ALS AND FAMILY FUNCTIONING

by

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Amyotrophic Lateral Sclerosis (ALS) is the most common progressive and fatal neurodegenerative disease and yet a cure remains unknown. Given the growing number of patients with ALS, it is imperative that behavioral health clinicians, medical providers, researchers, and policy makers are aware of their and their support persons' unique biological, psychological, social, and spiritual health concerns, the impact of the disease on family functioning, and the general care management of those living with ALS. This dissertation was written to help identify the impact of ALS on patients and their support persons' family functioning and biopsychosocial-spiritual (BPS-S) health. It includes three manuscripts: (a) a systematic review of the impacts of ALS on immediate family members' health and interactional patterns, (b) a descriptive phenomenological study designed to explore the lived experience of eight patients and nine support persons living with ALS, and (c) a perspective piece that presents an argument for requiring increased competencies in medical education and residency education programs around ALS diagnosis, treatment, and family-centered care. The systematic review resulted in 3,557 articles with 3,509 meeting exclusion criteria and 48 meeting inclusion criteria for full review. The articles in the systematic review demonstrated that previous literature on the BPS-S health outcomes of support persons of patients with ALS is disjointed, with the health components studied independent of one another rather than simultanesouly for a comprehensive

understanding. The literature reviewed also revealed a limitation in the amount of research that has focused on the impact of ALS on family functioning. The phenomenological study conducted as a part of this dissertation resulted in five themes relevant to patients and support persons' experiences living with ALS: (a) Dynamic transformations of relational systems, (b) Biological changes and well-being, (c) Emotional processes, (d) Impacts on spirituality, and (e) Healthcare system interactions. The novel findings of this study include: (a) a broader understanding of patients' with ALS (PALS) and SPs' experiences with the impact of ALS on family functioning due to the inclusion of multiple participants in the same interview; (b) participants explanation of progression of physical symptoms and subsequent decrease in verbal communication abilities resulting in a more isolated and frustrated state; (c) participants reporting on their ability to reach acceptance and choose positivity; (d) and the participant's expressing the need for medical providers outside of ALS clinics to have increased knowledge of proper ALS care. It is then recommended that future research should focus on the immediate and longitudinal whole health needs of PALS and SPs and their interactional effects, participant's perspectives on the lack of knowledge by non-ALS specialists, and studies that continue to include multiple family members and SPs conjointly with the PALS to ensure that more robust systemic perspectives are represented. The final manuscript is a perspective piece based on existing literature in combination with findings from this disseration. It is used to highlight the need for increased education among medical professionals on ALS diagnosis, interdisciplinary treatment, BPS-S and family-centered care

ALS AND FAMILY FUNCTIONING

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In Partial Fulfillment of the Requirements for the Degree

Doctor of Philosophy in Medical Family Therapy

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DEDICATION

For Will. You have made countless sacrifices in your support of my education throughout the years. I cannot thank you enough for your continuous patience, your steadfast voice of reason, and your ceaseless encouragement. I truly could not have completed this without you.

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To my uncle Paul, who stayed forever positive and kept us all laughing in spite of battling ALS. Your strength throughout your illness encourages me to keep fighting. A cure is out there and we'll never give up.

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PREFACE

In the summer of 2001, my mother's youngest brother was diagnosed with Amyotrophic Lateral Sclerosis (ALS) at the age of 40. At the time, my family, and virtually everyone we knew was unfamiliar with this disease. At 10 years old, I taught myself as much as I could about the disease and shared that information with as many others as possible. I did not want our family to feel alone in our experience and felt that if I could teach others about what we were going through, then we would no longer feel alone. After four and a half years of living with ALS, my uncle died from the disease and I lost the energy to fight against what I had come to consider a losing battle.

Twelve years following my uncle's death, in the summer of 2017, I was preparing to enter the Medical Family Therapy doctoral program at East Carolina University. I spent that summer attempting to determine what I hoped to focus on in terms of research during my time in the program. During that summer I found an old high school project in which I had to make a one-year, five-year, and ten-year plan for myself, which included assisting in finding a cure for ALS. Finding that document re-kindled my passion for research related to ALS. Although I would not be working on a cure, I knew that my research as a Medical Family Therapist would allow me to connect with families living with ALS and learn from them so that new interventions and therapies could be developed that might alleviate some of the suffering that comes from living with this terminal illness.

As I committed to this topic in the beginning of my time in the doctoral program, I quickly realized how much growth would need to happen for me to be successful in this endeavor. I was able to cling to the basic knowledge of ALS that I had learned several years ago and stretched that knowledge by centering each assignment around the topic so that I was

learning everything there was to know about the disease. This growth not only happened in my literal knowledge of ALS but grew my understanding of the impact this disease had on my own family and on myself. While enrolled in the doctoral program, I wrote several papers about ALS and completed a systematic review (Chapter 2) on the biopsychosocial-spiritual (BPSS; Engel, 1977; 1980) impacts of ALS on the patient and their familial caregivers.

The results of the systematic review demonstrated the void of research relating to how these BPSS (Engel, 1977; 1980) components interact with one another in families with ALS, how the majority of ALS research focuses solely on the patient, and that the qualitative research on ALS is quite scarce. I was able to determine then that a qualitative study would allow for these patients and their families to more accurately describe their experiences with ALS, expanding and enhancing what has already been done quantitatively. A qualitative study would allow these families to describe in their own words, the impact this disease has had on their family as a whole, a gap in the literature. It would give these families a voice.

Throughout my time working on this dissertation and following several conversations with my mentor, Dr. Jennifer Hodgson, I have come to understand that all of the hurdles, hiccups, and setbacks I have encountered during this journey are not all that important. What is important is reminding myself why I chose this area of research in the first place, to better understand and support the patients and families living with ALS. My hope is that my research will fill important gaps in the literature by providing a voice to patients with ALS and their families and support persons together. I also hope that this research will provide insight into how ALS is impacting the entire family unit from a BPSS perspective (Engel 1977; 1980) and how family functioning (Beaver, 1981) is changed by this progressive disease.

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CHAPTER 1: INTRODUCTION

Amyotrophic Laterals Sclerosis (ALS) is a relentlessly, progressive illness with no known cause or cure (Pagnini et al., 2012). Characterized by sudden onset, possible cognitive and behavioral impairment, and inevitable physical decline, the majority of ALS-related deaths occur within three years of onset (Galvin et al., 2016). The disease progression involves degeneration of the upper and lower motor neurons, which control muscular activity, causing progressive muscle weakness, atrophy or wasting away, and spasticity or continuously contracting muscles that leads to paralysis and eventual death (Calvo et al., 2015). Currently, the best treatment option involves palliative care (maintaining independence as long as possible, managing pain, and providing relief from stress and symptoms) (Aoun et al., 2013; Galvin et al., 2016), leaving the patient, family and healthcare system to navigate a difficult path of illness management. The following chapter will provide general information about the incidence rates, age onset and progression rates, symptoms, and diagnosis of ALS, as well as information on the impact of ALS on the systems surrounding them. It will conclude with a description of the proposed dissertation study, implications for the Medical Family Therapy field, as well as descriptions of each of the chapters included in this dissertation.

Incidence Rates

The lifetime risk of ALS has been estimated to be 1 in 400 (Larsson, Fröjd, Nordin, & Nygren, 2015) with approximately 5,600 patients newly diagnosed patients each year in the United States (Cruz et al., 2018). Incidence seems to occur more commonly in men than women, with an average male to female ratio of 1.5:1 (Kiernan et al., 2011; Wijesekera & Leigh, 2009). One possible explanation for the difference is women have protective hormonal factors leaving men at an increased likelihood of being exposed to the supposed risk factors (Wijesekera &

Leigh, 2009). Onset of the ALS typically occurs between the ages of 47 and 63 years (Kiernan et al., 2011) with only 5% of cases developing before 30 years (Wijesekera & Leigh, 2009). However, juvenile onset cases are on the rise.

Disease Onset and Progression

ALS is mercilessly progressive and terminal, with an average disease course of one to five years (Boillée, Vande Velde & Cleveland, 2006). As many as 50% of patients die within 30 months of symptom onset and up to 20% survive between five and ten years (Kiernan et al., 2011). Older age at symptom onset and respiratory muscle dysfunction have been related to shortened lifespan, while younger age at symptom onset and limb-onset of the disease are predictors of prolonged life spans (Kiernan et al., 2011). Patients with ALS (PALS) with bulbar onset (i.e., difficulties with speech and swallowing prior to difficulties with other body parts) have a median survival time of 26 months while PALS with limb onset (i.e., muscle atrophy of the legs and arms prior to other areas of the body) have a median survival of 32 to 33 months (Fujimura-Kiyono et al., 2011). It is also important to note that older adults and females tend to have higher rates of bulbar onset (Yates & Rafiq, 2016).

Symptoms

The most common presentation of ALS is limb-onset (70% of patients), which includes a combination of upper and lower motor neuron signs present in both the arms and legs (Kiernan et al., 2011). Upper motor neuron (UMN) difficulties may include spasticity, muscle weakness, and/or brisk, deep tendon reflexes. Lower motor neuron (LMN) include difficulties with muscle atrophy or deterioration and muscle weakness in the limbs. The alternate presentation is bulbar onset (25% of patients) where slow, labored, and distorted speech, gag and jaw jerks, tongue wasting, and dysphagia or difficulty swallowing symptoms appear. Due to the existence of

several disorders that mimic ALS, a thorough diagnostic assessment is required (de Carvalho et al., 2008; Kiernan et al., 2011; Wijesekera & Leigh, 2009).

Diagnosis

The diagnosis of ALS is based on the presence of several clinical findings (Kiernan et al., 2011). The 'El Escorial' diagnostic criteria was developed in 1994 but was revised during the 2000 Airlie Conference to increase the sensitivity of the criteria. Subsequently, it was renamed the Airlie House diagnostic criteria (Brooks, Miller, Swash, Munsat, 2000). The Airlie House criteria for an ALS diagnosis included: (a) the presence of LMN degeneration by clinical, electrophysiological or neuropathological examination, (b) UMN degeneration by clinical examination, and (c) progressive spreading of symptoms without evidence of other disease processes (Brooks, Miller, Swash, & Munsat, 2000). Based on these criteria, patients were classified into 'clinically definite,' clinically probable,' 'clinically probable-laborartory supported,' and 'clinically possible,' excluding the classification of 'clinically suspected,' which was previously used in the 'El Escorial' diagnostic criteria (Wijesekera & Leigh, 2009).

The most recent diagnositic criteria, the Awaji-shima criteria, was developed in 2006 in Awaji-shima, Japan and again simplifies the diagnositic criteria (Wijesekera & Leigh, 2009). The Awaji-shima criteria made the following two changes to previous diagnostic criteria, (a) electromyography (EMG) analyisis is no longer considered a secondary consideration, but carries the same weight as a physical exam, and (b) the Awaji-shima criteria expanded the kinds of EMG data that can indicate ALS to include fibrillations (loss of nerve connections) and fasciculations (earlier loss of nerve supply)(de Carvalho et al., 2008). These changes in the interpretation of data render obsolete the 'clinically probable-laboratory supported' category and have been found to improve diagnosis specificity (Carvallo & Swash, 2009; de Carvalho et al.,

2008). The complexity of how ALS presents and is diagnosed rivals in comparison to its overall impact on the patient and those positioned to provide essential caregiving.

ALS Caregiver Strain

Multiple studies on caregiver burden associated with ALS, confirm that caring for a loved one with a neurological illness is a source of psychological distress that may impair quality of life (Burke et al., 2015; Galvin et al., 2016; Lillo, Mioshi, & Hodges, 2012; Pagnini et al., 2010; Rabkin, Wagner, & Del Bene, 2000). Pagnini et al. (2010) reported that ALS caregivers experience extreme emotional and physical problems due to the steady progression of the disease and lack of effective treatment. Due to the average age of onset being between the ages of 47 and 63 years, caregivers are usually the patients' spouses, but at times this duty may fall on the children (Kiernan et al., 2011; Tramonti, Bongionni, Leotta, Puppi, & Rossi, 2015). Previous studies, specifically on families with ALS, have determined that there is a high need for ongoing family support and that studying the evolution of specific diseases may be helpful in evaluating the impact of the disease on family life (Martin & Turnbull, 2001; Tramonti et al., 2015). In fact, health researchers studying family functioning in families of terminally ill patients, found successful intervention may ultimately lead to reductions in reported caregiver burden and its negative consequences (Arestedt, 2013; Tremont et al., 2006). However, ALS research lacks connection to theories and research that helps make sense of the systemic and relational impact patients and their family members experience while navigating this progressive fatal disease.

Theoretical Conceptualization

The proposed dissertation will utilize general systems theory (von Bertalanffy, 1950; 1967), augmented by Beaver's systems model of family functioning (Beavers, 1981), and the Biopsychosocial-spiritual (BPS-S) framework (Engel, 1977; 1980; Wright, Watson, & Bell,

1996) to further understand the impact of ALS on family functioning and the BPS-S impact it has on PALS family members' health. While von Bertalanffy identified in general systems theory how living systems depend on the constant interchange of resources within its environments (von Bertalanffy, 1950; 1967), Beaver's systems model of family functioning helped to expand it by encouraging researchers and therapists to systemically study family functioning and competence in families (Beavers, 1981). The combination of general systems theory and Beaver's systems model along with the BPS-S framework facilitates a whole health (biological, psychological, social, and spiritual) view of ALS, increasing the comprehensiveness of research that is informing its treatment.

General Systems Theory

Through his general systems theory, Ludwig von Bertalanffy identified how living systems depend on the constant interchange of resources within its environments (von Bertalanffy, 1950, 1967). His ideas helped to think of systems as being more than the sum of their parts, but a product of the interaction within and among the parts and other systems surrounding it. He identified how the concepts of equifinality, equipotentiality, and homeostatic reactivity determine how and why systems initiate change, struggle to complete the process, and adapt accordingly (von Bertalanffy, 1967).

The utilization of general systems theory as a grounding theory for the current dissertation proposal emphasizes the importance of including the entire family system, to include the patient and family members, during the treatment of patients with ALS. Within general systems theory, equifinality suggests that the same final state may be reached from different initial conditions and in different ways (von Bertalanffy, 1967). Equipotentiality suggests that all parts of the system exhibit uniform potential at every point and homeostatic reactivity refers to

the mechanism within systems that seeks to maintain balance within the system (von Bertalanffy, 1967). These terms indicate that all family systems are able to reach a final preferred state from all varieties of initial starting points, that all members within family systems possess equal potential, and that the family system seeks to reach a healthy, balanced state. The inclusion of systems allows for the recognition that growth within the patient's system depends on the constant interchange of all of the available resources (von Bertalanffy & Sutherland, 1974).

Beavers Systems Model of Family Functioning

According to Beavers and Hampson (1993), the Beavers Systems Model was developed to help define core family functioning constructs and differentiate relationally and interactionally healthy families from less healthy families. Two of the central concepts of the Beavers Systems model are (a) family functioning, which includes the observable, live, interactive functioning within a family, and (b) family competence, which ranges from effective, healthy family functioning to severely dysfunctional patterns (Beavers & Hampson, 1993). It also includes the concept of family style, which refers to the degree of centripetal or centrifugal qualities in the family. Members in centripetal families seek satisfaction from within the family and pull inward and lean on one another when a stressor occurs; while, centrifugal family members look for satisfaction from the outside world and pull away from others within the family when stressors are experienced.

According to the Beavers Systems Model (Beavers, 1981) there is a continuum of family functioning. On one end are optimal families who try many approaches to resolve problems within the family, are usually capable of achieving intimacy, and quickly resolve conflict among them. On the opposite end are dysfunctional families who disengage from one another, have poor boundaries, and are extremely limited in negotiating stressors and stressful interactions with one

another. With families existing along this entire continuum, the model can be useful to understanding patterns of functioning and how different styles impact the management of ALS, identifying opportunities for further research and intervention. Combining this model with the biopsychosocial-spiritual framework (Engel, 1977, 1980; Wright et al., 1996), discussed below, allows for clinical work and interventions to be tailored to all families from optimal to severely dysfunctional with a focus on whole health.

Biopsychosocial-spiritual Framework

The Biopsychosocial (BPS) model is a systems-theoretical construct rooted in the writings of biologists and general systems theorists, Paul Weiss and Ludwig Von Bertanlanffy (Schubert, 2010). It proposes that biological, psychological, social components operate simultaneously in connected subsystems that influence all aspects of mental and physical health (Engel, 1977; 1980). The spiritual component was highlighted by Wright et al. (1996) and includes the importance of a person's spirituality in relation to their holistic health, giving way to the biopsychosocial-spiritual (BPS-S) framework.

It has become increasingly common for complex biomedical problems, such as ALS, to be studied by multidisciplinary teams of researchers (Suls, Krantz & Williams, 2013). As a result, multidisciplinary models of care have emerged reducing the risk of death from ALS by 45% at five years (Kiernan et al., 2011). According to Kiernan et al. (2011), multidisciplinary models of care are found in more specialized clinics as opposed to more general neurology clinics that might include treatment provided by neurologists only.

Currently, the majority of research regarding the treatment of ALS seems to primarily focus on biological treatments and clinical trials that seem to delay the physical progression of the disease (e.g., Kim et al., 2018; Luo et al., 2019; Rosenfield, 2018). Several researchers have

reviewed the impact of ALS on the psychological, social, and spiritual well-being of patients, yet there has been little to no investigation on the impact these treatments have on the entire family unit and their BPS-S health. Few research teams have investigated the BPS-S health of PALS and family members at the same time or examined the overall family functioning and health impact on family members, and have instead focused solely on the person with the ALS diagnosis or the individual family caregivers.

Purpose and Design

Although there is an abundance of literature available on ALS and PALS, there are clearly gaps in the literature regarding the impact ALS has on the whole health of the family, and on the overall functioning of families living with ALS. Whereas previous studies have investigated the individual BPS-S impacts of ALS on family members and PALS (Ozanne et al., 2015; Ozanne & Granneheim, 2017), none have identified how those health domains function synergistically. Furthermore, research looking at the BPS-S distinctions between bulbar and limb symptom onset seems largely understudied as well.

Ultimately, variations in the pattern of spread based on symptom are not well known, but the recognition of these different ALS subsets, bulbar and limb, is important as prognosis may vary depending on the subset (Predat & Bruneteau, 2008; Turner, 2010). Specifically, cases with bulbar onset are traditionally regarded as showing a poorer prognosis with associated symptoms including aspiration pneumonia, impaired quality of life, reduced will to live, and malnutrition (Fujimura-Kiyono et al., 2011; Lou, Moore, Gordon, & Miller, 2010; Marin et al., 2011; Tysnes, Vollset, Larsen, & Aarli, 2014). As the subsets present differently, a greater understanding of the pattern of symptom spreading for each will assist with developing more comprehensive and appropriate treatment plans (Fujimura-Kiyono et al., 2011).

Finally, only limited research has been conducted on how ALS impacts family functioning (Tramonti et al., 2014). To adequately address these gaps in the ALS literature, Beaver's systems model of family functioning (Beavers, 1981) and the BPS-S framework (Engel, 1977; 1980; Wright et al., 1996) were utilized within this dissertation study. If families are expected to provide essential ALS caregiving, addressing family functioning patterns and their influence is an important part of better understanding their lived experience with ALS.

Since little in know about how family functioning is impacted among families living with ALS, a qualitative design was used. More specifically, a phenomenological approach was utilized to explore the experiences of families living with ALS. The research question guiding this study was, "What are the experiences of patients - and their identified primary support persons - living with ALS and how has it impacted family functioning and health?" The study consisted of semi-structured phenomenological interviews with the PALS and the PALS' identified primary support person outside of their professional support system. The term support person(s) (SPs) is utilized throughout to identify any person outside of the PALS' professional support system that provides any support to the PALS. This qualitative study aligned with Husserl's (1970) descriptive phenomenological approach and utilized Colaizzi's (1978) method of data analysis.

Implications for the Medical Family Therapy Field

The research derived from the field of Medical Family Therapy (MedFT) illustrates considerable value as it contributes to an area of literature in which there is inadequate representation of the relational systemic voice. As a field, MedFT stresses the importance of collaboration between and among healthcare providers, the patient, and the patient's relationships. Furthermore, the competencies for family therapists working in healthcare settings

reinforces the importance of integrating and implementing systems theory, the BPS-S perspective, collaboration, ethics, diversity, in the delivery of integrated care (AAMFT, 2018). Given the definition and competencies listed, this dissertation provides clinical, research, and policy-based implications for the field of MedFT along recommendations with advancements in the training, clinical protocols, policies, and study of ALS.

Clinically, this dissertation argues that care should include family members and support persons in treatment to ensure treatment is provided from a relational systemic perspective (von Bertalanffy, 1950). It supports through a comprehensive systematic review and original study the need for ALS and non-ALS specialists to engage in higher levels of integration and researchers to conduct comprehensive BPS-S studies. Foundational to all of this are the main goals inherent to the field of MedFT: agency and communion (McDaniel et al., 1992). Agency is the active involvement in and commitment to one's own health care by making personal choices in dealing with illness and the health care system and communion is the sense of being cared for, loved, and supported by family members, friends, and medical providers (McDaniel et al., 1992). This dissertation is set to advance agency and communion by encouraging PALS to have a voice in their experiences with ALS and to access supportive systemic relationships to support them in those efforts.

From a research perspective, this dissertation was constructed to focus on the BPS-S health of patients with ALS and their family members/support persons. Currently, research studies conducted to study the family experience of ALS, with more than one family participant enrolled, are limited (e.g., Tramonti et al., 2014). As found from the systematic review conducted, research from this perspective would advance awareness regarding the BPS-S impacts of those living with ALS so policies and treatment plans can better align with a

comprehensive care framework grounded in systemic data. The original study conducted for this dissertation adds to the body of evidence that patients with ALS and their family members prefer and benefit from a comprehensive BPS-S focused care, experience changes in their family functioning as a result, and need their healthcare providers to collaborate effectively to help them in better managing it.

Finally, from a policy and education/training perspective, leaders in the field of MedFT could utilize the arguments presented in the dissertation to promote ALS family-centered policies and encourage more training about ALS during and beyond medical school. Specific policies informed by the systematic review and original study could be established for ALS treatment that utilize the BPS-S framework, offer family-centered care guidelines, strengthen the interdisciplinary team approach, and expand and enhance curriculum about ALS available to all levels of medical professionals.

Summary

This dissertation was designed to expand and enhance the ALS literature, furthering the research on how family functioning and BPS-S health is impacted. General systems theory (von Bertalanffy, 1950) augmented by Beaver's systems model of family functioning (Beavers, 1981) and the BPS-S framework (Engel 1977; 1980; Wright et al., 1996) formed the theoretical base for the study, attempting to explain how health and the functioning of families should be studied systemically and relationally. This focus will assist multidisciplinary clinicians in the development/implementation of future interventions for PALS and their families, generating evidence that is grounded in systemic and relational science and not just individually focused studies. Throughout this dissertation, the term support person was used to reference any family members, caregivers, or other important persons involved with the PALS. The following

chapters include information in regard to the anchoring literature, methodology, results, and implications for the field of Medical Family Therapy, as well as future ALS research, clinical applications, and policies.

The second chapter is a systematic review of the literature of the BPS-S impact on family members and caregivers of PALS and on the overall family functioning. The research question guiding the systematic review was, "How does ALS impact the immediate family members' health and interactional patterns?" In this chapter the support persons and family members are referred to as caregivers of PALS (CALS). The systematic review was conducted by the lead investigator and one co-investigator across three databases (PubMed, PsychINFO, and CINAHL Plus with Full Text) and resulted in 48 articles that met inclusion criteria. Of the final 48 articles included in the review, 21 studied at least one BPS-S variable of the caregiver's health, 13 used two, ten used three, and three used all four, but none of the articles identified how the BPS-S variables may be impacting one another. Additionally, only two articles included in the review addressed family functioning, with results indicating that some family members (15%) felt as though the disease had been detrimental to family relationships, while others (67%) felt as though the disease brought them closer together (Martin & Turnbull, 2001) and that family members tended to become problematically close (Tramonti et al., 2015). Overall, the results of this systematic review illustrate how family caregivers of PALS are impacted biologically, psychologically, socially, and spiritually. Based on the reviewed articles it is also clear that ALS has an impact on the family functioning although few studies have explored patterns related to subset of ALS onset, progression, and access to resources. These results reinforce the need for future studies to determine how family caregivers of PALS complete BPS-S health and how the family's functioning is impacted by and influences treatment planning for ALS.

The third chapter consists of a literature review of symptom onset and BPS-S symptoms in ALS. This chapter provides a synopsis on what information is currently available on bulbar and limb onset in ALS, to include any differences that PALS and their families may encounter in their ALS experience based on symptom onset. This chapter also presents information on how each component of the BPS-S framework is impacted in PALS and their family members and family caregivers. Physical symptoms represent the biological component, mental health symptoms (i.e.; anxiety and depression) represent the psychological component, social support the social component, and religious and spiritual changes the spiritual component. Finally, this chapter provides recommendations for future research informed by the literature reviewed.

