2007

Does Verbal Communication Impairment Affect Quality of Life in Amyotrophic Lateral Sclerosis Patients?

Jason Michael Duff
Philadelphia College of Osteopathic Medicine, jasondu@pcom.edu

Philadelphia College of Osteopathic Medicine

Department of Psychology

DOES VERBAL COMMUNICATION IMPAIRMENT AFFECT QUALITY OF LIFE IN AMYOTROPHIC LATERAL SCLEROSIS PATIENTS?

By Jason Michael Duff

Submitted in Partial Fulfillment of the Requirements of the

Degree of Doctor of Psychology

May 2007

PHILADELPHIA COLLEGE OF OSTEOPATHIC MEDICINE
DEPARTMENT OF PSYCHOLOGY

Dissertation Approval

This is to certify that the thesis presented to us by ___Jason Duff_____

on the __18th____ day of ____May_____, 2007, in partial fulfillment of the requirements for the degree of Doctor of Psychology, has been examined and is acceptable in both scholarship and literary quality.

Committee Members' Signatures:

Stephanie H Felgoise, PhD, ABPP, Chairperson

Robert A DiTomasso, PhD, ABPP

Dr Zachary Simmons

Robert A DiTomasso, PhD, ABPP, Chair, Department of Psychology

Acknowledgements

I am forever indebted to all of those who have guided, supported, mentored and encouraged me throughout this academic, professional and personal journey.

I would like to thank Dr. Stephanie Felgoise, Committee Chairperson, for her continued support in helping me to complete this project in such a timely manner. I appreciate the opportunity to have produced a body of work of which I am proud. This body of work will undoubtedly continue to provide me with compelling professional opportunities in the future. Dr. Felgoise's enthusiasm and passion surrounding ALS and the research process was exciting to be around.

I would like to thank Dr. Robert DiTomasso. Not only did the persistently hard-at-work Dr. DiTomasso make himself available to be on my committee, but he was also the motivating force behind my choice to pursue a Doctorate in Clinical Psychology at PCOM.

I am grateful to Dr. Zachary Simmons. He and his team's dedication towards this important topic has influenced and helped many people. I am delighted to be associated now with such special people.

I would also like to thank Dr. Arthur Freeman. From the start, he has been a tremendous influence. It has been my pleasure to work so closely with him over these past few years. His support and guidance especially regarding my ultimate decision to join the military was priceless. He continually spoke highly of me when he encountered both Navy and Air Force personnel.

Dr. Bruce Zahn perhaps has been the biggest influence in the sense that it is he who first recommended a military internship. His elegantly voiced opinion regarding the most significant decision I have ever made was truly valued.

To the entire faculty that I have worked with over the past few years: I appreciate every ounce of support, criticism, and kindness that was bestowed upon me.

I wish to extend heartfelt thanks to my colleagues who were always there for me: Sandy Capaldi, Brian Eig, Steve Timchak, and Donna Yaure. Your friendship, support and dedication through every daunting moment will never be forgotten.

I am grateful to my family who has always been there for me. Their steadfast belief in me has always provided a safety net to continue on and to see how far I could go. I

am the person I am today because of each and every one of you.

And finally, I express extreme gratitude to Patricia Anne Doyle. She has been unwavering in her support and belief in me. She has been an inspiration to me and I hope to always return the favor.

Abstract

Does Verbal Communication Impairment Affect Quality of Life in Amyotrophic Lateral Sclerosis Patients?

The purpose of this study was to examine the self-perceived QOL in ALS patients. Literature will be presented on the incidence, prevalence, prognosis, diagnosis and management of ALS, QOL studies for ALS, the role of the multidisciplinary team, the impairments and dysfunction that ALS patients experience, communication issues, and the development of ALS specific instruments to measure QOL.

The "bulbar dysfunction" that ALS patients experience in salivation management, speech, and swallowing were examined in detail. The objectives of this research study were to investigate the following hypotheses: 1. QOL will differ among ALS patients with varying levels of speech, swallowing, and salivation functioning, 2. Patients with less impairment in these aspects of physical functioning will report better QOL.

Archival data was obtained from a validation study for the ALSSQOL instrument that employed 7 university-based ALS centers. ANOVA revealed that self-reported QOL varied

according to level of functioning for speech F (4,333) = 5.13, p =.001; swallowing F(4, 333)= 6.88, p=.000; and salivation, F(4,333)= 3.75, p =.000.

This research is important because it showed that QOL is adversely affected by impaired communication abilities. Having this knowledge will allow mental health providers to tailor time-sensitive interventions more appropriately, perhaps enhancing ALS patients' QOL.

Areas of future consideration include utilization of the ALSSQOL for longitudinal studies and for investigation of ALS patients' mindsets as they prepare to experience each of the transitions during this predictable disease process.

Table of Contents

Tables

Page

Introduction and Literature Review

Statement of the Problem

Although it is an uncommon disease, Amyotrophic Lateral Sclerosis (ALS) has strong celebrity ties and often receives a fair amount of media attention. Famous sufferers of ALS, such as Lou Gehrig and Stephen Hawkings have created national and worldwide awareness of the disease. Despite its infrequency, ALS has attracted a great deal of attention in many different ways.

Recently ALS has been spoken about in the news because of reports that there is evidence of elevated incidences in those who served in the 1991 Gulf War. Research on this topic is ongoing.

Mitch Albom's bestseller, *Tuesday's with Morrie*, is a true story about a student rekindling his relationship with his former mentor after learning that the mentor has ALS.

In February of 2006, Oscar-nominated actor James Woods played a wheelchair bound doctor suffering from ALS on the hit medical drama, ER. As his character's condition worsens he becomes forced to utilize an electronic device to help him speak.

Although there has been increased attention on ALS in the news and entertainment industry, the attention that ALS has received from the medical community has been even more extensive in recent years. The increased focus has been on such issues as diagnostic considerations (Brooks, Miller, Swash & Munsat, 2000), multidisciplinary approaches to care (Lechtzin, Schmidt & Clawson, 2005; Traynor, Alexander, Corr, Frost & Hardiman, 2003), medical progress (Rowland & Shneider, 2001), and quality of life (QOL; Simmons, 2005; Lo Coco, Lo Coco, Cicero, Oliveri, Lo Verso, Piccoli, et al., 2005; Chio, Gauthier, Montuschi, Calvo, Di Vito, Ghiglione, et al., 2004; Bremer, Walsh, Simmons & Felgoise, 2004; Walsh, Bremer, Felgoise & Simmons, 2003).

Precise attention will be paid to the concept of quality of life throughout this text. QOL is a difficult concept to discuss because there is no consensual definition. Advertising executives, real estate agents and politicians alike inform the public about how they can improve quality of life by buying a specific product, living in a specified area or voting for a particular candidate. The concept of QOL is also used extensively throughout the healthcare system. Research can be found applying the concept in the fields of nursing, medicine and

other allied disciplines. Over the past three decades thousands of articles related to QOL have been published. A brief literature review revealed papers on QOL related to diabetes, obesity, pulmonary disease, cerebral vascular accident, systemic lupus, sarcoidosis, gastroesophageal reflux disease and an assortment of other illnesses of varying severity.

The purpose of the current study will be to examine the self-perceived QOL in ALS patients. Precise focus will be on verbal communication impairment and the effect that this impairment has or does not have on QOL. Literature will be presented on such issues as the incidence, prevalence and prognosis of ALS, the diagnosis and management of ALS, QOL studies for ALS, the role of the multidisciplinary team in working with ALS patients, the impairments and dysfunction that ALS patients experience, communication issues, and the development of ALS-specific instruments to measure QOL.

The "bulbar dysfunction" that ALS patients experience in salivation management, speech, and swallowing will be examined in detail. ALS presents in various ways. It is possible that some patients will have the symptoms of bulbar dysfunction early in the course of their illnesses,

on the other hand, others will not manifest such symptoms until later in the disease progression.

The objectives of this research study are to investigate the following hypotheses:

1. QOL will differ among ALS patients with varying levels of speech, swallowing, and salivation functioning,

2. Patients with less impairment in these aspects of physical functioning will report better QOL.

ALS Information

What is ALS?

Amyotrophic lateral sclerosis (ALS) is an inexorable, rapidly progressive, neurological disease that is consistently fatal. It affects nerve cells in the brain and the spinal cord that control voluntary muscle movement.

French neurologist Jean-Martin Charcot initially described the disease in the 19th century and named it amyotrophic lateral sclerosis. Amyotrophic refers to the muscle atrophy, weakness, and fasiculations that suggest disease of the lower motor neurons. Lateral sclerosis refers to the hardness observed when palpating the lateral

columns of the spinal cord in autopsy specimens (Rowland & Shneider, 2001).

Amyotrophic is derived from the Greek language, "A" meaning no or negative, "myo" referring to muscles, and "trophic" meaning nourishment; therefore, it literally means "no muscle nourishment" (The ALS Association, 2006). In the United States of America it is generally referred to as "ALS" or "Lou Gehrig's disease". Lou Gehrig, the "Iron Horse", was a professional baseball player for the New York Yankees who was the epitome of reliability and consistency. Gehrig set a record by playing in a consecutive streak of 2,130 professional baseball games throughout his career, despite sustaining 17 fractures in his hands, having severe back pain and suffering various other illnesses and minor injuries. A columnist once referred to him as a "symbol of indestructibility…a Gibraltar in cleats." However, in 1939 he hastily retired "for the good of the team" after he realized that he was not playing well and that something was wrong, physically. A few months later he was diagnosed with ALS; two years after the diagnosis he was dead. This point serves to elucidate the facts that not only does ALS fail to discriminate but also that it serves to severely debilitate in a relatively short time.

In the United Kingdom and elsewhere, the umbrella term motor neuron disease (MND) is more commonly used to describe this combination of upper and lower motor neuron dysfunction (Talbot, 2002). There are, however, forms of motor neuron degeneration that selectively affect upper or lower motor neurons; however, they will not be the focus of this discussion or investigation.

ALS results in the diffuse degeneration and death of motor neurons, leading to muscle atrophy, which invariably culminates in respiratory insufficiency and eventually death (Lechtzin, Schmidt & Clawson, 2005; Simmons, 2005). Before death, which at this time seems the unavoidable consequence of the disease, the patients' muscles gradually deteriorate, waste away, and twitch. Muscle strength erodes and all voluntary muscle control is ultimately lost. The inevitable respiratory insufficiency secondary to the impairment of the respiratory musculature is the most frequent cause of death in patients with ALS (Farrero, Prats, Povedano, Martinez-Matos, Manresa & Escarrabill, 2005).

The clinical presentation of ALS depends on the area of the nervous system that has been damaged. ALS can present primarily with limb involvement, yet bulbar

symptoms are exhibited initially in 19% to 25% of ALS cases (Walling, 1999).

Because of the harsh reality that ALS cannot be cured or its progression halted, the primary goal in treating patients with ALS is often palliative in nature. Optimizing quality of life is the main focus of patient management (Simmons, 2005).

Incidence, Prevalence and Prognosis

There has been a push in recent years to establish a national registry that would promote a better understanding of ALS. Legislation is currently pending to establish a national registry here in the United States. Among other things, a single registry would allow for data collection on incidence and prevalence, environmental and occupational factors that may be associated with the disease, the age, race/ethnicity, gender and family history of individuals diagnosed with the disease. Presently, it is estimated that as many as 30,000 Americans have the disease at any given time. The prevalence is believed to be six to eight persons per 100,000 (ALS Association, 2006). The incidence in the United States is somewhat over 5,600 newly diagnosed cases

per year. Lechtzin, Schmidt and Clawson (2005) report an estimated 1 death in every 800 adult males in the United States is attributable to ALS.

The worldwide incidence of ALS is estimated to be 0.6 to 2.6 cases per 100,000 people annually. The male to female ratio is 1.6:1, and the onset occurs most frequently in the sixth decade of life. However, there have been cases of ALS affecting teenagers and octogenarians (Lechtzin, Schmidt, & Clawson, 2005).

