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The Effects of Speech Impairment on Quality of Life over Time in Patients with Amyotrophic Lateral Sclerosis

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THE EFFECTS OF SPEECH IMPAIRMENT ON QUALITY OF LIFE OVER TIME IN PATIENTS WITH AMYOTROPHIC LATERAL SCLEROSIS

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Dissertation Approval

This is to certify that the thesis presented to us by Jacqueline Kelley Blessinger on the 26th day of May, 2015, 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.

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Abstract

Limited research exists on how speech impairments affect quality of life (QOL) factors over time in patients with amyotrophic lateral sclerosis (ALS). A review of the literature, including the history, disease course, and prevalence of ALS, is presented. Physical and psychosocial functioning, especially the decline of bulbar functioning's potential impact on QOL factors and communication style, are outlined. This study is a follow-up study on a study by Duff, who found a significant difference in QOL relative to level of bulbar functioning in a cross-sectional design. The current study used a longitudinal design to look at bulbar functioning, specifically levels of speech, and how that affects certain aspects of QOL over time. This study used archival data, collected from a multidisciplinary clinic at three time points, and evaluated the following measures: ALS Functional Rating Scale–Revised (ALSFRS–R) and the ALS–Specific Quality of Life Measurement–Revised (ALSSQOL–R). ALSSQOL–R domains of Negative Emotion, Interaction with People and the Environment, Intimacy, and Bulbar Function were analyzed by repeated measures tests, and correlated to speech impairment over time. Results indicated that as speech became significantly more impaired, no significant differences were observed on the Negative Emotion, Interaction with People and the Environment, and Intimacy subscales. However, as the ability to speak declines over time, participants' Bulbar Functioning subscale scores decreased. These findings suggest that increased bulbar symptoms are perceived as more problematic over time point. However, results also indicated that at the third time point (most recent clinic visit) the relationship between actual speech impairment and perceived bulbar functioning weakens.

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Chapter 1

Introduction

Statement of the problem.

Amyotrophic lateral sclerosis (ALS) is characterized as a degenerative motor neuron disease that causes progressive immobility and functional impairments to speech, breathing, and swallowing, leading to eventual death, typically from respiratory failure (Gordon, 2013; Lonergan, Mitsumoto, & Murray, 2014; Tramonti, Bongioanni, Di Bernardo, Davitti, & Rossi, 2012). To date, there is no cure for the disease, and thus treatment is largely palliative. Therefore, monitoring the patient's quality of life (QOL) becomes essential for accurate assessment and effective palliative treatment (Lou, Moore, Gordon, & Miller, 2010).

Knowledge of self-perceived QOL serves as an essential factor in treating patients with ALS because it helps providers and caregivers understand how impairments affect the many aspects of that person's life. The term QOL refers to health-related QOL or health status and encompasses a patient's perceptions of his or her functioning that includes, physical, emotional, and mental well-being (Felgoise et al., 2009). Measurement of health-related QOL emphasizes the patient's functional status (Tramonti et al., 2012). However, additional domains, such as employment, family, social relationships, and finances, also influence overall QOL (Felgoise et al.; Gibbons et al., 2013; Trail, Nelson, Van, Appel, & Lai, 2004).

Research with ALS patients has found that QOL does not relate to measures of physical functioning and strength; however, psychosocial concerns and existential

stressors are significantly affected (McLeod & Davis, 2007; Simmons, Bremmer, Robbins, Walsh, & Fischer, 2000).

The trajectory of the disease causes ALS patients to face many psychosocial challenges, including worries about family members, lack of caregiver support, emotional changes, the inability to continue working, and subsequent financial difficulties. These struggles are often compounded by existential factors, such as worries about illness progression, dependency on others, caregivers' well-being, outlook about the future, loneliness, and religiosity (Trial et al., 2004). Administering individual QOL assessments offers physicians and other healthcare professionals an understanding of patients' perceived well-being (Felgoise et al., 2009). These measurements become particularly useful for those patients with ALS who experience difficulties with communication.

The course of ALS compromises a patient's ability to speak, manage food consumption, and drink liquids due to impaired bulbar functioning. Bulbar functioning refers to the motor neurons located in the bulb region of the brain stem. This part of the brain controls the muscles responsible for chewing, swallowing, speaking, and maintaining an open airway (Kühnlein et al., 2008). The clinical factors of ALS reveal notable individual variability across patients and presentation of the disease over time (Chiò et al., 2009; Gordon, 2013; Lonergan et al., 2014). Some patients may experience bulbar symptoms early in the course of their illness, characterizing their ALS diagnosis as bulbar-onset, whereas others might not manifest bulbar dysfunction until later as the disease progresses (Gordon, 2013; Hardiman, 2011; Lonergan et al.).

Regardless of when bulbar symptoms present, disruption to this area of the brain often leads to a change in lifestyle and dependency on others. Researchers identified bulbar impairment as an independent predictor of psychological distress in patients with ALS (Goldstein, Atkins, Landau, Brown, & Leigh, 2006). Goldstein and colleagues suggested that patients presenting with bulbar impairment should receive additional assessment for possible depressive symptoms.

In an effort to ameliorate the physical and emotional distress experienced by patients with bulbar dysfunction, early detection of impaired speech and voice is essential (Kühnlein et al., 2008). A cross-sectional study by Duff (2008) evaluated QOL in groups with distinct differences in their level of speech impairment, according to the ALS Functional Rating Scale–Revised (ALSFRS–R). Duff indicated that after patients recognize some impairment, QOL stabilizes across the groups, suggesting the initial signs of impairment cause the greatest impact on ALS patients' QOL. These findings did not indicate a linear relationship between physical functioning and QOL, based on a cross-sectional study. Therefore, individual symptom progression and patient QOL require evaluation over time. Observing bulbar symptoms relative to QOL over time is necessary in order to provide targeted guidance, information, and preparation to assist patients before they experience levels of bulbar dysfunction that negatively impact QOL (Duff).

Earlier research supported Duff's (2008) conclusion proposed improving symptom control, and encompassed interventions directed at benefiting functional health status that are imperative to caring for patients with ALS (Neudert, Wasner, & Borasio, 2004). Addressing QOL factors prophylactically, incorporated as part of treatments, aims to benefit a patient's physical functioning and emotional well-being. Thus, treatment

needs to be timely and well-planned in order to have favorable effects on self-reported QOL in ALS patients (Duff). Therefore, it is essential to develop a better understanding of exactly when the decline in speech functioning occurs relative to its specifics effects on certain aspects of QOL such as Negative Emotion, Interaction With People and the Environment, Intimacy, and Bulbar Functioning over time.

Purpose of the study.

The present study continued research from an earlier cross-sectional study that revealed significant differences in QOL relative to level of bulbar functioning (Duff, 2008). Duff indicated that those ALS patients demonstrating initial signs of bulbar impairment had significantly lower ratings of QOL than patients with no functional impairment. Therefore, the objective of the current research was to determine how progressive speech decline in patients with ALS affects specific aspects of QOL as defined by the ALSSQOL–R: Negative Emotion, Interaction with People and the Environment, Intimacy, and Bulbar Functioning over time. Assessing patients' self-reported QOL longitudinally serves to advance the understanding of how speech impairment affects these areas of QOL. The goal of this study was to learn if, and how, the ability to speak influences individuals' Negative Emotion, Interaction with People and the Environment, Intimacy, and Bulbar Functioning over time and further analyze the findings of Duff's study with a longitudinal design.

Literature Review

History of ALS.

Interest in Amyotrophic Lateral Sclerosis (ALS) began at the end of the 19th century as the original example for supporting the research methodology of French neurologist Jean-Martin Charcot. Charcot extensively studied and defined the lesions of several spinal cord and cortical/subcortical syndromes. The core of his neurological research utilized a technique known as the "anatomo-clinical method," a two-part methodology to determine the correlation between clinical signs observed during life and anatomical lesions identified postmortem (Goetz, 2000).

Based on meticulous observation and extensive documentation, Charcot identified patients presenting with weakness but without sensory difficulties, epilepsy, or other possible motor disorder diagnoses. All of Charcot's patients presented with weakness, but only a few also demonstrated spasticity with contractures, whereas others were amyotrophic, with atrophy. He followed these patients clinically, documenting their progressive functional decline with written notes and medical drawings. This scrupulous documentation allowed Charcot to separate cases of acute-onset weakness from those that exhibited a slower and more progressive course of illness (Goetz, 2000).

At the autopsy step of his anatomic-clinical method, he found lesions typical of acute amyotrophy and a distinctive sclerosis of the spinal cord (Goetz, 2000). These cases became essential to supporting Charcot's thesis and represented the first cases of ALS (Goetz). More recently, there have been some discoveries in molecular and genetic medicine that have offered small contributions to understanding the disorder. However,

since Charcot's death in 1893, few additions and revisions have been made to his original work.

Disease course.

Today, ALS is understood as a disease of the motor neurons. Motor neuron disease refers to a group of disorders categorized by progressive degeneration of upper motor neurons (UMNs) and lower motor neurons (LMNs) in the brain and spinal cord (Gordon, 2013). The trajectory of ALS causes progressive and distinguishing symptoms that include weakness, spasticity, poor dexterity, involuntary muscle twitching, and the wasting of muscle groups (Lonergan et al., 2014). The disease is further characterized by a slow onset of anterior horn cell lesions of the spinal cord, beginning with the wasting of the small muscles in one or both hands.

Several variations/subtypes of the disease exist and are diagnosed from knowledge about the site of symptom onset and what motor neurons are prominently affected by the degeneration of this region. In the spinal form of ALS, the degeneration of the LMNs affects the lumbosacral segments. Symptoms manifest in patients as weakness, atrophy, and fasciculation (involuntary contraction of muscles). Simultaneously, UMNs are also disturbed, creating an automatic overreaction of reflexes and thus producing the classic ALS presentations of upper and lower motor neuron involvement (Goetz, 2003).

Onset of the disease in the brainstem creates lesions in the lower cranial nuclei that lead to progressive bulbar impairment. LMN degeneration is evidenced by weakness, atrophy, and fasciculation of the tongue. Observed weakness of voluntary palatal movement during simultaneous UMN involvement is evidenced by a very abrupt

gag reflex and jaw jerk (Goetz, 2003). However, in a variation of the disorder, primary lateral sclerosis, the UMNs are selectively affected at the onset and a gradual spastic paresis of all four limbs and pseudobulbar palsy is expressed (Goetz). Regardless of onset region, the disorder eventually involves motor neuron degeneration of the entire region of the spinal cord to the front of the brain. An ALS diagnosis is made by electromyography test (EMG needle test) in order to confirm or rule out widespread LMN disease (Gordon, 2013; Lonergan et al., 2014).

Incidence and prevalence.

In regards to incidence and prevalence of ALS based on United States population studies (ALS Association, 2015), approximately 30,000 people are living with ALS and slightly more than 5,600 people are diagnosed each year. ALS is 20% more common in males than females (ALSA). However, studies have shown that bulbar onset has a somewhat female predominance (Lonergan et al., 2014). The ALS Association reported the ages of onset as occurring between 40 and 70 years old, with the peak age of onset occurring at approximately 55 years old. For patients with ALS, Lonergan et al. indicated that peak mortality occurs between the ages of 65 and 75 years old. In comparison to other neurological disorders of aging, such as Parkinson's or Alzheimer's, the age of onset suggests that ALS is not a disease of aging, but rather a disease for which age is one of several risk factors (Chiò et al., 2009; Gordon, 2013; Hardiman, 2011; Lonergan et al.).

Nevertheless, the etiology of the disease remains largely unknown. Several studies claimed to have identified potential various risk factors. Clustering of ALS has identified among certain occupations, such as Italian soccer players (Gordon, 2013;

Hardiman, 2011) and U.S. veterans of the Gulf War (ALSA, 2015; Horner et al., 2003). However, researchers argue that further evaluation of risk factors requires large, unbiased, population-based case controlled studies because with the exception of smoking, definitive evidence of risk remains to be established (Hardiman).

Other studies suggest a possible inheritability of ALS. In fact, ALS is considered a complex genetic disorder in which multiple genes, coupled with environmental exposures, combine to make a person susceptible. However, few genetic or environmental risks have been identified to date (Gordon, 2011), as only between 5% and 10% of cases are inherited and are referred to as familial ALS (Gordon; Hardiman, 2011). As previously mentioned, the course of ALS is progressive and is generally associated with a life expectancy of approximately 3 to 5 years after symptom onset (Gordon; Hardiman). The range of survival extends up to decades for only approximately 5% of patients (Gordon).

Diagnosing ALS. Receiving an ALS diagnosis is a process and often described as a journey. The delay between symptom onset and date of diagnosis averages about 14 months (Kiernan et al., 2011; Leigh et al., 2003). The median time from the onset of symptoms until death has been found to be from 23 to 48 months (Lonergan et al., 2014; Simmons, 2005). The pathway to diagnosis is multifaceted and too frequently filled with uncertainty and unclear medical options. Further complicating the issue of diagnostic uncertainty is the issue of no confirmatory tests, scans, or clinical laboratory examinations for ALS.

In an effort to address this issue, many physicians utilize the El Escorial World Federation of Neurology Criteria. These criteria provide a standardized guide to clinical,

pathological, and electrodiagnostic examinations for diagnosing ALS (Lonergan et al. 2014). Originally, it was a guide designed as a research tool for clinical trials. However, the criteria are now used among most physicians for distinguishing those patients with possible symptoms of ALS (Gordon, 2013). El Escorial criteria divide the patients into those with definite, probable, possible, and suspected ALS, depending on the pattern of LMN and UMN degeneration relative to symptoms in bulbar, cervical, thoracic, and lumbosacral sites (Brooks, Miller, Swash, & Munsat, 2000; Lonergan et al.). Moreover, according to the criteria, the diagnosis of ALS requires the presence of the following symptoms:

(1) Signs of lower motor neuron degeneration by clinical, electrophysiological or neuropathological examination.

(2) Signs of upper motor neuron degeneration by clinical, electrophysiological or neuropathological examination.

(3) Progressive spread of signs within a region or to other regions, together with the absence of diagnostic imaging.

(4) Electrophysiological evidence of other disease processes that might explain the signs of LMN and UMN degenerations.

(5) Neuroimaging evidence of other disease processes that might explain the observed clinical signs. Following through these standardized steps helps physicians differentially diagnosis between other neurological diseases and ALS to make a confirmatory diagnosis (ALSA, 2015).

The initial step involves understanding a patient's history and conducting a physical examination, along with the appropriate neurological and clinical laboratory

examinations to determine the clinical significance of any finding. However, even after a patient completes the necessary diagnostic steps, the diagnosis often remains unclear. Findings from the physical and neurological examination only allow a clinical diagnosis of ALS to a particular degree of certainty (ALSA, 2015). These ALS diagnoses are described along a spectrum of certainty, ranging from a definitive ALS diagnosis to a diagnosis of less certainty.

