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Amyotrophic Lateral Sclerosis & Frontotemporal Impairment: A Qualitative Study of the Caregiving Experience

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Philadelphia College of Osteopathic Medicine

Department of Psychology

AMYOTROPHIC LATERAL SCLEROSIS & FRONTOTEMPORAL IMPAIRMENT:
A QUALITATIVE STUDY OF THE CAREGIVING EXPERIENCE

By Michelle L. Dube

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Dissertation Approval

This is to certify that the thesis presented to us by Michelle L. Dube on the 30th day of May, 2017, in partial fulfillment of the requirements for the degree of Doctor of Psychology, has been examined and is acceptable in both scholarship and literary quality.

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Abstract

Living with amyotrophic lateral sclerosis (ALS) is a complex and difficult experience that necessitates numerous lifestyle changes for the patient and caregiver. Although ALS was previously regarded as a disease that primarily affects the motor system and spares non-motor functions, it is now recognized that a sizable proportion of the ALS population demonstrates frontotemporal impairment (ALS+FTI), with a subset of the population meeting criteria for frontotemporal dementia. Clinically, ALS and FTI differ in many respects; however, both diseases are progressive and necessitate dependence on caregivers for assistance with activities of daily living and beyond. Although the relationship between ALS and FTI has received empirical attention, the experience of the caregiver remains largely unexplored. The aim of this qualitative study was to explore and describe caregivers' thoughts, feelings, behaviors, practical daily challenges, and rewards in caring for persons with ALS+FTI. Four female spousal caregivers participated in semi-structured interviews; they also completed self-report measures to assess quality of life, level of care, and FTI symptoms exhibited by the care recipients. Findings indicated that although each caregiver faced her own set of unique challenges, there were commonalities across narratives. Three major themes were identified: witnessing the effects of ALS+FTI, the impact of the caregiving experience, and coping with life as a caregiver. Findings were interpreted within the context of the existing caregiving literature and suggestions for improving FTI assessment within the ALS population were provided.

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Chapter 1: Introduction

Statement of the Problem

Amyotrophic lateral sclerosis (ALS) is a progressive neurodegenerative disease that is characterized by the degeneration of upper and lower motor neurons in the brain and spinal cord (The ALS Association, 2007). As the disease progresses, there is an irreversible loss of muscular functioning that limits or prevents the ability to move, speak, swallow, and, eventually, breathe (The ALS Association, 2007). Traditionally, it was believed that non-motor functions were spared by this disease (Pagnini et al., 2010); however, evidence suggests a sizable proportion of the ALS population experiences some degree of frontotemporal cognitive and/or behavioral impairment (Gibbons, Richardson, Neary, & Snowden, 2008; Grossman, Woolley-Levine, Bradley, & R. G. Miller, 2007; J. M. Murphy et al., 2016; Raaphorst, De Visser, Linssen, De Haan, & Schmand, 2010), with up to 14% of patients meeting criteria for frontotemporal dementia (FTD) according to Neary criteria (Neary et al., 1998; Phukan et al., 2012). Until recently, the association between ALS and frontotemporal impairment (FTI) has been largely unexplored.

Frontotemporal impairments are caused by the progressive degeneration of the brain's frontal and temporal lobes (Neary, Snowden, & Mann, 2005). They manifest as a spectrum of clinical deficits, including cognitive impairment, behavioral dysfunction, and/or FTD (Strong et al., 2009). FTD refers to a group of clinical syndromes that are characterized by profound changes in personality, cognition, executive functioning, language, and/or social conduct, while the memory is generally left intact (Merrilees, Klapper, J. Murphy, Lomen-Hoerth, & B. L. Miller, 2010; Phukan, Pender, & Hardiman, 2007). A majority of ALS patients who have frontotemporal deficits will not meet full

criteria for FTD according to Neary criteria (Neary et al., 1998). Nonetheless, they may present with frontotemporal deficits that are clinically significant, but not severe enough to warrant an FTD diagnosis (Strong et al., 2009; J. M. Murphy et al., 2007). Although these syndromes vary in severity and presentation, the presence of FTI within the ALS population (ALS+FTI) is associated with a variety of unique features and challenges that distinguish it from classic ALS.

Studies have indicated that patients with ALS+FTI have a shorter survival than those with classic ALS and are more likely to be noncompliant with recommended treatments, such as assisted ventilation (Hu et al., 2009; Olney et al., 2005; Strong et al., 2009). Evidence also suggests that ALS+FTI patients demonstrate a profound lack of insight and are significantly more apathetic in comparison to patients with classic ALS (Chio et al., 2010; Lomen-Hoerth et al., 2003; Woolley, Moore, & Katz, 2010). Moreover, there is a greater instance of impulsive and reckless behaviors, aggression, rigidity, and irritability within the ALS+FTI population (Merrilees et al., 2010). As a result, patients with ALS+FTI require more supervision than those with classic ALS (Lomen-Hoerth et al., 2003). Thus, the impact of FTI on the course of ALS has implications for both the patient and caregiver.

There is a considerable amount of research documenting the experiences of caregivers of persons with ALS and persons with FTI, but little work has been done to investigate the experiences of those who provide care to patients with ALS+FTI. Despite this gap in the literature, the existing research on ALS and FTI caregivers offers some insight into the potential challenges faced by caregivers of the combined ALS+FTI population. For example, caregivers of patients who were demented for shorter periods

of time had lower qualities of life than those providing care for longer periods of time (Nunnemann, Kurz, Leucht, & Diehl-Schmid, 2012). This finding is relevant to the ALS+FTI caregiving population because patients with ALS+FTI experience faster disease progression than classic ALS. In addition, the ALS and the FTI caregiving literature is replete with studies documenting the negative effects of caring for these populations, such as high levels of burden (Mioshi et al., 2013; Pagnini et al., 2010), depressive symptoms (Gauthier et al., 2007; Wong et al., 2012), strain and distress (Rabkin, Wagner, & Del Bene, 2000; Wong et al., 2012), and decreased quality of life (QOL; Roach, Averill, Segerstrom, & Kasarskis, 2009). The consequences of caregiving are evident in each population separately; therefore, caregiving for a loved one with concurrent ALS and FTI represents a significant challenge.

Although there are similarities in the experiences of ALS caregivers and FTI caregivers, there is also evidence to suggest differences between these caregiving populations. For instance, Chio and colleagues (2010) noted a difference between ALS and dementia caregivers, in that the psychological health of ALS caregivers was more affected by patients' levels of apathy rather than disinhibition, whereas the opposite was true for dementia caregivers. Other studies have reported higher levels of burden in dementia caregivers compared to ALS caregivers (Chio, Gauthier, Calvo, Ghiglione, & Mutani, 2005; Hecht et al., 2003). Taken together, these findings indicate that the experiences of ALS+FTI caregivers cannot be inferred from the existing literature and, therefore, are essentially unknown.

Although the current caregiving literature is informative, it is recognized that caregiving experiences are not universal (Clipp & George, 1993). Certainly, some

experiences and stressors may be shared throughout the entire caregiving population; however, each disease imposes its own unique challenges and demands upon caregivers that cannot be generalized across illnesses (Clipp & George, 1993). In recognition of this finding, an increasing amount of literature has become available on disease-specific caregiving experiences, especially within the context of caring for persons with medical conditions (e.g., cancer) and persons with psychiatric conditions (e.g., schizophrenia). It is without question that this shift in focus has increased the public's understanding of the heterogeneity of caregiving experiences; however, the literature seems to address medical conditions separately from psychiatric conditions and seldom addresses the experiences of those caring for persons with co-occurring medical and psychiatric symptoms. In the case of ALS+FTI caregivers, this becomes particularly relevant given the high probability that their loved ones will present with both medical and psychiatric symptoms, which may impact the caregiving experience. In light of this gap in the existing body of literature, this study sought to explore and describe lay-caregivers' thoughts, feelings, behaviors, practical daily challenges, and rewards in caring for persons with ALS+FTI.

Chapter 2: Literature Review

Amyotrophic Lateral Sclerosis

Amyotrophic lateral sclerosis (ALS) is one of the most common neuromuscular diseases worldwide (Armon, 2014). In the United States, approximately 12,187 Americans are living with a definite ALS diagnosis (Mehta et al., 2014). The prevalence is estimated to be 3.9 ALS cases per 100,000 persons in the general population (Mehta et al., 2014). Men are more likely to be affected by ALS than women (4.8 per 100,000 versus 3.0 per 100,000) and Caucasians are more than twice as likely to receive diagnoses in comparison to African Americans (4.2 versus 2.0 per 100,000; Mehta et al., 2014). ALS is more common in older adults, with individuals ages 70 to 79 demonstrating the highest rate of disease (17 cases per 100,000; Mehta et al., 2014).

There are two forms of ALS. The most common form is sporadic ALS and it accounts for 90% to 95% of all cases in the United States (Hartefeld, 2014). Sporadic ALS indicates that there is no clear cause for disease onset. The remaining 5% to 10% of cases are classified as familial ALS, which is the genetic or inherited form of the disease. In families who have a history of ALS, there is a 50% chance that the genetic mutation associated with ALS will be passed from parent to offspring (Hartefeld, 2014). Although the cause of ALS is not currently understood, it is believed that both environmental and genetic risk factors may be implicated.

ALS can be classified into four categories, which include classic ALS, primary lateral sclerosis (PLS), progressive bulbar palsy (PBP), and progressive muscular atrophy (PMA; Talbot, 2002). Classic ALS accounts for more than two thirds of individuals diagnosed with ALS, and is characterized by the progressive deterioration of both the

upper and lower motor neurons (Talbot, 2002). PLS is considered to be the least common type of ALS (Almeida et al., 2013). PLS is characterized by the degeneration of the upper motor neurons (Almeida et al., 2013). Approximately 25% of ALS cases are classified as PBP, which primarily affects the lower motor neurons in the brain stem (Talbot, 2002). As a result, persons with PBP generally present with difficulties in speaking, chewing, and swallowing (Talbot, 2002.). The remaining cases are classified as PMA, which is characterized by the progressive degeneration of the lower motor neurons in the spinal cord (Wijesekera et al., 2009). In most cases of PMA, if the upper motor neurons remain intact for at least 2 years, the disease is unlikely to spread beyond the lower motor neurons (Wijesekera et al., 2009).

Traditionally, ALS has been viewed as a disease that primarily affects the motor system and spares non-motor functions (Zago, Poletti, Morelli, Doretti, & Silani, 2011). Earlier versions of the diagnostic guidelines for ALS listed the presence of cognitive deficits as an exclusionary criterion for the diagnosis (Brooks, 1994). Interestingly, it was this feature of the disease that was once considered to be the silver lining juxtaposed against the devastation of ALS (Elman & Grossman, 2007). This viewpoint was best illustrated in a study that explored the positive life experiences of patients with ALS, in which one participant stated, “I continue to look beyond myself and the world . . . My world is only small physically but not mentally or spiritually . . . That is still the same” (Young & McNicoll, 1998, p. 39). For many individuals, cognitive and intellectual abilities become sources of hope and strength that help them to cope with the losses and challenges associated with ALS (Young & McNicoll, 1998). It is findings such as these

that become increasingly relevant when considering the implications of the presence of FTI in ALS.

A multisystem disease. Today, it is recognized that the neurodegeneration caused by ALS is not restricted to the motor cortex, but can also affect other areas of the brain, including the frontal and temporal lobes (Zago et al., 2011). ALS is now described as a multisystem disease that has the potential to affect both motor and non-motor functions (Zago et al., 2011). Indeed, evidence suggests that up to 50% of the ALS population demonstrates some degree of frontotemporal deficits over the course of the disease (Ringholz et al., 2005). For a subset of this population, deficits may be severe enough to warrant a diagnosis of FTD (Elman & Grossman, 2007). In one study, 52% of ALS patients met established criteria for FTD (Lomen-Hoerth et al., 2003); however, more conservative estimates indicate that FTD is present in approximately 14% of ALS patients (Phukan et al., 2012). The literature has documented enormous variations in the physical, cognitive, and behavioral presentations of ALS (J. M. Murphy et al., 2016; Zago et al., 2011).

ALS+FTI continuum. The frontotemporal deficits that occur within the ALS population appear on a continuum ranging from an absence of impairment to fulfilling the Neary criteria for FTD (Neary et al., 1998; Strong, Lomen-Hoerth, Caselli, Bigio, & Yang, 2003). Along this continuum are five categories of FTI classification: (a) ALS patients who are cognitively and behaviorally intact (classic ALS), (b) ALS patients with mild to moderate cognitive impairment, (c) ALS patients with mild to moderate behavioral impairment, (d) ALS patients with mild to moderate cognitive and behavioral impairment, and (e) ALS patients meeting criteria for FTD (Strong et al., 2009).

Although there are variations within each category, their purpose is to distinguish between the different clinical presentations and deficits noted within the ALS population.

Frontotemporal cognitive impairment. In the absence of a gold standard assessment battery or screening tool, cognitive impairment is diagnosed if the ALS patient scores below the fifth percentile on at least two measures of executive functioning (Phukan et al., 2012). It is estimated that up to 56% of the ALS population demonstrates cognitive impairments early in the course of the disease (Lule et al., 2014). ALS-associated cognitive impairments are related generally to frontally-mediated deficits in executive functioning; however, deficits in memory and language have also been observed (Phukan et al., 2012). Phukan and colleagues (2012) conducted a study to explore the clinical characteristics of cognitive impairment in ALS by administering a neuropsychological test battery that included a variety of tasks, such as the Stroop Colour-Word Test, Backward Digit Span, Wechsler Memory Scale-IV Logical Memory and Verbal Paired Associate subtests, Rey-Osterrieth Complex Figure Test, and Boston Naming Test. The cognitive impairments associated with ALS included deficiencies in verbal and nonverbal fluency response generation, working memory, cognitive flexibility, planning, problem solving, attention, judgment, and insight (Phukan et al., 2012).

The implications of ALS-associated cognitive impairment are numerous. For instance, executive dysfunction may render patients unable to manage finances, prepare meals, or care for themselves or their children (Merrilees et al., 2010). Safety issues become increasingly relevant as the patient experiences difficulties identifying ways with which to cope or avoid choking or falling after cognitive deterioration (Phukan et al., 2007). Deficits in insight and the ability to plan and problem solve become sources of

concern when faced with critical decisions about medical treatment, end-of-life wishes, and the patient's autonomy (Elman & Grossman, 2007; Lomen-Hoerth et al., 2003).

Similarly, language deficits may interfere with the patient's ability to effectively communicate his or her wishes or needs (Elman & Grossman, 2007). Moreover, cognitive impairment has also been associated with decreased treatment compliance (Kiernan, 2012).

Frontotemporal behavioral impairment. Significant changes in behavior that do not meet criteria for FTD and cannot be explained by other psychological factors are present in up to 50% of ALS patients (Gibbons et al., 2008). As is the case with cognitive impairment, there is no standardized battery to diagnose behavioral impairment within the ALS population; however, several studies have utilized self-report measures, such as the Frontal Systems Behavior Scale (FrSBe) and the Frontal Behavioral Inventory (FBI), to assess behavioral changes in this population (Woolley et al., 2010). The most frequently documented behavioral abnormalities in ALS patients include apathy, disinhibition, self-centeredness, irritability, and decreased insight (Lomen-Hoerth et al., 2003; Strong et al., 2009; Woolley et al., 2010).

