter 2: Development of the ALSSQOL-R

The original ALSSQOL was developed by using the McGill QOL Questionnaire<sup>2-5</sup>, the Idler Index of Religiosity<sup>6</sup>, and qualitative analysis of interviews with ALS patients. Additional tools used to establish the psychometric properties of the original ALSSQOL included the Schedule for the Evaluation of Individual Quality of Life-Direct Weighting (SEIQoL-DW)<sup>15</sup>, World Health Organization Quality of Life Instrument (WHOQOL-BREF)<sup>16</sup>, the Functional Assessment of Chronic Illness Therapy - Spiritual Well-Being (FACIT-Sp-12)<sup>17</sup>, and the Brief Symptom Inventory (BSI)<sup>18</sup>.

To test the psychometric properties of the ALSSQOL, a multi-site study was conducted resulting in the ALSSQOL-R containing 46 scored items and 4 non-scored items. The domains within the ALSSQOL were developed by principal component analysis identifying the 6 predominant domains of QOL for individuals with ALS.<sup>19</sup> A second multi-site validation study tested the reduced 50 item measure, confirmed the factor structure and established the psychometric properties for the ALSSQOL-R (see Chapter 6). A third study tested the ALSSQOL-R for reliability for self-reporting and computer administration validating that the instrument can be administered in multiple modes.<sup>20</sup>

## Chapter 3: Definition of Terms

**The Single-item Score** measures global QOL and is a summation of one's perceived state of physical, emotional, social, spiritual, and financial well-being.

**The Average Total ALSSQOL-R** score is the average of the 46 scored items of the ALSSQOL-R and is reported as a value between 0 and 10.

**There are six Domains within the ALSSQOL-R, each reported as a value between 0 (worse) and 10 (best):**

- 1. Negative Emotion** encompasses a variety of emotional states, including but not limited to depression and anxiety, experienced by the individual with ALS and pertaining to one's outlook about the future.
- 2. Interaction with People and the Environment** describes how individuals with ALS perceive and respond to friends and family, and how they experience their environment.
- 3. Intimacy** measures experience of, satisfaction with, and desire for social, emotional, and physical intimacy and sexual intercourse.
- 4. Religiosity** measures individuals' religious identification, use of religion as a source of comfort, use of prayer, and engagement in religious practice at home.
- 5. Physical Symptoms** measures the extent to which the following physical symptoms are perceived as problematic by the individual: ability to move, sleep, feeling physically terrible, fatigue, pain, bowel and bladder.
- 6. Bulbar Function** measures the extent to which the following bulbar symptoms are perceived as problematic by the individual: speaking, saliva, communication problems, mucous, and eating.

## Chapter 4: Test Administration and Scoring

### Test Administration

The ALSSQOL-R is designed to be a self-administered instrument but it can also be administered in an interview format, either via hard copy or on the internet using a survey software package. The instrument provides concise instructions for completion but a short introduction of its purpose with an individual or population is recommended. Completion averages 15 minutes per individual, with a range of approximately 10-25 minutes. It is recommended that the individual completing the instrument be in a private setting with minimal distractions. It is advisable to clarify that the ALSSQOL-R can easily be completed by the individual and does not require the caregiver to respond to the items.

If the ALSSQOL-R is administered in an interview format the interviewer should be instructed to present each item in an identical form, thereby reducing any possibility of biases or preference in answering the questions.

Individuals with physical limitations can respond to the items by use of pointing, nods or eye blinks.

The self-reported instrument should be reviewed by the administrator for missing items. Individuals should be queried for missing information.

### Administration of the ALSSQOL-R in a Multidisciplinary Clinic

The ALSSQOL-R can be used as a pre-clinic assessment. We recommend having the patient complete the instrument in their home via an internet based survey or with a paper and pencil version about 1-2 weeks before their scheduled appointment. For paper and pencil assessment, we recommend having the patient mail their assessment to the health care team prior to their appointment. The scores obtained from the ALSSQOL-R should be made available to the multidisciplinary team members prior to their visit with the patient.

### Scoring

Each item of the ALSSQOL-R is scored on a 0 to 10 scale, with 0 the least contribution to QOL, and 10 the greatest contribution to QOL. Several of the items require transposing (subtracting

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the score of the response from 10) prior to calculating a score, specifically items 1-10, 17, 22, 24, 25, 28, 31, 32, and 34. Thus, total scores range from 0 to 460 (10 x 46). An average total ALSSQOL-R score for each individual ranging from 0 to 10 can be determined by summing the scores for all questions for that individual, and then dividing by the number of questions answered by that individual. Subscores can be obtained by summing the responses for the questions within that domain and dividing by the number of questions answered within that domain. Please see Appendix B for a hand scoring sheet, and Appendix C for a computer syntax that can be used to generate automatic scoring with a statistical software package. If data is being used for research purposes and there are three or less missing scores on an individual's questionnaire, data can be imputed via statistical computation. If data is being used for clinical purposes, the subscales and total score should be computed by totaling the items within the subscale according to the above directions and dividing by the number of items answered. There is not an exact formulary to determine the validity of the measure with missing data, however, certain subscales (Religiosity, Bulbar Function) have fewer items, and therefore, more than one missing item per subscale could significantly affect the clinical utility of the scores. It is likely that Negative Emotion, Interaction with People and the Environment, Intimacy, and Physical Symptoms would still be valid with up to two items missing on each, since the Chronbach alpha coefficients are high and all items load significantly on the factor.

## Chapter 5: ALSSQOL-R Interpretation

The ALSSQOL-R provides insight into an individual's self-perceived QOL at any single point in time. The Single-item QOL question reflects individuals' global perception of QOL, and this score correlates highly with the Average Total ALSSQOL-R score. The QOL domains can be used to identify the importance of specific aspects of QOL. It is helpful to look at the profile of the score report in all of the domains. The domain scores can be examined for highs and lows, consistency within an individual profile, and trends over time. Average total and domain scores can also be compared to normative data for patients with ALS (Chapter 6: Tables 6.3, 6.3a, and 6.3b). Individual item responses provide details which may not be evident from the overall domain score and may thus provide individual clinical team members with further clarification and insight. The ALSSQOL-R can be administered at multiple visits, and values can be compared over time to identify changes. Examination of serial scores also can identify changes in response to interventions such as medications, counseling, adaptations, and in-home care.

### Domains

**Negative Emotion** has been shown to most contribute to individuals QOL.<sup>21</sup> Patients who score 6.23 or lower on this domain should be further evaluated for depression, suicidal tendencies, anxiety, concerns about the future and the meaning of one's life, and ability to cope, as discussed in the psychometrics section of this manual. Health care providers across a number of disciplines will find this domain useful. This domain is highly correlated with measures of depression, anxiety, and positive and negative coping skill sets.

**Interaction with People and the Environment.** Lower scores on this domain in comparison to those in Tables 6.3, 6.3a, and 6.3b may indicate the need to evaluate the patient's social environment, available social support and their interest in social interactions. This measure correlates strongly with measures of satisfaction with life, and perceived social support by family and significant others, but less so with friends.

**Religiosity** is only descriptive of a person's religious practices, and not of their spirituality. High or low scores compared to normative ALS data are not necessarily indicative of problems or strengths. Changes in this score over time may prompt further exploration. This domain may be used clinically with other domains to address spirituality and coping, bearing in mind that

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religiousness and spirituality differ from one another. It is particularly useful to look at Religiosity scores in the context of Negative Emotion scores. If Negative Emotion scores are high (indicating minimal problems in this area), follow up in the area of Religiosity may not be indicated because the person may be coping and adjusting well. If, however, individuals are distressed (low scores on Negative Emotion), it is helpful to review the Religiosity score. If it is low, then religious intervention should be explored as a possible means to improve overall QOL. However, if Religiosity is high despite low scores on Negative Emotion, the individual may be having a religious crisis or conflict, so that religious beliefs are associated with distress rather than comfort. This combined review is indicated because the Religiosity subscale correlated highly with the Faith in Illness Scale of the FACIT-sp, but not with the FACIT-sp Meaningfulness scale. Meaningfulness scale correlated strongly with Negative Emotion, suggesting a strong relationship between negative emotion and existential concerns (See psychometric section below).

**Intimacy** Lower scores compared to ALS norms may serve as a segue to conversations about relationships and communication between ALS patients and their significant others. Interventions may be guided toward enhancing physical, social, emotional, and sexual intimacy. High scores suggest patients have desire for and experience with the various expressions of intimacy. There are 4 additional items that are not included in the total score and which assess satisfaction with intimacy (Items 41, 44, 47, and 50). These items provide useful qualitative information.

