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GENERATIVE NAMING IN ALS: A LONGITUDINAL STUDY

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Abstract

Category specific deficits in naming and comprehension for living and non-living items exist in individuals with brain impairment. No research to date has examined the performances of individuals with ALS on generative naming tasks for these two different categories. The purpose of this study was to examine the nature of verbal category fluency performances for living and non-living items of individuals with ALS and control participants over time. A small sub-group of participants with ALS was significantly different than controls on the number of items generated for all categories with no difference between living and non-living categories. However, the sub-group did produce more errors in the living categories suggesting that the living categories could be more vulnerable to language impairment in ALS. Additionally, more participants with ALS produced semantically related errors than controls, suggesting language could be affected in these participants. Further research is warranted to provide more insight on the nature of categorical naming impairments in ALS.

Keywords: amyotrophic lateral sclerosis (ALS), category fluency, generative naming, language, cognitive impairment, category-specific deficits

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Introduction

Amyotrophic lateral sclerosis (ALS) is a progressive neurodegenerative disease that affects both upper and lower motor neurons. ALS affects the lower motor neurons (LMN) of the brainstem and spinal cord and the upper motor neurons (UMN) of the cerebral cortex (Brockington, Ince, & Shaw, 2006). The incidence of ALS is approximately 1.5 to 2.0/100,000 (Mitchell & Borasio, 2007) and is considered one of the most common adult onset neurodegenerative diseases (Strong, Grace, Orange, & Leeper, 1996). Males are affected more often than females with a ratio of approximately 1.6/1.0 (Mitchell & Borasio, 2007). ALS is an adult-onset disease with the age of onset between 55 to 75 years of age (Mitsumoto, Chad & Pioro, 1998). ALS is fatal, usually the result of respiratory failure, where death usually follows 3 to 5 years after the onset of symptoms. However, there are individuals who live much longer. Twenty percent of patients with ALS survive longer than 5 years and 10% survive longer than 10 years (Shoesmith & Strong, 2006).

There are three different types of ALS including sporadic ALS, familial ALS, and Western Pacific ALS. Sporadic ALS is the most common accounting for approximately 90% of ALS cases. Familial ALS occurs in 5 to 10% of cases (Mitchell & Borasio, 2007; Shoesmith & Strong, 2006) and is usually an autosomal-dominant pattern of inheritance (Mitchell & Borasio, 2007). Familial ALS differs from sporadic ALS in that it has a younger age of onset (i.e., approximately 47 years of age) and males are not more likely to develop it (Mitsumoto et al., 1998). Other than inheritance, there is no single clinical feature

that distinguishes sporadic ALS from familial ALS (Mitsumoto et al., 1998). The Western Pacific variant of ALS has a high incidence of occurrence in the Chamorro people on the islands of Guam, Rota and Tinian, on the Kii Peninsula in Japan and in West New Guinea (Mitsumoto et al., 1998). The variant of ALS found among the Chamorro people and on the Kii Peninsula has a male to female ratio of 1.8:1 and a mean age of onset of 46 years (Hudson, 1981). The ALS variant found in West New Guinea is identical with the exception of a higher incidence. Although the Western Pacific variant of ALS is clinically indistinguishable from sporadic ALS, it is distinct because it commonly occurs with a parkinsonism-dementia complex (Mitsumoto et al., 1998).

Clinical Features of ALS

ALS is a complex clinical syndrome because symptoms resulting from the degeneration of upper motor neurons and symptoms resulting from the degeneration of lower motor neurons can occur in combination and to varying degrees (Mitsumoto et al., 1998). Upper motor neuron degeneration causes the loss of dexterity, spasticity, weakness, hyperreflexia (i.e., increased muscle stretch reflexes), pathological reflexes such as the Hoffman or Babinski signs and clonus (Brockington et al., 2006; Mitsumoto, 1998). Degeneration of the lower motor neurons causes weakness, atrophy, fasciculations of the limbs and muscle cramping (Brockington et al., 2006; Mitsumoto et al., 1998).

ALS patients can first present with symptoms in the limbs (limb-onset) or in bulbar regions (i.e.,bulbar-onset). Approximately 75% of patients with sporadic ALS present with limb-onset while 21% present with bulbar-onset (Shoesmith &

Strong, 2006). Bulbar palsy affects the muscles of articulation, mastication and deglutition with symptoms that include mixed dysarthria, dysphagia, and sialorrhea (drooling) (Mitsumoto et al., 1998). When the lower motor neurons are affected it is referred to as flaccid or paretic bulbar palsy. In flaccid bulbar palsy, there is wasting of tongue musculature, flaccid tone and fasciculations (Darley, Aronson & Brown, 1975; Mitsumoto et al., 1998). Pseudobulbar palsy results from degeneration of upper motor neurons in bulbar regions and their descending corticobulbar tracts (Mitsumoto et al., 1998). Pseudobulbar palsy results in exaggerated snout and jaw reflexes and spasticity of muscles which cause slow repetitive movements of the tongue (Brockington et al., 2006). Emotional lability or uncontrollable episodes of laughing or crying also can be associated with pseudobulbar palsy (Brockington et al., 2006). Bulbar palsy in ALS is usually of a mixed profile comprising a mixture of upper and lower motor features (Darley et al., 1975).

The clinical features of ALS progression include weight loss due to amyotrophy, nutritional deficiencies as a result of dysphagia, head drooping due to weakening of neck musculature, and respiratory symptoms such as dyspnea and orthopnea (Brockington et al., 2006). Symptoms that result from nocturnal carbon dioxide retention (e.g., morning headaches, anorexia, and daytime somnolence) also can develop. In rare cases, patients with ALS can suffer sensory symptoms such as inconstant limb paresthesias, sensations of pulling, burning or formication involving a body part or a vague painful discomfort (Bonduelle, 1975; Mitsumoto et al., 1998).

Speech in ALS

Speech difficulties leading to decreased intelligibility are the result of the progressive degeneration of the oral, velopharyngeal, and laryngeal articulators (Bonduelle, 1975). The dysarthria that develops in patients with ALS is considered a mixed flaccid-spastic dysarthria which results from the degeneration of both upper and lower motor neurons (Darley et al., 1975). Individuals with bulbar involvement early in the presentation of ALS are more likely to exhibit a mixed dysarthria. The main speech characteristics related to the mixed dysarthria in patients with ALS are imprecise production of consonants and vowels, hypernasality, hoarse voice, short phrases and slow speaking rate (Darley et al., 1975). Additional speech characteristics can include harsh voice, strain-strangled voice, breathiness, audible inspiration, monopitch and monoloudness, low pitch, prolonged pauses between words and phrases, excess and equal stress, and nasal air emission (Darley et al., 1975). These characteristics manifest differently in each individual and can occur at different times throughout the course of the disease (Renout et al., 1995). As ALS progresses to advanced stages, speech becomes unintelligible and many patients become anarthric (unable to speak) (Mitsumoto et al., 1998).

Cognition and Neuroimaging in ALS

Traditionally ALS was thought to affect only upper and lower motor neurons and not to involve the cortical regions. Recent researchers however, who describe findings from a wide range of neuropsychological, neuroimaging and clinical studies, challenged this idea. It is now widely accepted that cognitive

impairment exists in ALS. Early studies by Gallassi et al. (1985, 1989) using neuropsychological tests demonstrated a slight but definite cognitive impairment in patients with ALS with spared memory function. Iwasaki, Kinoshita, Ikeda, Takamiya and Shiojim (1990) also detected cognitive impairment in their participants with ALS and concluded that memory, both immediate and delayed, were affected. David and Gillham (1986) used CT scanning and found mild degrees of cerebral atrophy in 57% of their participants, providing additional support that neurological impairments in ALS extend beyond the motor systems. They did not, however, indicate the extent and location of the atrophy.

Of these earlier studies, Poloni, Capitani, Mazzini, and Ceroni (1986) did not detect a cognitive impairment in their ALS participants. They determined that cognitive impairment in ALS is "a discrete, probably seldom occurring event" (Poloni et al., 1986, p. 259). However, their results can be attributed to using tests not sensitive to frontal lobe functioning (i.e., Wechsler Adult Intelligence Scale, digit span, spatial span on a block-tapping test, a prose-memory test [immediate and delayed] of a short tale, and a paired word-learning test) (Bak & Hodges, 2001).

Studies conducted over the past two decades provide further evidence that there is cognitive impairment in ALS. The authors of these studies used tests that are sensitive to frontal impairment and linked the cognitive impairment in ALS to frontal executive dysfunction (Abe et al., 1997; Abrahams et al., 1997; Abrahams et al., 2000; Hanagasi et al., 2002; Kew et al., 1993; Ringholz et al., 2005; Rippon et al., 2006; Strong et al., 1999; Talbot et al.,1995). The frontal

lobe-based abnormalities found after neuropsychological testing consisted of deficits in executive functioning (Abe et al., 1997; Abrahams et al., 1997; Hanagasi et al., 2002; Ringholz et al., 2005; Strong et al., 1999), deficits in attention (Abe et al., 1997; Hanagasi et al, 2002; Ringholz et al., 2005) and in some cases deficits in memory (Abrahams, Leigh, & Goldstein 2005, Iwasaki et al., 1990, Kew et al., 1993; Ringholz et al., 2005; Strong et al., 1999). Specific problems were found in the areas of mental flexibility (Strong et al., 1999), verbal and nonverbal fluency (Abrahams et al., 2000; Hanagasi et al., 2002; Kew et al., 1993; Ludolph et al., 1992; Racowicz & Hodges, 1998; Strong et al., 1999), working memory (Hanagasi et al., 2002; Ringholz et al., 2005), tests of sustained attention, response inhibition, and complex visuo-spatial processing (Hanagasi et al., 2002). Moreover, deficits were found on picture sequencing tasks, suggesting impairments in planning, organizing and self-monitoring (Talbot et al., 1995). Finally, difficulties with abstract reasoning, memory for both verbal and visual (pictorial) material (Mantovan et al., 2003) and impaired performance on the Wisconsin Card Sorting Test (WCST) which tests executive skills also were found (Strong et al., 1999).

Evidence from neuroimaging and neuropathological studies provides further support for frontal lobe dysfunction in patients with ALS. Frontal atrophy was found using both CT and MRI (Abrahams et al., 2004; Kiernan & Hudson, 1994; Poloni et al., 1986). Patients with ALS displayed impaired activation in the dorsolateral regions of the prefrontal cortex (middle and inferior gyri) and the anterior cingulated gyrus while performing a letter fluency task (Abrahams et al.,

2004). Additional abnormalities were revealed in the middle temporal gyrus, precuneus and inferior parietal lobes. Kiernan and Hudson (1994) used MRI and found cortical degeneration in the precentral gyrus and neurons that project to other motor regions of the frontal lobe.

Data from SPECT and PET studies add more evidence of impaired activation in frontal regions (Kew et al., 1993; Ludolph et al., 1992; Talbot et al., 1995). Talbot et al. (1995) showed reduced regional cerebral blood flow in the frontal and anterior temporal cortices while Kew et al. (1995) found lower regional cerebral blood flow in the medial prefrontal cortex and in the anterior cingulate cortex. Additionally, Ludolph et al. (1992) found a moderate decrease in cerebral glucose metabolism in the frontal cortex and in sub-regions such as the frontobasal cortex and the superior parieto-occipital cortex.

Cognitive impairment may be greater among those individuals with ALS who show bulbar involvement (Massman et al., 1996; Strong et al., 1999), although this is an inconsistent finding. Abrahams and colleagues (1997) explored the relationship of bulbar impairment with cognitive dysfunction in ALS by comparing patients who had pseudobular palsy to those without pseudobulbar palsy. Deficits were more pronounced for those with pseudobulbar palsy but were not limited to this group of patients. In a longitudinal study, Schreiber and colleagues (2005) found participants with bulbar profiles to be significantly more impaired than participants with spinal-onset at initial testing on tasks involving executive functioning. The degree of the significant difference increased over

time. They also found that participants with bulbar profiles were more impaired on memory tasks compared to those with spinal-onset.

Although the prevalence of cognitive impairment in ALS ranges widely in the literature, the number of individuals with ALS who exhibit cognitive impairment is reported to be as much as 35.6% (Massman et al., 1996). In two large studies conducted by Massman et al. (1996) and by Ringholz and colleagues (2005), which included 146 and 279 participants with sporadic ALS, respectively, the number of patients with cognitive impairment was 35.6% in the Massman et al. study and 30% in the Ringholz et al study. These results do not include participants who exhibit impairments significant enough to be classified as dementia.

ALS and Frontotemporal Dementia (FTD)

Dementia in individuals with ALS is a more recent conceptualization. Dementia is more severe than is cognitive impairment. A diagnosis of dementia requires multiple cognitive deficits of gradual onset and continual decline which include both memory and any one of the following: language problems, movement programming problems, agnosia, or disturbance in executive functioning. These deficits must cause significant impairment in social or occupational functioning and must represent significant decline from previous functioning. Additionally, deficits cannot be caused by other CNS conditions, systemic conditions known to cause dementia, substance abuse, delirium, or any other primary psychiatric disorder (DSM IV, 1994).

Although rare, an overt dementia is associated with all forms of ALS (Bak & Hodges, 1999; Hudson, 1981; Iwasaki et al., 1990; Kew et al., 1993; Neary et al., 1990; Rakowicz & Hodges, 1998; Ringholz et al., 2005). The development of dementia in ALS was noted as early as the late nineteenth century (Bak & Hodges, 2001). More recently ALS is commonly associated with a Parkinsonism-dementia on the Pacific Island of Guam in the Guamanian Chamorro population (Rodgers-Johnson et al., 1986). A similar association also was reported in the Kii Peninsula in Japan (Shiraki & Yase, 1975). Hudson (1981) determined that dementia is found not only in the Western Pacific variant of ALS, but also is found in both sporadic and familial types of ALS. However, he believed dementia to be a rare occurrence.

Authors of recent studies described changes in personality, behaviour, language and cognition in individuals with ALS that are consistent with those who exhibit a frontotemporal dementia (Bak & Hodges, 1997, 1999, 2001, 2004; Caselli et al., 1993; Neary et al. 1990; Rakowicz & Hodges, 1998; Ringholz et al., 2005; Rippon et al., 2006; Tsuchiya et al., 2000). Frontotemporal dementia results from degeneration of the frontal lobes, the anterior temporal lobes or both. Terminology used to refer to frontotemporal dementia is inconsistent in the literature. The term frontotemporal dementia (FTD) is used by some researchers and consists of both a frontal variant (fvFTD) and temporal variant (tvFTD) of the disorder (Perry & Hodges, 2000). Neary and colleagues use the term frontotemporal lobar degeneration (FTLD) which is considered a syndrome comprising the three clinical entities; frontotemporal dementia (FTD), primary

nonfluent aphasia (PA) and semantic dementia (SD). Kertesz proposed the term Pick's Complex which includes frontotemporal dementia (FTD), primary progressive aphasia (PPA) and semantic dementia (SD) as well as corticobasal degeneration (CBD), progressive supranuclear palsy (PSP) and frontotemporal dementia associated with motor neuron disease (FTD –MND) (2003).

