This paper presents a critical account of research into the non-motor symptoms associated with amyotrophic lateral sclerosis (ALS). Although research examining the cognitive and behavioural features of ALS has been extensively reported, social communication and emotion recognition changes have not been comprehensively explored. Furthermore, the current research and diagnostic criteria for diagnosing such changes, which have served as a useful conceptual model, do not provide a means of assessing the subtle cognitive changes reported in non-demented ALS patients. In addition, the impact and challenges of providing care for a person with a diagnosis of ALS who is also experiencing changes in cognition and behaviour have been explored only tangentially in the literature, and require immediate attention. We argue that the establishment of criteria to detect mild cognitive impairment may serve as a useful model to provide effective clinical interventions for both patients and caregivers at the earliest possible moment.

Key words: Frontotemporal dementia (FTD), Cognition, Behaviour, Social cognition, Emotion recognition, Caregiver burden
Amyotrophic lateral sclerosis (ALS) is a fatal neurodegenerative disorder of unknown aetiology (Kertesz 2011). It is characterised by the loss of upper motor neurons (UMN) and/or lower motor neurons (LMN) in the primary motor cortex, corticospinal tracts, brainstem and spinal cord (Wijesekera & Leigh, 2009). Traditionally, ALS was considered a progressive neuromuscular disorder restricted to the motor cortex. However, increasing evidence suggests that cerebral regions outside the motor cortex also atrophy in ALS (Strong, Grace, Freedman et al., 2009). Research has found that comorbid FTD and executive impairment in ALS are a negative prognostic indicator, associated with a shorter period of survival (Elamin, Phukan, Bede et al., 2011). Death generally ensues after two to three years, typically from respiratory failure as a result of bulbar paralysis (Mitsumoto, Przedbroski & Gordon, 2005). Approximately 5-10% of patients are thought to suffer severe cognitive and behavioural changes consistent with a profile of frontotemporal dementia (ALS-FTD), while other patients have been reported to display more subtle alterations in behaviour (ALSbi) or cognitive impairment (ALSci; Strong et al., 2009).

Research has charted the development of cortical degeneration in the anterior frontal and temporal cortex of ALS patients, reliably identifying changes in the domains of verbal fluency, abstract reasoning, judgment, and behavioural disinhibition (Abrahams, Leigh & Goldstein, 2005). Research has also revealed that ALS patients suffer deficits in social cognition, specifically the ability to infer mental states and discriminate between emotions (Zimmerman, Eslinger, Simmons & Barrett, 2007). Neuropsychological findings underscore the fact that the cognitive abilities of some ALS patients are impaired during disease progression, with cognitive and behavioural changes potentially impairing the patients’ ability to effectively communicate and engage in a wide range of situations and relationships. Such changes would be expected to negatively impact the patients’ quality of life and place a heavy burden upon those who care for them. Previous literature in dementia and progressive neurology has found increased caregiver burden and distress with increased severity of cognitive decline resulting in behavioural difficulties (Davis & Tremont, 2007), which encompass changes in personality, social interaction and communication, and emotional reactivity (Ballard, Lowery, Powell et al., 2000).

Although research into the cognitive and behavioural features of ALS has been extensively reported (Gibbons, Richardson, Neary et al., 2008; Raaphorst, De Visser, Linssen et al., 2010), social communication and emotion recognition changes have not been thoroughly explored. Social-emotional impairments can include alterations in social-interpersonal conduct, including impaired perspective-taking and impaired emotion recognition (Douglas, 2004). Currently, efforts to understand the range of social communication and emotion processing deficits in ALS are gaining momentum (Girardi, MacPherson & Abrahams, 2011; Meier, Charleston & Tippett, 2010). However, the impact of such deficits on caregivers...
of ALS patients has yet to be fully characterised and warrants immediate attention. The need for further research in this area is highlighted by the fact that the recently updated “good practice” guidelines, for supporting persons with ALS and their families (Miller, Jackson, Kasarskis et al., 2011), were unable to offer effective management techniques for dealing with cognitive and behavioural changes, due to a lack of evidence-based research within this specific area of ALS.

