Quality of Life in ALS: Often Discussed, Rarely Understood

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“The quality of life is more important than life itself.”

Alexis Carrel
Measuring Quality of Life in ALS
Why Measure QOL in ALS?

• Current treatments cannot stop or reverse ALS
  – Care centers on maintaining or improving QOL
• Future treatments may improve physical function but have negative side effects
  – The value of a treatment will be judged by how it impacts overall QOL.
How is QOL Measured in ALS?

- No consensus regarding the best instrument
- No consensus regarding which factors are most important
“Yes, it all comes down to quality of life. An inheritance sure would help.”
Why Do We Need Specific Instruments to Measure QOL?

Why Not Just Ask Patients to Assess their QOL?
QOL Instruments Add Value to Our Care

• QOL instruments help the care team understand our patients
  – What factors help/hurt QOL?
  – How do these factors change over time?
  – How can we as care providers use this information to maintain or improve QOL for our patients and their families?
Early Attempts at Measuring QOL in ALS

- Instruments were heavily weighted toward strength and physical function
- Inevitably declined over time as patients lost strength
Can Individuals With ALS Have a High QOL? Yes!

• QOL in ALS is underestimated by caregivers

• Caregivers rated patients with ALS as having less energy, greater suffering, and greater weariness than patients indicated for themselves

• The key to measuring QOL in people with ALS is to look beyond their physical abilities
QOL vs. Health-Related QOL vs. Health Status

- Health-related quality of life = health status
  - Measures health-related factors
  - May be physical or psychological
- Most questionnaires assessing health-related QOL do not measure overall QOL, which includes jobs, family, friends, other sources of emotional support, and considers the patient’s values, preferences, and expectations.
- Two individuals with the same health status may have very different qualities of life!!
Must QOL in ALS Inevitably Decline Over Time? No!

- Health-related QOL declines over time, but global QOL may be maintained by other factors
  - Measures of health-related QOL do not capture all factors which determine QOL in patients with ALS
Importance of Factors Other Than Strength and Physical Function in Determining Quality of Life in Individuals with ALS
Cross-Sectional Study of QOL: Penn State Data

- 96 patients with definite or probable ALS
- QOL, as assessed by the patient with ALS, did not correlate with strength and physical function, but depended on psychological and existential factors
- Spiritual factors and support systems played roles
Longitudinal Study of QOL: Penn State Data

- 60 patients with ALS assessed 3 times at 3 month intervals
- QOL is determined by factors other than physical function, and does not necessarily decline as patients lose function
Religiousness is related to quality of life in patients with ALS

Susan M. Walsh, RN, MSN, CS; Barbara A. Bremer, PhD; Stephanie H. Felgoise, PhD; and Zachary Simmons, MD
Role of Spirituality and Religion in QOL: Penn State Data

- 49 patients with ALS followed for one year
- QOL was not significantly correlated with religiousness at the onset of the study, but over time a significant relationship developed
- Religiousness plays an increasingly important role in maintenance of QOL for people with ALS as disease progresses
Schedule for the Evaluation of Individual QOL

- SEIQoL and SEIQoL-DW (direct weighted)
- QOL is determined by the individual
  - Does not decline as patients weaken
  - Factors of importance to patients in maintaining QOL evolve over time
SEIQoL-DW in ALS:
Penn State-Utah Collaborative Data
Findings

• Administered to 120 pts at 2 centers
• No significant relationship between SEIQoL-DW scores and physical function
• QOL as defined by the patient with ALS is determined largely by non-health-related factors
600 Cues elicited: Breakdown by Category

- Family/significant others – 31.17%
- Hobbies – 17.50%
- Religion – 12.17%
- Physical Function – 9.17%
- Friends – 7.83%
- Health – 6.17%
- Psychosocial or existential – 5.67%
- Finances – 3.83%
- Work – 3.50%
- Misc – 3.00% (largest single item was pets)
% of Subjects Nominating Each Cue

- Family/significant others – 90%
- Hobbies – 54.17%
- Religion – 55.83%
- Physical Function – 29.17%
- Friends – 39.17%
- Health – 27.50%
- Psychosocial/existential – 24.17%
- Finances – 18.33%
- Work – 15.83%
- Misc – 14.17%
Average Weight for Each Cue

- Family/significant others – 26.28%
- Hobbies – 14.81%
- Religion – 23.55%
- Physical Function – 16.23%
- Friends – 16.28%
- Health – 18.34%
- Psychosocial/existential – 17.51%
- Finances – 12.63%
- Work – 14.40%
- Misc – 15.94%
QOL in ALS: Conclusions from Penn State Studies