**Physical Symptoms and Bulbar Function.** It is important to realize that the physical and bulbar scores reflect the patient's perception of the impact of their physical symptoms on their QOL. QOL scores in the physical domain may not reflect the more objective measures of function such as the ALSFRS-R. High objective functional measurements may be associated with low scores on these two domains, and vice versa. Clinicians may use the scores to prioritize interventions for the patient based on the impact that the symptoms are having on the patient's QOL.

### Sample Patient Profiles

#### Patient #1

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BJ is a 43 year old man, married with two young adult children. His ALS Functional Rating Scale-Revised (ALSFRRS-R) score is 21/48. He uses an eye gaze system to communicate, supplemented by dysarthric speech. Despite his bulbar dysfunction, he has normal secretions and still has normal eating habits. He has no use of his arms and is totally dependent on his spouse for care. He is non-ambulatory and is in a power wheelchair for most of the day. He has no respiratory symptoms, although his forced vital capacity is 43% of predicted. He has recently begun using noninvasive positive pressure ventilation (NIPPV, often called a BiPAP device).

### Patient #1 QOL Assessment

Average Total QOL Score:	7.37/10
Negative Emotion:	7.31/10
Interaction:	8.36/10
Intimacy:	6.71/10
Religiosity:	8/10
Physical:	8.33/10
Bulbar:	6.2/10

Normative data for individuals with ALS is provided in Table 6.3. BJ's Total QOL assessment is slightly above the reported mean QOL assessment. Overall, looking at the QOL score profile for BJ, his scores are all relatively high. Despite his significant physical impairments, he reports his physical symptoms as 8.33/10 which indicates that physical symptoms are not perceived as a problem for BJ. His Bulbar score report is lower than the Physical score, and it is the lowest domain score in his profile. His bulbar symptoms should be explored at this ALS Clinic appointment. BJ's Negative Emotion score is slightly above the reported average score for this domain. His Interaction with People and the Environment Score is also high. BJ's Religiosity report is high, as is his Intimacy score. Despite his fairly advanced disease as measured by this ALSFRS-R, BJ reports a high QOL.

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### Patient #2

ML is a 46 year old woman, married with 2 young children. Her ALSFRS-R is 33/48. Her speech is intelligible with repeating. She has some problems with secretions. She has normal eating habits. ML is able to write legibly. She needs some assistance with activities of daily living. She walks with assistance and reports no respiratory problems. Her forced vital capacity is 98% of predicted.

### Patient #2 QOL Assessment

Average Total QOL Score:	5.41/10
Negative Emotion:	3.69/10
Interaction:	6.36/10
Intimacy:	5.85/10
Religiosity:	1.25/10
Physical:	7.83/10
Bulbar:	7.2/10

ML's average total QOL assessment is lower than average (Table 6.3). Viewing her QOL profile, the domain scores that are particularly low relative to average values for ALS patients include Negative Emotion, Interaction and Religiosity. During the ALS Clinic appointment, the team evaluated ML for depression, suicidal tendencies, anxiety, concerns about the future and the meaning of one's life, and ability to cope. After a comprehensive evaluation, it was determined that ML was depressed. Recommendations were made for ML to seek counseling, and a prescription for an antidepressant was provided. ML also reported that she was not as social as she used to be. ML had stopped joining her social groups for activities. Many of the social groups she had previously interacted with belonged to her religious congregation. This change in behavior was reflected in ML's reported low Interaction score. ML also reported low Religiosity. It was helpful to look at a previous QOL assessment for ML to examine if this domain

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score had changed from baseline. Previously, ML previously rated her Religiosity as 8/10. Her Religiosity score is clearly a change from baseline. The Pastoral Care counselor explored this with ML, who revealed that she felt abandoned by her religious congregation and was unable to participate in many of the congregation's activities. The counselor and ML discussed ways in which ML could participate in congregation activities to feel a better sense of belonging.

## Chapter 6: Psychometrics

### Introduction

This chapter presents the method used to validate the ALSSQOL-R and the results of the analyses conducted. As a result of a review of the literature and the preliminary test development and construction procedures described under “Development of the ALSSQOL-R,” the ALSSQOL has established content and face validity. Principal Component Analysis of the original ALSSQOL revealed six factors and the insignificance of 9 items which were subsequently deleted.<sup>19</sup> Because items of the original measure were removed and four items separated from scorable items, it was necessary to collect new data on the ALSSQOL-R to determine if the psychometric properties of the original measure were retained, and if the findings were reliable. Hierarchical multiple regression tested the six factors’ ability to predict global QOL as judged by the patient’s response to a single question derived from the McGill Quality of Life Instrument (MQOL-SIS). Furthermore, the current study included additional measures which allowed the construct validity of the ALSSQOL-R and its subscales to be tested. Construct validity was established by testing concurrent validity (convergent and discriminant validity) with “gold standard” measures relating to the factors of the ALSSQOL-R.

A description of the methods, factor structure tested by Confirmatory Factor Analysis, reliability and validity statistics are followed by normative data. The ALSSQOL-R’s relation to other standardized measures is described. This data substantiates the recommendations made in the clinical interpretation section above.

### Methods

#### Evaluation of the Psychometric Properties of the ALSSQOL

*Participant Inclusion/Exclusion Criteria.* A prospective study involving 12 American ALS clinics (Appendix D) recruited individuals 18 years or older, fluent in English at the 6<sup>th</sup> grade level or higher, with definite, probable, or probable laboratory-supported ALS by revised El-Escorial criteria.<sup>22</sup> Patients were excluded if they were judged by a physician or psychologist at the center to have a dementia sufficient to preclude the granting of informed consent and

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participation in the study. Approval for the study was obtained from the institutional review board at each of the centers, and all subjects provided informed consent.

## Instruments Used at all Sites

All sites collected demographic information and administered the following measures of QOL, function, and strength:

1. The ALS-Specific Quality of Life Questionnaire-Revised (ALSSQOL-R): The 50-item version of the ALSSQOL (46 scorable items plus 4 additional items) was administered to the patient in interview format by a psychologist, research coordinator, nurse, or trained psychology graduate student, with family members out of the room. The ALSSQOL-R total score represents the addition of all scorable items, after reverse-scoring is implemented. The ALSSQOLR Average Total Score is the ALSSQOL-R total score divided by 46.
2. The McGill Quality of Life Single-Item Scale (MQOL-SIS):<sup>2-5</sup> This global measure consists of a single question in which patients rate their QOL over the preceding 2 days on a 0-10 scale. Although it appears as the first question on the ALSSQOL-R, it is scored separately from the rest of the measure. To make it consistent with the rest of the ALSSQOL-R, the time frame for which QOL is judged by the patient was changed from 2 days to 7 days.
3. ALS Functional Rating Scale-Revised (ALSFRRS-R):<sup>23</sup> This is a 48-point scale consisting of 12 items which assess bulbar, limb, and respiratory function. Scores range from 0 (worst function) to 48 (best function).
4. Manual muscle testing: Strength in four muscle groups (arm abductors, wrist extensors, hip flexors, and ankle dorsiflexors, corresponding to proximal and distal upper and lower extremity muscle groups, respectively) was measured and recorded bilaterally for each patient using the Medical Research Council (MRC) Scale of 0-5.<sup>24</sup> A composite MRC score was calculated for each patient, consisting of the sum of the MRC scores in each of these 8 muscle groups divided by 8. Scores range from 0 (weakest) to 5 (strongest).