Despite recent advances in research and medical care, the prognosis unfortunately remains poor for patients with ALS. Simmons (2005) reports that the median time from onset of symptoms until death has been found to be 23 to 48 months. Five-year survival rates have been published, ranging from 9% to 40%, and 10-year survival rates are between 8% and 16%. The annual mortality rate attributed to ALS is two deaths per 100,000 in the U.S. (Forshew & Hulihan, 2005)

In 90% to 95% of all ALS cases, the disease presentation appears arbitrary with no clear associated risk factors, such as family history. However, approximately 5%-10% of all cases are inherited. This familial form of ALS requires only one parent to carry the

gene accountable for the disease (National Institute of Neurological Disorders [NIND], 2006).

Turner, Parton, Shaw, Leigh and Al-Chalabi (2003) put forth that those who present with initial bulbar onset, onset later in life or in the definite El Escorial category are not automatically precluded a long survival. However, those ALS patients who present at a younger age or with pure upper motor neuron signs initially have a better prognosis.

Diagnosis of ALS

Sadly, ALS can be a very difficult disease to diagnose. The average delay from symptom onset to a definitive diagnosis is about 14 months. Now and again there may be occasional incidences of a patient surviving 6 months or even less following his or her diagnosis (Leigh, Abrahams, Al-Chalabi, Ampong, Goldstein, Johnson, et al., 2003). Often patients' initial symptoms may lead them to believe that what they are experiencing is indicative of the normal aging process or arthritis and other such disorders. They may spend time researching possible etiology on the Internet. Quite possibly they may even

present to their primary care providers with vague somatic complaints that do not lead to the consideration of ALS as the source. The regrettable fact that there is no specific diagnostic test such as neuroimaging studies or specific laboratory procedures, makes it sometimes difficult to diagnose ALS even after the patient presents to the appropriate healthcare professionals. However, the presence of upper and lower neuron signs in a single limb is strongly suggestive of the disorder. Ultimately, the diagnosis can be made from a combination of clinical and neurophysiological assessments (Winhammer, Rowe, Henderson, & Kiernan, 2005).

Management Strategies for ALS

Breaking the News. The single most important indicator of how the client is going to react to the diagnosis and eventually collaborate with his or her healthcare providers is dependent upon the manner in which the diagnosis is conveyed by the physician (Simmons, 2005). Leigh and colleagues (2003) previously recognized this fact and reported that the experience of being informed that the

diagnosis is ALS shapes the subsequent relationships that the client will have with the healthcare team.

Respiratory Issues. The management of respiratory care is quite possibly the biggest challenge that the healthcare team faces in the care of ALS patients. The majority of ALS patients will die because of progressive respiratory failure. Thorough pulmonary evaluation is necessary because of the inconspicuous nature of presenting symptoms. For example, patients may have extremity involvement prior to overt respiratory issues. They may be confined to wheelchairs and be unable to exert themselves to the point of dypsnea (Lechtzin, Schmidt & Clawson, 2005). An early understanding of patients' preferences will make difficult decisions such as the initiation of invasive ventilation easier to broach, in a timely manner (Miller, Rosenberg, Gerlinas, Mitsumoto, Newman, Sufit, et. al., 1999).

Nutritional Issues. "Dysphagia" is the difficulty in swallowing or the inability to swallow. ALS patients with dysphagia are very likely to experience insufficient fluid intake and suboptimal caloric intake (Simmons, 2005; Lechtzin, Schmidt & Clawson, 2005; Miller, Rosenberg,

Gerlinas, Mitsumoto, Newman, Sufit, et al., 1999). Initial management includes the modification of food and fluid consistency. Patients with bulbar dysfunction generally present with jaw and tongue weakness and fatigue, drooling, choking, and slow intake of food; meal times must be handled deliberately. Malnutrition, dehydration, aspiration, weight loss, and infection become serious issues. Patients may lose weight because they eat more slowly and ultimately eat less than they did before. This occurs perhaps, even more in the presence of others due to the embarrassment caused by excessive saliva, sputtering, coughing, and food falling out of their mouths. They also often depend on others to assist them with eating; this is due to extremity weakness that leads to difficulty cooking and even feeding oneself (Leigh, Abrahams, Al-Chalabi, Ampong, Goldstein, Johnson, et al., 2003).

When the initial management issues are no longer effective then the insertion of a feeding tube, percutaneous endoscopic gastrostomy (PEG), must be considered (Simmons, 2005; Lechtzin, Schmidt & Clawson, 2005; Miller, Rosenberg, Gerlinas, Mitsumoto, Newman, Sufit, et al., 1999). A dietician best carries out the dietary assessments; however, close collaboration with

speech and language therapists is recommended when dealing with dysphagia (Leigh, Abrahams, Al-Chalabi, Ampong, Goldstein, Johnson, et al., 2003).

Psychological Issues. ALS is a disease that is clearly dynamic, yet maintains a fairly predictive course. This predictability allows for preparation of the many losses that will befall the ALS patient and his or her family. The role of a mental health professional is invaluable on the ALS multidisciplinary team due to the scope of real and perceived losses. Lechtzin, Schmidt and Clawson (2005) identify a succession of losses, such as independence, role identity, and future plans that put the patient at risk for periods of grief and mourning as he or she addresses each new phase of the disease. Grief is a complicated, individual process that cannot be well predicted. Yet healthcare professionals should be prepared for the patient and his or her support team to progress through varying periods of denial, anger, bargaining, depression and acceptance at his or her own rate and in no particular order (Kubler-Ross, 1973).

Lou, Reeves, Benice and Sexton (2003) affirm that depression is associated with a poor QOL and must be

treated as a priority. Viable interventions include both psychopharmacological and psychotherapeutic options. Support systems and available resources must be readily identified and then involved in the treatment process.

Emotional lability. ALS patients exhibit emotional lability that could be easily misconstrued as symptoms of a mood disorder. Pathological crying or laughing is observed in as many as 50% of ALS patients. The emotions exhibited are disproportionate or inappropriate to internal feelings or external stimuli. The abnormal affective display is not well understood. Although not definitively indicative of a mood disorder, antidepressants such as selective serotonin reuptake inhibitors (SSRIs) are often used to treat the emotional lability.

Pain Management. It is common for ALS patients to feel as if their pain will be protracted and uncontrolled (Hindelang, 2006). Between 40% and 73% of ALS patients experience pain in the later stages (Miller, Rosenberg, Gelinas, Mitsumoto, Newman, Sufit, et al., 1999). Pain in the early stages of ALS, although less frequent, still

occurs and is attributed chiefly to spasticity and cramping (Simmons, 2005).

Pain should be regularly assessed and the patient's perception of pain should be continually evaluated. Anxiety, stress, frustration, depression, poor sleep, and fatigue can all contribute to varying manifestations of pain.

End-of-Life Issues. The end of life is a very critical phase of treatment for ALS patients. Patients and families need to feel completely supported. Perhaps more than any other time during the course of treatment, religious and cultural beliefs need to be recognized fully.

There are a great number of ethical and legal issues involved when working with ALS patients. Healthcare teams should be fully aware of contemporary literature regarding euthanasia and physician-assisted suicides. Lechtzin, Schmidt and Clawson (2005) suggest discussing advance directives as soon as rapport has been established with the patient and family. Advanced directives are most likely in the form of a living will, a durable power of attorney for health care matters, or a note written and signed by the physician in the medical records, documenting the desires

of the patient or surrogate decision maker (Vasar, Weinacker, Henig & Raffin, 2002).

Caregiver Issues. Communication among the healthcare team helps to avoid any perceived ethical dilemmas that may arise. Depression and anxiety may be just as relevant for the caregivers as for the patients.

A recent trend has developed in which researchers have become increasingly interested in the QOL for caregivers of those with ALS. The strong connection between ALS patients and their caregivers, perhaps more so than in other dyads facing terminal illnesses, has lead the research to naturally progress in this direction.

Bromberg and Forshew (2002) put forth the notion that there is likely a response shift by patients that leads to an eventual difference between caregiver and patient, relative to the way in which QOL is viewed. They posit that during the course of the disease the patients change their terms or internal standards of reference used to judge QOL. In other words, what they previously viewed as important to maintaining QOL has now changed. This can actually be viewed as a positive coping mechanism. However, these

response shifts have made the measurement of QOL in ALS patients more difficult.

An often neglected area of discussion is the level of sexual activity for ALS patients. This is unfortunate because there is a realistic potential for normal sexual functioning. Sexual function is not directly affected; however, such issues as patient and partner passivity and decreased libido create obstacles. Most frequently the reasons reported for change in sexual activity include decreased physical strength and body image issues (Wasner, Bold, Vollmer & Borasio, 2004). Massage, shiatsu, reflexology and the scheduling of private time should be discussed as viable options for the couple. Encouraging the open discussion of such sensitive issues can serve to improve the quality of the couple's relationship and delay what may have already been viewed as a loss. This open discussion should take place before a significant loss of the patient's communication ability has taken place. It is a very sensitive and perhaps awkward topic to discuss and it may be made even more difficult to handle when complicated by an impaired ability to communicate clearly.

Bulbar Dysfunction in ALS

The term bulbar refers to the motor neurons located in the bulb region of the brain stem that control the muscles of chewing, swallowing, speaking and the capability of maintaining an open upper airway. The following pages will discuss the QOL issues that ALS patients must address when faced with bulbar dysfunction.

Excessive Salivation

Excessive salivation that is beyond a patient's ability to compensate for is known as sialorrhea. Sialorrhea can affect upwards of 20% of ALS patients. It can be socially debilitating and is particularly problematic for patients with significant bulbar involvement. The actual problem with sialorrhea is the inability to control and swallow one's saliva. Saliva production is actually not increased in ALS patients; rather, it is decreased (Miller, Rosenberg, Gelinas, Mitsumoto, Newman, Sufit, et al., 1999). It is unclear why saliva production is decreased. Weak muscles around the mouth, tongue, and throat can compromise the handling of

saliva in the mouth and impair the swallowing mechanism. It is quite possible that difficulty in swallowing leads ALS patients to restrict fluid intake, thus causing them to become dehydrated. Therefore they will produce less saliva. However, the little saliva that they do produce may still be difficult for them to manage. It is a difficult problem because once they become dehydrated feeding tubes may be placed to rehydrate and the sialorrhea can be exacerbated.

Sialorrhea can cause significant distress to the patient and caregivers because of the resulting drooling, choking, coughing, gagging, sputtering, and vomiting. The excessive drooling creates concern because ALS patients are at increased risk of aspiration, pneumonia, skin break down, and infections. When speech is impaired and communication devices are utilized, the excessive saliva can actually make the equipment wet and cause damage (Mathur & Vaughn, 2006).

Sialorrhea is categorized either as primary or as secondary. Primary sialorrhea causes drooling due to hypersecretion of the salivary glands. ALS patients' drooling is most likely due to secondary sialorrhea caused by impaired neuromuscular control with dysfunctional voluntary oral motor activity. The patient's inefficient

and infrequent swallowing further compounds the problem of sialorrhea (Mathur & Vaughn, 2006). Dehydration is a viable risk for ALS patients with salivation issues. Excess saliva can be further increased by anxiety, hunger and acid reflux. There are various medications and surgical procedures utilized to treat excess salivation.

Because of nasal congestion and fatigue of the jaw muscles, ALS patients may resort to breathing through their mouths more often than through their noses. This can create complications such as the thickening of their saliva. This, paired with the side effects of certain medications that also cause dryness and thickening, results in postnasal drip, chronic cough and the constant need to clear one's throat. In addition to these potential medical issues this writer believes that an ALS patient's QOL is adversely affected by complicated and awkward social situations created by their impaired saliva management.

Speech Impairment

There are many losses involved with ALS. The loss of one's mobility, independence, future, and what is arguably one of the most devastating losses, the ability to speak.