In the latter two syndromes, FTD refers solely to the behavioural manifestation and is similar to the frontal variant FTD (fvFTD) described by Perry and Hodges (2000). FTD or fvFTD is caused by bilateral atrophy of the frontal lobes and is characterized by behaviour, personality and social conduct changes (Neary et al., 1998). PA or PPA is caused by unilateral damage affecting the dominant hemisphere and is characterized by nonfluent language with aggrammatism, phonemic paraphasias or anomia (Neary et al., 1998). Semantic dementia or temporal variant FTD (tvFTD) is caused by atrophy of the anterior temporal lobes and is characterized by fluent, empty spontaneous language and loss of word meaning and comprehension (Neary et al., 1998; Perry & Hodges, 2000). Other terminology used for frontal and/or temporal lobe dementias include dementia of the frontal lobe (Neary et al., 1988) and frontal lobe degeneration of the non-Alzheimer's type (Gustafson, 1987). However, these terms are not used commonly in the current scientific literature. Due to the differing terminology in the literature used to describe frontal and/or temporal dementia the terminology used to describe the dementia found in ALS also is inconsistent. In this paper the original terminology used by the authors of the respective studies will be used in order to be consistent with their useage.

The association between ALS and a frontal lobe dementia was first documented in 1932 with symptoms such as emotional changes (e.g., violent behaviour, emotional lability, irritability and depression), personality changes (e.g., suspiciousness and social withdrawal) and cognitive deficits (e.g., memory, language and general intellectual deficits) (Van Braumuhl, 1932 as cited in Bak & Hodges, 1999). Similar symptoms were reported more recently and include an early change in personality and the expression of inappropriate social behaviours (Bak & Hodges, 2004; Neary et al., 1990). Some symptoms described in the literature include disinhibition and impulsivity, irritability, apathy, unconcern and economy of mental effort. A change in eating habits and stereotyped and perseverative behaviour also were noted and in some cases co-occurred with paranoia and hallucinations. Anosognosia (failure to recognize impairments) also has been reported (Portet, Cadilhac, Touchon, & Camu, 2001). Neuropsychological testing identified significant frontal-executive deficits (Bak & Hodges, 2004; Racowicz & Hodges, 1998; Portet et al., 2001). Specifically, impairments in memory, judgement, reasoning and behavioural dyscontrol have been shown (Portet et al., 2001).

A 'progressive aphasia' also has been reported in patients with ALS (Bak & Hodges, 2001, 1997, 2004; Caselli et al., 1993; Rakowicz & Hodges, 1998; Tsuchiya et al., 2000). Language impairments described as aphasia are thought to coincide with dementia or are the sole characteristic of cognitive change in ALS (Bak et al., 2001; Rakowicz & Hodges, 1998). The more generalized use of

the term 'aphasia' in participants with ALS is misleading and confusing. The language deficits found in ALS are discussed in greater detail below.

Frontotemporal dementia can precede, coincide with, or follow the diagnosis of ALS. In a recent study examining the overlap of ALS with FTD, five of 36 patients with FTD also met diagnostic criteria of definite ALS, while two had limb denervation. Although the remaining patients had normal electromyography (EMG) findings, 6 had trouble swallowing while 5 displayed fasciculations. The location of the fasciculations (i.e., upper or lower extremities) was not reported. One of the patients who exhibited fasciculations but who had a normal EMG developed definite ALS within a year (Lomen-Hoerth, Anderson, & Miller, 2002). Other cases are described in which patients first present with dementia symptoms characteristic of FTD including an early change in personality and inappropriate social conduct. Motor symptoms develop within 6 to 12 months after the onset of the frontal symptoms (Bak & Hodges, 2001; Neary et al., 1990). The reverse also has been described (Abe et al., 1997; Kew et al., 1993; Lomen-Hoerth et al., 2003; Racowicz & Hodges, 1998; Ringholz et al., 2005; Rippon et al., 2006). In a study by Portet et al. (2001) 7 patients with ALS exhibited a fronto-temporal dementia and in each case ALS was identified first. Dementia was secondary and was not present in the first 6-months of the disease.

Unfortunately, an exact prevalence of patients with ALS who have dementia is unknown. Early reports indicate 3 to 5 percent of patients with ALS have a co-existing dementia (Kew et al., 1993). More recent studies show the percentage of dementia in ALS to be up to 15 to 23% (Ringholz et al., 2005;

Rippon et al., 2006). Higher prevalence values were recorded by Lomen-Hoerth et al. (2003) who concluded that 18 out of 44 ALS patients without prior diagnosis of dementia met criteria for probable or definite FTLD.

The differing prevalence rates of dementia found in ALS can be attributed to several factors. One such factor is referral biases as some researchers study only ALS patients in their clinics whereas other investigators include selected participants referred from various motor disorders clinics (Murphy et al., 2007). Also, motor neuron disease can progress rapidly and cause death before frontotemporal dementia becomes apparent (Murphy et al., 2007). Furthermore, the differing prevalence rates could be explained by the range of criteria used to determine whether an individual has dementia as well as by the different thresholds for cut-off scores used to determine impairment (e.g, 1.5 or 2 SDs below normative values). Different dementia diagnostic criteria have included the Neary Criteria for FTLD (Neary, Snowden, Northen, & Goulding, 1988), the Clinical Dementia Rating Scale (Berg, 1988), and dementia based solely on neuropsychological testing. Barson, Kinsella, Ong and Mathers (2000) found that traditional research criteria for diagnosing dementia based on that of Mesulam (1985) revealed fewer individuals with dementia than when caregivers were given structured questionnaires which included specific features of frontotemporal dementia (2000).

Continuum and Progression of Cognitive Impairment in ALS

The scientific evidence is clear that cognitive impairment is present in ALS, but findings in the literature are inconsistent relative to its prevalence and

degree of impairment. Several authors documented only subtle cognitive impairments in ALS (Gallassi et al., 1985, 1989; Rottig et al., 2006). Subgroups of individuals with ALS who have been described in the literature consist of participants who present with mild, moderate and severe cognitive impairments (Abe et al., 1997; Ringholz et al., 2005). It is possible to conceptualize a continuum of cognitive impairment in individuals with ALS that ranges from mild levels of impairment all the way to overt, progressive dementia (Abe et al., 1997; Strong et al., 2003).

Lomen-Hoerth and Strong (2006) suggest that distinctions should be made to subtype the spectrum of frontotemporal change in ALS. They suggest three possible subtypes. The first subtype is ALS with cognitive impairment (ALSci), describing those individuals with ALS who have deficits on one or more neuropsychological measure testing executive function. The second subtype is behaviourly impaired (ALSbi) describing those individuals with ALS who display behavioural signs consistent with frontal lobe impairment but do not meet criteria for dementia. The third subtype is ALS-FTD for those patients who meet criteria for dementia as determined by the Neary Criteria (Neary et al., 1998). Interestingly, in a study of 23 participants with ALS, Murphy and colleagues (2007) divided participants into the specific subtypes and found 5 patients who met criteria for FTLD (2 with FTD, 2 with SD and 1 with PA), 4 who met criteria for ALSbi, 2 with ALSci and one who met criteria for Alzheimer's disease.

Another important question arises when considering the progression of cognitive impairment. It is uncertain at present whether patients with ALSci

represent the first stages of ALS with dementia. Recent studies explored this possible relationship (Abrahams et al., 2005; Kilani et al., 2004; Robinson et al., 2006; Schrieber et al., 2005; Strong et al., 1999). Several of the authors showed that cognitive impairment exists early in the disease along with impairments in verbal fluency (Abrahams et al., 2005; Kilani et al., 2004; Schrieber et al., 2005; Strong et al., 1999). Strong and colleagues (1999) conducted a study over 6-months and determined that ALS participants had more cognitive differences when compared to controls at the second time period than at the initial time period. Robinson et al. (2006) and Abrahams et al. (2005) also conducted longitudinal studies over 6 month periods. Robinson et al. (2006) concluded that cognitive impairment in individuals with ALS can develop over 6 months across several domains and Abrahams and colleagues determined that cognitive impairment progresses slowly. These results were confirmed by two longer studies by Kilani et al. (2004) and Schreiber et al. (2005) who followed participants with ALS for 12 month and 18 month periods, respectively. They found a slow progression of cognitive impairment or no progression at all.

Language in ALS without Dementia

Language in individuals with ALS who do not show dementia has not been studied in as much detail as has their cognitive functioning. Despite this disparity, the emerging literature shows that the language of persons with ALS is affected (Abrahams et al., 2004; Cobble, 1998; Strong et al., 1999). Language in participants with ALS but without dementia has been tested primarily within the context of a large battery of neuropsychological tests. Tests used include verbal

fluency (i.e., both letter - FAS - and category - animals, vehicles, etc.), the Computerised Sentence Completion Test (Abrahams et al., 2000) based on Part 1 of the Hayling Sentence Completion Test (Burgess & Shallice, 1997), the Graded Naming Test (McKenna & Warrington, 1983), the Boston Naming Test (BNT) (Kaplan, Goodglass & Weintraub, 1983), the Peabody Picture Vocabulary Test 3rd Edition (PPVT-III)(Dunn & Dunn, 1997), the Token Test (De Renzi & Vignolo, 1962) and verbal IQ determined by the Wechsler Adult Intelligence Scale – Revised (Wechsler, 1981).

The most common language deficit exhibited by individuals with ALS but without dementia is impaired performance on tests of word retrieval (Abrahams et al., 2004; Abrahams et al., 2000; Mantovan et al., 2003; Racowicz & Hodges, 1998; Strong et al., 1999). Naming deficits occur in both category verbal fluency tasks (i.e., naming animals) and letter verbal fluency tasks (i.e., naming words that begin with the letter "F") (Abrahams et al., 2000; Hanagasi et al., 2002; Strong et al., 1999) along with problems with confrontation naming (Abrahams et al., 2004 ; Hanagasi et al., 2002; Mantovan et al., 2003 ; Ringholz et al., 2005 ; Strong et al., 1999). Further analyses of naming problems revealed verbal paraphasias (Strong et al., 1999) and semantic paraphasias (Mantovan et al., 2003; Strong et al., 1999). Patients also make category-coordinate semantic errors or circumlocutions (i.e., providing details about an item without naming it) (Racowicz & Hodges, 1998). Additionally, patients showed reduced single-word vocabulary comprehension (Strong et al., 1999) and a moderate auditory comprehension impairment detected on the Token Test (Mantovan et al., 2003).

Cobble (1998) completed a comprehensive test of language comparing 9 patients with ALS to 9 healthy controls. She determined, that in some patients with ALS, a subtle language impairment is present but is revealed only on formal testing. A subgroup of patients showed impairments in naming, in auditory comprehension of complex sentences, in some semantic tests and in spelling.

Abrahams and colleagues (2000) studied the underlying cause of word retrieval deficits in individuals with ALS. The participants with ALS in their study performed poorly on some tests of intrinsic response generation (i.e., Written Verbal Fluency Test and category fluency). However, the participants with ALS did not differ from controls on phonological loop functions as tested by the Phonological Similarities Effect and Word length effect. Furthermore, they did not differ from controls on simple word retrieval as tested by a sentence completion test and the Graded Naming Test. Therefore, Abrahams and colleagues (2000) concluded that deficits on tests of verbal fluency are the result of higher order executive dysfunction and are not caused by a dysfunction in either the phonological loop or the lower order linguistic processes of simple word retrieval. In a more recent study, Abrahams and colleagues (2004) further investigated the word retrieval deficits found in participants with ALS using MRI to determine whether word retrieval problems and associated cerebral deficits were present only in letter fluency or whether they also were present in confrontation naming. They found that cerebral deficits were not specific to letter fluency but also were present in confrontation naming tasks. They concluded that some patients with ALS may have a sub-clinical language impairment.

Two longitudinal studies were conducted in which researchers examined the progression of language impairment in participants with ALS but without dementia over the disease progression (Abrahams, 2005; Strong et al., 1999). Investigators in both studies collected data at baseline and then again 6 months later. Strong and colleagues (1999) did not find any significant differences in the language performances of their ALS participants. Abrahams and colleagues (2005), however, found evidence of significantly slower word retrieval times on the Computerised Sentence Completion test at the 6 month period for the participants with ALS versus the controls. The participants with ALS had longer retrieval times at the 6 month period whereas the controls had shorter retrieval times.

Few studies have examined the communication or the discourse of persons with ALS. Cobble (1998) indicated that the ALS participants in her study did not experience overt word finding difficulty in everyday communication. However, she did not formally test communication or discourse. Strong et al. (1999) systematically tested communication and discourse in ALS. Discourse samples were obtained using a topic-directed interview (TDI) protocol based on Ripich and Terrell (1989) at two time periods over 6 months. Results indicated that ALS patients produced significantly fewer self-corrected utterances than the controls at the 6 month period.

Language in ALS with Dementia

The most often reported symptom in participants with ALS and dementia is reduced verbal output which can lead to complete mutism (Bak et al., 2001,

2004; Neary et al., 1990). Reduced verbal output has been reported in individuals with both the behavioural variant of FTD and the progressive aphasia described in the literature. The reduced verbal output was reported in early literature by Van Bogaert (1932) and Ziegler (1930) and was described as "speechlessness" and "loss of mind and inability to speak" (as quoted in Bak & Hodges, 2001). Bak and Hodges (2004) noted that the reduction in spoken language shown by the ALS participants in their study occurred after the development of dysarthria and was, therefore, not attributable to effortful dysarthric speech. Additional characteristics of language impairment include perseverations, echolalia and the use of stereotypic expressions (Bak & Hodges, 2001; Racowicz & Hodges, 1998).

Authors of recent studies of individuals with ALS methodically tested language and provided information about the language profile of participants with ALS and dementia (Bak & Hodges, 1997, 2001, 2004; Bak et al., 2001; Racowicz & Hodges, 1998). Assessments of language included both letter and category fluency tests, administration of the Picture Naming Test and Word-picture matching test from Hodge's semantic battery (Hodges, Salmon & Butters, 1991), The Graded Naming Test (McKenna & Warrington, 1983), the Pyramids and Palm trees test: three picture version (Howard & Patterson, 1992), the Test for the Reception of Grammar (Bishop, 1989), the Kissing and Dancing Test (Bak & Hodges, 2003), a word comprehension test, the Token Test (De Renzi & Faglioni, 1978), and a sentence repetition test.

Participants with ALS and dementia displayed impairment in auditory comprehension of syntax (Bak & Hodges, 2004; Cavalleri & De Renzi, 1994; Racowicz & Hodges, 1998). Naming and comprehension of both nouns and verbs also were impaired; however, impairment in naming and understanding verbs was significantly worse than naming and understanding nouns (Bak & Hodges, 1997; Bak et al., 2001; Hillis, 2004; Racowicz & Hodges, 1998). Anomia and semantic paraphasias were thought to be associated with ALS-FTLD (Bak & Hodges, 1997; Neary et al., 1990; Racowicz & Hodges, 1998).