Determining the degree of certainty from the neurological and physical examination is also important for diagnosing the specific type of ALS. As previously mentioned, obtaining a comprehensive history from the patient with regards to symptom disease onset, past medical history and injuries, and family medical history are essential to determining the etiology and pathological process of the motor neuron degeneration (ALSA, 2015). LMN and UMN degeneration progresses as various complex patterns in ALS patients and manifests itself through complex and impairing symptoms. In particular, knowledge of a patient's region of symptom onset offers the treatment team valuable information with regards to prognosis and disease course.

Physical functioning and strength.

Understanding a patient's prognosis and disease course supports physicians and other healthcare professionals in providing effective palliative care, while maximizing QOL. The trajectory of the disease causes ALS patients to experience increased limb weakness, decreased mobility, breathing difficulties, bulbar impairment, and nutritional concerns. Although it is an incurable disease, the progression of some symptoms can be abated and possibly even avoided with supportive therapies. Unfortunately, the literature on controlled trials of symptom management is limited (Gordon, 2011). Selection of

supportive therapies generally depends on physician experience, and although practice guidelines outline common strategies, management practices remain extremely varied (Gordon).

Discrepancies across treatment practices are necessary, given the individual variability among patients. Leigh et al. (2003) described the distinctions between the manifestations of ALS symptoms as, somewhat arbitrary. Given the ambiguity about and amount of time required for confirmatory diagnosis of ALS, this poses considerable difficulty for treatment.

Motor concerns. Many ALS symptoms affect patients more predictability and with less uncertainty. For example, spasticity is a symptom that patients with ALS experience. Spasticity is an increase of uncontrolled muscle tone that creates stiffness, impairing the ability to utilize limbs and facial muscles (Gordon, 2011). Protracted inactivity compounds the complications in patients with already severe muscle weakness and limited mobility. Prolonged inactivity results in functional deterioration of already weakened limbs due to contractures and worsening atrophy (Gordon). Corcia and Meininger (2008) reported that increased muscle tightness and functional disability has a major negative impact on QOL.

Nutritional concerns. Poor nutrition is a predictor of survival in ALS patients (Gordon, 2011), as malnourishment is a relevant contributing outcome for this population (Chiò et al., 2009). Bulbar dysfunction creates many additional challenges for patients, including difficulties swallowing, arm weakness that restricts lifting food to the mouth, and hypermetabolism, which causes a negative calorie balance (Chiò et al.). Thus, ALS patients with bulbar onset often have many additional concerns with regards to their

nutritional well-being. Poor nutritional symptoms place patients at risk for malnutrition, dehydration, and weight loss (Simmons, 2005). Accordingly, researchers emphasize strategies of nutritional maintenance as a predictor of survival with this patient population (Gordon; Simmons).

Monitoring weight loss is the most effective method for assessing caloric stability. Evaluation by a speech pathologist offers further information regarding risk of aspiration, ability to maintain nutritional balance, and compensatory strategies (Gordon, 2011). If a patient is no longer able to sustain a positive caloric balance via mechanical digestion, a gastrostomy is indicated to ensure sufficient nutrition (Gordon, 2013). However, the benefits of gastrostomy for survival and QOL are unknown (Limousin et al., 2010). Nonetheless, patients with ALS do experience significant nutritional concerns, and it is important to further examine this decline relative to QOL over time. Additional information about a patient's nutrition and other areas of physical functioning would allow for routine and individualized monitoring of treatment relative to symptom progression and QOL.

Physical functioning and strength maintenance.

As previously mentioned, strategies for treatment management tend to differ from clinician to clinician. Researchers briefly mention supportive therapies and strongly emphasize the use of medications to treat symptoms of motor concerns. In a literature review of management strategies for patients with ALS, Leigh et al. indicated an underutilization of opiates and reported that this class of medications for ALS patients demonstrated effectiveness in ameliorating breathlessness and anxiety (2003). Other reviews cautiously supported opiate medications for the sole purpose of pain

management. Simmons (2005) warned that prescribing opiates might cause depressed respiration, decreased airway protection, and suppressed coughing. In terms of other pharmacological therapies, there seems to be agreement across multiple reviews that benzodiazepines yield beneficial and efficacious results for managing symptoms of spasticity (Gordon, 2011; Hardiman, 2011; Leigh et al., 2003; Simmons, 2005). Few studies have compared the effects of this class of medication or antidepressants to QOL in patients with ALS (Pagnini, Manzoni, Tagliaferri, & Gibbons, 2014).

Measures of physical functioning and strength.

In order to provide effective and appropriate treatment strategies for ALS symptoms, patients require constant assessment and monitoring. Various measures help clinicians to evaluate patients' physical functioning and strength. Clinical trial outcome measurements of functioning and strength often include survival, manual muscle strength testing, maximal voluntary isometric contraction of muscle, pulmonary function testing, electrophysiological, and functional rating scales (Sorensen, 2004). Given that ALS causes progressive weakness and ultimately death, Sorensen argues the most accurate means of determining the extent and progression of the disease is to assess muscle strength.

However, measures of functional rating are more frequently employed as primary outcomes (Castrillo-Viguera, Grasso, Simpson, Shefner, & Cudkowicz, 2010). The Amyotrophic Lateral Sclerosis Functional Rating Scale–Revised (ALSFRS–R) is the most widely used and validated clinical rating for assessing ALS patients' level of functioning in four major domains: bulbar, gross motor, fine motor, and respiration (Cedarbaum & Stambler, 1999; see also Cedarbaum & Stambler, 1997). Researchers

aimed to determine how much change in the ALSFRS–R was necessary before patients perceived a clinical change (Gordon et al., 2007). Findings have indicated that the ALSFRS–R is an overall more reliable measure of function compared to the MMT, given that there are fewer missing values in ALSFRS–R scores. Furthermore, the results suggest patients may not be able to perceive intervention effects until the impact on ALSFRS–R is 9 points or more. These findings have suggested a greater need for understanding a patient's perception of functioning. There are limitations when using self-reported physical functioning. For example, items measuring functional impairment may not be reported as problematic compared to actual level of functioning.

In an earlier review of functional scales for the ALS population, the Brooks noted how rating scales provide a means of describing the clinical condition of a patient with a disease process (2002). These measures help clinicians identify areas of dysfunction due to the underlying disease process. Brooks further reported that all ALS functional rating scales to date included self-reported, rated, or measured items relating to four major demarcated domains of anatomical functioning: bulbar, respiratory, arms, and legs. The items provide a global measurement of the patient's condition (Brooks). Compared to other measures of functioning, the ALSFRS–R is the most widely used and accepted measure, as it is well validated for monitoring the functional status of patients with ALS. It is also utilized to monitor disease progression (Cedarbaum & Stambler, 1997).

Overall, the literature suggests the importance of research that focuses on facilitating a better understanding about the relative impact of physical functioning and strength impairment on the clinical state of the patient and, in turn, the effect of this clinical state on the well-being of the patient (Brooks, 2002). Therefore, it is important to

examine and discuss the aspects of psychosocial and emotional functioning of the ALS patient for the purposes of understanding the larger impact on communication dysfunction. Physical functioning and areas of emotional well-being can be assessed if a patient participates in treatment at a multidisciplinary clinic.

Multidisciplinary care.

Sustaining a patient's QOL by means of symptom management is the main objective of current ALS treatment. Multidisciplinary clinics that exclusively serve the ALS patient provide symptomatic and palliative care. The clinics provide management of nutrition and respiratory needs, opportunity for clinical trial enrollment, medication management, psychosocial supports, and community resource management.

Leigh et al. (2003) encourages early referral for team care as best practice. The goal of multidisciplinary clinics is to facilitate clinical expertise by sharing knowledge and resources that emphasize patient-centered care. Physicians that specialize in ALS are usually neurologists. These neurologists oversee a team of healthcare professionals who also share expertise in evaluating and managing patients with ALS (Simmons, 2005). Patients and families benefit from having questions addressed by the professionals from multiple disciplines in one visit. The clinic serves to mitigate two limiting factors for an ALS patient: time and energy (Gordon, 2011). A recent study has shown contrary results and indicates that, while conserving time and energy may be an advantage for attending a multidisciplinary clinic, for others the time and energy used for travel and an all-day appointment are perceived as disadvantages (Stephens, Young, Felgoise, & Simmons, 2015).

While the physician maintains responsibility for overall patient care, an ALS nurse provides nursing care. In addition, other allied health professionals provide specialized care to ALS patients, such as physical therapists who evaluate and manage limb strength, occupational therapists who manage smaller motor functions, dietitians who assess nutritional needs, speech pathologists who evaluate bulbar function, and respiratory therapists who aid in treating respiratory symptoms (Gordon, 2011). These specialties ensure that patients maintain, manage, and compensate for loss of functioning that influences activities of daily living. Many clinics also employee social workers who help patients and caregivers to coordinate with outside agencies, such as health insurance and disability, and to network with other community resources (Gordon). Other medical specialists, such as pulmonologists, gastroenterologists, psychiatrists, and psychologists, also help to address the specialized needs of ALS patients.

Patients are evaluated frequently in an effort to proactively detect and treat problems (Gordon, 2011). The assessment and delivery of aids and assistive devices requires that physiotherapists, occupational therapists, rehabilitation providers, and orthotics all work collaboratively. Implementation of these assessments and interventions would be better utilized if clinicians could target treatments that were timed to a patient's change in level of functioning and/or change in QOL.

Studies have shown that multidisciplinary ALS care prolongs survival, and ALS patients with multidisciplinary care have better mental QOL (Van den Berg et al., 2005, p. 1266). For those patients with bulbar onset who receive care at a multidisciplinary clinic, the difference in survival is even greater. Observed beneficial influences of the multidisciplinary clinic may be attributed to the combined effects of the comprehensive

services provided (McLeod & Clarke, 2007). Therefore, it is imperative to better understand the relationship between bulbar function and QOL to inform multidisciplinary teams of when and how to implement the multifaceted interventions needed to address QOL-related concerns.

Caregiver and social support.

Cognitive theory of psychological stress and coping is described as a reciprocal relationship between the person and environment, which is perceived by the individual as overwhelming or endangering to their well-being (Folkman, Lazarus, Gruen, & DeLongis, 1986). Researchers have widely demonstrated that social support and social relationships established prior to disease diagnosis strongly influence psychosocial and emotional health and enhance patient QOL (Goldstein et al., 2006; McLeod & Clarke, 2007).

Findings are different for the ALS patient population. Caregiver and social supports for ALS patients prove to be meaningful factors that contribute to maintaining adequate mental health functioning. Therefore, if patients have compromised speech or communication, this may negatively impact the caregiver-patient relationship and their ability to acquire necessary support. When speech is impaired, patients are forced to adopt alternative patterns and styles of communication and speech (McKelvey, Evans, Kawai, & Beukelman, 2012). Patients can no longer express themselves to caregivers in manner unique to that individual's style or pattern of communication. This unwanted change potentially impacts the caregiver-patient relationship and the patient's ability to acquire support effectively.

Matuz, Birbaumer, Hautzinger, and Kübler (2010) proposed that ALS represents a stressful life event. The researchers adapted Lazarus and Folkman's model of stress coping to chronic illness in order to examine psychosocial adjustment in ALS patients across four variables related to physical functioning, emotional well-being, social support, and coping. Matuz and colleagues hypothesized that the model would also successfully predict psychological adjustment to the diagnosis for patients with ALS. Results indicated that more severe physical impairment does not lead to decreased QOL and depressive symptoms (Matuz et al.). Findings showed that all together, perceived social support, appraisal of coping strategies, and sense of independence explained 61% of the variability in depressive symptoms. In terms of QOL, perceived social support, seeking support and information, and avoidance accounted for the remaining 56% of the variability (Matuz et al.).

Results from Matuz and colleagues also suggested the predictability of social support and coping appraisal and strategies in how patients adjust to the ALS diagnosis. Perhaps of most importance, findings highlighted that the quality of social support distinctly predicts both QOL and depressive mood (Matuz et al.). This study indicated that caregiver and social support play significant roles in the life of an ALS patient. Overall, the study proposed a novel model that integrated psychosocial adjustment to ALS as a means of guiding treatment. The ALS–Specific Quality of Life Scale–Revised (ALSSQOL–R) measures patients' perceptions of their interaction with people and their environment and intimacy. Scores from these domains may correspond with these variables and their impact. Likewise, bulbar functioning or impairment may have an impact on the ability to maximize benefits from available support persons.

Overall, the course of ALS causes a significant burden on multiple aspects of the patient's' physical, psychosocial, and emotional functioning, as evidenced throughout the literature. However, protective factors, such as perceived caregiver and social support, are seemingly protective factors for QOL depressive symptoms.

Intimacy. Unfortunately, ALS patients' intimacy and QOL remains largely unstudied. Intimacy is more commonly associated with constructs of sexuality and physical sexual relationships, but it is actually a much more complex aspect of people's lives (Amyotrophic Lateral Sclerosis Multidisciplinary Clinic, 2009). In studying the QOL of ALS patients, factors such as emotional support and relationships also look to incorporate aspects of intimacy. The ALS–Specific Quality of Life–Revised (ALSSQOL–R) instrument identifies intimacy as one of its domains. The Intimacy domain measures "the experience of, satisfaction with, and desire for, social, emotional, and physical intimacy and sexual intercourse" (Felgoise et al., 2011, p. 7). In a study of ALS patients' marital relationships, findings indicated that the quality of the marital relationship may be an influential factor in psychological status of both the ALS patient and their spouse (caregiver) and that addressing difficulties in the marital relationship is critical to preventing or addressing emotional distress (Atkins, Brown, Leigh, & Goldstein, 2010).

The results of the Atkins et al. (2010) study demonstrated that social connectedness, particularly the importance of intimacy and sexuality, played a role in psychosocial and emotional well-being for the ALS patient. This study raises several issues, such as the patients' level of speech dysfunction and the subsequent impact it has on all aspects of intimacy. However, the study did not discuss the impact of intimacy on

QOL if the patient is not married. If speech is compromised, and the ALS patient is not married, how does intimacy still act as a maintaining factor of emotional well-being?

In patients with ALS, the nature of the disease may impact sexual functioning. Impaired physical functioning affects the voluntary muscles that enhance sexual experiences and weaken respiratory functioning, which makes sexual activity a strain. Patients with ALS often experience fatigue. Patients reportedly experience anxiety issues due to concerns with self-esteem and decreased sexual confidence that may reduce sexual functioning and decrease intimacy (ALSMC, 2009). Some patients with ALS indicated that decreased sexual intimacy is associated with negative body changes, which disrupts their self-perception. These perceptions often increase sexual passivity. However, several studies found that for patients with ALS, intimacy is maintained, regardless of physical functioning, and that sexuality is important in the ALS population (Atkins et al., 2010; Rodriguez, 2010; Wasner, Bold, Vollmer, & Borasio, 2004).