The fourth chapter responds to the call for more research by laying out the methodology for this dissertation's phenomenological qualitative study. The qualitative inquiry method followed a Husserlian phenomenological approach (1970) with the interview data analyzed according to Colaizzi's (1978) phenomenological analysis method. The research question guiding the study was, "What are the experiences of patients and their identified primary support persons living with ALS and how has it impacted family functioning and health?" Purposive sampling (Patton, 1990) was used in the study to enroll participants from a Southeastern academic medical center ALS clinic. Once participants were enrolled in the study and had consented to participation, a semi-structured interview guide (Bernard, 1988) was used to provide an interview in which all participants addressed similar topics related to their experiences with ALS. Verification strategies used to ensure the trustworthiness of the data included: (a) credibility, (b) transferability, (c) dependability, and (d) confirmability (Lincoln & Guba, 1985).

The fifth chapter, written in manuscript format, reports the qualitative results of the dissertation's phenomenological study outlined in the previous chapter. Eight participant groups, consisting of one PALS and at least one identified primary support person outside of their professional support system participated in in-depth, open-ended, semi-structured interviews. The data was then analyzed using Colaizzi's (1978) method of data analysis, revealing 278 final significant statements which were then collapsed into 166 formulated meaning statements. These meaning statements were then grouped into 15 thematic clusters aligning under 5 emergent themes: (a) dynamic transformation of relational systems; (b) biological changes and well-being; (c) emotional processes; (d) spiritual adaptations and anchors; and (e) healthcare system interactions. An exhaustive description of the study's findings highlighted how relational systems adapted to the illness by either coming together or pulling away from one another, weathered difficult social and emotional challenges often accompanied by a sense of acceptance of the ALS journey, involved a sense of spiritual well-being, and expressed appreciation for ALS Clinics and their specialists. It also captures participants' desires for greater professional support during the diagnosis period and preparedness among providers outside of ALS clinics to detect, diagnose, and treat ALS, as well as and collaborate more often with ALS specialists.

The sixth and final chapter serves as the dissertation's implications chapter and is written as a policy manuscript to be submitted to a medical education peer reviewed journal. It utilizes information gathered from the participant interviews during the phenomenological study, as well as the systematic review, to establish an argument for the expansion of ALS related competencies in medical school, residency programs, and continuing education curriculums. A growing shortage of neurologists (Guttman et al., 2019), increasing number of patients with ALS (Arthur et al., 2016), and findings among existing studies reviewed and conducted in this

dissertation point to the need for a revision in the detection and treatment of ALS to maximize quality of life for PALS and their families/support persons. The chapter also recommends that ALS education be expanded and enhanced for those who are not ALS specialists. This education would include diagnostic awareness, symptom management, interdisciplinary collaboration methods, BPS-S and family-based care protocols, as a way to promote more seamless care between ALS Clinics and other specialists caring for the ALS patient and their family.

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CHAPTER 2: A SYSTEMATIC REVIEW OF THE IMPACTS OF ALS ON IMMEDIATE FAMILY MEMBERS' HEALTH AND INTERACTIONAL PATTERNS

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Amyotrophic lateral sclerosis (ALS), more commonly known in the United States as Lou Gehrig's disease, is a progressive, fatal neurological disease with no known cause or cure [1]. The disease involves degeneration of the upper and lower motor neurons, which control muscular activity, causing progressive muscle weakness, atrophy or wasting away, and spasticity or continuously contracting muscles [2]. In the United States as many as 30,000 live with ALS, with estimates of nearly 5,600 diagnosed annually [3] and an average age of onset between 47-63 years [5,6]. Although much is known about the impacts of ALS on the patient directly, little research has been done on the impact of ALS on family functioning and family member's biological, psychological, social, and spiritual health [2,4].

Studies focused on caregiver burden found caring for a partner or family member with a neurological illness is a source of psychological distress that may negatively impact one's quality of life [7-11]. For example, Pagnini et al. [10] reported that ALS caregivers often experience extreme emotional and physical problems due to the steady progression of the disease and delayed treatment. ALS interventionists recognize that caregivers need emotional, psychological, spiritual, and social support to help maintain their role and their own health [10, 12, 13]. However, role changes and restructuring in the family are commonly experienced as diseases progress [17]. For some families the illness may bring members closer together while others become more distant from one another [17-20]. Based on these studies, and others with similar findings, it is argued that family functioning in families of terminally ill patients may increase or

decrease caregiver burden and negative consequences [17, 19] and therefore is important to the development of research-informed protocols for assessing and treating families living with ALS [12].

Theory is critical for making sense of scientific observations. General systems theory [21, 22] identifies how living systems depend on the constant interchange of resources within its environments. Beaver's systems model of family functioning is an expansion on general systems theory, arguing that a more flexible and adaptive family is able to effectively deal with higher stress situations and is not bound to rigid behavior patterns and responses, allowing instead for more freedom to evolve and differentiate [23]. It is a method of explaining family competence and family style by examining family patterns. For example, families experiencing a health event may demonstrate centripetal behaviors (finds the most relationship satisfaction within the family) or centrifugal (finds the most relationship satisfaction outside the family) [23]. When faced with terminal illnesses such as ALS, centripetal families would likely respond by pulling inward and huddling together; whereas centrifugal families would likely respond by moving away and scattering apart [24]. Therefore, it is important to understand how family patterns and functioning impact patient and family members' health and what parts of health are most effected.

The biopsychosocial-spiritual (BPS-S) framework [14-16] helps researchers and clinicians make sense of the systemic implications of illness by viewing health through a more comprehensive lens. Approaching care from a BPS-S approach has emerged as a predictor of survival, reducing the risk of death in ALS patients by 45% at five years [5]. The combination of general systems theory and Beaver's systems model along with the BPS-S framework supports

understanding how ALS impacts the family system as a whole. It fills a gap in the science and literature by looking at health research more comprehensively.

This systematic review was designed to help answer the following research question: "How does ALS impact the immediate family members' health and interactional patterns?" More specifically, the aims of this review are to: (a) conduct a systematic review of available peer-reviewed literature where at least one component of the BPS-S framework was studied on at least one immediate family member of a person(s) with ALS (PALS), with immediate family members including the PALS' spouse or partner, parents, children and grandchildren, siblings, and self-identified family members, (b) identify themes for centripetal and centrifugal factors related to BPS-S health outcomes for the immediate family members of PALS, and (c) provide clinical and research recommendations to help understand and address BPS-S factors related to PALS and their immediate family members based on the family type, centripetal or centrifugal.

Methods

This systematic review was guided by Cooper's [25] seven-step systematic review model. Step-one [25] involved identifying the problem and formulating the research question as noted above. Step two [25] included systematically sifting through the existing literature using three academic databases (e.g., PubMed, PsycINFO, and CINAHL Plus with Full Text) to identify titles and abstracts that may be admitted into the review. Key terms were used in conjunction with medical subject headings (MeSH) during this step. To ensure consistency of the search strategy across databases, the lead author consulted with health science and information services' librarians to develop optimal MeSH and keyword search term strategies for each database (See Table 1, MeSH and Keyword Search Terms).

During step two [25] the lead investigator and a sub-investigator utilized the final MeSH and keyword search terms to conduct searches for articles within each search engine. From that initial search, 3,557 articles were identified as potentially appropriate. Each investigator was responsible for reviewing these article titles using Rayyan QCRI, an online tool designed to assist authors in the systematic review process [26]. When using this application, each investigator's decision was visible to the other investigator. Any decision discrepancies were automatically identified and discussed by both investigators so that a final decision could be reached. Upon conclusion of this step, 3,371 articles were excluded as their titles indicated they were not a good fit, 76 of those being excluded based on duplication, resulting in 110 articles admitted for abstract review.

Step three [25] included the identification of inclusion and exclusion criteria applied to potential articles identified from the searches. The inclusion criteria for title and abstract review were as follows: (a) articles that include at least one immediate family member of a PALS, (b) at least one BPS-S component of health studied, (c) qualitative and quantitative peer-reviewed journal articles, and (d) published in English. The exclusion criteria were: (a) cellular and genetic studies about ALS as the purpose of this systematic review was to focus on the personal level of the PALS and their family members rather than focusing on the cellular level of the disease, (b) biological intervention studies, and (c) non-peer reviewed grey literature (i.e., reports, policy documents, etc.), editorials and opinion writings, and conceptual articles. Subsequent to the abstract review, 44 additional articles were excluded and 66 were admitted for full-text review. Of note, when adequate information to determine inclusion/exclusion could not be obtained from the title and abstract, the investigators examined the article's full-text.

To confirm appropriateness for admission into this review, the two investigators both examined the first 20 of the 66 (30%) articles that were included for full text review. Of these 20 articles, the investigators were in agreement on 14 of the 20 (70%) and were in disagreement on six of the 20 (30%). The investigators shared their decision process with one another so that they were ultimately able to come to agreement on each of them. As the investigators encountered discrepancies or uncertainties on whether or not to include those articles, an investigator discussion was utilized to resolve them. The investigators then independently screened the remaining 46 articles to determine if they were eligible for inclusion based on the full text review, resulting in 22 of the 66 articles being excluded. Of those 22 articles, five did not meet the family member criteria, eight did not meet the BPS-S component criteria, two articles were based on genetic information only, five were in a language other than English, and two were not peer-reviewed. The reference lists of the remaining 44 articles were screened with twenty-two additional articles identified and four of the twenty-two being admitted following the abstract and full-text reviews for a final total of 48 articles admitted into the final review. There was 100% agreement from both investigators to include all 48 of the remaining articles for this review. A summary of the inclusion and exclusion review process can be found in Figure 1, PRISMA flowchart of paper selection process.

During the final steps [25], five investigators analyzed the articles and integrated the outcomes from each included study. The results of the study were broken down into each component of the BPS-S framework; biological, psychological, social, and spiritual health outcomes. Results in the psychological health component were separated by either symptoms of depression, anxiety, or overall burden and emotional well-being as these were the only specified psychological health outcomes discussed. The results relating to family functioning were

interpreted separately from the BPS-S outcomes. The investigators were each responsible for extracting the study characteristics and outcomes for 24 of the 48 final articles. In the following section, the results generated from this process are provided, as well as compared and contrasted to help identify where the science can be expanded and enhanced.

Results

In total, 48 articles published between 1988 and 2018 met the inclusion criteria from the 3,557 originally identified by keyword search in Pubmed, Psychinfo, and CINAHL. Following a discussion about study characteristics, findings related to the biological, psychological, social, and spiritual health outcomes of caregivers of ALS patients (CALS) are discussed. Results related to family functioning will then follow. As a reminder, patients with ALS will be referred to throughout the remainder of the article as PALS.

Study Characteristics

Across all 48 admitted studies, 4,037 CALS were enrolled as participants. Approximately 2,402 participants identified as female and 1,377 as male. Five articles did not report participants' gender [27-31]. Participants' ages ranged from 5 to 86 years old and family members' relationship to patient participants approximately consisted mainly of spouse or partner (78%) with the remainder being children (14%), parents (2%), siblings (1%), or other relatives (5%). However, not every article included complete demographic information on the PALS and CALS relationship [31, 32] and two of the articles did not include the age of the family member or caregiver [29, 33]. Only nine of the articles reported participant race/ethnicity with a total of approximately 546 white non-Hispanic participants, nine Black or African American participants, six Hispanic/Latino participants, and three unspecified race or ethnicity.

Due to the lack of consistent demographic information reported, the interpretation and application of it is limited.

Biological Health Outcomes

The question guiding this review, "How does ALS impact the immediate family members' health and interactional patterns?" called for a focus on the BPS-S health outcomes of the CALS. Out of the 48 articles reviewed, 17 included outcomes on the biological health impact of ALS on CALS. The majority of studies reported that CALS' physical well-being tended to decrease over the course of the disease and that the CALS often rated themselves with high scores of physical burden, indicating poorer physical health, on a variety of caregiver burden measures [6, 12, 29, 36-42]. Four out of 17 articles reported that CALS demonstrated poor physical health outcomes: CALS (a) worried over their own physical and biological health [10], (b) disclosed feelings of fatigue and somatic complaints [33], (c) described being physically tired most of the time [42], and (d) indicated declines in physical health occurring simultaneously to the PALS' declining physical health [44]. Although the studies revealed that family member and caregiver physical health decreased over the course of the disease, there was no mention of how the psychological, social, and spiritual health components were related to their biological health.

Three of the 17 articles did not reveal CALS as experiencing biological health decline. Olsson et al. [30] found no differences in the physical health of the CALS overtime in the entire next of kin group, while Trail et al. [13] found that 89% of CALS rated themselves as being in excellent health. In a study comparing health outcomes of CALS and PALS to health outcomes of the general Swedish population, Olsson et al. [29] found that there were no differences in physical health capacity between the CALS and the general Swedish population. These results may be due to the CALS being of a younger age, the course of the disease being short in length,

or even due to the family's ability to obtain assistive devices that eased the physical aspect of care. Although there is information available on how CALS' biological health is impacted by ALS, none of these studies included psychological health outcomes and how the two might relate to one another.

Psychological Health Outcomes

Of the 48 articles reviewed, 46 reported CALS experiencing challenging psychological outcomes. Due to the large number of articles that included psychological outcomes, these results were organized into the following categories: depression, anxiety, and overall emotional burden.

Depression

Out of the studies reviewed, 29 focused on depression among CALS. Results were mixed with 13 articles reporting CALS endorsed symptoms of depression and 16 reporting they did not. Of the 13 articles which focused on CALS' symptoms of depression, one article [45], found that nearly 50% of CALS identified symptoms of depression and 25% found that CALS had a score above the depression cut-off on various measurement tools. Furthermore, Watermeyer [46], found greater functional impairment among PALS was associated with higher caregiver depression; whereas, Chio et al. [47] found cognitive decline among PALS was directly related to depression in CALS.

Three studies found that CALS reported increased symptoms of depression as the disease progressed [7, 33, 36]. Chen et al. [28] found 24.7% of CALS were diagnosed with depression, and Olsson Ozanne et al. [48] found CALS endorsed worse depression symptoms than the general population. Specifically, a study conducted on the minor children of PALS [2], found

they experienced significantly higher levels of depression than children in a control group whose parents had no diseases.

To understand how 16 studies found depression to not be a significant outcome for CALS, one must look at how depression symptoms were assessed. First, studies utilized several measures to determine levels of depression. Rabkin et al. [44] used the PHQ-9 [49] and found 77% of CALS had no depression at the initial assessment and 90% had no depression at the last assessment, with no intervention provided between assessments. Galvin et al. [8] concurred and found that the mean depression scores of CALS was 5.9 on the HADS [50] (scores under 7 being within the normal range). Furthermore, 7 articles demonstrated that the majority of CALS had scores on the Zung Depression Scale [51], the Depression and Anxiety Scale [52], Beck's Depression Inventory [53] or Beck's Depression Inventory-II [54] that were lower than the cutoff for depression [9,37,47, 55-58].

Only two articles found CALS had no clinical depression [44] or depression symptoms [32]. Both of these studies had mean BDI scores for all CALS below the cutoff for depression. However, the participants in each of these studies were recruited from single multidisciplinary ALS clinics [32, 44]. These centers often offer more support, practical assistance, and a more positive approach for the PALS and their families, potentially diminishing any symptoms of depression [44]. Furthermore, more depressed CALS may be less likely to participate in projects described as studying how people cope with serious illness [44]. Additional studies (n=4) found that depression symptoms were not above the cutoff for depression in CALS [13, 29, 59, 60], that few CALS endorsed symptoms of depression [13], and that no significant differences in depressive symptoms in CALS were found over time [44].

The variations in outcome of these studies with 13 studies indicating levels of depression in CALS and 16 indicating no depression, may be a result of several components. The PALS and CALS with less depressive symptoms may have had higher levels of social support or spirituality, resulting in the relief of any depressive symptoms. There may have also been differences in age, gender, access to mental health services, or even access to other supportive devices between those with depressive symptoms and those without that may have eased these symptoms. In addition to studying levels of depression, several articles also focused on anxiety in CALS.

Anxiety

Unlike depression, all of the CALS studied reported experiencing some anxiety symptoms. Higher levels of anxiety were associated with higher levels of caregiver strain [35]. Goldstein et al. [61] found that the mean HADS scores did not indicate a diagnosis of anxiety for CALS although symptoms of anxiety were present. Garcia et al. [62], similarly found that 42% of CALS had scores above the lower cutoff to suggest presence of anxiety although not to the extent of warranting an anxiety diagnosis. A specific study on the impact of ALS on children demonstrated significantly higher anxiety scores for the children of a PALS than children in a control group with healthy parents [2].

While some articles evidenced lower CALS anxiety levels, there were no studies reporting no anxiety at all. Anxiety levels in four studies [9, 56, 60, 65] identified moderate anxiety levels in CALS. In one study [29] there were no differences found between PALS and CALS scores on the HADS [29]; however, one study noted CALS had higher levels of anxiety than the general population [58]. In reviewing these articles, it is clear that in every study, some of the CALS experienced symptoms of anxiety. However, what remains unknown is how the

additional BPS-S components influence one another and consequently symptoms of anxiety. Family functioning is also not studied to understand how family support and cohesion might alleviate anxiety symptoms. Following a focus on depression and anxiety symptoms, the remaining articles investigated overall quality of life, burden, and emotional well-being independent of depression or anxiety symptoms.

Caregiver Emotional Burden

Twelve of the articles included results on overall caregiver burden and emotional well-being. Patients often rated CALS as being more burdened than CALS rated themselves [70] and overall emotional burden was the domain with the lowest (worst) score on the Caregiver Burden Inventory [41]. One of the articles included general quality of life scores (QOL) with the summary mental capacity scores of CALS being significantly lower than the national standard [40]. Studies found that caregiver outcome scores resulted in increased psychological distress overtime [37] and that the severity of the disease was directly connected to caregiver emotional burden [60]. Two studies found female CALS reported having higher emotional burden than males [6, 39] and female partner emotional burden was significantly higher in adult child CALS [6]. Based on the results, it is clear that CALS are impacted emotionally in several ways. However, it remains unclear as to how the psychological health of the CALS are impacted by the other BPS-S health components. In addition to the reviewed articles that included biological and psychological health outcomes, 20 articles included in this systematic review focused on the social health outcomes of CALS.

Social Health Outcomes

Twenty of the 48 articles reviewed addressed social health outcomes of CALS. Twelve of these articles focused on non-specific social health outcomes such as noted restrictions on social

lives [8, 39, 43], limited personal time and social isolation [31, 36, 63, 64], disconnection with peers [34], and the need to confide in and receive social support from someone, with little time to make that happen [65]. Additional articles indicated that female CALS had worse overall social burden scores [41, 55] and that overall CALS' social health decreased over time [42].

Two of the 20 articles on social health reported marital intimacy implications. Atkins et al. [27] found that reduction in the everyday, interpersonal experiences in which the patient-spouse dyad had previously engaged contributed to reduced perceptions of the quality of their relationship. Further, Watermeyer et al. [46] reported that pre-illness marital intimacy scores were significantly higher than current scores and that when the PALS had greater behavioral symptoms, poorer outcomes in martial satisfaction were predicted.

An additional two articles focused primarily on the social health aspect of the PALS' family cohesion and overall family relationship satisfaction [41, 67]. Martin and Turnbull [67] studied the lasting social health impact on families after the loved one with ALS died. They collected information from surviving CALS using author designed questionnaires with no available psychometrics to support its reliability and validity. Results indicated that 15% of the 27 CALS characterized the disease as detrimental to family relationships, 18% felt that there was little change, and 67% believed the disease brought the family closer together [67]. Using the FACES III questionnaire [68], Tramonti et al. [41] studied the impact of ALS on family cohesion and adaptability. Results indicated that there was a high and increasing number of unbalanced families with "chaotically enmeshed" (highly adaptable with extreme closeness between family members) families being the most representative subtype based on the circumplex model [68]. Although there is no best level for relationships based on this circumplex model of family systems, families tended to have problems when they functioned at the extremes [68].

In addition to the previously described outcomes, Trail et al. [13] found that 6.8% of CALS studied chose worries about the patient's illness progression, concerns about the patient's swallowing and eating difficulties, and worries about loved ones' (PALS') emotional and physical well-being as their primary stressors. Hwang et al. [38] attempted to understand what might alleviate CALS burden so that they might have more personal time. The results of this study indicated that the use of the eye-tracking device (used by PALS for communication) allowed CALS to participate more often in social activities and in taking care of other family members. As mentioned previously, none of the articles that included the social component discussed how the social health of CALS may be impacted by the remaining BPS-S components. Therefore, it remains unknown how the social health of CALS may be impacted by their biological, psychological, or even spiritual health. After focusing on the biological, psychological, and spiritual health outcomes of CALS, the following section will discuss the remaining component of the BPS-S framework, spiritual health outcomes.

Spiritual Health Outcomes

Of the 48 articles reviewed, seven discussed the spiritual impacts on the CALS. Trail et al. [31] found that 63% of CALS continued to regularly attend church and that church was the CALS primary or even sole community activity. Calvo et al. [66] found that CALS' private religiousness was positively associated with caregiver quality of life and was the second most important predictor of quality of life after anxiety. Another study on the existential well-being and spirituality of PALS and their CALS resulted in higher existential well-being in the PALS than the CALS, but also showed that the higher levels of existential well-being in the PALS was directly related to higher existential caregiver well-being [69]. Similarly, Roach et al. [42] found a non-significant decrease in caregiver existential well-being subsequent to the formal diagnosis.

Although these few studies uncovered adverse spiritual impacts on the CALS, Murphy et al. [57] found spirituality was a major determinant of CALS quality of life while actively involved in the caregiving role. Finally, Trail et al. [13] found that 0% of CALS were concerned about their religiosity and were instead much more concerned about a sense of loneliness and time to themselves for hobbies and leisure activities. Again, based on the included articles, it seems as though spiritual health outcomes of CALS have not been considered simultanesouly with the biological, psychological, and social health of CALS. It is still unclear as to how the whole, BPS-S health and family functionoing of CALS is impacted by ALS.

Discussion

This systematic review was conducted to better understand the impact ALS has on CALS' BPS-S health and patterns of family functioning. Clinical and research recommendations are provided to demonstrate ways to apply the findings and advance the literature. Previous studies found that CALS are heavily impacted emotionally and physically by the steady progression of ALS, possibly due to caring for a partner or family member with a neurological illness negatively impairing quality of life [7-9] and family functioning [41, 67]. Supporting the value of studying BPS-S health factors simultaneously, alongside their family functioning, the Beaver's systems model of family functioning [23] and BPS-S framework [14-16] was applied to the findings as a way of interpreting the ALS literature in accordance with theory.

As a result of the systematic review, evidence was found that ALS has BPS-S impacts on CALS. While the 48 articles reviewed did include at least one BPS-S health outcome variable, several (n = 17) focused on biological health. They found CALS' physical well-being tended to decrease throughout the course of the disease resulting in higher scores of physical burden and poorer physical health on a variety of caregiver burden measures. The 46 articles that included

psychological health outcomes focused mainly on symptoms of depression (n=29), anxiety (n=11), and caregiver emotional burden (n=12).

Of the 20 articles that examined the social health component, the majority focused on non-specific social health outcomes such as restricted social lives [8, 39, 43], limited time for self, and social isolation [31, 36, 64]. Two studied marital intimacy, finding reductions in the patient-spouse relationship quality [27, 46]. Among the ALS literature reviewed, only two teams focused on family functioning [41, 67]. Both noted the majority of families pulled together (centripetal) during the illness experience; however, neither studied changes in family functioning patterns after the patient had died. Also, it is unclear what is happening among those families who push apart (centrifugal). In addition, researchers have not looked at the BPS-S health benefits or challenges experienced by centripetal or centrifugal family functioning patterns. In addition to articles on social health outcomes, seven included spiritual health outcomes. The vast majority of spiritual health outcomes revealed that ALS had no negative impacts on CALS' spirituality or existential well-being, however, these outcomes were not corroborated with any other biological, psychological or social health outcomes.

Future Research

Although each of the BPS-S components were addressed in some way throughout the reviewed studies, none attempted to understand how the components were influencing one another. It continues to remain unknown how the CALS' biological health is negatively or positively impacted by the amount of social support received or even by their spiritual beliefs. Likewise, higher levels of depression and anxiety were not related back to their own biological health, social support, or spiritual beliefs. In order to appropriately apply the BPS-S model, the components should not be studied in their own silos, but rather in the way it is designed as a

systemic whole [14-16]. Finally, as only one article studied the impact of ALS on the CALS following the PALS' death, this remains an area to be further explored [67].

Family functioning patterns among families with ALS are also inadequately represented. Although two of the included studies focused on family functioning [41, 67], the results of these studies do not represent all families with ALS. The results of these studies are also not expanded upon to determine what the health benefits or disadvantages are when the families become chaotically enmeshed. Furthermore, it remains unknown how family functioning may change within families throughout the duration of the illness or how family functioning may differ amongst families based on their ability to access additional supportive devices, participate in support groups, or even continue to participate in pre-illness routines (i.e., attending religious services, school events, family vacations, etc.). There is also space to discover how family functioning may change as the BPS-S health outcomes of the PALS and CALS change throughout the illness and even following the PALS' death.