Neuropathological evidence supports the position that there are language deficits in individuals with ALS and dementia wherein language areas in the cortex are affected by the ALS (Bak et al., 2001). In studying the brain and spinal cord tissue of 4 individuals with ALS and dementia, investigators found cortical changes in the frontal lobe and the rostral temporal lobe comprising mild gliosis and microvacuolation of cortical laminae II and III. Broca's area, which is involved in language, also was affected (Bak et al., 2001).

In summary, it is evident that cognitive impairment consistent with frontal lobar dysfunction exists in individuals with ALS; however, the prevalence and degree of impairment remain unknown. A dementia consistent with a frontotemporal dementia also is associated with ALS and similarly the prevalence reported in the literature is inconsistent. It is possible that there exists a continuum of cognitive impairment in ALS ranging from mild cognitive impairment to an overt dementia. Findings to date indicate that cognitive impairment can worsen as the disease progresses; however, further investigation is required.

The literature on ALS also shows that language is affected in individuals with ALS without dementia and in those with ALS with dementia. Word retrieval deficits are the most common in individuals with ALS without dementia while reduced verbal output is the most often noted symptom among individuals with ALS with dementia. Even though language performances have been examined in ALS, further investigation is warranted in order to provide a complete profile of the language deficits in ALS, especially across its progression.

Statement of the Problem

Verbal fluency is the most common language deficit found in individuals with ALS without dementia and has been found to be impaired early in the disease (Strong et al.,1999; Schreiber et al., 2005). Several studies show significant impairment in letter and category fluency among individuals with ALS, however, the possibility of specific category deficits has not been explored. To date only one study has commented on specific categorical deficits (Rakowicz & Hodges, 1998). One participant with ALS/aphasia showed more difficulty naming living creatures than man-made objects in both a confrontation naming task and a picture/word matching task.

Neuropsychological studies showed that individuals with brain damage can show selective deficits regarding specific semantic categories (Basso, Capitani & Laiacona, 1988; De Renzi & Lucchelli, 1994; Warrington & Shallice, 1984). The most common category specific deficit mentioned in the literature is impairments in the retrieval of knowledge concerning living versus non-living items (Basso, Capitani & Laiacona, 1988; De Renzi & Lucchelli, 1994; Hillis &

Caramazza, 1991; Moss & Tyler, 1998; Warrington & Shallice, 1984). In most cases category specific impairments for living items are more prominent than non-living items (Basso, Capitani & Laiacona, 1988; De Renzi & Lucchelli, 1994; Warrington & Shallice, 1984), although the opposite also has been found (Hillis & Caramazza, 1991; Moss & Tyler, 1998; Sacchett & Humphreys, 1992). Category specific deficits in both recognition and naming occurred in patients with herpes simplex encephalitis (Warrington & Shallice, 1984) and Alzheimer's disease (Laws, Crawford, Gnoato, & Sartori, 2006). Additionally, category specific object recognition was shown in individuals with schizophrenia (Laws, Leeson & McKenna, 2006).

There are a number of theories in the literature that attempt to explain these category-specific deficits. Caramazza and Shelton (1998) claim that the domain-specific knowledge framework best accounts for category-specific deficits. The domain-specific knowledge framework suggests that the semantic system is organized categorically due to evolutionary pressures. These pressures created specialized mechanisms that distinguish animate and inanimate items perceptually and conceptually. Thus, this knowledge is organized categorically in the brain. Conversely, the sensory/functional hypothesis (Gainotti & Silveri, 1996; Warrington & Shallice, 1984) proposes that sensory properties are important in identifying living items whereas functional properties are important in identifying non-living items. The proponents of this theory believed that processing information for categories, which rely on different properties of each category, is located in different areas of the brain (Warrington

& Shallice, 1984). Another theory that attempts to explain category-specific deficits is the unitary distributed system in which concepts are represented as patterns of activation over multiple semantic properties. Category-specific deficits arise because of the differences in the structure and content of concepts and not because there are divisions of conceptual knowledge in separate areas of the brain (Tyler & Moss, 2001).

In accordance with the first two theories described above, lesion and neuroimaging studies reveal that selected brain regions can be more or less involved in processing concepts in different semantic domains (Damasio, Grabowski, Tranel, Hichwa, & Damsio, 1996; Martin, Wiggs, Ungerleider, & Haxby, 1996; Hillis & Caramazza, 1991). In a review of the literature, Humphreys and Forde (2001) determined that processing for living items is affected by lesions to inferior occipital-temporal regions extending anteriorly into the temporal lobe. They also showed that processing for non-living items is affected by lesions associated with left temporal-parietal and parietal-frontal areas. However, inconsistencies in these patterns do exist.

Investigations examining category specific impairments in all the different clinical manifestations of FTLD do not exist in the literature; however, category specific deficits have been reported in selected cases of semantic dementia (Cardebat, Demonet, Celsis & Puel 1996; Lambon Ralph, Patterson, Garrard, & Hodges, 2003). Cardebat et al. (1996) described an individual with semantic dementia who showed increased impairment for naming animals versus naming objects in confrontation tasks, word-to-picture matching tasks and picture

categorization tasks. Lambon and colleagues (2003) studied 6 individuals with mild to moderate semantic dementia and found 1 participant who showed a category specific impairment for living items versus non-living artifacts on both receptive and expressive tasks. Building on these findings, exploration of the category fluency patterns in individuals with ALS can provide additional insight into whether the fronto-temporal lobe involvement found in ALS may be causally related to category specific impairments for either living or non-living items.

To date only one study has examined the change in category fluency performances in individuals with ALS without dementia (Abrahams et al., 2005). No change was found over a 6 month period. However, slower word retrieval times were found after 6 months, suggesting that language can become vulnerable as ALS progresses to later stages. Therefore, studies that examine performances over periods longer than 6 months are necessary to provide crucial information about category fluency performance towards the end of the disease.

Finally, it has been noted that individuals with ALS and bulbar involvement exhibit substantial impairments in cognition. Verbal letter fluency was significantly more impaired in bulbar-onset ALS than spinal-onset ALS with the difference increasing over time (Schreiber et al., 2005). However, category fluency performance of individuals with bulbar-onset ALS in comparison to performance of individuals with spinal-onset ALS has yet to be explored.

Purpose and Research Questions

The purpose of this study was fourfold. The first was to examine category fluency performances in both bulbar-onset and limb-onset ALS participants to

determine if category specific impairments exist for living or non-living items. The second was to determine whether category fluency performances of individuals with ALS change over time. Thirdly, the performance of individuals with bulbar-onset ALS on category fluency tasks was determined and was compared to the performances of those with limb-onset ALS. Finally, the nature of the errors produced on category fluency tasks by the participants with ALS was investigated and compared to controls.

The following research questions were addressed in this study;

1) Do participants with ALS display category specific impairments for living versus non-living items on category fluency tasks in comparison to control participants at each of four assessment time periods?
2) How do category fluency performances of individuals with ALS change over time?
3) How do individuals with ALS with bulbar involvement perform on category fluency tasks in comparison to individuals with limb-onset ALS and how do their performances change over time?
4) What is the nature of the errors on category naming fluency tasks among participants with ALS versus controls participants?

Method

Participants

Participants in this study included individuals with clinically definite ALS as determined by the El Escorial criteria (World Federation of Neurology Research Group on Neuromuscular Disease, 1994) with disease duration less than one

year from the time of clinical onset. Age-matched controls also were recruited. They were either the spouses or the relatives of the participants with ALS.

ALS Participants

ALS participants were recruited from the Motor Neuron Diseases (MND) Clinic at London Health Sciences Center, University Campus and were selected by the director of the MND clinic, Dr. M.J. Strong. Exclusion criteria included a history of other medical conditions, such as exposure to heavy metals (e.g., lead, aluminum, mercury), hypothyroidism, hypoparathyroidism, pernicious anemia, porphyria, alcoholism, AIDS or AIDS-related complex, or history of other neurological disease or psychiatric illness. A convenience sampling technique was used. All individuals with ALS willing to participate in the study were accepted. Data were collected from all participants at 6 month intervals. The group of participants was relatively the same for each time period with the sample size decreasing over time.

At baseline, Time 1, there were 14 participants with classical sporadic ALS and 2 participants with familial ALS. Five participants presented with bulbar signs and symptoms at disease onset while 11 participants presented with either upper limb (n=6) or lower limb (n=3) signs and symptoms at disease onset. There were 10 men and 6 women whose ages ranged from 34 to 68 years (M= 52.8 ± 9.15). Their years of education ranged from 10 to 22 years (M=14.9 ± 3.22). Number of years of education was not obtained for 1 participant and therefore was not included in the calculations. See Appendix A for a summary of the demographics of ALS participants at Time 1. There were no significant group

differences in age (t = -.386, p = .705, df = 14) and education (t = .725, p = .481, df = 13) between the participants with bulbar-onset ALS and those with limb-onset ALS at Time 1.

At Time 2 (6 months after Time 1), there were 10 participants with classical sporadic ALS. Four participants presented with bulbar signs and symptoms at disease onset while 6 presented with either upper limb (n=4) or lower limb (n=2) signs and symptoms at disease onset. There were 7 men and 3 women whose ages ranged from 35 to 60 (M= 51.1 ± 7.78). Their years of education ranged from 10 to 22 years (M = 15.2 ± 3.36). See Appendix B for a summary of the demographics of ALS participants at Time 2. There were no group differences in age (t = 1.15, p = .284, df = 8) and education (t = .218, p = .833, df = 8) between participants with bulbar-onset ALS and those with limb-onset ALS at time 2.

At Time 3, there were 7 participants with classical sporadic ALS. However, due to a malfunction in the audio equipment the scores for one of the participants were not recorded and therefore her data are not included. Of the 6 participants included at this time period, 1 participant presented with bulbar signs and symptoms at disease onset while 5 presented with either upper limb (n=3) or lower limb (n=2) signs and symptoms at disease onset. There were 4 men and 2 women whose ages ranged from 35 to 61 years (51.3 ± 10.3). Their years of education ranged from 10 to 18 years (14.2 ± 2.86). See Appendix C for a summary of the demographics of ALS participants at Time 3. T-tests could not be run to determine if there were statistically significant differences in age and

education for the bulbar-onset participant versus the limb-onset participants as there was only 1 participant in the bulbar-onset group. The individual with bulbar-onset ALS was 61 years of age and had 10 years of education. The ages of the limb-onset group ranged from 35 to 59 years (M = 49.4 $\pm$ 10.3) and their years of education ranged from 12 to 18 years (M = 15 $\pm$ 2.24).

At Time 4, there were 4 participants with classical sporadic ALS. Two participants presented with bulbar signs and symptoms at disease onset and 2 presented with either upper limb (n=1) or lower limb (n=1) signs and symptoms at disease onset. Two of the 4 participants were women. The ages of the 4 participants ranged from 44 to 61 years (M = 55.8 ± 7.89). Their years of education ranged from 10 to 18 years (M = 14.5 ± 3.42). See Appendix D for a summary of the demographics of ALS participants at Time 4. T-tests could not be run to determine if there were statistically significant differences in age and education for the bulbar-onset participant versus the limb-onset participants as there was only 1 participant in the bulbar-onset group. The individual with bulbar-onset ALS was 61 years of age and had 10 years of education. The ages of the limb-onset group ranged from 44 to 59 years (54 $\pm$ 8.66) and their years of education ranged from 14 to 18 years (16 $\pm$ 2.00).

At the final time period, Time 5, there were 2 participants with classical sporadic ALS. One participant presented with bulbar signs and symptoms at disease onset while the other participant presented with upper limb signs and symptoms at disease onset. They are both men and their mean age was 52 years ($\pm$12.7) years and their mean years of education was 12 years ($\pm$ 2.83).

See Appendix E for a summary of the demographics of ALS participants at Time 5. Table 1 contains summary demographic data for all ALS participants at each time period. T-tests could not be run to determine if there were statistically significant differences in age and education for the non-bulbar participant versus the limb-onset participant as there was only 1 participant in each group. The individual with bulbar- onset ALS was 62 years of age and had 10 years of education where as the participant with limb-onset ALS was 44 years old and had 14 years of education.

Control Participants

At Time 1, there were 7 men and 5 women control participants whose ages ranged from 34 to 63 years (M= 53.3 ± 8.03). Their years of education ranged from 10 to 19 years (M = 13.4 ± 2.60). See Appendix A for a summary of the demographic characteristics of control participants. There were no significant group differences in age (t = -.176, p = .862, df = 26) and education (t = 1.358, p = .187, df = 25) between the ALS and the control participants at Time 1.

At Time 2, there were 4 men and 3 women control participants. Their ages ranged from 45 to 63 years (M = 57.1 ± 3.93) and their years of education ranged from 10 to 18 years (M = 13.8 ± 2.67). See Appendix B for a summary of the demographic characteristics of control participants at Time 2. There were no significant group differences in age (t = - 1.881, p = .080, df = 15) and education (t = .925, p = .370, df = 15) between the ALS and control participants at Time 2.

Table 1

Summary Demographic Information for ALS and Control Participants at Times 1, 2, 3, 4 and 5

	Time[a]				
Measure	1	2	3	4	5
			ALS		
Sex	M=9 F=7	M = 7 F = 3	M = 4 F = 2	M = 2 F = 2	M = 2
Age of onset (yrs)	50.8 (9.65)	48.3 (8.69)	45.7 (11.5)	51.3 (8.30)	49 (12.73)
Age at testing (yrs)	52.8 (9.15)	51.1 (7.78)	51.3 (10.3)	55.8 (7.89)	53 (12.73)
Ed (yrs)	14.9 (3.22)	15.2 (3.36)	14.2 (2.86)	14.5 (3.42)	12 (2.83)
Site of onset	B=5, LL=4, UL=7	B=4, LL=2, UL=4	B=1, LL=2, UL=3	B=1, LL=1, UL=2	B=1, UL=1
Familial (F) /Sporadic (S)	F=2, S=14	S =10	S =6	S =4	S=2
			Control		
Sex	M = 7 F = 5	M = 4 F = 3	M = 4 F = 2	M = 2 F = 1	F = 1
Age (yrs)	53.3 (8.03)	57.1 (3.93)	56.7 (3.88)	59.3 (4.51)	
Ed (yrs)	13.4 (2.60)	13.8 (2.67)	13.1 (2.10)	14.7 (1.15)	

Note. B = Bulbar. LL = Lower Limb. UL = Upper Limb. [a]The difference between each time period is 6 month

At Time 3, there were 4 men and 2 women control participants. Their ages ranged from 54 to 64 years (M = 56.7 ± 3.88) and their years of education ranged from 10 to 16 years (M = 13.1 ± 2.1). Levene's test revealed violation of homogeneity of variances on measures of age (F=5.58, p=0.04) therefore, unequal variances were assumed when the t-test was performed. There were no significant group differences in age (t = -1.184, p = .264, df = 10) and education (t = .747, p = .474, df = 9.2) between the ALS and control participants at Time 3.

At Time 4, there were 2 men and 1 woman control participants whose ages ranged from 55 to 64 years (M = 59.3 ± 4.51). Their years of education ranged from 14 to 16 years (M = 14.7 ± 1.15). There were no significant group differences in age (t = -.696, p = .518, df = 5) and education (t = -.080, p = .940, df = 5) between the ALS and control participants at Time 4.