In a qualitative study of terminally ill cancer patients, Lemieux, Kaiser, Pereira, and Meadows (2004) showed that patients emphasized the importance of emotional connectedness through an alternative means of sexual expression, rather than intercourse. Gulledge, Gulledge, and Stahmann (2003) surveyed healthy men and women with regards to romantic physical affection type and relationship satisfaction. The results revealed that those married dyads who reported engaging in physically affectionate behaviors more frequently had more satisfying marriages than those married dyads who did not engage in physically affectionate behaviors. These displays of physical affection included holding hands, giving massages, hugging, kissing, and/or physical proximity. Therefore, displays of physical affection and relationship satisfaction are important

considerations when working with ALS patients and their partners, despite the decline in physical functioning. However, the extent to which bulbar impairment may impact patients' and caregivers' display or avoidance of intimacy is unknown.

Psychosocial and emotional functioning.

Impairments caused by ALS also have significant effects on patients' psychosocial and emotional functioning. In fact, psychosocial and emotional functioning appears to have a reciprocal relationship with physical functioning. Symptoms such as increased limb weakness, breathing difficulties, and bulbar dysfunction subsequently lead to significant changes in these areas of functioning. Impairment caused by the course of the disease usually requires increased dependency on others and substantial lifestyle changes. As such, the complexity of the disease creates a dynamic shift in familial and social relationships (Matuz et al., 2010).

Despite the progressive impairment of ALS, research has repeatedly shown that patients largely maintain their psychosocial and emotional health status. However, earlier findings tended to minimize the multiple psychosocial stressors and emotional challenges of living with ALS and described the coping abilities of the ALS patient as "heroically stoic," even in the face of imminent death (McLeod & Clarke, 2007, p. 5). This inaccurate conceptualization of patients seemingly mitigated the impact of the disease on psychosocial and emotional functioning, while also minimizing the true resiliency of ALS patients.

A study by Trail et al. (2004) identified ALS patients' greatest stressors and concerns impacting QOL across the domains of existential, psychosocial, and physical functioning. Findings were not significant across domains. However, significant

differences were observed at the individual domain levels for the following items: illness progression (existential), problems with speaking (physical functioning), and not being able to work (psychosocial level). Specifically, each patient identified concerns with illness progression and problems with speaking. Therefore, as functional impairment, particularly speech, worsens, perceptions of the impairments as they relate to QOL factors would likely be affected.

Negative emotion. Despite the devastating symptoms of ALS, prevalence of clinical depression in patients remains significantly low (Rabkin et al., 2005), but still higher than the general healthy population (Kurt, Nijboer, Matuz, & Kübler, 2007; Rabkin et al.). Others have argued that because self-report measures tend to emphasize physical symptoms such as fatigue, loss of appetite, and disturbed sleep, this may confound estimates of depression (Gibbons et al., 2011).

Therefore, it is vital to accurately identify patients with psychological morbidity because of the potential implications for improved prognosis with treatment (Felgoise et al., 2010; Pagnini et al., 2014). Clinically, psychological distress and morbidity are broadly defined constructs that include depression, hopelessness, hostility, and paranoia. However, in the ALS literature, psychological distress has largely focused on depression, anxiety, and hopelessness (Felgoise et al.). The relationship between psychological morbidity and bulbar function, specifically, has been largely unstudied.

Felgoise and colleagues found depression is the most studied aspect of psychological morbidity in ALS patients, although studies fail to identify clinical levels of depression. The depressive symptoms reported by and observed in ALS patients confounded their perception of illness, loss of favorable activities and independence

(Felgoise et al.). In addition to changes in emotional functioning, studies have shown that patients with ALS demonstrate certain changes in behavior, such as lack of inhibition, irritability, emotional blunting, lack of empathy, and apathy (Lillo, Mioshi, Zoing, Kiernan, & Hodges, 2011).

Therefore, it is necessary to understand clinical depression separate from adjustment to illness or poor QOL, as these challenges impact treatment delivery and patient care differently. The ALSSQOL–R Negative Emotion subscale assesses patients' various emotional states that include, but are not limited to, depression and anxiety experienced by the individual with ALS and pertaining to outlook about the future (Felgoise et al., 2011). Given that (Duff, 2008) found significant differences in QOL as speech impairment changed, expanding knowledge as to how these symptoms may change over time could impact many aspects of the patient's treatment.

Treatment of psychological and emotional needs.

Psychosocial, emotional, and psychological factors clearly play an influential role in the well-being and care of an ALS patient. Providing individualized support and addressing the affective needs of the patient includes psychotherapy and psychopharmacology. In the clinic setting, it is important for the treatment team to identify and address the needs of those patients presenting with emotional distress. Mental health professionals and other team members working with patients and family help identify problem-solving techniques in an effort to reduce emotional distress (Felgoise et al., 2010).

Research on empirically supported psychological treatments for patients with ALS is limited. There are some recommendations for the management of psychological

symptoms with medications. A review of the literature suggests tricyclic antidepressants and selective serotonin reuptake inhibitors are the drugs of choice (Gordon, 2013; Kurt et. al, 2007) for pharmacological treatment. However, other researchers offer a treatment approach that includes antidepressant medication in concert with professional counseling and the identification of community supports, such as social or religious support outlets (Simmons, 2005). Identification and treatment of patients' psychological and emotional needs is critical for maintaining QOL over time.

Quality of life.

Consistent with the biopsychosocial model of health, QOL helps to explain and measure an individual's perception of him or herself in relationship to the environment. The constructs of QOL demonstrate effects on health, either physically or mentally (CDC, 2011).

The World Health Organization (WHO) 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." This is a wide-ranging construct affected in a multifaceted manner by the person's physical health, psychological state, social relationships, personal beliefs, degrees of independence, and their relationship to significant features of the environment (WHO, 1997).

The definition provided by WHO reflects the increased interest in understanding mind-body relations. For ALS patients, QOL is determined by a comprehensive collection of factors and is maintained as patients' physical functioning declines (Robbins, Simmons, Bremer, Walsh, & Fischer, 2001; Walsh, Bremer, Felgoise, & Simmons, 2003). However, as Duff (2008) noted, previous research has evaluated

physical functioning only as an aggregate. The present study evaluates the effects of speech impairment on aspects of ALS patients' QOL, such as negative emotion, interaction with people and the environment, intimacy, and bulbar functioning, over time.

Assessment and measurement of QOL. Assessment of QOL is measured through a collection of items, scales, domains, and instruments. QOL measures can assess health either generally or specifically relative to a disease (McDowell, 2006). It is suggested that individuals be given the opportunity to report a rating of their self-perceived QOL. This global rating reflects disparate values and preferences of individual patients because QOL is something that is perceived by each patient independently. Elaborations on physical functioning constructs have tried to distinguish between physical functioning and health-related QOL, which reflects a patient's health status or physical health (McDowell). Instruments that focus on physical health, for example the SF-36, measure physical symptoms relative to overall functional ability, while also taking into account other nonmedical factors (Epton, Harris, & Jenkinson, 2009). In patients with ALS, many measures have been studied in order to determine which best assesses QOL. Epton and colleagues suggested that the measures were often heavily weighted towards physical functioning and failed to assess other important QOL constructs or were not useful in large samples (Epton et al.). Based on data from the McGill Quality of Life questionnaire, the ALSSQOL–R was developed to assess QOL across large samples in the following domains: Negative Emotion, Interaction with People and the Environment, Intimacy, Religiosity, Physical Symptoms, and Bulbar Function (Felgoise et al., 2011) The ALSSQOL–R was used as one of the measures in this study and will be discussed more extensively in the methods section.

Bulbar dysfunction.

As previously mentioned, *bulbar* is a term referring to the motor neurons located in the bulb region of the brainstem that are responsible for controlling the muscles of chewing, swallowing, speaking, and the ability to maintain an open airway. In patients with ALS, the uncontrolled increase in muscle tone creates spasticity of the facial muscles, causing bulbar and respiratory impairment. Bulbar symptoms at ALS onset are observed in up to 30% of patients (Kühnlein et al., 2008), and present most commonly as dysarthria, dysphagia, and sialorrhoea.

Dysarthria, dysphagia, and sialorrhoea. The combination of UMN and LMN dysfunction causes dysarthria, a spastic and flaccid paresis affecting the muscles of the face, tongue, and throat (Kühnlein et al., 2008; Simmons, 2005). Dysarthria is eight times more common in patients with bulbar onset and is experienced by bulbar onset patients with greater severity. Nonetheless, more than 80% of ALS patients, regardless of subtype, experience dysarthria during the course of the disease (Tomik & Guiloff, 2010).

Dysphagia, or difficulty swallowing, results from inverted cranial nerves that cause weakness or spasticity of the facial muscles (Kühnlein et al., 2008). Sialorrhoea occurs because of dysphagia and is defined as the uncontrolled and unintentional loss of saliva from the mouth (Young, Ellis, Johnson, Sathasivam, & Pih, 2011). It occurs in up to 20% of patients with ALS (Gordon, 2011). Sialorrhea creates significant respiratory difficulties for patients, and can be socially debilitating, as well.

Excess secretions from the mouth in concert with decreased bulbar functioning can lead to pneumonia and aspiration (Gordon, 2011). Loss of tone and decreased strength in muscles controlling the lips make it difficult for patients to breathe normally,

forcing them to breathe through their mouth rather than their nose. However, breathing from the mouth leads to thickened oral secretions and increased nasal congestion, which makes patients vulnerable to respiratory infections (Kühnlein et al., 2008). As stated earlier, the cause of death for most ALS patients is respiratory failure (Simmons, 2005).

The course of the disease eventually affects all muscles of the tongue, lips, palate, jaw, pharynx, larynx, and upper trunk. The degeneration of these muscles not only causes communication and respiratory dysfunction, but also compromises a patient's chewing and food transportation abilities (Kühnlein et al., 2008).

The degeneration of the LMN further compounds these issues, subsequently weakening the palate and causing substantial difficulties when attempting to swallow (Gordon, 2011). When the muscles involved in the swallowing process weaken, it affects the ability to maintain an adequate diet. Eating certain foods and drinking certain liquids becomes challenging, especially dry, crumbly solids or thin liquids, which can fall through the lips or back over the tongue into the unprotected airway, potentially causing the patient to choke (Carr-Davis, Blakely-Adams, & Corinbilt, 2007).

ALS patients experience sialorrhoea due to weakened facial muscles, and subsequently experience difficulties swallowing their saliva. This causes respiratory difficulties, such as coughing and choking. Patients additionally experience difficulties keeping saliva in their mouth. This struggle creates embarrassing social situations because they cannot control their excessive drooling (Carr-Davis et al., 2007).

Nutritionists and speech pathologists often administer assessments, recommendations, and treatments to help patients and their caretakers manage swallowing difficulties. Clinicians work in tandem to help ensure proper nutrition, while

also assisting patients to develop a program to compensate for swallowing and speaking challenges (Carr-Davis et al., 2007).

Communication functioning.

Patients with ALS face a variety of communication challenges due to progressive dysarthria as bulbar symptoms increase. Motor weakness and spasticity impact the facial and tongue movements needed for speech. Changes in functioning mainly affect the operations of the tongue that include: reduced range and rate of movement, reduction of tongue strength, smaller vowel space, and flattening of vowel formation and projections. As a result, communication becomes impaired, and ALS patients often experience a decreased ability to participate in social situations (Ball, Beukelman, & Pattee, 2004; Ball, Willis, Beukelman, & Pattee, 2001). These impairments directly reduce QOL in ALS patients (Mitsumoto & Del Bene, 2000; Tomik & Guiloff, 2010). Therefore, it is essential to understand the various aspects of communication, in terms of definitions, meaning, and the multiple modalities people utilize to exchange thoughts and feelings, and how this applies to the context of the ALS patient.

Defining communication. Communication is the act or illustration of conveying (Merriam-Webster Online Dictionary, n.d.). The act of communication is defined further as a process of transmitting information between individuals through a shared system of symbols, signs, or behaviors (Merriam-Webster Online Dictionary, n.d.). Communication is an imperative set of skills that people must learn in order to share ideas, feelings, desires, and needs physical material wants and needs. Communicating emotional expression is equally valuable to the individual.

In vocal communication, one person is required to play the role of the speaker while the other person participates as the listener. The role of the listener is required to distinguish between the different vocal sounds and information. When some sounds are too faint, vibrations occur too quickly, or noises are too small to be heard, a listener is unable to detect these differences. The limits beyond an individual's ability to hear are called thresholds (Miller, 1951). However, when speech is unintelligible, or compromised, it challenges the listener's thresholds. Miller qualitatively describes interrupted speech as seemingly hoarse or husky because the ear of the listener is attempting to reconstruct the fragmented pieces into the original sound. This can be a tedious task for both the speaker and listener, and as speech intelligibility declines, effective communication becomes significantly compromised.

As ALS patients become increasingly dysarthric, their speech intelligibility declines, and patients experience many challenges communicating effectively. Research suggests that when sentence intelligibility scores are greater than 81%, listeners reported that they are able to comprehend the content of the speaker's message; however, when speech intelligibility decreased below 81%, the ability to comprehend the speaker markedly declined (Beukelman & Yorkston, 1989). In regards to social context, speakers with ALS and their frequent listening partner had a high inter-rater reliability (Beukelman & Yorkston, 1989). The study found that the speaker and the listener both rated the speaker's ability for communication effectiveness similarly. The findings were consistent with clinical assessments and anecdotal observations of the subject's family members (Ball, Beukelman, & Pattee, 2004). Effective communication with family and friends likely remains intact despite the person's loss of intelligible speech and speech

rate. Impaired muscle tone of the face also affects ability to create and communicate nonverbal facial expressions.

Nonverbal communication. As previously described, the nature of the disease creates flaccidity and spasticity in the facial muscles that interfere with a person's ability to speak and swallow. This change in muscle tone also hinders ALS patients' ability to communicate nonverbal expressions. Nonverbal communications cue channels such as tone, kinetics (posture/body movements), spatial relations, gestures or facial expressions. The messages sent from the individual are either inadvertent or sometimes deliberate (App, McIntosh, Reed, & Hertenstein, 2011; Duff, 2008).

For ALS patients, nonverbal communication is a challenge and has several limiting factors. For example, patients will often struggle to form a smile and will find it difficult to deliver jokes and sarcasm with the appropriate tone and intent. Nonetheless, research suggests that physical touch supports intimate emotions (App et al., 2011). ALS patients' families and friends would benefit from utilizing nonverbal communication such as touch and presentation of self as effective means of communication to substitute or enhance impaired speech (Duff, 2008).

Conversation in the social context.

Pragmatics. Linguistics is the scientific study of language. Pragmatics is a branch of linguistics and is concerned with the relationship between sentences and the environment in which they occur (Merriam Webster Dictionary, n.d.). In order to understand the meaning of what is said, it is simply not enough to just know the meaning of the words or how they were strung together in a sentence. An individual also needs to know who uttered the sentence and in what context in order to make inferences regarding

what the speaker intended the hearer to understand (Birner, 2013b). Therefore, the study of pragmatics mainly focuses on the meaning, an important consideration when evaluating the conversational exchanges.