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## Instruments Used at Selected Sites:

At least two sites each collected data using the following additional measures to test the construct, convergent, and /or discriminant validity of the indicated factors of the ALSSQOL-R or the total ALSSQOL-R score:

1. Total ALSSQOL-R score and subscale scores. Participants either completed the World Health Organization QOL Instrument – Brief Version (WHOQOL-BREF, n=176) or the ALS Assessment Questionnaire 40 (ALSAQ-40, n=214). The WHOQOL-BREF is an abbreviated 26-item version of the WHOQOL-100, and was used in the validation of the original ALSSQOL. Four domains are assessed: physical health, psychological (including spirituality, religion, and personal beliefs), social relationships, and environment. Scores are calculated for each domain.<sup>16</sup> The ALSAQ-40 is a 40-item measure of self-reported health-related QOL with five subscales: Eating and Drinking, Communication, ADL/Independence, Physical Mobility, and Emotional Functioning. Construct validity, internal reliability, and sensitivity to change have been established for this measure.<sup>25</sup>
2. Negative Emotion. Participants in this arm of the study completed two measures of psychological distress, the Brief Symptom Inventory-18<sup>26</sup> (BSI-18; N = 74) and the Center for Epidemiological Studies- Depression Scale<sup>27</sup> (CESD; n =74), They also completed the Social Problem-Solving Inventory-Revised: Short Form<sup>28</sup> (SPSI-R; n = 73), a measure of social problem solving as a coping skill set. The BSI-18 is an 18-item self-report measure of symptoms of anxiety, depression, and somatization experienced by both medical and psychiatric patients. Items are rated on a five-point scale measuring degree of distress caused by each symptom (0, not at all; 4, extremely). This measure is often used as a screening tool with medical patients, as it has excellent psychometric properties and originated from the 53-item Brief Symptom Inventory.<sup>29</sup> The CES-D is a 20- item measure designed to assess depressed mood by having respondents record how often in the past week they have experienced listed symptoms (feelings or behaviors) according to a 4-point Likert Type Scale. This measure has established validity and reliability when used with a nonpsychiatric adult population.<sup>27</sup> This measure is particularly useful with

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medical populations because few vegetative symptoms are assessed. The SPSI-R is a 25 item measure of social problem solving which examines individual's ability to identify adaptive solutions to problems in daily living.<sup>28</sup> Social problem solving is conceptualized as a skill set that represents one's coping style and abilities. The SPSR-I has established normative scores for elderly adults, and has been widely used in research with patients with chronic illnesses such as cancer.<sup>30-31</sup> The measure is comprised of five scales including positive problem orientation, negative problem orientation, rational problem-solving, impulsive/carelessness style, and avoidance style.

3. Interaction with People and the Environment. This factor of the ALSSQOL-R was compared to the following measures completed by participants in this arm of the study: Satisfaction with Life Scale (SWLS; n = 79); Multidimensional Scale of Perceived Social Support (MSPSS; n = 79). The SWLS is a 5-item measure that has been widely used to measure global satisfaction with life in a variety of populations, including 4 groups of older adults (averaging approximately 53 to 75 years of age). Statements are endorsed according to a 5-point likert-type scale ranging from 1 (Strongly Disagree) to 7 (Strongly Agree). The SWLS has established positive psychometric properties, including reliability over time, and internal consistency.<sup>32</sup> The MSPSS is a subjective measure of perceived social support adequacy from family, friends, and significant other.<sup>33</sup> This measure includes 12 items assessing the extent to which support is received from these three sources on a 7-point Likert-type scale ranging from 1 (very strongly disagree) to 7 (very strongly agree). Higher scores are indicative of higher social support.
4. Religiosity. Participants in the "religiosity" arm of the study completed five brief measures of aspects of religiosity or spirituality: the Functional Assessment of Chronic Illness Therapy-Spiritual Well-Being Subscale (FACIT-SP-12; N = 138), the Idler Index of Religiosity (IIR; N = 139), and three scales from the Fetzer Institute report on the multidimensional measurement of religiousness/spirituality for use in health research: Forgiveness, Daily Spiritual Experiences, and Overall Self-Rank (N=140). The FACIT-Sp-12 is a 12-item measure of spirituality yielding two subscale scores (Meaning/Peace, Faith in Illness) and a total score. It was developed for

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evaluation of QOL in patients with chronic illness. Respondents indicate how true each statement is for them on a scale ranging from “0” (not at all) to “4” (very much).<sup>17</sup> The IIR is a four-item measure and consists of two items assessing public religiousness and two items assessing private religiousness. Total scores range from 4 (least religious) to 17 (most religious).<sup>6</sup> The Forgiveness, Daily Spiritual Experiences, and Overall Self-Rank scales were developed by a national working group supported by the Fetzer Institute in collaboration with the National Institute on Aging.<sup>34</sup> The forgiveness subscale includes three items and assesses the following dimensions of this construct: feeling forgiven by God, forgiving others, and forgiving oneself. This subscale was originated from the Judeo-Christian tradition, and therefore, this should be considered when interpreting its relation to the ALSSQOL-R constructs. Established psychometric properties of this subscale are limited, although theoretically and conceptually the scale appears to be well-developed. Daily Spiritual Experiences scale includes 6 items and represents the “individual’s perception of the transcendent in daily life and the perception of interaction with, or involvement of, the transcendent in life.”<sup>34</sup> The focus is on experience rather than cognitions, and the scale is intended to transcend religious boundaries. The Overall Self-ranking scale includes two items inquiring about the extent one self-reports being religious and/or spiritual. These three scales are imbedded within the Brief Multidimensional Measure of Religiousness/Spirituality and preliminary data suggests that within this measure adequate reliability and validity data exists for its use.

5. Intimacy. This subscale of the ALSSQOL-R was assessed using the Personal Assessment of Intimate Relationships (PAIR, n = 82). The PAIR is a measure of five types of intimacy, assessed by two 36 – item measures. One measure assesses perceived level of intimacy, and expected level of intimacy is measured by a second questionnaire. Administering both scales allow us a score to be determined for satisfaction by examining the discrepancy between the two. For the purpose of this study, only the actual level of intimacy was assessed. Data was only collected from the ALS patient, and not the partner. The following intimacies were measured: emotional intimacy, social intimacy, sexual intimacy, recreational intimacy, and

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intellectual intimacy.<sup>35</sup> The normative groups on which this measure was developed include individuals ranging in age from 18 to 61 years, recruited from community enrichment groups, and undergraduate class, and postgraduate extension classes. Reliability and validity data was established based on a sample of 192 couples who had been married between one and 37 years. This measure has been factor analyzed, and has established reliability and validity data resulting from the comparison of this measure to other couples' adjustment scales that are standard to the field.<sup>36</sup>

6. **Physical Functioning.** Measures to assess validity of this domain included several which are described above: the ALSFRS-R, Composite MRC, ALSAQ-40 Physical Mobility, Eating and Drinking, and ADL/Independence subscales, and the WHOQOL-BREF physical health domain.
7. **Bulbar Function:** Measures used to assess validity of this domain included several which are described above: the ALSAQ-40 Eating and Drinking subscale and Communication subscale, and items 1-3 of the ALSFRS-R.

### **Sample Size and Statistical Methods**

After collection at each center, data was sent to a central data collection center for entry and analysis. A multimethod, multimatrix design was implemented. A confirmatory factor analysis using Amos 16.0 was conducted to test the proposed model developed in the original ALSSQOLR validation by principal component analysis. Chronbach's alpha to determine internal consistency, Pearson's correlations, and measures of central tendencies and frequencies were conducted. Generally, the ratio of five cases (participants) per variable is recommended to have sufficient data for factor analysis.<sup>37</sup> As such, the number of participants required to perform factor analysis on the ALSSQOLR is 5 x 46, or 230 participants. Data collection for the ALSSQOLR exceeded this minimum requirement because larger samples of data on collateral measures were needed for validity testing of the factors, and only subsets of the overall sample completed these as described above. Hierarchical multiple regression analysis was conducted using subscale scores of the ALSSQOL-R as predictor variables and the MQOL-SIS as the criterion variable. Since the ALSSQOL-R and the MQOL-SIS were completed at each collection site, this

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data set provided an ample sample size for adequate power to conduct multiple regression analysis with an alpha set at .01 (by convention) and a medium effect size for more than the determined number of factors/subscales (i.e., a minimum of 147 subjects are recommended for multiple regression with 8 variables).<sup>38</sup> The number of subjects necessary for the factor analysis satisfied the number of subjects necessary to complete additional analyses of validity and reliability. Statistical analyses was performed using statistical software from SPSS, Inc (SPSS 17.0, Chicago, IL).

### **Testing of Validity, Reliability, and Reproducibility**

As a result of a review of the literature, initial test development and construction procedures described in the Development section, the ALSSQOL-R has established content and face validity. The MQOL-SIS was used to determine concurrent validity of the ALSSQOL-R. Although it is argued that the MQOL-SIS is a broader, global measure of QOL, QOL researchers recommend the use of global QOL measures in addition to health-related and/or psychosocial QOL measures.<sup>39</sup> Comparisons of the ALSSQOL-R to the WHOQOL-BREF domains and to the ALSAQ-40 served to test the convergent validity of the ALSSQOL-R by comparing its data to two additional reliable and valid measures of QOL. The ALSSQOL-R was compared to the BSI-18, CES-D, composite MRC, and ALSFRS-R to test discriminant validity, as positive QOL was hypothesized to represent more than the absence or inverse of psychological distress, physical strength, or physical function, as also supported by previous analysis of the original ALSSQOL. Each of the six factors of the ALSSQOL-R was compared to well-established measures with sound psychometric properties to test convergent validity and construct validity. Since limited psychological measures have been used with the ALS population, measures used to test Interaction with People and the Environment and Intimacy were chosen for analysis based on their construct and content similarity, although they had not been previously used with medical populations similar to the ALS population. All measures used for these comparisons are described above.