At Time 5 there was only one control participant. She was a woman who was 55 years of age and had 14 years of education. Table 1 contains summary demographic data for all control participants at each time of testing. T-tests could not be run to determine if there statistically significant differences in age and education for the control participant versus the participants with ALS as there was only 1 control participant and 2 participants with ALS. The participants with ALS were 44 and 62 years of age and had 14 and 10 years of education, respectively.

Procedure

The current study of generative naming in ALS was performed as part of a larger, multidisciplinary study examining the cognitive and language impairments

in individuals with ALS over time. ALS participants underwent neuropsychological testing, language and discourse testing, neuromotor/physiotherapy/pulmonary testing and perfusion computerized transaxial tomography (CT perfusion) at the St. Joseph's Health Care – St. Joseph's Hospital in London Ontario. Participants were tested at 5 different time periods; baseline (Time 1) and every six months thereafter over a 24 month interval. The same procedure was used at each time period and was performed in the same order with minor changes due to scheduling conflicts.

A comprehensive set of standardized and non-standardized language measures, non-standardized discourse tasks and questionnaires on pragmatics was administered. Testing was completed in approximately 1 to 1.5 hour sessions. All language and discourse testing was administered by the same individual and performed in the same order across all time periods with very few changes in the order of test administration. Participants were provided with rest breaks throughout the testing. All language and discourse measures were video recorded in a quiet room in the ALS Clinic on the seventh floor at the London Health Sciences Centre, University Hospital, London Ontario. Participants were asked to respond verbally unless they were unable to as a result of severe dysarthria or anarthria. Written responses were accepted when participants were anarthric. Only the procedures for the language measures, and not the discourse tasks, are described below because the language measures are central to this current study.

Standardized language tests included *The Peabody Picture Vocabulary*

test –III (PPVT-III) (Dunn & Dunn, 1997) and selected subtests from the *Arizona Battery for Communication Disorders (ABCD)* (Bayles & Tomoeda, 1991). Non-standardized measures include *The Action Naming* test (Obler & Martin, 1982), and category word fluency tasks (Rakowicz & Hodges, 1998). The *PPVT-III* and the *ABCD* possess strong reliability and validity scores. The *PPVT-III* has high internal consistency reliability with reliability coefficients of .92 to .98 and by split-half reliabilities ranging from .86 to .97. Additionally, the *PPVT-III* has high alternate forms reliability (.88 to .96), and test-retest reliability (.90 and above). The *PPVT-III* also has strong criterion validity shown by high correlations with the *Weschler Intelligence Scale for Children-Third Edition* (.82 to .92), the *Kaufman Adolescent and Adult Intelligence Test* (.76 to .91), the *Kaufman Brief Intelligence Test* (.62 to .82) and the *Oral and Written Language Scales* (.63 to .83). The *ABCD* has strong test-retest reliability revealed by the high probability of concordance for most subtests (.55 to .87), as well as strong internal consistency for most subtests (Cronbach's alpha of .5017 to .9853). Furthermore, it has strong criterion validity with high correlations to three well known measures of dementia severity: the *Global Deterioration Scale* (.63 to .82), the *Mini-Mental State Examination* (.62 to .85) and the Block Design subtest of the *WAIS-R* (.59 to .74). The *ANT* is a non-standardized test of confrontation naming of verbs with no published psychometric properties.

For the purpose of this study the procedure and analyses only involving the category word fluency tasks will be discussed in detail because they are the primary outcome measures. Category word fluency tasks used in this study

included 4 categories of living items (i.e., animals, birds, dogs, and water creatures) and 4 categories of non-living items (i.e., household items, vehicles, musical instruments, and tools). These categories are based on an adapted version of the Hodges, Salmon and Butters (1992) category fluency task conducted with individuals with Alzheimer's disease. All ALS and control participants were asked to generate verbally as many items as they could in one minute for each of the 8 categories. If participants were unable to respond verbally they were asked to write their responses. They were given an extra 30 seconds to account for writing time. Two participants provided written response at Time 3, one participant provided written responses at Time 4 and one participant provided written responses at Time 5. Number of items, number of errors, and the number of error types were recorded for each category for each participant at all Time periods.

Speech intelligibility and speech rate were rated by a group of three untrained listeners for each participant at each time period. Ratings were completed to determine the extent of motor speech problems due to dysarthria. Motor speech problems could impact language output and category fluency scores. A visual analogue scale (VAS) comprising a 100 mm line with anchors of "Completely Intelligible" to "Completely Unintelligible" was used to measure the participants' intelligibility at each time period (See Appendix F for the VAS scale of speech intelligibility). Additionally, a VAS comprising a 100 mm line with anchors of "Very Slow" to "Very Fast" with normal as the centre (i.e., 50 mm) was used to measure the participants' rate at each time period (See Appendix G the

VAS scale of rate).

The three raters of participants' speech samples were blinded to the objectives of the study and to the nature of the participants' diagnosis (i.e., ALS or control). They included 2, second-year and 1, first-year speech-language pathology graduate students from the University of Western Ontario. The three listeners rated the participants' speech intelligibility and speech rate using the visual analogue scales. They listened to a digitized audio sample of each participant's speech taken from the Topic Directed Interview task video recorded at each of the time periods. There were 60 samples in total. Samples were not evaluated for ALS participant 13 at Times 1 and 2, for ALS participant 11 at Times 3, 4 and 5, for control participant 9 at Time 3 and for control participant 10 at Time 3. ALS participant 13 did not complete the TDI task at Time 1 and wrote his responses at Time 2. ALS participant 11 wrote his responses at Times 3, 4 and 5. Due to malfunctions in the audio equipment, samples for control participants 9 and 10 at Time 3 could not be obtained. Ninety-three percent of the samples were 30 seconds long, 5% were less than 30 seconds and 2% were over 30 seconds long. The speech samples were presented to the three raters in a randomized order. It took the listeners between one hour and 6 minutes and one hour and 24 minutes to complete the task.

In order to determine intra-rater percent agreement each listener re-evaluated 10 speech samples for both intelligibility and rate. Agreement was calculated by determining the percentage of samples in which the score was within $\pm$ 10 mm of the original score given. Intra-rater percent agreement for

listener 1 was 70% for intelligibility and 70% for rate. Intra-rater percent agreement for listener 2 was 90% for intelligibility and 100% for rate. Intra-rater percent agreement for listener 3 was 90% for intelligibility and 100% for rate. Inter-rater reliability was determined by performing an intraclass correlation using SPSS 16.0 for windows. Inter-rater reliability was strong for both intelligibility (.875) and rate (.972). Intelligibility and rate scores were determined for each participant by calculating the mean scores given by the three listeners. Group comparisons were made to determine if there were differences in rate and intelligibility scores between the ALS participants and controls and between the ALS sub-groups (i.e., bulbar-onset vs. limb-onset). The results of these comparisons are reported at the beginning of the Results section.

Data Analyses

The words generated for the category fluency tasks were transcribed orthographically from the video recordings for all of the sessions for each participant. There was a malfunction in the audio equipment for two sessions at Time 3; therefore the data for these participants were obtained from the examiner's written records of the participants' responses. The raw scores for the number of correct items and the number of errors were recorded for each of the 8 sub-categories (i.e., animals, household items, birds, vehicles, water creatures, musical instruments, dogs and tools) for each participant at all time periods. See Appendices H, I, L, M, and N for the raw scores showing the number of correct items in the sub-categories for individual participants with ALS at Time 1, 2, 3, 4, and 5, respectively. See Appendices J, K, L, M and N for the raw scores

showing the number of correct items in the sub-categories for individual control participants at Time 1, 2, 3, 4 and 5, respectively. Unintelligible items were not included in the number of correct or incorrect items. There were no unintelligible items produced at Time 1, 4, or 5. There were 10 unintelligible items at Time 2 out of a total of 2227 items. ALS participant 12 produced nine unintelligible items that were spread fairly evenly across all sub-categories, with no more than two in one sub-category. ALS participant 6 generated one unintelligible item in the "dogs" sub-category. There was one unintelligible item at Time 3 out of a total of 1553 items. Control participant 9 produced an unintelligible item in the "dogs" sub-category.

Research Question 1

Research question one addressed whether individuals with ALS have category specific impairments for living and non-living items in comparison to control participants at Times 1, 2, 3, and 4. A repeated measures ANOVA was conducted with group (ALS vs. Control) as the between subject variable and number of items within a category (living vs. non-living) as the within subject variable at each of the first four time periods. An additional repeated measures ANOVA was conducted for each of the first four time periods to determine whether there were differences between or within groups for any of the 8 sub-categories. Group (ALS vs. Control) was the between group variable and sub-category (animals, household items, birds, vehicles, water creatures, musical instruments, dogs and tools) was the within group variable.

Research Question 2

Question 2 addressed how category fluency performances of individuals with ALS change with disease progression in comparison to controls. A regression analysis described by Kenny, Boger and Kashy (2002) was conducted to determine whether there were any between and within group differences in the rate of change in generative naming performance over time. Kenny et al. (2002) suggest using a regression analysis on each individual participant who participated at two or more time periods to determine slope values for each participant. The slope values from the within participant regression analyses indicate the individual rates of change in naming performance over time. A repeated measures ANOVA on the slope values for each participant was conducted to assess between and within group differences in rates of change. Group (ALS vs. Control) was the between subjects factor and the category (living vs. non-living) was the within subjects factor. An additional repeated measures ANOVA was conducted on the slope values with group (ALS vs. Control) as the between subjects factor and the sub-categories (animals, household items, birds, vehicles, water creatures, musical instruments, dogs, tools) as the within subjects factor.

Research Question 3

Research question 3 addressed whether there were differences in category naming of living or non-living items between the bulbar-onset ALS and limb-onset ALS groups and how the groups' performances change over time. A repeated measures ANOVA was conducted for each of the first two time periods

to determine whether there were any between or within group differences in the number of living and non-living items generated. Group (bulbar-onset vs. limb-onset) was the between subject variable and number of items within a category (living vs. non-living) was the within subject variable. An additional repeated measures ANOVA was conducted for each of the first two time periods to determine whether there were differences between or within groups for any of the 8 sub-categories. Group (bulbar-onset vs. limb-onset) was the between group variable and sub-category (animals, household items, birds, vehicles, water creatures, musical instruments, dogs and tools) was the within group variable. A repeated measures ANOVA was conducted on the slope values from the regression analysis (Kenny et al., 2002) to determine whether there were any between and within group differences in the rate of change in generative naming performance over time. Group (bulbar-onset vs. limb-onset) was the between subjects factor and the category (living vs. non-living) was the within subjects factor. An additional repeated measures ANOVA was conducted on the slope values with group (bulbar-onset vs. limb-onset) as the between subjects factor and the sub-categories (animals, household items, birds, vehicles, water creatures, musical instruments, dogs, tools) as the within subjects factor.

Significance Levels for Research Questions 1, 2, and 3

This is an exploratory study of individuals with ALS; therefore, for all repeated measures ANOVAs the alpha level was set at 0.05 to indicate statistical significance. Values between 0.05 and 0.10 were regarded as approaching significance. It is not uncommon in pilot studies to consider p-values that

approach significance, especially with small samples in clinical populations with high variability.

Research Question 4

Research question 4 addressed the nature of the errors made by the participants. All responses for each sub-category by each participant at all time periods were analyzed. Three examiners (EC, her advisor JBO, and post-doctoral fellow LJW) rated whether responses were correct items within each of the respective sub-categories. Items were considered errors by 2/3 agreement. The raters then sorted the errors according to the following error types: repetitions, phonemic/literal, verbal, semantically related, supraordinate semantically related, confabulations,whole/part and trade name. These error patterns are used commonly in studies of anomia. Error responses were considered to be a given error type with 2/3 agreement among the raters. Please see Appendix O for definitions of the error types. Calculations of the means, standard deviations, standard errors, and 95% confidence intervals were performed to determine significant differences between groups in percentage of errors produced and in types of errors produced.

Results

Intelligibility and Rate

Group comparisons were performed to determine if there were differences in mean intelligibility and rate scores between participants with ALS and controls. The means, standard deviations, minimum and maximum values for Time 1, 2, 3 and 4 are presented in Table 2. Recall that for intelligibility the anchor score of 0

Table 2

Intelligibility and Rate Measures for ALS and Control Participants at Times 1, 2, 3, and 4.

	ALS					*Control*				
Measure	*n*	*M*	*SD*	*Min.*	*Max.*	*n*	*M*	*SD*	*Min.*	*Max.*
					Time 1					
Intelligibility	15	22.24	21.47	1.33	71.67	12	2.64	2.39	0.33	6.67
Rate	15	34.40	8.67	16.00	48.67	12	45.42	5.89	35.33	52.00
					Time 2					
Intelligibility	9	32.30	32.15	0.33	82.33	7	2.43	2.00	0.00	5.00
Rate	9	34.81	11.12	17.00	54.33	7	45.57	4.63	40.67	53.67
					Time 3					
Intelligibility	5	14.13	13.24	3.33	34.67	4	2.42	1.10	1.33	34.67
rate	5	35.80	8.02	26.67	45.00	4	46.92	5.10	42.00	45.00
					Time 4					
Intelligibility	3	34.00	50.25	3.67	92.00	3	0.78	0.51	0.33	1.33
Rate	3	33.00	22.59	8.33	52.67	3	45.89	2.99	44.00	49.33

Note. For intelligibility the anchor score 0 means "Completely Intelligible" while the anchor score 100 means "Completely Unintelligible". For rate the anchor score 0 means "Very Slow" while the anchor score 100 means "Very Fast".

on the VAS means "Completely Intelligible" while the other anchor score of 100 on the VAS means "Completely Unintelligible". Also, for rate, the anchor score of 0 on the VAS means "Very Slow" while the other anchor score of 100 on the VAS means "Very Fast" with the centre point (i.e., 50) as normal. Levene's test revealed violation of homogeneity of variances for the intelligibility measure at Time 1($F(1, 25) = 15.28$, $p = 0.001$), Time 2 ($F(1, 14) = 40.50$, $p = 0.00$), Time 3 ($F(1, 7) = 8.90$, $p = 0.02$) and Time 4 ($F(1, 4) = 15.60$, $p = 0.017$), therefore non-parametric Mann-Whitney U tests were conducted. There were significant differences between intelligibility scores at Time 1 ($U = 22.50$, $Z = -3.296$, $p = 0.001$), Time 2 ($U = 12.0$, $Z = -2.064$, $p = 0.039$), Time 3 ($U = 1.50$, $Z = -2.091$, $p = 0.037$) and Time 4 ($U = 0.0$, $Z = -1.964$, $p = 0.05$). The ALS group had higher scores on the intelligibility measure at all time periods indicating that the group was more unintelligible than controls. One-way ANOVAs with rate as the between group variable were conducted and revealed that there were significant differences between the participants with ALS and controls at Time 1 ($F(1) = 14.10$, $p = 0.001$), Time 2 ($F(1) = 5.78$, $p = 0.032$) and Time 3 ($F(1) = 5.73$, $p = 0.048$). At all three time periods the ALS group had lower rate scores than controls. There were no significant differences in rate between ALS and control participants at Time 4 ($F(1) = 0.960$, $p = 0.383$). A correlational analysis using a Pearson Product Moment correlation also was conducted to determine if intelligibility and rate scores were correlated with the total number of items generated. The correlational analysis was conducted using scores for both participants with ALS and controls across all time periods. There was a small,

non-significant correlation between intelligibility and the total number of items generated for participants with ALS and controls (-0.113). There was a small, non-significant correlation between rate scores and the total number of items generated (0.167).