We review research examining the cognitive and behavioural alterations exhibited by persons with ALS, drawing particular attention to the current research and diagnostic criteria which have developed a useful conceptual model, but appears to lack sensitivity in detecting early signs of cognitive and behavioural alterations in non-demented ALS patients. Social cognitive deficits in ALS, which may serve as an early indicator of hallmark cognitive dysfunction, have yet to be fully characterised. Furthermore, there is limited research into the impact and challenges of providing care for a person with ALS who is also experiencing changes in cognition and behaviour. This is of particular importance as research in dementia and progressive neurology has firmly established that alterations in behaviour and emotional reactivity contribute to high levels of burden experienced by caregivers (Davis & Tremont, 2007). Only by examining the broader spectrum of symptoms associated with ALS from a biological, psychological, and social perspective can effective clinical interventions be delivered to both patients and those who care for them.

**CURRENT OPINIONS AND LIMITATIONS ON THE DIAGNOSIS OF COGNITIVE AND BEHAVIOURAL IMPAIRMENT IN ALS**

ALS is conceptually recognised to be a multisystem disorder in which motor symptom are a prominent feature, but in which non-motor symptoms are also frequently observed (Strong et al., 2009). When present, functional and structural changes in the frontotemporal region manifest as executive dysfunction and behavioural disinhibition, which are commonly observed in frontotemporal dementia (FTD; Ringholz, Apple, Bradshaw et al., 2005). Determining the presence of frontotemporal dysfunction in ALS is essential, as research consistently shows this clinical subgroup has a significantly shorter period of survivorship by at least one year compared to cognitively intact ALS patients (Miller, Jackson, Kasarskis et al., 2011).

Given the complexity and wide range of frontotemporal cognitive and behavioural syndromes observed in ALS, the research and diagnostic consensus criteria for diagnosing such change is consistent with three distinct variants (Strong et al., 2009):

1) [ALS-FTD] – Motor symptoms in conjunction with cognitive and behavioural changes consistent with a profile of frontotemporal dementia; characterised by an insidious onset with gradual progression, early decline in social inter-
personal conduct, early impairment in regulation of personal conduct, and impaired executive functioning;

2) [ALS-bi] – ALS with alterations in behaviour; requiring that the individual meet at least two non-overlapping supportive diagnostic features from either the Neary criteria and/or Hodge’s criteria;

3) [ALS-ci] and ALS with cognitive impairment, where the patient demonstrate cognitive decline on standardised neuropsychological testing two standard deviations below the mean, compared to age- and education-matched norms, on at least two distinct cognitive tests sensitive to executive functioning. Although preliminary consensus criteria have been established in an attempt to define non-motor ALS phenotypes (Strong et al., 2009), these have not yet been widely adopted by researchers, and time is needed to ascertain whether such classification will prove to be fruitful in further characterising cognitive and behavioural impairment in ALS.

Some studies have documented mild cognitive and behavioural changes in ALS patients over a six-month period (Murphy, Henry, Langmore et al., 2007; Robinson, Lacey, Grugan et al., 2006), while others have found no change over 12 months (Kilani, Micallef, Soubrouillard et al., 2004). Presently, it is not clear whether patients progress from ALSci or ALSbi to ALS-FTD. With respect to the rapidly progressive nature of ALS, research has yet to comprehensively examine the progression of cognitive impairment in ALS longitudinally. However, tracking the natural progression of cognitive decline in ALS is difficult due to progressive muscular deterioration and high mortality, which in turn affects sample sizes (Kilani et al., 2004).