This systematic reviewed highlighted that although researchers have been investigating BPS-S health outcomes of CALS, studies fall short in understanding the role healthy family functioning plays. Of the final 48 articles included in the study, 21 used one of the BPS-S variables, 13 used two, ten used three, and three used all four, again indicating a limitation of failing to incorporate each component of the BPS-S framework. Future studies should attempt to gather information related to each of the BPS-S health outcomes in a single study to more adequately determine how these individual components are impacting one another simultaneously.

Limitations

This systematic review has its limitations. First, the research team included two investigators who implemented safeguards such as the utilization of MeSH and keyword searches and crosschecked searchers to help reduce omission of relevant articles. However, these strategies do not eliminate the risk of overlooking or missing relevant articles including conference presentations, theses and dissertations, or non-peer reviewed research reports contained in gray literature [25].

Conclusion

Currently, there is a wealth of information on the individual BPS-S health outcomes of CALS. Research has shown that CALS are impacted biologically, psychologically, socially, and spiritually, although there has been no study that has reviewed each of these components simultaneously. Based on the reviewed articles it is also clear that ALS has an impact on the family's functioning although this research is limited. Future research is needed to determine how CALS' complete BPS-S health and how the family's functioning is impacted by ALS. Findings from this research can help to better address the BPS-S health needs of the entire family unit and can assist in the advocacy for more comprehensive family-based care for ALS families.

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Figure 1

PRISMA Flowchart of Paper Selection Process

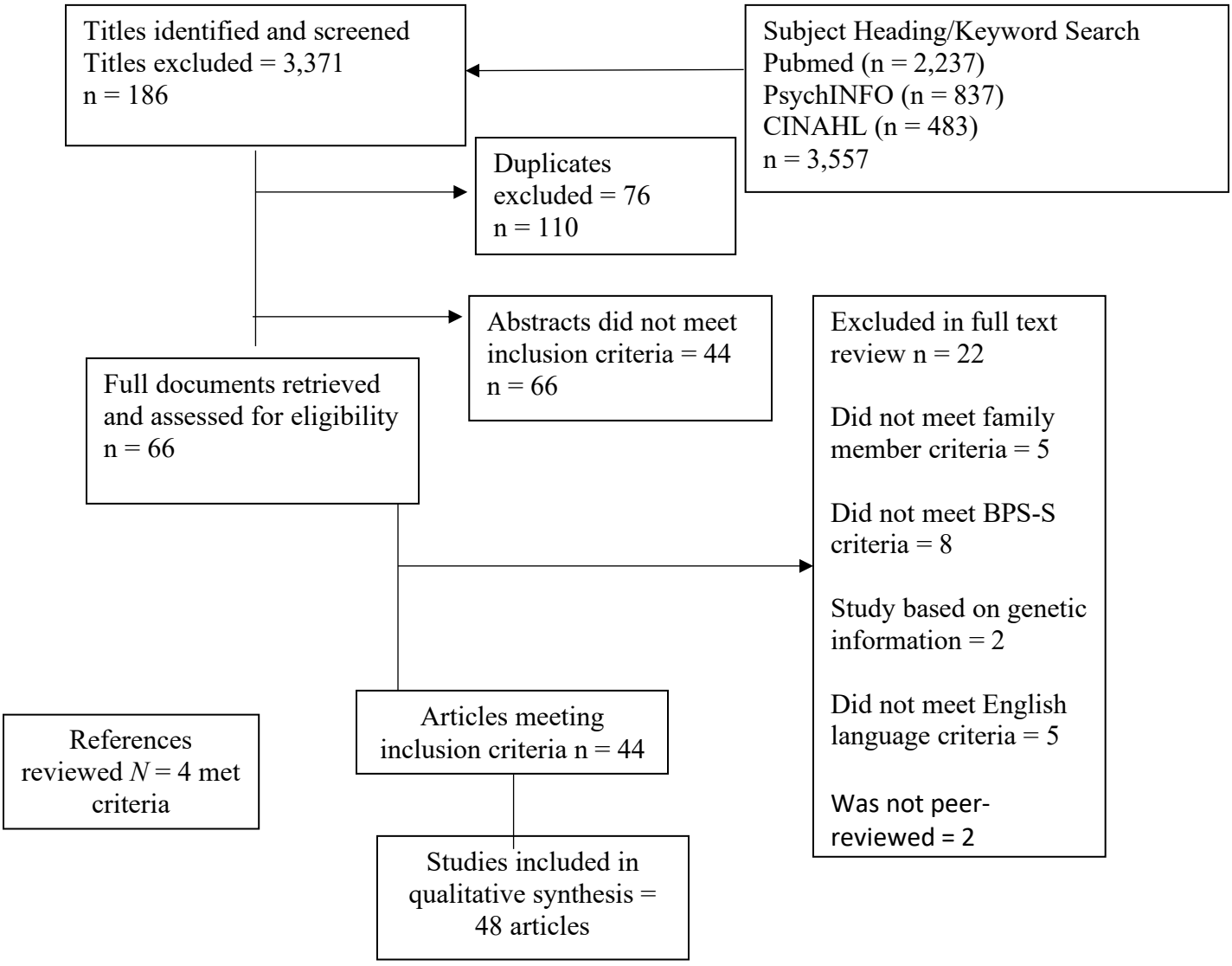

Table 1

MeSH and Keyword Search Terms

Database	MeSH and Keyword Search
Pubmed	"Amyotrophic Lateral Sclerosis"[Mesh] OR “Charcot Disease”[tiab] OR “Lou Gehrig Disease”[tiab] OR “Lou Gehrig's Disease”[tiab] OR “Lou-Gehrigs Disease”[tiab] OR “ALS”[tiab] OR “Gehrig's Disease”[tiab] OR “Gehrig Disease”[tiab] OR “Gehrigs Disease”[tiab] OR “Guam Form of Amyotrophic Lateral Sclerosis”[tiab] OR “Amyotrophic Lateral Sclerosis-Parkinsonism-Dementia Complex 1”[tiab] OR “Amyotrophic Lateral Sclerosis Parkinsonism Dementia Complex 1”[tiab] OR “Guam Disease”[tiab] OR “Amyotrophic Lateral Sclerosis With Dementia”[tiab] OR “Dementia With Amyotrophic Lateral Sclerosis”[tiab] OR “Motor Neuron Disease” [tiab] OR “Motor Neurone Disease” [tiab] OR “MND” [tiab] AND "Family"[Mesh:NoExp] OR "Nuclear Family"[Mesh:NoExp] OR "Siblings"[Mesh] OR “Parents"[Mesh:NoExp] OR "Spouses"[Mesh] OR Family[tiab] OR Families[tiab] OR “family member”[tiab] OR “family members and caregivers”[tiab] OR Stepfamily[tiab] OR Stepfamilies[tiab] OR “reconstituted family”[tiab] OR “reconstituted families”[tiab] OR Filiation[tiab] OR Kinship[tiab] OR Relatives[tiab] OR “Nuclear Families”[tiab] OR Child[tiab] OR Children[tiab] OR Kid[tiab] OR Kids[tiab] OR offspring[tiab] OR Sons[tiab] OR Son[tiab] OR Daughters[tiab] OR Daughter[tiab] OR Siblings[tiab] OR Sibling[tiab] OR Sisters[tiab] OR Sister[tiab] OR Brothers[tiab] OR Brother[tiab] OR Parent[tiab] OR Parenthood[tiab] OR “Step-parents”[tiab] OR “Step-parent”[tiab] OR “step

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CINAHL Plus with Full Text	S1: "Amyotrophic Lateral Sclerosis"[Mesh] OR TI (“Charcot Disease” OR “Lou Gehrig Disease” OR “Lou Gehrig's Disease” OR “Lou-Gehrigs Disease” OR “ALS” OR “Gehrig's Disease” OR “Gehrig Disease” OR “Gehrigs Disease” OR “Guam Form of Amyotrophic Lateral Sclerosis” OR “Amyotrophic Lateral Sclerosis-Parkinsonism-Dementia Complex 1” OR “Amyotrophic Lateral Sclerosis Parkinsonism Dementia Complex 1” OR “Guam Disease” OR “Amyotrophic Lateral Sclerosis With Dementia” OR “Dementia With Amyotrophic Lateral Sclerosis” OR “Motor Neuron Disease” OR “Motor Neurone Disease” OR “MND) OR AB (“Charcot Disease” OR “Lou Gehrig Disease” OR “Lou Gehrig's Disease” OR “Lou-Gehrigs Disease” OR “ALS” OR “Gehrig's Disease” OR “Gehrig Disease” OR “Gehrigs Disease” OR “Guam Form of Amyotrophic Lateral Sclerosis” OR “Amyotrophic Lateral Sclerosis-Parkinsonism-Dementia Complex 1” OR “Amyotrophic Lateral Sclerosis Parkinsonism Dementia Complex 1” OR “Guam Disease” OR “Amyotrophic Lateral Sclerosis With Dementia” OR “Dementia With Amyotrophic Lateral Sclerosis” OR “Motor Neuron Disease” OR “Motor Neurone Disease” OR “MND”) S2: "Family"[Mesh:NoExp] OR TI ("Nuclear Family"[Mesh:NoExp] OR "Siblings"[Mesh] OR “Parents"[Mesh:NoExp] OR "Spouses"[Mesh] OR Family OR Families OR “family member” OR “family members and caregivers” OR Stepfamily OR Stepfamilies OR “reconstituted family” OR “reconstituted families” OR Filiation OR Kinship OR Relatives OR “Nuclear Families” OR Child OR Children OR Kid OR Kids OR offspring OR Sons OR Son OR Daughters OR Daughter OR Siblings OR Sibling OR Sisters OR Sister OR Brothers OR Brother OR Parent OR Parenthood OR “Step-parents” OR “Step-parent” OR “step parent” OR Stepparent OR Stepparents OR Father OR Fathers

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CHAPTER 3: UNDERSTANDING SYMPTOM ONSET AND BIOPSYCHOSOCIAL-SPIRITUAL IMPACTS OF AYMOTROPHIC LATERAL SCLEROSIS

Amyotrophic Lateral Sclerosis (ALS) is a progressive, incurable disease that is likely to have a devastating impact on patients with ALS (PALS) and their families (O'Brien, Whitehead, Jack, & Mitchell, 2011). In ALS, muscle weakness continuously spreads to encompass all voluntary muscles, including respiratory muscles, leading to death within two to five years of symptom onset (Couratier et al., 2016). Currently, there are as many as 30,000 PALS living within the United States with approximately 5,600 individuals being newly diagnosed with ALS annually (Cruz et al., 2018). ALS has been characterized by sudden onset of progressive motor deficits that develop within weeks or months, possible cognitive and behavioral impairment and inevitable physical decline (Galvin et al., 2016). With no known cure, ALS is a fatal disorder with devastating symptoms that lead to eventual paralysis and death (Cruz et al., 2018).

ALS has traditionally been classified into either the sporadic or familial form (van Es et al., 2017). Approximately 90% of ALS cases are sporadic, meaning the cause of the disease remains unknown (Brooks, Miller, Swash, & Munsat, 2000). The remaining five to ten percent of cases are due to genetic mutations inherited from a family member. When two or more family members are diagnosed with ALS, the disease is considered familial. In addition to ALS cases being classified as sporadic or familial, the disease is often classified by site, pattern of onset, or by degree of upper motor neuron or lower motor neuron involvement (van Es et al., 2017). Although these classifications exist, variations in the pattern and spread of ALS are not well understood (Fujimara-Kiyono et al., 2011).

An additional area of ALS literature that has not been thoroughly understood from both the PALS' and their family members' perspective is quality of life through the lens of the

biopsychosocial-spiritual framework (BPS-S; Engel, 1977; 1980; Wright, Watson & Bell, 1996). The BPS-S framework proposes that biological, psychological, social, and spiritual components of health operate simultaneously in connected subsystems that influence all aspects of mental and physical health (Engel, 1977; 1980; Wright et al., 1996). Several researchers have reviewed the impact of ALS on individual components of the BPS-S framework for both PALS and family members and caregivers, with the results being somewhat varied (Makkonen et al., 2018; Olsson et al., 2010; Oyebode et al., 2013; Trail et al., 2003). For example, some research has shown that PALS and their caregivers report symptoms of depression (Chen et al., 2015), while other studies have shown that PALS and their caregivers report little to no symptoms of depression or anxiety (Rabkin, Albert, Rowland, & Mitsumoto, 2009).

Although research has been conducted on the BPS-S impact of ALS, little attention has been paid to all four domains simultaneously for patient and family members and how this might differ based on type of symptom onset, either bulbar or limb. Thus, this review will cover peer reviewed literature in attempt to highlight gaps and opportunities for future research. Ultimately, this review will (a) describe the staging systems for ALS, (b) explore the differences between bulbar and spinal onset, (c) detail the impact of ALS on PALS' and family members' BPS-S health, and (d) provide recommendations for future researchers who aim to optimize the care and quality of life for PALS and their family members.

Stages in ALS

Identified stages in ALS allow patients and caregivers to better understand the course of the disease and to better differentiate progression of the disease and subsequent quality of life (Chiò, Hammond, Mora, Bonito, & Filippini, 2013). Staging criteria for the disease help to provide a universal and objective measure of disease progression, resulting in benefits for patient

care, research classifications, and resource allocation (Roche et al., 2012). Various stages methods of ALS have been proposed, with the most widely studied methods being the Milano-Torino (MiTos) functional staging system (Chiò et al., 2013) and King's clinical staging system (Roche et al., 2012).

The King's system uses five stages (1 to 5) and is based on disease burden as measured by clinical involvement and significant feeding or respiratory failure (Roche et al., 2012). This staging system consists of stage 1: symptom onset (involvement of one central nervous system (CNS region), stage 2A: diagnosis, stage 2B: involvement of a second CNS region, stage 3: involvement of a third region, stage 4A: need for gastrostomy, and stage 4B: need for respiratory support (non-invasive ventilation). Symptom onset in stage 1 includes functional involvement by weakness, wasting away of muscles, spasticity, or continuously contracting muscles, dysarthria, or weakness in the muscles used for speech, or dysphagia, difficulty swallowing. Each of these stages occur in sequential order without reversion to earlier stages with the stages being reached at predictable, consistent times through the course of the disease. This staging system has been described as easy to use because it corresponds to information relevant to a neurologist and symptoms reported by the patient. While King's staging system focuses on anatomical disease spread and significant involvement of respiratory muscles, the MiTos staging system focuses on the distinction of functional capabilities during the spread of the disease (Fang et al., 2017).

The MiTos staging system is based on the validated ALS Function Rating Scale-Revised (ALSFRSR) and also identifies relevant stages of the disease (Chiò et al., 2013). This system uses six stages with stage 0 being normal function and stage 5 being death. Stage 1 includes the loss of independence in one domain of the ALSFRSR, stage 2 includes the loss of independence in two domains, stage 3, the loss of independence in three domains, and stage 4, the loss of

independence in four domains. Similarly to the King's staging system, this system also found that the distribution of patients across stages and the probabilities of transition between stages were consistent with sequential disease progression. Although these staging systems slightly differ, research supports the use of both systems (Fang et al., 2017).

Both the King's and MiTos staging systems summarize different aspects of patient information (Fang et al., 2017). King's system summarizes the clinical or anatomical spread of the disease while MiTos staging summarizes the functional burden of the disease. Although King's system is better able to differentiate early to mid-disease stage well, MiTos staging is able to differentiate late stages more effectively. Even though there are slight differences between these staging systems, research supports the use of both to describe ALS stages as together they provide a concise summary of disease spread and functional burden. Understanding the clinical stages of ALS is likely to be useful in the treatment of the disease. Similarly, understanding the differences in disease course based on symptom onset may be equally beneficial to PALS, their family members, and to the design and development of more beneficial research, treatment plans, interventions, and patient-family centered policies.

Symptom Onset

In order to more thoroughly understand the progression of ALS, it is important to consider the variations in the disease that occur based on symptom onset. The following section will provide an overview of (a) the different types of symptom onset in ALS, (b) how it progresses, (c) and information as to how each type is a predictor of prognosis. A characteristic feature of the ALS concludes that in 90-98% of cases the first symptoms occur in only one body region with subsequent progression confined largely to that same region before involvement of additional regions (van der Kleij, 2015). The disease is often classified by site or pattern of

symptom onset to include bulbar and spinal, or limb, onset in Classic ALS with research showing that the pattern of spread following symptom onset is a predictor of prognosis (van der Kleij, 2015; van Es et al., 2017).

Limb-onset

Approximately two-thirds (66%) of PALS have a spinal form of the disease (limb-onset; van Es et al., 2017). These PALS most often present with symptoms related to central muscle weakness and wasting with the symptoms starting either distally (farthest away from the point of origin) or primally (closest to the point of origin) in the upper and lower limbs. In limb-onset ALS, spasticity may develop in the weakened limbs, which can affect dexterity and gait. PALS may also notice muscle wasting before onset of weakness and some patients may even present with spastic paraparesis, progressive weakness and stiffness in the legs (Wijesekera & Leigh, 2009).

Weakness due to limb-onset ALS usually comes on slowly, with PALS noticing that these symptoms are exacerbated by cold weather (Wijesekera & Leigh, 2009). Following asymmetrical symptom onset, originally unaffected limbs develop eventual weakness and wasting, resulting in bulbar and eventually respiratory symptoms. Limb-onset ALS symptoms commonly also experience bladder dysfunction and sensory issues (i.e., numbness, tingling, and/or pain), along with cognitive and behavioral symptoms (i.e., dementia, reasoning and judgement impairments, and emotional regulation challenges). The remaining one-third (33%) of PALS present with bulbar symptoms.

Bulbar-onset

Bulbar-onset of the disease, and bulbar dysfunction overall, is often associated with poorer prognosis or outcomes (Felgoise, Zaccheo, Duff, & Simmons, 2016; Fujimara-Kiyono et

al., 2011; Green et al., 2013). Bulbar-onset most commonly targets muscles in the face, head, and neck (Simmons, 2005) and is more common in females and older age groups (Wijesekera & Leigh, 2009). Although it has been determined that individuals with ALS do not always notice the earliest changes in their bulbar function (Allison et al., 2017), people with ALS demonstrate various speech symptoms at the time of diagnosis (Makkonen, Ruottinen, Puhto, Helminen, & Palmio, 2018). The presence of bulbar-onset is associated with progressive reduction in speaking and articulatory rates (i.e., an increase in the number and duration of pauses during speech). The initial symptoms of bulbar-onset often include a slight, intermittent slurring of speech, a hoarse, nasal quality to the voice, a decrease in speech volume, slow and limited movements of the tongue, lips, and pharynx, resulting in progressively decreasing intelligibility and eventually complete loss of speech (Simmons, 2005). This loss of effective communication is often considered one of the worst aspects of the disease by PALS (Makkonen et al., 2018).

Previous research on bulbar and limb onset ALS has suggested that PALS' quality of life (QOL) can differ based on verbal speech ability (Felgoise et al., 2016). PALS with limb-onset are often referred to speech therapy sooner than those with bulbar-onset because they are usually already being monitored for other symptoms, resulting is substantially more time to organize communication aids for limb-onset patients (Makkonen et al., 2018). The resulting loss of the ability to communicate has been found to have devastating psychologic and social consequences (Simmons, 2005). This aspect of the disease, loss of speech, is often considered one of the worst aspects of the disease, with PALS most commonly asking how long they will be able to speak during clinical visits (Makkonen et al., 2018). Ultimately, changes in and loss of speech are found in both bulbar and limb-onset PALS, yet these changes tend to occur earlier in bulbar-onset PALS.

Overall, the type of initial onset of ALS, either limb or bulbar, is a major determinant of overall survival (Turner et al., 2010). Bulbar-onset of the disease, along with older age at symptom onset and earlier respiratory muscle weakness are most often associated with reduced survival (Kiernan et al., 2011). Limb-onset however, in addition to younger age at symptom onset and longer diagnostic delays, is a predictor of prolonged survival. Due to the symptom onset type being a predictor of outcomes in ALS, it is imperative that PALS and their support persons' experiences are more thoroughly understood to plan care more effectively (Turner et al., 2010). In addition to a scarcity of research comparing the lived experiences across ALS onset types, there are no known studies that have looked in to the psychosocial and spiritual experiences of PALS and their support systems regarding onset type, diagnosis experience, or disease progression. In that the BPS-S framework declares that each of the health components impact one another (Engel, 1977; 1980; Wright et al., 1996), it is essential that the simultaneous impact of each BPS-S component for both PALS and their support persons is adequately understood.

Biopsychosocial-spiritual Health

Exploring the impacts of ALS on PALS and their support persons through the BPS-S framework (Engel, 1977; 1980; Wright et al., 1996) assists researchers and clinicians in better understanding the systemic and relational implications of the disease by viewing health through a comprehensive lens. Utilizing the BPS-S framework has even emerged as a predictor of survival, with results indicating that the risk of death in PALS might be reduced by 45% at five years (Kiernan et al., 2011). Thus far, the research related to BPS-S health in ALS remains siloed with the majority of the research being independently patient-focused or independently focused on the support persons. However, a dearth of literature does explain how ALS effects the biological,

psychological, social, and spiritual, although these components have only ever been studied on their own, without determining how they might be influencing one another. The following section will provide a brief overview of the literature on BPS-S health in PALS and their support persons, as it exists, with the impacts on each component (biological, psychological, social, and spiritual) being explained independently of the others.

Biological

As ALS naturally results in progressive muscle weakness, it is clear that PALS are physically impacted by the disease in other ways as well (Creemers et al., 2015). They commonly experience malnutrition, weight loss, dehydration, and respiratory issues as their muscles begin to shut down (Simmons, 2005). PALS also report pain due to muscle spasticity and cramping. Although the support persons of PALS do not experience these same biological changes, they do experience other biological differences as the physical caregiving demands increase (Olsson, Markhede, Strang, & Persson, 2010; Pagnini et al., 2010).

Researchers found support persons often disclose feelings of fatigue and somatic complaints (Gelinas, O'Connor, & Miller, 1998; Roach, Averill, Segerstrom, & Kasarskis, 2009). They describe their physical health declining simultaneously to the declines in the PALS' physical health (Rabkin et al., 2009). Similar findings occur in the psychological literature noting evidence of a corresponding decline as the PALS' physical health deteriorates.

Psychological

The impact ALS has on PALS' psychological health varies from patient to patient (Simmons, 2005). Depression is one mental health condition that is commonly reported. Several factors seem to play a role in determining whether or not PALS become depressed, including individual pain levels, social support, and perceived family and caregiver burden. Depression

rates in PALS have ranged in various studies from 2% of PALS having major depressive disorder in one study (Rabkin, Wagner, & Del Bene, 2000) to anywhere from 11% to 75% having major depressive disorder in other studies. Anxiety has also been found to be very common in PALS with as many as 67% of PALS endorsing symptoms of anxiety (Chen et al., 2015). Symptoms of anxiety in PALS has been found to become increasingly severe as the disease progresses and has been found to determine subsequent depression in PALS (Atassi et al., 2011). Pseudobulbar affect and fronto-temporal dementia are additional conditions (impacting cognitive, emotional, and behavioral states) that are common in PALS (Ng, Rademakers, & Miller, 2015; Thakore & Pioro, 2017). Pseudobulbar affect manifests in uncontrollable episodes of crying or laughter, which are incongruent with the emotion or situation and has been found to be prevalent in PALS (Thakore & Pioro, 2017). Fronto-temporal dementia is a progressive brain disease with changes in behavior, personality, and language dysfunction due to loss of nerve cells that often occurs in PALS (Ng et al., 2015). Due to growing research on ALS' impact on patient's psychological health, it is important to expand the research to all areas of the BPS-S framework. It is also necessary to study not only PALS but their support persons as well so science can better inform treatment.

Due to the extensive time commitment of caregiving, its significant emotional burden, and due to the majority of ALS support persons being the PALS' spouses (Tramonti, Bongionni, Leotta, Puppi, & Rossi, 2015), extensive research has been conducted to evaluate the psychological health of support persons (Qutube, Lacomis, Albert, & Feingold, 2014). Similar to studies conducted with PALS, caregivers often experience extreme emotionality due to the steady progression of the disease and lack of effective treatment (Pagnini et al., 2010). Several studies (Chiò et al., 2010; Tremolizzo et al., 2016; Watermeyer et al., 2015) found nearly 50% of

support persons reported symptoms of depression and 25% had had a score above the cut-off for clinical depression on various measurement tools (Tremolizzo et al., 2016) with their symptoms of depression increasing as the disease progressed (Burke et al., 2018; Gauthier et al., 2007; Gelinas, et al., 1998). Interestingly, two research teams found little to no symptoms of depression (Qutub et al., 2014; Rabkin et al., 2009) with differences in cultural background, income, access to healthcare, level of support, sample size, or even the utilization of varying assessment tools potentially modifying the outcomes.

