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## Investigating the Psychological Effects Associated with Motor Neuron Degeneration in Persons Diagnosed with ALS

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# Investigating the Psychological Effects Associated with Motor Neuron Degeneration in Persons Diagnosed with ALS

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by

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An undergraduate honors thesis submitted in partial fulfillment of the

requirements for the degree of

Bachelor of Science

in

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Thesis Advisor

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## **Abstract**

Investigation of the psychological effects of a terminal condition, wherein current care plans are primarily focused on physical rehabilitation and pain management, is crucial. Amyotrophic lateral sclerosis enacts irreversible damage to the body of those diagnosed, resulting in neurodegenerative processes that cause paralysis and eventually respiratory failure. There is no known cure or cause, which eliminates many traditional treatment avenues. Though the disease wreaks havoc on the muscular systems of the body there are no direct effects that degrade mental processing, leaving the individual experiencing the condition an aware witness to their own functional loss. Creating a multidisciplinary approach inclusion of psychological care of both the person with the condition, as well as their family and caregivers, is integral to appropriately providing support. A community-based approach to caring and supporting those with ALS is integral to a comprehensive plan for addressing this terminal illness.

## Introduction

Amyotrophic lateral sclerosis (ALS) is a terminal neurodegenerative disease that has no known cure and no known cause (Caga, 2015). Though researchers know many of the deep neurological processes surrounding the condition, there still is no answer as to why the body systematically shuts down throughout disease progression. ALS is relatively rare; only approximately 15,000-30,000 Americans at any given time are living with the disease (“Epidemiology of ALS and Suspected Clusters”). Despite its rarity, ALS has slowly gained notoriety through prominent public exposure by way of physicist Stephen Hawking—who has lived with the condition for decades—and viral social media trend of 2014, the ALS Ice Bucket Challenge. The Ice Bucket Challenge raised money to fund ALS research by recording the dumping of ice-cold water over the participant’s head—a demonstration meant to briefly simulate the respiratory failure that characterizes the terminal end of ALS disease progression.

ALS initially expresses symptoms such as loss of breath, difficulty grasping objects, excess tripping, abnormal fatigue, or trouble controlling vocal pitch during speech. Following the initial presentation of these symptoms, the severity graduates to difficulty producing speech, difficulty swallowing, and the beginnings of degradation of motor movement in the limbs (the hands, arms, legs, and feet). In the final stages of the condition, the gradual breadth of muscle degradation progresses to reach the lung, chest, and face muscles. At this stage medical intervention is necessary to keep the individual breathing and adequately nourished if they are to survive (“Symptoms and Diagnosis”). As an incurable neurodegenerative condition, ALS exacts irreversible damage to the motor neurons in a person's body. Despite the gradual loss of motor neurons, the mental

process, all of the senses, eye movement, and bladder control are rarely affected in persons with the condition (“Symptoms and Diagnosis”). The individual with ALS remains witness to the degradation of their own motor function while having the capacity to comprehend the disease progression, emotionally feel the impact of the loss, and experience the sensory environment clearly but the ability to react in any physical respect degrades progressively over time.

ALS is part of a group of progressive neurological conditions called motor neuron diseases (MND). Symptoms of MNDs exhibit themselves through gradual degradation of cells controlling voluntary muscle function. In normative function, electrochemical messages from the upper motor neurons inside the brain transmit to lower motor neurons in the brain stem and spinal cord then activate outwards to particular muscles. Through expression of ALS symptoms, interruptions in the electrochemical signals between the lower motor neurons and muscles inhibit normal execution of muscle movement. Once these muscle functions are interrupted, they gradually deteriorate and never regain control—often developing an irrepressible trembling. Upon the disruption of signal between upper and lower motor neurons, a stiffness of limbs arises as muscles degrade (“Motor Neuron Diseases”).

While the symptomology of motor neuron degradation in ALS only directly affects the musculature of an individual, the possible reverberating psychological effects occurring from a terminal diagnosis must be incorporated into the multidisciplinary care plan adapted for treatment. The possible psychological side effects following diagnosis warrant investigation and incorporation into a multidisciplinary care plan suited not only to the individual diagnosed with ALS, but also their family and the caregivers involved.

The psychological impact of a terminal diagnosis on both individuals with the disease and their surrounding community has been thoroughly researched, but little consistent incorporation into treatment plans has been documented (Hodgen et al., 2015). Though mental health diagnoses such as anxiety and depression are rarely seen amongst individuals following the onset and diagnosis of their ALS, many specific symptoms associated with them—such as hopelessness and despair—are frequently endorsed by individuals throughout disease progression (Ng & Khan, 2011; Plahuta et al., 2002; Foley et al., 2014). Emerging patterns of hopelessness, demoralization, and grief are extra prevalent in persons diagnosed with ALS in comparison to populations with other terminal illnesses, especially in instances where diagnostic intervals (the amount of time between symptom onset and verified diagnosis) are longer (Caga et al., 2014). The involvement of personal factors—such as gender, age, socio-economic status (SES), race, religion, physical location, and past mental health diagnoses—heavily affects the vulnerability to psychological change post ALS diagnosis (Ng & Khan, 2011). Due to the multivariable complexity of ALS degradation, the psychological follow-up with the person diagnosed and their family by healthcare professionals is crucial to structuring an appropriate care plan of action.