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## Results

### Personal Characteristics of the Sample

389 patients were recruited, of whom 229 (58.9%) were men and 160 (41.1%) were women. Ages ranged from 27 to 88 years (median = 61.0, mean = 60.85, SD = 11.36). Duration of symptoms at time of enrollment in the study averaged 39.7 months (SD = 39.24; median = 26 months; range = 3 to 232 months; N=324). Length of time from diagnosis to completion of study questionnaires averaged 21.45 months (SD = 27.03; median = 12.5; range = 0 to 223 months; N= 362). Other demographic features of our patients are presented in Table 6.1.

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**Table 6.1: Demographics of ALSSQOL-R Validation Sample**

<b>Age (Males) (n = 227)</b>	
M=60.45, SD=11.45; Range 33-84.	
<b>Age (Females) (n=158)</b>	
M=61.42, SD=11.24; Range 27-88.	
<b>Education (n= 381)</b>	<b>Annual Household Income (n=375)</b>
Less than high school graduate: 21 (5.5%)	< \$20,000: 30 (8.0%)
High school graduate: 101 (26.5%)	\$20,000-39,999: 87 (23.2%)
Some college: 67 (17.6%)	\$40,000-59,999: 80 (21.3%)
2 year college degree: 20 (5.2%)	\$60,000-\$79,999: 55 (14.7%)
4 year college degree: 98 (25.7%)	\$80,000 or more: 80 (21.3%)
Graduate degree: 64 (16.8%)	Prefer not to answer: 43 (11.5%)
Trade/technical school: 10 (2.6%)	
<b>Living Arrangements (n=387)</b>	<b>Race (n=388)</b>
With a spouse or partner 298 (77%)	Caucasian: 352 (90.7%)
With another relative 21 (5.4%)	Hispanic: 11 (2.8%)
With a friend or other non-related individual 8 (2.1%)	African American: 16 (4.1%)
Alone 47 (12.1%)	Asian/Pacific Islander: 3 (.8%)
Long-term care facility 7 (1.8%)	Other: 3 (0.8%)
With significant other and another relative 6 (1.6%)	Preferred not to Answer: 3 (.8%)
<b>Marital Status (n=386)</b>	<b>Employment (n=375)</b>
301 (78%)	Employed part time: 28 (7.5%)
Divorced 38 (9.8%)	Employed full time: 50 (13.3%)
Separated 3 (0.8%)	Retired: 168 (48.8%)
Widowed 28 (7.3%)	Unemployed: 6 (1.6%)
Never married 16 (4.1%)	On disability: 107 (28.5%)
	Not employed prior to ALS: 14 (3.7%)

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Retired and Disabled: 2 (.5%)

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## Factor Analysis

The ALSSQOL-R consists of 46 items loaded on 6 factors that account for QOL; 4 contingency items not answered by all participants were not scored, but are clinically useful for monitoring patients individually. The six factors' labels and internal consistency appear in Table 6.2. (Comparison of Chronbach Alpha reliability estimates from the ALSSQOL to the ALSSQOL-R show that these statistics are stable and not significantly different.)

**Table 6.2: ALS-Specific Quality of Life Questionnaire (ALSSQOLR): Factors and Internal Consistency**

Factor (number of items)	Item Numbers on ALSSQOLR	Internal Consistency*
Negative Emotion (13 items)	11, 12, 13, 17, 18, 19, 21, 22, 25, 28, 31, 32, 34	0.91
Interaction with People and the Environment (11 items)	14, 15, 16, 20, 26, 27, 30, 33, 36, 37, 40	0.87
Intimacy (7 items)	39, 42, 43, 45, 46, 48, 49	0.81
Religiosity (4 items)	23, 29, 35, 38	0.92
Physical Symptoms (6 items)	1, 2, 7, 8, 9, 10	0.71
Bulbar Function (5 items)	3, 4, 5, 6, 24	0.83

\*Chronbach's alpha

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Confirmatory factor analyses were conducted using Amos 16.0 of SPSS 17.0. In evaluating each model, we generated goodness of fit statistics to determine how well our models (6 factors of QOL) matched the data; the model converged in 12 iterations. Of note, statistics used to measure goodness of fit comprise an area of hot debate and therefore are used with caution.<sup>40</sup> Three statistics of goodness of fit are presented: chi-square divided by degrees of freedom (i.e., chi-square/df), Comparative Fit Index (CFI), and Root Mean Square Error of Approximation (RMSEA). Desired values are as follows: a) for chi/square/df<2.0, b) for CFI>=.9, and c) for RMSEA<= .06,<sup>41</sup> although 0.1 or less is acceptable.<sup>42</sup> Our model resulted in goodness of fit indices as follows:  $X^2 = (2985.91/974) = 3.06$ ,  $p = .000$ . CFI = .766, and RMSEA = .069, 90% Confidence Interval: .066 - .072, (PCLOSE = .000). These results are reasonable, considering the sample size used for this analysis is relatively small. Table 6.21 shows factor loadings of items by factor, with the standard error of the estimate, and squared multiple correlations. This data supports the factor structure and shows the relationships among items and factors. A zero-order correlation table is represented in Table 6.22 to show the relationship between ALSSQOL-R items and the identified factor on which they load. Table 6.23 depicts the relationships between subscales, according to this model. As expected, most correlations are low, since the original measure was factor analyzed using principal component analysis with orthogonal varimax rotation.<sup>19</sup> Negative Emotion and Interaction with People and the Environment have a greater degree of shared variance due to the psychosocial nature of both of these constructs.

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**Table 6.21 . Maximum Likelihood Estimates, Squared Multiple Correlations, and Standardized Regression Weights for ALSSQOLR Items by Factors**

	Maximized Likelihood Estimates	Standard Error	Squared Multiple Correlations	Standardized Regression Weights
<b>Negative Emotion</b>				
ALSSQOLR 34	1.000		.624	.790
ALSSQOLR 28	.918	.068	.426	.652
ALSSQOLR 22	1.032	.058	.649	.806
ALSSQOLR 12	.769	.051	.542	.736
ALSSQOLR 32	.994	.060	.588	.767
ALSSQOLR 25	.791	.074	.286	.535
ALSSQOLR 31	.969	.069	.451	.672
ALSSQOLR 19	.662	.052	.417	.646
ALSSQOLR 11	.671	.058	.348	.590
ALSSQOLR 13	.858	.065	.426	.652
ALSSQOLR 21	.676	.063	.295	.544
ALSSQOLR 17	.618	.071	.196	.443
ALSSQOLR 18	.569	.064	.209	.457
<b>Interaction w/ People &amp; Environment</b>				
ALSSQOLR 33	1.000		.474	.689
ALSSQOLR 16	.879	.069	.540	.735
ALSSQOLR 26	.983	.076	.515	.718
ALSSQOLR 30	.853	.091	.253	.503
ALSSQOLR 15	.930	.081	.427	.653
ALSSQOLR 27	1.094	.091	.437	.661
ALSSQOLR 20	.882	.090	.296	.544
ALSSQOLR 36	.723	.073	.297	.545
ALSSQOLR 40	.803	.092	.230	.480
ALSSQOLR 37	1.033	.083	.481	.693
ALSSQOLR 14	.748	.068	.386	.622

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	Maximized Likelihood Estimates	Standard Error	Squared Multiple Correlations	Standardized Regression Weights
<b>Intimacy</b>				
ALSSQOLR 45	1.000		.612	.782
ALSSQOLR 42	.869	.070	.492	.702
ALSSQOLR 46	1.040	.071	.551	.742
ALSSQOLR 48	.761	.084	.227	.476
ALSSQOLR 43	.885	.077	.453	.673
ALSSQOLR 49	.678	.079	.214	.463
ALSSQOLR 39	.531	.062	.224	.473
<b>Physical Symptoms</b>				
ALSSQOLR 07	1.000		.289	.538
ALSSQOLR 08	1.123	.149	.295	.543
ALSSQOLR 10	1.242	.154	.407	.638
ALSSQOLR 02	1.236	.143	.468	.684
ALSSQOLR 01	.798	.126	.184	.429
ALSSQOLR 09	.909	.138	.197	.444
<b>Bulbar Function</b>				
ALSSQOLR 03	1.000		.260	.510
ALSSQOLR 05	1.408	.150	.501	.708
ALSSQOLR 24	1.522	.177	.414	.643
ALSSQOLR 04	1.593	.162	.664	.815
ALSSQOLR 06	2.041	.208	.716	.846
<b>Religiosity</b>				
ALSSQOLR 29	1.000		.780	.883
ALSSQOLR 35	1.019	.045	.727	.853
ALSSQOLR 23	1.078	.043	.816	.903
ALSSQOLR 38	1.077	.053	.656	.810