Conversational maxims. Consideration of conceptualizing conversational maxims relative to bulbar functioning and quality of life was first introduced by Duff (2008). The process of the conversation follows a series of cooperative efforts and turn-taking exchanges. The conversational process, according to Grice (1967), evolved based on the shared principle of cooperation. Each participant recognizes and has some awareness of a common purpose or set of purposes, or at least a mutually accepted direction; the purpose or direction may either be fixed from the start or may evolve throughout the conversational exchange (Grice). In an effort to provide greater specificity, Grice developed a set of guidelines for participants about the purpose or direction of the talk exchange in which they engage. As an addition to the cooperative principle, Grice put forth 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 refers to the amount of information the speaker gives. This maxim requires that the speaker is informative, sharing information relative to the current purposes of the exchange, and that the speaker's contributions are not overly informative (Birner, 2013a). As communication becomes increasingly impaired for the ALS patient, they might find it difficult to adhere to the maxim of quantity. Communication impairment makes it difficult for the ALS patient to make informative contributions; nonetheless, the ALS patient still maintains the ability to

exchange purposeful and relative information. However, the person with ALS is limited in the amount of information he or she can contribute.

Maxim of quality. The maxim of quality is the rule of giving truthful information. The speaker should not say what he or she believes to be false or what he or she cannot support with adequate evidence (Birner, 2013a). The accuracy of the ALS patient's statements should not be questioned because he or she has impaired speech. The issue of inaccurate interpretation is likely due to misinterpretation by the caregiver (Duff, 2008).

Maxim of relevance. The maxim of relevance requires the relationship between the speaker's current utterance and preceding utterances to be relevant to the current context and situation in which the conversation is taking place. Observation of this maxim allows a person to track meaning throughout extended conversations (Birner, 2013a). However, overt violations of the maxim of relevance can occur for the ALS patient who experiences symptoms of pseudobulbar affect. Laughing or crying are emotions often incongruent with the context of the conversation, and thus a violation of the maxim.

Maxim of manner. The maxim of manner requires the speaker to communicate information with brevity and clarity. It calls for avoiding ambiguities and obscurities when speaking (Birner, 2013a). The maxim of manner poses the most difficulty for adherence by ALS patients because they often have utterances of ambiguity and incomprehensibilities.

Observation of a maxim is defined as obeying the maxim rule; thus, a maxim that is not adhered to is defined as a non observance. If the speaker does not follow the rules of the maxim, the hearer then attempts to decide the speaker's implied meaning. First,

the hearer makes this assumption based on the assumption that both the speaker and hearer are observing the cooperative principle. At times, the speaker might violate a maxim, failing to observe it, but in a discreet manner, with the assumption that the listener will not realize the maxim is being violated. This violation of maxims is intended to mislead the hearer (Birner, 2013b). Speakers might violate a maxim principle intentionally in order to tell a joke or to avoid feelings of discomfort (Duff, 2008).

Birner discussed the concept of *to flout a maxim*, which he explained as the action of openly disregarding or purposefully defying the principle. In fact, the violation is generally so blatant that the listener is expected to be aware of and understand the intention of the violation. However, to flout a maxim for ALS patients with speech impairment is often uncontrollable. Even if an ALS patient intends to defy the maxim in order to tell a joke or imply sarcasm, the results of their delivery can be unpredictable. For example, if someone asks a patient "How are you today?", the patient may reply, "I am great," despite the fact he or she might not actually be feeling great. This provides an example of the speaker (patient) blatantly flouting the maxim of quality (truthfulness) that he or she intended the hearer to understand the meaning of the utterances not directly stated in words (Duff, 2008). Conveying sarcasm is difficult for patients with limited quantity of speech and impaired nonverbal communication.

The nature of conversation.

Some linguists describe participating in conversations as an artful dance. Tannen (1998) states how "people strive to capture the lilt, verbal twist, and the particulate nuance of what someone said in conversation" (p. 631). This artistic description of

conversation highlights how ordinary conversation shares the linguistic strategies often utilized in artful literary pieces.

The act of listening to the speaker in a conversation is not a passive process. Tannen (1998) identified a set of involvement strategies utilized for conversations that literary analysts have independently identified as significant in literary discourse: (a) Repetition in conversations provides a rhythm that helps to establish meaning through a series of contrasts and comparisons, (b) Dialogue, which is representative of the voice, and is responsible for creating rhythm and melodic cadence, as it establishes a dramatic scene for characters to interact with one another, and (c) Details offer added information, allowing the listener to employ their imagination and generate ideas about characters, emotions, and meanings.

However, for an ALS patient with impaired communication, it will be difficult to utilize repetition and dialogue, as the patient will struggle to create a rhythmic flow and express inflection during a conversation. As a result, this might further interfere with the ability to make comparisons or convey meaning to the listener. However, the exchange of meaning also requires consideration for the patient's cultural context.

Conversational style.

Tannen (2009) further noted that different cultural and regional backgrounds employ different linguistic cues to signal meaning, and therefore suggests referring to these cues as conversational styles relative to the values placed on the interactive goals. In the style Tannen refers to as "high-involvement," a person who values this interactive style shows that he or she is a good person by demonstrating connection to others by utilizing a fast rate of speech, short pauses, and paraphrasing to show attention and

enthusiasm. The high-involvement individual is generally described as relatively loud, with quick displays of listenership.

In the style referred to as high-considerateness, an individual demonstrates he or she is a good person by not imposing and utilizes a relatively slower rate of speech, longer pauses, avoids interrupting and overlapping talk, and seemingly understands displays of listenership (Tannen, 2009). Overall, Tannen believes, "conversational style is not something extra, added on like icing on a cake; it is the very stuff which the linguistic cake is made" (p. 251). Nevertheless, the study of pragmatics and the process of conversation are embedded with many intricacies and nuances.

Ventriloquizing. Tannen (2010), uses the word *ventriloquizing* to refer to examples where speakers frame their utterance as representing others' voices as their own. Ventriloquizing serves many purposes in a social context and mostly relates to the study of indirectness, defensiveness, and rapport. However, with regards to ALS patients, ventriloquizing offers an interesting perspective for those with impaired speech. ALS patients experiencing speech dysfunction often need the voice of others to represent their own as the disease progresses. Patients might require augmentative speech devices or caregivers to help support their daily communication needs.

Augmentative and alternative communication (AAC) methods.

Most of the aforementioned losses, such as mobility and autonomy, are inevitable; however, it is possible for a healthcare team to aid in maintaining the communication ability of ALS patients. Research showed that ALS patients who lost their ability to speak continued to report social closeness with another person (Murphy, 2004). Therefore, it is important to consider the abilities and expectations of all of those

involved in communicating with the patient when suggesting appropriate supplemental communication approaches and strategies.

Augmentative and alternative communication (AAC) refers to communication interventions that augment or assist existing speech by acting as an alternative to natural speech (Robarge, 2009). Exploring AAC options should begin when a patient's speaking rate has decreased and his or her speaking is unintelligible (Doyle & Phillips, 2001). Research suggests initiating an AAC intervention when the patient's rate of speech has declined to 50% below normal on reading tasks (Doyle & Phillips; Robarge).

However, the nature of the disease process is one of several factors that influence the acceptance and use of AAC and requires consideration when assessing and making recommendations (Doyle & Phillips, 2001). Acceptance and use of AAC are also influenced by factors such as expectations of the communication partners (Murphy, 2004; Robarge, 2009), the skills, needs, personality characteristics of the ALS patients, and the characteristics of the technology itself (Doyle & Phillips).

Attention to the patient's level of physical functioning is also important. Those patients described as technology savvy tend to recognize the benefits of ACC and tend to show more acceptance of and appreciation for devices with greater technology (Doyle & Phillips, 2001). Generally, clinicians consider two approaches to AAC: low technology and high technology. Low-technology approaches include writing or alphabet boards and other aids that do not require extensive training, whereas high-technology approaches include computer-based devices that involve voice output and computer-generated displays. These devices typically require some type of training in order to develop familiarity for efficient use, but also provide greater accessibility and modifications for

patients once they no longer have mobility in their limbs. Head tracking and eye tracking capabilities are among some of the options available to patients (Robarge, 2009).

In summary, the literature describes how speech dysfunction eventually affects all patients with ALS, regardless of the site of onset. Ball et al. (2003) reported that minimal data has been published on the effect of dysarthria in terms of successful communication in different social settings in patients with ALS. As previously noted, a recent study showed differences in the QOL among the levels of functioning in speech communication for patients with ALS (Duff, 2008). However, the design of that study was cross-sectional. In order to provide a better understanding of how speech impairment affects certain aspects of a patient's QOL as the disease progresses; it would be best to evaluate this information in a longitudinal design. Therefore, the current study used a longitudinal to examine how levels of speech affect certain aspects of QOL in ALS patients over time.

Chapter 2

Hypotheses

The first hypothesis examined the relationship between speech decline and ALS patients' self-reported emotional health and psychological well-being, as defined by the ALSSQOL–R over time.

H_1: As the ability to use speech declines over three time points, they will experience increased emotional and psychological distress, as reflected by a decline in Negative Emotion subscale scores.

The second hypothesis examined the relationship between speech decline and ALS patients' reported experiences interacting with, and willingness to participate in, the social environment, as defined by the ALSSQOL–R over time.

H_2: As the ability to use speech declines over three time points, they will have decreased Interaction with People and the Environment subscale scores.

The third hypothesis examined the relationship between speech decline and ALS patients' desire for intimacy, as defined by the ALSSQOL–R over time.

H_3: As the ability to use speech declines over three time points, Intimacy subscale scores will decrease.

The fourth hypothesis examined the relationship between speech decline ALS patients' perception of bulbar impairment as problematic, as defined by the ALSSQOL–R over time.

H_4: As the ability to use speech declines over three time points, the Bulbar Functioning subscale scores will decrease, as participants will perceive bulbar symptoms as increasingly problematic.

Chapter 3

Methodology

Overview.

This study used archival data from a multidisciplinary clinic in order to gain a better understanding of how speech impairment may impact selected domains of the ALSSQOL–R (Negative Emotion, Interaction with People and the Environment, Intimacy, and Bulbar Functioning) over time. The present study served as a follow-up to a previous cross-sectional study that revealed significant differences in QOL relative to the level of bulbar functioning in patients with ALS (Duff, 2008). The aim of this study was to longitudinally analyze the relationship between certain aspects of QOL, as defined by the ALSSQOL–R, and bulbar functioning, particularly speech impairment. The objective of the analysis was to develop a comprehensive understanding between the bulbar functioning domain, specifically the speech item score measured by the ALSFRS–R, and the relationship with QOL during the progression of the disease.

Design and design justification.

An objective measure of physical functioning completed by a clinic staff member (i.e., nurse or physician) and a self-reported QOL measure were collected at three different clinic visits or time points. A visit was defined as any clinic visit for which both the ALSFRS–R and the ALSSQOL–R were available. Analyzing data in a longitudinal study allows researchers to follow the trajectory of a disease. This offers an insightful analysis given that the data demonstrates the timing of disease onset relative to recent changes in the participant's condition (van Belle, Fischer, Heagerty, & Lumley, 2004).

Findings from longitudinal designs have shown that the collection of subject data from multiple time points is best analyzed with the use of repeated measures (van Belle et al.).

In order to study how the course of speech impairment may affect Negative Emotion, Interaction with People and the Environment, Intimacy, and Bulbar Functioning in patients with ALS, a within-subject repeated measures design was chosen. This type of design provided insight into the relationship between changes in level of speech impairment relative to different aspects of QOL during the trajectory of illness.

Participants.

A power analysis originally indicated 55 participants were needed to achieve significance. However, 52 participants were used in the final data set. Potential participants were those individuals who had received treatment for a diagnosis of ALS and completed the measures of interest to this study as part of their routine clinic visits no later than 2012.

Inclusion criteria.

Data obtained from the ALS clinic included those who met the following criteria: (a) consent to the ALS clinic data repository; (b) age 18 years or older; (c) clinically definite, probable, probable laboratory-supported, or possible ALS and; (d) completed the ALSSQOL–R and ALSFRS–R during three consecutive clinic visits (time points of data), with the most recent time point occurring no later than 2012.

Exclusion criteria.

Potential participants were excluded from the study if the measure of physical functioning indicated normal bulbar functioning. Patients excluded from the study if the bulbar domain on the ALSFRS–R equaled a total score of greater than 9 (minimal bulbar

dysfunction). Other exclusion criteria were: (a) significant cognitive impairment, as judged by the ALS clinic neurologist; (b) co-existing uncontrolled serious medical illnesses (e.g., cancer, heart disease) and; (c) co-existing uncontrolled psychiatric disorders other than depression (e.g., schizophrenia, personality disorders).

Procedures.

The population of interest was identified from the databases of an ALS clinic in central Pennsylvania from June 2014 to February 2015. Information obtained included date of birth, gender, ethnicity, marital status, date of symptom onset, date of diagnosis, history of familial ALS, and ALS Functional Rating Scale–Revised (ALSFRS–R) scores.

The data collection started at the most recent time point (time 3) and went back for two more time points (time 2 and time 1, respectively). The database was sorted, identifying those participants with the most severe impairment or no speech, and selected the subsets from them to ensure there were three retrospective time points.

Bulbar involvement, as reported on the ALSFRS–R, was verified to ensure it did not equal 9 or more for the summed scores on the items of speech, swallowing, and salivation at the final time point. Additional verification ensured that normal functioning (a score of 4) was not endorsed at the final time point.

After participants were identified, a physician reviewed the patient health records to ensure that they meet the criteria for a diagnosis of clinically definite ALS, clinically probable ALS, or clinically probable ALS, laboratory-supported. The patient database was viewed in reverse chronological order and those patients who completed three data points of the included measures were identified. A total of 307 patients were reviewed until the needed number of participants (N = 55) was obtained.

For those patients who met inclusion/exclusion criteria, data was extracted from the repository into a separate file. In addition, data was also collected from those patients who did not met inclusion/exclusion criteria, but were deceased and had not signed the repository registry form. Additional data, including ALSSQOL–R total and factor scores, were added from the ALS clinic database. After originally identifying 63 participants for the study, two were excluded because they did not meet ALS diagnostic criteria. Nine additional participants were excluded for one of the following reasons: updated ALSFRS–R bulbar functioning no longer met inclusion criteria, incomplete or missing measure, and too much time between ALSFRS–R and ALSSQOL–R dates.

Security. Personal health information such as Social Security numbers and home addresses were not collected. Therefore, there was minimal risk of loss of confidential health information for each participant. In regard to data collection, precautionary methods, such as assigning unique code numbers to identify subjects and securing of research documents further reduced risk.

Measures.

ALS Functional Rating Scale–Revised (ALSFRS–R). The Amyotrophic Lateral Sclerosis Functioning Rating Scale (ALSFRS–R) is a well-validated and reliable measure that has strong internal consistency (Cedarbaum & Stambler, 1997; Gordon, 2004). The instrument assesses activities of daily living and monitors functional status (Alkhatib, 2014). The rating scale identifies patients' fine and gross motor skills, as well as bulbar and respiratory functioning (Cedarbaum & Stambler, 1997). The ALSFRS–R also helps clinicians and researchers to monitor disease progression over time (Cedarbaum & Stambler).