Typical ALS care often begins within a framework meant to manage the physical loss individuals with the condition experience, but fails to provide psychological support. In order to adequately provide care and treatment for individuals with an ALS diagnosis, there must be a significant addition of psychological support structures into the care plan for the condition. Due to the physically degradative nature of ALS, the care team involved in treatment of the condition must be able to encompass treatment for failure of

all motor functions along with supporting the mental and emotional needs of the person with the condition. This is something not already universally applied within the multidisciplinary cannon ALS treatment. Because of the compounding complexity and variability of the rapid degradative functional motor loss, development of care plans must be uniquely tailored to the individual, their community, and their needs. Flexibility and communication from healthcare professionals must be heavily emphasized if a treatment plan is going to appropriately address the needs of the person experiencing the condition and their surrounding community.

The aspect of including the impact on family and caregivers should be prioritized within a multidisciplinary care model. Throughout the progression of the disease, the community surrounding individuals with ALS are exposed to the repercussions of the gradually degenerative effects of the disease. Because of the rapid loss of motor function throughout the advancement of the disease, care is required progressively more; as the capacity for healthy, safe autonomy begins to disappear, the increasing reliance on caregivers and relatives rises. Thus, a concurrently beneficial support system suited to caregivers, family, and the person experiencing ALS must be developed to maintain the health of all parties (Cipolletta et al., 2014). Recent studies suggest that from initial diagnosis, not only should systems be in place to support the person with the disease, but the community surrounding them. These family caregivers have an intimate knowledge of the progression of their relatives' condition and therefore should maintain consistent contact with diagnosticians to aid the understanding of the specific case (Hubbard et al., 2012; Hogden et al., 2015; Cipolletta et al., 2014).

The act of caregiving has both positive and negative qualities, with the potential to heavily impact the health of those involved—for the caregiver, families, and person with ALS. In order to combat the potential negative side effects of caregiving—such as increasing burden, stress from lack of resources and information, and failure of communication with healthcare professionals—a comprehensive psychologically oriented support model must be provided in order to care for the community surrounding an individual with ALS (Weisser et al., 2015).

This can be achieved in a variety of ways. Something like mindfulness training for caregivers and family has been examined; this is the practice of consciously and actively creating new observations of objects in an individual's awareness. Through this process, the individual practicing is able to cultivate awareness of change in perspective of the subject, rather than relying on the schema of old perceptions (Langer et al., 2010). Alternatively, peer support includes other community members who are living with or exposed to the condition. Social connectivity through support groups is a valid way to attain and retain structures of reliance and knowledge. Through the creation of a community of knowledge surrounding individuals with ALS, their caregivers, and their family, there is a real potential for improved quality of life (QoL) and prognosis from individuals diagnosed with ALS (Dawson et al., 2003; Pagnini et al., 2016; Weisser et al., 2015).

Another crucial aspect to improving QoL in persons with ALS is the immediate introduction of palliative care planning and measures after diagnosis. The terminality of the condition is often exhibited through rapid deterioration of muscular function, which can quickly limit communication capacity (Hodgen et al., 2015). In order to preserve the

integrity of the individual's independence, an emphasis on the importance of a pre-established plan for end of life (EoL) wishes is required. Palliative care should be used as an active tool throughout the neurodegenerative progression of ALS, aiding the management of symptoms (Dawson & Kristjanson, 2003). Developing a plan for end of life care following diagnosis is not only important for following the requests of the person with the disease, it relieves much of the decision-making pressures the family and caregivers may feel as communication degrades (Hodgen et al., 2015; Foley et al., 2014). Development of a solid palliative care plan is foundational to establishing a basis to move from throughout disease progression; determining the place of care, place of death, potential establishment of a Do Not Resuscitate order (DNR) or a Physicians Order for Life-Sustaining Treatment (POLST), or alternatively the depth of advanced care, and if assisted death is an option (depending on location) are only some of the ways this discussion comes into play early in the condition (Oliver & Turner, 2010).

This literature review will briefly address the variability of motor neuron degeneration, establish some of the possible psychological effects of the motor function degradation associated with the diagnosis of ALS, discuss the impact on the family and caregivers surrounding the individual with the condition, and develop a model flexible enough to mold to the multivariable composition of the disease, but specific enough to deal with the uniquely severe functional loss exhibited over its progression. The progression of ALS is erratic and outcomes can only be vaguely predicted through an analysis of the physical symptoms as they present; therefore a multidisciplinary model must retain the plasticity to adapt to the changing disease progression circumstances surrounding each case.

## Neurological Symptomology

### **Motor neuron degradation: a breakdown of functional loss**

The exact processes of motor neuron degeneration lies in the progressive and simultaneous decay of both upper motor neurons and lower motor neurons. Because body region of onset can vary, incorporating knowledge surrounding the severity of degradation based upon area initially affected is crucial in understanding the intensity and progressivity of each individual case. From clinical symptom onset, motor degradation is specific and distinct. Initial symptoms generally present universally across the condition. As physical deterioration cascades outward from the body area of origin, the disease's effect becomes more densely complex. Thus, it becomes highly important to understand the specific region of onset within each individual case. In a study on the focality of motor neuron degradation, researchers were able to evaluate the functional status in upper and lower motor neurons based on region of onset and the specific anatomical features included in said region. Their findings articulate the variability of severity in each body region, adding highly valuable data to inform the care process (see Appendix A) (Ravits et al., 2007).