Symptoms of anxiety in the support persons of PALS have also been studied extensively. The vast majority of research on symptoms of anxiety in the support persons of PALS has found that all support persons indicated at least some symptoms of anxiety (Creemers et al., 2016; Garcia et al., 2017; Grabler et al., 2018; Pagnini et al., 2012a). In addition to the psychological research on support persons of PALS covering anxiety and depression, this area of research has also focused on the overall caregiver burden and emotional well-being. The results of these studies have indicated that the psychological distress of these caregivers increases over time and that the severity of the disease has been directly connected to increased emotional burden in caregivers (Goldstein et al., 2006; Pagnini et al., 2012b). While there are extensive findings that endorse the notion that ALS psychologically impacts patients and their caregivers, previous studies havemainly focused on either the PALS or the support person, without investigating how the disease impacts social relationships.

Social

Social health is important to the overall quality of life (QOL) of PALS (Makkonen et al., 2018). For PALS, social integration is often impacted by impaired mobility and communication, even though positive effects on coping have been directly related to social involvement (Hecht et

al., 2002). Previous researchers found PALS may purposefully limit their social interactions due to embarrassments related to or fear of functional impairments (Kukulka, Washington, Govindarajan, & Mehr, 2019). In addition to previous social research on PALS, several research teams focused on non-specific social health outcomes of support persons caring for someone with PALS. Researchers found that support persons often report limited personal time and restricted social lives (Gauthier et al., 2007; Oyebode, Smith, & Morrison, 2013), disconnection from peers (Siciliano et al., 2017), and desire to confide in someone for social support but with little time to make that happen (Bolmsjö, & Hermerén, 2013). Ultimately, both PALS and their support persons seem to identify social groups and time with peers as being beneficial to their social health (Kukulka et al., 2019).

Due to PALS and support persons experiencing limited social interactions because of PALS' difficulties communicating and moving and support persons having little time to participate in social activities, interventions aimed at maintaining the social support of PALS and their support persons have proven to be effective (Isle et al., 2015; Loane & D'Alessandro, 2013). For example, PALS have been found to benefit socially from participation in on-line health related support groups as these groups allow them to participate regardless of the lack of mobility or communication skills (Loane & D'Alessandro, 2013). These on-line support groups have been found to essentially replace lost or deteriorating "real-life" social networks, with PALS positively benefiting from regular reminders that they are not alone in their ALS journey.

Likewise, support persons of PALS have also benefited positively from social support groups to include both face-to-face and online support groups (de Wit et al., 2018). Specifically, these support persons benefit from mutual support groups (Cipolletta, Gammino, Francescon, & Palmieri, 2018). Mutual support groups are informal networks of individuals who share a

common experience or issue and who can learn from each other's new adaptive behaviors. These groups allow support persons to interact socially with others who have experienced similar challenges due to the everyday care of a family member and can improve their knowledge of the disease, encourage the acceptance of difficult situations and aid the development of new coping strategies. As support persons of PALS often experience a lack of personal time for social interaction, a blend of online and face-to-face groups allow access to support at any time, effectively improving their social health (de Wit et al., 2018). Similarly to participation in social support groups, PALS' and support persons' spirituality and participation in religious practices has been found to be beneficial to their health.

Spiritual

Several studies have focused on the spiritual impact that ALS has on PALS and their support persons. It has been found that although overall QOL may not be correlated with religiosity early in the course of ALS, a significant relationship develops over the course of the disease (Bremer, Simone, Walsh, Simmons, & Felgoise, 2004). Murphy and colleagues (2000) found that PALS who were more spiritual or religious had more hope and were less likely to consider assisted suicide. Support persons reported attending church services and events to be the only community activity they were able to engage in regularly (Trail, Nelson, Van, Appel, & Lai, 2003). Calvo et al. (2011) found support persons' private religiousness was positively associated with QOL and was the second most important predictor of QOL after anxiety. Another study on the existential well-being and spirituality of PALS and their support persons demonstrated higher existential well-being in the PALS than the support persons (Pagnini et al., 2011). However, higher levels of existential well-being among PALS were directly related to higher existential well-being among caregivers.

Recommendations

To comprehensively assess the impact of ALS based on symptom onset and on the simultaneous impact of each BPS-S component and how those components potentially affect one another, additional research is needed in each of these areas. Previous literature has demonstrated that PALS have differing experiences based on bulbar or limb-onset, with bulbar-onset PALS often having poorer prognosis (Felgoise et al., 2016). The existing literature attempts to identify how the PALS and their support persons' BPS-S health is impacted (e.g., Makkonen et al., 2018; Olsson et al., 2010; Oyebode et al., 2013; Trail et al., 2003), yet it does not attempt to explain how each of these components potentially interact with, elevate, exacerbate, moderate, or even alleviate one another. In addition, few researchers have studied the comprehensive BPS-S health effects of ALS, with research sparse on how those health components are impacted by type of symptom onset. Finally, the current literature has shared how ALS impacts PALS and support persons, but studies contributing to this body of research have been done primarily with individuals. The comprehensive health of these individuals has yet to have been studied from the perspective of the entire family unit or support system.

Future studies focusing on comprehensive BPS-S health patterns and outcomes among PALS and their support persons' would allow for each of these existing, disconnected pieces to be conjoined. One place to begin would be to focus on how each BPS-S components of health interact with one another, while simultaneously considering how type of symptom onset may result in variations of these health components. It is also important to explore the experiences with ALS from the perspective of the entire support system or family unit. As ALS results in PALS becoming increasingly dependent on others, family members and additional support persons are often deeply involved with the disease (Tramonti, Bongionni, Leotta, Puppi, &

Rossi, 2015). Studying the experiences of the disease from a systemic perspective would allow for the previous research on the individual perspectives of PALS and their support persons to be combined and compared to one another (von Bertalanffy, 1967).

It may also prove beneficial for future research on these topics to be conducted using qualitative methods. Thus far the majority of ALS research has utilized quantitative methods. The use of qualitative methods would not only allow for multiple members of the PALS' system to share their own perspective of ALS but would also all for participants to elaborate on a variety of social determinants of health that might otherwise remain unknown when quantitative methods alone are use. Although it remains that ALS has no cure, future research on this topic can continue to provide valuable information and recommendations for multidisciplinary care that will optimize the quality of life for PALS and their family members.

Conclusion

Due to ALS having no known cure and no ability to stop the progression of the disease, the primary goal in caring for PALS and their support person has often been considered to be the optimization of QOL (Simmons, 2005). This review of the literature explored how QOL in PALS might differ based on symptom onset, finding that PALS with bulbar-onset have worse outcomes (Felgoise et al., 2016). The review of the literature also found that the results of previous studies on BPS-S health outcomes have been varied for both PALS and their support persons. When studying the impacts of ALS by symptom onset through the lens of the BPS-S framework, it is essential that the each of the BPS-S components are studied simultaneously. Given that the existing literature on the varying experiences of living with ALS based on symptom onset and BPS-S health outcomes remains limited, it is apparent that more research in this area of ALS is needed.

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CHAPTER 4: METHODOLOGY

Amyotrophic lateral sclerosis (ALS) is an incurable, progressive neurodegenerative disease characterized by a loss of motor neurons and increasing muscular weakness that inevitably leads to death (Knibb et al., 2016). The disease has long been recognized as a fatal condition, with median survival time estimated to be 20 to 48 months from symptom onset (Paulukonis, 2015). Since clinical manifestations and speed of progression for ALS vary widely among affected individuals, it is difficult to accurately predict the course of the disease, including the progression of disability for a given individual (Creemers, Grupstra, Nollet, van den Berg, & Beelen, 2015).

Patients diagnosed with ALS (PALS) gradually need increased assistance with their activities of daily living due to the disease's progressive course (Krivikas, Shockley, & Mitsumoto, 1997; Mockford et al., 2006). Eventually, ALS compromises their ability to speak and move without assistance (Rabkin et al., 2009). It has been estimated that ALS caregivers spend an average of 11 hours every day helping patients maintain a good quality of life (Krivikas, et al., 1997). As most caregivers are the patients' spouses, and at times even the patients' children (Kiernan et al., 2011; Tramonti et al., 2015), it is imperative to understand how families successfully navigate this disease and modify their roles and functioning to adapt.

Caregiver and family burden associated with ALS is known to negatively impact their physical (Hwang, Weng, Tsai, & Chang, 2014; Tramonti et al., 2015), emotional (Burke et al., 2017; Pagnini et al., 2012; Tremolizzo et al., 2016), social (Galvin et al., 2016; Sicilliano et al., 2017), and spiritual health (Roach, Averill, Segerstrom, & Kasarskis, 2009), yet little is known about how family functioning is impacted. Beavers systems model explains that negentropic (flexible and adaptive) families are more capable of negotiating and dealing effectively with

stressful situations (Beavers, 1981). This model further describes family compentence along the horizontal and the family style on the vertical axes. The horizontal axis, describing family competence, relates to the structure, available information, and adaptive flexibility of the system. Flexible and adaptive families can better negotiate, function, and effectively deal with stressful situations. Highly competent families have structure and the ability to change structures, as well as an approach to relationships that does not bind them to rigid behaviors. Less competent families tend to have poorer boundaries, lack communication, and are systemically rigid.

The vertical axis of the model, family style, describes families as either centripetal or centrifugal. Centripetal families are inner oriented with family members having difficulty separating from one another and feeling as though the family is the most trustworthy source of satisfaction. They are outer oriented with independence encouraged and satisfaction expected more often from the outside environment rather than from the family. Beaver's concept of centripetal and centrifugal family styles can be particularly useful in examining the impact that illnesses (i.e., ALS) have on family functioning.

Families of PALS are challenged by their need to adapt to new roles and reorganize their family structures as the disease progresses (Newby, 1996). Beavers and Voeller (1983) noted that chronic disease often exerts a centripetal pull on the family system with the family members focusing all role changes and uncertainty about the illness inward. The authors further argued that if during the onset of an illness the family gravitates toward a centrifugal style, it would likely result in a prolonged centrifugal period, with family members persistently pulling away from one another as the disease progresses (Newby, 1996). However, progressive diseases, such as ALS, are described as being inherently more centripetal in their effect on family stress (Newby, 1996). It is apparent that the centripetal and centrifugal reactions may vary greatly over

time, with that variability having important effects on the family's functioning throughout the illness. In addition to considering the family's style, either centripetal or centrifugal, it is also important to consider the PALS' type of symptom onset.

PALS primarily present with bulbar-onset disease (approximately 25%) or limb-onset disease (approximately 70%) with the remaining 5% presenting with initial trunk or respiratory involvement (Vucic, Burke, & Kiernan, 2007). Bulbar-onset consists of spastic dysarthria, slow, labored, and distorted speech often with a nasal quality, tongue wasting and weakness, and difficulty swallowing (Kiernan et al., 2011). PALS with bulbar-onset have obvious speech deterioration before definitive diagnosis (Makkonen et al., 2016). This loss of ability for effective communication may result in difficult psychological and social problems (Simmons, 2005), with PALS often considering the potential loss of speech to be one of the worst aspects of the disease (Hecht et al., 2002). Limb-onset consists of muscle spasticity, continuously contracting muscles, muscle weakness and wasting away, and fasciculations, spontaneous contractions causing flickers of movement under the skin (Kiernan et al., 2011). Bulbar-onset disease, as well as older age at symptom onset and early respiratory muscle dysfunction are independently associated with reduced survival, while limb-onset disease, as well as with younger age at symptom onset, and longer diagnostic delay are independently associated with prolonged survival (Talbot, 2009). Making sense of how these symptoms impact PALS and their families is important to advancing the research and treatment.

According to the Beaver systems model (Beavers, 1981), negotiation between and amongst family members is necessary for successfully managing stressful situations. An illness event may draw families closer together or push them farther apart. In addition to focusing on the impact of ALS on the family's functioning, the biopsychosocial-spiritual (BPS-S; Engel, 1977;

1980; Wright, Watson, and Bell, 1996) framework is used to study the comprehensive impact of ALS and the patient and their family system. It was developed as a scientific model that takes into account the missing elements of the biomedical model, including the psychological, social, and spiritual components of health. The framework suggests that all BPS-S components of health operate simultaneously and influence all aspects of mental and physical health. Thus far, the majority of ALS research on BPS-S health has been disjointed with studies focusing on only one or two health components at a time and failing to incorporate how those components may be impacting one another. The research also shows a lack of consideration as to how family member's BPS-S health may be impacting the family's functioning. The inclusion of the BPS-S framework, in conjunction with Beaver's systems model of family functioning allows for the opportunity to further investigate the simultaneous impact of ALS on a family's functioning and their overall BPS-S health.

This chapter presents the methodology for the dissertation investigating the experiences of patients and family members living with ALS and how family functioning and health are impacted. It is designed as a qualitative study, to be conducted with PALS and a primary support person, using Husserl's descriptive phenomenological method of inquiry (1901; 1970). This qualitative study provides a rich description on how family functioning styles are impacted by symptom onset. The following research question guided the study: "What are the experiences of patients - and their identified primary support persons - living with ALS and how has it impacted family functioning and health?"

Study Design

The qualitative study consisted of in-person, audio-recorded, in-depth, open-ended, phenomenological interviews. This qualitative study was designed in alignment with Husserl's

(1901; 1970) descriptive phenomenology. The research question guiding the study was: "What are the experiences of patients and their support persons living with ALS and how has it impacted family functioning and health?" It consisted of audio-recorded, in-depth, open-ended, semi-structured phenomenological interviews with PALS and their support person at a set time. The lead researcher conducted all interviews using a semi-structured interview guide (see Appendix E) to ensure all participants addressed similar topics and issues related to ALS and its impact on family functioning and BPS-S health. This study was approved by the East Carolina University and Medical Center Institutional Review Board (IRB) prior to participant recruitment (see Appendix A).

Setting

Patient participants were recruited from the Jim "Catfish" Hunter ALS Clinic at Vidant Medical Center, a Southeastern academic medical center. This center is one of only four multidisciplinary ALS clinics in the state of North Carolina and was opened in September 2008. Currently there are 25 patients at this clinic, with approximately 30 unique patients each year. As this ALS Clinic is located in rural, eastern, North Carolina, some of the current patients are from the neighboring eastern states, with a few patients traveling more than two hours to attend the clinic. The ALS clinic at Vidant Medical Center employees several multidisciplinary employees including, a neurologist, a neuropsychologist, a speech therapist, a dietician, a physical therapist, an occupational therapist, a respiratory therapist, an ALS association representative, and a local equipment consultant who patients meet with as needed.

The semi-structured interviews took place at the ALS clinic at Vidant Medical Center, via Web-Ex, and in participant's homes. In addition to conducting in-person interviews with PALS and at least one support person, any family members or support persons who wished to

participate, but were unable to be physically present, were given the option to utilize WebEx and participate from afar.

In person interviews were recorded using handheld, digital voice recorders. At the close of the interviews, the lead researcher immediately uploaded the audio-recorded interviews to a secure Piratedrive. In the event that support persons wished to participate but were unable to participate in person, secure, encrypted technology, through the use of CISCO WebEx, an online videoconferencing application, was utilized to conduct confidential video and/or audio-based interviews. Information about potential confidentiality risks of utilizing this technology was included in the consent forms, which participants read and signed prior to participating in the interviews. Immediately following interviews that utilized CISCO WebEx, the lead researcher downloaded and saved the audio/video recordings to a secure Piratedrive.

Participants

Participants consisted of a purposive sample of PALS and a member of their non-professional support system (e.g., partner, adult child, neighbor, extended family member, parent, etc.). Participants were considered eligible for participation in the study if they meet the following inclusion criteria: (a) English-speaking; (b) aged 18 or older; (c) the PALS has a formal diagnosis of ALS, and (d) both a PALS and at least one primary support person agree to participate in the study. Patient participants identifyed the support persons for possible inclusion in the study.

Exclusion criteria for this study included: (a) PALS' positive screening for frontotemporal dementia or other types of dementia as determined by Vidant ALS Clinic staff, (b) support person's cognitive impairment that would interfere with the ability to consent to participate, and (c) communication challenges that are unable to be resolved with technological

assistance already in use by the participant. Upon meeting the study criteria, participants were asked to consent to the study which includes their completion of a demographics survey and participation in a one-time, audio-recorded, in-depth, open-ended, semi-structured interview. Contact information was collected by a triangulated researcher who is employed by Vidant Medical Center and was used by the research team to schedule interviews and for member checking purposes. This information was stored on a separate password protected file on the lead researcher's Piratedrive. As part of the consent process, the lead researcher or triangulated researcher explained to participants that they were not obligated to participate and that they could elect to discontinue enrollment at any time without consequence to the care received at the ALS clinic. It was also included in the informed consent that none of the information would become part of their electronic health record and it would be deidentified to protect their confidentiality.

Throughout the qualitative study, participants were recruited, and interviews were conducted until thematic saturation was reached and no new themes emerged from participant interviews for both bulbar and limb onset groups (Glaser & Strauss, 1967). According to the literature, saturation depends on several factors, a large number of articles, book chapters and books suggest anywhere from five to 50 participants as adequate (Dworkin, 2012). In a study on sample size and saturation, Mason (2010) found that the most common sample sizes for phenomenological studies were 20 and 30 participants; however, Creswell (1998) suggested anywhere from five to 25 total interviews.

Researcher Roles

Lead Researcher

In a phenomenological study, the researcher suspends knowledge in an attempt to understand a phenomenon on a deeper level and to elicit rich and descriptive data unique to the

lived experiences (Merleau-Ponty, 1956). Within the Husserlian phenomenological approach, epoche, or bracketing, is a process utilized in setting aside one's own beliefs, feelings, and perceptions in order for the true, unbiased experiences of the phenomena to present themselves (Colaizzi, 1978; Streubert & Carpenter, 1999). Moustakas (1994), when focusing on Husserl's concept of bracketing, argued that researchers might embrace this idea when beginning a project by describing their own experiences with the phenomenon and bracketing their views before collecting participant data.

To fully engage in epoche, or bracketing (Tufford & Newman, 2010), the lead researcher engaged in reflexivity, suspending any pre-conceived notions, thoughts, and feelings around the possible lived experiences of family members of a PALS. Epoche is a concept in which investigators set aside their experiences as much as possible in order to take a fresh perspective toward the phenomenon under investigation. When engaging in reflexivity, the researchers should position themselves within a qualitative study, meaning that the researcher conveys their background, how it informs their interpretation of the information in a study, and what they have to gain from the study (van Manan, 2014). This means that the researcher is conscious of the biases, values, and experiences that they brings to a qualitative study (Hammersley & Atkinson, 1995). The reflexivity process in the current study included an ongoing reflexive journaling throughout the entirety of the dissertation process that assisted the lead researcher in identifying her beliefs, assumptions, and biases related to the dissertation topic (van Manan, 2014).

Triangulated Researcher

The triangulated researcher participated in the recruitment process and the data analysis process. Throughout the study the triangulated researcher assisted the lead researcher in these processes and triangulated the data as the lead researcher and the triangulated researcher

compared their data analyses. This data analysis comparison allowed the lead researcher and triangulated researcher to challenge one another's biases until agreements could be made about the findings and interpretations from the interviews. Any researchers, to include the lead researcher and any triangulated researchers, participating in data analysis prepared a bias statement of advance of participating in the data collection or analysis phases of the study. The lead researcher's and triangulated researcher's bias statements are included as an appendix (see Appendix G).

Data Collection and Procedures

The following section will include the procedures that the lead researcher engaged in to recruit, enroll, and consent participants. The specific details of the data collection process will also be included. Finally, the section will include a description of the plan for data analysis.

Recruitment, Enrollment, and Consent

Following approval from the university's Institutional Review Board, a two-pronged recruitment procedure was implemented. Individuals who meet the eligibility criteria were contacted from the Jim "Catfish" Hunter ALS Clinic by a triangulated researcher. This triangulated researcher is also an employee of the Jim "Catfish" Hunter ALS clinic and contacted the PALS by phone to inform them of the possibility of participating in the study using a standard telephone script (see Appendix B). The script that the triangulated researcher used to contact potential participants asked PALS to provide contact information for the support person(s) they wished to participate in this study with them. If these PALS wished to participate and agreed to have their contact information shared, the triangulated researcher passed this information on to the lead researcher who then contacted them to schedule an interview.

Recruitment also took place in-person at the ALS clinic. Once a month, the ALS clinic sees its patients. Once a patient is placed into an exam room, the same triangulated researcher entered the room and introduced the study to eligible PALS and any support persons in attendance. The study and consent process were explained to potential participants utilizing a recruitment script (see Appendix B). If the PALS expressed interest in participating, they were asked to identify a primary support person over the age of 18 who the patient identified as a member of their non-professional support system. If a primary support person was able to be identified who also met the study's inclusion criteria, the lead researcher scheduled an interview with them. If the PALS' primary support person was not onsite with the patient, the lead researcher attempted to contact that person with the PALS present and inquired to that person's willingness to participate. If the researcher was unable to contact that person, the PALS participant was encouraged to notify the potential primary support person in advance that they would be receiving a phone call from one of the study researchers so they were aware of its relevance and that a call would be coming in from one of the study researchers. If the primary support person was not available to answer the phone, the researcher left a voice message using a telephone script (see appendix B) that included the lead researcher's name, the name of the study, and a contact number that they could be reached at in response.

Following the explanation of the study, the consenting researcher addressed any questions or concerns expressed by either the PALS or the primary support person. After the study had been explained to participants and they expressed interest in participating, the lead researcher scheduled interviews to take place at the Vidant ALS Clinic, in their homes or via Web-Ex. Once the PALS, support persons, and lead researcher were together for the interview, the informed consent documents were provided to the participants prior to the start of the

interview, explaining that their participation was optional and that they were able to end their participation at any time with no consequences. Following the completion of the consent documents by both the PALS and all participating support persons, the participants were asked to complete the demographics survey. The interview began only after the consent documents and demographics survey were complete.

Data Collection

The current study involved data collection from qualitative interviews. To ensure confidentiality, the researcher assigned pseudonyms in place of participants' actual names to de-identify the data collected. Pseudonyms were used in the transcripts and in any presentations/publications that result from this study. Pseudonyms were assigned to participants at the time of their enrollment unless they were able to provide one themselves. Pseudonyms were added to the participant's contact information (i.e., phone number and email) and were stored in a password protected file on the lead researcher's secure Piratedrive. Note: if during the interviews, the participants mentioned actual names of anyone, those names were changed to pseudonyms as well to protect their identity. However, only the pseudonyms of consenting members were stored along with their actual names and contact information in a pass protected document. A master list of all pseudonyms used were stored in a separate pass protected file on the study's Pirate Drive as well. This master list helped the investigation team refrain from accidentally repeating a pseudonym during the transcription process.

Demographic Information

Demographic information was gathered from participants after the consent process. Information collected included age, sex, race/ethnicity, highest level of education, employment status, household income, relationship status, religious identification, approximate date of first

symptoms, symptom onset type (limb or bulbar) and date of diagnosis. The demographic survey information was collected REDCap electronic data capture tools hosted at East Carolina University (Harris et al., 2009). REDCap (Research Electronic Data Capture) is a secure, web-based application designed to support data capture for research studies (see Appendix D).

In-depth, Open-ended, Semi-structured Interviews

Prior to the start of the interview, participants were reminded of the study's purpose, the consent forms they initially signed, and their right to discontinue participation at any time without question or consequence to care being received at the ALS clinic. The interview was conducted during a one-time, hour long session with all participants simultaneously and was guided by a series of semi-structured interview questions (see Appendix E). Each interview began with the same grand tour question, "How do you believe ALS has impacted you and your support system?" The interview was digitally tape-recorded or recorded on WebEx and was uploaded immediately onto the study's Piratedrive, and transcribed verbatim using NVIVO Transcription. As some PALS may not have the ability to verbally communicate, these participants were given the option to write, type, or speak their responses. In the event that the PALS chose to write or type their responses, the lead researcher clarified and verablized their answers out loud so that they would be audio-recorded. Any interviews digitally recorded using a hand held device were immediately erased once the researcher confirmed it had successfully uploaded onto Pirate Drive. All consent documents, demographic data, pseudonym, and contact information files were stored in the study's assigned password-protected Piratedrive folder. This folder was only accessible to the researchers analyzing the data. It will be maintained for seven years by the faculty supervisor for this study in accordance with IRB regulations.

The lead researcher facilitated all interviews and utilized probing questions to encourage participants to expand upon on their answers to the grand question (Appendix E). Once the interview was complete, the lead researcher informed participants that they had the opportunity to verify their transcript and the results to help ensure the accuracy and trustworthiness of the findings (Lincoln & Guba, 1985). Participants who responded that they would like to participate in this part of the verification process, were provided with a copy of the transcripts by email and were asked to confirm that those results accurately reflected their own lived experiences. Participants who chose to participate were tracked on the same document that contains participant contact information and included which verification portions they chose to participate in, the transcripts, the results, or both. Those participants were asked to proof the transcripts and to add or change any information that they were not able to provide during the interview or that they thought of following the interview. Line numbers were added to each transcript for participants to refer to when offering added or edited text. Emailed transcripts were asked to be returned via email. Changes made to the transcripts by participants were added to the original transcripts and highlighted in yellow so that it was clear that the changes were added following the interview. Final transcripts were determined after all participants who wished to verify the results had either returned their copies or communicated verbally to the lead researcher that no changes were needed.

