Behavioral Changes in Amyotrophic Lateral Sclerosis: Effects on Caregiver Burden and Interventions By Healthcare Professionals

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Amyotrophic lateral sclerosis (ALS), also known as Lou Gehrig’s Disease, is a fatal, neurodegenerative disease that harms nerve cells in the brain and spinal cord, eventually causing the inactivity of voluntary muscles of the patient and extreme physical and behavioral impediments (Tramonti, Bongioanni, Di Bernardo, Davitti, & Rossi, 2012). Major physical disabilities that progressively occur include loss of function of speech, limbs, swallowing and breathing. The average onset age for ALS is 55-65 years, and only five percent of cases occur before age 30. For the majority of ALS patients, there is no known cause of the disease (nonfamilial). For five percent of cases, the disease is familial and caused genetically. The average survival period of a patient with ALS is 3.5 years (Boerner & Mock, 2012; Pagnini, 2013). Because medication has little to no effect on the progression of the disease, an interdisciplinary team of healthcare professionals is essential for the management of symptoms and the maintenance of quality of life. It is extremely difficult for patients and their caregivers to adjust to the severe deterioration and rapid progression of the disease (Goldstein & Abrahams, 2013).

Because the effects of ALS are so powerful and because there is no known cure, the role of the caregiver is crucial and has been studied extensively. The caregiver, usually the patient’s family member or spouse, experiences significant emotional, physical, social, mental, and financial changes (Lillo, Mioshi, & Hodges, 2012). Typically spending around eleven hours per day caring for their loved ones, the caregivers live intense lifestyles (Boerner & Mock, 2012). It is extremely difficult for caregivers to preserve their quality of life as they care for the patient,
and they often experience physical and emotional illness, leading to a harsh burden (Williams, Donnelly, Holmlund, & Battaglia, 2008). Many times, the tasks of the caregiver are so demanding that he or she may feel the need to lessen time at work to spend more time caring for the patient, leading to increased distress and burden (Pagnini, 2013).

The rapid progression of the disease and the intense demands on caregivers have led researchers to develop a model that can reduce caregiver burden by meeting the various needs of the caregivers. Based on the results of interviews with subjects in their study, Williams et al. (2008) proposed a qualitative approach to assess the needs of ALS family caregivers. A small sample of 19 families participated in two phases of interviews. In the first phase, information was collected based on responses to the prompt, “What are all your personal needs since the diagnosis of your family loved one with ALS? I am interested in hearing about all needs you are aware of from the time of diagnosis until now” (p. 280). The responses of 109 items were recorded and analyzed in the second phase of data collection. Each caregiver was given the following instructions: “Please sort the individual item cards in any way that makes sense to you” (p. 280). Caregivers were asked to rate the importance of the items from least important to most important. The results indicate that the caregivers’ needs can be described in a four stage model. During Stage 1 (Early Coping and Adjustment), caregivers demonstrate the need to help all family members adjust to the new life, while also maintaining as much normality as possible. In Stage 2 (Maintenance), caregivers shift their focus to continuing a slow pace and retaining as much control as possible. Stage 3 (Transition to End of Stage) is characterized by the caregiver beginning to focus on hospice care, as well as assuring that other family members stay involved. Stage 4 (Coping with Change and Loss) involves inevitable grief and acceptance of the
unpredictability of the disease. The proposed model can be very helpful in understanding that as the disease progresses, more intense demands are placed on the caregivers. However, the results are based on a small sample size and the sample was limited demographically. A bigger sample might increase the applicability of the model for other caregivers. The findings of this study can inform health professionals to provide interventions for the families while they care for terminally ill family members. Providing these interventions can increase the quality of life for both patients and caregivers. Future studies should attempt to address this and examine how healthcare providers can help reduce the caregiver burden. Based on this model, it is evident that ALS caregivers need early access to sufficient care tailored to their own needs in order to lessen their potential burden.

Given that ALS is a disease with rapid physical deterioration, most people would assume that caregiver burden is most closely related to the physical progression of ALS. However, it has been demonstrated that the physical degeneration is not the aspect that impacts the caregivers the most. Instead, the caregiver’s well-being closely correlates with the behavioral changes exhibited by the patient. Behavioral changes occur in about half of ALS patients. A study by Lillo and colleagues advances the notion that behavioral changes are common in patients with ALS. Ninety-two patients participated in the study and completed a survey, with the help of their caregivers, where the motor function, behavioral changes, depression, anxiety, and stress of the patients were analyzed. A high rate of behavioral changes was reported, and apathy was the most notable. Apathy, or a lack of motivation, is expected to correlate with depression. However, according to these results and those of previous studies, there is no correlation between apathy and depression, anxiety, or progression of the disease. Forty percent of participants were
apathetic, but less than twenty-five percent were significantly depressed. In a similar study by Gibbons and colleagues, it was noted that the lack of association between apathy and depression in ALS patients is likely due to the fact that apathy is simply a symptom of ALS for some patients, not an emotional reaction to having the disease. The results of these studies indicate the prevalence of behavioral changes in ALS patients and the need for healthcare professionals to pay just as much attention to these changes as to the physical changes. The findings provide strong evidence for the presence of behavioral changes in patients with ALS, but the impact of these behavioral symptoms on caregivers needs further analysis (Gibbons, Richardson, Neary, & Snowden, 2008; Lillo, Mioshi, Zoing, Kiernan, & Hodges, 2011).