The site of onset can also indicate the level of vulnerability an individual experiences, in respect to their survival outcome. There is a massive range of variability in survival intervals amongst individuals with the condition. In order to best aid the care plan, any influential variables must be included as areas for flexibility and personalization of support. For example, as demonstrated in Appendix A, bulbar-oriented ALS is a particularly aggressive variety of the condition (Ravits et al., 2007). Degeneration of both upper and lower motor neurons occurs much more intensely in the

instance of a bulbar onset—specifically in respect to the speed of respiratory failure. The bulbar region includes critical cortical areas such as the medulla, pons, cerebellum, and spinal cord, all of which are integral to respiratory function. With the degeneration of respiratory function, individuals with the condition face malnutrition due to muscle weakness and limited swallowing capacity, increased likelihood airway obstruction, and high risk of aspiration and pneumonia (Hadjikoutis & Wiles, 2001). Similarly, older individuals (80 years or greater) are often more vulnerable. These cases are especially important to quickly and comprehensively develop a care plan for because of the already established frailty of the individual (Chio et al., 2009).

Despite the needs to specify the case variability across ALS, there are some consistencies across the variability of ALS. Though motor neuron degeneration affects the muscular system of any individuals with the condition, it does selectively spare certain functions; bladder function located in Onuf's nucleus in the sacral cord tends to remain untouched by degradation, meaning those with ALS typically maintain control of their bowels throughout disease progression. Similarly, eye movements tend to maintain functionality throughout the entirety of the disease (Bruijn et al., 2004). Overall because of the differentiation between sensory motor neurons, those with ALS retain their perceptual capacity. The sensory experiences of sight, touch, hearing, taste, and smell are not damaged throughout the disease progression despite the surrounding functional loss (“Symptoms and Diagnosis”).

**ALS: a terminal condition**

The diagnosis of ALS is a terminal one; the rapid onset neurodegenerative condition is unavoidably fatal and exhibits particularly aggressive symptoms leading up to the death of the individual diagnosed. Through an attack on the motor neurons in the brain, which control voluntary muscle movement, the disease produces the degradation and eventual death of these brain cells. In turn, this commences a systematic shutdown of the body's voluntary motor function. Once motor neuron cells die, there is no regaining or replacing them. Through the death of these cells, ALS brings on slow onset paralysis and respiratory failure that eventually lead to the death of the person with the condition (Bungener et al., 2005). Despite a deep understanding of the genetic processes involved in the degradation of motor neurons in persons with ALS, this is superficial knowledge in comparison to the unknown root of the disease's irreversible processes. It is hypothesized that ALS is a multifaceted and multi-systemic disease when it comes to its neurological origins, but the true depth of its cause is yet to be completely explored (Musarò, 2013).

Thus when we discuss post-diagnostic care for individuals with ALS, for the sake of this literature review we will not be delving into the unanswered question of how to cure ALS. Instead of dwelling in the neurobiological reality of how ALS occurs, the purpose of this paper is to focus on the rehabilitative and palliative care measures that can be taken to improve QoL in persons diagnosed. While an informed perspective of the symptomology that occurs on a neurobiological level is helpful, a psychosocial approach to care is more beneficial to the current treatment of ALS. Psychosocial approaches can be used to improve post-diagnostic communication between medical professionals, those with ALS, their family, and their eventual caregivers, an area of the treatment of the

condition that family members and individuals with the condition have cited as dissatisfying (Paganoni et al., 2015). A combined focus on early stage preemptive physical strengthening, psychological support, and palliative care planning could possibly yield more satisfaction in the diagnostic process and help support the community for the future disease progression.

## **Psychology of persons diagnosed with ALS**

### **The influence of personal factors**

Understanding and addressing the psychological needs of individuals diagnosed with ALS is wrought with variability—each individual’s life, their family, and their chosen caregivers bring a wide range of personal factors to the process of disease progression. In order to best accommodate the needs of all parties, the identification and study of the influence of personal factors is crucial to providing adequate psychological support in symphony with the required physical care.

With the aid of a community-based ALS clinic out of Victoria, Australia, a study done by rehabilitation physicians identified personal factors that shape an approach to psychological care in ALS support groups (Ng and Khan, 2011). Their study was designed to use the International Classification of Functioning Disability and Health (ICF) in order to quantify the impact of progressively degenerative diseases, such as ALS (“International Classification of Functioning”). Based off an interview with each of the participants, researchers were able to successfully report on a massive multi-variable list of characteristics categorized into demographic factors, emotional states, coping strategies and styles, personality, beliefs, attitudes (of the individual with the condition),

and perceived social support, all of which have potential to inform the physical and psychological care involved in ALS treatment (Ng & Khan, 2011). Research using the ICF—in discourse with work done by Neurologist Dr. Adriano Chio using the ALS Functional Rating Scale (ALSFRS)—identifies crucial focal points to creating a physical and psychological basis of understanding a recently diagnosed individual. The work done identifies factors such as personality, socioeconomic status, dependence on others, self-efficacy, and attitude towards the biopsychosocial impact of neurodegeneration as focal in the discussion to best support the individual experiencing ALS and their surrounding community (2009).