Although diagnostic criteria are available, these criteria reflect a stage of significant cognitive impairment (i.e., 2 standard deviations below the mean). While helpful in conceptualising the spectrum of cognitive and behavioural impairment in ALS, the current diagnostic criteria are limited in that they have not yet addressed issues pertaining to the detection of early signs of cognitive decline in non-demented ALS patients. It is important to note that as frontal dysfunction is an insidious, progressive disorder, with no fixed events that clearly delineate its onset, it may be particularly challenging for clinicians to identify transition points from the asymptomatic phase to the symptomatic pre-dementia phase, or from the symptomatic pre-dementia phase to dementia onset in individual patients. To that end, the establishment of criteria akin to pre-Alzheimer’s disease (AD) Mild Cognitive Impairment (MCI), with a recommended cut off of 1 to 1.5 standard deviations below the mean (Albert, DeKosky & Dickons, 2011), may be a useful and sensitive conceptual model within ALS.

Furthermore, individual cognitive reserve must be considered when attempting to determine the presence of cognitive dysfunction in ALS. For example, individuals might be performing in the average range relative to their peers, yet this may still represent a significant decline if their premorbid abilities fell within the high average or superior range. In a recent study, Phukan and colleagues (2011) demonstrated that the Wechsler Test of Adult Reading (WTAR) was able to sig-
significantly distinguish between patients with ALS who were cognitively impaired and those who were cognitively intact. Accurately estimating levels of pre-morbid intelligence in ALS, particularly in pre-morbidly high functioning individuals who exhibit subtle reductions in cognitive ability, is an important consideration across both clinical and research domains. Detection of mild cognitive impairment promotes intervention at the earliest possible moment to assist both patients and caregivers to adequately prepare and plan for what lies ahead. Research has identified that taking active steps to prepare for death seems to represent positive coping rather than psychological distress (Rabkin, Wagner & Del Bene, 2000). In light of these findings, early interventions are important to allow both patients and caregivers to utilise what time they have left effectively, especially if faced with cognitive and/or behavioural deficits which may impede the patient’s ability to make complex end-of-life preparations or significant decisions about the welfare of their loved ones and their own interests.

COGNITION AND EXECUTIVE FUNCTION IN ALS

It is now commonly accepted that the prevalence of cognitive change in ALS is up to 50% (Hodges, 2008; Gibbons, Richardson, Neary et al., 2008), with published literature reliably identifying alterations in the domains of verbal fluency, abstract reasoning and judgment, language, and memory (Abrahams, Leigh & Goldstein, 2005). In a recent prospective population-based study, Phukan and colleagues (2011) found that 34.1% of non-demented ALS patients were cognitively impaired, while 13.8% of patients fulfilled the Neary criteria for frontotemporal dementia (FTD). The nature of the cognitive impairment was predominantly, but not exclusively, dysexecutive in nature, and many patients also showed impairments in language and memory function. The heterogeneous profile of cognitive deficits in ALS was highlighted in a recent meta-analysis of 16 studies exploring cognition in ALS (Raaphorst, De Visser, Linssen et al., 2010). This study found that non-demented ALS patients scored significantly lower on the Mini Mental State Examination (MMSE), in addition to measures of psychomotor speed, fluency, language, visual memory, immediate verbal memory and executive functioning. These findings illustrate the consistent presence of cognitive impairment in ALS. However, some studies have shown that highly utilised psychometric measures may be unsuccessful in determining the presence of very early signs of cognitive impairment in ALS (Girardi, MacPherson & Abrahams, 2011).

Similar to FTD, cognitive dysfunction in ALS may not be the most reliable early indicator of frontal pathology. For example, FTD patients with orbitofrontal lesions display few quantifiable cognitive deficits early in the disease (Gregory, Serra-Mestres & Hodges, 1999). This is problematic because widely-used cognitive screening instruments (e.g. MMSE) have been reported to be ineffective in detecting frontal dysfunction, where patients tend to perform within the normal range on a host of tasks thought to tap into executive functioning, such as verbal fluency – a measure that is frequently thought to signify the presence of compromised
frontal networks (Visskontas, Possin & Miller, 2007). Although this is not yet substantiated, it would seem reasonable to presume, given the overlap between ALS and FTD, that a similar pattern of cognitive impairment may emerge in cases of ALS-Sci and ALS-FTD, and subsequently render frontal executive tests less sensitive in detecting the hallmark pathology in the early stages of disease progression.