Group comparisons also were performed to determine if there were differences in mean intelligibility and rate scores between the ALS subgroups (i.e., bulbar-onset vs. limb-onset). One-way ANOVAs were conducted and revealed that there were no significant differences in intelligibility scores between the bulbar-onset and limb-onset groups at Time 1 ($F(1) = 1.44$, $p = 0.251$). There were significant differences in intelligibility scores at Time 2 ($F(1) = 18.38$, $p = 0.004$). The bulbar- onset group had higher intelligibility scores than the limb-onset group. Additional one-way ANOVAs with rate as the between group factor were conducted and revealed that there were no significant differences between bulbar-onset and limb-onset groups at Time 1 ($F(1) = 4.25$, $p = 0.06$) or Time 2 ($F(1) = 2.80$, $p = 0.138$).

Research Question 1

Research question 1 addressed whether individuals with ALS have category specific impairments for living or non-living items in comparison to control participants at Times 1, 2, 3, and 4. A repeated measures ANOVA was conducted for each of the first four time periods to determine whether there were any between or within group differences in the number of living and non-living items generated. Group (ALS vs. Control) was the between subject variable and number of items within a category (living vs. non-living) was the within subject

variable. The means, standard deviations, minimum and maximum values for Time 1, 2, 3 and 4 are outlined in Tables 3, 4, 5, and 6, respectively. There was no significant group by category interaction at Time 1 ($F(1, 26) = .99$, $p = 0.33$), Time 2 ($F(1, 15) = .49$, $p = 0.497$), Time 3 ($F(1, 10) = .43$, $p = 0.881$) or Time 4 ($F(1, 5) = .00$, $p = 0.962$). There was no main effect of group at Time 1 ($F(1, 26) = .06$, $p = 0.807$), Time 2 ($F(1, 15) = .25$, $p > 0.622$), Time 3 ($F(1, 10) = .024$, $p > 0.05$) or Time 4 ($F(1, 5) = .76$, $p = 0.423$). There was no main effect of category (living or non-living) for either the ALS or control group at Time 1 ($F(1, 26) = .03$, $p = 0.857$), Time 2 ($F(1, 15) = .11$, $p = 0.746$), Time 3 ($F(1, 10) = 1.2$, $p = 0.299$) or Time 4 ($F(1, 5) = 1.14$, $p = 0.335$).

An additional repeated measures ANOVA was conducted for each of the first four time periods to determine whether there were differences between or within groups for any of the 8 sub-categories. Group (ALS vs. Control) was the between group variable and sub-category (animals, household items, birds, vehicles, water creatures, musical instruments, dogs and tools) was the within group variable. There was no significant group by category interaction at Time 1 ($F(7, 182) = .78$, $p = 0.601$), Time 3 ($F(7, 70) = 1.02$, $p = 0.425$) or Time 4 ($F(7, 35) = .06$, $p = 1.0$). However, there was a significant group by category interaction at Time 2 ($F(7, 105) = 3.19$, $p = 0.004$). Figure 2 shows that the control group produced more correct items in the household items category than the ALS group. Additionally both groups produced more items for the animals and the household items categories than for the other 6 sub-categories. There

Table 3

Category fluency scores for ALS and Control Participants at Time 1

	ALS					*Controls*				
Categories	*n*	*M*	*SD*	*Min.*	*Max.*	*n*	*M*	*SD*	*Min.*	*Max.*
Animals	16	20.38	6.79	9	30	12	19.00	4.77	7	25
Household items	16	21.88	7.74	7	37	12	22.42	4.27	16	31
Birds	16	16.13	5.54	6	25	12	15.25	4.16	9	22
Vehicles	16	12.69	4.63	7	20	12	12.25	3.72	6	18
Water creatures	16	12.94	5.71	5	25	12	13.17	3.01	8	16
Musical instruments	16	14.94	5.48	4	24	12	13.25	3.62	7	20
Dogs	16	13.56	5.90	3	28	12	12.33	3.31	7	18
Tools	16	12.06	5.11	5	23	12	13.92	3.23	8	19
Living items	16	63.00	21.67	31	107	12	59.75	9.63	44	73
Non-living items	16	61.56	18.29	28	90	12	61.83	8.77	46	76
Total all categories	16	124.56	39.22	59	197	12	121.58	16.37	90	147

Table 4

Category fluency scores for ALS and Control Participants at Time 2

	ALS					*Controls*				
Categories	*n*	*M*	*SD*	*Min.*	*Max.*	*n*	*M*	*SD*	*Min.*	*Max.*
Animals	10	19.90	5.90	12	30	7	22.14	4.30	16	28
Household items	10	20.30	5.77	11	28	7	26.00	3.51	20	30
Birds	10	14.50	5.25	7	23	7	16.29	2.69	13	20
Vehicles	10	13.70	6.00	7	24	7	14.57	0.98	13	16
Water creatures	10	14.70	5.77	6	22	7	14.00	3.37	8	17
Musical instruments	10	14.40	4.35	7	20	7	12.43	3.36	7	17
Dogs	10	13.80	4.83	6	21	7	12.86	1.86	10	16
Tools	10	12.50	4.03	5	19	7	13.00	3.74	9	18
Living items	10	62.90	20.41	31	94	7	65.29	8.18	53	77
Non-living items	10	60.90	18.01	34	87	7	66.00	6.43	56	75
Total all categories	10	123.80	37.83	65	180	7	131.3	11.66	117	144

Table 5

Category fluency scores for ALS and Control Participants at Time 3

	ALS					*Controls*				
Categories	*n*	*M*	*SD*	*Min.*	*Max.*	*n*	*M*	*SD*	*Min.*	*Max.*
Animals	6	19.83	5.42	12	27	6	22.17	6.73	16	32
Household items	6	21.0	8.10	13	32	6	23.83	4.22	16	28
Birds	6	16.33	7.26	9	27	6	14.50	2.43	12	19
Vehicles	6	14.0	3.79	10	20	6	12.33	3.98	7	17
Water creatures	6	14.67	7.47	6	28	6	15.0	3.52	11	21
Musical instruments	6	15.17	6.05	8	25	6	13.0	4.65	8	19
Dogs	6	14.5	6.72	3	20	6	11.67	3.92	7	18
Tools	6	12.0	3.74	8	17	6	12.83	4.12	7	17
Living items	6	65.50	21.92	37	94	6	62.83	10.03	52	77
Non-living items	6	62.17	18.86	40	90	6	62.0	11.40	48	75
Total all categories	6	127.50	40.44	77	184	6	124.8	20.51	100	145

Table 6

Category fluency scores for ALS and Control Participants at Time 4

	ALS					*Controls*				
Categories	*n*	*M*	*SD*	*Min.*	*Max.*	*n*	*M*	*SD*	*Min.*	*Max.*
Animals	4	16.75	9.74	7	30	3	19.0	5.67	13	24
Household items	4	20.50	7.14	16	31	3	24.0	2.65	22	27
Birds	4	14.25	6.75	8	23	3	16.67	5.77	10	20
Vehicles	4	12.75	6.45	7	22	3	15.33	2.08	13	17
Water creatures	4	11.50	4.73	8	18	3	15.67	3.79	13	20
Musical instruments	4	11.75	4.57	7	18	3	15.33	4.16	12	20
Dogs	4	9.75	3.95	4	13	3	13.0	3.61	10	17
Tools	4	11.50	7.05	7	22	3	14.33	3.79	10	17
Living items	4	52.25	19.97	39	82	3	64.33	14.57	54	81
Non-living items	4	56.50	24.89	37	93	3	69.0	10.54	58	79
Total all categories	4	108.75	44.81	76	175	3	133.33	19.76	112	151

was a main effect of category at Time 1 ($F(7, 182) = 29.08$, $p = 0.000$), Time 3 ($F(7, 70) = 11.784$, $p = 0.000$) and Time 4 ($F(7, 35) = 6.452$, $p = 0.000$). Figures 1 and 3 show that a greater number of items were produced for the animals and household items categories than for the other 6 sub-categories for each group at Times 1 and 3. Figure 4 shows that a greater number of items were produced in the household items category at Time 4. There was no significant main effect of group at Time 1 ($F(1,26) = .061$, $p = 0.807$), Time 3 ($F(7, 10) = .02$, $p = 0.888$) or Time 4 ($F(7, 5) = .76$, $p = 0.423$).

The data in the figures are based on estimated marginal means (EMM). An EMM is an estimate of the population marginal mean of the dependent variables. Population marginal means are parametric functions which calculate means based on parameters of a linear model. They do not always equal the observed means (Searle, Speed & Milliken, 1980). These were used in the figures because these are the means used by SPSS when conducting a repeated measures ANOVA. The EMMs in this study are very similar to the observed means.

Sub-Analysis Comparing ALS Outliers to Control Participants

Box plots were used to examine patterns in the naming data. Figures 5, 6 and 7 show the ALS group had considerably larger variability in scores on the living, non-living and total of all categories measures at Times 1, 2, 3 and 4. Furthermore, it was noted that at Time 1 there were several participants with ALS who produced a number of total items that fell far below the mean.

Low performers. Based on visual inspection of the box plots, the ALS group was

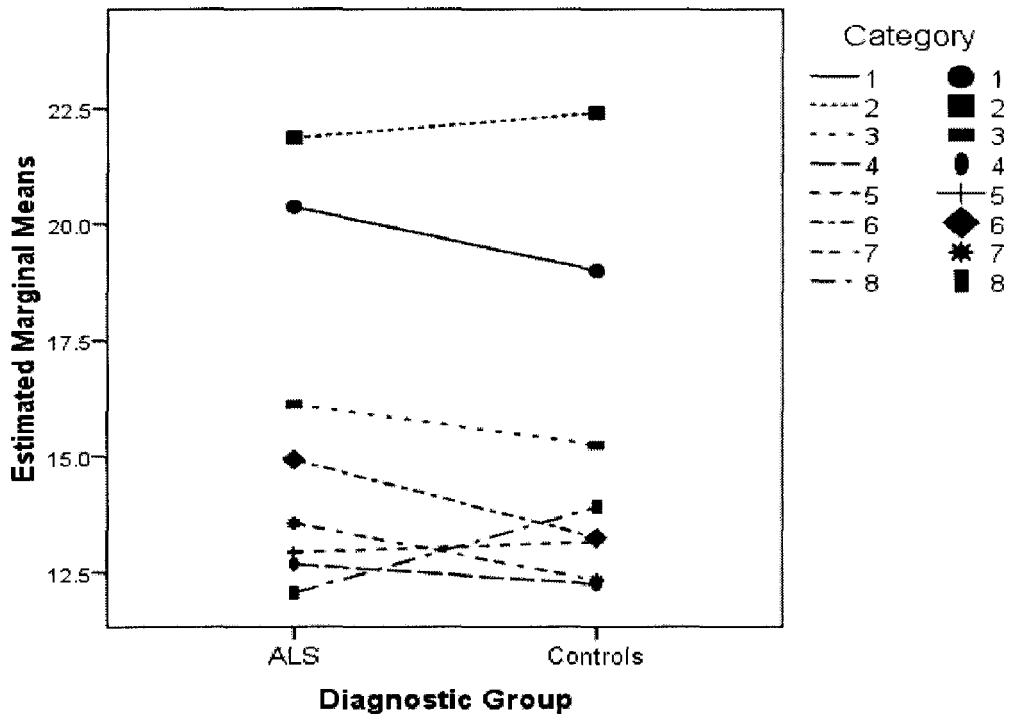

Figure 1. Estimated marginal means (EMM) by group for each category at Time 1. (1 = animals; 2 = household items; 3 = birds; 4 = vehicles; 5 = water creatures; 6 = musical instruments; 7 = dogs; 8 = tools)

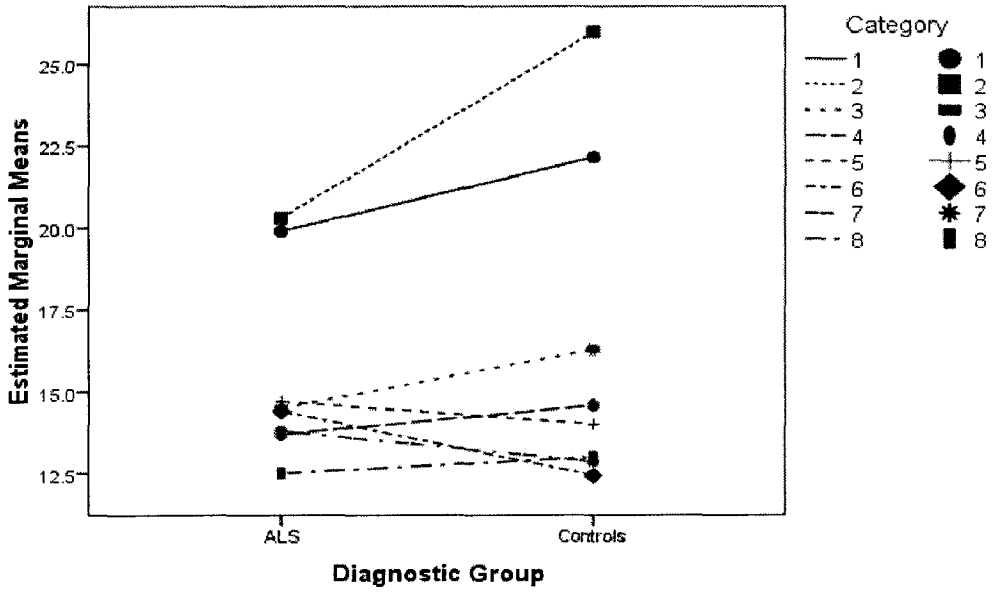

Figure 2. EMM by group for each category at Time 2. (1 = animals; 2 = household items; 3 = birds; 4 = vehicles; 5 = water creatures; 6 = musical instruments; 7 = dogs; 8 = Tools)