ALS-associated behavioral impairment manifest in a variety of ways. The ALS patient may seem uninterested in engaging in conversation with others or previously enjoyed activities (Chio et al., 2010). They may also demonstrate a lack of emotion or concern for others, even when others are very upset or overwhelmed (Chio et al., 2010; Woolley et al., 2010). It is not uncommon for ALS patients with behavioral impairment to be described as self-centered due to preoccupation with personal problems and needs, and a lack of consideration of the needs of others (Merrilees et al., 2010). In addition,

disinhibited individuals often make improper comments or behave inappropriately during social interactions (Chio et al., 2010; Woolley et al., 2010). Oftentimes, these individuals lack insight into their behavioral deficits (Lomen-Hoerth et al., 2003; Woolley et al., 2010).

Frontotemporal dementia. The cognitive and behavioral impairments associated with FTD are more extensive and severe than the frontotemporal deficits that do not meet FTD criteria. According to Neary criteria, a diagnosis of FTD requires insidious onset and gradual progression, early decline in social interpersonal conduct, early impairment in the regulation of personal conduct, early emotional blunting, and early loss of insight (Neary et al., 1998). There are three subtypes of FTD and they often overlap (Neary et al., 2005). The behavioral variant (bvFTD) is characterized by extreme impairments in thinking and behavior, such as emotional blunting, loss of insight, and problems with personal and social conduct (Merrilees et al., 2010; Strong et al., 2009). Disturbed eating behaviors and/or impaired hygienic practices are also common features of this subtype (Merrilees et al., 2010; Neary et al., 2005). The language variants of FTD include a non-fluent progressive aphasia (PNFA) subtype and a semantic dementia (SD) subtype (Neary et al., 1998; Neary et al., 2005). The former is associated with profound difficulties with word retrieval and speech production, and the latter is characterized by the loss of ability to name or comprehend the meaning of words (Zago et al., 2011; Strong, 2008).

Symptomatically, FTD is often mistaken for non-progressive psychiatric diseases including schizophrenia, obsessive compulsive disorder, borderline personality disorder, and bipolar disorder (Monji et al., 2014; Pressman & B. L. Miller, 2013; Shinagawa et al., 2014). In one study, 50% of patients with FTD were diagnosed incorrectly with a

psychiatric disturbance before receiving a FTD diagnosis (Pressman & B. L. Miller, 2013). In the early stages of disease progression, deficits present as disinhibition, apathy, stereotypic behaviors, social misconduct, and impulsivity. ALS+FTD patients may become insensitive, fail to empathize with others, violate social norms through inappropriate comments or behaviors, or become detached in relationships (Pressman & B. L. Miller, 2013). As the disease progresses, patients often become more irritable and, at times, explosive, and exhibit antisocial behaviors or commit criminal acts (Pressman & B. L. Miller, 2013). In contrast, others become overly friendly and trusting of others, which places them at an increased risk of mistreatment. Mental rigidity, intolerance for changes in routine or schedule, ritualistic behaviors, and hoarding are common (Merrilees et al., 2010; Shinagawa et al., 2014). In addition, individuals frequently demonstrate changes in eating habits and may feel compelled to put inedible objects in their mouths, which becomes particularly relevant for the ALS population (Lomen-Hoerth et al., 2003; Merrilees et al., 2010). For instance, gorging, a prominent symptom of FTD, can threaten the life of an ALS patient who is experiencing impaired swallowing (Lomen-Hoerth et al., 2003). In the end stages of the disease, individuals need help with basic activities of daily living and are often consumed by apathy and inertia, with some individuals becoming mute or immobile (Diehl-Schmid et al., 2013; Pressman & B. L. Miller, 2013).

In terms of their clinical presentation and conceptualization, ALS and FTI differ in many respects. Within the literature, ALS is conceptualized within a medical context, in which the physical consequences of the disease are the primary focus, whereas FTI is discussed generally within a psychiatric context with an emphasis on psychological sequelae. Nonetheless, both diseases are progressive and as individuals' physical health

deteriorates, they become increasingly dependent on others for assistance with activities of daily living and beyond. In most cases, these caregiving responsibilities fall on a family member or friend, who assumes the role of primary caregiver (Lim & Zebrack, 2004). Indeed, the demands associated with these caregiving populations are complex and ubiquitous, and oftentimes affect the caregiver's psychological and physical health negatively (Deeken, Taylor, Mangan, Yabroff, & Ingham, 2003; Diehl-Schmid et al., 2013). Additionally, it is important to attend to caregiving experiences because the physical, social, and emotional health of a caregiver has the potential to influence the quality of care provided to the patient, as well as the patient's physical and psychological well-being (Gater et al., 2014; Lim & Zebrack, 2004).

Caregiving

In the United States, the healthcare system has become increasingly focused on developing strategies for providing cost-effective treatment and care to individuals with physical and/or mental illnesses (Lim & Zebrack, 2004). This has resulted in several changes within medical practices, such as shorter inpatient hospital stays and a shift to the use of outpatient services, which are less costly than around-the-clock medical care (Deeken et al., 2003). For individuals with chronic illnesses, there has been an increased reliance on home-based care, in which the majority of the caregiving responsibilities fall on patients' families or friends (Gater et al., 2014; Lim & Zebrack, 2004).

A caregiver is an unpaid individual who assists a person who is elderly, ill, and/or disabled with activities of daily living and/or medical care on a daily basis. According to a study conducted by the National Alliance for Caregiving and the American Association of Retired Persons (AARP; 2009), more than 65 million individuals in the United States

provide uncompensated care to disabled, ill, and/or elderly family members or friends. Depending on the illness or disability, the responsibilities associated with caregiving may range from transportation and meal preparation to more challenging tasks, such as bathing, toileting, feeding, and transferring the individual in and out of bed (Elliott & Rivera, 2006; National Alliance for Caregiving & AARP, 2009). On average, caregivers spend approximately 20 hours per week performing caregiving duties; however, caregivers who cohabit with care recipients spend an average of 39.3 hours per week in their role as caregivers (National Alliance for Caregiving & AARP, 2009). Most individuals serve as caregivers for 4.6 years.

The caregiving experience is complex and intense. Caregivers are often encumbered by the challenge of balancing their multiple roles and non-caregiving responsibilities with the demands associated with caring for their loved ones. Given the amount of time and energy required to care for a loved one, it is not surprising that caregivers are at high risk of diminished QOL (National Alliance for Caregiving & AARP, 2009).

Quality of life. The World Health Organization (WHO) defines QOL as “individuals’ perceptions of their position in life in the context of culture and value systems in which they live and in relation to their goals, expectations, standards and concerns” (The WHOQOL Group, 1996, p. 3). It is viewed as a multidimensional construct that encompasses individuals’ subjective evaluations of their physical health (e.g., pain, energy, fatigue), psychological health (e.g., positive and negative feelings, spirituality, religion, personal beliefs), social relationships (e.g., personal relationships, practical social supports), and environment (e.g., financial resources, recreation and

leisure, home environment; The WHOQOL Group, 1996). QOL does not merely represent the symptoms of a disease; rather, it reflects an individual's perceptions of the effects that a disease, impairment, or intervention has had on his or her physical health, psychological well-being, social relationships, and environment.

Many studies address the adverse consequences of caregiving on various aspects of QOL. In regard to psychological health, caregiving has been associated with increased stress, emotional distress, burden, and strain (Bennett, Fagundes, & Kiecolt-Glasser, 2013; Glozman, 2004; Pagnini et al., 2010). Within the physical health domain, research indicates caregiving has a negative effect on the immune system, which increases a caregiver's susceptibility to infection, cardiovascular disease, and stroke (Bennett et al., 2013; Lambert, Jones, Girgis, & Lecathelinais, 2012; Pagnini et al., 2010). The social and environmental domains are also affected, as evidenced by caregivers' reports of decreased social satisfaction, and increased social isolation and financial strain (Glozman, 2004). In contrast to these findings, some caregivers report positive experiences associated with caregiving, including shifts in personal and family values, greater feelings of closeness, and improved family and personal relationships (Perlesz, Kinsella, & Crowe, 1999; Reinhard, Samis, & Levine, 2014).

As mentioned previously, caregiving experiences cannot be generalized across illnesses because each disease imposes its own unique challenges and demands upon caregivers (Clipp & George, 1993). For instance, chronically ill individuals often require permanent assistance. Therefore, the caregiving experience is not transitory; rather, it represents a permanent lifestyle change that transforms the lives of both the caregiver and care recipient. In contrast, caregiving for an individual undergoing treatment for an

illness is frequently temporary; however, the caregiving experience is complicated by the severe and sudden onset of treatment side effects, which places a high demand on the caregiver at the outset of the caregiving experience. Furthermore, caregiving within a medical context may affect a caregiver's QOL differently than caregiving within a psychiatric context. As such, it is important to address QOL across caregiving populations to truly appreciate the caregiving experience and its implications. Table 1 depicts a summary of biopsychosocial challenges across caregiving populations (see Appendix A).

Implications of Caregiving: Chronic Medical Conditions

Amyotrophic lateral sclerosis. The role of caregiver for a patient with ALS represents an unremitting commitment with constantly changing responsibilities (Aoun, Connors, Priddis, Breen, & Colyer, 2012). Although ALS caregiving can be rewarding, caregivers are often faced with increasingly challenging demands, including physical tasks, financial costs, providing emotional support, managing medical equipment, and assisting with nearly all activities of daily living (Aoun et al., 2012). As ALS progresses, the patient and caregiver's home becomes inundated with medical supplies and equipment, and an increasing amount of the caregiver's time is devoted to caregiving tasks (Rabkin & Albert, 2009). Approximately half of all caregivers spend 12 hours per day caring for an ALS patient (Aoun et al., 2012), with caregivers of ALS patients with assisted ventilation devices providing an average of 14 hours each day (Kaub-Wittemer, von Steinbüchel, Wasner, Laier-Groeneveld, & Borasio, 2003). Oftentimes, caregiving responsibilities are performed at the expense of the caregiver's physical, psychosocial, and financial well-being (Aoun et al., 2012; Galvin et al., 2016; Rabkin & Albert, 2009).

Physical concerns. A majority of caregivers experience difficulty caring for themselves as a result of the tremendous time commitment associated with caring for their loved ones (Aoun et al., 2013). A review of the literature indicates that ALS caregivers are concerned about their own physical health (Aoun et al., 2013; Pagnini, 2013), with one study finding that up to 42% of caregivers reported that they felt unwell physically (Krivickas, Shockley, & Mitsumoto, 1997). According to Aoun and colleagues (2013), the most commonly expressed physical concerns of ALS caregivers were physical exhaustion, fatigue, and insomnia.

Psychosocial concerns. A review of the literature indicates that the psychological well-being of ALS caregivers is affected negatively by the caregiving experience, and worsens over time (Gauthier et al., 2007). One study revealed nearly half of ALS caregivers reported feeling unwell psychologically (Krivickas et al., 1997). Indeed, increased rates of psychological distress, depression, anxiety, burden, and stress have been documented throughout the research (Aoun et al., 2013; Gauthier et al., 2007; Pagnini, 2013). It has been indicated that as the dependency needs of care recipients increase, ALS caregivers experience higher rates of anxiety, insomnia, and decreased QOL (Aoun et al., 2012). In addition, higher levels of depression have been associated with increasing caregiving intensity, supervision time, patient criticism, and frequent changes and losses experienced by ALS caregivers (Aoun et al., 2013). When compared to caregivers of patients with similar neurodegenerative diseases, ALS caregivers reported the lowest QOL (Aoun et al., 2012). Furthermore, several studies have documented the positive influence of social support on the ALS caregiving experience. Increased social support and patient supportiveness are associated with more positive

feelings about the caregiving experience, decreased burden, and reduced psychological distress in ALS caregivers (Aoun et al., 2013; Pagnini, 2013; Rabkin, Albert, Rowland, & Matsumoto, 2009).

The financial costs associated with ALS are tremendous, and the financial burden can be devastating. In the end stages of the disease, costs associated with medical care and mechanical ventilation devices can exceed \$200,000 per year (Rabkin & Albert, 2009). Although health insurance coverage may alleviate some of the financial strain, many families are forced to pay out of pocket or rely on charitable donations to cover the costs of medications, home health assistants, household renovations, and long term mechanical ventilation devices (Obermann & Lyon, 2015; Rabkin & Albert, 2009). Furthermore, given the increasing demands placed on the caregiver, these financial concerns are compounded by reductions in annual income due to decreased work hours for both the patient and caregiver.

Despite the preponderance of literature documenting the adverse impact of caregiving for ALS patients, there is evidence to suggest not all caregivers are affected negatively by their caregiving responsibilities. Indeed, even under the most stressful circumstances, some ALS caregivers do not experience clinically significant depressive symptoms (Rabkin et al., 2009). Furthermore, findings have shown a subset of the ALS caregiving population views their experiences as personally satisfying and maintains a positive outlook by finding meaning in their roles as caregivers (B. Miller & Lawton, 1997).

Spinal cord injury. Caregivers play an active role in the recovery of family members after spinal cord injury (SCI; Arango-Lasprilla et al., 2010). SCI is often

associated with motor and sensory impairments, such as paralysis or weakness, depending on the site and severity of the damage (Arango-Lasprilla et al., 2010). In moderate to severe cases, the caregiving demands can be extensive, with estimates ranging from an average of 11.3 hours of care per day to 80 hours per week (Arango-Lasprilla et al., 2010). Informal caregivers for individuals with SCI usually assist with activities of daily living and beyond, including bathing, dressing, feeding, transferring, and toileting (Dreer, Elliott, Shewchuk, Berry, & Rivera, 2007).

Physical concerns. SCI caregivers report lower overall physical health status while caregiving (K. T. Lucke, Coccia, Goode, & J. F. Lucke, 2004). The most common physical complaints associated with SCI caregiving include body pain, decreased energy levels and vitality, and moderate fatigue (Blanes, Carmagnani, & Ferreira, 2007; K. T. Lucke et al., 2004). These caregivers experience higher rates of physical illness and health than the general population and report more frequent visits to doctors' offices (Blanes et al., 2007). In an attempt to alleviate some of the physical demands of caregiving, SCI caregivers often hire professional caregivers or attendants to assist with caring for their family members (K. T. Lucke et al., 2004).

Psychosocial concerns. SCI caregivers describe their caregiving responsibilities as relentless, and often experience difficulty balancing their caregiving roles with their personal responsibilities (Dickenson, O'Brien, Ward, Allan, & O'Carroll, 2010; Ellenbogen, Meade, Jackson, & Barrett, 2006). The demands associated with caregiving often leave the caregiver with limited time to engage in social or leisure activities or attend appointments with his or her own physicians (Ellenbogen et al., 2006). SCI caregivers report elevated levels of stress, physical and emotional burden, depression,

anxiety, anger, resentment, and dissatisfaction with life (Arango-Lasprilla et al., 2010; Schulz et al., 2009). In a study conducted by Schulz and colleagues (2009), SCI caregivers' burden and depression scores were slightly higher than those reported by caregivers of Alzheimer's patients. This caregiving population is also faced with the overwhelming financial costs related to the need for special transportation services and vehicles, medical supplies and equipment, and adapting their homes to accommodate their family members' needs (Arango-Lasprilla et al., 2010; Ellenbogen et al., 2006). SCI caregivers often feel overwhelmed by their caregiving roles, but many caregivers also describe positive experiences while caring for their loved ones, such as the development of stronger bonds with their loved ones and becoming more aware of each other's needs (Dickenson et al., 2010). These caregivers report feelings of success and achievement in their ability to maintain and strengthen their relationships (Dickenson et al., 2010).