Normal performances on tests of executive function rely on the preservation of the lateral prefrontal cortex (Knight & Stuss, 2002). In FTD, when performance falls within a normal range, cortical degradation may be present in orbitofrontal or dorsomedial regions (Hodges, 2008). These regions show susceptibility to degradation early in ALS, with research noting deficits in social communication and emotion recognition well before signs of overt executive dysfunction are detectable (Girardi, MacPherson & Abrahams, 2011). As it stands, more rigorous means of screening for early signs of frontotemporal change are needed, especially in non-demented ALS patients.

SOCIAL COGNITION AND EMOTION PROCESSING

Deficits in social cognition and emotion recognition, which encompass impaired perspective-taking and emotion recognition, are reported in ALS (Girardi, MacPherson & Abrahams, 2011; Meier, Charleston & Tippett, 2010). Despite the presence of social-emotional impairments in ALS, including alterations in social-interpersonal skills, neither the recently updated good practice guidelines for caring for persons with ALS (Miller, Jackson, Kasarskis et al., 2009), nor the current research and diagnostic criteria for diagnosing cognitive and behavioural change in ALS, have acknowledged such changes. Given the continued documentation of social cognitive impairments in ALS, the current research and diagnostic criteria may require updating to consider deficits in social cognition a core disease element.

Tests of social cognition have been shown to be sensitive in detecting compromised frontal networks much earlier than traditional psychometric measures (Girardi et al., 2011). However, the most widely employed and recommended measure for establishing the presence of a frontal dysfunction in ALS is the presence of executive dysfunction, evident in tests of verbal fluency. In a sample of non-demented ALS patients examined by Girardi, MacPherson, and Abrahams (2011), patients performed within the normal range on a host of executive tests, including verbal fluency, while displaying a significant impairment on social cognitive tasks. This suggests that cortical regions responsible for comprehension of social cues may in fact be susceptible to compromise early in the disease process and may subsequently serve as the most reliable early indicator of cognitive impairment in ALS.

Research has established that tests of social cognition appear to be of particular value for the detection and early diagnosis of frontal dysfunction where formal and highly utilised psychometric measures fail to detect alterations in non-demented ALS patients (Girardi, MacPherson & Abrahams, 2011). The static nature of social cognitive measures utilised in ALS research (e.g., Facial Expressions
of Emotions Test, Reading the Mind in the Eyes) do not capture the unstructured, non-artificial and dynamic scenarios that are conveyed in everyday social interactions via emotional, prosodic and facial morphology. Complex social communication, such as sarcasm, requires an understanding of both the latent and actual content in order to evaluate both the facts surrounding the scenario and the inferences made by the speaker (McDonald, 1999). Similar to cortical areas implicated in social cognitive dysfunction in ALS, individuals with ventromedial and orbitofrontal lesions have been reported to perform poorly on tests of sarcasm recognition, which correlates with a reduction in empathy and inferring affect (Shamay-Tsoory, Tomer & Aharon-Peretz, 2001). Studies have repeatedly demonstrated that poor social skills are linked to a breakdown in relationships and social isolation (Halford & Hayes, 1995; Travis & Singmen, 1998). Though not yet investigated, it would seem reasonable to presume that deficits in social cognition would lead to similar outcomes in ALS.