Demographically, age, gender, race, educational status, and socioeconomic status (SES) all are informative variables in following progression and experience of ALS in persons diagnosed with the disease (Chio et al., 2009; Ng & Khan, 2011). Most significantly, age heavily factors into the survival interval for individuals diagnosed with the condition. Though individuals with symptom onset before 40 years old have a significant likelihood of living up to a decade following ALS manifestation, the prognosis for persons diagnosed at or around the age of 80 is often less than two years from disease inception; a time well underneath the average bounds for survival time at symptom onset, which tends to be approximately 3-5 years (Rosengren et al., 2015; Simmons et al., 2000). The importance of introducing palliative care and psychological intervention as quickly as possible following diagnosis is emphasized here; the older an individual is, the more vulnerable they become as the disease progresses. And that's without factoring in any other demographics with the potential to influence vulnerability.

Gender does not have significant influence over the average survival interval of ALS (Chio et al., 2009). It is noteworthy, however, to state that older women have a higher likelihood of manifesting the rapidly progressive bulbar-oriented focus of the disease, which makes them more vulnerable to aggressive forms of the condition that degrade autonomic function much quicker (2009). This only further reinforces the necessity for the construction of psychosocial support structures for the women and their communities. Furthermore, in an analysis of the biographies of four women who had ALS, research uncovered relevant information surrounding their gender roles and psychology. Many of their experiences articulated the frustrations coming out of the rapid loss in communication capacity typical of the condition, reporting the horrifically patronizing ways healthcare professionals would “pat them on the head or cheek” in substitution for genuine compassion and attempt at communication. Others wrote of the difficulty of feeling useless in their condition and not fulfilling the requirements of being a wife.

Though not fully documented in the research on ALS, I believe these sentiments reflect particular vulnerability on women who are used to occupying spaces of control around the home—a space they are likely to be located within until the disease reaches its final stages. Their accounts of ALS progression compound the previously mentioned loss of control into anxieties they found in struggling to maintain family structure, even as they were in the final stages of their life (Rosengren et al., 2015). Research conducted on individuals with Parkinson’s Disorder articulates women’s higher tendency towards depressive symptoms than men, as well as more intense pain levels and cramping. While this may not directly correlate to ALS, it is important to consider the results of the study

of PD because of the closeness between the two conditions. When a discussion amongst the communities looking into PD cites a gendered experience of the condition, the presence of the same effects could very easily exist within ALS community. As research identifies, sex and gender are crucial components to understanding health and disease in the later stages of life (Solimeo, 2008). The lack of comprehensive, localized investigation into the effects of sex and gender on ALS progression indicates a gap in the current research. Based off of the effects demonstrated within PD community, it would be remiss if the ALS healthcare community ignored this avenue of research.

Data are not as numerous for race, educational status, or SES, but their inclusion in the profile of individuals diagnosed with ALS increase the capacity for understanding their collective backgrounds. Limited research points to a higher rate of ALS occurrence amongst Caucasian populations than other non-white racial groups, but this could also be a byproduct of the lack of diversity amongst studies on the condition (Plathuta et al., 2002; Ng & Khan, 2011). Further research by Plathuta in 2002 points to a significant combined influence of age, gender, marital status, and education on hopelessness, but discounts their individual roles in informing psychological care beyond this correlation; I argue that their lack of deeper analysis on the dynamics of each of these categories may have shorted their capacity to truly evaluate the importance of each (Plathuta et al., 2002).

### **Illness beliefs**

In the studies of the beliefs of individuals with ALS and other terminal conditions, a developed understanding of their perspectives on their conditions can aid the treatment of the disease. Research on Parkinson's disease has alluded to the power of perceived

control over daily symptoms and overall progression of terminal conditions, stating that these factors hold sway over both person experiencing the illness and caregivers (Wallhagen & Brod, 1997). While the symptoms of Parkinson's mostly manifest in tremors of extremities, slowness of movement, inflexibility of upper body, or lessened balance and coordination, ALS most often exhibits initially in speech difficulties, trouble grasping objects, or general fatigue before progressing to large-scale system failures ("What is Parkinson's disease?"; "Symptoms and Diagnosis"). Though Parkinson's disease by no means has an identical symptomology, treatment plan, or genetic expression to ALS, the similarities between both neurodegenerative conditions allow for some generalizability. The diseases are both terminal and classified as neurodegenerative; therefore they both have the potential to inform the experiences of the individuals affected by these illnesses.

Further research done on Parkinson's disease has illuminated the importance of positive perceptions of terminal illness. By examining populations diagnosed with Parkinson's, some newfound information about negative illness perceptions reshaped understanding of the disease. The results in the Hurt et al. (2015) study yielded that younger persons with Parkinson's had a higher likelihood of demonstrating negative perceptions of their condition, heavily influencing the likelihood for depression found in these younger subpopulations. Research also found that in-person completion of the questionnaire involved in the study (as opposed to online) yielded less symptoms reported on average per individual. Additionally, the reaction to discussion of treatment to control symptoms was also found to negatively affect the responses of persons with Parkinson's disease and their perception of their condition. Overall, the studies points to

irrelevancy in positive belief, but instead perception of control (Hurt et al., 2015). All of these findings linked together point to a necessity for development of models inclusive of younger populations. Centering communication on EoL wishes early in disease progression establishes control in decision-making for persons of all ages with ALS, this will help aid overall illness perception.