Cancer. As is the case for persons with ALS and SCI, relatives and friends are often the primary source of support and care for individuals diagnosed with cancer (Kim & Schulz, 2008). The challenges associated with cancer caregiving are vast, and are further complicated by the unpredictable nature of the disease course, symptom severity, and patient response to treatment, if available (Kim & Schulz, 2008). Cancer patients often require assistance sporadically and for limited periods of time, with the majority of care needs occurring after surgery, during treatment, or at the end of life (Kim & Shulz, 2008). Thus, caregivers must be able to adapt to constantly changing routines, treatment needs, home care procedures, and symptom severity (B. A. Given, C. W. Given, & Sherwood, 2012).

Similar to other chronic or terminal illnesses, cancer caregivers offer assistance with transferring, feeding, dressing, showering, preparing meals, coordinating appointments, and clinical care tasks, such as medication administration (Deshields et al., 2012; Kim & Schulz, 2008). On average, cancer caregivers provide 54.6 hours of care per week to their ill relatives or friends (Deshields et al., 2012). In a study comparing the caregiving experiences of cancer, dementia, and frail elderly caregivers, Kim and Shulz (2008) found that cancer caregivers provided the greatest amount of care per week; however, in contrast to dementia caregivers, cancer caregivers provided more medically-based care activities, such as changing catheters or colostomy bags (Kim & Shulz, 2008). Oftentimes, these caregivers are forced to perform clinical care tasks that would otherwise be performed by trained medical professionals (van Ryn et al., 2010). Nonetheless, cancer caregivers must learn to monitor their loved ones for signs of treatment side effects, help manage their fatigue, pain, or nausea, decide whether more medications are needed, administer medications, judge whether a doctor's care is required, and change bandages (van Ryn et al., 2010). Given these demands, it is understandable that caregiving can affect the caregiver's physical and psychosocial health.

Physical concerns. Caregiving for a loved one with cancer can be detrimental to the caregiver's physical health. These caregivers often report increased physical health issues and lowered physical functioning after caregiving (Girgis et al., 2013; B. A. Given et al., 2012). In addition, pain, headaches, insomnia, fatigue, altered appetite, weight gain, high blood pressure, and altered immune response are commonly associated with caring for a loved one with cancer (Girgis et al., 2013; B. A. Given et al., 2012).

Psychosocial concerns. One of the most distressing aspects of cancer caregiving is observing the overt signs of physical pain, discomfort, and suffering exhibited by the person with cancer (Deshields et al., 2012; Kim & Shulz, 2008). At times, this distress becomes so great that the caregiver demonstrates higher distress levels than the person with cancer (Girgis et al., 2013). Cancer caregivers are also at an increased risk for high levels of strain, burden, anxiety, depression, worry, anger, posttraumatic stress, and diminished QOL (Girgis et al., 2013; B. A. Given et al., 2012; van Ryn et al., 2010).

In a study investigating the long-term effects of cancer caregiving, Kim and colleagues (2014) revealed that caregivers demonstrated high levels of clinical depression 5 years after the initial cancer diagnoses for both current and bereaved caregivers. In addition, findings from several studies reported high levels of social isolation and financial stress in cancer caregivers. In regard to the former, many caregivers described a loss of social support following cancer diagnoses and many reported being the sole caregiver of their loved ones (Girgis et al., 2013; van Ryn et al., 2010). In regard to the latter, caregivers reported a loss of work hours or complete loss of employment due to the demands of caregiving (B. A. Given et al., 2012). This loss of income becomes a significant source of concern for families because of the astronomical expense for medical treatments, which can cost upwards of \$100,000 per year (B. A. Given et al., 2012). Unfortunately, in addition to caregiving demands and the potential loss of loved ones, many families are forced to sell their homes, take out loans, or declare bankruptcy to manage their medical expenses (B. A. Given et al., 2012). Nevertheless, in the midst of these difficult times, some caregivers maintain a high QOL and describe an array of positive experiences associated with caregiving, such as becoming a stronger and more

empathic person, growing emotionally and spiritually, having a greater appreciation for life, strengthening bonds with family, and a deeper sense of purpose in life (Kim, Shulz, & Carver, 2007).

Implications of Caregiving: Chronic Psychiatric Conditions

Dementia and frontotemporal impairment. Caregivers of FTI patients face a series of unique challenges that distinguish their experiences from those of dementia caregivers. For instance, since FTD is less common and not as well understood as other forms of dementia, FTD caregivers experience more difficulty accessing social and professional supports than do caregivers of other types of dementia (Riedijk et al., 2006). In addition, the lack of knowledge about FTI frequently results in improper diagnosis of the patient, which delays the patient and caregiver in discussing plans for the future and may lead to inappropriate treatment recommendations (Pressman & B. L. Miller, 2013). Beyond diagnosis and treatment, family members often do not have a choice in undertaking their caregiving roles because many community-based programs or long-term facilities refuse to admit patients with FTI due to inadequate understanding of the disease and the challenges associated with caring for these individuals (Morhardt, 2011). Thus, FTI caregivers are often left without alternatives for care or support while caring for loved ones.

The challenges associated with providing home-based care for patients with FTI are vast. Caregivers have to constantly supervise and monitor patients due to the behavioral problems associated with FTI. For example, FTI caregivers often resort to locking kitchen cabinets to prevent patients from overindulging or choking on food (Pressman & B. L. Miller, 2013). In the later stages of disease, the demands of

caregiving increase as their loved ones require assistance with the most basic activities of daily living (Diehl-Schmid et al., 2013). Certainly, the demands placed on FTI caregivers are profound and likely affect many aspects of the caregivers' lives; however, a review of the literature reveals a paucity of research on FTI caregivers. Thus, in an attempt to convey the caregiving experiences of FTI caregivers, the remainder of this review will focus on dementia caregivers, in general, and will make reference to FTI caregivers if information is available.

Caring for a loved one with dementia is associated with long hours and physically and emotionally demanding tasks (Brodaty & Donkin, 2009). Family members must learn to manage the frequently changing demands and unpredictable problem behaviors of their loved ones (Tremont, 2011). Dementia caregivers assist their loved ones with an array of daily activities, such as dressing, bathing, and managing finances and treatment needs (Brodaty & Donkin, 2009). On average, dementia caregivers provide at least 40 hours of care per week to their loved ones (Brodaty & Donkin, 2009), and often do so at the expense of their own needs (Tremont, 2011).

Physical concerns. In comparison to non-caregivers, dementia caregivers experience a greater number of physical health problems and report worse physical health overall (Brodaty & Donkin, 2009). One study indicated that the behavioral problems of the care recipient predict the physical health of the caregiver and are associated with greater levels of exhaustion and fatigue (Hooker et al., 2002). Other studies linked the physical health of the caregiver to the severity of cognitive and behavioral impairment of the care recipient, cohabitation with the care recipient, longer duration of caregiving, and less informal support (Tremont, 2011). Dementia caregivers demonstrated a range of

physical health problems, such as cardiovascular problems, poor immune response, poor sleep patterns, and higher rates of chronic conditions, smoking, and alcohol intake (Brodaty & Donkin, 2009).

Psychosocial concerns. Research indicates that caregiving for a person with dementia is more stressful than caring for someone who is elderly and medically compromised (Tremont, 2011). In addition, studies suggest dementia caregiving is more burdensome than caregiving for other populations (Brodaty & Donkin, 2009). The most common symptoms of psychological distress in dementia caregivers include high levels of depression and anxiety (Tremont, 2011). Socially, dementia caregivers frequently report feeling isolated as a result of the time demands associated with caregiving and the need to sacrifice hobbies and leisure activities, reduce or terminate employment, and restrict social interactions or gatherings with friends and families (Brodaty & Donkin, 2009). Dementia caregivers who were satisfied with their social lives and interactions with others exhibited fewer psychological symptoms than those who felt socially isolated (Brodaty & Donkin, 2009).

The literature reveals FTD caregivers are highly stressed and burdened (Diehl-Schmid et al., 2013). In fact, burden and stress are higher among FTD caregivers than other dementia caregivers, including Alzheimer's disease (Diehl-Schmid et al., 2013; Rosness, Haugen, & Engedal, 2008). A study on FTD caregivers found that caregiver burden was connected to patient need for supervision at home, lack of manners, aggression, changes in personality, and difficulties swallowing (Diehl-Schmid et al., 2013; Rosness et al., 2008). For FTD caregivers, the most troubling aspects of caring for individuals with FTD were reported as dealing with the loss of their loved ones, disease

course and progression, and feelings of hopelessness (Diehl-Schmid et al., 2013). Moreover, the patient-caregiver relationship is affected by presence of FTD, with one study reporting that 50% of relationships worsened, 30% stayed the same, and 20% improved (Diehl-Schmid et al., 2013).

In contrast, although there has been tremendous focus on the adverse impact of dementia caregiving, it is important to understand that not all caregivers experience burden and distress (Tremont, 2011). A study conducted by Sanders (2005) revealed 50% to 90% of caregivers reported positive experiences, such as feelings of accomplishment, personal and spiritual growth, increased faith, and enjoyment of the time spent bonding with the care recipients. Other studies indicated that caregiving made them feel needed and useful, and helped them develop positive attitudes, appreciate life, and strengthen their relationships with their loved ones (Tremont, 2011).

Schizophrenia. Schizophrenia is associated with a deterioration of the frontal and temporal lobes that manifests as deficits in executive functioning, memory, and verbal response fluency (Chan et al., 2014). Thus, individuals with schizophrenia who live in the community are often highly dependent on their caregivers for assistance with basic needs (Chan et al., 2014). Caregivers provide assistance with tasks, such as managing appointments, medication, personal hygiene, and meal preparation (Gater et al., 2014). It is estimated that schizophrenia caregivers provide an average of 6 to 9 hours of care per day (Awad & Voruganti, 2008). Furthermore, it is not uncommon for these caregivers to become overwhelmed with their responsibilities and, consequently, seek the help of formal paid assistants (Gater et al., 2014).

Physical concerns. Research indicates that caregivers of persons with schizophrenia face significant physical health risks and frequently report hospitalizations or visits to emergency rooms (Awad & Voruganti, 2008; Mitsonis et al., 2012). The most common physical complaints involve physical exhaustion, including feeling drained, worn out, and tired (Gater et al., 2014). These caregivers obtain less sleep and experience difficulties falling and staying asleep (Gater et al., 2014).

Psychosocial concerns. Awad and Voruganti (2008) revealed the burden of caring for people with chronic psychiatric disorders is similar to the burden of caring for people with chronic neurological disorders; however, their findings also indicate that schizophrenia caregivers demonstrate higher levels of subjective burden than caregivers of persons with neurological disorders because of their experiences with stigma, inadequate social support, and lack of acceptance from society. Studies also suggest that feelings of embarrassment, shame, and frustration are common in schizophrenia caregivers because of their loved ones' unusual behaviors at home and in public (Gater et al., 2014). These caregivers also sacrifice their social lives due to limited time, exhaustion, fear of leaving their loved ones unattended, and feeling too embarrassed to invite friends or families to their homes (Gater et al., 2014). Moreover, individuals living with schizophrenia frequently depend on their caregivers for financial support (Gater et al., 2014). Financial strains are often exacerbated by caregiver loss of income as a result of frequent absence or the decision to discontinue their employment to care for their loved ones (Gater et al. 2014). Despite these challenges, schizophrenia caregivers also report positive experiences. These caregivers enjoyed the experience because they were able to ensure their family members were safe, received quality care, and felt understood

(Gater et al., 2014). Furthermore, caregivers described the experience as rewarding and satisfying, and as helping them to become more patient, grow into better people, and share their love with the care recipients (Gater et al., 2014).

Bipolar disorder. Individuals diagnosed with bipolar disorder (BP) frequently exhibit impaired cognitive functioning in a variety of domains, such as executive function, attention, psychomotor speed, learning, and memory (Martinez-Aran et al., 2004). Similar to individuals diagnosed with schizophrenia, the deficits observed in BP are believed to be associated with abnormalities within the frontal and temporal lobes, hippocampus, cerebellum, and amygdala (Martinez-Aran et al., 2004). Depending on the severity of illness, persons with BP may rely on family members to provide partial or total management of their care (Goossens, Van Wijngaarden, Knoppert-Van Der Klein, & Van Achterberg, 2008). BP caregivers are frequently called upon to manage their loved ones' finances, medical and psychological treatment, medication regimens, problem behaviors, and legal issues (Dore & Romans, 2001). Although the amount of care provided varies, BP caregivers report up to 32 hours of care and/or contact with their ill relatives per week (Goossens et al., 2008). As is true in other caregiving populations, caring for a family member with BP can be demanding and distressing, which has the potential to affect the health and QOL of the caregiver negatively (Reinares et al., 2006).

Physical concerns. There is evidence to suggest caregiving for a loved one with BP can affect the caregiver's physical health negatively. A study conducted by Ogilvie and colleagues (2005) revealed that BP caregivers report increased physical illness following the onset of their caregiving responsibilities. More specifically, BP caregivers

describe a wide range of physical concerns, such as increased tension, physical exhaustion, tiredness, insomnia, muscle pain, and mental fatigue (Tranvag & Kristoffersen, 2008). As such, BP caregivers report using healthcare services more often than the general population (Perlick, Hohenstein, Clarkin, Kaczynski, & Rosenheck, 2005).

Psychosocial concerns. Caregivers of loved ones with BP experience rates of burden that are at least as high as those reported by schizophrenia caregivers (Perlick et al., 2005). This level of burden is often influenced by the symptoms exhibited by the individual diagnosed with BP, such as the presence of aggressive or violent behavior, impulsive spending, depression, suicidal ideation, irritability, hyperactivity, and social withdrawal (Reinares et al., 2006). In addition to burden, BP caregivers report high levels of stress, depression, anxiety, frustration, and social isolation (Ogilvie, Morant, & Goodwin, 2005; Steele, Maruyama, & Galynker, 2010), which makes them twice as likely to use mental health services than the general population (Steele et al., 2010). A frequent consequence of caregiving for a loved one with BP is strained relationships between the caregiver and care recipient, family members, and/or friends (Dore & Romans, 2001). A survey of spousal caregivers indicated that many believed they would not have entered into relationships with the person diagnosed with BP if they had been more knowledgeable about the illness and its associated consequences (Dore & Romans, 2001). Moreover, many caregivers reported feeling embarrassed by their loved ones' illness and endorsed experiencing stigma because of it (Dore & Romans, 2001).

In contrast, some BP caregivers report positive experiences with caregiving, such as feeling closer with the care recipients (Dore & Romans, 2001). Other caregivers

believed their caregiving roles helped them to improve their listening skills and become more nurturing people (Dore & Romans, 2001). Despite the challenges associated with BP caregiving, studies suggest that many caregivers are reasonably happy and want to continue living with and caring for their loved ones (Goossens et al., 2008; Steele et al., 2010).