The ALSFRS–R includes a total of 12 items that assess level of functioning across the domains of: Bulbar, Gross Motor, Fine Motor, and Respiration. Each item is scored on a 5-point scale from 0 (*unable/dependent*) to 4 (*none/normal*) and then totaled to represent a score between 0 (*poor functioning*) and 48 (*best functioning*). For the purposes of this study, particular attention was given to the bulbar domain that encompasses items on impaired speech, swallowing, and salvation.

ALS–Specific Quality of Life (ALSSQOL–R) Instrument–Revised. The ALS–Specific Quality of Life Instrument–Revised is a validated instrument for measuring overall QOL in individuals diagnosed with ALS. The ALSSQOL–R has shown strong reliability and validity with independent factors that revealed strong internal consistency. The measure consists of 50 total items, 46 scored and 4 non-scored items, and can generally be completed by most individuals in 15-20 minutes. The 46 scored items contribute to each of the six domains that represent the following aspects of QOL: Negative Emotion, Interaction with People and the Environment, Intimacy, Religiosity, Physical Symptoms, and Bulbar Functioning (Felgoise et al., 2011). The ALSSQOL–R produces a single-item score, an average total score, and six subscale scores, with regards to each of the six domains, an average score is calculated, and ranges from 0 (least contribution to QOL) to 10 (greatest contribution to QOL).

Each of the 50 individual items is rated from 0 (*strongly agree*) to 10 (*strongly disagree*) by the patient. The items that comprise the ALSSQOL–R have been shown not to correlate with each other due to the orthogonal varimax rotation principle utilized when constructing the test's original version (Felgoise et al., 2011; Simmons et al., 2006). Several of the items require transposing, subtracting 10 from the response because those

items contain negative statements. Moreover, it is important to note that the Negative Emotion domain has been shown to contribute the most to QOL. Of note, according to the measure's manual, patients who score 6.23 or lower on the Negative Emotion domain warrant further evaluation for depression, anxiety, and possible suicidal tendencies, and likely have distressing concerns about their future (Felgoise et al.).

Statistical analysis.

The ALSFRS–R speech item scores for each time point and the subscale scores from the ALSSQOL–R (Negative Emotion, Interaction with People and the Environment, Intimacy, and Bulbar Functioning) were analyzed using a within-subject repeated measures test for each of the hypotheses, and the ALSFRS–R speech item scores. Post-hoc analyses were conducted and the Average Total ALSSQOL–R, Physical Symptoms, and Religiosity overall scores were extracted and analyzed for the three time points.

Chapter 4

Results

Study demographics.

Descriptive statistics were calculated for all participants within the overall sample (N = 52) and summarized the basic features of the data. The mean age of the sample was 61.52 (SD = 10.63). Means, medians, and frequencies were used to describe the main characteristics of the sample (Table 1). Any variable with missing data was replaced by using series means.

Table 1

Study Demographics

	%	*n*
Gender		
Male	57.7	30
Female	42.3	22
Ethnicity		
Caucasian	94.2	49
African American	3.8	2
Southeast Asian	1.9	1
Marital Status		
Married	76.9	40
Single	15.4	8
Divorced	1.9	1
Widowed	5.8	3
Familial ALS		
No	75.0	39
Yes	9.6	5
Unknown	15.4	8
Site of Symptom Onset		
Limb	46.2	24
Bulbar	28.8	15
Limb, Bulbar	11.5	6
Unknown	13.5	7

Time. Data was collected at three time points. Table 2 shows the mean and standard deviation for this variable. Several participants demonstrated variability in the time between their visits to the clinic (time between time 1 and time 2). The length of time in between time 1 and 2, for one participant was 9-month difference, compared to one participant that had a 12-month (1 year) difference between these two time points. Similarly, this variability in time was also observed between time points 2 and 3. One participant had a one-month difference (less than the average) between these visits, where as another subject had an 11-month difference.

Table 2

Mean Difference in Time Points/Clinic Visits

	M	*SD*	Minimum	Maximum
Difference between time 2 and time 1	3.87	2.28	2	12
Difference between time 3 and time 2	3.73	1.67	2	11
Difference between time 3 and time 1	7.98	2.95	5	18

Note. Time in months.

Estimated time since diagnosis. The estimated time since diagnosis was calculated by examining the difference between date of the most recent ALSFRS–R assessment and date of diagnosis. Generally, date of symptom onset is used to generate this variable, but this date was unavailable for most patients. The estimated time since diagnosis variable had missing values, as the date of diagnosis was not obtainable for

three participants from the ALS database. The missing values were replaced with series means. The average time since diagnosis was 27.24 months (2.27 years) and the standard deviation was 26.80 (2.23 years). The shortest estimated time since diagnosis the sample was 181 months (15.08 years). The longest estimated time since diagnosis was 188 months (15.66 years) and was reported by only one participant.

Time near death. The time near death variable is the difference in time (months) between date of death and the date of the most recent ALSFRS–R assessment (time 3). Table 3 shows the sample size, mean, and standard deviation of participants who passed away after the third time point.

Table 3

Time Between Dates of Death and Completed ALSFRS–R at Time 3

	n	*M*	*SD*
Time near death	23	4.65	2.80

Note. Time in months.

Descriptive statistics of the Amyotrophic Lateral Sclerosis Functional Rating Scale–Revised (ALSFRS–R).

ALSFRS–R data was collected at three time points for all participants. Means, medians, and score minimums and maximums were used to describe physical functioning over time. Results are summarized and outlined in further detail in Tables 4 to 19.

Total ALSFRS–R scores. The average total ALSFRS–R scores ranged from 27.00 to 17.98 over three time points. A decline in mean scores indicated an overall decline in functional impairment over three time points. Table 4 shows the mean, standard deviation, and minimum and maximum scores of participants' average total functioning over time.

Table 4

Total ALSFRS–R Scores (Scale of 0 to 48)

	M	*SD*	Minimum	Maximum
Time 1	27.00	9.54	6.00	44.00
Time 2	22.13	8.91	6.00	40.00
Time 3	17.98	8.06	5.00	41.00

Total bulbar functioning scores. The bulbar functioning domain includes the items speech, swallowing, and salivation and is scored on a total of 12 points. Table 5 shows the mean, standard deviation, and minimum and maximum scores. Over time, mean scores ranged from 4.87 to 2.96, revealing a decline in bulbar functioning. The direction of these scores demonstrated that bulbar functioning has a linear relationship with disease progression. The itemized means, standard deviations, and itemized scores for speech, salivation, and swallowing are provided in Tables 6, 7, and 8.

Table 5

Total Bulbar Functioning Scores (Scale of 0 to 12)

	M	*SD*	Minimum	Maximum
Time 1	6.23	2.91	1.00	12.00
Time 2	5.09	2.84	.00	10.00
Time 3	3.82	2.67	.00	8.00

Speech item scores. Speech is a measurement of the level of speech impairment. Table 6 shows the mean, standard deviation, and minimum and maximum scores. Over time, average speech item scores ranged from 1.00 to 1.75. Results indicated that initially, 28.8% of participants had a detectable speech disturbance, 21.2% had loss of useful speech, and 3.8% of participants still had normal speech processes. At the most recent time point, no participant possessed normal speech processes (0%), and 42.3% of participants had loss of useful speech. This indicates that at time 3, there is limited variability in the range of speech functioning. Examination of these means over time suggests that as the disease progresses, normal speech processes almost diminish entirely.

Table 6

ALSFRS–R Bulbar Domain: Speech

	M	*SD*	Minimum	Maximum
Time 1	1.75	1.20	0	4
Time 2	1.40	1.14	0	4
Time 3	1.00	1.03	0	3

Table 7

ALSFRS–R Bulbar Domain: Salvation

	M	*SD*	Minimum	Maximum
Time 1	2.44	1.26	0	4
Time 2	2.02	1.15	0	4
Time 3	1.54	1.04	0	3

Table 8

ALSFRS–R Bulbar Domain: Swallowing

	M	*SD*	Minimum	Maximum
Time 1	2.04	1.20	0	4
Time 2	1.67	1.31	0	4
Time 3	1.29	1.24	0	3

Table 9

ALSFRS–R Fine Motor Domain: Handwriting

	M	*SD*	Minimum	Maximum
Time 1	2.37	1.299	0	4
Time 2	1.85	1.289	0	4
Time 3	1.42	1.391	0	4

Table 10

ALSFRS–R: Number of Patients With and Without Gastrostomy

	n	%
Time 1		
With	13	25.0
Without	39	75.0
Time 2		
With	20	38.5
Without	32	61.5
Time 3		
With	27	51.9
Without	25	48.1

Table 11

ALSFRS–R Fine Motor Domain: Cutting Food and Handling Utensils in Patients Without a Gastrostomy

	M	*SD*	Minimum	Maximum
Time 1	1.95	1.47	0	4
Time 2	1.41	1.27	0	4
Time 3	.92	1.22	0	4

Table 12

ALSFRS–R Fine Motor Domain: Cutting Food and Handling Utensils in Patients With Gastrostomy

	M	*SD*	Minimum	Minimum
Time 1	1.15	1.07	0	4
Time 2	1.10	1.25	0	4
Time 3	.81	1.24	0	4

Table 13

ALSFRS–R Gross Motor Domain: Dressing and Hygiene

	M	*SD*	Minimum	Minimum
Time 1	1.87	1.36	0	4
Time 2	1.38	1.29	0	4
Time 3	.98	1.18	0	4

Table 14

ALSFRS–R Gross Motor Domain: Turning In Bed and Adjusting Bed Clothes

	M	*SD*	Minimum	Maximum
Time 1	2.17	1.48	0	4
Time 2	1.62	1.40	0	4
Time 3	1.25	1.28	0	4

Table 15

ALSFRS–R Gross Motor Domain: Walking

	M	*SD*	Minimum	Maximum
Time 1	2.21	1.94	0	4
Time 2	1.65	1.23	0	4
Time 3	1.42	1.14	0	4

Table 16

ALSFRS–R Gross Motor Domain: Climbing Stairs

	M	*SD*	Minimum	Maximum
Time 1	1.37	1.51	0	4
Time 2	1.15	1.41	0	4
Time 3	.65	1.14	0	4

Table 17

ALSFRS–R Respiratory Function Domain: Dyspnea

	M	*SD*	Minimum	Maximum
Time 1	2.69	1.25	0	4
Time 2	2.44	1.24	0	4
Time 3	2.35	1.27	0	4

Table 18

ALSFRS–R Respiratory Function Domain: Orthopnea

	M	*SD*	Minimum	Maximum
Time 1	3.02	1.55	0	4
Time 2	2.56	1.60	0	4
Time 3	2.27	1.68	0	4

Table 19

ALSFRS–R Respiratory Function Domain: Respiratory Insufficiency

	M	*SD*	Minimum	Maximum
Time 1	3.33	1.02	1	4
Time 2	3.10	1.14	1	4
Time 3	2.94	1.13	1	4

Descriptive statistics for the Amyotrophic Lateral Sclerosis Quality of Life Scale– Revised (ALSSQOL–R).

ALSSQOL–R data were collected at three time points for all participants. Means, medians, and score minimums and maximums were used to describe the scores for the subscales of the ALSSQOL–R over time. Results are summarized in Tables 20 to 26.

Average Total ALSSQOL–R. The Average Total ALSSQOL–R score represents an individual's self-reported overall QOL. Table 20 shows the mean, standard deviation, and minimum and maximum scores for the Average Total ALSSQOL–R scores at each time of the three points. Mean scores ranged from 6.66 to 6.32, indicating a decline in total QOL. These scores suggest that over time, QOL is lower than the normative mean (*M* = 6.83; *SD* = 1.2*)*.

Table 20

Average ALSSQOL–R Total Score

	M	*SD*	Minimum	Maximum
Time 1	6.66	1.12	4.14	9.60
Time 2	6.61	1.21	3.72	9.57
Time 3	6.32	1.12	3.17	9.36

Negative Emotion. The Negative Emotion domain score measures various self-reported emotional states, including but not limited to anxiety and depression, and how patients with ALS feel about their future (Felgoise et al., 2011). Table 21 shows the mean, standard deviation, and minimum and maximum scores. Mean scores ranged from 6.60 to 6.35 revealing a minimal decline. Compared to the normative sample (*M* = 6.67; *SD* = 2.0), these scores suggested that study participants endorsed their emotional states and future outlook over time similarly. As previously mentioning, the

ALSSQOL–R manual suggests that those patients with scores 6.23 or lower on this domain receive further evaluation for depression, suicide, anxiety, worries about their future, and issues related to coping (Felgoise et al.). The average scores over the three time points were never below the 6.23 score threshold warranting additional concern for psychosocial evaluation.

Table 21

ALSSQOL–R Negative Emotion

	M	*SD*	Minimum	Maximum
Time 1	6.60	1.73	.92	10.00
Time 2	6.65	2.07	.00	10.00
Time 3	6.35	1.78	.00	9.58

Interaction with People and the Environment. The Interaction with People and the Environment domain score describes self-reported perception and response to friends and family, and how patients with ALS participate in their environment over time (Felgoise et al., 2010). Table 22 shows the mean, standard deviation, and minimum and maximum scores. Mean scores ranged from 8.02 to 7.68, demonstrating a minimal decline. Compared to the normative sample (M = 8.17; SD = 1.5), these scores revealed that over time participants perceived their social support, availability of social support, and interest in engaging in their environment somewhat similarly. Therefore, these

scores suggest that participants do not perceive and respond to social supports or rate their level of interest in participation in social activities as a significant concern.

Table 22

ALSSQOL–R Interaction with People and the Environment

	M	*SD*	Minimum	Maximum
Time 1	8.02	1.32	4.45	10.00
Time 2	7.76	1.96	.00	10.00
Time 3	7.68	1.90	.00	10.00

Intimacy. The Intimacy domain score assesses experience, satisfaction, and desire for intimacy in the areas of social, emotional, and physical intimacy, and sexual intercourse (Felgoise et al., 2011) over time. Table 23 shows the mean, standard deviation, and minimum and maximum scores. Mean scores ranged from 5.38 to 5.49, demonstrating a nominal increase in these scores. Compared to the normative sample (M = 5.85; SD = 2.1), these scores indicated that participants self-report a similar pattern of desired intimacy. Results indicated that as the disease progressed and functional impairment declined, and participants need for intimacy increased.