In the bulbar form of ALS, speech problems are most common and can present as slurring, hoarseness or decreased volume. The initial symptoms are often a slight, intermittent slurring that can occur when the client is tired or has been speaking a lot on a particular day. Over time the ALS patient's speech slowly becomes more and more unintelligible as the functioning of the tongue, lips and pharynx become more impaired (Simmons, 2005).

The classification of ALS is determined by the site of involvement (upper motor neuron versus lower motor neurons) and depends upon the involvement of spinal nerves or bulbar nerves. The bulbar symptoms are the initial manifestations in 19 to 25% of all ALS cases (Walling, 1999). Bulbar involvement in ALS is most often a combination of upper and lower motor neuron dysfunction. This combination results in mixed dysarthria with both spastic and flaccid components affecting muscles of the face, tongue, and throat (Simmons, 2005). Dysarthria is a neurologically based speech disorder that results in weakness or spasticity of the lips, tongue, jaw movement, soft palate and respiratory muscles (Carr-Davis, Blakely-Adams, & Corinbilt, 2005). Dysarthric speech is characterized by problems with articulation, volume and quality of speech, and prosody (speech rate, rhythm and

naturalness; Yorkston, 1999). Unfortunately many ALS patients can develop anarthria (loss of motor ability to speak) within just a few months (Leigh, Abrahams, Al-Chalabi, Ampong, Goldstein, Johnson, et al., 2003).

Stages of Severity of Dysarthria in ALS. Yorkston and colleagues (1993) put forth a five-stage model for the severity of dysarthria in ALS. For each of the five stages there are treatment interventions outlined. The underlying strategy for each stage is to maintain functional communication with natural speech or augmentative communication strategies, regardless of the severity of the speech disorder. The five-stage model is as follows (Yorkston, Strand, Miller, Hillel & Smith, 1993):

Stage 1: Normal speech processes.

Stage 2: Detectable speech disturbances.

Stage 3: Behavioral modifications.

Stage 4: Use of augmentative communication.

Stage 5: Loss of useful speech.

Individuals in the first stage do not demonstrate overt speech changes. Normal rate and volume of the patient's speech is preserved. The level of change may be noticeable only to the client or perhaps to his or her

spouse (Yorkston, Beukelman & Ball, 2002). Interventions at this time should be focused around providing information to the patient and family. This first stage can be used as a preparatory time of education regarding the disease course. The inevitable loss of speech must be fully explored and the patient and family should be made aware of the choices they will have to make in the future. Ideally the decision-making relative to communication devices should occur while the patient still retains the ability to speak (Yorkston, Beukelman & Ball, 2002).

The second stage of dysarthria in ALS is marked by detectable speech disturbances. Although there are overt changes that listeners will recognize, the patient's speech does remain intelligible. However, these changes in rate, articulation and resonance will be exacerbated during periods of fatigue or stress (Yorkston, Beukelman & Ball, 2002). Environmental control is among the interventions to be utilized during this stage of impairment. Background noise may lead to the ALS patient's having to speak louder. If this is even possible, the experience can be very fatiguing. Both background noise and distance may need to be reduced when interacting with ALS patients (Yorkston, Beukelman and Ball, 2002). Turning off the television and

any other mechanical equipment, such as kitchen appliances and non-vital medical equipment may be necessary for a brief period during predetermined conversation time. Effort should be placed on establishing clarity when conversing with an ALS patient. It is vital to confirm the context of the discussion prior to changing a topic or ending a conversation. Additionally, ALS patients will find it very difficult to partake in group discussions and may tend to shun social gatherings altogether. Another relevant issue is the fact that a great number of ALS patients and their partners are elderly and may have experienced some level of hearing loss (Yorkston et al., 2002). Hearing loss is certain to make the deciphering of impaired speech even more difficult.

Stage three is marked by reduction in intelligibility. Frequent communication breakdowns will occur in stage three, particularly while in adverse listening conditions. Messages will often need to be repeated to resolve these communication breakdowns (Yorkston, Beukelman and Ball, 2002). Interventions during this stage focus on behavior modifications. Optimizing speech performance and using compensation techniques are the main themes in stage three. However, environmental control is once again stressed

during this stage of impairment. The optimization of speech performance starts by encouraging the ALS patient to slow his or her rate of speech (Yorkston, Beukelman and Ball, 2002; Yorkston & Beukelman, 2000). ALS patients are encouraged to exaggerate their speech. In particular, words that are important to the context of what they are trying to say must be overemphasized. Incorporated into the stage three approach is training in energy conservation. Attention is given to avoiding fatigue because it can have a substantial impact on the quality of an ALS patient's speech. Whenever possible the ALS patient should refrain from attempting communication in less than ideal situations. The goal is to avoid a reduction in the efficiency of the patient's speech muscles. This may involve recognition on the part of the listener to suggest such actions as changing locations; an example of this may be finding a quieter area in a restaurant.

Stage four is marked by the patient's reliance on augmentative systems. Devices are used either as the patients' primary or secondary means of communication. When natural speech is not understood the patient uses his or her device as a supplement. Augmentative systems may be used to provide information about a topic, present the

first letter of a word, or to "verbalize" a word or phrase that is unusual or difficult to enunciate (Yorkston & Beukelman, 2000).

Although the patient can still speak using residual speech, the transition to augmentative communication approaches is complete during this stage. This transition period is extremely crucial in the management of patients with ALS (Yorkston, Beukelman and Ball, 2002; Yorkston & Beukelman, 2000). It is fundamental that the patient and family are supported and educated appropriately during this stage of speech impairment. There may be confusion, frustration, anger, and even guilt ("Why did I not speak my mind when I had the chance?").

The fifth stage is characterized by the loss of functional speech. The ALS patient is now at a point during which the ability to use natural speech as a functional means of communication has been lost. At this point the patient has become totally dependent upon augmentative communication strategies (Yorkston, Beukelman and Ball, 2002; Yorkston & Beukelman, 2000).

Individuals will have to use a variety of strategies to meet their communication needs. Included among these strategies is the development of a non-fatiguing and

reliable means of indicating "yes" or "no". This is an absolutely mandatory component of an effective management plan for dysarthria. Eye gaze is often utilized as a method of selection when the ALS patient has lost the ability to move his or her head or his or her hands in a dependable manner. This may involve the family, partner or caregivers being trained to recognize and decipher the intricate and subtle aspects of eye gaze communication techniques. Additional strategies to be utilized during the fifth stage include low-tech picture and alphabet boards. Still other strategies involve the use of progressive, computer-based systems (Yorkston, Beukelman and Ball, 2002; Yorkston & Beukelman, 2000).

Swallowing Dysfunction

Weakness and poor control of the mouth and throat muscles can result in difficulty swallowing, or dysphagia. ALS patients with dysphagia are at risk for insufficient nutrition and dehydration that can exacerbate muscle atrophy, weakness, loss of coordination and fatigue. Swallowing is a complex process that occurs in three different stages: oral phase, pharyngeal phase, and

esophageal phase. During the first phase (oral phase) the tongue moves the food around the mouth for chewing and then towards the throat. The second stage (pharyngeal phase) begins when the swallowing reflex is triggered by food moving to the back of the mouth. The food then passes through the pharynx that connects the mouth and the esophagus. The larynx closes tightly and breathing is ceased to prevent food or liquid from entering the lungs. If food does enter into the airway it is known as aspiration. This is a dangerous situation for the ALS patient. Food or liquid entering the lungs can lead to the development of a bacterial infection and subsequently what is known as aspiration pneumonia, which can further complicate the swallowing process. The third stage (esophageal phase) is the point at which food enters the esophagus and is carried towards the stomach. Dysphagia can occur during any of these different stages.

There are varied muscles that are part of the swallowing process; therefore, the presenting problems can vary depending on the particular muscles that have been affected. Hillel and Miller (1989) have identified a pattern of affected musculature during the course of bulbar ALS. Their research identified the fact that the pattern of

dysfunction begins with the tongue and lips. This can lead to the possibility of oral stage difficulties, such as sucking, chewing and moving the food or liquid around the mouth towards the throat (Watts & Vanryckeghem, 2001).

Dysphagia in ALS usually begins with a noticeable variation in the ability to manage certain foods. The initial changes are usually reported as difficulty managing foods that require a lot of chewing (meat), difficulty with foods that fragment easily (crackers), and problems with thin liquids (especially room temperature water; Hillel, Dray, Miller, Yorkston, Konikow, Strande, et al., 1999).

The five stage model is as follows (Hillel & Miller, 1989):

Stage 1: Normal eating habits

Stage 2: Early eating problems

Stage 3: Dietary consistency changes

Stage 4: Needs tube feedings

Stage 5: Nothing per oral

Quality of Life

Definitions

Various definitions exist to explain what is meant by QOL. Additionally, there has been no clear consensus about which instruments provide the best assessment of QOL, despite the fact that it is widely recognized as an important area of focus.

In 1997 the World Health Organization defined QOL as "individuals' perception of their position in life in the context of the culture and value systems in which they live and in relation to their goals, expectations, standards and concerns. It is a broad ranging concept affected in a complex way by the person's physical health, psychological state, level of independence, social relationships, personal beliefs and their relation to salient features of their environment" (WHO, 1997).

This is an idea that is different from those that have been put forth previously in the literature, mainly in those studies outside of ALS research in which greater emphasis has been placed on physical functioning as an indicator of QOL. An important caveat here is that those in good health generally underestimate the QOL of those with chronic diseases. Also noteworthy is the fact that two individuals with similar pathology may have completely different QOLs. Simmons and colleagues have posited that

QOL in ALS is determined by a broad array of factors and is maintained as a patient's physical function declines (Walsh, Bremer, Felgoise, & Simmons, 2003; Robbins, Simmons, Bremer, Walsh & Fischer, 2001). However these studies have looked at physical functioning only as an aggregate. The current study will look specifically at the effects of impairment in communication on ALS patients' QOL.

Quality of Life Instruments

World Health Organization (WHO): WHOQOL-100/WHOQOL-BREF. As mentioned previously, there has been an expanded focus in the measurement of health beyond traditional health indicators such as mortality and morbidity. There is now increased attention given to such issues as disease impact, impairment of daily activities and behaviors (World Health Organization Group, 1998).

This new focus has led to the World Health Organization Quality of Life (WHOQOL) project, initiated in 1991. WHO's commitment towards promoting cross cultural holistic health care led to interest in developing a QOL assessment instrument that would function effectively as an

international measure for QOL. The instrument which was eventually developed, the WHOQOL-100, assesses individuals' perceptions from a frame of reference that incorporates their cultures and value systems and their personal goals, standards and concerns. The initial conceptual framework for the WHOQOL-100 proposed that the 24 facets relating to QOL should be grouped into 6 domains. However, subsequent analysis of the data showed a four-domain solution to be more appropriate. The resulting instrument based on this four-domain structure is the WHOQOL-BREF, composed of 26 items, which measure the following broad domains: Physical Health, Psychological Health, Social Relationships, and Environment. This new broad focus marks the emergence of measures assessing the impact of disease; however, they do not encompass the idea of QOL.

McGill Quality of Life Questionnaire. The McGill Quality of Life (MQOL) Questionnaire is relevant to all phases of the disease course for people with a myriad of life-threatening illnesses. The MQOL was initially designed to measure the subjective well-being of cancer patients and HIV infected patients by reflecting the

patient's experienced QOL (Robbins et al., 2001; Cohen, Mount, Strobel, & Bui, 1995). The authors of the MQOL attempted to address the identified shortcomings of other QOL instruments; therefore, the MQOL differs from previous QOL questionnaires in three ways. The first difference is that the physical domain was considered an important but not predominant aspect of the measure. Second, the positive contributions to QOL were measured. Last, the existential domain that reflects a patient's perception of purpose, meaning in life and the capacity for personal growth was measured. The MQOL consists of 16 items organized into five subscales: Physical Symptoms, Physical Well-Being, Psychological, Existential, and Support. The 16 Items are scored from 0 (worst) to 10 (best; Robbins et al., 2001; Cohen, Mount, Strobel, & Bui, 1995).