A discourse has begun to develop surrounding illness beliefs in ALS. Preliminary research delves into the personal factors shaping an individual's experience with the condition—such as gender, race, age, educational status, and socioeconomic status. Researchers have been able to place a correlation between individual's risk to depression, anxiety, fear, and stress and an interaction of personal factors (Ng & Khan, 2011). Similar research looking directly into hopelessness in individuals with ALS addresses specific predictors of negative affect following diagnosis. Through predictors such as the health locus of control and feelings of purpose in life, there can be indications of hopelessness in a person with an ALS diagnosis (Plahuta et al., 2002). In application on ALS, the results can encourage further development of support systems, like Hodgen's model, that help to promote control in the lives of persons with the condition. This research also lends itself to development of furthered understanding in caregivers and family members involved in the lives of persons with ALS. By instigating control on the parts of the individuals with the disease, it gives more reliable guidelines for the caregivers and family surrounding the people living with the condition.

## **Stress & coping with functional loss**

The action of ALS works as a rapid operational loss, regressing an individual's physical capacity until the collapse of respiratory function. With the process of the disease comes a consistent adjustment and re-adjustment of physical ability as the loss of motor function progresses. Alongside this physical loss is a subsequent psychological strain that accompanies the condition. As the individual comes to terms with the progressive physical loss, research has identified that they are concurrently affected by despair, powerlessness, anger, hopelessness, and loss of identity as components of their suffering. In order to balance meaningfulness over the difficulty found in suffering from functional loss, the individual diagnosed with ALS must have the psychological resources to support them. Frustration and despair have the capacity to arise out of the complications surrounding functional loss, but similar experiences can also be attributed to absence of adequate communication, information, or clear responses to questions about the illness. These findings indicate that not only are personal factors integral to an individual's experience of the condition, but also the solid communication with healthcare professionals, family, and caregivers about the disease (Rosengren et al., 2015).

Participants' testimony from a separate study confirms the importance of internal personality traits and externally supportive relationships with relevance to distress and suffering. Those diagnosed with ALS with a concurrent positive quality of life (P-QoL) cite feeling more optimistic, more control, and less vulnerable to the decline of their muscular function when engaging in social and physical activities while their health permits. Mitigating stress and coping becomes easier when engaging in physical activity

(while able) and maintaining contact with community. Expectedly, as with other research findings, those with more severe disease progression in the negative quality of life group (N-QoL) were more likely to experience increased sadness, anger, and frustration (Nelson et al., 2003). If N-QoL persists, individuals with ALS, their family, and their caregivers report a significant increase in the level of anxiety they feel towards the future; this warrants a higher necessity for reliance on caregivers and family sooner, in comparison to those with P-QoL. Alternatively, if adequate support and coping strategies are provided to mitigate the experience of physical deterioration and emotional distress a weight can be taken off the community's shoulders by alleviating the experience of psychological burden.

In order to support individuals recently diagnosed with ALS, a bolstering of their social support structures is crucial. Beyond the impact of being diagnosed with a terminal condition, the intricacy of losses surrounding the progression of the condition demand the development of coping strategies (Jakobssen Larsson et al., 2016). The use of problem-focused and emotion-focused coping strategies have been shown to increase the perception of control in the lives of those with the condition, a key component to aiding management of hopelessness, despair, powerlessness, and loss of identity that occur (King et al., 2009). Specifically, the use of problem-focused strategies—which aim to manage the source of stress—have been shown to elaborate on the control that can be expected in ALS and how to achieve it. These strategies then not only aid sustenance of an individual's locus of control, but also serve to improve communication of information on the condition (Jakobssen Larsson et al., 2016).

The complexities of realizing the coping process make it difficult to identify direction of influence. Research demonstrates correlations between differing coping strategies, psychological health, and physical function. Though, the difficulty of solidifying this relationship comes when considering direction of influence: are the coping strategies informing the psychological health and physical function or is it vice-versa? Jakobssen-Larsson's research cites these are findings that must be delved into further to provide knowledge on how best to support those with ALS and their communities (2016).

### **Comorbidity between ALS and mental illness**

Preliminary research into the manifestation of mental illness, such as depression and anxiety, in individuals with ALS is fairly uncommon. Though anxiety and depression are not prevalent amongst populations with ALS, there are significant behavioral impairments that go along with presentation and identification of ALS. The behavioral impairments (such as hopelessness, anger, and feelings of despair and isolation) indicate psychological and social shifts following diagnosis that are important to address. These shifts often manifest over the six months following diagnosis and can be accompanied by anosognosia relative to the physical degeneration that is characteristic of the condition (Bungener et al., 2005). While helpful preliminary findings, the act of studying ALS longitudinally gives a more complete picture of disease progression.