Implications of Caregiving: Chronic Medical and Psychiatric Conditions

The caregiving literature addresses medical conditions separately from psychiatric conditions and seldom addresses the experiences of those caring for persons with co-occurring medical and psychiatric symptoms. A recent study compared the experiences of caregivers providing care for family members with cognitive conditions, behavioral health conditions, or both, with that of family caregivers providing medically-based care for individuals without co-occurring conditions (Reinhard et al., 2014). Caregivers of individuals with combined conditions reported performing medical or nursing tasks as well as assisting with basic activities of daily living (e.g., bathing, feeding) and instrumental activities of daily living (e.g., transportation, cooking; Reinhard et al., 2014). The study's findings revealed that caregivers of people with combined conditions provided care for longer periods of time, viewed caregiving responsibilities as more challenging and time consuming, reported more pressure to assume the role of primary caregiver or believed they did not have a choice in caring for their family members, and experienced higher levels of stress, depression, and strain than caregivers of individuals without combined conditions (Reinhard et al., 2014). Based on these findings, it appears the caregiving experiences of those providing care to persons with co-occurring medical and psychiatric symptoms are different than the experiences of medical caregivers or

psychiatric caregivers. Thus, research on the combined caregiving population, such as those providing care to ALS+FTI patients, warrants further exploration.

HIV-associated dementia complex. HIV-Associated Dementia Complex (HAD) is a progressive neurological condition, similar to subcortical dementia, that affects individuals who are infected with human immunodeficiency virus (HIV; Nath et al., 2008). HAD is characterized by frontotemporal impairment that causes severe motor, cognitive, and behavioral impairments (Morrow, Allen, & Campbell, 1997), which resemble those experienced by ALS+FTI patients. In that regard, behaviorally, HAD patients demonstrate impaired judgment in terms of impulsivity and poor decision-making; personality changes including irritability, apathy, and decreased social interest; mood disturbance related to depression, anxiety and extreme mood swings; and less commonly, psychotic behavior (Meadows, Le Marechal, & Catalan, 1999; Morrow et al., 1997). Cognitively, HAD is associated with executive dysfunction, memory loss, and decreased concentration and processing speed (Prachajkul & Grant, 2003; Nath et al., 2008). Lastly, the motor deficits that characterize HAD may include musculoskeletal pain, muscle wasting, limb weakness, tremors, loss of balance and coordination, and gait difficulties (Nath et al., 2008; Prachakul & Grant, 2003). With disease progression, persons with HAD demonstrate increasing levels of confusion, lethargy, weakness, and memory loss (Family Caregiver Alliance, 2012).

Family caregivers are instrumental in aiding persons with HAD (Leblanc, London, & Aneshensel, 1997). It is estimated that HAD caregivers spend an average of 8.5 hours per day in their caregiving roles, with the amount of time spent caring increasing with disease progression and behavior problems (Prachakul & Grant, 2003).

Caregiving for persons with HAD is associated with a wide range of biopsychosocial consequences that impact the caregiving experience and the caregiver's QOL; however, these family members are often overlooked by health professionals and loved ones because the focus generally remains of the ill patients (Stetz & Brown, 2004).

Physical concerns. HAD caregivers often experience weight loss, fatigue, and physical exhaustion due to the demands of their caregiving roles (Stajdugar & Davies, 1998). These physical consequences have been associated with the severity of the patient's illness, number of symptoms, duration of caregiving, and the presence of behavior problems (Prachakul & Grant, 2003). HAD caregivers often describe the physical demands of caregiving as overwhelming, ongoing, and never-ending (Stajduhar & Davies, 1998).

Psychosocial concerns. The literature suggests HAD caregivers frequently experience high levels of psychological distress, anger, fear, stress, and burden (Prachakul & Grant, 2003; Stajduhar & Davies, 1998). In regard to psychological distress, sadness, depression, and feelings of hopelessness are most commonly reported by HAD caregivers, with a small proportion of caregivers experiencing suicidal ideation (Prachakul & Grant, 2003). HAD caregivers frequently endorse the patients' memory and behavior problems as the most challenging aspects of the caregiving experience (Flaskerud, Carter, & Lee, 2000). Studies also suggest these caregivers experience a significant degree of social isolation due to stigmatization, withdrawal of support systems, and adversarial relationships with healthcare providers (Morrow et al., 1997; Stetz & Brown, 2004). Nevertheless, despite limited social support, evidence suggests HAD caregivers view their experiences as more rewarding and less distressing than

caregivers of persons with HIV (without dementia) or cancer (Flaskerud et al., 2000; Stetz & Brown, 2004). Furthermore, some found positive meaning in the caregiving experience and reported feeling useful, fulfilled, stronger, and more spiritual (Prachakul & Grant, 2003).

Traumatic brain injury. Individuals who have suffered a traumatic brain injury (TBI) experience long-term and sometimes permanent cognitive, behavioral, physical, and emotional impairments (Corrigan & Hammond, 2013). The cognitive deficits associated with TBI may include memory loss, decreased attention and concentration, executive dysfunction, and slowed processing speed (Degeneffe, 2001). Individuals with TBI also demonstrate behavioral impairments such as impulsivity, disinhibition, agitation, and withdrawal, and may exhibit aggressive behaviors toward others (e.g., hitting, pushing, swearing) and changes in personality (Degeneffe, 2001; Sutter, 2014). Physically, hemiparesis, sensory impairments, bowel and bladder dysfunction, and seizures commonly occur after TBI (Degeneffe, 2001). The expression of deficits post-TBI varies for each individual depending on location and severity of the injury (Degeneffe, 2001). For moderate to severe cases, rehabilitation can last from years to a lifetime (Rotondi, Sinkule, Balzer, Harris, & Moldovan, 2007); therefore, home-based care is usually necessary, with a majority of this responsibility falling on family members (Degeneffe, 2001; Kreutzer et al., 2009).

Family caregivers of TBI patients provide an average of 54 (Sutter, 2014) to 60 hours of care per week (Arango-Lasprilla et al., 2011). TBI caregivers provide assistance with activities of daily living, instrumental tasks, emotional support, and managing appointments and finances (Kreutzer et al., 2009; Rotondi et al., 2007). Additionally,

TBI caregivers are faced with the challenges of managing problem behaviors and inappropriate sexual expressions, monitoring medications, and addressing issues regarding alcohol and drug use (Degeneffe, 2001). As a result of these demands, caregivers report little time for themselves (Sutter, 2014).

Physical concerns. TBI caregivers report decreased health related QOL and poor general health after assuming their caregiving roles (Arango-Lasprilla et al., 2011). More specifically, caregivers describe experiencing bodily pain related to physically demanding caregiving tasks, such as washing, dressing, and transferring their loved ones (Arango-Lasprilla et al., 2011). Additionally, TBI caregivers report sleep disturbances, fatigue, decreased energy and vitality, and changes in appetite (Arango-Lasprilla et al., 2011; Sutter, 2014; Wells, Dywan, & Dumas, 2005).

Psychosocial concerns. Findings from a study that compared the experiences of caregivers of persons with TBI, dementia, multiple sclerosis (MS), and SCI revealed that TBI caregivers demonstrated the greatest amount of mental health problems, burden, and unmet needs, and were least satisfied with their lives compared to dementia, MS, and SCI caregivers (Kreutzer et al., 2009; Sutter, 2014). A review of the literature indicates TBI caregivers demonstrate clinically significant levels of depression and anxiety, poor QOL, anger, and high levels of social isolation and burden (Kreutzer et al., 2009; Sutter, 2014; Wells et al., 2005). Remarkably, one study reported that high levels of perceived burden in TBI caregivers can last for as long as 7 years post-injury (Davis et al., 2009).

Consistent with the FTI and HAD caregiving literature, the presence of cognitive and/or behavioral changes in the persons with TBI appears to be a significant predictor of a

caregiver's well-being, mental health, perceived burden, and QOL (Sutter, 2014; Wells et al., 2005).

Findings from a study conducted by Wells and colleagues (2005) revealed that poor impulse control on the part of the patient was associated with greater caregiver anger and hostility, and poor planning or a lack of ability to foresee the future resulted in the caregiver feeling more negatively about the caregiving experience. In addition, the patient's inability to empathize or appreciate the caregiver's stress resulted in significantly reduced caregiver ratings of overall satisfaction with life. Physical restrictions and memory loss were poor predictors of caregiver well-being (Wells et al., 2005), which seems to be consistent with the literature (Davis et al., 2009; Marsh, Kersel, Havill, & Sleigh, 1998; Sutter, 2014). Thus, the presence of behavioral impairments in TBI patients adversely affects almost all aspects of the caregiving experience and frequently disrupts family dynamics and relationships (Wells et al., 2005). Behavioral impairments have also been associated with reduced social functioning and increased social isolation and withdrawal on the part of TBI caregivers (Wells et al., 2005). In comparison to dementia, SCI, and MS caregivers, TBI caregivers reported the most unmet financial and social support needs (Sutter, 2014), with high levels of impairment in social functioning reported in caregivers of TBI patients with behavioral impairment (Wells et al., 2005).

In contrast to the adverse consequences of caregiving, TBI caregivers also report positive caregiving experiences. For instance, some find their caregiving roles empowering because of the amount of help they offer to their loved ones (Degeneffe, 2001), and some caregivers report feeling more confident as a result of their ability to

provide care for ill family members (Wells et al., 2005). Overall, Wells and colleagues (2005) reported that caring for disabled family members was perceived by most TBI caregivers as a more positive than negative experience.

Amyotrophic lateral sclerosis and frontotemporal impairment. Caregivers of individuals with ALS+FTI are faced with the challenge of providing care for loved ones with at least two incurable, progressive illnesses. The combined impact of these diseases affects almost all aspects of the patient's physical, cognitive, social, and emotional health, which has the potential to intensify the caregiving experience and impact the caregiver's health. In an isolated study, Chio and colleagues (2010) found that the presence of frontotemporal neurobehavioral symptoms in ALS patients had a profound negative impact on caregivers' QOL, psychological health, and perceived burden. Other studies revealed that the burden associated with caregiving for an ALS patient with cognitive and behavioral impairments is significant and likely more stressful than caregiving for a physical disability in the absence of psychiatric symptoms (Zago et al., 2011). Furthermore, a column published by Simmons (2007) discussed the complications associated with caring for a cognitively impaired ALS patient while making life-altering medical decisions regarding advance directives, assisted ventilation, and feeding tubes. Despite the paucity of research on this population, the few studies that have explored FTI within ALS reveal that the challenges faced by these individuals and their caregivers are unique and not yet understood.

Summary

The body of literature that has investigated the experiences of the ALS population and FTI population separately has provided insights into each disease's unique impact on

the patient and caregiver. In the literature, ALS is conceptualized within a medical context, whereas FTI is generally discussed within a psychiatric context. Despite this separation, both diseases are characterized as progressive illnesses that often result in the need to enlist family caregivers to assist their loved ones with activities of daily living and beyond. Although the relationship between ALS and FTI has recently received attention in the empirical literature, the experience of the caregiver remains largely unexplored.

The purpose of this qualitative study was to explore and describe lay-caregivers' thoughts, feelings, behaviors, practical daily challenges, and rewards in caring for persons with ALS+FTI. It also offers suggestions for assessing FTI within the ALS population to facilitate early identification of FTI and assist caregivers in managing problematic and challenging behaviors exhibited by ALS+FTI patients. It is hoped that the information obtained from this study increases understanding of the unique experiences and challenges faced by this caregiving population and, ultimately, contributes to the development and implementation of biopsychosocial interventions that address the specific needs of individuals caring for persons with ALS+FTI.

Chapter 3: Research Questions

The goal of this study was to provide answers to the following questions:

1. What are the thoughts, feelings, and behaviors of lay-caregivers who provide care for persons with concurrent ALS and FTI?
2. What are the practical daily challenges and rewards in caring for persons with ALS+FTI?
3. What methods can be used to evaluate FTI in ALS patients?

Chapter 4: Method

Research Design

A grounded theory approach to qualitative methodology was used to explore and describe caregivers' experiences in caring for individuals with ALS+FTI. A qualitative approach was selected for this study because of its emphasis on answering questions of "what," "how," or "why," as opposed to questions of "how much" or "how many," which are often addressed by quantitative research (Green & Thorogood, 2009, p. 5).

Qualitative research seeks to bring individual experiences to life by providing in-depth descriptions of how a particular experience is perceived and endured, as well as by conveying the meaning of the experience for those who are living it (Kazdin, 2002).

Ultimately, qualitative research aims to provide a deep, empathic understanding of human experience, whereas quantitative research seeks to measure a construct of interest (Green & Thorogood, 2009; Kazdin, 2002). Additionally, qualitative approaches are often selected when there is a paucity of research in the area of interest, thereby providing investigators with a method to explore and understand an understudied phenomenon (Strauss & Corbin, 1998). Thus, a qualitative approach was well suited to this study's overarching goal of understanding the experiences of ALS+FTI caregivers.

Participants

The sample consisted of four female ALS+FTI caregivers. Participants were recruited through an ALS clinic at a medical facility in central Pennsylvania. The ALS clinic serves a predominantly Caucasian population and offers a multidisciplinary approach to patient care. The average clinic appointment lasts approximately 2 to 3 hours.

Recruitment. Caregivers whose partners fit this study's criteria were initially contacted by letter to ask about interest in participating in the study. Follow-up calls were made by the researcher, and the clinic nurse also e-mailed participants to assess interest. Informed consent was obtained from caregivers, and consenting participants were interviewed by telephone.

Inclusion criteria. In order to be eligible for participation in the study, caregivers were required to be age 18 or older and a lay-caregiver. A lay-caregiver is a family member or friend who is not hired to care for the patient and assists with medical and activities of daily living. Caregivers were also required to identify as the primary lay caregiver for an individual diagnosed with ALS+FTI (i.e., must provide the majority of daily care to the individual on a daily basis). In addition, caregivers were eligible if they were providing care for individuals aged 18 or over who were diagnosed with ALS+FTI according to the following criteria: (a) meets El Escorial criteria for clinically probable or definite ALS, as determined by the treating neurologist, (b) meets criteria for FTI as determined by an ALS-Specific composite score of 77 or below on the Edinburgh Cognitive and Behavioural ALS Screen (ECAS), and (c) The symptoms associated with FTI are considered to be clinically significant, as determined by the treating neurologist.

Exclusion criteria. Caregivers who were unable to speak and read English and/or were unable or unwilling to provide informed consent were excluded from participation in this study. In addition, caregivers of patients who were unable to

complete the ECAS were excluded. Patients were excluded if they were unable to speak English.

Materials

Personal information questionnaire: Caregiver. Each participant completed a questionnaire designed by the researcher to collect demographic data and other information about the caregiver. A wide range of data was collected, including the caregiver's gender, age, race, household income, relationship to ALS patient, number of years providing care to patient, patient descriptives, whether there was an aide in the home, type of equipment and/or respite care utilized, etc. The questionnaire took approximately 5 minutes to complete.

Caregiver interview. A semi-structured interview designed by the researcher was used to gather information about the caregiver's subjective experiences while caring for an ALS patient with cognitive and/or behavioral impairment. More specifically, the caregiver was asked to describe in her own words her thoughts, feelings, behaviors, practical daily challenges, and rewards in caring for a person with ALS+FTI. The semi-structured interview included open-ended questions to facilitate discussion and gather information about the constructs of interest, while also allowing caregivers the opportunity to respond without constraint. In the event that insufficient information was provided, questions were followed up by prompts to increase the chances that information about the constructs was elicited. Therefore, if the caregiver offered insufficient information, the interviewer prompted the caregiver to encourage a detailed response (see Appendix B).