Sickness Impact Profile. The Sickness Impact Profile (SIP) is a general quality of life scale. It consists of 136 items that measure 12 distinct domains of QOL: Ambulation, Movement and Mobility, Body Care, Social Interaction, Communication, Alertness, Emotional Behavior, Sleep, Eating, Work, Household Management and Recreation.

The SIP can be administered by an interviewer or can be taken by the patients themselves. Although it is easy to administer and score, it is relatively time-consuming, taking approximately 30 minutes to complete. Patients identify statements that most accurately communicate their experiences. All of the 136 items are weighted, depending on the severity of dysfunction. A higher score indicates greater dysfunction. The SIP, however, is a generic instrument that is applicable to a variety of diseases and not specific to the distinct issues that ALS presents.

Sickness Impact Profile/ALS-19. A subset of the SIP is the SIP/ALS-19. It is an example of a modified, established instrument utilized to address the unique aspects of ALS (Bromberg & Forshew, 2002). The SIP/ALS-19 is a questionnaire that consists of 19 items chosen from the full 136-item SIP. These 19 items were chosen because they were believed to have the greatest impact on QOL. The scores on this instrument correspond closely to measures of strength and function (Robbins, Simmons, Bremer, Walsh & Fischer, 2001). The SIP/ALS-19 was developed because of its ability to predict changes in function as measured by the Tufts Quantitative Neuromuscular Examination (TQNE). The

TQNE is a standardized tool for measuring muscle strength and pulmonary function in patients with ALS (McGuire, Garrison, Armon, Barohn, Bryan, Miller, et al., (1996).

ALS Assessment Questionnaire. The ALSAQ-40 is a disease-specific, health-related QOL instrument for use in patients with ALS or other motor neuron diseases. Patients are asked to give one of five possible answers to a series of forty questions. The instrument covers five dimensions of health status that are affected by ALS: Physical Mobility; Activities of Daily Living and Independence; Eating and Drinking; Communication; and Emotional Functioning (Jenkinson, Peto, Jones, & Fitzpatrick, 2003).

Criticism of the SIP, SIP/ALS-19 and the ALSAQ-40 includes the fact that they are heavily weighted toward physical functioning and therefore fail to capture other factors related to QOL (Simmons et. al., 2006). The measures correspond too closely to measurements of strength and function; consequently, when the inevitable physical decline begins, so do the patient's QOL scores as measured by these instruments.

Schedule for the Evaluation of Individual Quality of Life. The Schedule for the Evaluation of Individual Quality of Life (SEIQoL) and the shorter SEIQoL-Direct Weighting (SEIQoL-DW) assess individualized QOL using a semi-structured interview technique. The SEIQoL is a measure designed to elicit the value systems of individual respondents and to quantify QOL. This instrument is an interview-administered measure with three structured stages. Initially, respondents identify five domains of life that they recognize as important in relation to their present QOL. They then indicate the relative weight of each of these "elicited cues" in regard to their QOL. The SEIQoL has demonstrated the importance of nonphysical factors in assessing QOL. However, the instrument is designed to attain a greater level of individual subjectivity than is achieved with generic measures of QOL. Therefore it may not be useful when assessing large samples (Simmons et al., 2006).

Development Towards an ALS Specific Tool

ALS-specific Quality of Life (ALSSQOL) Instrument. The Amyotrophic Lateral Sclerosis-Specific Quality of Life

instrument (ALSSQOL) was developed to reflect overall QOL; it is a measure that has proved valid and reliable across large samples of ALS patients. The ALSSQOL, which asks wide-ranging questions, is a 59-item questionnaire that uses a 0 to 10 point scale for each item. The least desirable situations are scored as a 0, and the most desirable are scored as a 10, resulting in scores ranging from 0-590 (Simmons et. al., 2006).

The ALSSQOL was based on many of the principles of the MQOL; however, the ALSSQOL inquires more broadly about spirituality and religiousness. In addition to asking questions regarding physical symptoms, religiousness and spirituality, the ALSSQOL inquires about intimacy, loneliness, relationships, environment, social interaction, values, coping and interests, and desires/goals (Simmons et al., 2006).

The ALSSQOL was developed at Penn State Hershey Medical Center. The evaluation of the ALSSQOL's psychometric properties was a prospective study involving seven university-based ALS centers. The ALSSQOL appears to be a valid instrument demonstrating concurrent, convergent, and discriminant validity for the overall instrument. Convergent validity was demonstrated for its subscales. The

ALSSQOL takes on average 15 minutes to complete with a range of approximately 10 to 25 minutes (Simmons et al., 2006).

ALSSQOL-Revised. Validation studies are currently underway for a shortened version of the ALSSQOL. Presently a multi-center study to validate a shorter, 50-item version of the ALSSQOL is being conducted. Thus far the ALSSQOL-R has shown promise as an effective instrument for measuring QOL in ALS patients. This time there are nine university-affiliated ALS clinics involved in the study. In addition to validating the shorter 50-item ALSSQOL, the current studies aim to determine the relationship, if any, between hope, optimism, social problem-solving skills, relationship satisfaction, religiosity/spirituality and the QOL in caregivers of patients with ALS. The researchers intend to explore further the possibility of using the ALSSQOL-R via methods other than personal interview only; these include home completion of written or electronic questionnaires and possibly even telephone administration (Simmons et al., 2006).

Communication

Definitions

"Communication" is defined as the exchange of information between individuals, for example, by means of speaking, writing, or using a common system of signs or behavior (Merriam-Webster Online Dictionary, n.d.). Communication is one of the most difficult, yet necessary, skills that we as human beings learn. Communication occurs continuously throughout the course of one's day. Expressing one's needs, feelings, ideas, preferences, and opinions allows people to control and modify their environments. Any change in an individual's ability to speak can greatly impact these everyday expressions and impair that control. ALS patients have to modify how, when, and where they speak in order to be well understood; therefore, their sense of control can vacillate from one situation to the next.

Assessing QOL in people with communication impairment is often difficult in health related research. Hilari and Byng (2001) claim that the materials used to assess QOL can often be linguistically complex. They also report that the way in which the materials are administered does not

usually facilitate the sharing of personal experiences by those with communication disabilities.

Nonverbal Communication

The exchange of thoughts and feelings does not always occur through the use of language. Nonverbal communication such as gestures, kinesics (body motions/posture), facial expressions, spatial relations, touch and display (presentation of self) are effective means of communication. It is necessary to stress to patients and to their families the importance of utilizing these nonverbal techniques to substitute for the loss of speech or to enhance impaired speech. Nonverbal communication techniques can, however, be very limiting. Nonverbal communication can be imprecise and interpretation is difficult especially if the two (or more) parties involved are not very familiar with one another. Additional impediments to the use of nonverbal communication include the difficulty in presenting complex information and the inability to communicate sarcasm. The need to be in fairly close proximity to those with whom you wish to communicate is essential. Nonverbal communication is effective only in

person and therefore text and telephone communication is prevented.

Verbal communication is impaired in more than 80% of patients with ALS over the course of the disease (Leigh, Abrahams, Al-Chalabi, Ampong, Goldstein, Johnson, et al., 2003). ALS affects the strength and coordination of a patient's breath, vocal cords, tongue, lips and jaw, thereby creating challenges to his or her communicative ability (Carr-Davis, Blakely-Adams, & Corinbilt, 2005).

Dysarthria is usually the earliest symptom presenting in patients with bulbar onset. As mentioned previously, it is unfortunately common for ALS patients to become anarthric within just a few months (Leigh, Abrahams, Al-Chalabi, Ampong, Goldstein, Johnson, et al., 2003).

Communication can be unfavorably affected when a patient experiences the psuedobulbar symptoms of ALS. The exaggerated emotional responses they experience can be very disruptive and can hinder their ability to share or exchange information effectively with others.

Pragmatics

Herbert Paul Grice, an American philosopher-linguist, was best known for his contributions to the study of meaning within language. Grice's work is considered one of the foundations of the modern study of pragmatics. Pragmatics, a subfield of linguistics developed in the 1970s, is the study of how people use language. Every communicative act or speech act has two intents or meanings. One is the informative intent or the sentence meaning, and the other is the communicative intent or speaker meaning. Pragmatics is primarily concerned with bridging the explanatory gap between the sentence meaning and speaker's meaning. "Meaning is not something which is inherent in the words alone, nor is it produced by the speaker alone, nor by the hearer alone. Making meaning is a dynamic process, involving the negotiation of meaning between speaker and hearer, the context of the utterance (physical, social and linguistic) and the meaning potential of an utterance" (Thomas, 1995, p. 22).

The ability to produce and understand a communicative act necessitates knowledge about social distance, social status between the involved speakers, and cultural knowledge. People constantly change their use of language in response to different contexts. When the ability to

communicate is impaired, as it is in patients with ALS, the opportunity to adapt to different settings and situations is severely challenged.

Thomas (1995) states that speakers frequently mean more than simply what their words actually say. This may be a difficult aspect of communication for ALS patients because the message they may be trying to convey is not always facilitated by their non-verbal techniques. The physical impediments that ALS patients experience can make it difficult to control such aspects of communication as inflection, body positioning, gesturing, and touching. Because of muscle weakness, the facial expressions of ALS patients can be difficult to interpret correctly. So even when a patient's words are successfully understood there may still be information that he or she wishes to express that has been missed or misconstrued. Expressions rely on fairly minute differences in the proportion and positioning of facial features. Therefore, family, friends and healthcare providers of those with ALS must be extremely sensitive when attempting to interpret facial expressions.

Maxims of Conversation

Grice (1967) proposed that conversations develop on the basis of a cooperative principle. This principle assumes that both speaker and hearer converse with a willingness both to deliver and to interpret a message. It is this cooperation that leads to successful communication (Thomas, 1995). For communication to be truly effective, it must be guided by specific rules. Therefore, in addition to the cooperation principle, Grice put forth a set of four conversational maxims: the maxim of quantity, the maxim of quality, the maxim of relevance and the maxim of manner.

Maxim of Quantity. The maxim of quantity requires the speaker to give the right amount of information when he or she speaks. This speaker is expected to be not too brief or too verbose. This rule dictates that a contribution to a conversation be as informative as required, but that it should not be more informative than is appropriate for the given situation.

Maxim of Quality. The maxim of quality is a matter of giving truthful information. One should not say what he or she believes to be false or what he or she cannot provide adequate evidence to support. Truthfulness is regarded as

the most important maxim on which all the others are dependent. However, the veracity of an ALS patient's statements should not be questioned simply because he or she has impaired speech. It is most likely to be an issue of inaccurate interpretation on the part of the listeners.

Maxim of Relevance. The maxim of relevance requires the speaker to be relevant to the context and situation in which the conversation is currently taking place. It is important to recognize that the parties involved in the conversation determine relevance. The pathological laughing or crying that results from pseudobulbar affect may lead to overt violations of the maxim of relevance when ALS patients are displaying emotions that are clearly out of context.

Maxim of Manner. The maxim of manner calls for speaking with brevity and clarity, yet maintaining logical order when conversing. It dictates that one should avoid obscure expressions and ambiguity. The maxim of manner is perhaps the most difficult maxim for adherence by ALS patients because of their potentially ambiguous or unclear

utterances.

A maxim that is not adhered to is known as a non-observance. If the speaker breaks a maxim the hearer then attempts to discern the speaker's implied meaning. The hearer does so based on the assumption that both speaker and hearer are observing the cooperative principle and are interested in communicating effectively. Often a speaker will intentionally fail to observe a maxim. This may occur to create a humorous situation or to avoid discomfort. Unfortunately, the breaking of a maxim for an ALS patient may not be volitional. Rather, the violation may be unavoidable, given speech and physical limitations. Even when an ALS patient does intend to break the rules and imply a meaning beyond the literal sense, the results can be unpredictable. For example, if asked, "How are you today?" a patient may reply, "I am great" even though he or she may clearly not be feeling great. However, this could be an example of the speaker (patient) so blatantly breaking the rule of quality (truthfulness) that he or she intended the hearer to pick up the meaning of the utterance not directly stated in words. For those whose quantity of speech may be limited and whose non-verbal techniques are impaired, it could be difficult to convey such sarcasm if

they should have to explain themselves further.