Table 23

ALSSQOL–R Intimacy

	M	*SD*	Minimum	Maximum
Time 1	5.38	2.20	.00	10.00
Time 2	5.46	2.53	.00	10.00
Time 3	5.49	2.49	.00	10.00

Religiosity. The Religiosity domain describes self-reported religious identity. The domain score represents how individuals use religion as a source of comfort, their use of prayer, and their participation in religious practices at home (Felgoise et al., 2011) over time. Table 24 shows the mean, standard deviation, and minimum and maximum scores. Mean scores ranged from 6.86 to 6.95, showing an increase in Religiosity domain scores. Compared to the normative sample (M = 6.58; SD = 3.4), these scores indicate that participants' identification of religiousness is relatively average. For this domain, the ALSSQOL–R manual emphasizes that low scores do not indicate concerns, and high scores do not suggest strengths. Evaluating Religiosity scores at the individual level in the context of Negative Emotion scores is advised (Felgoise et al., 2010).

Table 24

ALSSQOL–R Religiosity

	M	*SD*	Minimum	Maximum
Time 1	6.86	3.215	.00	10.00
Time 2	6.73	3.421	.00	10.00
Time 3	6.95	3.330	.00	10.00

Physical Symptoms. The Physical Symptoms domain score measures the degree to which participants endorse the following physical symptoms as problematic over time: ability to move, sleep, feeling physically terrible, fatigue, pain, and bowel and bladder function (Felgoise et al., 2011). Table 25 shows the mean, standard deviation, and minimum and maximum scores. Mean scores ranged from 6.61 to 5.75, revealing a decline for this subscale. These scores are comparable to those of the normative sample (M = 6.70; SD = 1.8). Participant's scores declined over time, suggesting that with disease progression and increased functional impairment, patients perceive experiencing physical symptoms as increasingly concerning.

Table 25

ALSSQOL–R Physical Symptoms

	M	*SD*	Minimum	Maximum
Time 1	6.61	1.49	3.67	9.33
Time 2	6.32	1.50	2.50	9.17
Time 3	5.75	1.52	2.83	10.00

Bulbar Functioning. The Bulbar Functioning domain score assesses the extent to which participants self-report the following bulbar symptoms as problematic over time: speaking, saliva, communication problems, mucus, and eating (Felgoise et al., 2011). Table 26 shows the mean, standard deviation, and minimum and maximum scores. Mean scores ranged from 5.05 to 3.74. Compared to the normative sample (M = 7.26; SD = 2.4), these scores indicate that participants perceived problems with regard to bulbar functioning as a concern, especially over the trajectory of the disease.

Table 26

ALSSQOL–R Bulbar Functioning

	M	*SD*	Minimum	Maximum
Time 1	5.05	2.13	1.00	8.75
Time 2	4.63	2.09	.40	9.00
Time 3	3.74	2.13	.00	8.40

Correlations.

Pearson product-moment correlations were conducted to investigate the relationship between the ALSFRS–R speech item score and each of the ALSSQOL–R subscales at each of the three time points. No significant correlations were found between speech impairment (times 1, 2, or 3) and the ALSSQOL–R subscales other than Bulbar Functioning.

Correlational analyses evaluated the relationship between speech impairment at time 1 and Bulbar Functioning, as defined by the ALSSQOL–R at each time point. Speech impairment at time 1 showed a statistically significant and strong correlation with Bulbar Functioning at time 1, $r(50) = .585$, $p < .001$. Similarly, speech impairment at time 2 was strongly correlated with Bulbar Functioning at time 2, $r(50) = .491$, $p < .001$. The relationship between speech impairment at time 3 and Bulbar Functioning at time 3 revealed a weak correlation, $r(50) = .244$, $p = .08$. Given that there is minimal variability in speech functioning (Table 6) at time 3, this provides some explanation for the

weakening of the correlation at this time point. Additionally, the relationship between actual functioning and perceived functioning at time 3 suggests that the two variables do not decline systematically. Despite the significant loss of speech at the third time point, participants likely have accepted and adapted to this loss. Findings suggest that as patients with ALS experience loss of functional speech, they no longer perceive this loss as problematic. Given that patients with no or minimal speech have likely been forced to adapt and identify alternative means of expressing speech.

Repeated measures analyses.

A within-subjects repeated measures test was conducted with three time points for each of the dependent variables. The repeated measures test analyzed change in the speech item score, Negative Emotion, Interaction with People and the Environment, Intimacy, and Bulbar Functioning in patients with ALS. An alpha level of .05 was used for all repeated measures analyses and the Bonferroni correction was used to adjust for multiple comparisons. Post hoc analyses were conducted to identify differences in Average Total QOL, Religiosity, and Physical Symptoms at the three clinic visits.

Negative Emotion. A repeated measures analysis test was used to determine if Negative Emotion scores changed over time. For this analysis, Mauchly's test of sphericity indicated the assumption of sphericity had not been violated, $\chi^2(2) = .561$, $p = .75$. Results did not indicate any significant differences in the mean Negative Emotion scores over time, $F(2, 102) = .987$, $p = .38$, partial $\eta^2 = .019$. The means and standard deviations for this domain from time 1 to time 3 are presented in Table 21. Negative Emotion mean scores had a slight increase from time 1 to time 2, followed by a decline from time 2 to time 3. Although an examination of the means from

each time point failed to show statistical significance. Similarly, polynomial contrasts did not indicate a significant linear trend, $F(1, 51) = 1.33$, $p = .25$, partial $\eta^2 = .025$, nor a significant quadratic trend, $F(1, 51) = .70$, $p = .40$, partial $\eta^2 = .01$. Figure 1 shows the direction of mean scores for the Negative Emotion domain over time. Given that there was no statistically significant difference between the means at the three time points ($p > .05$), the null hypothesis cannot be rejected. Thus, over time participants' self-reported emotional health and psychological well-being does not significantly change.

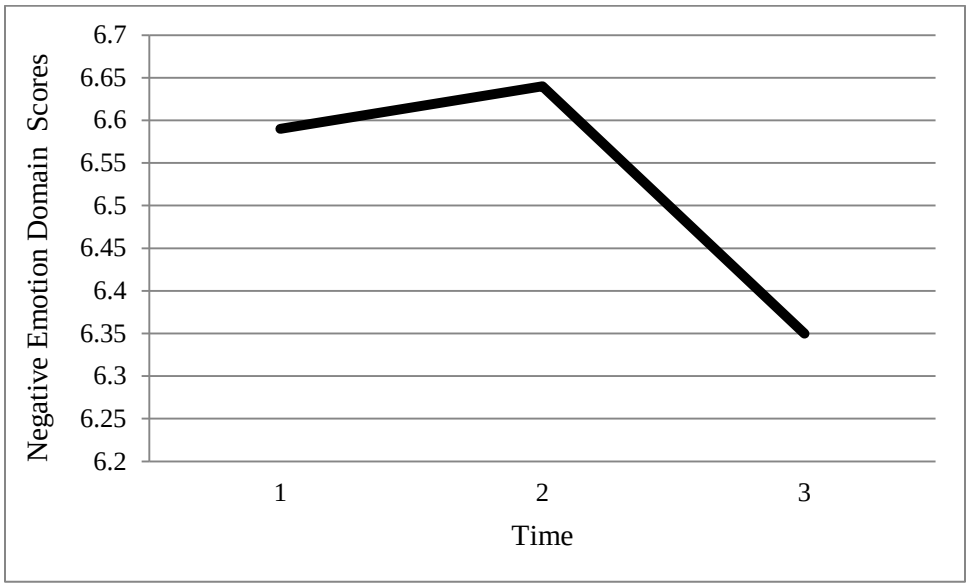

Figure 1. ALSSQOL–R: Negative Emotion domain scores over time. Line graph displaying the change in mean subscale scores for the Negative Emotion domain over three time points (clinic visits) and shows an initial increase and then small decrease in scores.

Interaction with People and the Environment. In order to assess change in self-reported scores of Interaction with People and the Environment over time, a repeated measures analysis test was conducted. The assumption of sphericity was met, as indicated by Mauchly's test of sphericity $\chi^2(2) = .285$, $p = .86$. No statistically significant changes were observed across the three time points for this domain, $F(2, 102) = 1.32$, $p = .21$, partial $\eta^2 = .02$. The means and standard deviations for each of the three time points are presented in Table 22. Examining the mean scores from time 1 to time 2 revealed no significant decrease, and the decline in mean scores from time 2 to time 3 is also not significant. Figure 2 shows the direction of mean scores over time for this domain. Polynomial contrasts did not reveal any statistically significant trends. Given that there was no statistically significant difference between the means at the three time points ($p > .05$), the null hypothesis cannot be rejected. Therefore, as the diseases progresses over time, participants' experiences with and willingness to engage in the social environment does not significantly change.

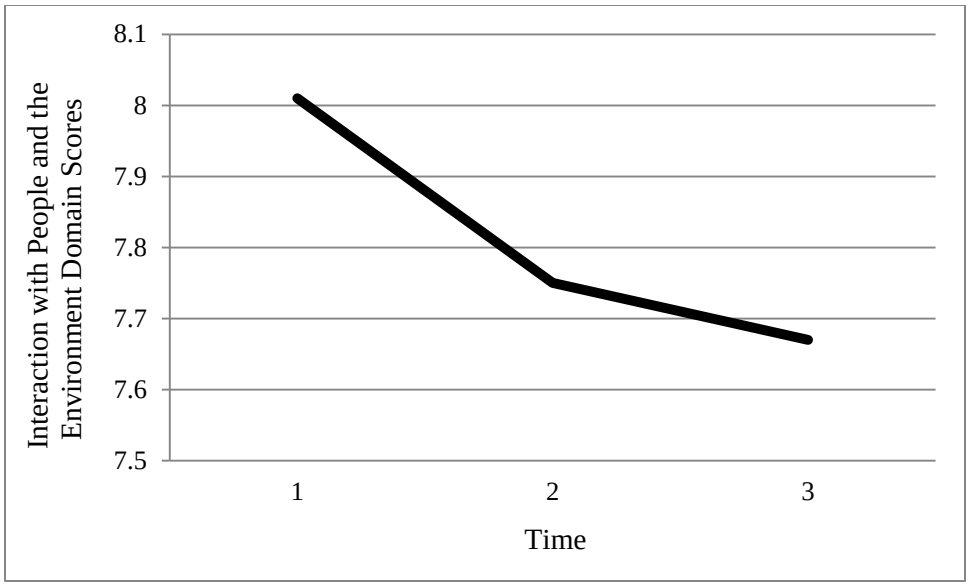

Figure 2. ALSSQOL–R: Interaction with People and the Environment domain scores over time. Line graph displaying a small decline in the subscale scores for the Interaction with People and the Environment domain over three time points (clinic visits).

Intimacy. Conducting a repeated measures test assessed change in the average Intimacy subscale scores over time. Mauchly's test of sphericity indicated the assumption of sphericity was not violated, $\chi^2(2) = 1.440$, $p = .41$. Findings suggested no significant differences in self-reported Intimacy over time, $F(2, 102) = .022$, $p = .93$, partial $\eta^2 = .001$. The means and standard deviations for the three time points are presented in Table 23. Although the changes in the Intimacy scores were not significant, the results suggested that despite loss in functional impairment, participants reported a greater desire for intimacy. No significant differences were observed from time 1 to time 2 or time 2 to time 3. Figure 3 shows the direction of mean scores for the Intimacy domain over time. According to polynomial contrasts, the direction of scores failed to reveal any significant trends. Given that there were no

statistically significant differences between the means over time ($p > .05$), the null hypothesis cannot be rejected. Thus, participants' desire for intimacy does not significantly change over time.

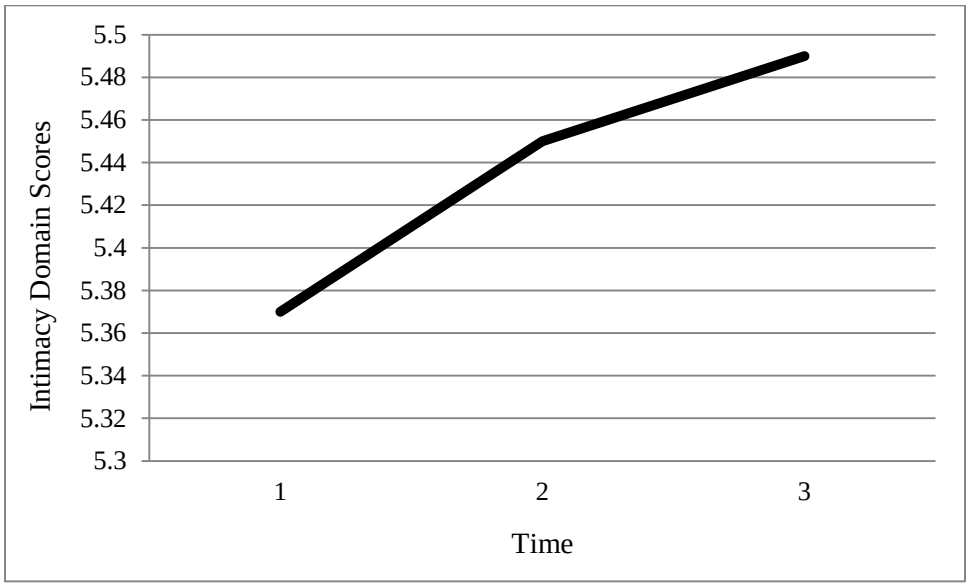

Figure 3. ALSSQOL–R: Intimacy domain scores over time. Line graph displaying a nominal increase in subscale scores for the Intimacy domain over three time points (clinic visits).

Bulbar Functioning. A repeated measures analysis of the ALSSQOL–R Bulbar Functioning domain was conducted in order to assess differences self-perceived functioning at three different time points. For this analysis, Mauchly's test of sphericity indicated the assumption of sphericity was met, $\chi^2(2) = 4.66$, $p = .097$. Results revealed that participants' perception of their bulbar functioning changed over time, $F(2, 102) = 13.25$, $p < .001$ partial $\eta^2 = .21$. The average scores and standard deviations for Bulbar Functioning, as defined by the ALSSQOL–R are presented in Table 26.

Examination of theses means suggests that participants rated their bulbar functioning as increasingly troublesome over time. Figure 4 depicts the direction of mean scores for the Bulbar Functioning domain over time. Statistically significant differences were found between the average scores for time 1 and time 3, $p < .001$, and also found between the average scores for time 2 and time 3, $p = .006$. These results demonstrate that as the impairment worsens over time (from time 2 to time 3), participants perceive the impact of symptoms as more problematic. Polynomial contrasts supported this, indicating a significant linear trend, $F(1, 51) = 21.31$, $p < .001$, partial $\eta^2 = .30$. Therefore, over time, participants' perception of their bulbar impairment as problematic changes significantly.