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**Table 6.22 Correlations between ALSSQOLR Items and Factors**  
(Completely Standardized Solution)

Item Number	Negative Emotion	Interaction w/ People & Environment	Intimacy	Physical Symptoms	Bulbar Function	Religiosity
ALSSQOLR 18	.457					
ALSSQOLR 17	.443					
ALSSQOLR 21	.544					
ALSSQOLR 13	.652					
ALSSQOLR 11	.590					
ALSSQOLR 19	.646					
ALSSQOLR 31	.672					
ALSSQOLR 25	.535					
ALSSQOLR 32	.767					
ALSSQOLR 12	.736					
ALSSQOLR 22	.806					
ALSSQOLR 28	.652					
ALSSQOLR 34	.790					
ALSSQOLR 14		.622				
ALSSQOLR 37		.693				
ALSSQOLR 40		.480				
ALSSQOLR 36		.545				
ALSSQOLR 20		.544				
ALSSQOLR 27		.661				
ALSSQOLR 15		.653				
ALSSQOLR 30		.503				
ALSSQOLR 26		.718				
ALSSQOLR 16		.735				
ALSSQOLR 33		.689				
ALSSQOLR 39			.473			
ALSSQOLR 49			.463			

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	Negative Emotion	Interaction w/ People & Environment	Intimacy	Physical Symptoms	Bulbar Function	Religiosity
ALSSQOLR 43			.673			
ALSSQOLR 48			.476			
ALSSQOLR 46			.742			
ALSSQOLR 42			.702			
ALSSQOLR 45			.782			
ALSSQOLR 09				.444		
ALSSQOLR 01				.429		
ALSSQOLR 02				.684		
ALSSQOLR 10				.638		
ALSSQOLR 08				.543		
ALSSQOLR 07				.538		
ALSSQOLR 06					.846	
ALSSQOLR 04					.815	
ALSSQOLR 24					.643	
ALSSQOLR 05					.708	
ALSSQOLR 03					.510	
ALSSQOLR 38						.810
ALSSQOLR 23						.903
ALSSQOLR 35						.853
ALSSQOLR 29						.883

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**Table 6.23 Correlations between Factors on ALSSQOL-R (p = .000 for all)**

	Negative Emotion	Interaction w/ People and Environment	Religiosity	Intimacy	Physical Symptoms	Bulbar Function
Negative Emotion		.591	.170	.288	.502	.113
Interaction			.331	.439	.270	.111
Religiosity				.298	.051	-.074
Intimacy					.150	.116
Physical						.263
Bulbar						

### Normative Data

Normative data is presented for the entire sample (See Table 6.3) and by gender (See Table 6.3a, Table 6.3b), although the only statistically significant differences between means by gender were on Interaction with People and the Environment and Religiosity subscales. Analyses comparing subscale and total scores on the ALSSQOL-R across three age groups (less than 45 years, 45.1 through 65, 65.1 through oldest) revealed no differences across groups, except in bulbar function, therefore, stratifying the sample to provide age-based normative data would not add value. Likewise, ALSFRS-R scores were not correlated with ALSSQOL-R subscales or total scores, except for Physical Symptoms and Bulbar Function, thus, stratifying the sample by physical functioning levels also does not seem to add value.

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**Table 6.3 Normative/Mean scores of strength, function and quality of life in total patient sample completing the ALSSQOLR**

Measure	Mean (SD)	Range
ALSFRS-R (N=385)	(33.11) 7.95	6-48
Composite MRC (N=389)	3.59 (1.1)	0-5.0
MQOL-SIS (N=385)	6.85 (1.99)	0-10
ALSSQOL-R average total score (46-items) (N=377)	6.83 (1.2)	3.30-9.52
ALSSQOLR-Negative Emotion Subscale (N=388)	6.67 (2.0)	.77-10
ALSSQOLR-Interaction w/People & Evt (N=388)	8.17 (1.5)	2.64-10
ALSSQOLR-Intimacy (N=392)	5.85 (2.1)	.43-10
ALSSQOLR-Religiosity (N=388)	6.58 (3.4)	.00-10
ALSSQOLR-Physical Symptoms (N=392)	6.70 (1.8)	1-10
ALSSQOLR-Bulbar Function (N=392)	7.26 (2.4)	.00-10
ALSSQOLR-ALSSQOLR Total (N=377)	314.40 (56.7)	152.00-438.00

ALSFRS-R = ALS Functional Rating Scale Score-Revised; MRC = Medical Research Council; MQOL-SIS = McGill Quality of Life Single Item Scale; ALSSQOL-R = ALS-Specific Quality of Life-Revised Instrument

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**Table 6.3a Normative/Mean scores of quality of life in male patient subsample completing the ALSSQOL-R**

Measure	Mean (SD)	Range
MQOLSIS (N=227)	6.80 (1.90)	1-10
ALSSQOL-R average total score (46-items) (N=217)	6.80 (1.22)	3.30-9.30
ALSSQOLR-Negative Emotion Subscale (N=225)	6.77 (2.0)	.77-10
ALSSQOLR-Interaction w/People & Env't (N=225)	7.36 (1.5)	2.64-10
ALSSQOLR-Intimacy (N=228)	5.99 (2.1)	.86-10
ALSSQOLR-Religiosity (N=226)	6.15 (3.5)	.00-10
ALSSQOLR-Physical Symptoms (N=228)	6.75 (1.7)	1.17-10
ALSSQOLR-Bulbar Function (N=228)	7.30 (2.4)	.60-10
ALSSQOLR Total (N=217)	276.00 (56.14)	152.00-428.00

ALSRFS-R = ALS Functional Rating Scale Score-Revised; MRC = Medical Research Council; MQOLSIS = McGill Quality of Life Single Item Scale; ALSSQOL-R = ALS-Specific Quality of Life-Revised Instrument

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**Table 6.3b Normative/Mean scores of quality of life in female patient subsample completing the ALSSQOL-R**

Measure	Mean (SD)	Range
MQOLSIS	6.92 (2.11)	0-10
ALSSQOL-R average total score (46-items) (N=156)	6.86 (1.26)	3.67-9.52
ALSSQOLR-Negative Emotion Subscale (N=159)	6.52 (2.1)	.92-10
ALSSQOLR-Interaction w/People & Env't (N=159)	7.64 (1.5)	3.27-10
ALSSQOLR-Intimacy (N=160)	5.64 (2.2)	.43-10
ALSSQOLR-Religiosity (N=158)	7.22 (3.2)	.00-10
ALSSQOLR-Physical Symptoms (N= 160)	6.58 (1.8)	1.00-10
ALSSQOLR-Bulbar Function (N=160)	7.15 (2.5)	.00-10
ALSSQOLR Total (N=156)	315.78 (57.87)	169.00-438.00

ALSRFS-R = ALS Functional Rating Scale Score-Revised; MRC = Medical Research Council; MQOL-SIS = McGill Quality of Life Single Item Scale; ALSSQOL-R = ALS-Specific Quality of Life-Revised Instrument

### Validation

Multiple correlations are presented in Table 6.4. With regard to concurrent validity, the ALSSQOL-R correlated positively with the MQOL-SIS, suggesting that 58% of the variance in the single-item self-report of QOL is explained by the ALSSQOL-R. Thus, the ALSSQOL-R explained more of the variance in the MQOL-SIS than the ALSAQ-40 total scale score, any of the WHOQOL-BREF domains, or the ALSAQ-40 subscales (Table 6.5). Convergent validity was demonstrated via the positive correlation of the ALSSQOL-R average total score with the four WHOQOL-BREF

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domains Table 6.4). When testing for discriminant validity, the ALSSQOL-R average total score revealed very weak correlations with manual muscle testing as measured by the composite MRC ( $r = 0.094$ ,  $p = 0.071$  (ns)) and with the ALSFRS-R ( $r = 0.22$ ,  $p=0.000$ ), similar to the correlations of the MQOL-SIS with composite MRC ( $r = 0.18$ ,  $p = 0.000$ ) and with the ALSFRSR ( $r = 0.19$ ,  $p = 0.000$ ). This is consistent with our previous studies<sup>43,44</sup> and validation of the original ALSSQOL,<sup>19</sup> which indicated that QOL is largely determined by factors other than physical functioning. Additional testing of convergent validity by comparison of the six ALSSQOL-R subscales to other measures revealed a number of significant correlations (Table 6.6).