Turn-Taking

Another communication issue that is relevant in regard to ALS patients is the turn-taking behaviors displayed during conversations. Sacks, Schegloff and Jefferson (1974) were the first to recognize that conversations were highly organized social activities. They put forth two types of turn-allocation techniques: a) the current-speaker chooses the next speaker, or b) the next-speaker is allocated by self-selection. The time interval between the current-speaker finishing and the next-speaker starting is known as reaction time latency. Initiative time latency is the duration between a current-speaker's completion and the start of a follow-up utterance by the same speaker (Sacks, Schegloff and Jefferson, 1974). As would be expected, there are various cultural norms dictating what is appropriate during these aforementioned intervals. Only by actively paying attention, looking and listening, can a prospective next-speaker appropriately incorporate himself or herself into the conversation. These are particularly important matters to acknowledge when attempting to converse with ALS

patients who have speech problems. Muscle weakness, aches, cramps, spasms, spasticity, excessive salivation and generalized fatigue can lead to conversational lulls that create communication discontinuity. To interject or change the subject of the conversation before the patient has had an opportunity to express him or herself completely can be construed as dismissive, ignorant or indifferent. Because there may be possible extended reaction time latency, ample opportunities must be provided for the patient to respond to previous statements or to introduce new topics.

Augmentative and Alternative Communication (AAC) Methods

Although most of the aforementioned losses (mobility, independence, future) are inevitable, it is possible that the healthcare team can aid ALS patients in maintaining their ability to communicate. Augmentative and Alternative Communication (AAC) refers to communication approaches that augment or supplement existing speech or act as an alternative to natural speech. Through AAC devices and compensatory strategies, ALS patients can learn to communicate very effectively. Among ALS patients with impaired verbal communication, 80% are so disabled that

they need AAC strategies to meet their daily communication needs (Yorkston, Beukelman & Ball, 2002).

AAC can refer to any method that makes communication less challenging and more manageable. People use multiple communication strategies throughout the course of a day to emphasize and clarify their intentions. ALS patients may not be fortunate enough to have various strategies to choose from during daily discourse. AAC may be used along with gestures and facial expressions so that ALS patients may emphasize and clarify their messages. There are three different designations for AAC strategies:

1. No-tech strategies
2. Low-tech strategies
3. High-tech strategies

No-tech Strategies. These strategies are those that do not require the use of technology. Strategies include sign language, gestures, and eye gaze. Talking slowly and exaggerating one's movements when possible are effective no-tech interventions.

Low-tech Strategies. Low-tech refers to communication strategies that employ items not requiring the use of electrical power. Examples of low-tech strategies include manual communication boards and eye gaze boards. These may

often be the preferred methods because higher levels of energy, technical skills and knowledge are not needed as they would be with the more sophisticated equipment. The use of simple communication boards with alphabet letters, words and phrases, or symbols and pictures representing specific messages can be very effective low-tech strategies.

High-tech Strategies. High-tech refers to communication strategies such as computer-assisted devices. These devices can include a wide range of computerized systems that can amplify the patients' voices or can create their own synthesized voices. Personal computers can be modified and used as communication devices simply by installing communication software.

Research Hypothesis

There are two hypotheses for the current study. They are as follows:

1. QOL will differ among ALS patients with varying levels of speech, swallowing, and salivation functioning,

2. Patients with less impairment in these aspects of physical functioning will report better QOL.

Significance of the Research

The current study examined the self-perceived QOL in ALS patients. Precise focus was on communication impairment and the effect that it has or does not have on QOL. This research is important because it is expected the findings will show that QOL is adversely affected by impaired communication abilities. Having this knowledge will allow mental health providers to tailor time-sensitive interventions more appropriately, perhaps enhancing ALS patients QOL.

Methodology

Participants

A total of 342 ALS patients from seven university-based ALS centers chose to participate in the ALSSQOL study. There were 226 men (66%) and 116 women (34%) involved. The patients' ages ranged from 27 to 87 years old (median = 58.0, mean = 57.6, standard deviation = 12.8). At the inception of the study, the symptom duration varied from 1 month to 237 months (median = 24.5, mean = 37.4, SD = 37.4). There was symptom duration of <60 months in 84% of patients and <120 months in 95% of patients (Simmons et al., 2006).

Criterion for Inclusion in the Study

From these centers, all patients who were willing to participate were included if they met the following criteria: 1) age 18 years or older; 2) clinically definite ALS, clinically probable ALS, or clinically probable ALS, laboratory-supported; 3) fluency in English at the sixth grade level or higher (Simmons et al., 2006).

Criterion for Exclusion in the Study

Patients were excluded if an evaluating physician or psychologist determined that they suffered from a dementia or other cognitive impairment that was sufficient to preclude the granting of informed consent and participation in the study (Simmons et al., 2006).

Overview

IRB approval was obtained at each of the independent institutions. Research assistants at the multiple sites were responsible for data collection duties that included introducing the study to patients and caregivers, obtaining/verifying informed consent and administering the questionnaires. Ideally it was a trusted member of the healthcare team, such as the physician or clinic nurse, who would initially approach the patient and introduce the basics of the study. The patients may even have received correspondence from the ALS clinic prior to his or her scheduled appointment. The various ways in which patients responded to the questionnaires included answering orally,

through pointing to the varied responses or through blinking. As the research assistants read through the options, the patients would have blinked upon hearing their desired responses or blinked the number of times necessary to indicate their choices. By administering all of the patient questionnaires in interview format, participation was increased and the burden on the patients was kept to a minimum.

Description of Measures and Forms

ALS Functional Rating Scale (ALSFRS)/ALSFRS-Revised. The ALSFRS is a validated rating instrument for monitoring the progression of disability in patients with ALS. It is a 5-point scale consisting of 10 items, each of which is scored from 0 (unable/dependent) to 4 (none/normal). Individual item scores are then summed to produce a reported score between 0 (worst) and 40 (best). The rating scale evaluates bulbar function, motor function, and respiratory function. The ALSFRS shows close agreement with objective measures of muscle strength and pulmonary

function. The scale shows test-retest reliability and has proved to be consistent (Cedarbaum & Stambler, 1997).

However, one identified weakness of the original ALSFRS was the disproportionate amount of weight it allotted to limb and bulbar dysfunction over respiratory dysfunction. The revised version the ALSFRS-R incorporates additional assessments of dyspnea, orthopnea and the need for ventilatory support. The ALSFRS-R retains the properties of the original scale and shows strong internal consistency and construct validity. Although both instruments are on a 5-point scale, the revised version consists of 12 items, each of which is scored from 0 (unable/dependent) to 4 (none/normal). Summed scores total between 0 (worst) and 48 (best). The rating scale evaluates bulbar function, motor function, and respiratory function. The ALSFRS and the ALSFRS-R are validated clinical rating scales. They have been shown to track progression of disability accurately in ALS and have even been shown to be a predictor of survival in ALS. Both instruments take just a few minutes to administer; this is accomplished easily at each clinic visit by a neurologist or a nurse (Cedarbaum, Stambler, Malta, Fuller, Hilt, & Thurmond, 1999).

For all of the variables on the ALSFRS, a score of a 4 indicates normal functioning. A score of 3 for speech indicates detectable speech disturbances. A score of 2 identifies intelligible speech that requires repeating. Speech that is combined with non-vocal communication is scored a 1. Loss of all useful speech is scored a 0.

A score of 3 for salivation indicates slight, but definite excess of saliva in mouth. These patients may have problems with nighttime drooling. A score of 2 is given when there is moderately excessive saliva that may lead to minimal drooling. A score of 1 is assigned when there is marked excess of saliva with some drooling. Marked drooling that requires the constant use of a tissue or handkerchief would be assigned a score of 0.

When assessing the ability to swallow, a score of 3 would be given for early eating problems with occasional choking episodes. Dietary consistency changes would necessitate a score of 2. A score of 1 would be assigned when a supplemental feeding tube needs to be put in place. A score of 0 is given when it is determined that the patient can no longer eat anything by mouth and he or she begins to receive parenteral (via intravenous or

intramuscular route) or enteral (using the gastrointestinal tract) feedings.

All the participants in the ALSSQOL validation studies were administered the ALSFRS. Three of the ten items on the ALSFRS were examined specifically for the current study: Swallowing, Speech and Salivation.

Manual Muscle Test. Manual muscle testing (MMT) is an important part of the physical examination. It is a technique that is beneficial for establishing a diagnosis, prognosis, and treatment of neuromuscular and musculoskeletal disorders. It is the most frequently used method for documenting impairments in a patient's muscle strength (Cuthbert & Goodheart, 2007).

MMT is a scored neurological examination. The full test, which examines 34 muscles, scores these muscles from 0 to 5. For the purposes of our study, four muscle groups were assessed: Arm (shoulder) abductors, wrist extensors, Hip flexors and ankle dorsiflexors. All MMT assessments are conducted bilaterally. This is important because ALS can affect muscle groups incongruently.

The scale is composed both of subjective and of objective factors. The subjective criteria include both the

standard amount of resistance as determined by the examiner and the amount of resistance the client can tolerate. The objective criteria for establishing an MMT score include the patient's abilities in completing the available range of motion, holding the designated position and moving against gravity.

Cuthbert and Goodheart (2007) assert that MMT is both a science and an art and must be performed according to precise testing protocol. Proper positioning, consistent timing and pressure, nonpainful contact, and the avoidance of preconceived impressions regarding outcome must be employed throughout the MMT.

The grading system for the MMT comprises both a numerical score and a qualitative score. The numerical scores range from 0 to 5. The qualitative scores, with their corresponding numerical scores, are as follows: Zero (no activity/0), Trace activity (1), Poor (2), Fair (3), Good (4), and Normal (5). The following more specific scale is put forth by the Medical Research Council (MRC): No contraction (0), Flicker or trace contraction (1), Active movement with gravity eliminated (2), Active movement against gravity (3), Active movement against gravity and

resistance (4), and Normal power (5; Great Lakes ALS Study Group, 2003).

Modifications to the measurements are possible for grades 3, 4 and 5. Plus and minus indicators modify the whole number scores by a third of a grade. Grade 5 can be modified only by a minus sign (5- = 4.67); however, grades 3 and 4 can be modified either by a plus or by a minus. The purpose of the plus and minus modifiers is to represent a muscle that is slightly stronger or weaker than the whole number would indicate.

Statistical Analysis

The data was entered into a computer-based statistical package, Statistical Package for the Social Sciences (SPSS, 15.0, 2006), to conduct statistical analyses. The various statistical analyses of the data included 1) one-way analysis of variance (ANOVA), 2) descriptive statistics, 3) frequency statistics, 4) univariate analysis of variance for between-subject factors for all three independent variables (speech, swallowing, and salivation), and 5) the Tukey HSD test, which was used in the post-hoc tests of the three independent variables.

A one-way ANOVA is used to test for differences among three or more independent groups. When statistically significant, the ANOVA will indicate that there is a difference somewhere; however, the specific pairs of means that are significantly different are not known. To determine this information a post hoc test is used. To analyze our results further and to find out where the differences between means lie, the Tukey HSD test was utilized. The Tukey HSD test is used for testing the significance of unplanned pairwise comparisons. When multiple significance tests are completed, the chance of finding a "significant" difference just by chance increases. Tukey's HSD test is a method of ensuring that the chance of finding a significant difference in any comparison is maintained at the alpha level of the test.

Descriptive statistics are used to describe the basic features of the data in a study. Descriptive statistics (such as means, medians and modes) were utilized to describe the personal characteristics of the sample.