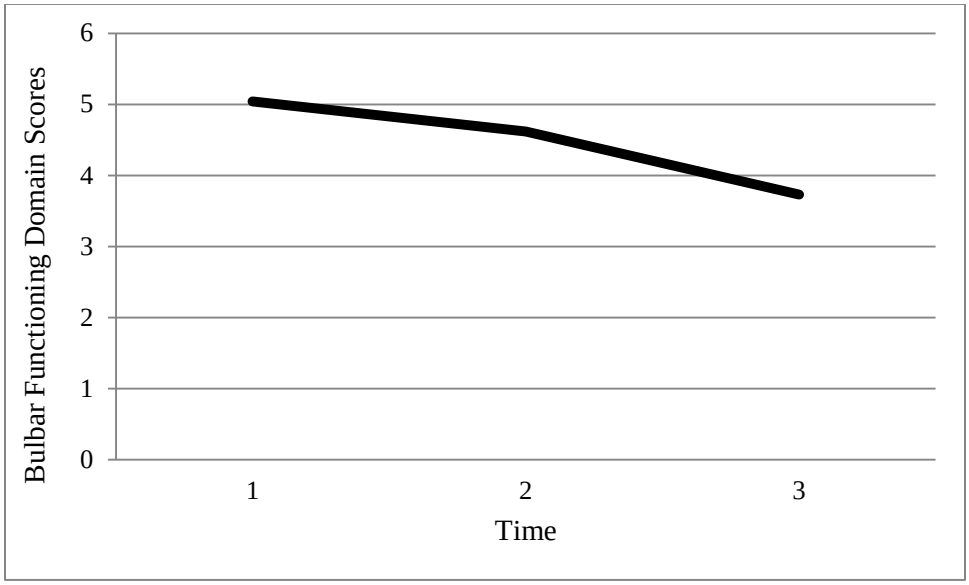

Figure 4. ALSSQOL–R: Bulbar Functioning domain scores over time. Line graph displaying a significant decline in the subscale scores for the Bulbar Functioning domain over three time points (clinic visits), as the line depicts a significant linear trend.

ALSFRS–R: speech item scores. A repeated measures analysis of the ALSFRS–R speech item scores, with Huynh-Feldt correction, was conducted to assess differences in the ability to speak at three different time points. Mauchly's test of sphericity was significant, violating the assumption of sphericity, $\chi^2(2) = 9.7295$, $p < .05$, and as such degrees of freedom were corrected accordingly. Results indicated that speech significantly declined over time, $F(1.75, 89) = 22.86$, $p < .05$, partial $\eta^2 = .31$. Examination of means revealed that the ability to speak declines significantly from time 1 to time 2, $p = .006$, and continued to significantly decline from time 2 to time 3, $p < .001$. Polynomial contrasts indicated a significant linear trend in a negative direction, $p < .001$. The means and standard deviations of speech over time are presented in Table 6. Figure 5 depicts the direction of mean scores for the ALSFRS–R: speech item score over time in relationship to the change in QOL mean scores. The findings from the repeated measures analysis of the Bulbar Functioning domain suggested that ALS patients endorse symptoms of functional impairment as more problematic than concerns related to their psychological and emotional well-being.

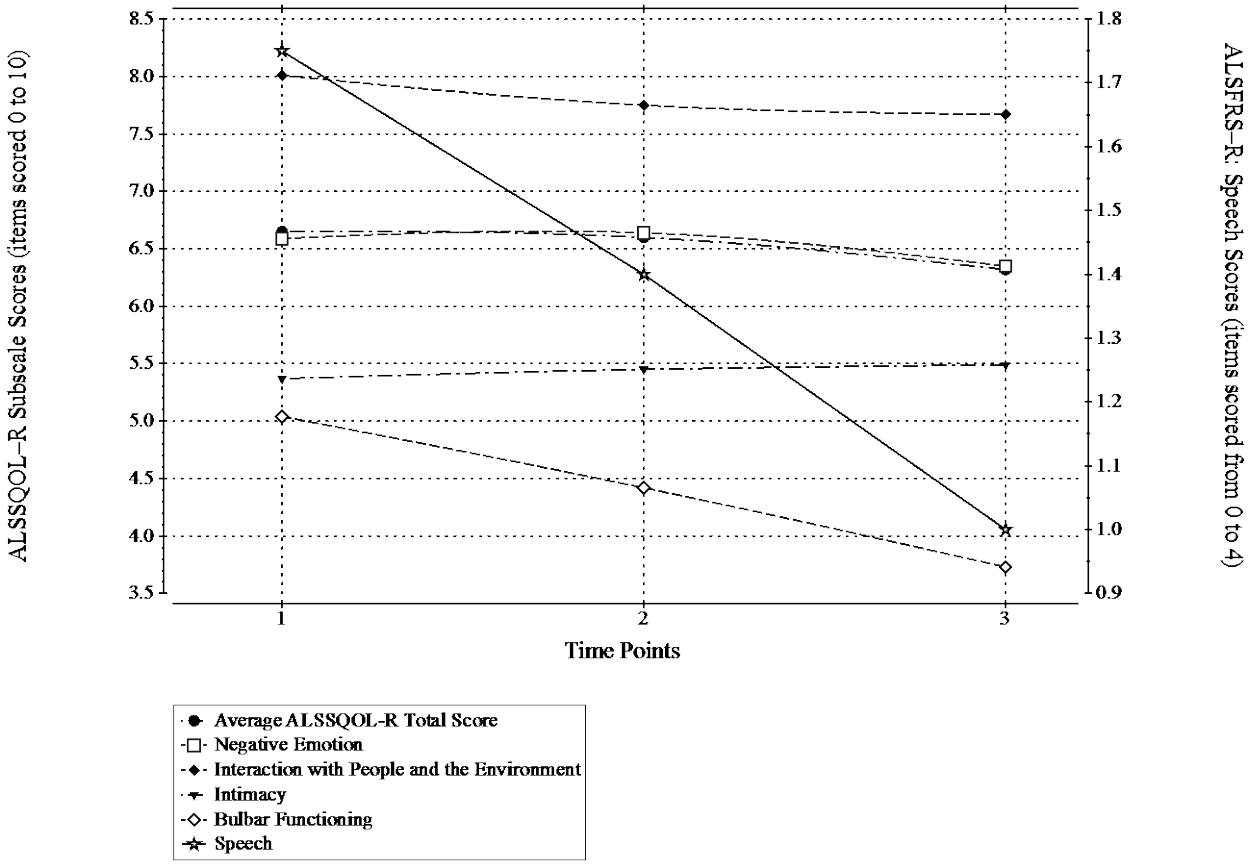

Figure 5. Selected ALSSQOL–R domain scores verses the ALSFRS–R: speech item scores over time. This graph shows the change in the selected subtest scores from the ALSSQOL–R and from the ALSFRS–R speech item scores over three time points (clinic visits).

Post hoc analyses.

In an effort to further understand how speech impairment impacts all aspects of QOL, including a patient's total QOL score over time, post hoc analyses of the remaining ALSSQOL–R subscales were conducted.

Physical Symptoms. A repeated measures analysis of the Physical Functioning domain was conducted in order to measure differences between how participants perceived physical symptoms over time. Mauchly's test of sphericity indicated the assumption of sphericity was met, $\chi^2(2) = 4.171$, $p = .124$. Results found that self-reported Physical Symptom scores significantly changed over time, $F(2, 102) = 8.767$, $p < .001$, partial $\eta^2 = .15$. The means and standard deviations are presented in Table 25. Examination of these means revealed a significant decline from time 1 to time 3, $p = .002$. From time 1 to time 2, the decline is not significant, although from time 2 to time 3, there is a continuous decline in scores that is statistically significant, $p = .002$. Polynomial contrasts supports this directionality of scores, as a significant linear trend is observed, $F(1, 51) = 13.371$, $p < .001$, partial $\eta^2 = .21$. Over time, participants perceive their physical symptoms as problematic.

Religiosity. A repeated measures analysis of the Religiosity domain was conducted in order to examine differences in participants' identification of religion and use of religion over time. For this analysis, Mauchly's test of sphericity indicated the assumption of sphericity was met, $\chi^2(2) = 4.535$, $p = .104$. Findings revealed no significant differences in Religiosity domain scores over time, $F(2, 102) = .161$, $p = .85$, partial $\eta^2 = .003$. The means and standard deviations are presented in Table 24. An examination of means and standard deviations for the Religiosity domain between each of the time points did not reveal any significant findings. Polynomial contrasts did not suggest a significant linear trend, $F(1, 51) = .054$, $p = .818$, partial $\eta^2 = .001$, nor a significant quadratic trend,

$F(1, 51) = .268$, $p = .60$, partial $\eta^2 = .005$. Over time, identification of religiosity does not change.

Average Total ALSSQOL–R. In order to assess differences in total quality of life scores at three points, a repeated measures test was conducted. Mauchly's test of sphericity indicated the assumption of sphericity was met, $\chi^2(2) = 1.946$, $p = .37$. Results revealed significant differences in Average Total ALSSQOL–R scores over time, $F(2, 102) = 5.37$, $p < .05$, partial $\eta^2 = .10$. The average scores and standard deviations for the Average Total ALSSQOL–R mean scores over time are presented in Table 20. Examination of these means from time 1 to time 3 revealed a significant decline in Average Total ALSSQOL–R scores, $p = .02$. No significant change was observed from time 1 to time 2, but the decline in average scores from time 2 to time 3 indicated a significant change, $p = .04$. Polynomial contrasts support this, as a significant linear trend is observed, $F(1, 51) = 7.96$, $p = .007$, partial $\eta^2 = .13$. Figure 6 shows the direction of the Average Total ALSSQOL–R scores over time. Average QOL declined over time, although decreased QOL scores is likely due the statistically significantly change in perception of Physical Symptoms and Bulbar Functioning as problematic.

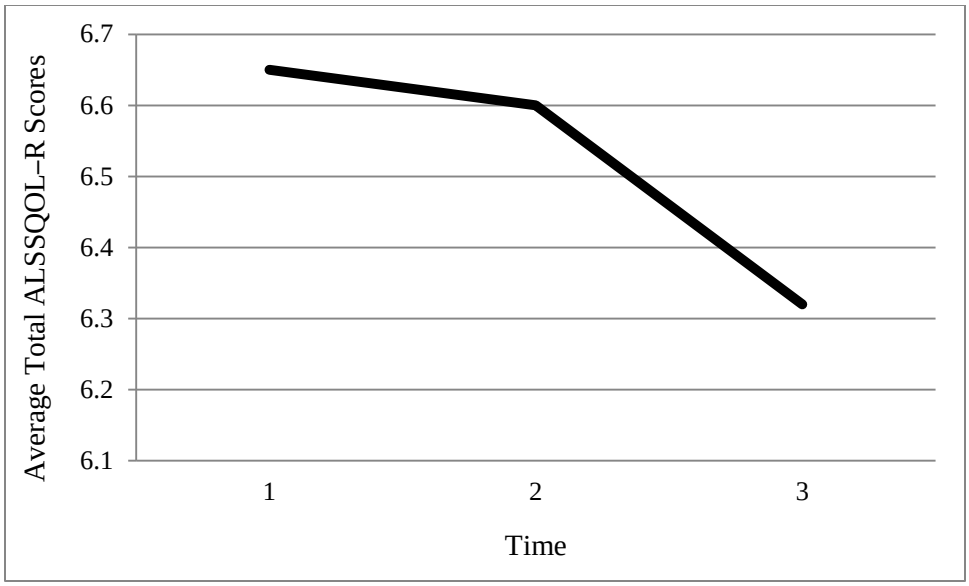

Figure 6. Average Total ALSSQOL–R scores over time. This line graph depicts a statistically significant change in Average Total ALSSQOL–R scores over three time points (clinic visits).

Chapter 5

Discussion

Summary of findings.

The sample used in this study appears to represents the national demographic of ALS patients, as characterized by age, gender, time since diagnosis, and region of symptom onset (ALSA, 2015). The mean age for this sample was 61.52, and according to ALS Association, the average age at diagnosis is 55 years old (2015), but the disease can develop in people between the ages of 40 and 70. In our sample, the estimated time since diagnosis averaged 27.35 months (2.27 years). This mean reflected for life expectancy for persons with ALS, which averages from 2 to 5 years from the time of diagnosis; however some variability in this length of time is noted (ALSA).

As previously discussed, this research was a continuation of an earlier cross-sectional study that found significant differences in QOL relative to level of bulbar functioning (Duff, 2008). The findings of the cross-sectional study showed that ALS patients demonstrating initial signs of bulbar impairments self-reported significantly lower QOL when compared to those patients with no functional impairment, indicating a nonlinear relationship between QOL and physical functioning (Duff). In contrast to the previous study, the current research examined the relationship longitudinally of the specific bulbar function of speech relative to certain aspects of QOL. There are limited studies examining bulbar functioning, in particular speech impairment, as it relates to QOL factors. Therefore, any information with regards to the timing of treatment interventions relative to level of bulbar impairment provides potentially novel information for the QOL and ALS literature.

The goal of the study was to answer the question: How do changes in speech impairment over time affect certain aspects of QOL in patients with ALS? Longitudinal assessment of participants' functional abilities and QOL occurred at three time points (clinic visits). The average difference between each time point ranged from 3.73 and 3.97 months. As expected, participants' total ALSFRS–R scores demonstrated a significant decline in overall physical functioning over time. Compared to other studies using the ALSFRS–R to assess physical functioning over time, this sample's mean scores were significantly lower (Cupp et al., 2011; Miano, Stoddard, Davis, & Bromberg, 2004).

Consistent with previous research findings, the relationship between physical functioning and QOL demonstrated a lack of meaningful correlations (Cupp et al., 2011; Robbins et al., 2001). No relationship between physical functioning and QOL was observed at times 2 or 3, although a moderate relationship between the two was identified at time 1. Examining correlations between physical functioning and QOL with greater detail allowed for a more critical analysis of the relationships. Observing how speech impairment relates to the participants' perceptions of these symptoms revealed a strong relationship (ALSSQOL–R: Bulbar Function) at the first two time points. Studies have suggested stronger correlations with those items and QOL measures related to physical functioning, such as the SF-36 (Tramonti et al., 2012). However, at time 3, there was a weak correlation between speech impairment (ALSFRS–R: speech item score) and the ALSSQOL–R: Bulbar Function. Examination of the speech item scores of the ALSFRS–R revealed minimal variability in speech functioning at time three.

Through the development of four hypotheses, the author predicted that as participants' ability to speak became increasingly impaired, they would have lower scores

on the following ALSSQOL–R subscales for Negative Emotion, Interaction with People and the Environment, Intimacy, and Bulbar Functioning. The hypotheses were not supported, with the exception of H_4 (perception of bulbar functioning). In terms of the other hypotheses, it was incorrectly proposed that a linear relationship exists between speech and QOL over time. This contrasts with the findings from Duff (2008), which found the relationship between physical functioning and QOL were not linear and that individual symptom progress required specific examination. Given that these findings have shown changes in QOL scores relative to levels of speech impairment, it was predicted that the trend this relationship might also be observed in a longitudinal design (Duff; Hillemacher et al., 2004).

Given that only one of the hypotheses was supported and three of the proposed hypotheses were unsupported, additional post hoc analyses were conducted to measure possible changes over time in the other QOL domains, Physical Symptoms and Religiosity, and Average Total ALSSQOL–R score.