ALS Functional Rating Scale – Revised (ALSFRS-R). The ALSFRS-R is a 12-item clinician-administered instrument designed to assess activities of daily living, motor function, respiratory function, and bulbar symptoms in patients with ALS (Brooks et al., 1996; Cedarbaum et al., 1999). Each item is rated individually on a 5-point Likert scale, with 0 representing an inability to perform the specific action and 4 indicating normal ability. A total score is derived from the sum of the ratings, with lower total scores corresponding to more severe impairment. This instrument can be used to monitor response to treatment or disease progression and takes approximately 10 minutes to administer.

Edinburgh Cognitive and Behavioural ALS Screen (ECAS). The ECAS is a brief multidimensional screener designed specifically for ALS patients (Abrahams, Newton, Niven, Foley, & Bak, 2014). It assesses the following domains: executive functions (e.g., digit span backward, alternation), fluency (e.g., verbal fluency, restricted verbal fluency), language (e.g., naming, spelling), memory (e.g., immediate recall, delayed recall, recognition), and visuospatial functioning (e.g., dot counting, number letter location). The ECAS is designed to be administered by healthcare professionals and takes approximately 15 to 20 minutes to complete. The ECAS yields an ALS-Specific score (derived from the total score of executive function, language, and fluency domains), an ALS Non-Specific score (derived from the total score of memory and visuospatial functioning domains), and an ECAS Total score (combined total score across all domains). Each domain and total score has a specific cut-off score for abnormality, which is based on values that are two or more standard deviations below the mean score. The ALS Non-Specific score was designed to differentiate ALS-specific cognitive and

behavioral impairments from the impairments commonly associated with other disease processes, such as Alzheimer's disease.

Edinburgh Cognitive and Behavioural ALS Screen (ECAS) Carer Interview.

The ECAS Carer Interview is a 10-item checklist that assesses for the presence of behavioral change in the patients across five domains: behavioral disinhibition, apathy, loss of sympathy/empathy, perseverative/stereotyped behavior, hyperorality/altering eating behavior (Abrahams et al., 2014). Each endorsed symptom is scored as a 1, with a maximum total score of 10. In addition, caregivers are asked to respond to three questions that assess for the presence of psychotic symptoms. Each endorsed symptom is scored as a 1, with a maximum total score of 3. All questions are based on recent diagnostic guidelines for FTD. The presence of behavioral change in three or more domains indicates the possible presence of the behavioral variant of FTD. Completion of this measure takes approximately 5 minutes.

World Health Organization Quality of Life – Brief Form (WHOQOL-BREF). The WHOQOL-BREF is a 26-item self-report instrument that assesses subjective QOL in four domains: physical health (e.g., energy, pain), psychological health (e.g., negative/positive feelings, spirituality, personal beliefs), social relationships (e.g., personal relationships, social support), and environment (e.g., financial strain, recreation, access to healthcare; WHO, 1998). Each item is rated on a five-point Likert scale. Individual scores for each domain are calculated and converted to transformed scores ranging from 0 to 100, with higher scores representing higher QOL. The instrument also yields overall health and overall QOL scores. Completion time ranges from 5 to 10 minutes.

Level of Care Index. The Level of Care Index is a 3-item self-report instrument that measures the total number of hours of care provided per week and the types of care provided by the caregiver (V. Murphy, Felgoise, Walsh, & Simmons, 2009). The Level of Care Index assesses the number of hours spent assisting with direct tasks (e.g., dressing, bathing, feeding, toileting) as well as indirect tasks (e.g., grocery shopping, transportation, meal preparation, financial planning). Scores on this measure range from 1 to 13, with a combined total score of 13 representing the greatest amount of care provided by caregivers. The Level of Care Index takes less than 5 minutes to complete.

Procedure

The clinic neurologist, nurses, and staff identified eligible caregivers and patients. Once identified, the clinic staff contacted potential participants by e-mail and letter, and the researcher followed up on these contacts with telephone calls to assess interest and provide caregivers with a description of the study, requirements for participation, and potential risks and benefits of participating in the study. Individuals who were willing and able to participate scheduled a convenient date and time to speak over the telephone. At that time, this researcher reviewed the informed consent form with the caregiver and addressed her questions before obtaining consent from the caregiver over the telephone. A telephone consent form was filled out for each participant. Once the caregiver consented, she was interviewed by the researcher using a semi-structured interview format (see Appendix B). Interviews times ranged from 20 to 45 minutes. All interviews were recorded on two digital recorders. The researcher recorded field notes and personal reactions before, during, and after the interview.

Upon conclusion of the interview, the caregiver was asked to complete a personal information questionnaire, an index of objective demands of caregiving, the ECAS Carer Interview, and a QOL questionnaire. These questionnaires were sent by either e-mail or mail, depending on each caregiver's preference. The rationale for administering these measures was to obtain information regarding the demands of the current caregiving situation, the level of behavioral change noted in the patient, and each participant's subjective report of QOL.

All caregivers were debriefed immediately after the interview to minimize the potential for residual psychological distress following the interview. All participants were provided with contact information for staff at the ALS clinic in the event that further debriefing or assistance was required. All interviews were transcribed verbatim by the researcher within 2 weeks of the interview.

Data Analysis

The grounded theory method of qualitative analysis was utilized to analyze the data that emerged from the caregiver interviews. Grounded theory is an inductive process in which theory is generated systematically from raw data (Green & Thorogood, 2013). Thus, grounded theory research does not rely on a specific theory to guide the data collection procedures; instead, the theory is generated from the data (Green & Thorogood, 2013). In order to provide meaning to the caregiving experience, grounded theory was used to increase understanding of the unique experience of each caregiver and to identify connections in their experiences.

As suggested by Strauss and Corbin (1998), the data were analyzed using three coding procedures: open, axial, and selective coding. Open coding refers to the analytic

process of breaking down the data into key concepts and categories (Strauss & Corbin, 1998). The axial coding process consists of identifying the relationships between key concepts or categories and connecting the categories to subcategories (Strauss & Corbin, 1998). Lastly, a selective coding process was used to identify core variables and categories that were present within the data (Strauss & Corbin, 1998). As such, this method was selected in hopes of identifying the commonalities, or themes, that emerged across ALS+FTI caregivers.

To increase the validity of the data analysis, a validation team consisting of three doctoral level psychology students (including the researcher) was formed to achieve triangulation. Each team member was provided with copies of the transcripts and interview questions. Each coder reviewed and coded the transcripts individually. Subsequently, a telephone conference was scheduled to discuss, collaborate, and validate the findings from the transcripts. Field notes were recorded to document each coder's strategies, reactions, and thoughts throughout the coding process. Once all transcripts were coded, the researcher generated themes from the data. Finally, the team arranged a final feedback teleconference to compare, contrast, and review the themes and patterns that were generated from the data as a whole.

Descriptive Statistics

The frequency, means, and ranges for the data gathered from the demographics questionnaires were calculated for each category. In addition, the frequency and mode scores for the instruments completed by the caregivers were calculated. Each caregiver's responses on the WHOQOL-BREF were scored and interpreted based on the normative data provided in the manual (WHO, 1998).

Chapter 5: Results

Descriptive Statistics: Demographics and Objective Measures

Three caregivers completed the personal information questionnaire and objective measures. One caregiver did not return her packet. Of the three caregivers who completed the personal information questionnaire, all were married, Caucasian females who were providing care for spouses. The mean age was 60.7 (range 44 to 70). Two caregivers were retired and one was on leave from work. Additionally, two caregivers reported having a bachelor's degree and the third received her high school diploma. The average length of time as a caregiver was 2.7 years (range 2 to 4 years). Descriptive statistics for the objective measures are summarized in Tables 2, 3, and 4 in Appendix A.

Qualitative Data Analysis: Interviews

All interviews were conducted over the telephone due to geographic constraints. To protect the identities of participants, the interview data were deidentified and each caregiver was assigned a pseudonym. Using grounded theory, three major themes were identified across caregiver narratives: witnessing the effects of ALS+FTI, the impact of the caregiving experience, and coping with life as a caregiver. There were also commonalities within each theme; therefore, several subthemes were identified and incorporated into the presentation of results.

Witnessing the effects of ALS+FTI. The distress associated with observing the unremitting physical deterioration of one's spouse was a prominent theme across caregivers in this study. The emotional pain and struggle inherent in this experience was portrayed clearly in each caregiver's narrative. For example, Mrs. A. shared the following experience:

When I see my husband get a little worse, then I see myself get more and more depressed . . . That's my main thing. I will watch him fall asleep on the sofa [and] it's unbelievable how his legs don't stay still. Oh, it's horrible! Sometimes I'll see him in the fetal position [because] his legs hurt and he's trying to get comfortable . . . It sucks.

Similarly, while discussing the physical demands of caregiving, Mrs. D. stated, "Watching my husband lose so much . . . Emotionally, it is harder for me than the physical part to be honest. The physical part I can deal with, but watching him lose something daily is harder." Seventy-five percent of caregivers specifically mentioned the patients' increased need for sleep as a prominent symptom of ALS. Some caregivers spoke about how their spouses were forced to abandon pieces of their identities as their physical functioning declined. For instance, Mrs. C. explained her husband's shifting identity concurrent with physical changes:

I've seen him go from someone [who] was an athlete in high school and his whole life has been about athletics, and in college he was an athlete . . . he used to have really big calf muscles and now he doesn't. That has really impacted me and our daughters.

The difficulty in watching one's spouse's physical deterioration was compounded by the onset of FTI symptoms. Caregivers described a range of symptoms that were consistent with the cognitive and/or behavioral variants of FTI. Specifically, Mrs. A. reported strictly cognitive symptoms, Mrs. B. and D. described symptoms consistent with the behavioral variant, and Mrs. C. reported cognitive and behavioral symptoms. Notably, Mrs. C. and D.'s descriptions of their spouses' symptoms were characteristic of

individuals with FTD; however, since their spouses had not been recently evaluated, the diagnosis was unable to be confirmed. Nevertheless, both caregivers noted a progression of symptoms since the last assessment.

Frontotemporal cognitive impairment. Mrs. A. reported her spouse experienced memory loss and confusion:

His memory is going pretty bad now . . . The other day he tried to start my car with his car keys and he yelled, ‘Honey, honey, honey, come here! There’s something wrong with your car!’ and here he had his car keys . . . He is not turning the oven off, like the burners . . . He even told me one time he got confused driving [and] he was by himself.

Mrs. A. stated her spouse often had trouble keeping track of the date and was also forgetful in terms of entering a room and forgetting what he was looking for, as well as leaving clothing in the laundry for extended periods of time. Nevertheless, she noted her spouse was aware of this cognitive decline, which made it easier for her because he did not become frustrated when she provided corrective feedback (i.e., “He doesn’t yell at me or anything. If you said, ‘Turn the burner off’ [to other guys], they would probably say, ‘F you’ or ‘Do it yourself.’ [Mr. A.] is not that way. He really isn’t”).

Mrs. C. also noticed cognitive changes, particularly confusion and disorientation. She recalled, “One morning last week, he said, ‘Where am I? I know I am not in my room,’ and I said, ‘Yes, [Mr. C.]. You’re in your room,’ and he said, ‘Nope, I was out in the driveway.” She added her spouse’s condition vacillates between periods of disorientation/confusion and lucidity.

Frontotemporal behavioral impairment. Mrs. B. and D. discussed their struggles with their spouses' lack of concern for their well-being despite being significantly overwhelmed and consumed by caregiving responsibilities. Mrs. B. described this difficulty:

He knows that I sit around with hot water bottles and pain patches all the time, but he never addresses knowing that I'm having problems or hurting. In all honesty, I don't think he understands how hard it is for me or how difficult he makes things for me. He just doesn't seem to care about the impact on me.

Likewise, Mrs. D. shared a similar experience:

He would get angry with me and was not empathic toward me. He would say to me things like, 'You're not sick,' or 'Don't complain.' He wouldn't allow me to say anything that was sad in my life or he would get angry. That's the first thing I noticed.

Mrs. B. and D. also reported their spouses became increasingly self-centered, demanding, and ungrateful, which made the experience particularly difficult. Mrs. D. struggled greatly with her spouse's lack of gratitude:

I think it's always easier when somebody is happy with you; it's easier to take care of them . . . I feel very horrible talking about him in this manner because he was an amazing person, but since the disease did this to him, he isn't grateful. I can't ever do enough to please him and I am killing myself basically. I am working so hard and it is a difficult task when you are doing everything you can and you're exhausted and you go to bed and you're like . . . there is no gratitude. There is no gratitude there.

Mrs. B. discussed similar challenges. She indicated she hired a part-time caregiver to help her manage Mr. B.'s care, but Mr. B. would not allow the caregiver to assist him:

[Mr. B.] was quite demanding and insisted that I do everything, and I mean everything, like the caregiver would see it because [Mr. B.] wouldn't let him do anything, but as soon as I walked in the door, he wanted everything done . . . His expression to me was, 'That's what you're here for.' . . . I'm trying not to force anything on him that he's not ready to accept, like he still wants to use the board to slide in and out of bed, but it would be easier for me if we use the sling. I'm probably at the point where I'm going to tell him that's it, whether you like it or not, that's it, because it's very hard on me . . . very hard on me physically. He can't see that this is difficult for me too or that I am tired. All he thinks about is what he is going through.

Both Mrs. B. and D. observed changes in their spouses' temperaments. They noted challenges navigating their spouses' anger and irritability. Mrs. B. stated that Mr. B. "can have a temper and I figured if I crossed him then it would make things harder . . . I just didn't want to add to it." Mrs. D. explained, "My husband also loses patience if he can't get something or have something his way so he takes his fists like a kid and slams them on the table."

Another symptom reported by caregivers was a lost interest in relationships and tendency to become withdrawn from others. Mrs. D. described this element of the disease:

It has also withdrawn him, extremely, from all of us . . . He does not pay attention to us . . . He does not interact with the family, particularly in the last 6 months.

He sits with his headphones on. He sits in the room with us, but he has his headphones on in his own world and it is hurtful to everybody.

Two caregivers, Mrs. C. and D., reported their spouses exhibited symptoms of behavioral disinhibition in terms of hypersexuality and making inappropriate comments to others. In the case of Mrs. C., her spouse's sexual proposition toward a young female turned into a legal matter in which charges were filed against Mr. C. She noted, "It has just been devastating to our family and it's just been a nightmare and it has not been resolved." She explained, "I just accepted that he is going to say some things and do some things that are different," and added that her spouse has also made inappropriate racial comments. Likewise, Mrs. D. reported she suspected something was wrong with her spouse when professional caregivers refused to return to their home. She stated she was unaware of what was happening until "Finally, finally, the agency told me that my spouse was making passes at these women." She had been informed that her spouse was making inappropriate comments and engaging in sexual behaviors while in the presence of the caregivers. Mrs. D. added, "On top of that, he had a money thing where he wanted money . . . He wanted cash because he wanted to offer [the caregivers] money." Mr. D. also became "obsessed" with watching pornography. Mrs. D. explained, "It was confusing to me because he knew [what] he was doing wrong, but he couldn't stop doing it so it was pretty awful. It was pretty awful."

Notably, neither Mrs. C. nor Mrs. D. recognized their spouses' behaviors as symptoms of FTI. They described confusion and disbelief regarding these behavioral changes. In both caregivers, their spouses' sexual misconduct prompted them to consult

with medical providers at the ALS clinic, who provided them with education about FTI.