Inferential statistics are used to determine conclusions that reach beyond the immediate data. These types of statistics are used to draw inferences from a sample about a population. The statistical analyses of the

inferential data collected during the validation of the ALSSQOL included analysis of variance (ANOVA) and were used during this current study.

Results

Descriptive Statistics

Age. The ages for the study sample ranged from 27 to 87 years old. The mean of the sample was 57.57 years old. The standard deviation was 12.847 years.

Duration of Illness. The minimum duration of ALS symptoms at the time of enrollment in the study was 1 month. The longest duration of ALS symptoms in the sample was 237 months (19.75 years). The mean duration of symptoms was 37.37 months (3.11 years) for our study sample with a standard deviation of 37.40 months (3.11 years). There were bi-modal duration results for the sample. Symptom durations of 13 months and 16 months were reported by 13 patients.

Gender. Subjects were excluded from the final analysis if there was any data that was missing. Gender data was missing on 14 of the subjects so that information from 342 valid subjects was used. The gender distribution for the total sample (n = 342) consisted of 226 males (66.1%) and 116 females (33.9%).

Marital Status. Marital status distribution consisted of 274 individuals who were married (80.1%), 31 who were divorced (9.1%), 15 who were never married (4.4%), 3 who were separated (0.8%), and 19 who were widowed (5.6%).

Race. The total sample population (n= 342) showed a race distribution of 317 subjects who were Caucasian (92.7%), 7 who were Hispanic (2.0%), 10 subjects who were African American (2.9%), 6 who were Asian/Pacific Islander (1.8%), 1 who indicated "Other" (0.3%) and 1 who indicated "Prefer not to answer" (0.3%).

Manual Muscle Testing Scores. The total number of patients with valid MMT scores was 340. A total of eight muscle tests were conducted on each of these clients. They included bilateral assessments of the arm (shoulder) abductors, wrist extensors, hip flexors and ankle dorsiflexors. The minimum MMT score was 0 (no contraction), which indicates that no contractile activity can be felt in the gravity eliminated position. The maximum MMT score was 5 (normal power), which indicates that the patient can hold

the position against maximum resistance and through complete range of motion.

Amyotrophic Lateral Sclerosis Functional Rating Scale (ALSFRS). Valid results were obtained for 342 of the subjects in the sample population. The minimum score for the ALSFRS was 1, and the maximum score recorded was 40. The mean score for the ALSFRS was 26.96. The standard deviation was 7.157.

Table 1

Dependent Variable (Speech): ALSSQOL total 46 items

	(I) Speech	(J) Speech	Sig.
Tukey HSD	0	1	.981
		2	.556
		3	.682
		4	.007*
	1	0	.981
		2	.965
		3	.995
		4	.249
	2	0	.556
		1	.965
		3	.988
		4	.227
	3	0	.682
		1	.995
		2	.988
		4	.004*
	4	0	.007*
		1	.249
		2	.227
		3	.004*

* The mean difference is significant at the .05 level.

Table 2

Dependent Variable (Salivation): ALSSQOL total 46 items

	(I) Salivation	(J) Salivation	Sig.
Tukey HSD	0	1	.817
		2	.994
		3	1.000
		4	.822
	1	0	.817
		2	.283
		3	.556
		4	.016*
	2	0	.994
		1	.283
		3	.918
		4	.826
	3	0	1.000
		1	.556
		2	.918
		4	.061**
	4	0	.822
		1	.016*
		2	.826
		3	.061**

* The mean difference is significant at the .05 level. ** Approaching statistical significance.

Table 3

Dependent Variable (Swallowing): ALSSQOL total 46 items

	(I) Swallowing	(J) Swallowing	Sig.
Tukey HSD	0	1	1.000
		2	.929
		3	.999
		4	.517
	1	0	1.000
		2	.880
		3	.993
		4	.210
	2	0	.929
		1	.880
		3	.254
		4	.000*
	3	0	.999
		1	.993
		2	.254
		4	.008*
	4	0	.571
		1	.210
		2	.000*
		3	.008*

* The mean difference is significant at the .05 level.

Table 4

ALSFRS Items by ALSSQOL Total

		N	Mean	SD	Minimum	Maximum
Speech	0	25	301.92	60.08	176.00	403.00
	1	15	312.27	74.79	162.00	440.00
	2	45	323.47	49.94	218.00	404.00
	3	117	318.49	58.28	182.00	429.00
	4	136	343.99	55.70	209.00	454.00
Total		338				
Salivation	0	11	317.91	50.48	207.00	403.00
	1	20	294.55	59.22	176.00	394.00
	2	37	326.03	43.71	218.00	403.00
	3	78	316.37	63.15	162.00	429.00
	4	192	337.01	57.74	203.00	454.00
Total		338				
Swallowing	0	10	314.20	41.70	239.00	380.00
	1	18	313.22	56.15	207.00	394.00
	2	41	298.15	54.40	176.00	398.00
	3	104	319.33	57.23	162.00	440.00
	4	165	343.15	57.46	205.00	454.00
Total		338				

Table 5

ALSFRS Descriptives

		N	Mean	SD	Minimum	Maximum
Speech	0	25	18.04	8.97	1	32
	1	15	22.60	7.38	7	34
	2	45	24.49	7.35	8	37
	3	117	26.82	6.44	11	39
	4	136	30.13	5.12	14	40
Total		338				
Salivation	0	11	22.09	6.73	10	32
	1	20	18.50	9.28	1	33
	2	37	25.22	7.36	2	36
	3	78	25.81	7.45	8	37
	4	192	29.00	5.75	11	40
Total		338				
Swallowing	0	10	13.30	10.02	1	27
	1	18	20.28	7.45	7	31
	2	41	22.93	6.33	6	35
	3	104	26.53	6.76	11	38
	4	165	29.88	5.18	12	40
Total		338				

Table 6

Correlations

		ALSFRS	ALSSQOL Total 46 Items
ALSFRS	Pearson Correlation	1	.257**
	Sig. (2-tailed)		.000
	N	342	342
Total 46 items	Pearson Correlation	.257**	1
	Sig. (2-tailed)	.000	
	N	342	342

** Correlation is significant at the 0.01 level (2-tailed).

Table 7

One-Way ANOVA: Between-Subject Factors

		Sum of Squares	df	Mean Squares	F	Sig
Speech	Between Groups	31.669	4	7.917	5.130	.001
	Within Groups	513.966	333	1.543		
	Total	545.635	337			
Salivation	Between Groups	23.523	4	5.881	3.751	.005
	Within Groups	522.112	333	1.568		
	Total	545.635	337			
Swallowing	Between Group	41.624	4	10.406	6.875	.000
	Within Groups	504.011	333	1.514		
	Total	545.635	337			

Discussion

The goal of this investigation was to determine whether or not QOL differs among ALS patients with varying levels of impairment in bulbar functioning (speech, swallowing, and salivation) and to verify whether or not patients with less impairment in these aspects of physical functioning will report better QOL. The results of this study that have been presented in the previous chapter demonstrate that there are significant differences in QOL, relative to level of bulbar functioning. This chapter will summarize the study and discuss the research findings. Study limitations and directions for future research on QOL and ALS are recommended.

Discussion of Research Findings

A weak relationship exists between physical functioning and QOL ($r = .257$; $p = \leq .001$). However, when comparing those with no impairment in bulbar functioning to those with various levels of bulbar impairment, the greatest difference in QOL is between those ALS patients who have no impairment and those who are demonstrating the

first signs of impairment. In other words, there was significant difference or differences approaching significance across all three items (speech, salivation and swallowing) when comparing groups with no impairments (ALSFRS scores of 4) to those with early, slight or detectable disturbances (ALSFRS scores of 3). There was basically no significant difference between varying groups who have evidence of some impairment. After some impairment is recognized, QOL is fairly stable across the groups. This suggests that the initial signs of impairment have the biggest impact on ALS patients' QOL. The three ALSFRS items of concern are discussed specifically in the following paragraphs.

Through post hoc testing it was discovered that in regard to speech functioning there was a significant difference (.007) in QOL for ALS patients with no functional impairment (ALSFRS score of 4; mean ALSSQOL score = 343.99) when they are compared with those having no functional speech (ALSFRS score of 0; mean ALSSQOL score = 301.92).

A significant difference (.004) was also observed in QOL for ALS patients with no functional impairment (ALSFRS score of 4) when compared with those patients initially

experiencing detectable speech disturbances (ALSFRS score of 3; mean ALSSQOL score = 318.49). This can be perceived as a crucial intervention time in working with the ALS patient. A change in the patient's self-perceived QOL was readily identified; therefore, planning and education should target this time period specifically.

As previously discussed, there was an observed difference approaching statistical significance (.061) between those with no impairment in salivation management and functioning (ALSFRS score of 4; mean ALSSQOL score of 337.01) and with those having slight, but definite excess of saliva in their mouths (ALSFRS score of 3; mean ALSSQOL score of 316.37).

Post hoc testing further demonstrated, that relative to salivation management and functioning, there was a significant difference (.016) in QOL for ALS patients when comparing those with no functional impairment (ALSFRS score of 4; mean ALSSQOL score of 337.01) with those having marked excess of saliva with some drooling (ALSFRS score of 1; mean ALSSQOL score of 294.55). However, those groups with marked drooling who require constant tissue or handkerchief use (ALSFRS score of 0) had a mean ALSSQOL score of 317.91. This is a higher ALSSQOL score than those

with marked excess of saliva with some drooling (294.55) and those with slight, but definite excess of saliva in their mouths (316.37). This may be evidence of acceptance on behalf of the ALS patients regarding their progression of salivation-related symptoms.

Post hoc testing of swallowing function demonstrated significant differences (.008) between those ALS patients who have normal eating habits with no impairment (ALSFRS score of 4; mean ALSSQOL score of 343.15) and those who displayed early eating problems such as occasional choking (ALSFRS score of 3; mean ALSSQOL score of 319.33).

However significant differences (.000) were also observed when comparing those who have normal eating habits and no impairment (ALSFRS score of 4; mean ALSSQOL score of 343.15) with those experiencing dietary consistency changes (ALSFRS score of 2; mean ALSSQOL score of 298.15). This appears to indicate that when the ALS patients initially have to make changes in the types of food they consume, it is a significant event with definite changes in self-reported QOL that should be recognized and addressed. Noteworthy is the fact that those with supplemental tube feedings (ALSFRS score of 1; mean ALSSQOL score of 313.22) and those who eventually receive nothing by mouth report

higher QOL (ALSFRS score of 0; mean ALSSQOL score of 314.20) than those who have to make dietary consistency changes (ALSFRS = 2; ALSSQOL mean = 298.15). This can possibly be explained by the continued acceptance of ALS patients regarding the progression of their swallowing symptoms and the fact that QOL may improve after they receive tube feedings because they are no longer choking, sputtering, and aspirating their food. This data may have an impact on the way in which the medical community treats ALS patients with swallowing impairments. Interventions meant to delay the introduction of a feeding tube may not necessarily be improving an ALS patient's QOL.

These results are important because there is not a linear relationship between physical functioning and QOL; therefore, the individual symptom progression needs to be examined specifically. There appear to be identified areas during which healthcare providers can intervene with ALS patients. Guidance, targeted information and preparation can be provided to the patients to assist them before they experience the levels of bulbar dysfunction that were identified as impacting QOL in the greatest degree. Problem solving skills can be introduced or reinforced. During these preliminary transition times decision clarification

can be obtained regarding difficult choices that the patient and family may have to make. Isolation and confusion are potential obstacles during these transitions from one level of functioning to another. Timely and well-planned interventions may have favorable effects on ALS patient self-reported QOL.

ANOVA revealed that self-reported QOL varied according to level of functioning for speech $F(4,333) = 5.13$, $p = .001$; swallowing $F(4, 333) = 6.88$, $p = .000$; and salivation, $F(4,333) = 3.75$, $p = .000$. The between-group differences for speech, swallowing and salivation are all significant ($p < .05$).