**Table 6.4. Correlations (Pearson product moment coefficients) between various quality of life measures in patients with ALS**

	MQOL-SIS	ALSAQ-40	WHOQOL-Physical Health	WHOQOL-Psychological	WHOQOL-Social Relationships	WHOQOL-Environment
<b>ALSSQOL-R</b>	0.58	-.049	0.54	0.68	0.58	0.46
	P=0.000	P=0.000	P=0.000	P=0.000	P=0.000	P=0.000
<b>MQOL-SIS</b>		-0.37	0.50	0.55	0.36	0.31
		P=0.000	P=0.000	P=0.000	P=0.000	P.000

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MQOL-SIS = McGill Quality of Life Single Item Scale; SEIQoL-DW – Schedule for the Evaluation of Individual QOL, Direct Weighting; WHOQOL = World Health Organization Quality of Life Instrument; ALSSQOL = ALS-Specific Quality of Life score

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**Table 6.5 Correlations (Pearson Product-Moment Coefficients) between MQOLSIS and ALSAQ-40 subscales**

ALSAQ subscale	Pearson <i>r</i>	<i>p</i>
Eating and Drinking		NS
Communication		NS
Physical Mobility	-0.222	P=0.000
ADL index	-0.258	P=0.000
Emotional Reactions	-0.507	p=0.000

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**Table 6.6. Correlations (r=Pearson product moment coefficients) between various ALSSQOLR subscales and other measures in patients with ALS**

ALSSQOLR subscale	Measure used for convergent validity	Correlation coefficient (Pearson)*	Number of subjects
Negative emotion	WHOQOL-BREF psychological domain	0.719	172
	ALSAQ40 emotional reactions scale	-0.784	211
	CES-D	-0.843	72
	BSI depression scale	-0.788	72
	BSI anxiety scale	-0.590	72
	BSI somatization scale	-0.424	72
	SPSI-R NPO	-0.431	71
	SPSI-R PPO (discrim val)	-0.220, p = .67	71
	Interaction with people and the environment	WHOQOL-BREF social relationships	0.614
WHOQOL-BREF environment		0.515	162
Satisfaction with Life Scale (SWLS)		.536	77
MSPSS Significant Other		.409	77
MSPSS Family		.513	77
MSPSS Friends		.280	77
MSPSS total score		.484	77
Intimacy	WHOQOL-BREF social relationships	0.410	166
	PAIR emotional intimacy subscale	0.310, p = .005	82
	PAIR social intimacy subscale	0.409	81
	PAIR sexual intimacy subscale	0.521	76
	PAIR intellectual intimacy subscale	0.391	81
	PAIR recreational intimacy subscale	0.369	82
	PAIR conventional intimacy subscale	0.376	81
	Religiosity	Fetzer total score	-0.770
	Fetzer forgiveness scale	-0.476	140
	Fetzer Daily Spiritual	-0.734	139

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	Experience		
	Fetzer Overall Self-Rank	-0.462	140
	Idler public subscale	0.563	139
	Idler private subscale	0.802	139
	Idler total score	0.707	139
	Facit Meaningfulness	0.351	139
	Facit Faith total	0.766	139
	FACIT SP 12 item scale	0.581	138
Physical symptoms	WHOQOL-BREF physical health	0.638	173
	ALSAQ40- physical mobility	-0.432	212
	ALSAQ40-eating and drinking	-0.179, p = .009	213
	ALSAQ40-ADL index	-0.367	213
	ALSFRSR	0.374	384
	Composite MRC	0.222	388
Bulbar function	ALSFRSR	0.384	384
	ALSAQ40-eating and drinking	-0.696	212
	ALSAQ40-communication	-0.764	213

\*p<0.001 for all correlations in table, except where noted.

As expected, the ALSSQOL-R factor “Negative Emotion” strongly correlated with the WHOQOL-BREF’s psychological domain, with the CES-D and the BSI depression and anxiety subscales. Analyses of the Negative Emotion scores by the ALS patients in comparison to their BSI-18 depression, anxiety, and somatization subscale scores suggest that patients who score 6.23 or lower on the Negative Emotion subscore should be further evaluated for depression and anxiety. This cutoff score was derived by the following procedure and analyses. The ALSSQOL-R data was separated into male and female subsets so that BSI-18 scores could be compared to the norm scores provided in the BSI-18 manual, which is stratified by sex. The ALSSQOL-R datasets were then filtered to contain only persons who scored  $\geq 7$  on the BSI-18 depression scale, and subsequently the BSI-18 anxiety scale and the BSI-Global Severity Index, which is the equivalent to the transformed T score on the BSI-18 of 63 (or greater). This score is thought to identify “caseness” (when 2 subscales of the BSI or the total BSI-Global Severity Index are 63 or

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over) as compared to the Community Sample.<sup>29</sup> Caseness suggests that these persons are a “positive case” or “at risk” for depression, anxiety, or psychological distress. The ALSSQOL-R score of 6.23 represents a conservative cutoff score equal to or under which most all persons scoring in the “caseness” range on the BSI would be captured. Of note, there was a slight discrepancy between male and female cutoff scores, but given the relatively small samples from which this data was drawn (males, N=48; females, N= 21), and compared to similar data from the original ALSSQOL and BSI dataset, it seemed prudent to choose the more conservative cutoff that would be less likely to miss distressed individuals at the risk of possibly screening a few additional patients than needed.

The moderate strength of the correlations between the factor “Interaction with People and the Environment” and two of the WHOQOL-BREF subscales suggests that while there are similarities, the ALSSQOL-R subscale provides slightly different information. This subscale correlated moderately and significantly with the SWLS and the significant other and family MPSS scales, as expected, but only had a weak correlation with the MPSS friends scale. These relationships suggest that the ALSSQOL-R Interaction subscale may include these constructs within this broader scale, but offers additional information as well.

The ALSSQOL-R “Intimacy” subscale has the strongest significant relationship with the PAIR Sexual Intimacy subscale, and shares low to moderate variability with other subscales of the PAIR, and the WHOQOL Social Relationships domain. However, when individual items of the ALSSQOLR were analyzed in comparison to PAIR subscales, and when items were combined according to desire, experience, and satisfaction with each type of intimacy (emotional, physical, sexual), the satisfaction items correlated most strongly with the PAIR subscales. Analyzing item to subscale correlations may inflate the chance of a Type I statistical error, and is interpreted with caution. However, this post hoc analysis was conducted to aid in understanding the ALSSQOLR Intimacy subscale more clearly. It is concluded that this subscale is possibly offering unique information (as provided by the additional questions on the ALSSQOL-R Intimacy subscale) about intimacy in this population compared to other available measures. Alternatively, it is noted that the ALS participants reported higher mean scores on the Satisfaction with

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Intimacy scale, suggesting that either the comparative norm scores of the PAIR samples are not appropriate for comparison to an older population with medical concerns, such as individuals with ALS, or the nature of administering the PAIR via interview has negatively affected the validity of the measure by, perhaps, eliciting inflated (“fake good”) responses. Thus, while it appears that the PAIR suggests the ALSSQOL-R Intimacy subscale focuses highly on sexual intimacy, further evaluation of this construct in future studies is indicated.

The ALSSQOL-R “Religiosity” component strongly correlated with several of the FACIT-SP and Fetzer subscales and the Idler subscales and total score. However, Religiosity had only small correlations with the Fetzer Forgiveness and Facit Meaningfulness subscales, offering some discriminant validity information. Further analysis revealed that the ALSSQOL-R Religiosity subscale correlated highly with the Faith in Illness Scale of the FACIT-SP-12 ( $r = .77, p = .000, N=139$ ), but not with the FACIT-SP-12 Meaningfulness scale ( $r = .35, p = .000, N=139$ ). Examination of the items on these FACIT-SP-12 scales provide a context for understanding these statistics. The Faith in Illness subscale offers statements for endorsement as follows, “I find comfort in my faith or spiritual beliefs,” “I find strength in my faith or spiritual beliefs,” “My illness has strengthened my faith or spiritual beliefs,” “I know that whatever happens with my illness, things will be ok.” The Meaningfulness scale includes the following statements, “I feel peaceful,” “I have reason for living,” “My life has been productive,” “I have trouble feeling peace of mind,” “I feel a sense of purpose in my life,” “I am able to reach down deep into myself for comfort,” “I feel a sense of harmony within myself,” “My life lacks meaning and purpose.” The Meaningfulness subscale correlated strongly with the ALSSQOL-R Negative Emotion subscale, suggesting a strong relationship ( $.729, p = .000, N = 139$ ) exists between negative emotion and existential concerns. The Faith subscale had only a small correlation with Negative Emotion ( $.391, p = .000, N=139$ ).