Data Analysis

This study employed Colaizzi's (1978) data analysis methodology for phenomenological studies. Colaizzi's method was chosen for the current study as it provides researchers with clear, logical, and sequential steps, and increases the reliability and dependability of the obtained results (Wirihana et al., 2018). It differs from other methods of phenomenological data analysis

as it provides participants the opportunity to validate the findings to ensure accuracy and credibility. Following Colaizzi's method of analysis, the following steps were taken: (a) read the transcripts; (b) identify significant statements; (c) form meaning statements from the significant statements; (d) cluster themes from the meaning statements and form emerging themes; (e) create and exhaustive description; and (f) validate the findings. Reflexive journals were used throughout the analysis process to allow the researchers to continuously critically examine their roles in the research process and how their biases may affect the data (Orange, 2016). This reflexivity enhanced several areas of the study to include data collection, data analysis and ethics.

Reading the Transcript

In following the first steps of Colaizzi's (1978) analysis method, the researchers read and re-read each transcript to obtain a global sense of the phenomenon prior to identifying themes from the recorded interviews. The researchers bracketed ideas and potential biases that came from these readings in their reflexive journals.

Identifying Significant Statements

The second step consisted of extracting phrases or sentences (i.e., significant statements) directly from each transcript that pertain to the phenomenon being studied. To ensure that researchers participating in the data analysis part of the study were interpreting the data similarly, researchers coded the first transcript individually and then met to address any discrepancies. If an agreement could not be reached, a member of the researcher team served as a peer debriefer who was brought in to resolve any disagreements regarding the interpretations of the interview. Disagreements between the researchers were documented in the audit trail to include when the disagreement took place and what was done to resolve it. The peer debriefer

was only consulted when the lead researcher and triangulated researcher had attempted to reach an agreement but were unable to do so on their own.

Forming Meaning Statements

In the third step, researchers developed meaning statements from the significant statements (Colaizzi, 1978). They established fidelity to the participant's experiences by ensuring that the statements illuminated the meanings hidden within the context of the transcripts (Colaizzi, 1978). These meaning statements were formulated by using "creative insight" to interpret the meaning of the participants' statements.

Clustering Themes

The fourth step consisted of significant statements and their formulated meanings being grouped into themes and then into thematic clusters (Colaizzi, 1978). Themes and thematic clusters were validated against the original transcripts to ensure that the interpretation of the data could be traced successfully back to the raw data (i.e., participant interview responses). During this process, investigators tracked thematic clusters back to themes, meaning statements, and significant statements. A spreadsheet containing the line number(s) associated with each significant statement, among with its corresponding meaning statement, theme, and thematic cluster was maintained by the lead researcher to ensure interpretations were trustworthy and could be accounted for in the analysis.

Creating an Exhaustive Description

In the fifth step, all findings were integrated into an exhaustive description pertaining to the phenomenon under investigation. This was accomplished by the researchers involved in the analysis re-examining the transcripts, thematic clusters, and themes many times to study any contradictions or different perspectives and to ensure the interpretation of the exhaustive

description is thorough. This exhaustive description was written to capture all of the themes listed from participant's transcripts in the prior steps and included a context that unified them into one cohesive summary of the participants' lived experiences.

Validating the Findings

To confirm that the exhaustive description and fundamental structure of the phenomenon accurately depicts the participants' experiences of the phenomenon, the participants were asked to confirm their validity (Colaizzi, 1978). This step is similar to member checking, defined by Lincoln and Guba (1985) as the most crucial technique for establishing credibility, whereby data, interpretations, and conclusions are tested with members of the stakeholding groups from whom the data were originally collected. In this step, the lead researcher reached out to participants who expressed interest in validating the statement of identification and exhaustive description by telephone or email. The lead researcher asked whether or not the exhaustive description adequately represented the participant's experiences. If contacted by email, participant's were asked to return their responses by email. The responses from this step were also included in the study findings (Colaizzi, 1978).

Verification Strategies

This study followed Lincoln and Guba's (1985) recommendations for trustworthiness to establish credibility, transferability, dependability, and confirmability of its findings. Credibility was established through the use of triangulation, peer debriefing, and member checking. Thick descriptions were used to ensure transferability. Finally, dependability was ensured through the use of an inquiry audit and confirmability was ensured through the use of an audit trail, including a reflexive journal that was used to record ideas about potential themes and to check biases. These processes are further explained in the following paragraphs.

Credibility

Credibility is defined by Lincoln and Guba (1985) as the confidence in the "truth" of the findings, which can be established through activities that make it more likely that credible findings and interpretations will be produced and through activities that provide for the direct testing of findings. Lincoln and Guba suggested four modes of triangulation, to improve the probability that findings and interpretations will be found credible. One of these four modes, the use of different investigators, ensures that one team member is kept more or less honest by additional team members. In the current study, credibility was ensured through triangulation, peer debriefing, and member checking.

Triangulation was achieved by interviewing multiple participants and by including a triangulated researcher in the data analysis process. The triangulated researcher participated at an equal level to the lead researcher in the coding process. Researchers independently read through and coded each interview transcript prior to joining together to discuss and resolve any differences in coding. If the lead researcher and triangulated researcher were unable to resolve disagreements, a peer debriefer was consulted.

Peer debriefing was also used to establish credibility. Peer debriefing consisted of a peer researcher reviewing the lead and triangulated researchers' analyses. This was done in the hopes of resolving discrepancies and confirming that the interpretation and analysis process was correctly being followed and was absent from researcher biases. The peer debriefer was involved at the end of each data analysis phase, when the lead researcher and triangulated researcher were unable to resolve disagreements on their own.

Finally, member checking, wherein the data, interpretations, and conclusions are tested with members of the group from whom the data was collected, was used as a crucial technique

for establishing credibility (Lincoln & Guba, 1985). Member checking occurred in this study following the transcription of interviews and following the data analysis as it exists as the final step of Colaizzi's (1978) data analysis. The lead researcher reached out to participants who agreed to participate in the member checking process by telephone or email to ask whether or not the transcriptions and exhaustive description adequately represented the participant's experiences. Again, the participants who agreed to participate in member checking were tracked on the same document as participant contact information and included if they chose to participate in verification of the transcriptions, final results, or both.

Transferability

Transferability is the extent to which the findings can be applied to other cases beyond the current study (Lincoln & Guba, 1985). Thick descriptions are thoroughly detailed presentations of the findings that enable a reader to determine whether or not the findings are transferable to other cases. The lead researcher used thick descriptions in the writing of the results by describing the findings in great detail, using participants' words, phrasing, and direct quotes.

Dependability

Dependabiliy shows that the findings are consistent and can be repeated (Lincoln & Guba, 1985). In the current study, dependability was established through the use of an inquiry audit, in which a researcher who was not involved in the data collection and anlaysis processes examined these processes and the results of the study. An inquiry audit comfirmed the accuracy of the findings and ensured that the findings are supported by the data. A member of the research team who was not involved in the data collection or data analysis portions of the current study completed the inquiry audit.

Confirmability

According to Lincoln and Guba (1985) confirmability is the extent to which the findings of a study are shaped by the respondents and not by researcher bias, motivation or interest. Audit trails were used to ensure confirmability in the current study (Lincoln & Guba, 1985). The audit trail consisted of a research log and the lead researcher's reflexive journal. The audit trail materials were electronic documents that were stored in the study's Piratedrive. The research log included a continuous documentation of the entire research process to include any decisions made regarding the protocol and any contacts and communication with others involved in decisions regarding this study. The reflexive journals consisted of the lead researcher and triangulated researcher participating in continuous journaling throughout the research process to identify their own experiences and biases related to the study. This included the lead researcher's thoughts related to the understanding of the participants, insights and biases related to the participants and data, and an on-going process of bracketing those biases by setting aside those experiences and thoughts as much as possible to take a fresh perspective of the phenomenon under investigation (Moustakas, 1994). Several entries from the reflexive journal are included as an appendix (see Appendix H).

Ethical Considerations

The design of this study included several safeguards to ensure the well-being of the participants and be in adherence with the ethical guidelines of the American Association for Marriage and Family Therapy (AAMFT, 2015) and East Carolina University's, University and Medical Center Institutional Review Board. The current study involved minimal risk to participants including time and energy spent completing the surveys and participating in the interviews. Any potential discomfort or emotions that arose from the interview questions related

to the ALS experience were validated by the lead researcher during the interview and community-based resources were provided for additional support if needed (see Appendix F). These minimal risks were accounted for with appropriate safeguards intended to reduce them.

Participants were informed that they were free to pause their participation in interviews or that they could withdraw from participation entirely, at any time during the study if they felt overwhelmed and unable to continue. Safeguards implemented during the recruitment, enrollment, and data collection processes included the de-identification of participants using pseudonyms and password protected files stored on the University's IRB approved Pirate Drive server. Lastly, this researcher commited to following the study through to dissemination of its results. It is important to honor the participants' time and contributions by having and following through with a dissemination plan. This plan may include professional presentations and publications and will include all dissertation committee members unless they opt out of each opportunity as it is presented.

Summary

This qualitative study was designed to investigate the impacts of ALS on family functioning and members' BPS-S health. Utilizing qualitative methods allowed for further exploration of how the type of symptom onset, either bulbar or limb, and BPS-S components may impact the family functioning and health of PALS and their participating support persons. The results of this study will inform future ALS research studies, as well as help craft policy and make programmatic recommendations designed to improve and enhance the BPS-S health of the family unit.

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CHAPTER 5: THE EXPERIENCE OF ALS THROUGH A SYSTEMIC LENS: A PHENOMENOLOGICAL STUDY

There are approximately three to five new cases per 100,000 people diagnosed with Amyotrophic Lateral Sclerosis (ALS) each year in Europe and the United States (Brown & Al-Chalabi, 2017). ALS is a progressive, neurodegenerative disease, marked by muscle weakness, swallowing difficulties, slow or slurred speech, respiratory failure and eventual death (Edge et al., 2019). This non-curative, progressive, neurodegenerative disease most commonly begins with symptoms and weakness in the limbs (limb onset), with about one third of the cases presenting with difficulty chewing, speaking, or swallowing (bulbar onset; van Es et al., 2017). However, as this disease leads to a loss of abilities, the support system steps into a critical role of assisting their loved one with managing its impacts (Krivikas, Shockley, & Mitsumoto, 1997; Mockford et al., 2006; Rabkin et al., 2009).

Caregivers spend an average of 11 hours every day helping patients maintain a good quality of life (Krivikas et al., 1997). Due to the essential caregiving needs of patients with ALS, family members reported negative health impacts that are physical (Hwang, Weng, Tsai, & Chang, 2014; Tramonti et al., 2015), emotional (Burke et al., 2018; Pagnini et al., 2012; Tremolizzo et al., 2016), social (Galvin et al., 2016; Sicilliano et al., 2017), and spiritual (Roach, Averill, Segerstrom, & Kasarskis, 2009) in nature. However, each of these studies has focused on the biological, psychological, social and spiritual (BPS-S) domains individually versus interactively, and without considering how the family's style of functioning impacts it.

The biological, psychological, social, and spiritual (BPS-S) framework (Engel, 1977; 1980; Wright, Watson & Bell, 1996) helps researchers and clinicians make sense of the comprehensive implications of illness by viewing health through a more systemic lens. Its intent

is to encourage researchers and clinicians to assess, understand, and treat systems as a whole versus only focusing on one individual or health domain. Beavers systems model of family functioning (Beavers, 1981) identified two family types that emerge in response to stressors like a terminal illness. Centripetal families respond by pulling inward and huddling together; whereas centrifugal families psychologically, socially, and physically create distance (Kelsey-Smith & Beavers, 1981). Thus far, the ALS research on families focuses on individual family members' experiences. For example, spouses reported experiencing fear, lack of understanding (Ozanne & Graneheim, 2017), and even isolation (Ozanne, Graneheim, & Strang, 2015) in their caregiving roles. What is unknown is how caregivers' BPS-S health is impacted by the systemic family functioning within the family. Exploring the impact of ALS, utilizing a systemic measure of family functioning and the BPS-S components of health by using multi-person data, fills a gap in the science and literature by looking at health research and caregiving resources more comprehensively.

To gain a more thorough understanding of the experiences of living with ALS and the impacts of the disease on the patients with ALS' (PALS) and their family members' BPS-S health and family functioning, a descriptive phenomenological design (Husserl, 1970) was utilized in the current study. A phenomenological design was chosen to explore the lived experiences of PALS and their support persons (SPs) and further the research on how family functioning is impacted, as well as family members' BPS-S health. The following section provides a detailed description of the phenomenological method used in this study.

Method

Descriptive phenomenology, as understood by Husserl (1970) focuses on the commonalities of lived experiences, concepts, or phenomena amongst several individuals. Its

basic purpose is to collect individual narratives and analyze them to capture the universal essence among their lived experiences. The following research question was selected for the current study, "What are the experiences of patients - and their identified primary support persons - living with ALS and how has it impacted family functioning and health?" Approval was obtained from the East Carolina University and Medical Center Institutional Review Board (IRB) for this qualitative study (see Appendix A).

Participants

Purposive sampling was used to enroll participants from a Southeastern academic medical center ALS clinic. The ALS Clinic serves approximately 30 unique patients each year. PALS were considered eligible for participation in the study if they met the following inclusion criteria: (a) English-speaking; (b) aged 18 or older; (C) the PALS has a formal diagnosis of ALS, and (d) both a PALS and at least one primary SP agreed to participate in the study. Patient participants identified SPs for possible inclusion in the study. SPs were considered eligible for participation based on the following inclusion criteria: (a) English-speaking; (b) aged 18 or older; (c) is a SP of a PALS who has a formal diagnosis of ALS; and (d) both a PALS and at least one primary SP agreed to participate in the study.

Exclusion criteria for PALS included: (a) PALS' positive screening for frontotemporal dementia or other types of dementia as determined by ALS Clinic staff; and (b) communication challenges unable to be resolved with technological assistance already in use by the patient participant. In addition, the one exclusion criterion for SPs involved any cognitive impairment that would interfere with the SP's ability to consent to participate. It is important to note that participant recruitment and data collection was completed prior to the start of COVID-19 pandemic in the United States. Investigators believe given the nature of this study, the pandemic

would have altered participant responses related to family functioning and social distancing requirements.

Following the enrollment of participants based on inclusion and exclusion criteria, the lead researcher then conducted the audio-recorded, in-depth, open-ended, semi-structured interviews until saturation of themes was reached. Saturation of themes was determined to have occurred when the analysis of additional data from participant interviews yielded no new themes (Glaser & Strauss, 1967). Out of nine eligible PALS, eight participated in the study. One eligible patient was not enrolled because the lead researcher was unable to contact the individual to schedule an interview. All nine PALS had at least one SP participate in the interview, while one PALS chose to have two SPs participate. Participants in the study included five male PALS, aged 59 to 76, and three female PALS, aged 43, 48 and 79, with two male SPs, aged 44 and 49, and seven female SPs, aged 49 to 75 (see Table 1 for participant demographics). Of the included PALS, one had a known diagnosis of bulbar onset ALS, five had a known diagnosis of limb onset ALS, and two shared that they did not know their exact onset of ALS.

Data Collection and Procedures

Eligible patients were recruited using a two-pronged recruitment process. A recruitment script was used in either approach to ensure all eligible participants heard the same information. The first recruitment strategy took place over the phone. A triangulated researcher, who participates as a secondary researcher in a study in order to validate findings, (Lincoln & Guba, 1985) employed within the clinic, would screen for PALS who met the inclusion criteria and contact them to determine if they were interested in enrolling in the study. Interested PALS were asked to identify a SP who might also be interested in participating. This potential SP participant

was contacted by the lead researcher to explain the study's intent and if willing to participate, schedule the interview.

The second recruitment strategy took place in-person at the ALS Clinic. Once a month the ALS clinic devotes a full day to seeing new and returning PALS. The study's triangulated researcher would screen PALS for eligibility and any SPs in attendance. If they agreed to participate, their contact information was shared with the lead researcher who contacted them at a later date to schedule an interview.

Prior to beginning the in-depth interviews, PALS and all participating SPs first completed the consent document and demographics survey. Of the eight interviews conducted, one took place via video teleconference, two were conducted at the ALS Clinic, and five were conducted in patient's homes. This strategy was done to help accommodate the varied physical and communication abilities and accessibility needs of each participant.

At the beginning of each interview, the lead researcher invited participants to choose pseudo names for themselves to de-identify the collected data and protect their confidentiality. If the participants did not wish to select their own pseudo names, the lead researcher chose for them. Following the interviews, the interviews were transcribed verbatim using NVivo qualitative data analysis software (QSR International Pty Ltd. Version 12, 2018). Once the interviews were transcribed, participants who expressed interest in member checking opportunities were emailed their transcripts and asked to add to or clarify any responses so as to better reflect their experiences. Of the seven participant groups who expressed interest in this member checking process, one participant shared additions to be added to the interview transcript. These were added to the original transcripts and were highlighted in yellow to identify them as additions. The lead researcher, with the help of a triangulated researcher and a peer

debriefer, coded the transcripts until thematic saturation was reached. The peer debriefer is a noninvolved professional with whom the researchers discuss and resolve difficult processes throughout the study (Lincoln & Guba, 1985). Although all interviews began with the same grand tour question: "How do you believe ALS has impacted you and your support system?" the lead researcher used an interview guide to provide a semi-structured interview experience in which all participants addressed similar topics related to their BPS-S experiences with ALS (See Appendix E for Interview Guide).

Data Analysis

Following each interview, the lead researcher and co-researcher applied Colaizzi's (1978) phenomenological data analysis methodology to the data. The clear, sequential steps outlined in Colaizzi's method increases the reliability and dependability of the results (Wirihana et al., 2018). In following Colaizzi's data analysis method, the following steps were taken: (a) read the transcripts; (b) identify significant statements; (c) form meaning statements from the significant statements; (d) cluster themes and identify emergent themes; (e) create an exhaustive description, and (g) validate the findings. It was determined to merge the exhaustive description with Colaizzi's step of forming a statement of identification in this study based on previous literature (Kristianingrum, Wiarsih, & Nursasi, 2018; Yambo et al., 2016; Yodchai, Hutchinson, & Oumtanee, 2018).

The first step consisted of identifying and coding significant statements. The lead researcher and a triangulated researcher both worked through all transcripts and identified an initial 304 significant statements, resolving any disagreements as they were presented. Duplicate significant statements were removed, resulting in a total of 278 significant statements. The researchers then formed meaning statements from the significant statements. The triangulated

researcher participated in 50% of meaning making statements from the significant statements resulting in 278 meaning statements, with 98% inter-rater agreement. The researchers discussed any disagreements until agreements were reached. Finally, 112 duplicate meaning statements were removed, resulting in 166 final meaning statements at 100% inter-rater agreement.

The lead researcher and triangulated researcher categorized meaning statements by thematic clusters, resulting in 15 thematic clusters with 97.5% inter-rater agreement. The 15 thematic clusters were then reduced to 5 emergent themes (Table 2). As the two researchers were unable to decide on the inclusion of a fifth theme, impacts on spirituality, a peer debriefer was consulted to help reach a decision. Selected examples of narratives and emergent theme formation are included in Table 3. Lastly, an exhaustive description was developed to capture all emergent themes in one comprehensive narrative.

Following the creating of the statement of identification, the results were validated using a second level of member checking. Participants who wished to participate in the second level of member checking were emailed the exhaustive description and final statement of identification developed from the findings. They were requested to provide feedback of either agreement or disagreement with the summative results. Out of the 15 participants who wished to participate in the member checking process, 7 confirmed that the exhaustive description appropriately reflected their lived experience. The remaining participants could not be reached for confirmation.

Verification Process

Since qualitative studies determine trustworthiness of its findings differently than quantitative, Lincoln and Guba's (1985) recommendations for increasing confidence in qualitative results was applied. Credibility (i.e., internal validity) was established through the utilization of a triangulated research, peer debriefer, and use of a two-step member checking

strategy. Transferability (i.e., external validity) was established through the utilization of thick descriptions. This involved writing of the results using participants' exact words, phrasing, and direct quotes so consumers of the study could determine its applicability to their context. Dependability (i.e., reliability) was monitored through the use of an inquiry audit. Each member of the research team who was not involved in the data collection and analysis processes examined these processes and the results of the study to ensure all findings were grounded in the actual participant data and all methods were followed appropriately. Audit trails were used to ensure confirmability (i.e., objectivity) in the current study and consisted of a research log and the lead researcher's reflexive journal stored in a pass protected file saved to a secured and encrypted server. The reflexive journals were utilized by the researchers to engage in the bracketing of biases. The researchers engaged in bracketing throughout the analysis process to ensure that the findings were true to participant experiences and were not altered by researcher bias. The research log included a continuous documentation of the entire research process and the reflexive journals consisted of the lead researcher and triangulated researcher participating in continuous journaling throughout the research process to identify their own experiences and biases related to the study.

Findings

Eight interviews were completed with 17 participants, eight PALS and nine SPs. Participant groups is used throughout to describe the groupings of participants in interviews with the PALS and at least one SP. Interviews with the seven dyadic units and one triad yielded 278 final significant statements which were reduced to 166 formulated meaning statements. In response to the original research question, "What are the experiences of patients - and their identified primary support persons - living with ALS and how has it impacted family functioning

and health?" five themes emerged when meaning statements were collapsed into thematic clusters. The themes capture a common, recurring pattern in the data that are organized around a central concept, while the thematic clusters exist within a theme, sharing the central concept but focusing on one specific element of the theme (DeSantis, & Ugarriza, 2000). The five themes are as follows: (a) dynamic transformation of relational systems; (b) biological changes and well-being; (c) emotional processes; (d) spiritual adaptations and anchors; and (e) healthcare system interactions. These themes were initially identified by the lead researcher and triangulated researcher after analyzing the data. The study's peer debriefer assisted in combining and naming of the five final themes after reviewing the actual data and discussing it with the research team. The following section provides an in-depth description of each theme, including participants' quotes. This section concludes with an exhaustive description that captures the lived experiences of PALS and their SPs.

Theme 1: Dynamic Transformations of Relational Systems

All participant groups reported changes in their relational systems with six participant groups sharing that ALS has brought them closer together and two participant groups sharing that their ALS experience has not pulled them apart, but it has not necessarily brought them closer together either. Three thematic clusters emerged within this theme that reflect how dynamic relationships are altered in response to the intrusion of ALS. These three clusters include social persons becoming closer, social persons pulling away, and challenges with social interactions and support.

Thematic Cluster 1a: Support Persons Becoming Closer

Participants expressed how relationships with family members and friends had grown emotionally closer or improved since living with ALS. When discussing how ALS brought

relationships closer together, SP Sharron reported, “I think it's brought us closer. I feel like we're more. We've grown closer almost because we've had to.” Emotional closeness seemed to be a binding force that strengthened the families. PALS Sarah said, “It's brought us closer…More of a stronger heart bond and not so much as a physical one.” The impact of ALS not only brought already close members together, but also extended emotionally distant members the opportunity to heal longstanding wounds. SP Kathy shared, “For 20 years, she [PALS] didn't speak with her [daughter]. So, this is sort of like, you know mom has ALS, we've got to go see her. So, it opened up the love line again. Everybody came.”

In addition to family members and how closeness intensified, participants noted the positive impact friends had as well. SP Rosie said, “We have a lot of friends and they visit, and they've gone through the part where he could speak some. Then he couldn't speak. But they still come and it's good to have them visit.”

Thematic Cluster 1b: Support Persons Pulling Away

In addition to ALS having brought the family and other SPs closer together, participants also experienced friends and some family members grew distant. SP Sharron observed, “A lot of his friends don’t come around and stuff anymore. And um, he doesn’t get out much, you know, to go see them.” Understanding why this was happening was attributed to very different causes. SP Rosie explained that the physical effects of losing speech was a notable cause for changes in their social network. She said, “Some [friends] that they'll visit with me and just speak to him and leave because they know he can't talk. In fact, he has a brother that didn't come from the longest time.” However, others observed that family members who struggled with the diagnosis itself pulled away much sooner. SP Ruby shared…

> I still don't think he's [my son] totally accepted that he's, his father has ALS. We have a walk every year and the first couple of years he didn't come at all. He did come twice, and he says, "I'm not coming anymore", then I thought, you know, that to me is just telling me something. It just it really upsets me because everybody that has got ALS, their family's there. I have more friends come than I have family.

PALS Frank expanded on this and the hopefulness that he believes is at the root of his adult child's distancing behavior. He explained, "My daughter. I don't think she really understands a lot about me. I mean, she thinks I'm to get rid of this [feeding tube]. I don't think I'll ever get rid of this.

Thematic Cluster 1c: Challenges with Social Interactions and Support

In addition to observations of family and members of the support system being brought closer together or apart after an ALS diagnosis, participants discussed challenges when attempting to increase social interactions and support. Challenges that inevitably lead to feelings of social isolation. SP Tonya explained that many challenges with socialization begin with identifying a location that will meet their needs. She said, "We have to plan any trip outside our home to ensure our destination is accessible and [that we] know where an accessible bathroom is." PALS Frank explained, "It's hard for people to understand what I need to get in and out of the car and up the steps." They also noted how hard it is to move about in crowds and navigate physical spaces to socialize with others. For SP participants even leaving the PALS alone to socialize seemed unrealistic. SP Daniel shared, "I want to go out sometimes, you know, like go to town for a couple hours, but I don't feel good going when I get there because …there's nobody here [with her]."