The mean age for the sample is fairly consistent with the general population of ALS patients. Most people who develop ALS are between 40 and 70 years of age. The average age at the time of diagnosis is 55; however, cases do occur in men and women in their twenties and thirties (ALS Association, 2007).

The mean duration of symptoms was 37.37 months (3.11 years) with a standard deviation of 37.40 months (3.11 years). The average life expectancy of a person with ALS is 24 months (2 years) to 60 months (5 years) from time of diagnosis. The ALS Association (2007) reports that 50% of

those diagnosed with ALS will live at least 36 months (3 years) or more after the initial diagnosis. Twenty-five percent will live 60 months (5 years) or more after diagnosis. Only 10% of those diagnosed with ALS will live (120 months) 10 years and beyond after the initial diagnosis. The mean duration of symptoms for this sample population is 37.37 months. Therefore the mean duration for the sample falls within the average life expectancy of all persons with ALS. However, there appear to be obvious outliers (e.g. 237 months) in this sample in regard to duration. It should also be noted that the subjects' duration of symptoms was obtained through self-report and not through medical records review. Additionally, subjects were not followed until death for this study.

The male to female ratio of 66.1% to 33.9% in the sample appears to be representative of the general ALS population. According to the ALS CARE Database, 60% of the people with ALS in the Database are men (ALS Association, 2007).

In the sample population (92.7%) and in the ALS CARE Database (93%), a large majority of the ALS patients are Caucasian. This data is representative of patients seen in many university-based, multidisciplinary ALS clinics;

however, the data may or may not be indicative of the general ALS population. The fact remains that it is not known whether or not race and ethnicity determine an individual's level of risk in developing ALS (Logroscino & Armon, 2007). It may be that minorities are underrepresented in these studies because they do not have the opportunities or even the desire to partake in such studies. They may have limited access to such care. Perhaps Caucasians for one reason or another (environment, genetics, lifestyle, etc…) are just more likely to develop ALS. Even though ALS may be more prevalent in one race or ethnicity than another, it does, however, occur throughout the world and affects people of all races, ethnic backgrounds and socioeconomic categories. Essentially, ALS strikes without prejudice. It is recognized that in this particular study there is a majority of English speaking, Caucasian clients. Conceivably, this may be an accurate representation of the general ALS population.

The mean MMT score for the sample was 3.69 with a standard deviation of 1.10. This is interpreted to mean that the average ALS patient in this sample received a grade indicating slightly less than active movement against gravity and resistance (4- = 3.67).

The MMT data examined for this particular study did not include the composite scores for the eight muscle tests. Total scores are not known. Furthermore, it is not known specifically the types of physical impairments that the sample population was experiencing. There is no doubt that they were varied. It is not known whether or not a particular patient was dealing primarily with upper or lower limb involvement. Nor was it known whether or not he or she had significant differences between right and left sided strength. It is expected that a patient with lower limb MMT scores of 3 or less would generally need assistance with walking. Additionally, upper limb MMT scores of 3 or less would most certainly indicate that a patient needed assistance with completing his or her activities of daily living.

ALSFRS comparisons are made with the patient's status prior to the onset of the disease, not with status at the last visit. The patients in this study sample demonstrated the greatest bulbar impairments in their speaking ability (mean = 2.96). Swallowing ability (mean = 3.17) was the second most impaired of the bulbar functioning. The least amount of impairment was in patients' ability to control saliva (mean = 3.24).

Limitations of the Study

There are a number of potential limiting factors identified for this study. Potential limiting issues include the fact that our data was obtained from the ALSSQOL validation study, conducted across seven university-based ALS Centers. The population involved in the study was made up of volunteer patients; these may not be representative of the typical ALS patient.

From the clinics, there were a limited number of patients (>15%) who declined to participate in the study; this, therefore, is still an issue to be considered. Those who chose not to participate could very well have had a QOL different from those included in the study (Simmons et al., 2006).

The ALSSQOL were administered in a personal interview format to patients who might have been fatigued, hurried, upset, nervous, or otherwise distracted, despite the researchers best attempts to optimize the environment. All of the sites involved in the study are multidisciplinary settings. The patients may come into the office so rarely

that many providers are scheduled to see them on the same day.

Another limitation that threatens the ability to generalize from the study's results (external validity) is the fact that the majority of the patients were English speaking, Caucasian clients without dementia or without considerable cognitive deficits. It may be difficult to infer with any certainty that the underrepresented populations who have ALS, such as minorities, non-English speaking patients, and those with cognitive impairments would benefit in the same manner from the results of the ALSSQOL studies.

Recommendations for Future Study

An area of future consideration should be the effect that the loss or impairment of handwriting to communicate has on an ALS patient's QOL.

Bulbar symptoms can present at the onset of ALS or towards the end of the disease course. Future research should examine how the timing of onset affects the QOL of ALS patients.

Another area of consideration for future study would be the utilization of the ALSSQOL for longitudinal studies. We are currently comparing individuals who are in myriad stages of ALS. It would be scientifically relevant to follow a number of individuals and to administer the ALSSQOL at various points throughout the disease progression to track their self-reported QOL.

Future studies should also include an examination of the significance of each change for ALS patients as they progress from normal level of functioning on the ALSFRS (item score 4) through each level of impairment (item scores 3, 2, 1, and 0). Researchers should investigate ALS patients' mindsets as they prepare to experience each of the transitions during this fairly predictable disease process.

Conclusions

In spite of the aforementioned limitations, it appears that this study's hypotheses have led to significant results. QOL does appear to be affected by the varying levels of impairment in speech, in salivation and in swallowing that accompany ALS. It is too broad a statement

to insist that QOL is not related to physical function. As this research has demonstrated, there are times when ALS patients self-reported QOL is adversely affected by physical impairments. However, the relationship between QOL and physical functioning is not linear; therefore, specific interventions should be planned for patients during the problem areas that have been identified. The expectation exist that future studies will expand upon these results.

References

American Academy of Neurology (2006). Manage ALS from the beginning: Care makes a difference. *AAN Guideline Summary for Clinicians*. Available: http://www.aan.com/professionals/practice/pdfs/manage_als.pdf

Amyotrophic Lateral Sclerosis Association (2007). What is ALS?, Initial Symptoms of the Disease, Who Gets ALS?, Diagnosing ALS. [On-line]. Available: http://www.alsa.org

Bremer, B. A., Simone, A. L., Walsh, S., Simmons, Z., Felgoise, S. H. (2004). Factors supporting quality of life over time for individuals with amyotrophic lateral sclerosis: The role of positive self-perception and religiosity. *Annals of Behavioral Medicine, 28*, 119-125. Retrieved from Ovid on August 20, 2006.

Bromberg, M. B. & Forshew, D. A. (2002). Comparison of instruments addressing quality of life in patients with ALS and their caregivers. *Neurology, 58*, 320-322.

Brooks, B. R., Miller, R. G., Swash, M., & Munsat, T. L. (2000). El Escorial revisited: Revised criteria for

the diagnosis of amyotrophic lateral sclerosis. *Amyotrophic Lateral Sclerosis & Other Motor Neuron Disorders, 1,* 293-299. Retrieved from Ovid on August 22, 2006.

Carr-Davis, E. M., Blakely-Adams, C., & Corinbilt, B.(2005). Adjusting to swallowing and speaking difficulties. *Living With ALS: Manual 5.* Amyotrophic Lateral Sclerosis Association.

Cedarbaum, J. M. & Stambler, N. (1997). Performance of the Amyotrophic Lateral Sclerosis Functional Rating Scale ALSFRS) in multicenter clinical trials. *Journal of Neurological Sciences, 152,* S1 - S9. Retrieved from Ovid on August 20, 2006.

Cedarbaum, J. M., Stambler, N., Malta, E., Fuller, C., Hilt, D., Thurmond, B., et al. (1999). The ALSFRS-R: a revised ALS functional rating scale that incorporates assessments of respiratory function. *Journal of Neurological Science, 169,* 13-21. Retrieved from Ovid on August 20, 2006.

Chio, A., Gauthier, A., Montuschi, A., Calvo, A., Di Vito, N., Ghiglione, P., et al. (2004). A cross sectional study on determinants of quality of life in ALS. *Journal of Neurology, Neurosurgery, and*

Psychiatry, 75, 1597-1601. Retrieved from jnnp.bmjjournals.com on September 7, 2006.

Ciechoski, M. A. (2005). Coping with Change. *Living With ALS: Manual 2*. Amyotrophic Lateral Sclerosis Association.

Cohen, S. R., Mount, B. M., Strobel, M. G., and Bui, F. (1995). The McGill Quality of Life Questionnaire: a measure of quality of life appropriate for people with advanced disease. A preliminary study of validity and acceptability. *Palliative Medicine, 9*, 207-219.

Cone, J. D. & Foster, S. L. (1993). *Dissertations and theses from start to finish: Psychology and related fields*. American Psychological Association: Washington, DC

Cuthbert, S. C. & Goodheart, G. J. (2007). On the reliability and validity of manual muscle testing: A literature review. *Chiropractic & Osteopathy, 15*. Published online at: http://www.pubmedcentral.nih.gov/picrender.fcgi?artid=1847521&blobtype=pdf

DePaul, R. & Kent, R. D. (2000). A longitudinal case study of ALS: Effects of listener familiarity and proficiency on intelligibility judgments. *American*

Journal of Speech-Language Pathology, 9, 230-240. Retrieved from Ovid on August 21, 2006.

Farrero, E., Prats, E., Povedano, M., Martinez-Matos, J. A., Manresa, F., & Escarrabill, J. (2005). Survival in amyotrophic lateral sclerosis with home mechanical ventilation: The impact of systematic respiratory assessment and bulbar involvement. *Chest, 127*, 2132-2138. Retrieved from www.chestjournal.org on August 20, 2006.

Forshew, D. & Hulihan, S. (2005). What's it all about? *Living With ALS: Manual 2*. Amyotrophic Lateral Sclerosis Association.

Friedrich, M. J. (2005). Experimental therapies hold promise for treating amyotrophic lateral sclerosis. Journal of the American Medical Association, 293, 1048-1049. Downloaded from www.jama.com on August 21, 2006.

Great Lakes ALS Study Group (2003). A comparison of muscle strength testing techniques in amyotrophic lateral sclerosis. *Neurology, 61*, 1503-1507. Retrieved from the Ovid database on February 20, 2007.

Grice, H. P. (1975). William James Lectures, Harvard University, published in part as "Logic and

conversation." In P. Cole and J. L. Morgan (Eds.), *Syntax and Semantics*, vol. 3. Speech Acts (pp. 41-58). New York: Seminar Press.

Hilari, K. & Byng, S. (2001). Measuring quality of life in people with aphasia: The Stroke Specific Quality of Life Scale. *International Journal of Language & Communication Disorders, 36*, 86-91. Retrieved from the Ovid database on November 20, 2006.

Hillel, A. D., Miller, R., Yorkston, K., McDonald, E., Norris, F. H., & Konikow, N. (1989). The amyotrophic lateral sclerosis severity scale. *Neuroepidemiology, 8,* 142-150.

Hillel, A., Dray, T., Miller, R., Yorkston, K., Konikow, N., Strande, E., et al. (1999). Presentation of ALS to the otolaryngologist/head and neck surgeon: Getting to the neurologist. *Neurology, 53*, 22-25. Retrieved from the Ovid database on November 20, 2006.

Hindelang, M. (2206). To help you live until you die: Patient education in hospice, palliative, and cancer care. *Home Healthcare Nurse, 24*, 142-144.

Jenkinson, C, Peto, V., Jones, G., & Fitzpatrick, R.

(2003). Interpreting change scores on the amyotrophic lateral sclerosis assessment questionnaire (ALSAQ-40). *Clinical Rehabilitation, 17*, 380-385.

Kubler-Ross, E. (1973). On Death and Dying. London: Tavistock.

Lechtzin, N., Schmidt, E. P., & Clawson, L. L. (2005). Approach to the patient with amyotrophic lateral sclerosis. *Clinical Pulmonary Medicine, 12*, 168-176. Retrieved from Ovid on August 20, 2006.