An ecologically valid measure of social cognition (The Awareness of Social Inference Test, TASIT; see McDonald, Flanagan & Rollins, 2003), based on video vignettes of actors making sincere, sarcastic or paradoxically sarcastic statements (e.g., a seemingly true, yet contradictory statement), has revealed that patients with frontotemporal atrophy performed well below expected levels when interpreting complex social stimuli, such as sarcasm and identifying negative emotions (Kipps, Nestor, Acosta-Cabronero et al., 2009). Deficits such as these correspond to atrophy in the right lateral orbitomedial – temporal lobe – amygdala network, but not orbitofrontal or dorsolateral neocortical networks (Kipps et al., 2009). It is important for research to employ these ecologically valid measures to determine the scope of social cognitive deficits in ALS and examine the potential impact that these impairments may conceivably place on patients and caregivers.

**BEHAVIOUR**

When present, behavioural changes in ALS tend to reflect the spectrum of symptoms seen in the behavioural variant of FTD (bvFTD; Murphy, Vanderpool & Lomen-Hoerth, 2006). Studies of carer ratings of gross behavioural changes, using behavioural inventories designed to tap frontal lobe dysfunction, consistently report that a large majority of ALS patients exhibit apathy, irritability, mental rigidity, disinhibition, and diminished social judgement (Lomen-Hoerth, Murphy, Langmore et al., 2003), elevated scores in self-centeredness, and a reduction in empathy (Gibbons, Richardson, Neary et al., 2008). However, by the time individuals with ALS are reported to display significant behavioural changes, they are typically in the middle phase of the disease and display significant motor impairment (Gibbons et al., 2008), or meet the criteria for ALS-FTD (Lomen-Hoerth et al., 2003), making behavioural interventions extremely difficult given their lack of insight and cognitive rigidity (Miller, Jackson, Kasarskis et al., 2009).
While not yet reported in ALS, very early alterations in behaviour and social conduct are difficult to detect in cases of FTD, with literature reporting that individuals who go on to develop definite FTD are often initially misdiagnosed with a range of psychiatric or affective disorders (Mendez, Shapira, Woods et al., 2008; Passant, Elfgren, Englund et al., 2005). In a recent study, Rosness and colleagues (2008) revealed that arriving at a clinical diagnosis of FTD took approximately four years in a Norwegian sample and five years in a sample of Swedish patients. Overall, 71% of individuals who went on to develop definite FTD initially received a non-dementia diagnosis. While not yet empirically investigated, it would seem reasonable to assume that detecting behavioural alterations in ALS would be equally, if not more, challenging – particularly as the motor symptoms associated with ALS have the potential to overshadow behavioural problems. Furthermore, clinicians rely primarily on information provided by caregivers to corroborate the presence of behavioural symptoms in ALS (Gibbons, Richardson, Neary et al., 2008; Lillo, Mioshi, Zoing et al., 2011). This may not be reliable, however, as research has noted that loss of physical function in ALS is distressing to caregivers (Pagnini, Rossi, Lunetta et al., 2010), and may subsequently impact upon accurate reporting of behavioural alterations in the early stages of disease progression.

The recommended criteria for diagnosing behavioural dysfunction in ALS are the same criteria used to diagnose FTD (Strong et al., 2009). However, these criteria have been reported to be ineffective at detecting hallmark behavioural alterations in the early stages of FTD (Piguet, Hornberger, Shelley et al., 2009). Unlike Alzheimer’s disease, where a diagnosis is arrived at in a timely fashion, due to the disease’s hallmark amnestic presentation (Dubois, Feldma, Jacova et al., 2007), the reverse pattern is reported in attempting to diagnose FTD, due to the subtle and insidious nature of the disease, which does not significantly interfere with daily living in the initial stages (Piguet et al., 2009). Although the clinical features of bvFTD are well established, a recent retrospective clinical review by Kipps, Hodges, and Hornberger (2010) identified patients who fulfil diagnostic criteria for FTD but do not appear to progress clinically. The authors argued that the widely used criteria established by Neary and colleagues (1998) lack both sensitivity and specificity in distinguishing static from progressive FTD, particularly in the early stages of the disease. This is particularly worrying, since the criteria for diagnosing FTD are used to diagnose behavioural dysfunction in ALS.