Over the past decade, research on the effects of behavioral changes on caregiver burden is becoming more acknowledged as the prevalence of the behavioral changes has gained a great deal of attention (Aoun et al., 2012). The purpose of a study conducted by Chio et al. (2010) was to evaluate the frequency of behavioral changes in ALS patients and the toll of these changes on caregiver burden. Seventy patients and their caregivers participated in the study and were assessed in a series of interviews and tests, including the Frontal Systems Behavior Scale (FrSBe), the Mini Mental State Examination (MMSE), and the Zung Depression Scale (ZDS). Similarly to the studies of Lillo (2011) and Gibbons (2008), the results showed that half of the participants displayed behavioral impairments and that these impairments impacted the caregivers’ depression, burden, and quality of life. The causes of caregiver burden have been mainly focused on progression of the patient’s physical status, but these results now indicate that the behavioral changes of the patient play a much more significant role in caregiver burden and depression. Apathy and executive dysfunction, which is a disruption to management and
regulation of cognitive functions, took the largest negative toll on the status of the caregivers. The authors concluded that because of the effects of patient behavioral changes on caregiver burden, it is critical that the caregivers are educated on the likelihood of their loved one undergoing these behavioral changes before they occur in order to lessen the burden that will come with the changes. It is again emphasized that healthcare professionals must be aware of this need for caregiver education and care, so interventions can be implemented.

Similarly to Chio and colleagues (2010), Lillo et al. (2012) explored caregiver burden and the dependency on behavioral changes rather than physical changes. Data was collected from 140 ALS patients and caregivers. Behavioral changes of the patients were measured by the CBI-R, and caregiver burden was measured by the Zarit Burden Interview (ZBI). Again in conjunction with Chio, the results of the assessments conveyed that behavioral impairments have a stronger impact on caregiver burden than progression of the patients’ physical impairments. Specifically, disinhibition and impulsivity were the behavioral changes that had the largest impact on the caregivers, while apathy is the most common in the majority of other studies. Notably, physical function had no impact on caregiver burden.

Further evidence of the negative impact of behavioral impairment on caregiver burden was presented in an 11-month study conducted by Cui and colleagues. Sixty-five ALS patients participated in a series of neurological, laboratory, neuroimaging, and electrophysiological exams. In this group of participants, irritability was the most prevalent behavioral change out of the 43% of patients who expressed behavioral impairments, compared to apathy being the most frequent in the majority of studies. Abnormal behaviors presented to be strongly associated with caregiver burden. The caregivers who reported previous emotional burden were the most
affected by the behavioral impairments. Along with the studies of Lillo et al. (2012) and Chio et al. (2010), the findings document the relationship between behavioral changes in ALS patients and increased caregiver burden (Cui et al., 2015).

Behavioral changes have been clearly demonstrated to impact caregiver burden, but the next logical step is providing interventions for caregivers to prepare them for the burden and hopefully lessen it. Healthcare providers are in a unique position to significantly influence the burden of the caregivers because they are in such close contact with the patients and caregivers throughout the entire course of the disease. However, the healthcare providers are often unaware of the severe impact of behavioral changes. Thus, informing and educating them is the key for possible interventions. Several studies have suggested the importance of potential interventions, but there still remains a large gap in this area of ALS research as the research on behavioral changes is relatively new. In their paper, Merrilees and colleagues provide a guide for healthcare providers help minimize the behavioral impairments that are likely to occur. Methods of dealing with executive deficits, apathy, irritability, poor judgement, impulsivity, loss of insight, emotional changes, social disinhibition, and aggression are summarized. The five main methods of helping patients and caregivers cope with abnormal behaviors are environmental, behavioral, pharmacological, physical, and internal to the caregiver. The guide outlines possible interventions for the negative behaviors through the five methods. While the authors focus on interventions for lessening patient symptoms, they clearly conclude the utmost crucial need of involving healthcare providers with interventions for lessening caregiver burden (Merrilees et al., 2010). According to Lillo et al. (2012), many healthcare providers who care for ALS patients are not aware of the impact on family caregivers. Therefore, healthcare providers need to be
educated on specific interventions that are necessary to help caregivers build their coping skills and lessen their burden.

Despite the evidence on burden and distress experienced by caregivers, there are limited studies on interventions for families caregivers. Based on their research, Aoun et al. (2012) suggested interventions, similar to those of Merrilees and colleagues (2010), to improve quality of life for ALS caregivers. Aoun and colleagues stress the importance of “providing education to health-care providers in order that they may better meet the emotional and practical needs of family caregivers” (p. 443). There is a lack of research on interventions that will improve the experience of caregiving for ALS families and also improve the care of ALS patients.

The most prominent studies in the field of behavioral changes of ALS patients and caregiver burden suggest the presence of behavioral impairments in ALS patients and the negative relationship those impairments have with caregiver burden. However, the key factor that is missing in this field of research is educating healthcare providers on how, when, and why to implement interventions for caregivers of ALS patients with behavioral symptoms. Further research is needed to prevent caregivers from experiencing such a large burden associated with the behavioral changes of their loved ones. If effective interventions are developed that include health care providers helping ALS caregivers through the caregiving process, this burden that has been extensively studied can be alleviated.


Lillo, P., Mioshi, E., & Hodges, J. (2012). Caregiver burden in amyotrophic lateral sclerosis is more dependent on patients’ behavioral changes than physical disability: A comparative


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