Mrs. D. described this revelation:

I didn't really understand it at first because I thought it was more of: he is trapped in this body and he is angry . . . I wish I would have known what to look out for earlier so we could have discussed options or medication sooner. I didn't know what to expect.

Across all clinical presentations, caregivers discussed how they endured the loss of the very essence of their loved ones with the progression of FTI. Caregivers indicated this aspect of their spouses' conditions was the most difficult to witness, more so than the physical losses, which may be unique to ALS+FTI caregivers. Mrs. D. shared her experience with the emotional loss:

My husband suffers from dementia and that is emotionally the worst. [That is] the hardest thing for me because he is not himself. He was an awesome, cool guy. People loved my husband. He had a great personality and was such a fighter . . . You're not only losing the person, you're losing the person you love. It has been watching the man I love—I don't know him anymore. You lose that person even before they die and that's a huge challenge.

The impact of the caregiving experience. All carers' narratives conveyed the intensity of the caregiving experience by offering a snapshot of the widespread impact and daily challenges they faced. Within this theme, several subthemes were identified: progression of the disease, physical challenges, psychological struggles, social functioning, and practical challenges.

Disease progression. Caregivers in this study frequently mentioned disease progression within the context of their caregiving responsibilities. They noted that as their spouses' diseases progressed, the intensity of the experience increased. Although some caregivers reported very few responsibilities (i.e., "I help him get out of bed, [but] other than that, he can do everything himself"), others described themselves as "consumed" by their caregiving tasks (i.e., "I am 100% consumed. I don't have a life at this point. The ALS and [my spouse] consume me . . ."). Numerous tasks were listed as caregivers recounted their daily routines. For instance, Mrs. C. stated, "My husband cannot do anything for himself. He can't stand, he can't walk, he can't dress [or shower] himself, he is incontinent, he can't feed himself [so] I do all that. I shave him, I brush his teeth; everything, I do." Caregivers indicated the experience also intensified with the progression of FTI because there was an increased need to monitor and supervise their spouses. Mrs. A. offered, "I have to watch him more, especially with the oven . . . It changes the way you live. The way of life." Likewise, Mrs. C. explained a similar experience with monitoring her husband:

When a friend would come and visit, I used to go off in another room, but now I stay in the room and I listen to make sure of what he says . . . Now, I am much more careful . . . I would never leave the room.

Caregivers in this study also discussed the practical challenges associated with disease progression. With more advanced levels of disease, new challenges arose that rendered familiar ways of completing tasks and assisting their loved ones ineffective. As such, caregivers had to determine whether new medical equipment was necessary and

whether modifications to the environment were required to meet their spouses' needs.

Mrs. C.'s statement captures this experience:

Unfortunately, our bedroom is on the second floor of our house. It became a struggle for him to get up the stairs and so then we got a stair lift, which he was able to use for a certain amount of months, and then it got to where I was not able to get him on the stair lift. It was getting dangerous and he fell one time with just me trying to get him on the stair lift and so then we moved his bedroom down to our living room, which of course made it easier, but then we just had a powder room on the first floor so for months he wasn't able to take a shower. I would just sponge bathe him [so] we had a room built on the back of our home and we had a shower built on the back of the powder room . . . All of these things progressed where, at first, all I had to do was make sure he didn't fall and could get into his regular wheelchair, and then . . . get on the portable toilet and everything, [but] then it got to where he had to use a board to go from his bed to the toilet or from his bed to the wheelchair, and then we got a power wheelchair, [but then] it got to where we couldn't do it with the board so now I use the Hoyer lift for everything.

Relatedly, Mrs. B. shared how her physical surroundings became inundated with equipment over the course of her spouse's disease: "Here's what we have: he has [a] \$50,000 power chair, we have ramps out of the house [and] ramps into the garage; we have a wheelchair van, we have a hospital bed, we have stair lifts, [and] a walk-in shower . . ." Caregivers in this study conveyed that ALS is a disease involving continuous change and adjustment. One can see how managing these daily challenges can have an impact on a caregiver's health and well-being.

Physical challenges. Most caregivers in this study mentioned the physical demands and challenges they faced while providing care to their loved ones. A common subtheme expressed by participants was the weight differential between caregiver and patient in terms of the patient weighing significantly more than the caregiver. Caregivers reported significant physical difficulties transferring and positioning their spouses, which was extremely taxing on the caregivers' bodies. Mrs. B. described this challenge:

I'm having increased discomfort with my hip and my leg on my right side because I'm tugging and he weighs about 220 pounds and I weigh 140 so he's heavy . . . I have to lift his legs in the bed and lift them out of the bed . . . [It's] very hard on me physically.

Notably, Mrs. B. added, "I'm a retired RN so I have the knowledge and know exactly everything to do; however, to a point that doesn't help anything. You still have the lifting and maneuvering . . ." Mrs. C. and D. shared similar experiences, respectively:

The most difficult for me to manage is not having the physical strength to do things . . . [He] weighs 212 pounds and I weigh about 120 pounds so it's becoming more and more difficult for me. It used to be that he could help me. I mean, I could help him turn over and he could give me some help . . . Now, we can't do that. I just don't have the power to turn him over.

We were adding a full bathroom [to our first floor] so for a period of a few months, I had to help him up and down the stairs, and physically, my back would hurt, my knees would hurt, my feet would hurt. I injured my foot . . . It is really tough on the body—lifting him in and out, there is definitely a physical aspect

there. I'm semi-fit, but I am not a huge woman. He is 175 pounds and I definitely hurt.

In addition to physical pain, all caregivers in this study reported feeling physically tired or exhausted. Two out of four participants reported developing new medical conditions and/or health concerns following the onset of caregiving, such as migraines, weight gain, and carpal tunnel syndrome. Others indicated that preexisting health conditions (e.g., arthritis) were exacerbated by their caregiving duties.

Psychological struggles. Results of this study revealed that psychological struggles were reported consistently across all narratives. Caregivers unanimously described the experience as “difficult” and noted increased frustration and decreased patience. Mrs. A. stated, “We have been having our moments where I get snappy or he gets snappy . . . I'm snapping and I never snap, which isn't good. It gets to everything.” Mrs. C. also discussed psychological struggles:

It's a very difficult thing . . . Sometimes I get very frustrated . . . Sometimes I get impatient . . . everything is new and I am not good with change and I've had to change so much. We have had to change our home and I've had to change everything and some things I haven't dealt with as well as I would have liked.

Caregivers also reported feeling depressed (100%) and discouraged (75%), which for some, increased over time. Mrs. B. explained, “The first year I endured then I realized I needed somebody to help me cope. [I experienced] crying spells, discouraged, anger, great anxiety at some points.” Most caregivers (75%) indicated they were stressed and overwhelmed. Caregivers also discussed the emotional challenges associated with their spouses' FTI symptoms, describing these experiences as “hurtful” and “maddening.”

Interestingly, when asked directly about the impact of caregiving on their psychological health, caregivers often expressed feeling guilty about their emotional challenges or reactions to the experience, noting that they were not the ones with the ALS diagnosis. Caregivers reported “regret” and made statements such as “I shouldn’t be like that.”

Social functioning. Caregivers in this study stressed that there was little, if any, time to engage in social or leisure activities outside of their caregiving responsibilities. When asked about her social life, Mrs. B. replied, “There isn’t anymore. There just isn’t. I cannot leave Mr. B.” Mrs. D explained, “I basically do not stop from the time I get up until the time I go to bed . . . I don't have time to exercise. I don't have the time to basically do anything for myself . . . I cannot leave him.” Both Mrs. A. and C. mentioned engaging in fewer recreational activities because their spouses were unable to participate. Mrs. C. stated, “We don't have activities like we used to have . . . I never dreamed that our retirement would be like this. I thought we would be traveling and doing things.”

Most caregivers in this study (75%) discussed the challenges of maintaining relationships with friends and families. It appeared these struggles were not only related to a lack of time, but also families’ and friends’ difficulties understanding and appreciating the intensity of the caregiving experience. Loss of friendships were noted across caregivers, including Mrs. D.:

It’s very isolating. Over time, people stop calling. Some people can’t handle it and some people just go away simply because they ask you to do things and you can’t because you can’t get away . . . They stop including you in things because they know you can’t so it is a very lonely place . . . A lot of people don’t know

what to say to me so they avoid me. You learn who your friends are, let me just say that, you do learn.

Mrs. B. shared her challenges:

I haven't spent personal time with my big sister who lives nearby since last summer because I've used the caregiver only to do my necessary errands and then one day of Christmas shopping so . . . Here's a way to put it: If I could have more me time, I think that would be better for him. I wouldn't get as discouraged or feel so overwhelmed, but that is a very difficult thing to do.

Additionally, Mrs. C. reported that her spouse's FTI symptoms contributed to the loss of a relationship with a "very close family friend." Similar to other caregivers, she also mentioned that she did not feel understood by others, but clarified she was able to identify with individuals who were enduring similar challenges:

But the caregiver thing, it's very difficult, very difficult, and like I said, I don't think people understand how difficult it is. I have a few people in my circle of friends that do understand; my best friend is going through a lot with her husband . . . very similar to what I have.

Interestingly, many caregivers shared their struggles in soliciting help and support from friends and families, which contributed to their feelings of isolation and loneliness.

Unanswered calls for help. Caregivers discussed that when they attempted to reach out to friends and families for support or assistance with providing care for their loved ones, people were often unwilling or unavailable. For instance, Mrs. C. indicated this was a particularly frustrating and surprising aspect of her experience because she had

been assured by her spouse's providers that friends and family would likely assist with caregiving tasks. She noted:

Several times they made the expression, "When friends and family find out what's going on, they're just going to jump on the bandwagon and pitch in with [Mr. C.]," and I'll be honest, that's not true. They don't . . . Even when I've asked someone to fill in here once in a while so I can have more than 1 hour to run an errand or something, I got all kinds of unbelievable excuses with no compassion whatsoever and that was hard because now I have to live with their reactions and it's hard to do.

Mrs. D. also indicated she received minimal assistance from her spouse's family:

[My] family doesn't live in the area so it's basically me doing everything.

Occasionally, my husband's family comes up for a few hours a day and relieves me for 2 nights (*laughs*). I got 2 nights of vacation for a year, but they stayed with him 2 nights the entire year . . . So you become bitter.

She added that even when she reached out to family or friends for emotional support, they were not always willing or able to do so. She explained, "I wish more people would let me talk, but people don't have time for me . . . I would call my sister-in-law and she would say, 'I just don't have time. I'll call you later,' and they don't."

The difficulties in obtaining assistance extended beyond friends and families.

Indeed, all caregivers repeatedly discussed their struggles obtaining professional help and resources from healthcare agencies due to limited availability, poor quality, or lack of financial resources. Mrs. B. highlighted this:

It took me, believe it or not, over 6 months to get 11 names of caregiving agencies and of those 11 there were only two possibilities and that's difficult . . . [The ALS clinic says] that there's a lot of groups out there, but the groups say, 'We don't have enough staff, we can't take more on' or 'We have a 20-pound weight limit.' . . . I have thousands of hours on the phone and on the internet . . . I've talked to all the government agencies in the area [but] many of them won't even respond to you. You have to beg and plead for everything you get. It's sad, very sad. The help should be more easily obtained, but it's not. It's very discouraging.

She added that even when she was able to find professional caregivers who were available, they were not able to assist with many tasks. She stated, "They just sit there and are available, but they don't really do anything. It's very difficult." Mrs. C. also expressed frustration regarding this process, noting that she had contacted an agency that promised to send a part-time caregiver, but did not communicate when these services would begin. Moreover, caregivers pointed out the expenses associated with hiring professional caregivers and explained that even though they recognized they needed more help, they were unable to afford it. Mrs. D. stated, "I think I am doing too much . . . It's very expensive, that's the problem." In the absence of support from families, friends, and professional agencies, caregivers stated they had no choice but to "do it all" themselves. Often, this meant caregivers had to relinquish roles and attempt to balance their responsibilities outside of their caregiving tasks.

Balancing other roles and responsibilities. Caregivers spoke about their difficulties managing other roles and responsibilities outside of their caregiving demands. For instance, Mrs. B. reported she was so consumed by her role as a caregiver that she

unintentionally neglected other responsibilities, such as housekeeping and bill paying. Some caregivers were forced to give up their careers, whereas others explained they were unable to dedicate as much time to their roles as mothers. For example, Mrs. D. asserted, “I have to give so much to my husband that [my children] kind of have to take second place, sometimes out of necessity, because I am stretched extremely thin.” She also explained that she was forced to relinquish her role as a wife to fulfill her role as a caregiver. She expressed, “You become less of a wife and more of just a caregiver . . . I would say that’s something you can add that is really difficult.” Mrs. C. shared how performing certain caregiving tasks (e.g., toileting) was particularly unpleasant for both her and her spouse, albeit necessary, as she was the only person available to assist him. Similarly, Mrs. B. indicated that her interactions with her spouse revolved around caregiving tasks and offered, “We don’t talk anymore . . . this hideous disease does cut into the partnership, it really does.”

All caregivers in this study discussed the devastation and widespread impact of ALS on their physical, psychological, and social functioning. They described the practical challenges faced on a daily basis, as well as the need to adapt and adjust constantly to the progression of the disease. In providing an account of their experiences, caregivers spoke about the various coping strategies they implemented to sustain themselves throughout their journey as caregivers.

Coping with life as a caregiver. Most caregivers struggled to cope with the impact of the caregiving experience. As Mrs. D. stated, “Mentally, trying to cope with all this, it throws me for a loop. That is the most difficult part for me.” Despite their

challenges, caregivers identified strategies to cope, including seeking mental health treatment and relying on their spiritual or religious beliefs.

Mental health treatment. Two caregivers (50%) sought community mental health services to cope with the stressors and emotional consequences of caregiving. Both caregivers indicated they were being treated with antidepressants, whereas only one of them was involved in counseling. For the caregiver who participated in counseling, she explained she wished she had sought psychological services sooner because she “waited too long” and was under significant psychological stress at the onset of therapy. Nevertheless, she reported the counseling sessions helped her to engage in self-care and manage her stress.

Religion/spirituality. Two caregivers (50%) relied on their faith and spiritual beliefs to sustain them, including Mrs. C.:

I will tell you I could not get through this without my church and my faith, and I believe that God is helping me through this and if I did not have that, I don't know how I would get through it. I rely on prayer a lot. I read the Bible. I do devotionals a lot . . . My faith has gotten stronger. I've turned everything over to the Lord . . . I know the Lord is going to get us through this and so that's how I do it. I have turned it over to the Lord.

Likewise, when Mrs. B. shared how she has coped with her role as caregiver:

I would say it's my spiritual time. I read the Bible and spiritual books every morning and I have several people that I listen to [on the internet]. It would be my spiritual background that probably keeps me going the most.

Summary

As emphasized by all caregivers in this study, the subjective experience of each ALS+FTI caregiver is different. Nevertheless, this study's findings revealed that although every caregiver faced her own set of unique challenges, there were commonalities across the caregivers' narratives. These themes were identified as witnessing the effects of ALS+FTI, the impact of the caregiving experience, and coping with life as a caregiver. The following section focuses on synthesizing and interpreting these findings within the context of the existing literature.