The hypothesis that speech decline would be linked to negative emotion scores was not supported at a statistically significant level. Results failed to demonstrate a decline in Negative Emotion subscales scores over time to indicate increased emotional distress with disease progression. Other studies reported similar findings, indicating that over time, patients tend not to show significant changes in scores of psychological health, well-being, and self-esteem (Goldstein et al., 2006). Similar findings were obtained in a retrospective study by Cupp et al. (2011) that assessed the impact psychological health of patients with ALS over time by specifically examining the Negative Emotion subscale of the ALSSQOL–R. The study further evaluated the impact of antidepressants,

gastrostomy, and noninvasive positive pressure ventilation (NIPPV) interventions on psychological well-being. Findings demonstrated no correlation between psychological health (Negative Emotion subscale scores) and physical functioning in cross-sectional analysis of visit groups. Further observation of the data over time showed that psychological health remains stable despite the severe decline in functioning (Cupp et al., 2011). This may be explained by the fact that several symptoms of depression (fatigue, loss of energy, psychomotor retardation, etc.) overlap with the physical symptoms of ALS, and if patients have has lost the ability to communicate effectively, expressing how they are experiencing symptoms differently might be extremely challenging.

Similarly, the hypothesis that speech decline would be linked to lower social environment scores was not to be supported. Findings showed that participants Interaction with People and the Environment subscale scores slightly decreased over time, but not significantly. This finding is supported by the Lazarus and Folkman model of stress coping, suggesting reciprocity between the relationship of social support and symptoms of depression and anxiety. This theory has been suggested in other studies showing that social supports act as maintaining factors for patients with ALS (Trail et al., 2004). In fact, Lulé and colleagues (2013) have shown that social interactions and the support of friends for ALS patients are approximately twice as important in determining QOL for ALS patients then for cancer patients, and family members are equally important.

The hypothesis the speech decline would be linked to a decreased for intimacy also failed to demonstrate significance. Results showed that participants' desire for intimacy increased over time, although not at a level of significance. This positive direct

relationship between intimacy and time is consistent with previous studies that found that sexuality (Wasner et al., 2004) and intimacy rating are high, regardless of level of functioning (Rodriguez, 2010).

The prediction correctly indicated that as speech declines over time, associated symptoms are perceived as increasingly problematic. This was determined by decline in scores on the Bulbar Functioning subscale. From time 1 to time 2, the decline in scores was unremarkable, but the Bulbar Functioning subscale scores significantly decreased from time 2 to time 3. These findings were inconsistent with findings from an earlier study that showed speech impairment was related to depression symptoms (Hillemacher, et al. 2004).

Similar results were found in post hoc analysis of the Physical Symptoms subscale scores. The changes in mean scores over time indicated that as impairment increased with disease course, physical symptoms were perceived as increasingly troublesome. Again, a similar pattern was observed: from time 1 to time 2, the decline in scores was unremarkable, but the Physical Symptoms subscale scores significantly decreased from time 2 to time 3. In an earlier study, Ganzini, Johnston, and Hoffman (1999) identified pain and physical symptoms as correlates of quality of life and suffering, suggesting that patients easily identify physical symptoms of suffering as problematic, but symptoms of poor emotional well-being are more difficult to identify.

Examination of the Average Total ALSSQOL–R scores suggested a decline in QOL over time. From time 1 to time 2, the decline in scores was unremarkable, but QOL significantly decreased from time 2 to time 3. These results tend to contradict previous research, which has shown that global or overall QOL does not decline in patients with

ALS relative to physical decline. However, it is acknowledged that there is great variability amongst these studies due to the measures used and definitions of constructs (McElhiney et al., 2014; Pagnini et al., 2014).

Lastly, the results failed to show a significant difference or change in Religiosity subscale scores over time. As previously mentioned, high or low scores do not necessarily suggest impairment, but provide information about patients' religious identity (Felgoise et al., 2011). Other studies have identified the importance of faith for patients with ALS (Foley, Timonen, & Hardiman, 2014) and have shown that spirituality is a maintaining factor of QOL, especially as the disease progresses (Walsh et al., 2003).

Significance of findings.

Findings of the present study have shown that the level of speech impairment does not change QOL in the areas of negative emotions, social interactions, and intimacy. However, results did demonstrate that as speech declines over time, perception of bulbar functioning, physical symptoms, and overall QOL are impacted. These findings, in concert with findings from previous studies, support the notion that loss of speech in patients with ALS perhaps causes some change in decision making and perceptions of illness (Hillemacher et al., 2004; McKelvey, Evans, Kawai, & Beukelman, 2015), but certainly requires further understanding and exploration.

Limitations.

The present study has identifiable limitations. The sample consisted of patients receiving care from a multidisciplinary treatment clinic. Therefore, the findings will be specific to those individuals with ALS treated at a multidisciplinary clinic and may not be generalizable to the ALS population at large. Although the sample size was only few

participants smaller than the power analysis indicated, the study was greatly underpowered.

Limitations of the study also included identification of participants from a database. Discrepancies were noted between completed measures of functioning and QOL and clinic visits, as some patients completed one measure but not the other, and thus were excluded. Several participants were excluded because their bulbar functioning improved at the later at the third time point.

In terms of other individual ALS patient variables, the study did not take into account common treatments with known effects on psychological health, and/or noninvasive ventilator support. Other variables that were not controlled for were marital status, although this information was known, and additional information about the primary caregiver, which could have offered greater knowledge about both the functional capacity and speech.

Another limitation of this study is the difficulty in tracking use of devices to aid communication. Anecdotal information indicated that with the availability of technology such as smartphones and tablets, patients have increasingly utilized these devices as communication aids, rather than using traditional assistive augmentative communication devices. Therefore, it would be essential for clinicians and for future research to examine how augmentative speech devices, assistive speech devices, tablets, smartphones, etc. affect quality of life, once a patient has lost functional speech.

Implications of research findings.

The findings from this study have implications for further QOL research in the ALS patient population, and thus implications for medical professionals, psychologists, and ALS caregivers. Knowing that ALS patients perceive decline in bulbar function and physical symptoms as problematic over time, but do not have the functional ability to communicate the effects of this loss may help improve treatment implementation. Continuing to study the timing of when decline of the impairment occurs jointly with the study of patient perceptions and will likely prove beneficial for providers, patients, and their caregivers. Furthermore, understanding how actual loss of functioning versus perceptions of the loss affect decision making and how these variables influence QOL might be extremely valuable to both future researchers and patient care.

As previously discussed, research has previously revealed significant differences in QOL relative to level of bulbar functioning. The greatest differences in QOL are noted when comparing ALS patients with no impairment in bulbar functioning to those with the first signs of impairment. Those with the first detectable disturbances of bulbar impairment have lower self-reported QOL than patients with greater impairment (Duff, 2008). Therefore, the early signs of bulbar symptoms might prove a crucial time in working with ALS patients, their families, and treatment teams (Duff).

Timely psychoeducation for the patient, family, and other caregivers can helps provide an understanding of what can be expected in terms of QOL at each specific time point of functional decline throughout the course of the disease. Preparing a patient, family, and other caregivers offers the opportunity for prophylactic psychological and emotional treatments, prior to the onset of severe impairment, such as cognitive and

behavioral interventions. The literature shows that problem-solving techniques help the patient and family to reduce emotional distress (Felgoise et al., 2010). Thus, teaching patients problem-solving techniques in addition to coping skills to reduce discomfort and to manage pain and frustration prior to a change in functioning may help prepare the patient and improve QOL.

Further, psychoeducation needs to emphasize the importance of interpersonal relationships and the role of family, caregivers, and social support, given that ALS causes loss of or difficulty in communication and mobility, which can lead to limited social interaction. Several considerations need to be taken into account regarding intimacy, social skills, and conversational skills training for both the patients and caregivers. Psychoeducation should inform partners about the physical decline of their loved one, being careful when engaging in physical affection, and taking advantage of days when physical health is optimal (Gott, Hinchliff, & Galena, 2004; Rodriguez, 2010). Psychoeducation is most beneficial when it emphasizes alternative means of sexual expression, such as emotional connectedness.

As the disease progresses, emotional connectedness and the support received from caregivers and other interpersonal relationships is instrumental for maintaining QOL. Rodriguez (2010) demonstrated a relationship between sharing information with loved ones and to how intimacy is maintained in patients with ALS, regardless of functioning.

Therefore, ALS patients should be prepared in participating and engaging in social interactions, especially with the onset and progression of bulbar impairment, may be challenging. As speech becomes increasingly compromised, patients would benefit from a set of strategies to compensate for the inevitable communication deficits. Family

members, friends, and others caregivers would also benefit to learn a set of strategies to help prepare for the patient's eventual speech decline.

It is important for members of a patient's support network to understand that communication impairment makes it difficult for the ALS as her or she becomes limited in the amount of information they can contribute to a conversation. The patient's family also needs to understand that that accuracy of statements should not be questioned because he or she has impaired speech. The issue of inaccurate interpretation is likely due to the listener, or misinterpretation by a caregiver.

Understandably, this can create many frustrating situations for all parties involved. This provides further evidence for the importance and necessity for having someone accompany the patient to doctors' appointments and why it is important for both the caregiver and patient to take notes while at the appointment. However, if the patient and those in his or her support network can anticipate loss in speech, it will increase preparedness to allow for the implementation of adequate education, planning, and evidenced-based interventions.

Furthermore, the ALS patient who experiences symptoms of pseudobulbar affect often demonstrates pathological laughing or crying. Similarly, psychoeducation with regards to the associated symptoms of pseudobulbar affect, and what patients and families can expect, is important to understanding that at these times patients' emotional affect might seem incongruent with the present situation, due to loss of functioning. Providing psychoeducation, in concert with cognitive and behavioral techniques, can aid in assuaging any potential distress.

As the disease progresses, patients will become increasingly dysarthric and will demonstrate utterances of ambiguity and incomprehensibilities. Psychoeducation should be provided, as it is important for others to understand that unintelligible speech is not necessarily a reflection of cognitive abilities. Patients' support networks should offer them the opportunity to communicate through alternative mean and/or devices. Patients and their caregivers should discuss their treatment prior to loss of speech functioning in order to prepare for how patients plan to communicate with others, especially communication of their medical needs.

Nonverbal communication is an important aspect of how people interact and express ideas with one another. For some patients with ALS, nonverbal communication does become compromised, but can also play a vital role in maintaining effective social relationships and intimate emotional expression (Joubert, Bornman, & Alant, 2012). In preparing for loss of nonverbal communication, psychoeducation helps to advise patients and their support network that bulbar dysfunction will compromise their ability to deliver a joke, intend sarcasm, and smile. Therefore, patients and their social networks may wish to create hand gestures for when the patient indicate sarcasm and other nonverbal cues to others.

However, it is also important for those close to the patient to know that nonverbal communication also plays an important role in facilitating and maintaining emotional support for ALS patients. Physical touch supports intimate emotions (App et al., 2011). ALS patients' families and friends would benefit from utilizing nonverbal communication, such as touch and presentation of self, as effective means of communication to substitute or enhance impaired speech (Duff, 2008). As reiterated

throughout, all of these techniques are best implemented when identified for targeted time points when a change in the patients' self-perceived QOL may occur.

Future research directions.

Unfortunately, this study was not able to advance the cross-sectional findings from Duff (2008) in order to demonstrate the changes in QOL relative to loss of speech over time. Ideally, a follow-up study might consider a hybrid of the Duff and Cupp et al. studies (2011). Findings from Cupp et al. showed that patients with ALS could experience increased emotional distress with the onset of each loss of function, followed by an improvement in psychological health as they achieve acceptance. These results supported those shown by Duff, although assessing over time. However, only changes in negative emotion relative to global physical functioning were examined. Cupp and colleagues noted the intervals of 3 to 4 months between clinic visits might be too great to detect the quick and possibly small spikes in emotional distress. Therefore, a combined cross-sectional and longitudinal design with shorter intervals might offer further understanding about the timing of decline in QOL with physical impairment.

However, QOL in ALS research would benefit from the exploration of other variables, including response shift, acceptance, coping, and coping versus perceived decline. A somewhat recent trend in ALS and other chronic illness QOL literature is the concept of response shift. The term response shift is derived from statistics and attempts to account for the change on QOL measures over time. Response shift is broadly defined as how a person reevaluates values and expectations and essentially redefines the meaning of their QOL because of their illness (Sprangers & Schwartz, 1999). Subsequently, these reevaluations influence how patients might respond to items on the

same measure of QOL at a different (later) time. Although one might argue that it is an academic and theoretical epiphenomenon and is really no different than constructs of acceptance and/or coping. Admittedly, measuring change in these constructs relative to change in functioning would be difficult over time because they are theoretical in nature. Attempting to assess change in cognition rather than change in behavior is challenging because it is difficult to quantify an individual's thoughts, attitudes, or perceptions. More studies using qualitative methodology could provide valuable insight into the individual variability in ALS symptoms using identified themes and generalizing to the larger ALS population.

In terms of coping and coping verses perceived change, exploring these concepts further could offer a better understanding of the factors that maintain QOL over time for patients with ALS. According to the literature, coping and perceptions of change and change versus coping are often related to social contexts, decision making, and theories of loss and acceptance (Foley et al., 2014). Similarly, further exploring these concepts qualitatively, while also studying problem-solving theories with regard to timing of patients' decisions and whether they perceive interventions such as a gastronomy or NIPPV medically necessary. With regard to loss of speech, advancing the research on perceptions and attitudes and decision-making about use of assistive devices would also be beneficial. However, because it seems assistive devices are underutilized, little is known about their impact on QOL. Advancing technologies and use of smartphone and tablets may be another avenue for future study.

Conclusions.

The degenerative impairments of ALS significantly impact nearly every aspect of a patient's life. However, research has shown that despite the catastrophic effects of the disease, patients' QOL is maintained over time. Similar findings were observed in this study. The examination of speech impairment was not found to correlate with QOL over time, with the exception of participant's perception of bulbar impairment as problematic. At the first two time points, there was a strong correlation between impairment and perception, but at the third time point the relationship weakens. These findings suggested that while speech functioning does not vary, patients' QOL does, therefore, the two variables did not correlate. It is possible that the variability in QOL may be due to a third variable such as social problem-solving skills and/or coping. As such, some patients with ALS may demonstrate better coping than others, and therefore, may have a better perceived QOL, despite their speech impairment. Others may not have adequate problem-solving skills to cope with their speech impairment, and therefore, may have poorer QOL.

Findings also indicated that over time ALS patients do not experience changes in their emotional health or psychological well-being, social or environmental interactions, religiosity, or their desire for, and satisfaction with intimacy. These results are consistent with the literature. However, the findings of this study also revealed that over time, patients with ALS perceive bulbar impairment and physical symptoms as increasingly problematic. Therefore, based on the findings of this present study and the previous cross-sectional study (Duff, 2008), expanding research to target detection of initial impairment and the possible association with constructs such as, acceptance, coping, and

perception of change will help to provide a better understanding of and knowledge about patients' problem solving and decision making.

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