When using longitudinal methodology, the danger of persistent hopelessness in persons with ALS becomes more apparent. Through these broader, longer studies, the influence of psychosocial factors truly arises. Despite most of the cross-sectional research looking into disease-related variables, less common longitudinal research is able to more

readily measure the impact of psychosocial factors over an individual's interaction with the disease. Though previous diagnoses of mental illness has little influence over an individual's reaction post-diagnosis, those with longer diagnostic intervals, individuals over the age of 65, and those from lower socio-economic platforms have been cited widely across ALS literature to be more vulnerable (Caga et al., 2015; Jakobssen Larsson et al., 2016). Analysis of the relationship between hopelessness and degree of suffering echoes the correlation between N-QoL and perception of disease progression, as each increases, the other follows. The real question lies in what the direction of influence is. Nonetheless, individuals with ALS report finding relief from the hopelessness and anxiety of the condition when they focus locus of control internally. Instead of attempting to articulate control over external factors like their health and the effects on surrounding community, individuals with ALS are encouraged to focus on internal aspects like self-care, mindfulness, and self-empowerment. Strategies focused on removing control over external stimuli have been shown to boost subjective well being (Nelson et al., 2003, Plahuta et al., 2002).

In order to address the multifaceted degeneration associated with ALS, a multidisciplinary care program is encouraged. Research shows that this approach to care not only emphasizes including specialist doctors in disease progression, but also provides room for inclusion of psychological and community care. Despite not demonstrating massive amounts of common associated psychological phenomena, providing support for people with ALS has the potential to positively affect QoL, perception of control, and overall social community. Setting up psychological support structures as close temporally

to diagnosis as possible should be included as part of palliative care program that is developed for disease progression planning.

### **Psychological support structures**

The psychological and economical impact of ALS often begins far before diagnosis. Individuals with undiagnosed motor neuron degeneration commonly mistake early symptoms for a whole range of natural life occurrences—such as aging, result of injury, or other conditions. Therefore the importance of action as early after diagnosis as possible is crucial to support and care. Early physical and psychological action is integral, not only does it improve survival intervals, but also persons with ALS may already be harboring some side effects from pre-diagnosis symptoms. Furthermore, research also suggests that the impact of personal factors on QoL should be weighed heavily in the care process (Ng & Khan, 2011). Psychological issues, social support, and spirituality have all been shown to influence QoL more highly than the actual physical state of an individual with ALS, thus highlighting their importance in the care plan. Support looks different for every case, but as previously identified, creating an internal locus of control amongst individuals with ALS has the potential to pave the way for a more positive experience through disease progression (Plahuta et al., 2002; Simmons et al., 2000).

Communication is the key to solidifying the preferred method of support for the person diagnosed. Self-empowerment has been shown through research to be heavily influential in the psychosocial strengthening in the cases of persons with ALS. For individuals with the condition, their families, and the care community, self-empowerment establishes the flexibility needed as the disease symptomology becomes more violent. As

degeneration continues, the importance of maintaining self-empowerment and self-care is integral to the health of those involved (Ng & Khan, 2011).

In examination of active mindfulness practice, researchers negatively correlated feelings of burden, depression, and anxiety with positively correlated quality of life. Mindfulness practice aids the transition onto internal rumination. Through the use of meditation and conscious attention, researchers were able to identify the beneficial elements of this practice on the lives of those with terminal illness. Parallel to the existing effects in those without terminal illness, mindfulness practice amongst those with ALS was correlated with increased positive mood change and outlook, decreased anxiety surrounding future events, and an overall renewed sense of peace. The practice was also cited by some participants to bring them closer to their spirituality or religion (Chadwick et al., 2008). Both an independent and group task, mindfulness practice is continually appealing because of its accessibility. This makes it well suited for inclusion in a multidisciplinary care model, because it can be assembled in nearly any environment and maintain success amongst those participating.

## **Family & caregiver involvement**

### **Burden**

I have discussed the aspects of ALS that can be focused upon in order to improve the quality of life in persons with the disease, but as with support of a patient there must be support of the surrounding community of family and caregivers. The act of caring for a person experiencing a neurodegenerative disease has the high potential to be extremely emotionally and physically demanding. As neurodegenerative symptoms progress, there is an innate necessity for more involvement on the part of the caregiver and family.

Throughout the progression of ALS, symptoms graduate from being mildly difficult to navigate autonomously to severe enough that independence is impossible.

Comprehensive research providing data on the experiences of family or caregivers is integral to a fully functional model of the disease. In order to enact proper care, these people must be appropriately supported by a structure that also prioritizes their health, as well as the individuals that have the condition.

In a meta-ethnography done in 2012, an investigation into family member's perspectives on the terminality of their relative's condition shed light on the breadth of current research. Out of a sample of 212 research papers on neurodegenerative diseases, 84 were excluded because of a distinct lack of any discussion surrounding family member's perspective. Furthermore, from those initial 212, only nine ended up meeting the criteria for the synthesis conducted by the reviewers. The results yielded from this meta-ethnography led to a deepened understanding of the weight of the emotions involved in terminal neurodegeneration. The ethnographical research demonstrated an "intimate awareness of changes in the relative" (Hubbard et al., 2012, p 112), suggesting that family members are aware of the differing progressive aspects of the disease, deterioration in functional abilities, and the understanding that their relative will in fact die. Relatives acutely observe changes in their family member's state, but reviewers articulate that there was a lack of relationship between their perspective and the support they were receiving (Hubbard et al., 2012).