In response to the finding uncovered by Kipps, Hodges, and Hornberger (2010), revisions to the FTD criteria were undertaken in an attempt to improve sensitivity and specificity and reduce the ambiguity of behavioural descriptors and the inflexible application of the criteria (see Rascovsky, Hodges & Knopman et al., 2011). Of 137 cases evaluated using both the revised and previously established 1998 diagnostic criteria, 118 (86%) met criteria for possible FTD, and 104 (76%) met criteria for probable FTD. In stark contrast, the proportion of cases fulfilling the 1998 criteria (53%) was significantly lower. The increased sensitivity of the revised FTD criteria is thought to reflect the optimised diagnostic features,
less restrictive exclusion features and particularly, a flexible structure that accommodates distinction in the symptom profile at presentation. Use of revised criteria for FTD diagnosis appears to have the potential to improve identification of the syndrome, particularly in the early stages when interventions are most likely to be effective (Rascovsky, Hodges, Knopman et al., 2011). To date, no empirical investigation has been conducted to determine whether the newly proposed FTD criteria are more accurate than the 1998 criteria in establishing the presence of behavioural alterations at an earlier stage in ALS. In light of these findings, the current criteria for diagnosing behavioural impairments in ALS may benefit from review, with the incorporation of the newly proposed criteria potentially providing a more sensitive indicator for detecting hallmark FTD-like behavior in ALS, and thus supporting early intervention when behavioural-modifying therapies are most likely to be beneficial.

**CAREGIVER BURDEN**

An estimated 2.4% of Australian adults (approximately 474,600 primary caregivers) provide ongoing substantial unpaid assistance to a family member (Edwards, Higgins, Gray et al., 2008). Compared to familial carers of patients with other diseases, those who care for individuals with dementia are twice as likely to be providing a higher amount of home care (approximately 40+ hours a week of direct care; Epstein-Lubow, Davis, Miller et al., 2008). This is especially true for individuals caring for sufferers of FTD, for they are faced with patients exhibiting marked behavioural and cognitive alterations that are poorly understood by the general public and the health care system alike (Riedijk, De Vugt, Duivenvoorden et al., 2006). Similar findings are reported in studies of ALS, where carers are placed under high levels of physical and emotional stress due to the debilitating nature of the disease, and are once again thought to provide a high amount of home care (a median of 11 hours a day; Krivickas, Shockley & Mitsumoto, 1997). While research has investigated the impact of caring for individuals with ALS, the primary focus of such investigations thus far has been on motor symptoms associated with disease progression and the impact they place on caregivers (Chio, Gauthier, Calvo et al., 2005). Literature addressing the impact and challenges of providing care for a person with a diagnosis of ALS who also experiences changes in cognition and behaviour have been explored only tangentially (Gordon, Wand, Doorish et al., 2007). While Gordon et al. (2007) revealed that cognitive and behavioural changes and more severe forms of ALS were associated with depressive symptoms in caregivers, factors such as caregiver burden and anxiety were not explored. A further limitation of this study was that the authors did not elaborate on the specific types of cognitive and behavioural changes that contributed to depressive symptoms in caregivers. Studies in dementia have shown that factors associated with frontal lobe dysfunction (e.g., disinhibition, apathy, executive dysfunction) are highly predictive of caregiver burden after controlling for disease severity and depression (Davis & Tremont, 2007).
Given that behavioural problems in dementia are some of the strongest predictors of caregiver burden, investigating the scope and contribution of specific behaviours exhibited in ALS has the potential to allow clinicians to implement and target appropriate interventions for managing problematic behaviours.