The ALSSQOLR “Physical Symptoms” subscale positively correlated with the WHOQOL-BREF physical health domain, but only slightly with the composite MRC score, and some of the ALSAQ-40 subscales. It is important to recognize that the ALSSQOL-R Physical Symptoms

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subscale reflects “How much of a problem” the physical symptoms are for individuals, rather than how much of a physical disability they have, according to objective measures.

“Bulbar Function” as measured by the ALSSQOLR subscale, correlated strongly with the ALSAQ40 Communication subscale and with the ALSAQ-40 Eating/Drinking Subscale. The correlation between the Bulbar Function subscale of the ALSSQOL-R and the total ALSFRS-R score was weaker, as expected, since the total score includes other physical symptoms.

Using hierarchical multiple regression analysis with ALSSQOL-R subscale scores as predictors, and the MQOL-SIS as the criterion variable, Negative Emotion, Physical Symptoms, and Intimacy explained 37%, 2%, and 1%, respectively, of the variance in self-reported quality of life ( $F(3, 371) = 82.44, p = .000$ ); Satisfaction with People and the Environment, Religiosity, and Bulbar Function did not enter into this equation. These findings are similar to the analyses conducted with the original ALSSQOL. However, these variables are still considered significant contributors to the understanding of QOL, as indicated by ALS patients in previous interviews and studies.

## **Chapter 7: Concluding Comments**

This manual describes the ALSSQOL-R scale designed to assess QOL in individuals with ALS. The ALSSQOL-R has been developed and validated for clinical and research use. The ALSSQOL-R is a tool for assessing QOL and the contributing domains at a single point and over time in ALS individuals and groups. Use of this tool will allow further questions to be asked about the impact of ALS on QOL, the relationships within each domain and between domains over time, and the effects of clinical interventions on QOL for individuals and groups.

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## Appendix A: ALSSQOL-R Instrument

### ALS-Specific Quality of Life Questionnaire-Revised

Participant ID# \_\_\_\_\_

Date: \_\_\_\_\_

#### Instructions:

The questions in this questionnaire begin with a statement followed by two opposite answers. Numbers extend from one extreme answer to its opposite. Please circle the number between 0 and 10 which is most true for you. There are no right or wrong answers. Completely honest answers will be most helpful.

#### EXAMPLE:

	Not at All										Extremely
I am hungry.	0	1	2	3	4	5	6	7	8	9	10

If you are not even a little bit hungry, you should circle 0.

If you are a little hungry (you just finished a meal but still have room for dessert), you might circle 1, 2, or 3

If you are feeling moderately hungry (because mealtime is approaching), you might circle 4, 5, or 6.

If you are very hungry (because you haven't eaten all day), you might circle a 7, 8, or 9.

If you are extremely hungry, you should circle 10.

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**BEGIN HERE:**

Please assess your overall quality of life over the past week (7 days):

Very bad	Excellent
Considering all parts of my life – physical, emotional, social, spiritual, and financial – over the past week, the quality of my life has been.	
0	1 2 3 4 5 6 7 8 9 10

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Please rate the following symptoms and experiences according to how much of a problem each one has been for you. Please respond about how you have felt or what you have experienced over the past week using the scale provided.

		No Problem											Tremendo us Problem
1.	Pain	0	1	2	3	4	5	6	7	8	9	10	
2.	Fatigue	0	1	2	3	4	5	6	7	8	9	10	
3.	Eating	0	1	2	3	4	5	6	7	8	9	10	
4.	Excessive Saliva	0	1	2	3	4	5	6	7	8	9	10	
5.	Mucous in My Throat	0	1	2	3	4	5	6	7	8	9	10	
6.	Speaking	0	1	2	3	4	5	6	7	8	9	10	
7.	My Strength and Ability to Move	0	1	2	3	4	5	6	7	8	9	10	
8.	Sleep	0	1	2	3	4	5	6	7	8	9	10	
9.	Bowel and Bladder (Constipation, Diarrhea, Poor Control)	0	1	2	3	4	5	6	7	8	9	10	

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Please rate the following statements according to how strongly you agree or how strongly you disagree with each of them. Please respond about how you have felt or what you have experienced over the past week.

			Strongly Disagree										Strongly Agree
10.	I have felt physically terrible.	0	1	2	3	4	5	6	7	8	9	10	
11.	My life has been purposeful and meaningful.	0	1	2	3	4	5	6	7	8	9	10	
12.	I have been coping well with my illness.	0	1	2	3	4	5	6	7	8	9	10	
13.	I believe I have control over my life.	0	1	2	3	4	5	6	7	8	9	10	
14.	When I have thought about my life, I thought that my life to this point has been worthwhile.	0	1	2	3	4	5	6	7	8	9	10	
15.	The world has been caring and responsive to my needs.	0	1	2	3	4	5	6	7	8	9	10	
16.	I have felt supported.	0	1	2	3	4	5	6	7	8	9	10	
17.	ALS has interfered with the important things in my life.	0	1	2	3	4	5	6	7	8	9	10	
18.	The past week has been a gift.	0	1	2	3	4	5	6	7	8	9	10	
19.	I have felt good about myself as a person.	0	1	2	3	4	5	6	7	8	9	10	
20.	When I have thought about my whole life, I thought that I have achieved my life's goals.	0	1	2	3	4	5	6	7	8	9	10	
21.	Whatever the future holds, I know that things will be ok.	0	1	2	3	4	5	6	7	8	9	10	

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Please rate the following statements according to how much you have felt or experienced what is described. Please respond about how you have felt or what you have experienced over the past week.

		0	1	2	3	4	5	6	7	8	9	10	Very Much
22.	I have been depressed.	0	1	2	3	4	5	6	7	8	9	10	
23.	My religion has been a source of strength or comfort to me.	0	1	2	3	4	5	6	7	8	9	10	
24.	Communication has been a problem.	0	1	2	3	4	5	6	7	8	9	10	
25.	When I have thought of the future, I have been afraid.	0	1	2	3	4	5	6	7	8	9	10	
26.	Relationships with those closest to me have been satisfying.	0	1	2	3	4	5	6	7	8	9	10	
27.	I have been interested in other people or things.	0	1	2	3	4	5	6	7	8	9	10	
28.	I have been nervous or worried.	0	1	2	3	4	5	6	7	8	9	10	
29.	I consider myself to have been religious or spiritual.	0	1	2	3	4	5	6	7	8	9	10	
30.	I enjoyed spending time with other people.	0	1	2	3	4	5	6	7	8	9	10	
31.	I have felt helpless.	0	1	2	3	4	5	6	7	8	9	10	
32.	I have felt hopeless.	0	1	2	3	4	5	6	7	8	9	10	
33.	I have enjoyed the beauty of my surroundings.	0	1	2	3	4	5	6	7	8	9	10	

Please rate the following statements according to how often you have felt or experienced what is described. Please respond about how you have felt or what you have experienced over the past week.

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		Never										Very Often
34.	I have felt sad.	0	1	2	3	4	5	6	7	8	9	10
35.	I have prayed to God.	0	1	2	3	4	5	6	7	8	9	10
36.	I have laughed.	0	1	2	3	4	5	6	7	8	9	10
37.	I was excited about or looked forward to something.	0	1	2	3	4	5	6	7	8	9	10
38.	I have engaged in religious practices in my home or in my community.	0	1	2	3	4	5	6	7	8	9	10

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The following statements are about social contact (for example, visits from family and friends). Please think about your experiences with or how you have felt about social contact in the past week, and use the scales provided below to respond.

39.	My desire for social contact has been strong.	Strongly Disagree	0	1	2	3	4	5	6	7	8	9	10	Strongly Agree
40.	Family and friends have visited me.	Never	0	1	2	3	4	5	6	7	8	9	10	Very Often
41.	Visits from family and friends have been satisfying.  (If you have not had any visits, please leave the response section blank).	Not at All	0	1	2	3	4	5	6	7	8	9	10	Very Much

The following statements are about emotional intimacy (for example, sharing deep, private thoughts; feeling connected). Please think about your experiences with or how you have felt about emotional intimacy in the past week, and use the scales provided below to respond.