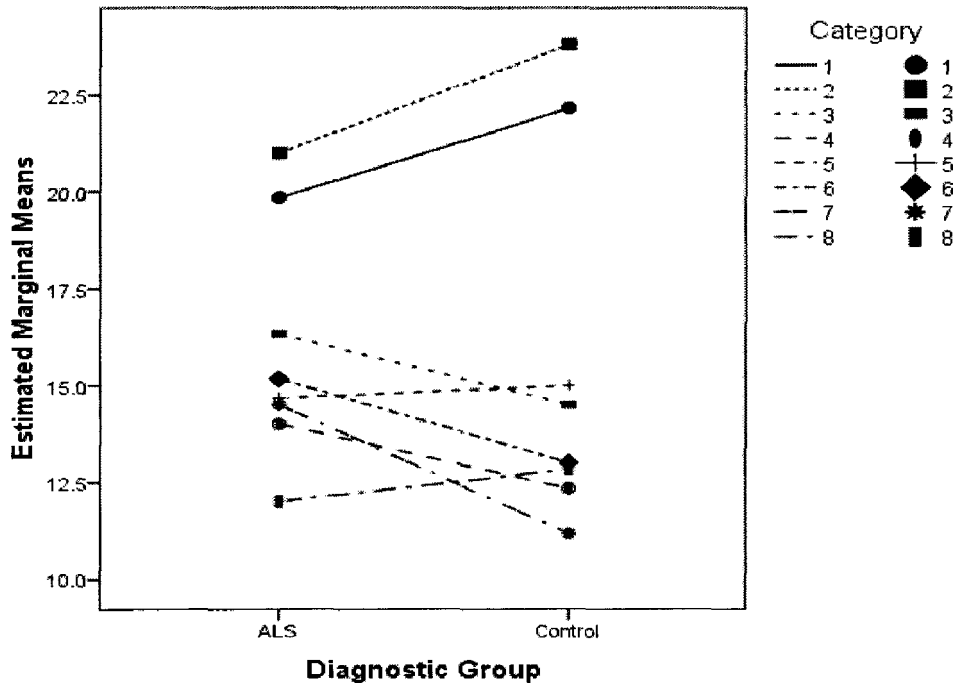

Figure 3. EMM by group for each category at Time 3. (1 = animals; 2 = household items; 3 = birds; 4 = vehicles; 5 = water creatures; 6 = musical instruments; 7 = dogs; 8 = tools)

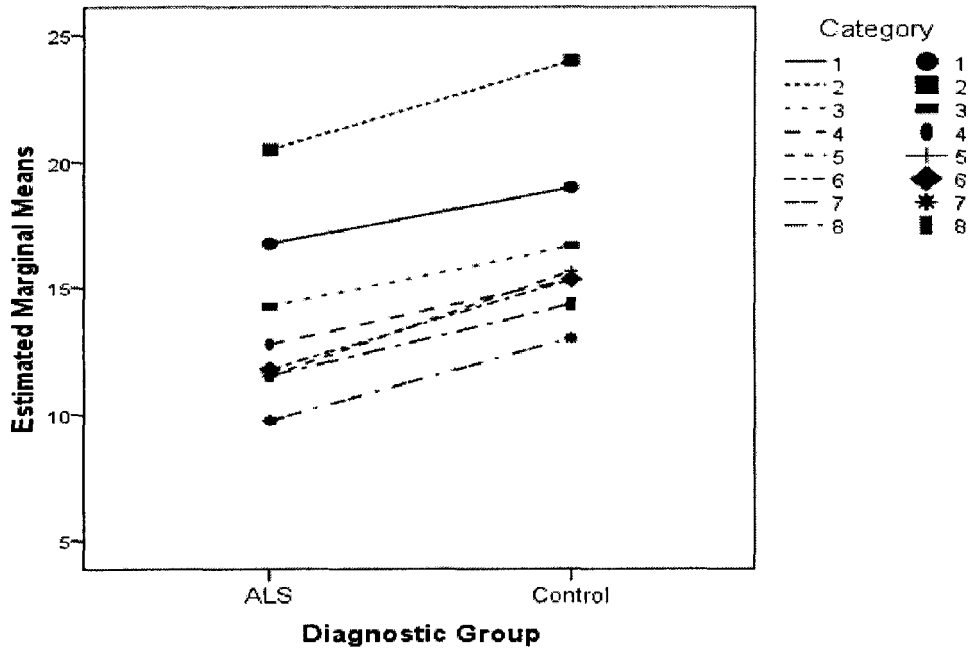

Figure 4. EMM by group for each category at Time 4. (1 = animals; 2 = household items; 3 = birds; 4 = vehicles; 5 = water creatures; 6 = musical instruments; 7 = dogs; 8 = tools)

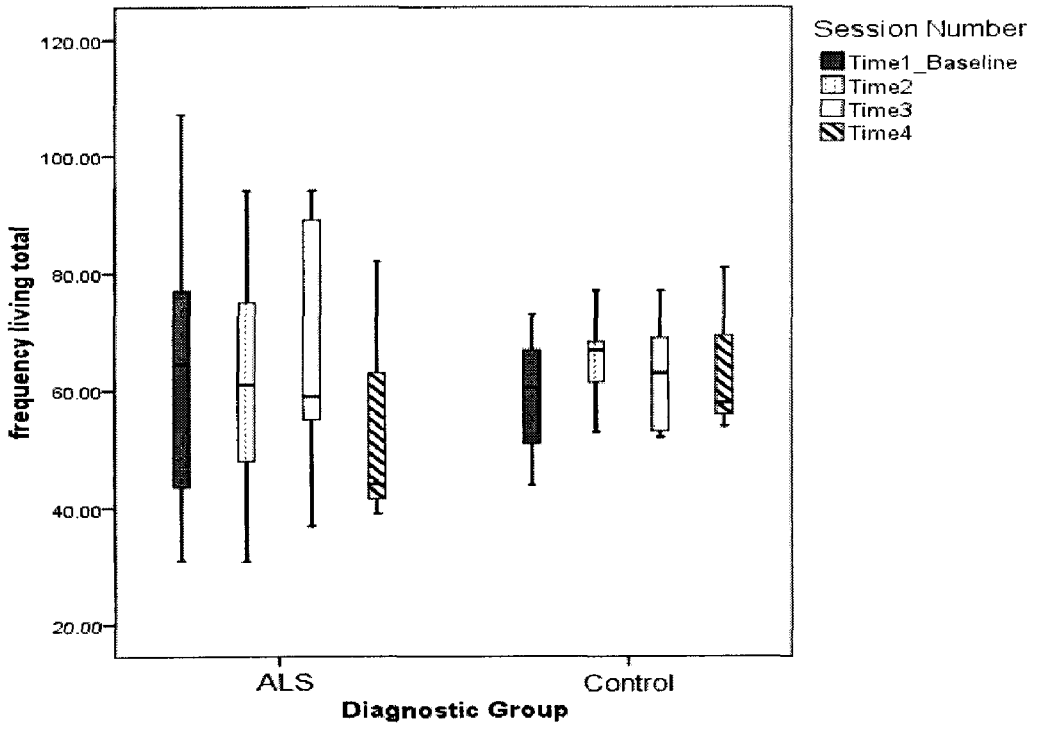

Figure 5. Plots of the means and variations of the total number of items within the living categories for each group at Times 1, 2, 3, and 4.

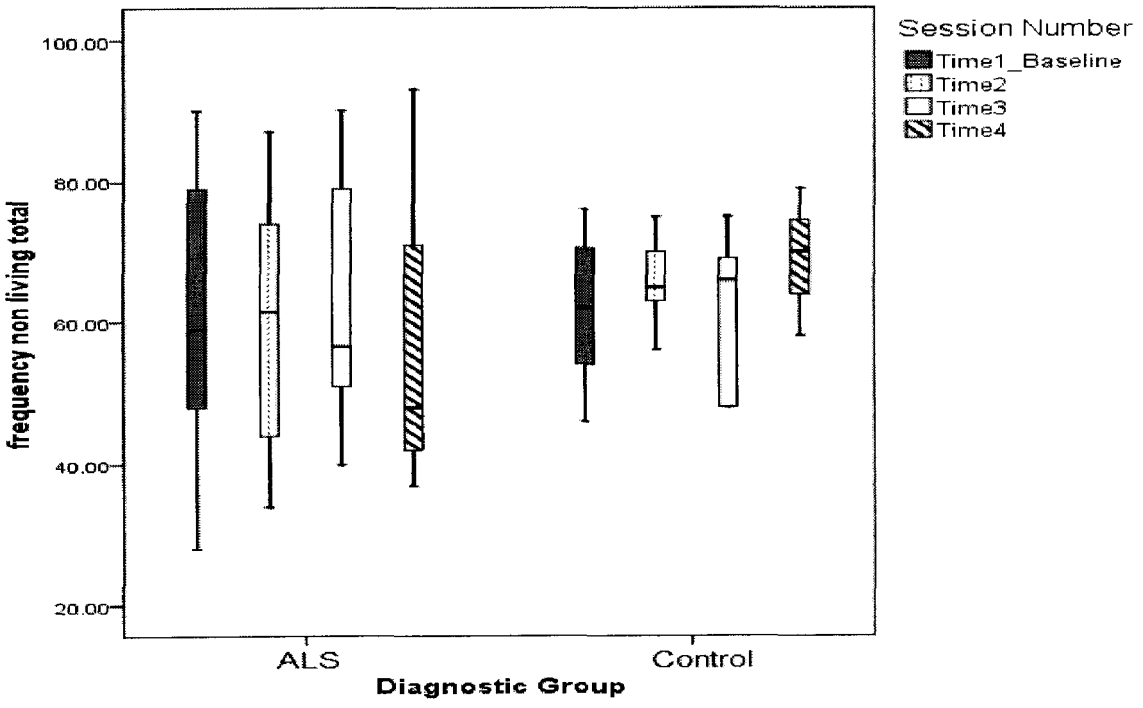

Figure 6. Plots of the means and variations of the total number of items within the non-living categories for each group at Times 1, 2, 3, and 4.

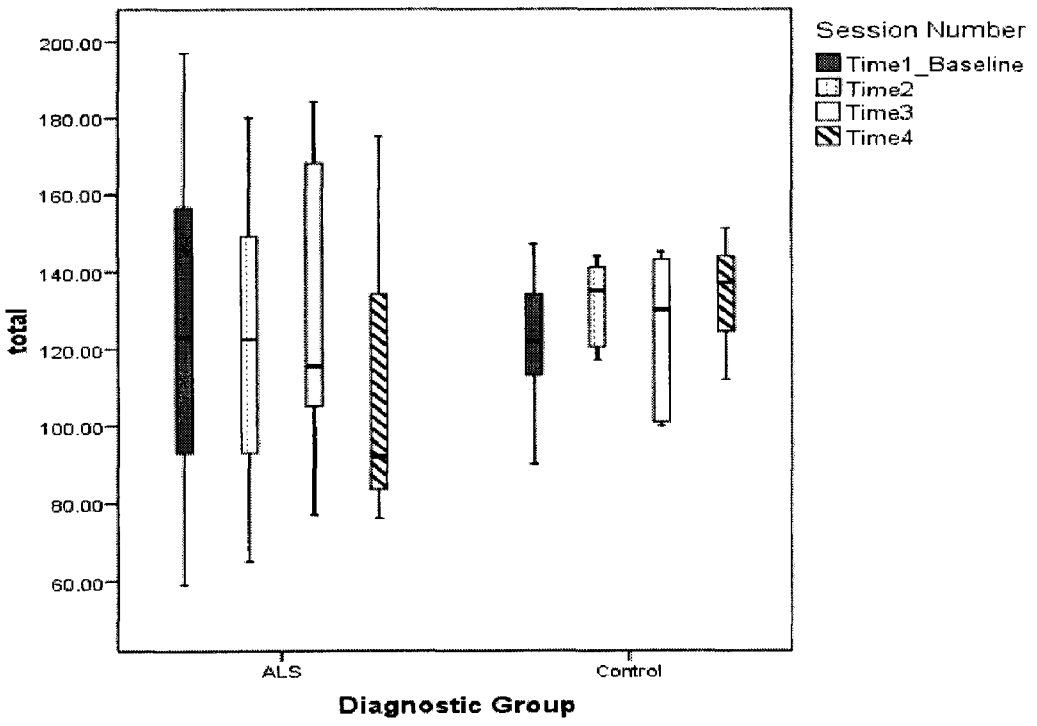

Figure 7. Plots of the means and variations of the total number of items within all categories for each group at Times 1, 2, 3, and 4.

divided systematically into two groups using a cut-off of one standard deviation below the mean on the total number of words generated across all categories. This was done to determine if there was a sub-group of participants with ALS demonstrating impairments in generative naming tasks. The cut-off of only one standard deviation below the mean was used because this is a pilot study of category naming in ALS. Furthermore, a similar cut-off has been used in a previous exploratory study examining language and cognition in ALS (Strong et al., 1999). Three ALS patients (19%) fell below the cut-off at Time 1. This group was labelled ALS group 1. ALS group 2 included the 13 remaining ALS participants who did not fall below the cut-off value of one standard deviation. Sub groups were formed for Time 1 only due to the drop out of participants at the later time periods. At Time 2 only 10 participants with ALS remained in the study and only 1 participant fell below the cut off of one standard deviation below the mean. At Time 3 only 6 participants with ALS remained in the study with 1 participant falling below the cut-off. The participant who fell below the cut-off at Times 2 and 3 was participant 11. He also was included in the low performing sub-group at Time 1. At Time 4 only 4 participants remained in the study and none fell below the cut-off.

ALS group 1 included participants 4, 11, and 16. ALS participant 4 was a right-handed man with limb-onset ALS. At Time 1 he was 47 years old and it had been 16 months since the onset of ALS. He had 15 years of formal education. He received a score of 1.33 on the measure of intelligibility (0 = completely intelligible and 100 = completely unintelligible) and a score of 42.33 on the

measure of rate (0 = very slow, 50 = normal and 100 = very fast). ALS participant 11 was a right-handed man with bulbar-onset ALS. At Time 1 he was 60 years old and it had been 26 months since the onset of ALS. He had 10 years of formal education. He received a score of 61.33 on the measure of intelligibility (0 = completely intelligible and 100 = completely unintelligible) and a score of 16 on the measure of rate (0 = very slow, 50 = normal and 100 = very fast). ALS participant 16 was a right-handed man with limb-onset ALS. At Time 1 he was 68 years old and it had been 29 months since the onset of ALS. He had 10 years of formal education. He received a score of 71.67 on the measure of intelligibility (0 = completely intelligible and 100 = completely unintelligible) and a score of 27.33 on the measure of rate (0 = very slow, 50 = normal and 100 = very fast).

Analyses were conducted to determine if there were differences between group means on the total number of words generated across all sub-categories for ALS group 1, ALS group 2 and the control group. Levene's test revealed violation of homogeneity of variances ($F(2, 25) = 4.89$, $p = 0.016$) therefore non-parametric Mann-Whitney U tests were conducted. There was no significant difference between ALS group 2 and the control group ($U = 58.5$, $Z = -1.061$, $p = 0.289$). Therefore, removing the 3 participants (i.e., ALS group 1) from the entire ALS group did not change the overall characteristics of the ALS group. There was a significant difference between ALS group 1 and the control group on the total number of words generated across all categories ($U = 0.0$, $Z = -2.603$, $p = .009$).

A repeated measures ANOVA was conducted to compare ALS group 1 to the control group on the number of items generated in living and non-living categories at Time 1. There was no significant sub-group by category interaction ($F(1, 13) = .08$, $p = 0.785$). There was no main effect of category for living or non-living items ($F(1, 13) = 0.15$, $p > 0.706$). There was a main effect of sub-group ($F(1, 13) = 20.33$, $p = 0.001$). As shown in Figure 8, the control group produced more items than the ALS group 1 on both living and non-living items.