Theme 2: Biological Changes and Well-being

Both PALS and SPs shared how living with ALS had impacted them biologically. Participants spoke about how they have experienced the progression of ALS symptoms and specifically the challenges associated with changes in communication. Additional biological changes included general changes in physical well-being to include fatigue, aches and pains, lack of exercise, and even difficulty concentrating. Thematic clusters within this theme include: (a) participants' reactions to the slower progression of symptoms, (b) communication challenges that result as symptoms progress, and (c) general biological and physical challenges.

Thematic Cluster 2a: Responses to Slower Progression

Several participants shared experiences with slower symptom progression. SP Tanya spoke about the unpredictable progression of her husband's ALS and the gratitude she had for more time with him. She shared…

> When he was diagnosed his doctor told him he had about two years to live. That was seven years ago so we're very fortunate his ALS progresses slowly...this allowed us to be more prepared for the day to day and the future.

Others noted that the ALS symptom onset type (i.e., bulbar or limb) made a difference in their physical adjustment to ALS, preferring limb over bulbar onset. SP Kathy shared, "[PALS Sue] feels like this [limb onset] gives her a longer life by starting in her legs." Bulbar onset appeared to be the ALS pathway that was more disruptive and challenging to participant groups.

Thematic Cluster 2b: Communication Challenges

Distinct from the physical challenges of bulbar onset, were the actual communication challenges experienced as symptoms progressed. SP Linda explained what it has been like for her husband to no longer have the ability to communicate verbally. She shared, "The

communication thing, it just shuts you off." PALS Jon, who communicates with an electronic device shared, "I am used to talking as a former teacher and principal. I cannot talk to my dear wife or anyone else." Finally, SP Claire spoke of the challenges they face with the PALS' inability to communicate, "You don't know what's going on and we can't help him and he's trying to tell us. You don't know what to do and I feel so sorry for him because he's trying to tell you."

Thematic Cluster 2c: General Physical Challenges

In addition to experiencing physical challenges related to symptom progression and communication difficulties, participants also expressed challenges with their general physical well-being in addition to the physical progression of ALS. PALS Frank said, "My back hurts so much sometimes I just have to sit. Because I feel like I'm gonna be so weak in the next minute that I'm gonna fall. PALS Jon explained for his wife, "It is hard on Linda as it fatigues her. She has to pick up the load I can no longer carry." Furthermore, SP Rosie shared how her caregiving responsibilities had impacted her biological well-being, "I'm tired. Having the nurses is a blessing through the day. It's kind of rough to do nights and days, so I try to do my stuff and rest a little bit and I'm still not ready for the nighttime."

Theme 3: Emotional Processes

Throughout the interviews, participant groups expressed a variety of feelings related to their journeys with ALS. Participants' responses included denial about having ALS as well as anger and frustration over the continuous progression of symptoms, subsequent changes in ability, and unrelenting adaptations to communication challenges. Participants also shared moments of depression and other difficult emotional processes related to the diagnosis period with all reaching a place of acceptance of the illness. Thematic clusters that emerged in this

theme included feelings of (a) denial, (b) anger and frustration, (c) depression and other emotional difficulties, and (d) acceptance of living with ALS.

Thematic Cluster 3a: Denial

In the beginning stages of living with and adjusting to a diagnosis of ALS, participants shared feelings of denial about the diagnosis or about decreasing biological functioning and abilities. PALS Lisa shared that she was not convinced of her shocking diagnosis when she stated, "That's one of the reasons I haven't accepted the diagnosis. I know something's wrong, but other than my speech, I have the justification for everything except my speech." Other participants did not entirely deny the diagnosis but were in greater denial over the PALS' increasing lack of ability. PALS Donald said, "I get frustrated a lot. I have to sit down and think about it, what I used to do and what I can't do," while PALS Frank explained his process of coming to terms with his limitations, "Some days I can get up and get in this [walker] and walk around and go outside and do that. But then some days I depend too much on the cart and I'll sit there, and I get comfortable." Just as participants shared periods of denial throughout their journey with ALS, they frequently spoke about their anger and frustration as well.

Thematic Cluster 3b: Anger and Frustration

Anger and frustration seemed to be commonly experienced among participants. These experiences were more pronounced with the progression of symptoms as challenges could no longer be ignored. Participants expressed feelings of anger in response to changes in biological functioning, SP Rosie said, "Well, I want to be angry at times. And, I think he (PALS Ronald) does too." Interestingly, expressions of frustration were more tied to relational issues such as lack of recall related to caregiving needs or PALS' inability to communicate what is needed. PALS Frank explained, "It's frustrating for her [his wife and SP] to remember everything [I

need]," SP Claire shared, "It can be really frustrating when I don't know what he wants, and he can't communicate that with me." Several other participants used words other than anger to express their mood state. For example, SP Daniel explained, "Well, I ain't angry. It just frustrates me."

Thematic Cluster 3c: Depression and Emotional Processes

As participant groups continued to explain their experiences of living with ALS, their shared experiences with denial, anger, and frustration often then turned into conversations about depression and other emotional difficulties. SP Linda spoke openly about her and her husband's depression, stating, "He was terribly, he was depressed. I was depressed." Observations of depression noted that it appeared to worsen as ALS progressed. PALS Sue explained that "It's not easy as it gets worse." While resources aimed at providing support to ALS patients and caregivers are beneficial, SP Sharron shared how that can also be very depressing. She shared, "very depressing because everyday somebody is diagnosed, and everyday people are losing somebody…so it's hard to read about that." While depression and other difficult emotions were identified throughout the ALS journey, several participants also expressed acceptance of living with ALS.

Thematic Cluster 3d: Acceptance

All participants expressed feelings of acceptance over living with ALS. They noted how it brought out their more positive characteristics and lead to their ability to make meaning out of it. PALS Frank shared how he grew to accept living with ALS. He noted, "I guess I'm getting used to it. I'm probably a strong-willed type person. I don't let things bother me that much. I try and take it in and then just say, that's it. Nothing I can do." PALS Sarah shared similar sentiments, "So it's just accepting the journey I have, and I have a purpose." The identity of

being a caregiver was not something that SPs thought they would hold, until ALS was diagnosed in the PALS. SP Linda shared how she learned to cope with the disease's progression, make adjustments, and manage the disappointments that came with its harsh reality. She shared…

> You learn to cope with the changes. And you adapt and you get through with that and so your life just kind of well, it's kind of a new world. But you, you know, you make the adjustments and you just go on and you kind of try to get through the disappointments. Things that you had planned to do, you'd hope to do, aren't going to happen now.

Theme 4: Impacts on Spirituality

While purpose was noted within the aforementioned thematic cluster of acceptance, it also tied into how participants made sense of their experiences with ALS. Some saw it as their way to make meaning out a devastating illness. With regard to religious affiliation, most participants identified as Christians, except for one PALS who did not identify as a religious or spiritual person. When discussing how their spirituality or faith had been impacted throughout their journey with ALS, two thematic clusters emerged to include, participants faith having been maintained and their faith having been questioned.

Thematic Cluster 4a: Faith Maintained

For some participants, living with ALS had either no impact on their faith or had made their faith stronger. When explaining the impacts ALS had on spirituality, PALS Jon shared, "I have a strong sense of another life beyond this one. This ALS has made that more real to me and I am at peace." PALS Sue explained why her faith and relationship with God had gotten better, "Because I'm living proof of what happened, the bad and ugly that happened to me in two years. But this [situation] is the best." SP Kathy further expressed how her faith has given her strength as a caregiver, "So here's my thing, when I pray, which I pray a lot, God talks to me and He says,

‘Kathy, you can do this. It's only for a moment.’” Although several PALS and SP participants explained that their faith had increased or had given them strength, others expressed that they had struggled with their faith since living with ALS.

Thematic Cluster 4b: Faith Questioned

Participants shared that living with ALS had resulted in strains on their faith. SP Rosie explained that the inability to physically attend church had been difficult for them and their faith, “I don't know if you're church going people but when you, you know, you can believe and you can be a Christian, but it really takes being at the church, being with people to get fulfilled.” SP Linda shared her own struggle with her faith, “It's just kind of hard to understand how things work out the way they do. Trying to find peace has been one of my seemingly things, one of those things that I just can't quite get.” However, PALS Bryan found that time alleviated some of the doubt, “At first our faith was in doubt. However, with time we have come to terms with having an uncertain future and learned to cherish every minute together.”

Theme 5: Healthcare System Interactions

Due to the nature of ALS, all participants reported that a part of their experience of living with ALS included being seen routinely in the healthcare system, resulting in this fifth emergent theme. Visits included time spent in primary care offices, specialists’ offices, assisted care facilities, and ALS clinics. Participants expressed pleasure with medical care received at the ALS clinics, they shared that additional support, knowledge, and guidance around certain issues would have helped more with their ALS experience. Thematic clusters within this theme include additional education for health care providers, the need for additional support from medical providers, and the appreciation of ALS clinics.

Thematic Cluster 5a: Additional Education for Healthcare Providers

Participants explained that they often had interactions with medical professionals who were not familiar with ALS. PALS Sarah explained, “A lot of just people who don’t specifically work closely with ALS know it’s a motor neuron disease, but they don’t know the effects of it.” Frank, another PALS, shared similar sentiments, “Even when I go to the hospital there, sometimes a doctor will be talking to you and he had no idea what was going on with me.” SP Daniel, explained that in addition to traditional medical providers need further education, others involved in medical care need additional knowledge as well, “The rescue squad and the fire departments need to know who in their area has it so they'll know how to take it when they get there.”

Thematic Cluster 5b: Additional Support Needed from Medical Professionals

Participants also repeatedly spoke about their need for additional support from medical providers during and following the diagnosis period. SP Steve shared, “Once you’re diagnosed with ALS, a lot of medical doctors won't even see you for anything else.” SP Rosie provided insight into the perceived lack of support during the diagnostic period:

> It was just frustrating, challenging too, that we had to find the doctor, had to get it going, get his diagnosis and, I don't know. I just felt like they [medical providers] just weren't helping us like they should. We had a problem that needed to be seen about and they just weren't doing anything about it.

When explaining the additional support needed from non-ALS medical providers, SP Kathy shared, “If they could just be a more, empathetic, instead of just seeing the symptoms of the disease instead of the person” and Lisa, a PALS, explained that she just needed for the providers to listen, “Just listen. They [the providers] don’t listen.”

Thematic Cluster 5c: Appreciation of ALS Clinics

Participant groups expressed their appreciation of and satisfaction specifically with the ALS clinic. SP Ruby responded, “They [the ALS Clinic] were excellent. I mean, I would go back there before anything” and PALS Sarah reported, “I’m glad to see everybody because I don’t have to go days on end to different offices. I’m thankful for that.” SP Kathy appreciated how personable the providers are. She said, “So we really feel like the line of communication through ALS is completely wide open, I can call there. I have personal numbers they give me.” SP Daniel shared, “I think they [the ALS Clinic] have done a damn good job with all the knowledge they’ve got.”

Exhaustive Description

Following the identification of themes from participants’ interview transcripts, an exhaustive description was developed. This exhaustive description is intended to capture all of the emergent themes and accurately represent the richness of the PALS and SP’s lived experiences (Colaizzi, 1978). The exhaustive description for this study is as follows:

PALS and SPs living with ALS experienced transformations of their relationships with family members and friends subsequent to receiving the diagnosis. Relationships between the PALS and SP often became closer throughout the ALS journey as PALS and SPs spent more time together and adjusted to patient and caregiver roles. Relationships with friends also increased in emotional closeness following diagnosis. In instances when social supports pulled away both emotionally and physically, it seemed as though these persons were unfamiliar with the nature and progression of the disease or simply did not know how to respond emotionally to the PALS’ progressive symptoms. As the disease progressed, social outings became more difficult physically and this led to a sense of social isolation for both the PALS and SPs.

PALS and SPs shared various difficulties related to the physical progression of ALS. General challenges in physical well-being were tied to fatigue and aches and pains associated with living with ALS. As symptoms progressed, participants reported greater difficulties with physical challenges, such as lifting and moving, as PALS required greater assistance with the increasing lack of mobility and with verbal communication. This loss of communication led to feelings of being shut off from the rest of the world. Several participant groups expressed how fortunate they felt to experience slower progression or to experience symptoms that began in the limbs versus starting with loss of speech.

Emotional challenges were experienced by all participant groups. PALS and SPs shared feelings of denial and shock over a diagnosis of ALS, often followed by feelings of anger and frustration in living with the disease. Participants reported experiencing moments of depression and other emotional difficulties as they coped with the loses that came with symptom progression. With the ever-changing progression of symptoms, participants shared that moments of emotional difficulties were followed by a peak and adjustment to physical changes until additional loss of ability brought about another round of emotional difficulties. With time, several PALS and SPs expressed that they had come to accept their journey with ALS having moved past the emotional difficulties, either through attempting to continuously find a new normal, learn and adapt to ever changing abilities, recognizing their purpose in living with ALS, or by simply choosing to stay positive.

Participants communicated the impacts that living with ALS had on their spiritual well-being. Several participants conveyed that the diagnosis of ALS had not given them reason to question their faith with several reporting that the diagnosis had in fact made their faith even stronger. Some participants did express that they had struggled with their faith and finding peace

with the diagnosis with the majority of those struggles fading with time. A few of the participants also found their faith to be a coping source as they found comfort in prayers and talking with God.

PALS and SPs living with ALS conveyed the importance of their interactions with healthcare providers throughout their illness journeys. Specialists in ALS clinics were valued for their knowledge of the disease and for their continuous communication with PALS and their families. However, participants also conveyed a strong need for increased support from medical professionals, especially during the diagnosis period. Participants shared their frustrations over the inability to reach a diagnosis in a timely manner and over non-ALS medical provider's inability to recognize the first symptoms of ALS that may have led to a faster diagnosis. Similarly, participants expressed frustration with the general lack of knowledge about ALS in medical providers outside of ALS clinics, sharing that first responders, primary care providers, and even health providers in assisted living facilities should have at least a general knowledge of ALS and the proper care needed for PALS and SPs. Despite participant frustration with the lack of ALS knowledge and need for additional support from providers during and following diagnosis, it appears as though ALS clinics were ultimately seen by PALS and SPs valuable resources. ALS clinics seemed to provide a sense of responsiveness, critical care, and knowledge, especially when participants were experiencing relationships with family members and friends pulling away.

Discussion

The purpose of this study was to explore the experiences of PALS and their identified primary SPs living with ALS and how it has impacted family functioning and health. Previously, two studies have attempted to understand the impact that ALS has on family dynamics and

functioning. One study (Tramonti et al., 2014), including PALS and family caregivers, focused on family cohesion and adaptability. The study included individual responses to the FACES III Questionnaire (Olsson, 1986) and did not allow participants to provide verbal explanations to their responses. Furthermore, the participants completed the questionnaire individually, suggesting a potential limitation as the findings are based on singular experiences without considering multiple family perspectives simultaneously (Fingerman, 2001). Martin and Turnbull (2001) conducted a study on overall family relationship satisfaction after experiencing the loss of a family member with ALS. They found 67% of participants believed that the disease brought the family closer together. However, PALS were not participants and data was collected by only one member of the family system, again failing to consider multiple family perspectives. Additionally, the study fails to provide insight into the family system's experience while living with ALS and does not consider all BPS-S components. Considering these previous studies, the current study attempted to add to the literature by including multiple members of ALS support systems in singular in-depths interviews that allowed participants to explain their own experiences related to the impacts of ALS on their family functioning and BPS-S health.

One novel finding of including several members of the same support system in one interview is the broader understanding of the PALS and SPs experiences with the impacts of ALS on family functioning. While previous studies focused on the impacts of ALS on the individual patient (e.g., Mock & Boerner, 2010; Pagnini et al., 2012) or family caregiver (e.g., Tramonti et al., 2015; Siciliano et al., 2017), this qualitative study approached the topic from a systemic perspective (von Bertalanffy, 1950) including multiple members of the system. Some participants in the current study found that their experiences with ALS brought the family system closer together. This finding was consistent with what Beavers and Voeller (1983) noted when

they developed their theory that illness can ignite a centripetal pull, leading the family unit to be more insular and interdependent. Additional participants seemed to experience centrifugal patterns (Beavers, 1981) within their support system, sharing that some SPs stepped away emotionally and physically due to not knowing how to offer support to those living with ALS.

The findings of the current study present an additional novel finding when considering participant explanations of the impact of ALS on their BPS-S health. Previous literature on BPS-S health (Engel, 1970; 1980; Wright et al., 1996) of PALS and SPs reveals changes over the course of the disease to include increased fatigue and declining physical health simultaneous to the progression of ALS (Gelinas, O'Connor, & Miller, 1998; Oyebode, Smith, & Morrison, 2013), depression and general emotional difficulties (Gauthier et al., 2007; Miyashita et al., 2011) and the importance of spirituality and faith (Calvo et al., 2011; Trail et al., 2003). However, in the current study, participants also added specific perspectives as to how the progression of physical decline and subsequent decrease in verbal communication abilities left the families psychosocially in a more isolated and frustrated state. Although participants in the current study also shared their own experiences with depressive symptoms and general experiences with the emotional processes, all participants spoke of their ability to reach acceptance of the disease, which is not reported in previous studies. Finally, in agreement with the previous BPS-S literature on ALS (Calvo et al., 2011; Trail et al., 2003), participants shared how their continued involvement in church and other spiritual activities is important to their well-being.

A final novel finding of the current study is the shared experience of participants expressing the need for medical providers outside of ALS clinics to have increased knowledge of ALS and how to properly care for PALS. Although previous literature has studied the

importance of multidisciplinary ALS clinics (Kiernan et al., 2011), the research does not seem to explore the lack of ALS related education for medical providers outside of those ALS clinics. In the current study, participants frequently expressed their perceived need for increased education for non-ALS providers based on poor experiences with primary care providers, hospital providers, and providers in assisted living facilities. They shared providers refusal to care for the PALS simply because of a lack of knowledge on how to properly do so and when they did receive care outside of the ALS clinic, there seemed to be no communication between outside providers and ALS clinic providers concerning that care.

Future Research

Based on the findings of the current study, the following research recommendations should be considered. The participants shared experiences of their connected BPS-S health (Engle, 1970; 1980; Wright et al., 1996) revealing the importance of treating PALS and their SPs' BPS-S health components simultaneously as these components are inherently connected and continuously interacting. For example, PALS and SP participants shared that biological challenges were directly related to their emotional and social well-being, which in turn impacted their spirituality, revealing first-hand the interactions between each of the BPS-S components. Future studies are needed that focus more on the immediate and longitudinal whole health needs of PALS and SPs, their interactional effects, and how ALS clinics that provide direct care for PALS and SPs impact short and long term BPSS health outcomes.

Participants' perspectives surrounding the lack of ALS knowledge by non-ALS specialists should also be studied further. Similar to the recommendations of a recent study on neurology residents' ability to empathetically deliver a diagnosis of ALS (Schellenber et al., 2014), participants in this study endorsed the need for more education to non-ALS specialists on

the diagnosis, treatment, and BPS-S health impacts of ALS on the patient and family system. Research could include Delphi studies (Dalkey & Helmer, 1963), where experts in ALS from a variety of specialties (e.g., medicine, behavioral health, pulmonary, occupational therapy) identify competences that non-ALS specialists should have to help expedite the diagnosis of ALS and BPSS care provided to PALS and SPs.

Finally, future studies on the experiences of living with ALS should continue to study family members and SPs conjointly to ensure that more robust systemic perspectives are represented in the literature, pivoting away from relying exclusively on individually oriented research to inform treatment. Previous studies largely focused on the perspective of one family member or the PALS to serve as the voice of the family system (e.g., Martin & Turnbull, 2001). This bias creates doubt in the trustworthiness of the findings and their transferability. Qualitative researchers have long argued that individual family members have different perspectives on the same experience (Reczek, 2014). The data provided from individual family members is therefore quite limited, with interviews including multiple members of the family providing the best understanding of the family relationship and family dynamics (Fingerman, 2001).

Limitations

There are several limitations to this study. Firstly, the participants included in the study are both racially and regionally homogenous. The lived experiences of these participants may apply to other PALS and their SPs; however, generalization of findings is not entirely generalizable based on the demographics of current participants. Secondly, several participants expressed that their families had become much closer throughout their journey with ALS, reflecting a sense of centripetal families resulting from ALS (Beavers, 1981). However, it could

be argued that centrifugal families, families who have become distant throughout their journey with ALS, may have had no desire to participate.

An additional limitation exists in the related experiences of the lead researcher. The lead researcher experienced ALS first hand as a family member was diagnosed with and died from ALS. Although both the lead and triangulated researchers participated in continuous bracketing of bias throughout the analysis process, it is possible that the lead researcher's own related experiences resulted in biases that could have entered into the interpretation of the data.

Furthermore, the exclusion of PALS with frontotemporal dimentia (FTD) is an additional limitation. Current research finds that as many as 50% of PALS have symptoms of (FTD) (Ferarri et al., 2011), meaning that several potential participants were automatically excluded from the current study. Although these PALS were excluded due to the challenges involved with participation and communication, it is possible that additional themes may have emerged from these PALS' and SPs' experiences.

Finally, in phenomenological studies, it is recommended to conduct additional interviews past the point of saturation to confirm that all themes have been revealed (Morse et al., 2002). These additional interviews were unable to be completed due to the inability to recruit additional eligible participants amid the COVID-19 pandemic and social distancing practices. Although a triangulated researcher and peer debriefer concluded that saturation was achieved, it is possible that additional themes might be revealed, and additional interviews would have confirmed this.

Conclusion

The utilization of a phenomenological method allowed for the researchers to understand and describe the experiences of PALS and SPs living with ALS in an effort to reduce their explanations into one universally exhaustive description. Findings underscored the value of

social relationships, specifically close familial, centrifugal, relationships (Beaver, 1981), the importance of treating PALS and their SPs from a BPS-S framework (Engel 1977; 1980; Wright et al., 1996) and the importance of continuous support and collaboration from and between both ALS specific and non-ALS medical providers with greater ALS education needed for the non-ALS providers. In providing care for PALS and their SPs, medical providers should capitalize on the benefit of centrifugal family relationships, intervening when families seem to be pulling away. Moreover, non-ALS providers of PALS should attempt to gain an understanding of the basic care needs of these PALS and their SPs, making sure to remain in communication with ALS clinic providers. Caring for PALS and their SPs with their family relationships and BPS-S health in mind, should result in greater health and well-being for all.