Specifically in families of persons with ALS, there is a very visceral discussion of grief: "every day (the grief) is with you... the whole time it never leaves you. Then you go through the different stages, when they stop walking, when they're in chairs. You get

used to a new stage and then you carry on, then the next one comes,” (Dawson & Kristjanson, 2003, p. 39). The progression of ALS exhibits itself as a series of crises that potentially lead to the death of the individual with the condition. In order to minimize the risk and stress on the family members and other caregivers, there has to be a comprehensive system to support those involved in instances of emergency. Most notably identified by family members and caregivers were the requirements for need-based assessment on a regular basis in the late stages of ALS. They also expressed need for a more cohesive structure of supportive care services without the gaps that currently exist, inclusion into the case management discussion that embraces the experiences of children related to the person with the condition, further emphasis on quality of life and future planning, incorporation of palliative care into the end of life plan, and extensive training in hospitals about dealing with individuals admitted with ALS (Dawson & Kristjanson, 2003). Through these experiences articulated by family members and caregivers of persons with ALS or other neurodegenerative diseases, there is a clear absence of support. The focus of ALS care rests in the physical treatment of the symptoms and less in the synthesis of a well-rounded plan to maintain QoL, as the person with ALS would want. If steps are going to be made to create a model for the progression of ALS, there must be an inclusion of support of family and caregivers.

### **Positive aspects of caregiving**

While the act of caring for a person diagnosed with ALS can be a taxing responsibility—both physically and emotionally—there are some positive aspects to the role of caregiver for both family and care team. In research done in 2015, positive reward was defined as “positive psychological experiences” (Weisser et al.) that were capable of

benefiting both individual with the condition and caregiver. A compilation of rewards listed after interview analysis with caregivers lists “being helped, receiving psychological support, feeling rewarded, and being able to enjoy moments of respite either alone or with friends,” (Weisser et al., 2015) as integral aspects to the positive care experience. The depth of connection built by being in a supportive and caring relationship with an individual with a terminal condition, such as ALS, was cited to have also brought the community closer together. Not only does this include the caregiver and family dynamic, but also a greater network of close friends and extended family rallying around a person with ALS to create supportive community (Weisser et al., 2015).

Interestingly, when describing the most challenging aspects of caregiving for an individual with ALS, the aspects listed often relate to the most rewarding. The distinction between burden, need, or reward is described quite fluidly throughout interviews with the caregivers. This particular aspect of caregiving is integral for inclusion into a multidisciplinary model. Because of the fluidity of psychological need, a model supporting the caregivers of those with ALS cannot address their states in a linear fashion. The model itself must be flexible enough to adapt to the specific burden, needs, and rewards a caregiver—or family member—is experiencing at any particular time in disease progression (Weisser et al., 2015).

Alternatively, there has to be some sensitivity to the specific methodology of psychological support for caregivers and family members. While family-centered support groups reported mostly positive experiences when becoming involved in self-help community meetings, non-relative caregivers often report negatively on the experience. Most commonly they cite feeling distressed being amongst a community of individuals

that are also either witnessing the deterioration of others or have neurodegeneration themselves (Locock & Brown, 2010). In these instances of differing reactions to support groups, there must be a secondary system of psychological care that is available to those who do not benefit from the vulnerability of an exposed group setting.

### **Healthcare system's failures**

The final aspect of family and caregiver dynamic I want to address is their relationship with the complexity of the condition, and thus the healthcare needs surrounding it. As the multisystem muscle degeneration and failure occurs across disease progression, the reliance on the healthcare system becomes greater. Without the adequate structure to support family and caregivers, the risk of becoming lost in the complexity of the healthcare system and ALS runs high.

In research interviews with family and caregivers, the lack of support from the medical community is cited as one of the greatest stresses on their roles. The access to services and information was reported as unsatisfactory in interviews with families of individuals living with ALS. Through the lack of support or guidance from the healthcare system, families often learned independently by way of their personal experience or the Internet. Articulating the frustrations of understanding what to do following diagnosis a wife of someone with the condition said, "they [health care professionals] gave me no information, no one explained to me what I should have done, no one explained to me what was the duration of the disease, what would happen," (Cipolletta and Amicucci, p. 5, 2014). The complete absence of information and aid not only endangers the individual diagnosed with ALS, but also automatically creates distance between the healthcare system and the community dealing with the psychological repercussions of diagnosis.

With the structure of a healthcare model, the hope is that it will not only promote a fully encompassed perspective of ALS, but also a better communication system between all parties. Without it, the community of those diagnosed with ALS run the risk of becoming unpaid laborers of the healthcare system—independently seeking out information, managing a multifaceted care plan with multiple doctors, and creating distance between medical professionals (Cipolletta and Amicucci, 2014).