A repeated measures ANOVA was conducted with sub-group (ALS group 1 vs. Control) as the between subject factor and category (animals, household items, birds, vehicles, water creatures, musical instruments, dogs and tools) as the within subject factor for Time 1. There was no significant sub-group by category interaction ($F(7,91) = 1.10$, $p = 0.372$). There was a significant main effect of category ($F(7, 91) = 5.91$, $p = 0.0$). As shown in Figure 9 both groups produced more items for animals and household items than for the other 6 categories. There was a significant main effect of group ($F(1, 13) = 20.33$, $p = 0.001$). The control group produced more items than the ALS group 1 on all 8 sub-categories (see Figure 9).

Group comparisons also were calculated to determine if there were differences in intelligibility and rate scores between ALS group 1 and control participants. Intelligibility scores for ALS group 1 ranged from 1.33 to 71.67 ($M = 44.78 \pm 37.98$) and rate scores ranged from 16 to 42.33 ($M = 28.55 \pm 13.21$). Levene's test revealed violation of homogeneity of variances for the intelligibility measure ($F(1, 13) = 59.32$, $p = 0.0$), therefore non-parametric Mann-Whitney

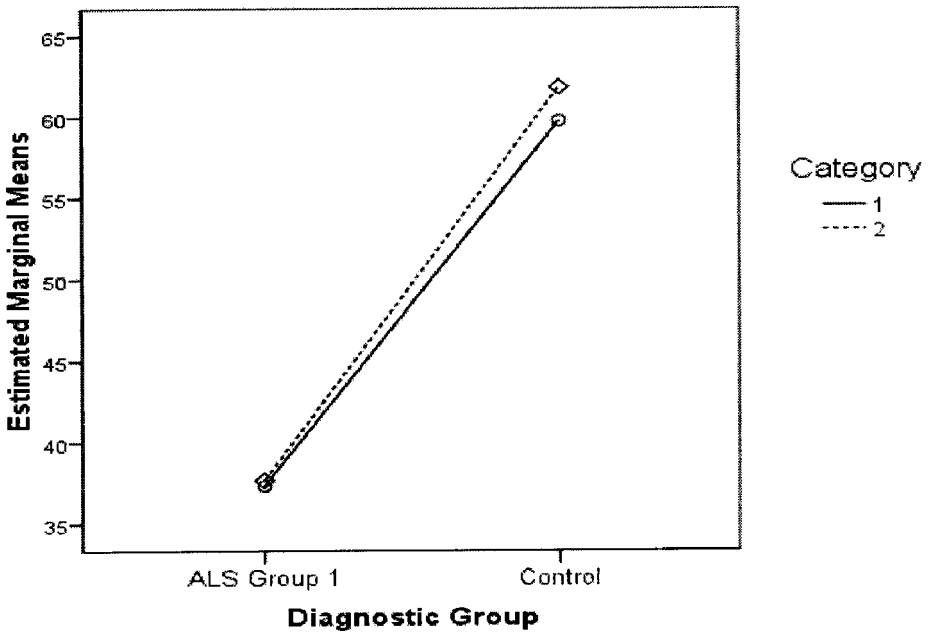

Figure 8. EMM by group for living and non-living categories. (1 = Living; 2 = Non-living)

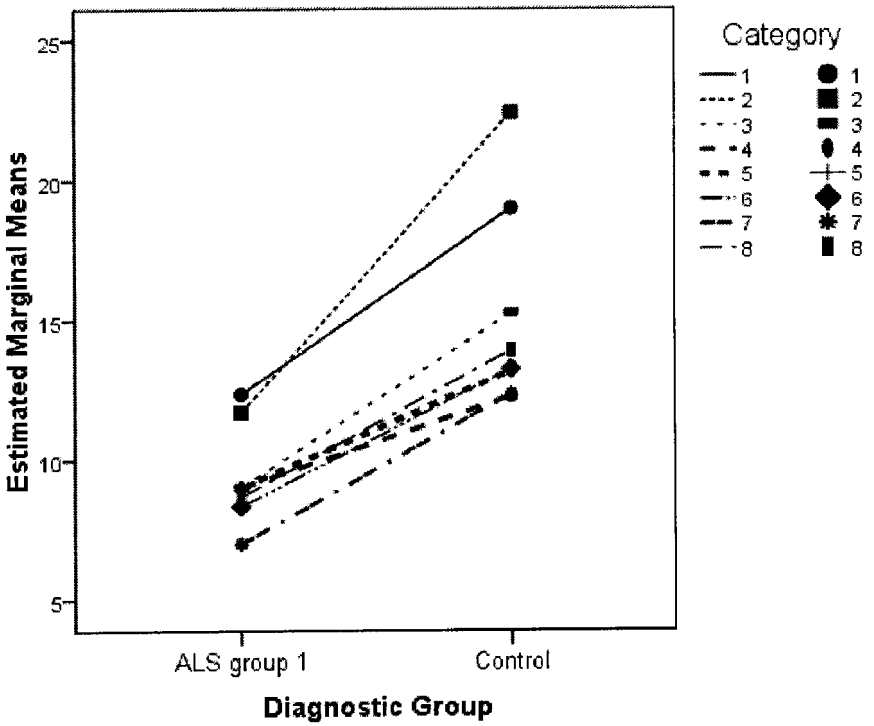

Figure 9. EMM by group for each category. (1 = animals; 2 = household items; 3 = birds; 4 = vehicles; 5 = water creatures; 6 = musical instruments; 7 = dogs; 8 = tools)

U tests were conducted. There were no significant differences in intelligibility scores between the two groups (U = 6.50, Z = -1.664, p = .096). A one-way ANOVA with rate as the between group factor was conducted and revealed a significant difference between ALS group 1 and control participants ($F(1)$ = 12.15, p = 0.004).

High performers. Within the original group of ALS participants at Time 1 there also was a group of high performers. Three participants performed 1 standard deviation above the mean on the total number of words generated across all categories. This ALS group included participants 2, 6 and 12. ALS participant 2 was a right-handed man with limb-onset ALS. At Time 1 he was 51 years old and it had been 23 months since the onset of ALS. He had 15 years of formal education. He received a score of 4.67 on the measure of intelligibility (0 = completely intelligible and 100 = completely unintelligible) and a score of 40.67 on the measure of rate (0 = very slow, 50 = normal and 100 = very fast). ALS participant 6 was a right-handed man with bulbar-onset ALS. At Time 1 he was 53 years old and it had been 11 months since the onset of ALS. He had 22 years of formal education. He received a score of 38.33 on the measure of intelligibility (0 = completely intelligible and 100 = completely unintelligible) and a score of 28 on the measure of rate (0 = very slow, 50 = normal and 100 = very fast). ALS participant 12 was a right-handed woman with bulbar-onset ALS. At Time 1, she was 48 years old and it had been 11 months since the onset of ALS. She had 17 years of formal education. She received a score of 18.33 on the measure of intelligibility (0 = completely intelligible and 100 = completely

unintelligible) and a score of 39.33 on the measure of rate (0 = very slow, 50 = normal and 100 = very fast). All three of these ALS high performers generated an average of 31 more words than the highest number of words generated by the control group. Therefore, since they performed substantially better on the task, no statistical analyses were performed on comparisons with the control group. As previously mentioned, sub-groups were not formed for the later time periods due to the high attrition rate.

Group comparisons also were calculated to determine if there were differences in intelligibility and rate scores between the high performers and the control participants. Intelligibility scores for the high performers ranged from 4.67 to 38.33 (M = 20.44 $\pm$ 16.93) and rate scores ranged from 28 to 40.67 (M = 36.00 $\pm$ 6.96). Levene's test revealed violation of homogeneity of variances for the intelligibility measure ($F(1, 13) = 19.08$, $p = 0.001$), therefore non-parametric Mann-Whitney U tests were conducted. There were significant differences in intelligibility scores between the two groups ($U = 3.5$, $Z = -2.099$, $p = 0.036$). A one-way ANOVA with rate as the between group factor was conducted and revealed a significant difference between the high performers and control participants ($F(1) = 5.78$, $p < 0.032$).

Summary

Overall, the analysis for research question 1 showed that, at Time 1 there was a sub-group of 3 participants with ALS whose scores were statistically lower than the control group on all verbal category fluency measures. However, their scores did not differ for living and non-living categories.

Research Question 2

Research question 2 addressed how category fluency performances of individuals with ALS change with disease progression in comparison to controls. A regression analysis was conducted within each participant who participated at more than one time period (Kenny et al., 2002). The slope values from the regression analysis for each participant are shown in Appendix P. Additionally, the means, standard deviations, minimum and maximum slope values for each category are shown in Table 7. A repeated measures ANOVA was conducted on the slope values from the regression analyses with group (ALS vs. Control) as the between subjects factor and category (living vs. non-living) as the within subjects factor to determine whether there were any between and within group differences in the rate of change in generative naming performance over time. There was no significant group by category interaction for the slope values ($F(1, 15) = .92$, $p = 0.354$). There was no significant main effect of category ($F(1, 15) = 2.18$, $p = 0.160$). Also, there was no significant main effect of group ($F(1, 15) = .40$, $p = 0.537$). An additional repeated measures ANOVA was conducted to determine whether there were differences in slope values between or within groups for any of the 8 sub-categories. Group (ALS vs. Control) was the between group variable and sub-category was the within group variable. There was no significant group by category interaction for the slope values ($F(7, 105) = 1.73$, $p > 0.110$). There was no significant main effect of category ($F(1, 105) = .51$, $p > 0.827$). Also, there was no significant main effect of group ($F(1, 15) = .61$, $p = 0.446$).

Table 7

Slope values for ALS and Control Participants

	ALS					*Controls*				
Categories	*n*	*M*	*SD*	*Min.*	*Max.*	*n*	*M*	*SD*	*Min.*	*Max.*
Animals	10	-0.17	0.56	-1.50	0.56	7	0.06	0.31	-0.43	0.41
Household items	10	-0.05	0.46	-1.00	0.53	7	0.05	0.35	-0.52	0.50
Birds	10	-0.19	0.61	-1.00	0.88	7	0.21	0.63	-0.33	1.50
Vehicles	10	-0.17	0.43	-1.00	0.25	7	0.16	0.60	-0.91	1.00
Water creatures	10	-0.02	0.36	-1.00	0.25	7	0.43	0.52	-0.50	1.00
Musical instruments	10	0.17	0.46	-0.68	1.00	7	0.07	0.72	-1.50	0.58
Dogs	10	0.06	0.43	-.500	0.75	7	-0.15	0.49	-0.88	0.50
Tools	10	0.24	0.63	-0.32	1.67	7	-0.23	0.75	-1.00	1.00
Living items	10	0.00	0.15	-0.17	0.28	7	0.13	0.12	-0.11	0.25
Non-living items	10	-0.03	0.32	-0.74	0.50	7	-0.04	0.25	-0.39	0.24
Total all categories	10	0.03	0.08	-0.08	0.17	7	0.06	0.10	-0.07	0.23

Summary

Overall, the analysis for research question 2 showed that there was no difference in the rate of change in generative naming performance between the ALS and control participants on any of the category fluency measures.

Research Question 3

Research question 3 addressed whether there were differences in category naming of living or non-living items between the bulbar-onset ALS and limb-onset ALS groups. Question 3 also addressed how the ALS sub-groups' performances change over time.

Differences in Category Naming

A repeated measures ANOVA was conducted for each of the first two time periods to determine whether there were any between or within group differences in the number of living and non-living items generated between the bulbar-onset ALS and limb-onset ALS groups. Group (bulbar-onset vs. limb-onset) was the between subject variable and number of items within a category (living vs. non-living) was the within subject variable. There was no significant ALS onset group by category interaction at Time 1 ($F(1, 14) = .01$, $p = 0.942$) or Time 2 ($F(1,8) = 1.47$, $p = 0.260$). There was no significant main effect of category (living or non-living) for either the bulbar or limb onset groups at Time 1 ($F(1, 14) = .35$, $p = 0.565$) or Time 2 ($F(1, 8) = .43$, $p = .532$). There was no main effect of group at Time 1 ($F(1, 14) = .25$, $p = 0.626$) or Time 2 ($F(1, 8) = .21$, $p > 0.658$).

An additional repeated measures ANOVA was conducted for each of the first two time periods to determine whether there were differences between or within

groups for any of the 8 sub-categories. Group (bulbar-onset vs. limb-onset) was the between group variable and sub-category was the within group variable. There was no significant ALS onset group by category interaction for Time 1 ($F(7, 98) = .41$, $p = 0.893$) or Time 2 ($F(7,56) = .10$, $p = 0.443$). There was a main effect of sub-category at Time 1 ($F(7, 98 = 14.09$, $p = .000$) and Time 2 ($F(7, 56) = 13.96$, $p = .000$). Figures 10 and 11 show that a greater number of items were generated for the animals and household items categories than for the other 6 sub-categories by both the bulbar and the limb onset groups at both time periods. There was no significant main effect of group at Time 1 ($F(1,14) = .25$, $p = 0.626$) or Time 2 ($F(1,8) = .21$, $p = 0.658$).