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Table 1

Participant Demographics

	PALS (n = 8)	**SPs (n = 9)**
Age	*M* = 65	*M* = 57
Gender	Male: 5 Female: 3 Other (transgender, gender neutral, etc): 0 Prefer not to answer: 0	Male: 2 Female: 6 Other (transgender, gender neutral, etc): 0 Prefer not to answer: 0
Highest level of school completed	Highschool: 4 Some college, but no degree: 0 Associate's Degree: 0 Bachelor's Degree: 3 Master's Degree: 1	Highschool: 1 Some college, but no degree: 3 Associate's Degree: 3 Bachelor's Degree: 2 Master's Degree: 0
Current employment status	Paid Employee: 0 Disabled: 3 Retired: 5	Paid Employee: 5 Disabled: 0 Retired: 4
Employment status at symptom onset	Paid Employee: 3 Disabled: 0 Retired: 5	Paid Employee: 5 Disabled: 0 Retired: 4
Range of current household income	$20,000 to $99,999	$20,000 to $99,999
Health insurance status	Medicaid or Medicare: 7 Tricare: 1 Private: 0 Uninsured: 0	Medicaid or Medicare: 4 Tricare: 1 Private: 4 Uninsured: 0
Relationship status	Single, never married: 0 Committed Relationship: 0 Married: 7 Widowed: 0 Divorced: 1 Legally Separated: 0	Single, never married: 0 Committed Relationship: 0 Married: 8 Widowed: 1 Divorced: 0 Legally Separated: 0
Time in relationship	0 to 3 years: 0	0 to 3 years: 0

	3 to 5 years: 1 5 to 10 years: 0 10 to 15 years: 1 15 to 20 years: 1 20+ years: 4 N/A: 1	3 to 5 years: 1 5 to 10 years: 0 10 to 15 years: 1 15 to 20 years: 1 20+ years: 5 N/A: 1
Number of children	*M* = 2	*M* = 2
Number of children living in the home	*M* = 1	*M* = 1
Religious affiliation	Christianity: 7 Other: 1	Christianity: 9 Other: 0
Time since first symptoms	*M* = 5 years	N/A
Time since diagnosis	*M* = 3.5 years	N/A
Onset Type	Bulbar: 1 Limb: 5 Unknown: 2	N/A

Table 2

Selected Examples of Narratives and Emergent Theme Formation

Significant Statements	Formulated Meanings	Thematic Clusters	Emergent Themes
"I think it's brought us closer. I feel like we're more. We've grown closer almost because we've had to because. You know. It's hard for people to be around people that are dying."	ALS has brought our family closer together.	Support Persons Becoming Closer	Dynamic Transformations of Relational Systems
"Maybe some of both. Some that they'll visit with me and just speak to him and leave because they know he can't talk. In fact, he has a brother that didn't come from the longest time."	Some friends or family don't know what to do or say so they don't visit	Support Persons Pulling Away	Dynamic Transformations of Relational Systems
"I don't get to, you know, do some things by myself or go out with friends as much or out by myself as much anymore."	As a caregiver I don't have as much social interactions.	Challenges with Social Interactions and Support	Dynamic Transformations of Relational Systems
"When he was diagnosed his doctor told him he had about two years to live. That was seven years ago so we're very fortunate his ALS progresses slowly."	We're fortunate for his slow progression.	Responses to Slower Progression	Biological Changes and Well-being
"But it's like I said, without communication it's made it hard."	The lack of communication makes it hard.	Communication Challenges	Biological Changes and Well-being
"But yeah, I've had just aches and pains. I mean, it's more than I've had before, and I know it's lack of exercise.	ALS physically means aches and pains.	General Physical Challenges	Biological Changes and Well-being
"That's one of the reasons I haven't accepted the diagnosis. I know something's wrong, but other than my speech, I have the justification for everything except my speech."	I haven't accepted the diagnosis.	Denial	Emotional Processes
"Emotionally, it can be really frustrating when I don't know what he wants, and he can't communicate that with me."	ALS is frustrating when I can't understand what he needs since he can't talk.	Anger and Frustration	Emotional Processes

“We both have less patience and can be more depressed.”	We’ve become less patient and more depressed.	Depression and other emotional processes	Emotional Processes
“At first there was so much uncertainty, but over the years we have learned to expect change. We try to plan ahead by talking about how we’ll handle different situations. We try to stay positive and remember we’re prepared and can handle situations. It usually works.”	We’ve learned to expect changes, stay positive.	Acceptance	Emotional Processes
“At first our faith was in doubt. However, with time we have come to terms with having an uncertain future and learned to cherish every minute together.”	With time our faith has allowed us to come to terms with uncertainties	Faith Maintained	Impacts on Spirituality
“At first our faith was in doubt. However, with time we have come to terms with having an uncertain future and learned to cherish every minute together.”	With time, our faith has allowed us to come to terms with uncertainties.	Faith Questionined	Impacts on Spirituality
“So, a lot of just people who don't specifically work closely with ALS know it's in a motor neuron disease, but they don't know the effects of it. And they look at you like you're crazy you can't move your foot.”	Those who don’t work closely with ALS don’t know enough about it.	Additional Education for Health Care Providers	Health Care System Interactions
“It was just frustrating, challenging too, that we had to find the doctor, had to get it going, get his diagnosis and. I don't know. I just felt like they just weren't helping us like they should.”	The diagnosis period is challenging.	Additional Support Needed from Medical Professionals	Health Care System Interactions
“Well, we love (the ALS clinic) because it’s smaller and more personal.”	We love the ALS clinic because it is more personal.	Appreciation of ALS Clinics	Health Care System Interactions

Table 3

Emergent Themes and Thematic Clusters

Thematic Clusters	Emergent Themes
• Support Persons Becoming Closer • Support Persons Pulling away • Challenges with Social Interactions and Support	Dynamic Transformations of Relational Systems
• Responses to Slower Progression • Communication Challenges • General Physical Challenges	Biological Changes and Well-being
• Denial • Anger & Frustration • Depression and other emotional processes • Acceptance	Emotional Processes
• Faith Maintained • Faith Questioned	Impacts on Spirituality
• Additional Education for Health Care Providers • Additional Support Needed from Medical Professionals • Appreciation of ALS Clinics	Health Care System Interactions

CHAPTER 6: AN ARGUMENT FOR INCREASING THE KNOWLEDGE OF ALS IN MEDICAL SCHOOL AND GRADUATE MEDICAL EDUCATION PROGRAMS

(This manuscript was prepared in accordance with the submission reqirements for the *Journal of Graduate Medical Education.)*

Amyotrophic lateral sclerosis (ALS) is the most common progressive and fatal neurodegenerative disease[1]. Currently, 300,000 individuals are living with ALS in the United States[2] with 7,000 newly diagnosed each year[1]. Characterized by motor neuron degeneration, progressive wasting away and weakness of skeletal muscles, ALS leads to eventual paralysis and death within three to five years of symptom onset[1,3]. Even though ALS is becoming increasingly common[4], the preparation of specialists to staff ALS Clinics and healthcare professionals to care for patients and families living with ALS is limited. The following article shines a light on (a) a growing shortage of neurologists and ALS specialists, (b) the need for increased collaboration between ALS specialists and non-ALS specialists, and (c) opportunities for updating and expanding the ALS curriculum offered in medical school and graduate medical education programs.

Shortage of Neurologists and ALS Specialists

While incidence rates of ALS are projected to increase[4], there is a corresponding shortage of medical students going into neurology residencies and who specialize in caring for patients with ALS[5]. Neurophobia, a term coined in 1994, is used to describe medical students' fear of neurology[6.] This fear often begins early in medical school as neurology is thought of as the most difficult subject to master[7]. Neurophobia, along with the negative perception of the complexity of the field of neurology, has led to a growing shortage of neurologists with one recent study finding that only 2.8% of medical students indicated intent to enter a neurology residency[5]. This

shortage of neurology residents only compounds the challenge of increasing the number of ALS specialists to staff ALS clinics.

Multidisciplinary ALS clinics, in which care exclusively caters to patients with ALS, emerged in the mid-1990s[8]. Since their emergence, research on ALS clinics has shown to reduce the risk of death from ALS by 45%, five years following diagnosis[9]. Unfortunately, these multidisciplinary ALS clinics have also been found to have disjointed collaboration and transitions between healthcare providers[10], difficulty sharing a diagnosis of ALS with the patient and family[3], and a lack of support in delivering evidenced-base care[11]. These limitations support the need for not only focusing on increasing the competencies among medical students and neurology residents in caring for ALS patients and families, but also support enhancing the education of all specialists (e.g., primary care) who manage health issues for these patients outside of ALS clinics.

For example, primary care providers (PCPs), play an important role in the care management of patients and families living with ALS[12]. They are more prone to have biological, psychological, social, and spiritual historical knowledge about their patients and families, are often more easily accessible for patient care visits, and are leading the advancement of integrated care in the patient centered medical home[12,13]. Although PCPs prove valuable in the general care management of ALS, when patients seek care for other healthcare needs or due to inability to travel to ALS clinics, they expressed frustration at PCPs lack of knowledge related to ALS care[14]. Capitalizing on the strengths of PCPs and enhancing their knowledge about ALS, would position them to better recognize its initial symptoms, facilitate timelier referrals to neurology for diagnosis, and provide care that encompasses the patient's entire biopsychosocial-spiritual health (BPS-S)[12,14,-17]. Due to the median length of survival from symptom onset being between two

and four years[12], a faster diagnosis would potentially allow for a greater quality of life and enhanced symptom management.

The need for increased attention to caring for ALS patients and families in medical schools, residency programs, and through continuing medical education forums is directly grounded in research with patients and families diagnosed with ALS. One study conducted by Schellenberg et al.[18], found neurology residents' communication skills and levels of empathy when delivering a diagnosis of ALS were less than optimal for patients. Patients reported dissatisfaction with the fast pace of these appointments, frustrations with their inability to ask questions, and/or lack of opportunity to confirm understanding of their diagnosis and its prognosis. Participants in an additional study further reported a desire for increased knowledge on the care management of ALS by medical providers outside of ALS clinics, greater support during and following the diagnosis period, and a greater inclusion of family members in treatment[14].

To ensure that medical providers outside of ALS clinics, have a base knowledge on the general care of patients with ALS, it is suggested that specific education pertaining to ALS is developed. Identifying gaps and implementing curriculum modifications would help expedite ALS diagnoses, improve care, and provide opportunities for greater collaboration. Due to the varying accrediting bodies in medical and residency education, which all have different standards, and in order to ensure that these additions are practical, it is further suggested that the recommended components be added to exisiting coursework or lectures. For example, each of these components could be introduced during a neurology lecture combined with a standardized patient simulation that includes a patient with ALS. These curriculum components may include:

(a) general care management of ALS, (b) interdisciplinary and team-based care models, (c) family-centered care approaches, and (d) BPS-S treatment planning.

General Care Management of ALS

To ensure that medical providers outside of ALS clinics have a base knowledge, it is recommended that basic education on ALS include awareness to: (a) prevalence rates, (b) ALS diagnostic criteria, (c) varied presenting symptoms, (d) patterns of illness progression, (e) recognition of familial and sporadic ALS, (f) known environmental risk factors[12], (g) empathetic delivery of an ALS diagnosis[18], and (h) symptoms associated with the various onset types, limb, bulbar, or respiratory/trunk onset[1]. According to Nold[12], base knowledge of these topics would allow providers outside of ALS specialty clinics to swiftly identify ALS and provide optimal care. It would also open up the opportunity to share care using interdisciplinary models of care.

Interdisciplinary Care

Although multidisciplinary clinics have proven highly effective in providing care for patients with ALS[3], providing care from an interdisciplinary perspective might alleviate the challenges associated with multidisciplinary clinics[19]. In contrast to multidisciplinary clinics, interdisciplinary clinic teams meet regularly to discuss and collaboratively set treatment goals. They also jointly carry out treatment plans and are ideally on the same hierarchical level with a high degree of communication and cooperation among the team members. Including education on care from an interdisciplinary team-based perspective would expand those trained to treat ALS thereby alleviating patients' previous frustrations with providers' limited communication skills[18] within and outside of ALS clinics[14]. With this information in mind, specific educational competencies should minimally include: (a) the value of collaborative care, (b) the creation of joint treatment goals, and (c) the importance of communication between and amongst various

providers. This focused effort to remove barriers in collaborating would help ensure that all team members, patients and family members are coordinating and exchanging information fluidly[19].

Family-Centered Care

Increased education on family-centered ALS care would allow for the mitigation of previous frustrations with providers' inability to provide empathetic support to patients with ALS and their families[18]. Family-centered care includes respect for patients' values and preferences, the integration of care provided by various specialists and medical professionals, information about the disease provided in individualized ways, physical comforts from pain, emotional support, and the inclusion of family and friends[20]. When given the opportunity to share their own lived experiences with ALS, patients and their families (which may include non-related support persons) shared suggestions for improving family-centered ALS care[14]. They desired increased communication between their ALS specialists and non-specialist providers (i.e., PCPs), more attention given to family members and support persons, and increased emotional support during and following the diagnosis period. Williams et al.[14] found patients and families living with ALS wanted their providers to be skilled in: (a) designing individualized treatment plans, (b) initiating and participating in integrated interdisciplinary care, (c) providing emotional support, and (d) incorporating involved family members and friends in understanding the illness and care management needed. Patients and families argued for providers who could consider the BPS-S health of a patient and their caregiver(s) to successfully treat the system as a unit.

BPS-S Care

The majority of research regarding the treatment of ALS seems to primarily focus on biological treatments and clinical trials that seem to delay the physical progression of the disease

[21-23]. Previous research on the BPS-S[15-17] health of patients with ALS and their support persons has been disjointed, focusing on one individual component at a time. Thus far, research has failed to determine how each component impacts one another simultaneously[14]. Increased education on the BPS-S framework would assist providers in better understanding the systemic and relational implications of the disease by viewing health through a comprehensive lens. Specific competencies to be achieved related to the BPS-S framework would include education on: (a) general systems theory[24] as the theoretical construct of the BPS-S framework, (b) Engel's[15,16] development of the biopsychosocial framework, (c) the subsequent addition of the spiritual component[17], and (d) techniques for providing care to patients and families living with ALS.

Conclusion

Since ALS was first described by French neurologist Jean-Martin Charcot 150 years ago, there continues to be no known cause or cure for this progressive neurodegenerative disease. ALS clinics have been integral in advancing the diagnosing process and care of patients with ALS. The essential nature of these clinics incidentally creates pressure for ALS clinics to adequately meet all healthcare needs appropriately suited for primary care, behavioral health, occupational therapy, and even speech therapy. However, there is evidence that merits the expansion of competencies in medical school, graduate medical education, and continuing education programs to include interdisciplinary, family-centered approaches which incorporate knowledge on the general care of ALS from a BPS-S perspective.

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APPENDIX A: IRB APPROVAL

EAST CAROLINA UNIVERSITY
University & Medical Center Institutional Review Board
4N-64 Brody Medical Sciences Building· Mail Stop 682
600 Moye Boulevard · Greenville, NC 27834
Office **252-744-2914** · Fax **252-744-2284** ·
rede.ecu.edu/umcirb/

Notification of Initial Approval: Expedited

From: Social/Behavioral IRB
To: Rachel Williams
CC: Jennifer Hodgson
Date: 11/18/2019
Re: UMCIRB 19-002186
ALS and family functioning

I am pleased to inform you that your Expedited Application was approved. Approval of the study and any consent form(s) occurred on 11/14/2019. The research study is eligible for review under expedited category # 6,7. The Chairperson (or designee) deemed this study no more than minimal risk.

Changes to this approved research may not be initiated without UMCIRB review except when necessary to eliminate an apparent immediate hazard to the participant. All unanticipated problems involving risks to participants and others must be promptly reported to the UMCIRB. The investigator must submit a Final Report application to the UMCIRB prior to the Expected End Date provided in the IRB application. If the study is not completed by this date, an Amendment will need to be submitted to extend the Expected End Date. The Investigator must adhere to all reporting requirements for this study.

Approved consent documents with the IRB approval date stamped on the document should be used to consent participants (consent documents with the IRB approval date stamp are found under the Documents tab in the study workspace).

The approval includes the following items:

Name	Description
Data Collection Sheet	Data Collection Sheet
Dissertation Proposal	Study Protocol or Grant Application
Email Script for Support Persons participating via WebEx	Recruitment Documents/Scripts
HIPAA Authorization	HIPAA Authorization
In Person Recruitment Script	Recruitment Documents/Scripts
Informed Consent Form	Consent Forms
REDCap Demographics Survey	Surveys and Questionnaires
Request for Preparatory Review of PHI	HIPAA Authorization
Semi-Structured Interview Guide	Interview/Focus Group Scripts/Questions
Telephone Identified Support Person Recruitment Script	Recruitment Documents/Scripts
Telephone Recruitment Script	Recruitment Documents/Scripts
Telephone Script for Interview Scheduling - Patient	Recruitment Documents/Scripts
Telephone Script for Interview Scheduling - Support Persons	Recruitment Documents/Scripts

For research studies where a waiver of HIPAA Authorization has been approved, each of the waiver criteria in 45 CFR 164.512(i)(2)(ii) has been met. Additionally, the elements of PHI to be collected as described in items 1 and 2 of the Application for Waiver of Authorization have been determined to be the minimal necessary for the specified research.

The Chairperson (or designee) does not have a potential for conflict of interest on this study.

EAST CAROLINA UNIVERSITY
University & Medical Center Institutional Review Board
4N-64 Brody Medical Sciences Building· Mail Stop 682
600 Moye Boulevard · Greenville, NC 27834
Office **252-744-2914** · Fax **252-744-2284** ·
rede.ecu.edu/umcirb/

Notification of Amendment Approval

From: Social/Behavioral IRB
To: Rachel Williams
CC: Jennifer Hodgson
Date: 12/4/2019
Re: Ame1 UMCIRB 19-002186
UMCIRB 19-002186
ALS and family functioning

Your Amendment has been reviewed and approved using expedited review on 12/3/2019. It was the determination of the UMCIRB Chairperson (or designee) that this revision does not impact the overall risk/benefit ratio of the study and is appropriate for the population and procedures proposed.

Please note that any further changes to this approved research may not be initiated without UMCIRB review except when necessary to eliminate an apparent immediate hazard to the participant. All unanticipated problems involving risks to participants and others must be promptly reported to the UMCIRB. The investigator must submit a Final Report application to the UMCIRB prior to the Expected End Date provided in the IRB application. If the study is not completed by this date, an Amendment will need to be submitted to extend the Expected End Date. The investigator must adhere to all reporting requirements for this study.

Approved consent documents with the IRB approval date stamped on the document should be used to consent participants (consent documents with the IRB approval date stamp are found under the Documents tab in the study workspace).

The approval includes the following items:

Document	Description
Informed Consent Form(0.03)	Consent Forms
Revised Data Collection Sheet (0.01)	Data Collection Sheet

For research studies where a waiver of HIPAA Authorization has been approved, each of the waiver criteria in 45 CFR 164.512(i)(2)(ii) has been met. Additionally, the elements of PHI to be collected as described in items 1 and 2 of the Application for Waiver of Authorization have been determined to be the minimal necessary for the specified research.

The Chairperson (or designee) does not have a potential for conflict of interest on this study.

APPENDIX B: RECRUITMENT, TELEPHONE, AND EMAIL SCRIPTS

In-person Recruitment Script

Hello, Ms./Mr. {insert name]. My name is Rachel Williams and I am a student at East Carolina University. I am working on a study to better understand how ALS impacts families and each member's biological, psychological, social, and spiritual health. This information will help us to improve the quality of care available to patients and families impacted by ALS. I am wondering if it would be alright if I sit down with you and tell you a little more about this study so you can make a decision about whether or not you would like to participate?

[If the participant agrees for the PI to continue with the recruitment process, she will do so. If the patient does not agree, the PI will thank the patient for his or her time and leave the room].

The study has a few different parts to it. If you agree to participate today, I will ask you to sign a form stating that you agree to participate. I will then give you a brief survey to complete using this computer that helps us understand more about each person who agrees to participate. The survey will ask questions about you such as your age, gender, education level, and relationship status. The survey will take about 5 minutes to complete. The final part is an hour-long, audio-recorded interview that will be done at a later date with myself and members of your support system/family of your choosing. Therefore, in order to participate you will need to provide the name and contact information for at least one non-professional support person in your life that might also be willing to participate. We can either contact that person together at this time or the research team can contact them at a later time, so long as you are willing to inform that we will be contating them concerning their participation in this study. In participating in the interviews, we have technology so that you or any members of your family do not even

have to leave your homes to participate. You just need access to a phone or computer with a camera.

Please know that your decision whether or not to participate will not affect your care at the ALS clinic in any way. We want you to feel comfortable saying "no" if you do not wish to participate. Also, if you do decide to participate, know that you may choose to stop participating in the study at any time. It is your choice and you always have the right to change your mind.

Do you have any questions about the information I have shared with you? Do you have any concerns?

[PI will pause for questions and concerns. If participant has questions or concerns, PI will answer and address them to the best of her ability. If patient does not have questions or concerns, PI will continue with recruitment process].

Would you like to participate? If so, we will need you to sign this informed consent document to show your agreement.

[If participant agrees, they will sign the informed consent document a computer will be made available or the REDCap survey link sent to the patient participant via mail. If the patient declines to participate, the PI will thank them for their time and leave the room].

Telephone Indetified Support Person Recruitment Script

Hello, Ms./Mr. {insert name]. I am contacting you because someone close to you with ALS has requested your participation in a study about ALS. The study is designed to help understand the patient and support person's experience of ALS so that we may work to improve the care provided to all involved. I would like to explain the study to you further and hope that you might be available to join the person who recommended you for an interview. My name is Rachel Williams and I can be reached at the following number, 252-737-1415 or by email at williamsra11@students.ecu.edu.

Telephone Recruitment Script

Hello, Ms./Mr. {insert name]. The following information is provided to you as an opportunity to participate in a study that will help us to improve the quality of care to patients and families impacted by ALS. This study will help us to better understand how ALS impacts families and each member's biological, psychological, social, and spiritual health.

The study has a few different parts to it. If you agree to participate today, you will be asked to sign a form stating that you agree to participate. You will complete a brief survey to helps us understand more about each person who agrees to participate. The survey will ask questions about you such as your age, gender, education level, and relationship status. The survey will take about 5 minutes to complete. The final part of the study is an hour-long, audio-recorded interview that will be done at a later date with the lead researcher and members of your support system/family of your choosing. Therefore, in order to participate you will need to provide the name and contact information for at least one non-professional support person in your life that might also be willing to participate. In participating in the interviews, we have technology so that even participates who do not live in the area may also participate.

Please know that your decision whether or not to participate will not affect your care at the ALS clinic in any way. We want you to feel comfortable saying "no" if you do not wish to participate. Also, if you do decide to participate, know that you may choose to stop participating in the study at any time. It is your choice and you always have the right to change your mind.

If you would like to participate we ask that you provide contact information that will be shared with the lead researcher who will only use this information to contact you to schedule an interview. If you have any questions or concerns the lead researcher, Rachel Williams, can be contacted by email, williamsra11@students.ecu.edu or by phone at 252-737-1417.

Telephone Script for Interview Scheduling - Patient

Hello, Ms./Mr. {insert name]. I am contacting you based on the interest in participating in a study about ALS that you shared with our triangulated researcher, who shared your contact information with me. As a reminder, this study is designed to help understand the patient and support person's experience of ALS so that we may work to improve the care provided to all involved. The next step in the study is to schedule an interview with yourself and any members of your non-professional support system to be held at the Vidant ALS Clinic. We can schedule the interview now or you can contact me at a later date to do so. My name is Rachel Williams and I can be reached at the following number, 252-737-1415 or by email at williamsra11@students.ecu.edu.

Telephone Script for Interview Scheduling – Support Persons

Hello, Ms./Mr. {insert name]. I am contacting you based on the interest you expressed about participating in a study about ALS, as a support person to someone who has ALS. As a reminder, this study is designed to help understand the patient and support person's experience of ALS so that we may work to improve the care provided to all involved. The next step in the study is to schedule an interview with yourself, the patient with ALS, and any additional members of the patient's non-professional support system to be held at the Vidant ALS Clinic.. We can schedule the interview now or you can contact me at a later date to do so. My name is Rachel Williams and I can be reached at the following number, 252-737-1415 or by email at williamsra11@students.ecu.edu.

APPENDIX C: INFORMED CONSENT & HIPAA AUTHORIZATION

Informed Consent to Participate in Research

Information to consider before taking part in research that has no more than minimal risk.

Title of Research Study: How ALS Impacts Family Functioning and Health

Principal Investigator: Rachel E. Williams, MS, LMFTA
Institution, Department or Division: Human Development and Family Science
Address: 114 Redditt House, East Carolina University, Greenville, NC 27858
Telephone #: (252) 737-1415

Participant Full Name: _____Date of Birth: _____
Please PRINT clearly

Researchers at East Carolina University (ECU) Vidant Medical Center and Vidant Medical Groups study issues related to society, health problems, environmental problems, behavior problems and the human condition. To do this, we need the help of volunteers who are willing to take part in research.

Why am I being invited to take part in this research?

You are being invited to take part in this research because you area or you are a member of a patient's support system. The decision to take part in this research is yours to make. We hope that the information that you provide will help us learn how to better support and assist families living with ALS.

If you volunteer to take part in this research, you will be one of about 40 people to do so.

Are there reasons I should not take part in this research?

I understand I should not volunteer for this study if I am: (a) a non-English speaker, (b) under 18 years of age, and/or (c) have screened positive for Frontotemporal Dementia or any other forms of dementia by the clinic staff at the Vidant ALS Clinic.

What other choices do I have if I do not take part in this research?

Participation in this research project is voluntary. You may choose not to participate at any time with no impact on the treatment you are receiving at the Vidant ALS Clinic.

Where is the research going to take place and how long will it last?

The research will occur at the following times and places: Vidant Medical Center and over the phone. Your contact information will be collected by the triangulated researcher when you express interest in participating in this study. Your contact information will be shared with the lead researcher who will use it to schedule an interview with you and your participating support person. For the interview, you will need to come to the Vidant ALS Clinic one time during the study. Prior to the start of the interview, you will be asked to fill out a brief survey, which should take 5-10 minutes, at the Vidant Medical Center. You may complete it using your smartphone or a computer made available to you. During the interview we will ask you if you would like to participate in the verifying of the results. This portion of the study is

optional, can be completed at home, and would take approximately 30 minutes. The total amount of time you will be asked to volunteer is between 30 minutes and two hours depending on the length of time it takes to complete the interview and on your participation in reviewing the results for accuracy.

What will I be asked to do?

You will be asked to do the following:

- Complete the University & Medical Center Institutional Review Board (UMCIRB) HIPAA Privacy Authorization Form, which provides your authorization to use and disclose your protected health information for research. This form will be provided to you prior to the start of the interview.
- Complete a brief online survey that asks about your demographic information (age, gender)
- As the patient, you will provide the name and contact information of at least one member of your support team to participate in an interview with you.
- Participate in a one-time, audio-recorded, interview. During the interview you will be asked to share your experiences of living with ALS. The audio-recorded interviews will be typed up for analysis and will be kept in a password-protected file. Audio files will be password-protected and stored on a secure server at East Carolina University.
- At the end of the interview you will be asked if you would like to be involved in the process of double-checking the results ("member checking") which will take place in two portions. The first portion will include checking your typed-up interview for accuracy and the second portion will include checking the results to see how well they match your experience. If you are interested in participating in either of these portions of member checking, we will provide you with the transcripts and/or results by mail or e-mail, whichever is your preference. Participating in checking the typed-up responses from the interview will give you an opportunity to share any additional information you may not have thought of during the interview. Participating in checking the results will give you the opportunity to make sure those results are accurate or help us make corrections. Audio files will be erased after they are typed, and the member checking process is completed. The typed interviews will be stored electronically on a secure server at East Carolina University, which is password-protected. The typed interview documents will be kept in a locked filing cabinet in a locked office. Documents will be shredded and electronic files containing identifying information will be erased seven years after the project is completed.