- Psychological, existential, support, and spiritual or religious factors play a significant role in determining QOL in these patients, while strength and physical function are less important.
- The existing tools which we have used do not capture all factors which determine QOL in patients with ALS, and should be modified.
- But, the SEIQoL-DW is not a useful tool for large studies.
  - This led to the development of a new instrument.
The ALS-Specific Quality of Life Questionnaire

ALSSQOL
ALSSQOL – General Description

• A 59-item questionnaire using a 0 to 10-point scale for each question
• Inquires more broadly into physical symptoms and spirituality/religiousness, and asks questions about intimacy, loneliness, relationships, environment, social interaction, values, coping, and interests/desires/goals
Validation of ALSSQOL

• Seven university-based ALS clinics recruited 342 persons with ALS
• 6 categories were identified as being important to QOL. Questions were asked in each category
  – Negative Emotion (examples to follow)
  – Interaction with People and the Environment (examples to follow)
  – Intimacy
  – Religion
  – Physical Symptoms
  – Bulbar Function
Negative Emotion: Examples

- My life has been purposeful, meaningful
- I have been coping well with my illness
- I believe I have control over my life
- ALS has interfered with important things
- I have been depressed
- I have felt helpless
- I have felt hopeless
- I have been nervous or worried
Interaction with People and the Environment: Examples

- Relationships with those closest to me have been satisfying
- I’ve been interested in other things, people
- I enjoyed spending time with other people
- I’ve enjoyed beauty of my surroundings
- I was excited about or looked forward to something
- Family and friends have visited me
ALSSQOL - Conclusions

• The 46-item ALSSQOL shows promise as a useful instrument for measuring QOL in patients with ALS
• We are organizing a multicenter study to validate this shorter instrument, which we hope may be generally useful for assessing QOL in individuals with ALS
What Next?

• Assessment of individuals with ALS who do not attend multidisciplinary clinics: what is their QOL like?
  – Web-based. A trial is ongoing
  – In collaboration with the ALS Association

• Goal – To design interventions
  – This is the purpose of all this work!!
  – We are looking at ways to improve QOL, based on what we now know is important
Caregiver QOL: Penn State Data

- Social problem-solving skills and spirituality were the best predictors of caregiver QOL.
- Highest QOL was found in caregivers who viewed problems as challenges rather than threats.
- The level of care provided to ALS patients was not a predictor of caregivers’ QOL.
- Interventions aimed at strengthening ALS caregivers’ social problem-solving abilities could be beneficial.
ALSA Certified Centers: Core Team

- Neurologist
- Nurse
- Physical therapist
- Occupational therapist
- Speech and language pathologist
- Registered dietician
- Respiratory therapist
- Psychologist or psychiatrist
- Social worker
Welcome!

Comprehensive, multidisciplinary treatment for ALS patients and their families
Effect of a multidisciplinary amyotrophic lateral sclerosis (ALS) clinic on ALS survival: a population based study, 1996–2000

B J Traynor, M Alexander, B Corr, E Frost, O Hardiman

J Neurol Neurosurg Psychiatry 2003;74:1258–1261
Multidisciplinary ALS Clinics: Effect on Survival

- 82 Irish patients with ALS attending multidisciplinary clinic and 262 followed in “general neurology clinics.”
- Prognosis in multidisciplinary clinic was better
  - Median survival was 7.5 months longer
  - Reduction in mortality: 29.7% at 1 year, 10.7% at 2 years
- For bulbar ALS, differences were more striking
  - Median survival was 9.6 months longer
  - Reduction in mortality: 39% at 1 year, 18% at 2 years
Multidisciplinary ALS care improves quality of life in patients with ALS

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Multidisciplinary ALS Clinics: Effect on Quality of Life

- 133 Dutch patients with ALS who received care from a multidisciplinary ALS team and 75 patients receiving general ALS care (general practitioner, physician in a nursing home, or rehabilitation specialist)
- The mental health overall was better in the multidisciplinary group as assessed by the mental health section of the SF-36
“We must ever be mindful that our function as physicians is to cure when we can; if we cannot cure, let us treat if we can; if we cannot treat, let us comfort.” - Milhorat
“When [no therapy] avails to ward off the fatal ending, it is still no small portion of [the physician's] art to rid his patient's path of thorns if he cannot make it bloom with roses.” - Alfred Stille, address to the medical classes of the University of Pennsylvania in 1884
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