Chapter 6: Discussion

The purpose of this study was to explore and describe lay-caregivers' thoughts, feelings, behaviors, practical daily challenges, and rewards in caring for persons with ALS+FTI, as well as to offer suggestions for how to assess FTI within the ALS population. More specifically, this study sought to provide preliminary answers to the following questions: (a) What are the thoughts, feelings, and behaviors of lay-caregivers who provide care for persons with concurrent ALS and FTI? (b) What are the practical daily challenges and rewards in caring for persons with ALS+FTI? (c) What instruments can be used to assess FTI in the ALS population? A total of four ALS+FTI caregivers participated in this study; three prominent themes and several subthemes were identified within their narratives.

Analysis of the caregivers' narratives revealed similarities in their subjective experiences and thoughts about their roles as ALS+FTI caregivers. All participants described caregiving as "difficult" and frequently discussed the losses they witnessed and experienced while fulfilling their roles as caregivers. Numerous losses were mentioned, including the loss of their identities as working professionals and/or wives, loss of relationships, and loss of their loved ones in terms of their "essence" (physically and cognitively). Regarding the loss of the person, ALS+FTI caregivers reported experiences that were consistent with the findings of a recent study conducted by Galvin and colleagues (2016), in which ALS caregivers reported losing "the person they knew and the life they may have shared together" (p. 5). This appeared to be a particularly salient aspect of the experience for ALS+FTI caregivers in this study. Participants stated it was more difficult for them to cope with their loved ones' cognitive and behavioral

impairments than any other aspect of the disease. It was also notable that while describing these experiences and losses, caregivers often spoke about their husbands using the past tense, even though the care recipients were still alive. This was a common theme noted throughout many of the transcripts.

Caregivers also appeared to be confused by the initial emergence of behavioral symptoms and personality changes because they were unaware of, or did not fully understand, the symptoms of FTI or how they would manifest in their loved ones. This lack of recognition or understanding of FTI symptoms often contributes to delays in diagnosis and the implementation of behavioral management strategies, which has been discussed in the FTI caregiving literature (Pressman & B. L. Miller, 2013), and is consistent with the experiences of the participants in the current study. For at least two participants in this study, the FTI diagnosis appeared to offer partial relief because they were provided with an explanation for their spouses' personality and behavioral changes, as well as strategies to manage the problematic behaviors (e.g., psychotropic medication to address hypersexuality).

Caregivers also demonstrated commonalities in their feelings about the experience, both psychologically and physically. In terms of their psychological experiences, all caregivers endorsed depression or depressive symptoms and most indicated they felt overwhelmed, consumed, and distressed by their responsibilities. Caregivers also reported feelings of frustration and anger, particularly in regard to the patients' FTI symptoms, such as lack of gratitude, selfishness, demandingness, and abnormal behaviors. This finding is consistent with the available literature on ALS+FTI caregiving experiences, which indicates neurobehavioral symptoms are related to

caregiver burden and psychological distress (Andrews et al., 2016; Chio et al., 2010; Galvin et al., 2016; Lillo, Garcin, Homberger, Bak, & Hodges, 2010). Many caregivers reported handling the caregiving responsibilities “alone,” which was associated with feelings of loneliness and resentment toward family members who were often perceived as unwilling to help. Participants expressed that they did not feel understood by friends and family members, which appeared to contribute to feelings of isolation. Caregivers reported similar experiences with professional services, and commented on the immense amount of time they invested in identifying and securing these resources. Physically, the demands of caregiving proved to be painful and taxing on caregivers’ bodies. Caregivers reported difficulty sleeping, as well as feeling tired, exhausted, and drained.

Behaviorally, caregivers reported the demands of caregiving increased with the progression of the disease, whereby more advanced stages placed greater pressure on each caregiver. Like caregivers of patients with physical illnesses (Arango-Lasprilla et al., 2011), ALS+FTI caregivers assisted with a range of activities, from helping their loved ones out of bed to transferring, bathing, and feeding; however, ALS+FTI caregivers also reported that the emergence of FTI symptoms required them to provide more direct supervision to their loved ones, which is reported commonly in the psychiatric caregiving population (Diehl-Schmid et al., 2013). As such, the experiences of ALS+FTI caregivers appear consistent with the literature on caregivers of patients with both physical and psychiatric symptoms (Andrews et al., 2016; Reinhard et al., 2014). Caregivers also discussed the continuous need to reevaluate and adapt their approaches to caregiving, as well as their physical environment based on their loved ones’ disease progression. Participants communicated that as they dedicated more time to their

caregiving responsibilities, they faced isolation and disengagement from social activities and relationships. As such, they described having little time to engage in leisure activities or take care of their personal needs.

Most caregivers identified coping with the complexities of caregiving as challenging. Some caregivers relied on their faith and/or religion to sustain them, whereas others sought treatment with antidepressants and/or outpatient mental health counseling. One caregiver reported that social support, particularly from a trusted companion, was particularly helpful because this provided an outlet for her to express her frustration and struggles as a caregiver. As such, caregivers employed different methods to cope with their experiences.

A range of practical daily challenges were mentioned across caregivers in this study. Notably, in the advanced stages of disease, caregivers explained how the physical demands exceeded their physical abilities and posed significant challenges for them on a daily basis. This concern has been documented in the literature, indicating this is a common challenge faced by ALS caregivers (Galvin et al., 2016). Participants also commented on the difficulties associated with identifying and receiving assistance from professional caregiving agencies or other community resources (e.g., equipment suppliers). These accessibility challenges were compounded by financial limitations, whereby caregivers acknowledged the need for additional help but were unable to afford the cost of these services. As such, caregivers in this study were tasked with the competing demands of performing their caregiving duties while also tending to their other responsibilities, such as childcare and bill paying, with minimal support.

Interestingly, when asked about the rewarding aspects of caregiving, most caregivers (75%) indicated they did not perceive any aspect of the experience as rewarding. Nevertheless, one caregiver mentioned specifically that her ability to bring her spouse joy or happiness was rewarding, whereas two additional caregivers shared similar sentiments (i.e., importance of making their spouses happy) throughout their interviews, but did not specifically identify this as rewarding. Similarly, half of this study's sample denied any type of personal growth from the experience. Regarding the caregivers who reported personal growth, one caregiver described feeling emotionally stronger (i.e., "a stronger, more caring person") and the other noted religious/spiritual growth.

Limitations and Directions for Future Research

One of the primary limitations of this study pertains to the small sample size and homogeneity of the participants, which limits the generalizability of the findings. There was little to no variance in geographical location, race, ethnicity, or socioeconomic status, as participants were predominantly Caucasian and all were recruited from the same multidisciplinary clinic in central Pennsylvania. Factors such as age, education, race, culture, and socioeconomic status may influence caregiving experiences; therefore, future studies designed to address the experiences of ALS+FTI caregivers should employ sampling strategies that increase the diversity and representativeness of their samples. In addition, participants in this study participated on a voluntary basis, which may have contributed to the homogeneity of the sample due to the self-selection process. For example, caregivers who were particularly stressed and/or burdened may have opted out of the interview.

Additionally, all data collected from this study were self-reported and may have been affected by factors such as social desirability or defensive reactions. For example, caregivers may not have been willing to admit certain negative feelings or experiences to the researcher during the interview, particularly FTI symptoms, which have been associated with caregiver embarrassment and shame within the literature. Moreover, the interviews were conducted over the telephone, which increases the likelihood that the patient may have been nearby while the caregiver responded to questions. As such, the results of this study should be interpreted with this information in mind.

As is true with all qualitative research, there is an issue of researcher bias or expectancy effects. In this regard, data analyses are subject to mistakes and bias, especially when considering the potential for the researcher's understanding of the literature to unintentionally shape the way in which analyses are performed. Although efforts were made to increase the current study's credibility and validity with a validation team, these methods may not eliminate bias entirely.

The current study may have also been limited by the lack of objective neuropsychological data to document the cognitive and/or behavioral impairments of care recipients. Nevertheless, since the aim of this study was to explore the caregiving experiences of ALS+FTI caregivers rather than evaluate FTI objectively, the screening instruments employed in this study were deemed sufficient to provide evidence for the presence of FTI in care recipients. Future studies may include neuropsychological data to determine the degree and severity of FTI in the care recipient and enhance the participant selection process.

Recommendations for the Assessment of FTI in ALS Patients

Although the caregiver sample was not secured within the allotted timeframe as proposed for this study, it is notable that recruitment challenges appeared to be related to difficulties identifying patients who met this study's inclusion criteria for FTI.

Consistent with this challenge, the literature stresses the need to develop a battery of tests that can be administered flexibly to ALS patients to evaluate for the presence, severity, and type of cognitive and/or behavior impairment (Goldstein & Abrahams, 2013; R. G. Miller et al., 2009). As such, there has been increased attention to the development of screening measures for this population; however, not all instruments are considered reliable and valid assessment tools. Therefore, the following discussion will focus on offering recommendations to promote the development of an assessment battery to assist in the evaluation of FTI in ALS patients. The recommendations provided in this review are derived from the empirical literature, as well as from suggestions and requests of caregivers in the current study. This supplemental battery may assist medical professionals and psychologists in identifying FTI earlier within the disease process, which has the potential to contribute to better outcomes for patient care and the experiences of caregivers.

Rationale. Although cognitive and/or behavioral impairments are prominent in the ALS population, there is evidence to suggest that cognitive screening is not practiced routinely within specialized ALS clinics (American Academy of Neurology [AAN], 2012). This is concerning, given the impact such symptoms can have on the patient and caregiver. It is possible that the hesitation to evaluate FTI within a healthcare clinic is related to a generalized uncertainty about how to perform such an assessment efficiently

and effectively. Indeed, the literature indicates there is no gold standard approach for assessing FTI in ALS patients, and there is little consensus amongst healthcare professionals about which instruments or screening batteries to administer (R. G. Miller et al., 2009). As such, providers are left with little guidance on how and when to evaluate FTI in their ALS patients.

The standard approach to assess cognitive functioning is neuropsychological assessment (Floris et al., 2012). Neuropsychological assessment is a performance-based method for assessing various cognitive ability areas, including memory, executive functioning, language, attention and concentration, motor functioning, reasoning, problem-solving, and processing speed (Harvey, 2012). Neuropsychologists often perform these assessments using a battery approach, in which several tests are administered to evaluate multiple cognitive ability areas, as well as personality/psychological functioning. The neuropsychological test battery can be individualized and tailored to the problems and needs of the examinee or a standardized battery can be administered in which the same group of tests are administered to all examinees regardless of their presenting problems (Harvey, 2012). An individual's performance on these tests is compared to a normative sample, or reference group, that consists of individuals who are similar to the examinee in age, sex, race, and educational background (Harvey, 2012). These normative comparisons inform the evaluator whether the examinee is performing at a level that would be expected given his or her demographic makeup. Self- and observer-rating scales may also be included in a neuropsychological test battery to evaluate mood, behavior, adaptive functioning, and other disorder-specific symptoms.

Although neuropsychological assessment is the gold standard approach to the assessment of cognitive functioning, standard assessment measures may not be suitable for use within the ALS population (Floris et al., 2012; Goldstein & Abrahams, 2013). In fact, almost all psychological tests have been developed for use with physically intact examinees and many of these tests require a written or verbal response. Due to the speech and motor deficits inherent in ALS, the patient may not have the ability to complete tasks, or if he or she is able to complete them, his or her performance would likely be hindered by these deficits (Goldstein & Abrahams, 2013; Hu et al., 2013). As such, ALS patients may present as more impaired than they are because their performances are compared to those of physically healthy individuals. Moreover, neuropsychological testing is likely too time-consuming to implement in specialty clinics or healthcare settings, and patients may lack the stamina to complete such testing (Hu et al., 2013). In addition, the administration and interpretation of neuropsychological tests must be completed by a clinical neuropsychologist; therefore, routine healthcare providers are unable to administer these test batteries to patients. As such, the provider would have to refer the patient for an evaluation, which can be time-consuming and expensive.

Although it is recognized that it is impractical for ALS clinics to refer every patient for neuropsychological testing, there is consensus on the need to identify cognitive and behavioral impairment in ALS patients as early as possible (Abrahams, 2013; Strong et al., 2009). As an alternative to neuropsychological testing, screening measures designed specifically for use with the ALS population have been developed and published (Abrahams, 2013; Goldstein & Abrahams, 2013). Nevertheless, there is

ongoing debate about whether these measures are sensitive enough to identify cognitive and behavioral impairment in this population. Some studies advocate for the continued use of neuropsychological assessment (Strong et al., 2009), whereas others argue that screening measures are sufficient to diagnose FTI (Abrahams et al., 2000). Given that each approach has strengths and weaknesses, the current study recommends a gradual or hierarchical approach.

Baseline assessment. The prevalence and potential impact of cognitive and behavioral change on the ALS patient's disease process necessitates that healthcare providers monitor patients for changes and/or impairments in functioning within the clinic (Goldstein & Abrahams, 2013). One way to monitor change over time is to evaluate a patient's cognitive functioning at the onset of treatment or soon after the ALS diagnosis is given so that a cognitive baseline can be established. Baseline data act as a benchmark, or reference point, for future comparison. With repeated cognitive assessment, providers can compare recent performance to the patient's baseline data to identify changes in functioning. This is particularly beneficial within the ALS population because, in the absence of baseline data, providers must rely solely on norm-referenced scores, which may increase the risk of false-positive errors in ALS patients without FTI or false-negative errors in high functioning ALS patients with mild FTI (Goldstein & Abrahams, 2013; Harvey, 2012). In other words, rather than comparing the ALS patient's performances to a heterogeneous normative sample, the patient's baseline data are used for comparison purposes. Baseline data are the most reliable sources of comparison for monitoring change in individuals over time. For this reason, this study recommends that ALS clinics administer a brief cognitive assessment to all patients who

present for treatment, including those who do not complain of cognitive or behavioral change. There are a number of screening measures that can be used for this purpose.

Screening batteries. After a baseline assessment is performed, it is recommended that cognitive and behavioral impairment be evaluated at least once annually (AAN, 2013; R. G. Miller et al., 2009); however, given the variability in rate of FTI progression, it is recognized that some patients may require more frequent assessment. For example, studies have identified risk factors for developing FTI symptoms within the ALS population, such as patients with bulbar onset disease, dysarthria, low education, older age, and family history of dementia (Elman & Grossman, 2007). As such, ALS patients with one or more of these risk factors may require evaluation more often, such as every 3 to 6 months.

Given the limitations of neuropsychological testing within this population, the current study asserts that screening measures are appropriate for routine evaluation of cognitive and behavioral functioning in ALS patients. Screening batteries are designed to assess for the presence, severity, and type of cognitive and/or behavioral impairment (Abrahams et al., 2014). Administration time varies depending on the instrument, but requires significantly less time than standard neuropsychological batteries. The results of the cognitive screening determine whether additional testing is warranted (Michels, Tiu, & Graver, 2010). For instance, if there is no indication of significant cognitive or behavioral change, the patient will continue to be screened on an annual basis and no additional testing is required; however, if there are abnormalities identified with the screening tool, a neuropsychological evaluation may be warranted, particularly if the patient and/or caregiver has expressed concern about these matters (Michels et al., 2010).

These recommendations are documented and supported in the literature (AAN, 2013; R. G. Miller et al., 2009).

As previously stated, there is no consensus regarding the best screening tests for impairment in ALS (R. G. Miller et al., 2009); however, screening tools designed specifically for the ALS population have been developed. The literature reveals that some of these tools have been subjected to empirical investigation and deemed valid and reliable for use within this population (Abrahams et al., 2014; Millet et al., 2013).