## **Construction of a model**

ALS presents a significant challenge for end of life care. Previously developed established models for terminal cancer, chronic disease management, or end of life care do not completely address the complexity of ALS as a treatment plan should—making them inadequate for use on the condition. According to research done on ALS, the average life expectancy following symptom onset is approximately two to five years (Chico et al., 2009; Hodgen et al., 2015); this expresses a narrow window for a rapidly progressive disease that exhibits severe expression of symptomology at the terminal end of the condition. Despite the clear physical symptoms of ALS, diagnoses rarely happen at the onset of symptoms. Additionally tightening the window for decision-making action are the communication difficulties that occur in nearly all persons with the condition because of the restriction of movement in the vocal cords, tongue, and throat. In order to mitigate the strain felt by individuals diagnosed with ALS, their family, and their caregivers, research has begun to delve into setting up effective and patient-centered decision-making models for the ALS community. Research shows that the early introduction of palliative care measures helps ease the burden of both the individual with

ALS and the surrounding community members—usually family, friends, and caregivers (Chio et al., 2009).

Research has initiated investigation into the development of a model inclusive of persons with ALS, caregivers, and familial input to create a multidisciplinary care program for those diagnosed with the condition. The researchers involved were motivated to create early decision-making processes that lessened the strain on persons with ALS and those surrounding them (Hodgen et al., 2015). Because of the symptomology of ALS there is a constant practice of bouncing around between different medical doctors. The action of having to constantly move from hospital to private practice to other different healthcare offices adds only more stress to the treatment of this already distressful condition. Through the methodology within this study, the researchers were able to establish a rough idea of the most effective and preferred treatment options for individuals with ALS. Following the semi-structured interviews, four interwoven disease-progressive stages aided the development of a specialized multidisciplinary model strictly for the ALS community. As persons with ALS move in and between these stages, the model suggests support of the types of decision-making processes the involved individuals are moving through based on the progression of the disease (Hodgen et al., 2015).

Perhaps most integrally located within the discussion is the following statement: “effective and patient-centered decision-making is enacted collaboratively by patients, [caregivers], and health professionals within the environment of the specialized multidisciplinary clinic,” (Hodgen et al., 2015, p 1779). The development of Hodgen’s model of decision-making processes is integral, but the formation of multidisciplinary

healthcare centers that are inclusive of support communities directly exposed to ALS implements structure into practice. Through support of persons with ALS, the strain of the decision-making process is minimized and the surrounding caregivers and family are able to respect a plan created by and for the individual progressing through the condition.

Thus when developing multidisciplinary, multifaceted model to support the communities of individuals with ALS, it is integral to provide a basis for the person diagnosed with the condition to have control over their EoL conditions. Providing physical and psychological support around autonomous control for as long as possible will help set up a plan for disease progression, introduction of palliative care, and the necessary emotional care for the entire community. Given the nature of the condition, this has to be highly flexible. The intervals from diagnosis to death range from less than a year to decades, which emphasizes the need to have discussions surrounding end of life care as soon as possible.

As discussed in the literature review, communication is a key weakness cited in the relationship between the community surrounding the person diagnosed and the healthcare system. From this initial conversation (or conversations), the needs of the person living with ALS and the caregiving community can be assessed and weighed into consideration. Moving forward, the healthcare team should be able to adequately develop a plan based on their wishes and work with them going forward as muscle degeneration progresses.

ALS is a vastly complex disease. As demonstrated by the literature discussed within this piece, there are several broad areas of study where research is being done. But

there is very little that attempts to synthesize a care strategy inclusive of planning for the progressive muscular degeneration characteristic of the condition, palliative care steps needed for a terminal condition, and psychological support for the person diagnosed with ALS and their community. Most frequently, because of the visceral physical manifestations, ALS is treated solely on a physical level and the psychological health of those with the condition and their community is often put on the back burner. This piece addressed the variables affecting the under acknowledged emotional care of an individual living with ALS, as well as the community that supports and cares for them.

In creating models for a multisystem disease, our medical communities must develop a multisystem, comprehensive approach that suits the needs of each individual case. Applying a restrictive, ill-fitting methodology to the treatment and support of those with the disease only results in the erasure of important components of ALS care-- psychological care, caregiver support systems, early palliative care and end-of-life discussions, and many other aspects of the condition that do not deal directly with muscular degeneration. The complexity of a terminal condition should be inherently inclusive of psychological support, especially if it includes the level of complexity found in ALS symptomology; to disregard this aspect of care could be neglectful of the individual with the condition, their family, and their caregiver(s). Hopefully, by creating a system of solid early communication with medical professionals, plans prioritizing personal control and decision-making, physical and psychological care, and consistent check-ins with medical professionals will pave the way for a better supported process.

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## Appendix A

### Articulation of LMN and UMN Severity Scores in Relationship to Region of Onset

**Table** Composite severity scores showing distribution of motor neuron deficits indexed to

Body region of onset	Body region evaluated	Composite LMN severity scores
Bulbar (n = 29)	Bulbar*	102
	Both arms	40
	Both legs	8
Arms (n = 34)	Onset arm (focus)	85
	Contralateral arm	41
	Ipsilateral leg	14
	Contralateral leg	6
	Bulbar	6
Trunk (n = 6)	Trunk*	24
	Both arms	17
	Both legs	8
Legs (n = 29)	Onset leg (focus)	66
	Contralateral leg	37
	Ipsilateral arm	19
	Contralateral arm	16
	Bulbar	4

\*Scores are doubled to normalize with limbs.

LMN = lower motor neuron; UMN = upper motor neuron; NA = not applicable.

Ravits et al., 2007