42.	My desire for emotional intimacy has been strong.	Strongly Disagree	0	1	2	3	4	5	6	7	8	9	10	Strongly Agree
43.	I have shared emotional intimacy with others.	Never	0	1	2	3	4	5	6	7	8	9	10	Very Often

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44.	Emotional intimacy with others has been satisfying.  (If you have not shared emotional intimacy, please leave the response section blank).	Not at All	0	1	2	3	4	5	6	7	8	9	10	Very Much
-----	--	------------	---	---	---	---	---	---	---	---	---	---	----	-----------

The following statements are about physical intimacy (for example, touching, hugging, kissing). Please think about your experiences with or how you have felt about physical intimacy in the past week, and use the scales provided below to respond.

45.	My desire for physical intimacy has been strong.	Strongly Disagree	0	1	2	3	4	5	6	7	8	9	10	Strongly Agree
46.	I have shared physical intimacy with others.	Never	0	1	2	3	4	5	6	7	8	9	10	Very Often
47.	Physical intimacy with others has been satisfying.  (If you have not shared physical intimacy, please leave the response section blank).	Not at All	0	1	2	3	4	5	6	7	8	9	10	Very Much

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The following statements are about sexual intercourse. Please think about your experiences with or how you have felt about sexual intercourse in the past week, and use the scales provided below to respond.

48.	My desire for sexual intercourse has been strong.	Strongly Disagree	0	1	2	3	4	5	6	7	8	9	10	Strongly Agree
49.	I have shared sexual intercourse with others.	Never	0	1	2	3	4	5	6	7	8	9	10	Very Often
50.	My sexual relationship has been satisfying.  (If you have not had sexual intercourse, please leave the response section blank).	Not at All	0	1	2	3	4	5	6	7	8	9	10	Very Much

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## Appendix B: ALSSQOL-R Hand Score Sheet

Page 1

Subject ID#:

Date Obtained: \_\_/\_\_/\_\_\_\_

1. 10 - __ =	26.
2. 10 - __ =	27.
3. 10 - __ =	28. 10 - __ =
4. 10 - __ =	29.
5. 10 - __ =	30.
6. 10 - __ =	31. 10 - __ =
7. 10 - __ =	32. 10 - __ =
8. 10 - __ =	33.
9. 10 - __ =	34. 10 - __ =
10. 10 - __ =	35.
11.	36.
12.	37.
13.	38.
14.	39.
15.	40.
16.	41. do not use this value
17. 10 - __ =	42.
18.	43.
19.	44. do not use this value
20.	45.
21.	46.
22. 10 - __ =	47. do not use this value
23.	48.
24. 10 - __ =	49.
25. 10 - __ =	50. do not use this value
Subtotal for Column =	Subtotal for Column =
	Total ALSSQOL-R =
	AVG ALSSQOL-R = Total/46 =

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Page 2

Subject ID#:

Date Obtained: \_\_/\_\_/\_\_\_\_

Average Subscores:

Negative Emotion:  $(11 + 12 + 13 + 17 + 18 + 19 + 21 + 22 + 25 + 28 + 31 + 32 + 34) / 13.$

= \_\_\_\_\_

Interaction with People and the Environment:  $(14 + 15 + 16 + 20 + 26 + 27 + 30 + 33 + 36 + 37 + 40) / 11.$

\_\_\_\_\_

Intimacy:  $(39 + 42 + 43 + 45 + 46 + 48 + 49) / 7.$  = \_\_\_\_\_

Religiosity:  $(23 + 29 + 35 + 38) / 4$  = \_\_\_\_\_

Physical Symptoms:  $(1 + 2 + 7 + 8 + 9 + 10) / 6.$  = \_\_\_\_\_

Bulbar Function:  $(3 + 4 + 5 + 6 + 24) / 5.$  = \_\_\_\_\_

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## Appendix C: ALSSQOL-R Computer Score Syntax

Reverse Code ALSSQOLR Items.

RECODE

q1 q2 q3 q4 q5 q6 q7 q8 q9 q10 q17 q22 q24 q25 q28 q31 q32 q34

(0=10) (1=9) (2=8) (3=7) (4=6) (5=5) (6=4) (7=3) (8=2) (9=1)

(10=0) .

EXECUTE .

Calculate Average Total ALSSQOLR Score.

COMPUTE Avg TotalQOL = SUM(q1,q2,q3,q4,q5,q6,q7,q8,q9,q10,q11,q12,q13,q14,q15,q16

,q17,q18,q19,q20,q21,q22,q23,q24,q25,q26,q27,q28,q29,q30,q31,q32,q33,q34,q35

,q36,q37,q38,q39,q40,q42,q43,q45,q46,q48,q49) / 46 .

EXECUTE .

Compute Negative Emotion Factor Score.

Compute Negative\_Emotion = (q11 + q12 + q13 +q17 +q18+q19 + q21 +q22 + q25 + q28 + q31 + q32+q34)  
/ 13.

EXECUTE.

Compute Interaction with People and the Environment Score.

Compute Interaction = (q14 + q15 + q16 + q20 + q26 + q27 + q30 + q33 + q36 + q37 + q40) / 11.

EXECUTE.

Compute Intimacy Score.

Compute Intimacy = (q39 + q42 + q43 + q45 + q46 + q48 + q49) / 7.

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EXECUTE.

Compute Religiosity Score.

Compute Religiosity =  $(q23 + q29 + q35 + q38) / 4$ .

EXECUTE.

Compute Physical Symptoms Score.

Compute Physical =  $(q1 + q2 + q7 + q8 + q9 + q10) / 6$ .

EXECUTE.

Computer Bulbar Function Score.

Compute Bulbar =  $(q3 + q4 + q5 + q6 + q24) / 5$ .

EXECUTE.

## Appendix D: Items by Factor

### Negative Emotion

My life has been purposeful and meaningful.

I have been coping well with my illness.

I believe I have control over my life.

ALS has interfered with the important things in my life.

I have felt good about myself as a person.

When I have thought about my whole life, I thought that I have achieved my life's goals.

Whatever the future holds, I know that things will be ok.

I have been depressed.

When I have thought of the future, I have been afraid.

I have been nervous or worried.

I have felt helpless.

I have felt hopeless.

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## Interaction with People and the Environment

When I have thought about my life, I thought that my life to this point has been worthwhile.

The world has been caring and responsive to my needs.

I have felt supported.

I have felt good about myself as a person.

Relationships with those closest to me have been satisfying.

I have been interested in other people or things.

I enjoyed spending time with other people.

I have enjoyed the beauty of my surroundings.

I have laughed.

I was excited about or looked forward to something.

Family and friends have visited me.

## Intimacy

My desire for social contact has been strong.

My desire for emotional intimacy has been strong

I have shared emotional intimacy with others.

My desire for physical intimacy has been strong.

I have shared physical intimacy with others.

My desire for sexual intercourse has been strong.

I have shared sexual intercourse with others.

# ALS Specific Quality of Life: User's Guide

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## Religiosity

My religion has been a source of strength or comfort to me.

I consider myself to have been religious or spiritual.

I have prayed to God.

I have engaged in religious practices in my home or in my community.

## Physical Symptoms

Pain

Fatigue

My Strength and Ability to Move

Sleep

Bowel and Bladder (Constipation, Diarrhea, Poor Control)

I have felt physically terrible.

## Bulbar Function

Eating

Excessive Saliva

Mucous in My Throat

Speaking

Communication has been a problem.

### **Appendix E: Participating ALS Clinics and Directors**

Penn State Hershey Medical Center: Zachary Simmons, MD

John P. Murtha Neuroscience and Pain Institute: Zachary Simmons, MD

Drexel University, Philadelphia, PA: Terry Heiman-Patterson, MD

Hennepin County Hospital, Minneapolis, MN: Ezgi Tiryaki, MD

Johns Hopkins University: Lora Clawson, MSN, CRNP

Mayo Clinic Jacksonville: Kevin Boylan, MD

Providence ALS Center, Portland, OR: Kimberly Goslin, MD

Stanford University, Stanford, CA: S. Charles Cho, MD

University of Kentucky, Lexington, KY: Edward Kasarskis, MD

University of Pennsylvania: Leo McCluskey, MD

University of Utah: Mark Bromberg, MD

University of Vermont: Rup Tandan, MD