Edinburgh Cognitive and Behaviour ALS Screen (ECAS). Please refer to the Materials section in Chapter 4 for a description of this measure and the related Carer Interview. The ECAS was designed to minimize the impact of physical disability on performance and to maximize its utility in patients with a range of physical impairments by providing interchangeable tests for patients with speech or motor impairment (Abrahams et al., 2014). In a recent study, the ECAS was validated against a standardized neuropsychological battery to assess its sensitivity and specificity (Pinto-Grau et al., 2017). The neuropsychological test battery included measures such as the Rey Complex Figure Test (Meyers & Meyers, 1996), Boston Naming Test (Kaplan, Goodglass, & Weintraub, 1983), Wechsler Test of Premorbid Functioning (Wechsler, 2011), Psycholinguistic Assessments of Language Processing in Aphasia (Kay, Lesser, & Colheart, 1996), and select subtests from the Delis-Kaplan Executive Function System (Delis, Kaplan, & Kramer, 2001). Psychometric properties of the ECAS indicate that it is a highly sensitive measure of overall cognitive decline; the ECAS total and composite scores were highly correlated with findings from the neuropsychological test battery (Pinto-Grau et al., 2017).

Interestingly, the literature indicates that ALS patients do not demonstrate a significant learning effect when administered the ECAS within a 6-month timeframe (Burkhardt, Neuwirth, & Weber, 2016). As such, if providers prefer to administer this instrument for screening or baseline testing, it can be administered every 6 months; however, if repeat testing is required prior to the 6-month timeframe, a parallel version of the ECAS can be administered and used interchangeably with the original version (Crockford et al., 2015). The availability of alternate forms helps to address the issue of practice effects for individuals who require evaluation at shorter time intervals (i.e., every 3 to 6 months). At the very least, the current study recommends that both the patient and carer instruments be administered on an annual basis unless otherwise indicated (e.g., report of symptoms within the 1-year period, presence of risk factors).

ALS Cognitive Behavioral Screen (ALS-CBS). The ALS-CBS has been validated against formal neuropsychological testing and was designed to assess executive and behavioral functioning in the ALS population (Woolley et al., 2010). The cognitive section is completed by the patient, and the behavioral component is completed by the caregiver. The test is administered in 5 minutes and yields a total cognitive score that is derived from four subtests: initiation and retrieval, concentration, attention, and tracking monitoring. It identifies patients with cognitive and/or behavioral changes with 71% specificity and 85% sensitivity (Woolley et al., 2010). If providers choose to administer this instrument for screening purposes, it is recommended it be administered annually to patients and caregivers.

Other screening instruments. In addition to the ECAS and ALS-CBS, several additional ALS-specific screening tests have been developed to evaluate FTI in ALS

patients, including the Penn State Screen of Frontal and Temporal Dysfunction Syndromes (Flaherty-Craig, Brothers, Dearman, Eslinger, & Simmons, 2009), the Cambridge Behavioral Inventory (Lillo et al., 2010), the ALS Frontotemporal Dementia Questionnaire (Raaphorst et al., 2010), and the University of California San Francisco Screening Battery (UCSF; J. M. Murphy, Ahmed, & Lomen-Hoerth, 2015).

Additionally, a telephone-based screening measure based on existing and validated screening measures has been developed recently and demonstrated findings that were equivalent to in-person testing of cognitive functioning, but not behavioral functioning (Christodoulou et al., 2016). The availability of telephone-based screening measures offers a convenient and efficient method of evaluating ALS patients by eliminating the need to for patients to be present physically in healthcare professionals' offices. As such, individuals who require more frequent screening can be evaluated without having to travel.

Neuropsychological assessment. A neuropsychological assessment is recommended if cognitive and/or behavioral impairments are noted on a screening measure or if a patient or caregiver is reporting symptoms in the absence of impaired performance on screening batteries. When planning the neuropsychological battery, it is important to balance the need for comprehensive screening with the needs and abilities of the ALS patient. Generally, it is recommended that the neuropsychological test battery assess multiple domains to increase the evaluation's sensitivity to detecting FTI and to thoroughly evaluate the cognitive domains that are typically impaired in ALS patients with FTI (i.e., executive functioning, personality/behavior; Strong et al., 2009). This type of battery will help the neuropsychologist determine whether the patient's pattern of

impairment is consistent with that of an ALS+FTI patient or if the impairment is related to other impairments, such as depression, pseudobulbar affect, or Alzheimer's disease (Giordana et al., 2011; Strong et al., 2009). Although this approach is ideal, it is often impossible because ALS patients do not have the stamina to complete multiple hours of testing (Flaherty-Craig et al., 2009). As such, the current study recommends that neuropsychologists begin with a brief battery that targets the cognitive domains that were impaired on screening measures or the areas that are of concern to the patient, caregiver, or healthcare provider. ALS patients are more likely to tolerate brief testing sessions, thereby producing valid and reliable estimates of their cognitive abilities.

The literature indicates that neuropsychological assessment of FTI in ALS patients should assess executive functioning thoroughly, as this is a hallmark impairment noted in ALS+FTI patients that can help differentiate between competing diagnoses (Strong et al., 2009). Generally, it is recommended that these batteries include at least one measure of verbal fluency (Strong et al., 2009), which can be adapted if speech difficulties are present (described below in detail). A thorough assessment should also include interviews with the patient and caregiver, as well as informant-report measures for the caregiver to complete about the patient's behavioral and emotional functioning (Strong et al., 2009). All evaluations will likely require adaptations from standard tests to minimize the impact of speech and motor impairments or other confounding variables (e.g., fatigue, frustration) on performance. Suggestions for adapting standard tests are described below.

Consistent with ALS+FTI research, this study recommends that standard fluency measures be administered as part of the neuropsychological test battery, including letter,

category, and design fluency tests (Strong et al., 2009). For ALS patients who do not demonstrate speech impairment, the Controlled Oral Word Association Test (COWA; Bechtoldt, Benton, & Fogel, 1962) is a valid and reliable measure that can be administered easily to evaluate letter fluency. For ALS patients who exhibit mild speech impairment but are able to complete the test, a modified version of the COWA can be administered (see Abrahams et al., 2000). In the adapted version, the patient is given 1 minute to produce as many words as he or she can that begin with a certain letter of the alphabet. There are three trials with the letters P, R, and W (see Benton & de Hamsher, 1976 for normative data). Thereafter, the examiner times the examinee as he or she reads the words he or she produced aloud as quickly as he or she can. To account for speech difficulties, a scoring correction is used in which the reading trial time is subtracted from the total generation time and the difference is divided by the total number of items produced during the generation trials.

For ALS patients with severe speech impairments, a written version of the verbal fluency test can be administered reliably (Abrahams et al., 1996; Thurstone & Thurstone, 1962). In this adaptation, the patient is asked to write as many four-letter 'S' (excluding proper nouns) words as he or she can in 5 minutes, then as many four-letter 'C' (excluding proper nouns) words as he or she can in 4 minutes. Then, the examinee is given a rest from motor tasks. After rest, the examiner times the examinee as he or she copies the list of 'S' and 'C' words as quickly as he or she can on a separate piece of paper to determine his or her writing speed. A correction procedure for oral/written fluency is used to calculate the Written Verbal Fluency Index (see Abrahams et al., 1996 for more information). The copy time is subtracted from the total time and the difference

is divided by the total number of items generated ([time allowed for test - time to copy words]/total number of words produced). The same procedure can be used for the category fluency test. In this regard, the examinee is given 2 minutes to write items belonging to a category. There are four trials: animals, colors, fruits, and towns. Again, the examinee is given a rest period and, subsequently, is timed as he or she copies the words he or she produced on a separate sheet of paper. The same formula is used as above with larger numbers corresponding to greater impairment.

The design fluency test can also be adapted to meet the needs of ALS patients. For individuals without significant motor impairment, standardized measures of design fluency can be administered without adaptation (e.g., Delis-Kaplan Executive Functioning Scales); however, if mild motor impairment is evident, design fluency can be adapted to correct for motor slowness (see Abrahams et al., 2000; Jones-Gotman & Milner, 1977). First, the examinee is asked to draw as many different nonsense/meaningless drawings as he or she can in 5 minutes. Second, the examinee is asked to draw as many four-line nonsense designs as he or she can in 4 minutes. Then, the examinee is timed as he or she copies the drawings from the previous two trials. The examiner keeps track of the number of rule breaks by counting the number of unacceptable drawings produced (i.e., drawings with more than four lines on the second task or nameable drawings on the first) and perseverative errors (i.e., repeated drawings). The calculation presented in the previous paragraph is used to correct for motor impairments.

Other commonly used tests can also be adapted to control for speech and motor deficits. For example, the Trail Making Test (Reitan, 1955) can be used to evaluate

cognitive flexibility in persons with motor impairment by computing the ratio between Part A and Part B (see Machts et al., 2014; Strong et al., 2009 for more information). Another option is to administer the oral version of the Trail Making Test (Ricker & Axelrod, 1994); however, validity studies and normative data are sparse so caution must be exercised when interpreting this measure. Researchers have demonstrated success by modifying components of standardized batteries, such as the COGNISTAT (Kiernan, Mueller, Langston, & Van Dyke, 1987). For instance, Flaherty-Craig and colleagues (2009) modified the aural comprehension commands to non-verbal commands (e.g., “blink twice, smile, then open your mouth”). Similarly, the Boston Diagnostic Aphasia Examination (Goodglass, Kaplan, & Barresi, 2000) can be adapted for speech or motor impairment by changing the verbal communication to eye blinks, thumbs-up, or nodding (Lezak, Howieson, Bigler, & Tranel, 2012). In addition, Abrahams and colleagues (2000) demonstrated success with the use of the National Adult Reading Test (NART; Blair & Spreen, 1989) to estimate premorbid intellectual functioning in ALS patients. They also used the Raven’s Standard Progressive Matrices (SPM; J. Raven, J. C. Raven, & Court, 1998) to measure current intellectual functioning and stated this measure was less vulnerable to the effects of motor impairment when compared to other commonly used tests of intelligence.

Neuropsychologists should also consider alternative ways to adapt the testing procedures in general. For instance, inquiring about the patient’s “best time of day” or scheduling multiple sessions may help to address issues with low stamina (Lezak et al., 2012). Patients who experience muscle spasms or cramping often benefit from rest breaks with opportunities to move and stretch; therefore, examiners should be alert for

signs of pain (Lezak et al., 2012). It is beyond the scope of this study to identify adaptations for all commonly used cognitive tests or test procedures; however, examiners are encouraged to consult the literature for possible modifications to other standardized tasks.

Most recently, studies have begun to explore and test the utility of eye-tracker controlled testing modalities as an alternative to paper-and-pencil tests (Poletti et al., 2017). Poletti and colleagues (2017) adapted cognitive tests of language, attentional abilities, executive functioning, and social cognition for eye tracking (ET) control, and tested this modality in a sample of healthy participants. Their findings revealed significant correlations between the ET-based battery and standard paper-and-pencil tests (e.g., Digit Sequencing Task, Montreal Cognitive Assessment), which suggests its potential for use with the ALS population. This assessment approach is in its infancy; however, it offers a promising solution to the difficulties associated with evaluating FTI in patients who have speech and motor impairments. Moving forward, it will be important for healthcare professionals and researchers to continue to explore how technology can be used to adapt existing cognitive measures. Furthermore, large-scale studies exploring cognitive performances within the ALS+FTI population could also address these limitations through the establishment of normative data for comparison. This represents an ambitious undertaking that will likely be time consuming; therefore, if scoring corrections or administration adaptations are unavailable at present, neuropsychologists are encouraged to rely upon demographically-corrected normative data (i.e., corrected for sex, race, age, and education) as comparison groups until more appropriate normative data are published. Similarly, subtests from well-established and

empirically validated test batteries, such as the Delis-Kaplan Executive Functioning System or Wechsler Memory Scale – Fourth Edition, can be administered, but it is essential for neuropsychologists to recognize the likely underestimation of the patient's abilities within the testing report. Lastly, it is recommended that continued efforts be placed on the development of parallel forms for existing screening batteries. These endeavors will increase the repertoire of neuropsychological tests that can be administered to ALS patients and, ultimately, will assist in the identification of FTI within the ALS population.

Education for patients and caregivers. Determining when to educate patients and caregivers about the possibility of FTI requires clinical judgment. There is research demonstrating that experienced clinicians are able to accurately judge the psychological well-being and coping ability of their ALS patients after one interview (Hugel et al., 2010). Nevertheless, patients and caregivers require and seek information at different paces and stages of illness (Goldstein & Abrahams, 2013; Wicks & Frost, 2008). Therefore, a standardized timeline is inappropriate and providers are encouraged to use their clinical judgment to determine when it is best to share information with patients and caregivers.

It is essential for healthcare providers to be sensitive to the individual needs and preferences of the patient and caregiver; however, it is also important for providers to educate their patients about the potential development of FTI symptoms, even if the news is difficult to deliver. Research indicates providers often do not educate their ALS patients and caregivers about the possibility of FTI symptom development (Wicks & Frost, 2008). In one study, patients and caregivers were given a test to evaluate their

knowledge of the physical and psychological components of ALS (Wicks & Frost, 2008). Findings indicated that both groups were knowledgeable about the physical components of the disease, but scored poorly on items testing their knowledge of psychological aspects of ALS. It is possible that clinicians hesitate to provide information because they do not want to burden patients or caregivers; however, when patients and caregivers were asked about their preferences for receiving information about FTI symptoms, a majority of individuals indicated they wanted to be told about the possibility of FTI symptoms (Goldstein & Abrahams, 2013; Wicks & Frost, 2008). For instance, in a study conducted by Goldstein and Abrahams (2013), the majority of patients indicated they wanted information about cognitive dysfunction (66%) and emotional dysregulation (73%), and the majority of caregivers stated they wanted to know about the possibility of cognitive dysfunction (73%) and pseudobulbar affect symptoms (80%). These findings are consistent with similar studies (Wicks & Frost, 2008), as well as the results of the current study. As such, the literature supports that withholding information is not always beneficial for patients and caregivers.

Given these findings, the current study recommends that a brief overview of FTI symptoms be provided to patients and caregivers following the completion of baseline assessments. The amount of information provided will be determined by the experienced clinician and response of the patient and caregiver; however, it is suggested that overall warning signs be reviewed to assist caregivers in identifying symptoms of FTI early in the disease process. Finally, if cognitive and/or behavioral impairment is identified through testing, healthcare professionals should provide a more detailed overview of those symptoms and discuss behavioral management strategies with caregivers.

Resources for support and information should also be provided to facilitate adjustment to life as an ALS+FTI caregiver.

Implications and Concluding Thoughts

It is hoped that the information obtained from this study increases understanding of the unique experiences and challenges faced by ALS+FTI caregivers. The current findings have important implications for the development and implementation of biopsychosocial interventions that address the specific needs of individuals caring for persons with ALS+FTI; however, additional studies are needed to increase understanding of the experiences of a broader sample of ALS+FTI caregivers. Moreover, unexpected challenges from this study uncovered the need for the development of empirically validated test batteries designed to assess FTI within the ALS population. Early identification of symptoms may facilitate adjustment to life as an ALS+FTI caregiver, and may also streamline treatments to address problematic and challenging behaviors exhibited by ALS+FTI patients. Finally, it is hoped that this study's findings increase physicians' understanding of the caregiving experience so they can educate caregivers about the challenges of ALS+FTI caregiving and provide them with informal and formal resources for support early in the disease process.

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