Leigh, P. N., Abrahams, S., Al-Chalabi, A., Ampong, M-A., Goldstein, L. H., Johnson, J., et al. (2003). The management of motor neuron disease. *Journal of Neurology, Neurosurgery, and Psychiatry, 74*, 32-47. Retrieved from jnnp.bmjjournals.com on August 20, 2006.

Lo Coco, G., Lo Coco, D., Cicero, V., Oliveri, A., Lo Verso, G., Piccoli, F., et al. (2005). Individual and health-related quality of life assessment in amyotrophic lateral sclerosis patients and their caregivers. *Journal of the Neurological Sciences, 238*, 11-17. Retrieved from Ovid on October 18, 2006.

Logroscino, G. & Armon, C. (2007). Amyotrophic lateral sclerosis: A global threat with a possible difference in

risk across ethnicities. *Neurology, 68*, 17. Downloaded from www.neurology.org on May 2, 2007.

Lou, J.S., Reeves, A., Benice, T. & Sexton, G. (2003). Fatigue and depression are associated with poor quality of life in ALS, *Neurology, 60*, 122-123.

Mathur, N. N. & Vaughn, T. L. (2006). Drooling. *eMedicine from WebMD*. Retrieved from http://www.emedicine.com/ent/topic629.htm

Mathy, P., Yorkston, K. M., & Gutmann, M. L. (2000). AAC for individuals with amyotrphic lateral sclerosis. In Beukelman D, Yorkston K., & Reichle J. (Eds.), *Augmentative and Alternative Communication in Adults with Acquired Neurologic Disorders*. Baltimore: Paul H Brookes Publishing Co.

Merriam-Webster Online Dictionary (n.d.). Retrieved November 22, 2006 from http://www.merriamwebster.com/dictionary/communication

McGuire, D., Garrison, L., Armon, C., Barohn, R., Bryan, W., Miller, R., et al. (1996). Relationship of the Tufts Quantitative Neuromuscular Exam (TQNE) and the Sickness Impact Profile (SIP) in measuring progression of ALS. SSNJV/CNTF ALS Study Group. *Neurology, 46*, 1442-1444.

Miller, R. G., Rosenberg, J. A., Gelinas, D. F., Mitsumoto, H., Newman, D., Sufit, R. et al. (1999). Practice parameter: The care of the patient with amyotrophic lateral sclerosis (an evidenced-based review): Report of the quality standards subcommittee of the American academy of neurology, *Neurology, 52*, 1311-1329. Downloaded from www.neurology.org on August 23, 2006.

Rippon, G. A., Scarmeas, N., Gordon, P. H., Murphy, P. L., Albert, S. M., Mitsumoto, H., et al. (2006). An observational study of cognitive impairment in amyotrophic lateral sclerosis. *Archives of Neurology, 63*, 345-352. Retrieved from Ovid on August 20, 2006.

Robbins, R. A., Simmons, Z., Bremer, B. A., Walsh, S. M., & Fischer, S. (2001). Quality of life in ALS is maintained as physical function declines. *Neurology, 56*, 442-444. Retrieved from Ovid on August 20, 2006.

Rowland, L. P. & Shneider, N. A. (2001). Amyotrophic lateral sclerosis. *New England Journal of Medicine, 344,* 1688-1700. Retrieved from Ovid on August 21, 2006.

Sacks, H., Schegloff, E. A. & Jefferson, G. (1974). A simplest systematics for the organization of turn-taking

for conversation. *Language, 50*, 696-735.

Simmons, Z. (2005). Management strategies for patients with amyotrophic lateral sclerosis from diagnosis through death. *The Neurologist, 11*, 257-270. Retrieved from Ovid on August 20, 2006.

Simmons, Z., Bremer, B. A., Robbins, R. A., Walsh, S. M., & Fischer, S. (2000). Quality of life in ALS depends on factors other than strength and physical function. *Neurology, 55*, 388-392. Retrieved from Ovid on August 20, 2006.

Simmmons, Z., Felgoise, S. H., Bremer, B. A., Walsh, S. M., Hufford, D. J., Bromberg, M. B., et al. (2006). The ALSSQOL: Balancing physical and nonphysical factors in assessing quality of life in ALS. *Neurology, 67*, 1659-1664. Retrieved from Ovid on December 4, 2006.

Squires, N. (2006). Dysphagia management for progressive neurological conditions. *Nursing Standard, 20*, 53-57. Retrieved from Ovid on August 21, 2006.

Stambler, N., Charatan, M., Cedarbaum, J. M., & ALS CNTF Treatment Study Group. (1998). Prognostic indicators of survival in ALS. *Neurology, 50*, 66-72. Retrieved from Ovid on August 20, 2006.

Statistical Program for Social Sciences (SPSS Graduate

Pack, 2006). (Version 15.0), Chicago, IL: SPSS, Inc.

Tabachnick, B. G. & Fidell, L. S. (2001). *Using Multivariate Statistics, 4th edition*. Allyn & Bacon, Boston.

Talbot, K. (2002). Motor neuron disease. *Postgraduate Medical Journal, 78*, 513-519. Retrieved from jnnp.bmjjournals.com on August 20, 2006.

Thomas, J. (1995). *Meaning in interaction: An introduction to pragmatics.* Harlow: Pearson Education.

Traynor, B.J., Alexander, M., Corr, B., Frost, E., & Hardiman, O. (2003). Effects of a multidisciplinary amyotrophic lateral sclerosis (ALS) clinic on ALS survival: a population based study, 1996-2000. *Journal of Neurology, Neurosurgery, and Psychiatry, 74*, 1258-1261. Downloaded from www.jnnp.bmj.com on November 16, 2006.

Vasar, L. T., Weinacker, A. B., Henig, N. R. & Raffin, T. A. (2002). Ethical issues in the long-term management of progressive degenerative neuromuscular diseases. *Seminars in Respiratory and Critical Care Medicine, 23*, 307-314.

Walling, A. (1999). Amyotrophic lateral sclerosis: Lou

gehrig's disease. American Family Physician, 59, 1489-1499. Retrieved on August 23, 2006 from http://www.aafp.org/afp/990315ap/1489.html

Walsh, S. M., Bremer, B. A., Felgoise, S. H., & Simmons, Z. (2003). Religiousness is related to quality of life in patients with ALS. *Neurology, 60*, 1527-1529. Retrieved from Ovid on August 20, 2006.

Wasner, M., Bold, U., Vollmer, T. C., & Borasio, G. D. (2004). Sexuality in patients with amyotrophic lateral sclerosis and their partners. *Journal of Neurology, 251*, 445-448.

Watts, C. R. & Vanryckeghem, M. (2001). Laryngeal dysfunction in amyotrophic lateral sclerosis: A review and case report. *BMC Ear, Nose and Throat Disorders, 1*. Published online and retrieved on December 12, 2006 from http://www.biomedcentral.com/1472-6815/1/1

WHOQOL: Measuring Quality of Life. Geneva: World Health Organization; 1997

Winhammer, J. M. C., Rowe, D. B., Henderson, R. D., & Kiernan, M. C. (2005). Assessment of disease progression in motor neuron disease. *The Lancet Neurology, 4*, 229-238. Retrieved from Ovid on August 20, 2006.

World Health Organization Group (1998). Development of the World Health Organization WHOQOL-BREF quality of life assessment. Psychological Medicine, 28, 551-558.

Yorkston, K. M. & Beukelman, D. R. (2000). Decision making in AAC intervention. In Beukelman D, Yorkston K., & Reichle J. (Eds.), *Augmentative and Alternative Communication in Adults with Acquired Neurologic Disorders*. Baltimore: Paul H Brookes Publishing Co.

Yorkston, K. M., Beukelman, D. & Ball, L. (2002). Management of dysarthria in amyotrophic lateral sclerosis. *Geriatrics & Aging, 5*, 38-41. Retrieved: http://www.geriatricsandaging.com/PDF/PDFJanFeb2002/0501dysarthria.pdf

Yorkston, K. M., Beukelman, D. R., Strand, E. A., & Bell, K. R. (1999). *Management of motor speech disorders in children and adults*. Austin, TX: Pro-Ed

Yorkston, K. M., Strand, E., Miller, R., Hillel, A., & Smith, K. (1993). Speech deterioration in amyotrophic lateral sclerosis: Implications for the timing of intervention. *Journal of Medical Speech-Language Pathology, 1,* 35-46.

Appendix A

Sample Questions from the ALSSQOL

Negative Emotion:

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.**

		Not at All										Very Much
22.	I have been depressed.	0	1	2	3	4	5	6	7	8	9	10
31.	I have felt hopeless.	0	1	2	3	4	5	6	7	8	9	10

Intimacy:

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.

		Not at All										Very Much
42.	My desire for emotional has been strong.	0	1	2	3	4	5	6	7	8	9	10

Satisfaction with Relationships and the Environment:

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.**

		Not at All										Very Much
26.	Relationships with those closest to me have been satisfying.	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

Religiosity:

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.**

		Not at All										Very Much
23.	My religion has been a source of strength or comfort to me.	0	1	2	3	4	5	6	7	8	9	10

Physical Functioning:

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.**

		Not at All										Very Much
10.	I have felt physically terrible.	0	1	2	3	4	5	6	7	8	9	10

Bulbar Functioning:

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

		Not at All										Very Much
3.	Eating.	0	1	2	3	4	5	6	7	8	9	10
4.	Excess saliva.	0	1	2	3	4	5	6	7	8	9	10
6.	Speaking.	0	1	2	3	4	5	6	7	8	9	10

Appendix B

ALS Functional Rating Scale

1. Speech
4 Normal speech process
3 Detectable speech disturbance
2 Intelligible with repeating
1 Speech combined with non-vocal communication
0 Loss of useful speech

2. Salivation
4 Normal
3 Slight but definite excess of saliva in mouth; may have nighttime drooling
2 Moderately excessive saliva; may have minimal drooling
1 Marked excess of saliva with some drooling
0 Marked drooling; requires constant tissue or handkerchief

3. Swallowing
4 Normal eating habits
3 Early eating problems-occasional choking
2 Dietary consistency changes
1 Needs supplemental tube feeding
0 NPO (exclusively parenteral or enteral feeding)

4. Handwriting
4 Normal
3 Slow or sloppy; all words are legible
2 Not all words are legible
1 Able to grip pen but unable to write
0 Unable to grip pen

5a. Cutting Food and Handling Utensils (patients without gastrostomy)
4 Normal
3 Somewhat slow and clumsy, but no help needed
2 Can cut most foods, although clumsy and slow; some help needed
1 Food must be cut by someone, but can still feed slowly
0 Needs to be fed

5b. Cutting Food and Handling Utensils (alternate scale for patients with gastrostomy)
4 Normal
3 Clumsy but able to perform all manipulations independently

2 Some help needed with closures and fasteners
1 Provides minimal assistance to caregiver
0 Unable to perform any aspects of task

6. Dressing and Hygiene
4 Normal function
3 Independent and complete self-care with effort or decreased efficiency
2 Intermittent assistance or substitute methods
1 Needs attendant for self care
0 Total dependence

7. Turning in Bed and Adjusting Bed Clothes
4 Normal
3 Somewhat clumsy, but no help needed
2 Can turn alone or adjust sheets, but with great difficulty
1 Can initiate, but not turn or adjust sheets alone
0 Helpless

8. Walking
4 Normal
3 Early ambulation difficulties
2 Walks with assistance
1 Nonambulatory functional movement
0 No purposeful leg movement

9. Climbing Stairs
4 Normal
3 Slow
2 Mild unsteadiness or fatigue
1 Needs assistance
0 Cannot do

10. Breathing
4 Normal
3 Shortness of breathe with minimal exertion (walking, talking, etc…)
2 Shortness of breathe at rest
1 Intermittent (e.g. nocturnal) ventilatory assistance required
0 Ventilator dependent