What might I experience if I take part in the research?

There are possible risks (the chance of harm) when taking part in this research. This research will require your time and energy to complete. Some participants who have completed surveys and interviews on topics of mood, social support, and spirituality may experience discomfort with answering questions. Some participants may also experience negative feelings (such as anger, fear, grief, or sadness) from being asked to recall and talk about a difficult time (such as your experiences with ALS). Please know you may always ask to "skip" questions you are not comfortable with answering. You can also ask to stop a survey or interview at any time.

In the case that you experience negative emotions or symptoms of anxiety or depression throughout the study, we will help connect you with behavioral health resources that you may choose to use.

What are the possible benefits I may experience from taking part in this research?

We do not know if you will get any benefits by taking part in this study. Hopefully, this research will help us learn more about how to best serve patients with ALS and their support systems. There may be no personal benefit from your participation, but the information gained by doing this research may help others in the future. However, other people who have participated in this type of research have

experienced benefit from being able to talk about their experience with researchers. By participating in this research study, you may also experience these benefits.

Will I be paid for taking part in this research?
We will not be able pay you for the time you volunteer while being in this study.

Will it cost me to take part in this research?
It will not cost you any money to be part of the research. This research study is not for profit and is being completed as part of the Principal Investigator's degree requirements. All researchers are donating their time to the study.

Who will know that I took part in this research and learn personal information about me?
ECU and the people and organizations listed below may know that you took part in this research and may see information about you that is normally kept private. With your permission, these people may use your private information to do this research:

- The University & Medical Center Institutional Review Board (UMCIRB) and its staff have responsibility for overseeing your welfare during this research and may need to see research records that identify you.
- Any agency of the federal, state, or local government that regulates human research. This includes the Department of Health and Human Services (DHHS), the North Carolina Department of Health, and the Office for Human Research Protections.
- People designated by Vidant Medical Center and Vidant Health.
- If you are a patient at ECU or Vidant, a copy of the first page of this form will be placed in your medical records.

How will you keep the information you collect about me secure? How long will you keep it?
Your name will only be attached to this informed consent document and a list kept by the Principal Investigator of the study participants. A list of study participants with contact information and your pseudo name used for the interview will be listed in a password-protected file on a secure server at East Carolina University. The audio file of the interview will be password-protected and stored on a secure server at East Carolina University. Hardcopies of your typed interview will be stored in a locked file cabinet. Your name will not be attached to these materials. Audio files will be erased after the interviews are typed and the member checking process is completed. After this study is complete, documents will be shredded and electronic files containing identifying information will be erased seven years after the project is completed.

What if I decide I don't want to continue in this research?
You can stop at any time after it has already started. There will be no consequences if you stop and you will not be criticized. You will not lose any benefits that you normally receive.

Who should I contact if I have questions?
The people conducting this study will be able to answer any questions concerning this research, now or in the future. You may contact the Principal Investigator, Rachel Williams, at (252) 737-1415 (Fridays, 9AM-5PM) or the Principal Investigator's Supervisor, Jennifer Hodgson, PhD, at (252) 328-1349 (Monday-Friday, 9AM-5PM).

If you have questions about your rights as someone taking part in research, you may call the Office of Research Integrity & Compliance (ORIC) at phone number 252-744-2914 (days, 8:00 am-5:00 pm). If you would like to report a complaint or concern about this research study, you may call the Director for

Human Research Protections, at 252-744-2914 and the Vidant Medical Center Risk Management Office at 252-847-4473.

Is there anything else I should know?

The following research results will be provided to you: The results from the interviews will be shared with you as part of the member checking process. This will allow you to verify if that the results accurately reflect your experiences with ALS.

I have decided I want to take part in this research. What should I do now?

The person obtaining informed consent will ask you to read the following and if you agree, you should sign this form:

- I have read (or had read to me) all of the above information.
- I have had an opportunity to ask questions about things in this research I did not understand and have received satisfactory answers.
- I know that I can stop taking part in this study at any time.
- By signing this informed consent form, I am not giving up any of my rights.
- I have been given a copy of this consent document, and it is mine to keep.

_____ _____

Participant's Name (PRINT) **Signature** **Date**

☐ The participant is unable to sign and date due to the physical progression of ALS, however the participant has given consent through verbal or nonverbal communication.

Person Obtaining Informed Consent: I have conducted the initial informed consent process. I have orally reviewed the contents of the consent document with the person who has consented above and answered all of the person's questions about the research.

_____

Person Obtaining Consent (PRINT) **Signature** **Date**

_____

Principal Investigator (PRINT) **Signature** **Date**
(If other than person obtaining informed consent)

UMCIRB HIPAA Privacy Authorization

East Carolina University (ECU)/Vidant Medical Center (VMC): Research Participant Authorization to Use and Disclose Protected Health Information for Research

For use only with the research consent form for UMCIRB#: 19-002186
Principal Investigator: Rachel Williams
Title: How Amyotrophic Lateral Sclerosis Impacts Family Functioning and Health

Location where research will be conducted
The members of the research team will conduct the research study at:
☐ East Carolina University (ECU) ☒ VMC ☐ ECU & VMC ☐ Other

When taking part in research, protected health information (PHI) is collected, used, and shared with others who are involved in the research. Federal laws require that researchers and health care providers protect your PHI. Also, federal laws require that we get your permission to use collected PHI for the research. This permission is called authorization.

In order to complete the research project in which you have decided to take part, the research team needs to collect and use some of your PHI as described below.

What types of protected health information (PHI) about me will be used or disclosed?
(Select all that apply.)

ECU Health Care Component:
[☐] ECU Physicians
[☐] School of Dental Medicine
[☐] Speech, Language, and Hearing Clinic
[☐] Human Performance Lab
[☐] Physical Therapy
[☐] Student Health
[☐] Other ECU Health Entity
(please list):

Vidant Health Entity:
[☐] Entire Vidant Health system
[☒] Vidant Medical Center
[☐] Other Vidant Health Entity
(please list):

Type of ECU Records:
[☐] Medical/clinic records
[☐] Billing records
[☐] Lab, Pathology and/or Radiology results results
[☐] Mental Health records
[☐] PHI previously collected for research
[☒] Records generated during this study
[☐] Other:

Type of Vidant Records:
[☐] Medical/clinic records
[☐] Billing records
[☐] Lab, Pathology and/or Radiology

[☐] Mental Health records
[☐] PHI previously collected for research
[☐] Records generated during this study
[☒] Other: Name, Telephone Number, and Email

Who will use or disclose my PHI?
[☒]Principal Investigator
[☒]Other members of the research team
[☐]Other providers involved in your care during research procedures, outpatient/inpatient stays during which research is being performed, or physician office visits during which research is being performed.

Who will receive my PHI?
[☐] Sponsor or other funding source to provide oversight for entire research project
[☒] Research investigators to conduct and oversee the research project
[☒] Principle Investigator and research team members to participate in the various research activities
[☐] FDA or other regulatory agencies to provide regulatory oversight
[☒] UMCIRB to provide continuing review of the research project
[☒] Institutional officials in connection with duties for monitoring research activity
[☐] Other providers involved in your care during research procedures, outpatient/inpatient stays during which research is being performed, or physician office visits during which research is being performed.
[☐] Researchers at other sites—List sites:
[☐] Data and Safety Monitoring Board and its staff
[☐] Contract Research Organization and its staff
[☐] Other

We will share only the PHI listed above with the individuals/agencies listed above. If we need to share other PHI or if we need to send PHI to other individuals/agencies not listed above, we will ask for your permission in writing again

How my PHI may be released to others:
ECU and VMC are required under law to protect your PHI. However, those individuals or agencies who receive your PHI may not be required by the Federal privacy laws to protect it and may share your PHI with others without your permission, if permitted by the laws governing them.

What if I do not sign this form?
You will not be eligible to participate in this study if you do not sign this Authorization form.

How may I revoke (take back) my authorization?
You have the right to stop sharing your PHI. To revoke (or take back) your authorization, you must give the Principal Investigator your request to revoke (or take back) your authorization in writing. If you request that we stop collecting your PHI for the study, you may be removed from the study. If you are removed from the study, it will not affect your ability to receive standard medical care or affect payment, health plan enrollment or benefit eligibility. PHI collected for the research study prior to revoking (or taking back) your Authorization will continue to be used for the purposes of the research
study. Also, the FDA (if involved with your study) can look at your PHI related to the study even if you withdraw this authorization.

Restrictions on access to my PHI:
You will not be able to see your PHI in your medical record related to this study until the study is complete. If it is necessary for your care, your PHI will be provided to you or your physician.
How long may the PHI about me be used or disclosed for this study?
Research information continues to be looked at after the study is finished so it is difficult to say when use of your PHI will stop. There is not an expiration date for this authorization to use and disclose your PHI for this study.

If you have questions about the sharing of PHI related to this research study, call the principal investigator, Rachel Williams**,** at 252-737-1415. Also, you may telephone the University and Medical Center Institutional Review Board at 252-744-2914. In addition, if you have concerns about confidentiality and privacy rights, you may phone the Privacy Officer at Vidant Medical Center at 252-847-3310 or the Privacy Officer at East Carolina University at 252-744-5200.

Authorization

To authorize the use and disclosure of your PHI for this study in the way that has been described in this form, please sign below and date when you signed this form. A signed copy of this Authorization will be given to you for your records.

Name of Participant or Authorized Representative (print) Signature Date

---

If an Authorized Representative has signed on behalf of a Participant please print on the line above the authority of the Legal Representative to do so (*such as parent, court-appointed guardian, or power of attorney*).

Person Obtaining Authorization Signature Date

APPENDIX D: REDCap SURVEY

Demographics

For Patients and Support Persons:

1. How old are you?

   _____

2. To which gender identity do you most identify?
   A. Female
   B. Male
   C. Other (e.g., transgender, gender neutral, etc. Please specify: _____)
   D. Prefer Not to Answer

3. What is your race? (Select all that apply)
   A. American Indian or Alaskan Native
   B. Asian
   C. Black or African American
   D. Hawaiian or Other Pacific Islander
   E. White
   F. Other (Please specify:_____)
   G. Prefer not to answer

4. What is your ethnicity?
   A. Hispanic or Latino/Spanish Origin
   B. Not Hispanic or Latino/Spanish Origin
   C. Prefer not to answer

5. What is the highest level or school you have completed your highest year of schooling completed?
   A. Less than high school
   B. High School/GED
   C. Some College, but no degree
   D. Associate degree in college (2-year)
   E. Bachelor's degree in college (4-year)
   F. Master's degree
   G. Doctoral degree
   H. Professional degree (JD, MD)

6. Which statement best describes your current employement status?
   A. Working
      a. paid employee
      b. self-employed
   B. Not working
      a. temporary layoff from a job

b. looking for work
c. retired
d. disabled
e. other
f. student

7. What was your employment status at time of symptom onset?
A. Working
a. paid employee
b. self-employed
B. Not working
a. temporary layoff from a job
b. looking for work
c. retired
d. disabled
e. other
f. student

8. What is your current yearly household income?
A. Less than $10,000
B. $10,000 to $19,999
C. $20,000 to $29,999
D. $30,000 to $39,999
E. $40,000 to $49,999
F. $50,000 to $59,000
G. $60,000 to $69,999
H. $70,000 to $79,999
I. $80,000 to $89,999
J. $90,000 to $99,999
K. $100,000 or more

9. What is your current health insurance status?
A. Uninsured, Self-pay
B. Insured by Medicaid or Medicare
C. Insured by private insurance (i.e., Blue Cross & Blue Shield of NC, Cigna)
D. Insured by Tricare

10. What is your current relationship status?
A. Single, never married
B. In a committed relationship
C. Married or civil union
D. Widowed
E. Divorced
F. Legally Separated
G. Other (Please Specify: _____)

11. If you are currently in a relationship, how many years have you been in that relationship?

A. 0 to 3 years
B. 3 to 5 years
C. 5 to 10 years
D. 10 to 15 years
E. 15 to 20 years
F. 20 or more years
G. Not applicable

12. Do you have any children/stepchildren?

A. Yes (how many: _____?)
B. No

13. How many of your children/stepchildren are living? _____ deceased?_____

14. If you have children/stepchildren, how many of those children currently live with you?

A. 0-2
B. 3-5
C. 5 or more

15. If you have children, how many of those children do not currently live with you?

A. 0-2
B. 3-5
C. 5 or more

16. What is your religious affiliation, if any?

A. Agnostisicm
B. Atheism
C. Buddhism
D. Christianity
E. Hinduism
F. Judaism
G. Islam
H. Other (Please specify: _____)
I. Prefer not to answer

17. Do you have a history of mental illness?

A. Yes (Please describe:_____)
B. No

18. Did you experience physical limitations of impairments prior to the onset of ALS?

A. Yes (Please describe:_____)
B. No

For Support Persons Only:

19. What is your relationship to the patient? For example: friend, parent, partner, child, etc.

_____

20. Do you and the patient reside together?

A. Yes
B. No

For Patients Only:

21. When did you first notice symptoms of ALS? _____ (approximate date)

22. What date were you diagnosed with ALS? _____

23. What as your onset type:

C. Bulbar
D. Limb
E. Other (please specify_____)
F. Unknown

24. What is your relationship to the primary support person? For example: friend, parent, partner, child, etc.

_____

25. Do you and the primary support person reside together?

G. Yes
H. No

APPENDIX E: INTERVIEW GUIDE

Introduction

Thank you for meeting with me today. I am a doctoral student in medical family therapy at East Carolina University and would like to better understand the experiences of families living with ALS. This project is part of my requirements for my degree but this topic is very personal and important to me as well. I am meeting with you because I am particularly interested in what it is like for patient and members of their support system to live with ALS and how it impacts the way you all function together.

There are no right, or wrong answers and you may share anything related to your experience with ALS as it comes to mind. What you share with me today will only be used for this study. I will not use anyone's actual name or any information that may be used to identify you when I prepare the results for publication and presentation. The consent form you signed when we first met is your written agreement for the interview today.

You may decide not to continue with the interview at any time and you may choose to skip a question if you do not feel comfortable answering it. If you wish to skip a question or stop entirely, please let me know and we will move to the next question or stop the interview completely. I will be recording the interview so that I can type up what was discussed and study the responses to the questions later. I do ask that you try not to use anyone's real name as much as possible but rather refer to that person by your relationship with or to them. For example, my sister, doctor, friend, dad, etc.

Before we begin, do any of you have a fake name that you would like for me to use when I type up the interview? If not, I can assign you one later.

This interview will take about an hour and will be audio recorded. Do you have any questions before we begin?

Grand Tour Question

How do you believe ALS has impacted you and your support system?

Probing Questions

- What has having ALS meant to each of you?
- Why have you chosen to participate in this study?
- What are some challenges each of you have faced?
    - How has ALS impacted you physically (as the support person)?
    - How has ALS impacted you emotionally?
    - How has ALS impacted you socially?
    - How has ALS impacted you spiritually?
- Describe in what ways, if any, the ALS symptom onset type (i.e., bulbar or limb) has impacted you and your support system?
- What are some changes your family has encountered?
    - Have you become closer?
        - In what ways?
    - Have you drifted apart?
        - In what ways?
- Can you describe a time when your or your support system felt most challenged during the course of your illness?
- What positive things have you all experienced as a result of ALS?

- What services (or assistance) have you needed but have not received to help you and your support system manage ALS?
    - What could health professionals like doctors and nurses have offered after diagnosis and throughout your journey with ALS?
- What else would you like to share about your experience with ALS?

APPENDIX F: COMMUNITY BASED RESOURCES

Participation in the interview may evoke feelings of anxiety (feeling worried or nervous), depression (feeling down, sad, or hopeless), or emotional distress (feeling angry, fearful, frustrated, or greif-stricken). During the interview today, some of your answers let me know that you may be experiencing some of these feelings. I encourage you to do the following to make sure you get the help you need:

1. Let your trusted family members and friends know that you are experiencing feelings of anxiety, depression, or emotional distress.
2. Contact a behavioral health provider with whom you can meet and discuss your feelings.

Here are some local resources:

A. The Jim "Catfish" Hunter ALS Clinic, 2310 Stantonsburg Road in Greenville, NC. The clinic is in the Outpatient Rehabilitation Building adjacent to Vidant Medical Center. You may contact Natalie Cox at **252-847-1779** to schedule an appointment.

B. The Greenville area ALS support group meets the 3rd Wednesday of every month from 6:30-8:30 PM at the Vidant Rehabilitation Center Classroom, 2100 Stantonsburg Rd, Greenville, NC. The contact person is Katie Leegins-Vinson, LCSW, kleegins-vinson@alsnc.org

C. ECU Family Therapy Clinic, 612 East 10th St., Greenville, NC (252) 737-1415. Provides therapy to individuals, couples, and families on a sliding-fee scale.

D. Trillium Health Resources, 201 W. 1st, Greenville, NC, (866) 998-2597. A local governmental agency that manages mental health, substance use, and intellectual/developmental disability services in eastern North Carolina.

If you ever feel you are in crisis and need help immediately, here are some important contacts:

A. REAL Crisis Intervention (252) 758-4357. 24/7 crisis line. Provides over-the-phone counseling and referrals.

B. Call 911 or go to your nearest emergency room.

APPENDIX G: STATEMENTS OF BIAS

Rachel Williams

I, the lead researcher, have attempted to recognize and set aside any prior beliefs, biases, and preconceived notions related to individuals and families impacted by ALS. Specifically, I have realized that a motivating factor in conducting research on these topics is my own lived experience of my uncle's diagnosis of and subsequent death due to ALS. I have recognized that the experiences I and my family have lived through may share some similarities with others impacted by the disease, but that ultimately, my own experience remains just that, my own.

In recent years, I have also become more involved with local area ALS support groups. These experiences have allowed me to grow in my understanding of how different experiences with this disease vary from person to person and family to family. These experiences have also provided several opportunities for to immerse myself within the ALS culture, so to speak, to observe and interact with people who are currently living through these experiences every day, and to ultimately set aside my biases and beliefs in real time. Ultimately, the death of my uncle to ALS, and the resulting distressing experiences my family lived through ingnited the spark that has resulted in the current study on ALS. However, more recent real life experiences and educational opportunities have allowed me to approach this topic from a position of inquisitiveness, without the need to overshadow other's experiences and meaning making of those experiences with my own.

In allowing the phenomenon to speak of itself (Colaizzi, 1978), I recognize my positions as a researcher and as a white, college-educated female and how these positions are pertinent to the conceptualization of this study. Furthermore, I acknowledge my own experiences with ALS and am committed to maintaining an awareness of those experiences throughout the study in

order to manage the undue influences the biases resulting from these experiences may have on the study in general. In asking questions to gain a greater understanding and including a methodological component that explores participants' experiences as they understand them (phenomenology), I hope to promote the voices of PALS and their support persons.

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Olivia Riser

As the triangulated researcher, I recognize my position as a British, mixed race, college educated female living in North Carolina and how it may have affected perspective on ALS and subsequent disparities in several ways. Firstly, I realized that although my secular upbringing was common place in Great Britain, this was not necessarily the case in North Carolina and the surrounding southern states. A degree in Anthropology facilitated the my understanding of the significance and function of religion; from its place within a global society to its place within local interactions, such as individuals and their health.

Secondly, having been brought up in Great Britain, I faced a drastically different racial climate compared to my current place of residence. Since living in North Carolina, I have become cognizant of the many disparities and inequalities people of color are subject to, especially in health care settings.

APPENDIX H: REFLEXIVE JOURNAL SELECTED ENTRIES

1/3/2020

"After I got my transcript finished from the first interview, I've read over it and found that I need to go deeper with my questions. I'm finding that I ask the initial question, and receive an answer, but don't tend to ask them to explain more or provide examples. I'm wondering if I'm subconsciously keeping myself from asking the more difficult questions? Am I a bit fearful of the answers? Am I a bit fearful of how "real" and close to home the answers might be? Am I a bit fearful of how it might be a little difficult for me to hear how the families are struggling? I'm not sure. I'm just thankful of my awareness of this and will make a concerted effort to dig deeper in future interviews.

2/7/20

"This was my longest interview so far and I've realized that for the past few interviews I've really been able to go through the entire interview guide and then explore other areas as needed, without fear of what the answers will be. I was really touched by the fact that this patient hadn't had a relationship with her children for several years, but that bc of her illness they have rekindled their relationships and their love."

3/4/20

"I've been reading through several of the transcripts just now. I've read through at least four of them and then have gone back to re-read them and pull out the significant statements. I've had a bit of a difficult time doing this. Some of the pieces of the interviews are really heavy, or at least they feel heavy to me. At the moment, I'm taking a step back and will make sure that I only work

through these one at a time. I want to continue to make sure that whenever I'm working through this analysis that I'm able to honor the participant's experiences and not let my own experience get in the way."

3/31/20

Olivia and I have worked together through all of the steps of the analysis. She has previously shared with me that she does not have any prior experiences with anyone with ALS, which has helped me to bracket any biases that may have come up by chatting through things with her. I felt really good about our thematic saturation when she shared with me her experiences with analyzing the data from another study. She shared that she felt like all of our data fit very well into our themes.

APPENDIX I: QUALITY ASSESSMENT FOR NON-RANDOMIZED ARTICLES IN SYSTEMATIC REVIEW

Author Year	Selection Bias	Study Design	Confounders	Blinding	Data Collection Method	Withdrawals and Dropouts	Global Rating
Adelman et al., 2004	2	3	3	3	1	3	*
Alankaya et al., 2015	2	3	3	3	1	N/A	*
Atkins et al., 2009	2	3	3	3	1	3	*
Bolmsjö et al., 2003	2	3	3	3	3	N/A	*
Bruletti et al., 2014	2	3	3	3	1	1	*
Burke et al., 2017	3	3	3	3	1	3	*
Calvo et al., 2011	2	3	3	3	1	N/A	*
Calvo et al., 2015	3	3	3	3	1	N/A	*
Chen et al., 2015	3	3	3	3	1	N/A	*
Chiò et al., 2005	3	3	3	3	1	N/A	*
Chiò et al., 2010	3	3	3	3	1	N/A	*
Creemers et al., 2016	1	1	3	3	1	2	*
Galvin et al., 2016	3	3	3	3	3	N/A	*
Garcia et al., 2017	2	3	3	3	1	1	*
Gauthier et al., 2006	3	3	3	3	1	3	*
Gelinas et al., 1998	3	3	3	3	1	N/A	*
Goldstein et al., 1998	3	3	3	3	1	N/A	*
Goldstein et al., 2006	2	3	3	3	1	3	*
Hecht et al., 2003	2	3	3	3	1	N/A	*
Hwang et al., 2014	3	2	3	3	1	N/A	*
Lillo et al., 2012	3	3	3	3	1	N/A	*
Lo Coco et al., 2005	3	3	1	3	1	N/A	*
Martin et al., 2002	2	3	3	3	3	N/A	*
Miyashita et al., 2009	2	3	3	3	3	N/A	*
Miyashita et al., 2011	3	3	3	3	1	N/A	*
Mock et al., 2010	2	3	3	3	3	N/A	*
Murphy et al., 2008	2	3	3	3	1	N/A	*
Olsson et al., 2010a	2	3	3	3	1	N/A	*

Olsson et al., 2010b	2	3	3	3	1	2	*
Olsson Ozanne et al., 2010	2	3	3	3	1	N/A	*
Oyebode et al., 2013	3	3	3	3	3	N/A	*
Ozanne et al., 2014	2	3	3	3	2	N/A	*
Pagnini et al., 2010	2	3	3	3	3	N/A	*
Pagnini et al., 2011	2	3	3	3	1	N/A	*
Pagnini et al., 2012a	2	3	3	3	1	N/A	*
Pagnini et al., 2012b	3	3	3	3	1	N/A	*
Qutub et al., 2014	2	3	3	3	1	N/A	*
Rabkin et al., 2000	2	3	3	3	1	3	*
Rabkin et al., 2009	3	3	3	3	1	1	*
Roach et al., 2009	3	3	3	3	1	3	*
Sciliano et al., 2017	2	3	3	3	1	N/A	*
Trail et al., 2003	3	3	3	3	1	N/A	*
Trail et al., 2004	3	3	3	3	3	N/A	*
Tramonti et al., 2014a	2	3	3	3	1	1	*
Tramonti et al., 2014b	2	3	1	3	1	N/A	*
Tremolizzo et al., 2016	2	3	3	3	1	N/A	*
Vignola et al., 2008	3	3	3	3	1	2	*
Watermeyer et al., 2015	2	3	3	3	1	N/A	*
Note. *** = Strong Rating; ** = Moderate Rating; * = Weak Rating							