Research has established that communication difficulties between caregivers and care recipients contribute to the level of burden (Savundranayagam, Hummert & Montgomery, 2005). Caregivers of dementia patients frequently report that significant difficulties arise as a result of a severe breakdown in the care recipient’s interpersonal communication, inability to read social cues, and social faux pas (Murray, Schneider, Banerjee et al., 1999). According to caregivers, communication breakdown is the most difficult aspect of caregiving (Murray et al., 1999). Furthermore, research in traumatic brain injury has found impaired communication skills were a direct consequence of frontal system dysfunction and a significant factor in interpersonal difficulties, difficulties maintaining social relationships and reduced quality of life (Douglas, 2004). Disturbances in social aspects of behaviour, such as social inappropriateness, have been reported in ALS (Hodges, 2008), and thus specific investigations of deficits in social cognition and social communication are warranted. Given that research has shown that alterations in both behaviour and emotion recognition have been linked to early signs of cognitive dysfunction in ALS, examining social-emotional communication has the potential to provide novel insights into the nature of emotion recognition and behavioural dysfunction in a sub-population of persons with ALS and the subsequent impact that places on caregivers.

Burden, depression and anxiety are associated with alterations in behaviour, social conduct (Davis & Tremont, 2007), and social cognitive deficits (Ballard, Lowery, Powell et al., 2000). Given that research has documented alterations in emotion recognition in persons with frontotemporal dementia (FTD; Snowden et al., 2005) and ALS (Lule, Anders, Kassubek et al., 2005; Zimmerman, Eslinger, Simmons et al., 2007), it would seem reasonable to predict a relationship between impaired emotion recognition, social communication (i.e., both verbal and gestural interactions) and behaviour disturbances. Literature in dementia and progressive neurology has found increased caregiver burden and distress with increased severity of behavioural difficulties (Davis & Tremont, 2007), which encompass changes in ‘personality,’ ‘social interaction and communication’ and ‘emotional reactivity’ (Ballard et al., 2000). Currently there is little published research that investigates the impact of social communication and emotion processing deficits on caregivers. Only by quantitatively examining the nature of impairments in emotion recognition and social communication and investigating how such difficulties impact on caregivers can appropriate evidence-based education or intervention programs be established.
CONCLUSION

The last few years have seen a considerable amount of research centered on understanding the spectrum of cognitive and behavioural dysfunction between ALS and FTD, and the impact that such alterations have on care-givers. However, a number of issues pertaining to the diagnostic criteria must be addressed before research can effectively begin to assess and intervene clinically within this population. As they stand, the criteria for diagnosing both cognitive and behavioural impairment in ALS appear insensitive to subtle changes early in disease progression and thus require revision to improve sensitivity to early alterations in cognition and behaviour. Whether or not cognitive impairment is an inevitable hallmark of ALS, or more strictly present in a sub-group, remains to be determined. However, one thing is certain: the current criteria for diagnosing cognitive impairment in ALS reflect a stage of considerable impairment, and as such, fail to capture more subtle changes in non-demented ALS patients. The establishment of criteria to detect mild cognitive impairment may serve as a useful model to provide effective clinical interventions to both patients and caregivers at the earliest possible moment.

Changes in social cognition and emotion recognition, which are related to increased caregiver burden and distress as a result of behavioural difficulties (Savundranayagam, Hummert & Montgomery, 2005), have also not yet been extensively explored in ALS. Research has effectively illustrated that where highly utilised psychometric measures fail to detect cognitive change in both ALS (Girardi, MacPherson & Abrahams, 2011) and FTD (Gregory, Serra-Mestres & Hodges, 1999), measures of social cognition may be more sensitive in detecting change at an earlier stage in disease progression (Girardi et al., 2011; Meier, Charleston & Tippett, 2010). Clearly delineating the extent of cognitive and behavioural impairment in ALS has important implications. Given the overwhelming burden endured by caregivers when dealing with ALS (Chio et al., 2005) and FTD (Riedijk, De Vugt, Duivenvoorden et al., 2006), it would be expected that providing care for patients who experience both motor and cognitive decline would be even more challenging and further add to the level of burden and psychological ill health in caregivers.

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StaioS et al., ALS carers and non-motor changes


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