Change in Category Naming over Time

A repeated measures ANOVA was conducted on the slope values from the regression analysis (Kenny et al, 2002) to determine whether there were any between and within group differences in the rate of change in generative naming performance over time. Group (bulbar-onset vs. limb-onset) was the between subjects factor and category (living vs. non-living) as the within subjects factor. There was no significant ALS onset group by category interaction for the slope values from the regression analysis ($F(1, 8) = .21$, $p = 0.658$). There was no significant main effect of category ($F(1, 8)= .07$, $p = 0.800$). There were no group effects on slope values for living or non-living categories ($F(1, 8) = .01$, $p = 0.915$). A repeated measures ANOVA also was conducted on the slope values with group (bulbar-onset vs. limb-onset) as the between subjects factor and the sub-categories as the within subjects factor. There was no significant ALS onset

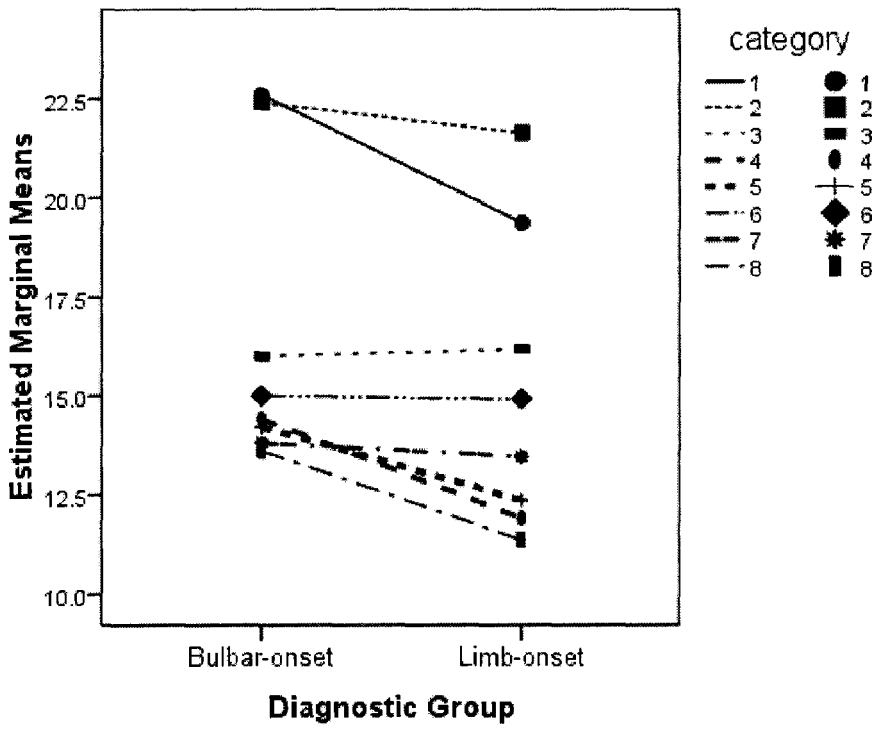

Figure 10. EMM by group for each category at Time 1. (1 = animals; 2 = household items; 3 = birds; 4 = vehicles; 5 = water creatures; 6 = musical instruments; 7 = dogs; 8 = tools)

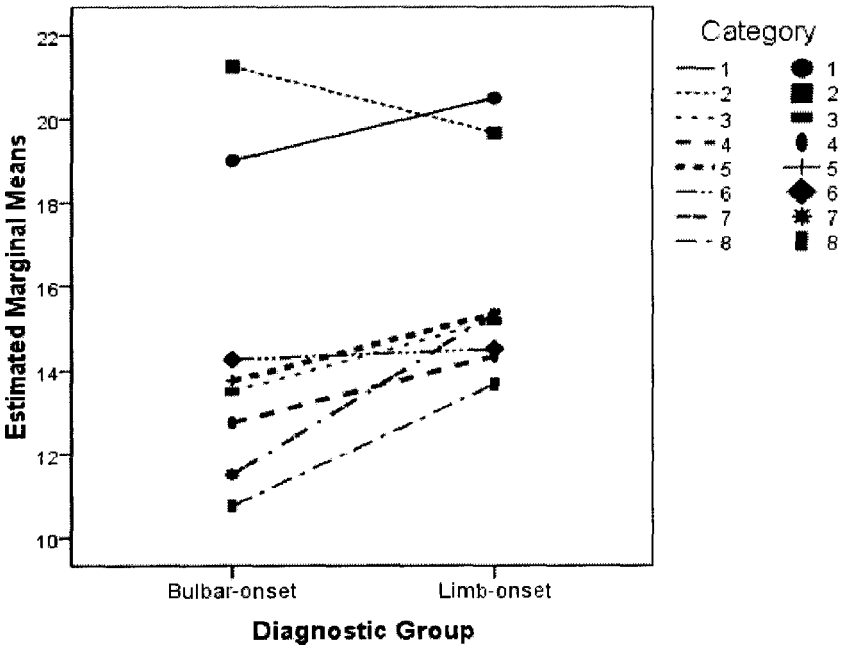

Figure 11. EMM by group for each category at Time 2. (1 = animals; 2 = household items; 3 = birds; 4 = vehicles; 5 = water creatures; 6 = musical instruments; 7 = dogs; 8 = tools)

group by sub-category interaction ($F(7, 56) = .80$, $p = 0.590$). There was no main effect of sub- category ($F(7, 56) = .98$, $p = 0.454$). There was no significant main effect of group ($F(1, 8) = 1.00$, $p = 0.346$).

Summary.

Overall, the analysis for research question 3 showed that there was no difference between the number of items produced by the bulbar-onset or limb-onset ALS groups for either living or non-living categories. Furthermore, the rate of change in generative naming performance between the two groups did not differ for any of the category fluency measures.

Research Question 4

Research question 4 addressed ALS vs. control group differences in the nature of the errors produced on category fluency tasks. The number of errors was recorded for all participants for each category at each time period. The percentages of the total errors generated for each category for both ALS and control groups were calculated for Times 1, 2, 3 and 4 (see Table 8). Additionally, the mean, standard deviation, standard error and 95% confidence intervals of the error percentages for Times 1, 2, 3, and 4 were calculated. No overlap in the 95% confidence intervals indicates a significant difference at the $p \leq 0.05$ level. There were no significant differences in mean percentage of errors for living or non-living categories between the two groups at any time period (see Figure 12). Furthermore, there were no differences between percentages of errors produced for living or non-living categories within either group.

Further calculations were made to investigate the types of errors being

Table 8

Percentage of errors in each category for ALS and Control participants at Times 1, 2, 3, and 4

	ALS				*Controls*			
Categories	Time 1	Time 2	Time 3	Time 4	Time 1	Time 2	Time 3	Time 4
Animals	2.45	1.97	1.65	2.90	2.15	3.13	0.75	1.72
Household items	3.31	1.99	0	2.38	3.24	2.67	2.72	1.37
Birds	5.15	2.03	3.92	3.39	2.66	3.39	1.14	0
Vehicles	4.60	2.84	3.45	0	2.65	0.97	6.33	0
Water creatures	4.17	3.92	1.12	4.17	2.47	4.85	2.17	4.08
Musical instruments	2.85	4.00	2.15	2.08	3.05	6.45	4.88	6.12
Dogs	3.13	2.13	3.33	7.14	0.67	1.10	1.47	0
Tools	3.98	5.30	1.37	6.12	3.47	2.15	4.94	2.27
Living items	3.63	2.48	2.49	4.13	2.05	3.18	1.31	1.53
Non-living items	3.62	3.37	1.58	2.51	3.13	2.94	4.37	2.36
Total all categories	3.63	2.92	2.05	3.33	2.60	3.06	2.85	1.96

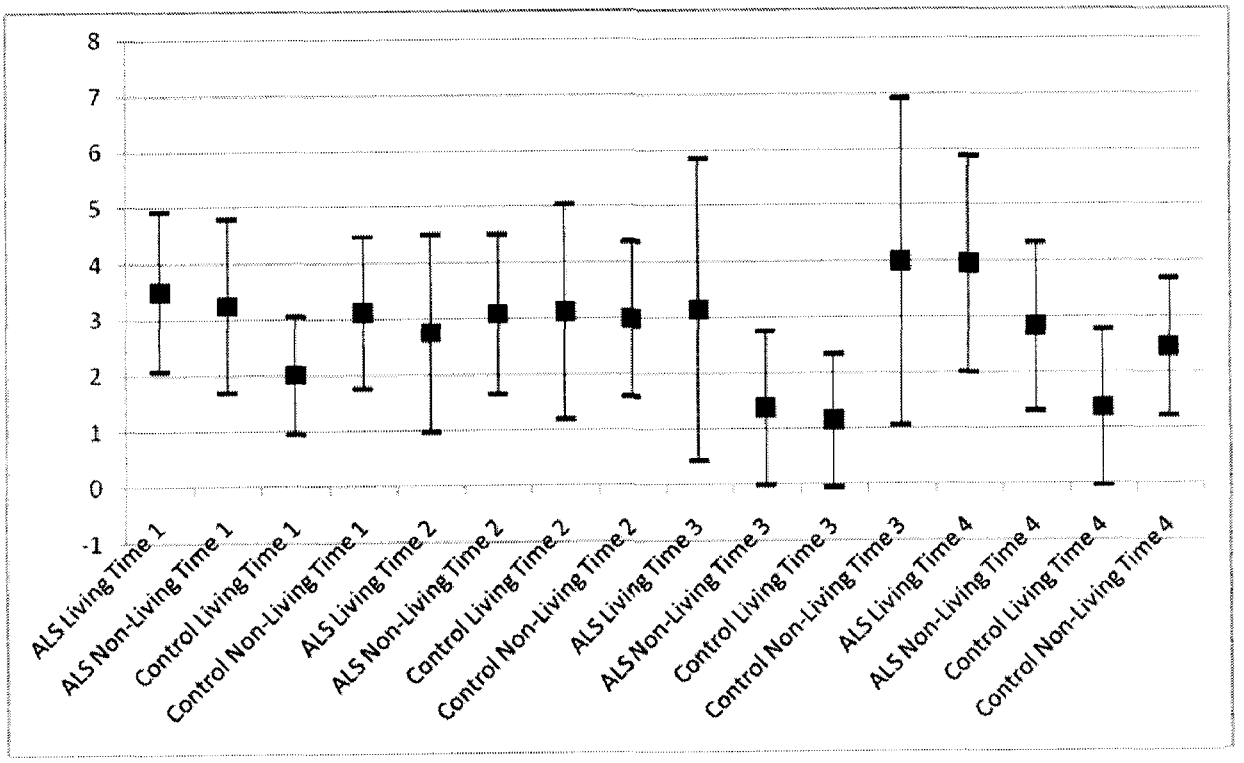

Figure 12. 95% confidence intervals for the percentages of errors produced for the living and non-living categories at Times 1, 2, 3 and 4 for ALS and Control groups.

made by both groups. Calculations of the mean, standard deviation, standard error and 95% confidence intervals revealed that there were no significant differences between the percentages of each error type made by the two groups at Times 1, 2, 3 or 4. However, during examination of the data it was noted that at Time 1 only 1 of the control participants (8%) produced semantically related errors where as 7 of the ALS participants (44%) produced these errors. Furthermore, 11 of the 16 participants with ALS (69%) produced errors other than repetitions whereas 5 of the 12 control participants (41%) produced errors other than repetitions. Similarly, at Time 2, 7 of the 10 ALS participants (70%) produced errors other than repetitions whereas 4 out of 7 control participants (57%) produced errors other than repetitions. At Time 3, 5 of the 6 participants with ALS (83%) produced errors other than repetitions whereas 3 of the 6 control participants produced errors other than repetitions (50%).

Low Performers

The nature of the errors made by ALS group 1 (i.e., low performers) at Time 1 was examined. Calculations of the means, standard deviations, standard errors and 95% confidence intervals are shown in Table 9. ALS group 1 produced fewer errors than the control group in 4 of the 8 sub-categories which included household items, vehicles, water creatures, and musical instruments. Furthermore, ALS group 1 did not produce any errors in 5 of the 8 categories which included water creatures, dogs, household items, vehicles, and musical instruments. When participants in ALS group 1 did

Table 9

Error percentages for ALS Group 1 and Control Participants at Time 1

	ALS Group 1					*Controls*				
Categories	*n*	*M*	*SD*	*SE*	*95% CI*	*n*	*M*	*SD*	*SE*	*95% CI*
Animals	3	7.25	6.34	3.66	(0.08,14.4)	12	1.90	3.05	0.88	(0.17,3.62)
Household items	3	0	0	0	(0,0)	12	3.18	4.03	1.16	(0.91,5.46)
Birds	3	5.81	5.04	2.91	(0.10,11.5)	12	2.87	4.51	1.30	(0.32,5.42)
Vehicles	3	0	0	0	(0,0)	12	2.96	4.81	1.39	(0.24,5.69)
Water creatures	3	0	0	0	(0,0)	12	2.43	3.83	1.10	(0.26,4.59)
Musical instruments	3	0	0	0	(0,0)	12	2.94	4.75	1.37	(0.25,5.63)
Dogs	3	0	0	0	(0,0)	12	0.49	1.70	0.49	(-0.47,1.5)
Tools	3	3.03	5.25	3.03	(-2.9,8.97)	12	2.70	5.25	1.52	(-0.27,5.7)
Living items	3	4.42	1.91	1.10	(2.26,6.58)	12	2.01	1.84	0.53	(0.97,3.05)
Non-living items	3	0.74	1.28	0.74	(-.71,2.19)	12	3.12	2.39	0.69	(1.77,4.47)
Total all categories	3	2.65	1.27	0.73	(1.21,4.09)	12	2.62	1.79	0.52	(1.60,3.63)

produce errors they tended to be within the living categories. There was a significant difference between the percentages of errors for living items and the percentage of errors for non-living items for ALS group 1 (see Figure 13). They produced a higher percentage of errors for the living categories than for the non-living categories. Furthermore, participants in ALS group 1 only produced 2 types of errors; repetitions and semantically related errors. However, only 1 of the 3 ALS group 1 participants produced a semantically related error.

High Performers

There were no significant differences between the percentages of errors produced by the high performers and the control group for either living or non-living categories (see Figure 14). The high performers produced errors in all categories except for animals. Furthermore, there were no differences in the types of errors produced by the high performers in comparison to controls.

Bulbar-onset and Limb-onset ALS Groups

The number and types of errors produced by the bulbar-onset ALS group in comparison to the limb-onset group at Time 1 and 2 also was examined. Calculations of the means, standard deviations, standard error and 95% confidence intervals revealed that the bulbar-onset group did not differ from the limb-onset group in the percentage of errors produced for either living or non-living categories at Time 1 or at Time 2 (see Figure 15). Furthermore, the number of errors between living and non-living categories

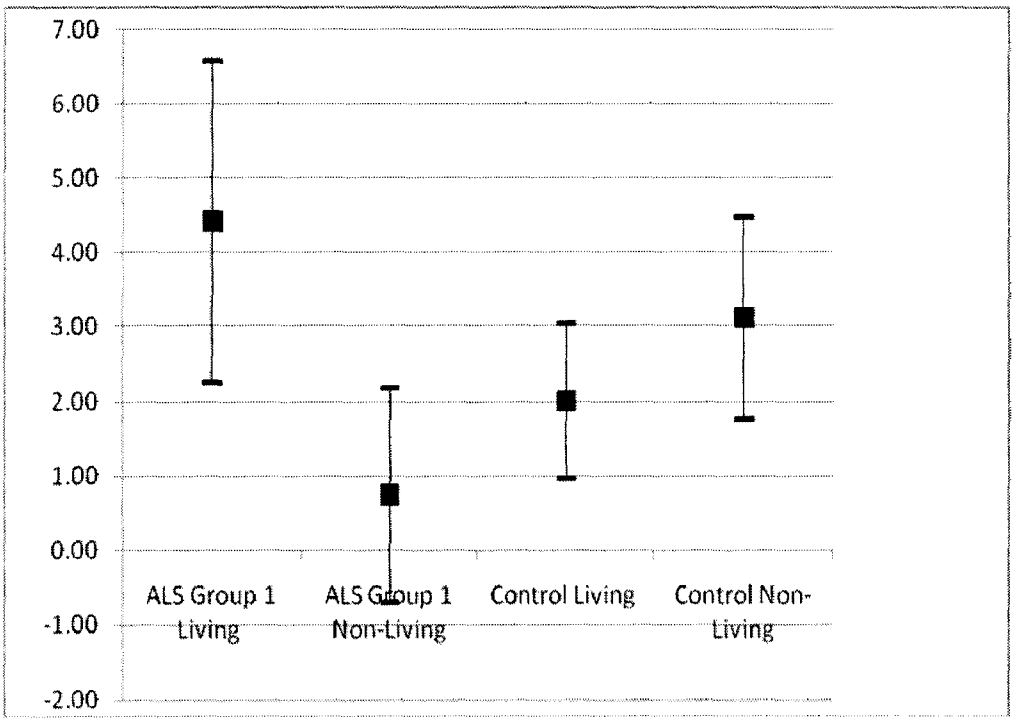

Figure 13. 95% confidence intervals for the percentages of errors produced for the living and non-living categories for ALS group 1 and the control group at Time 1.

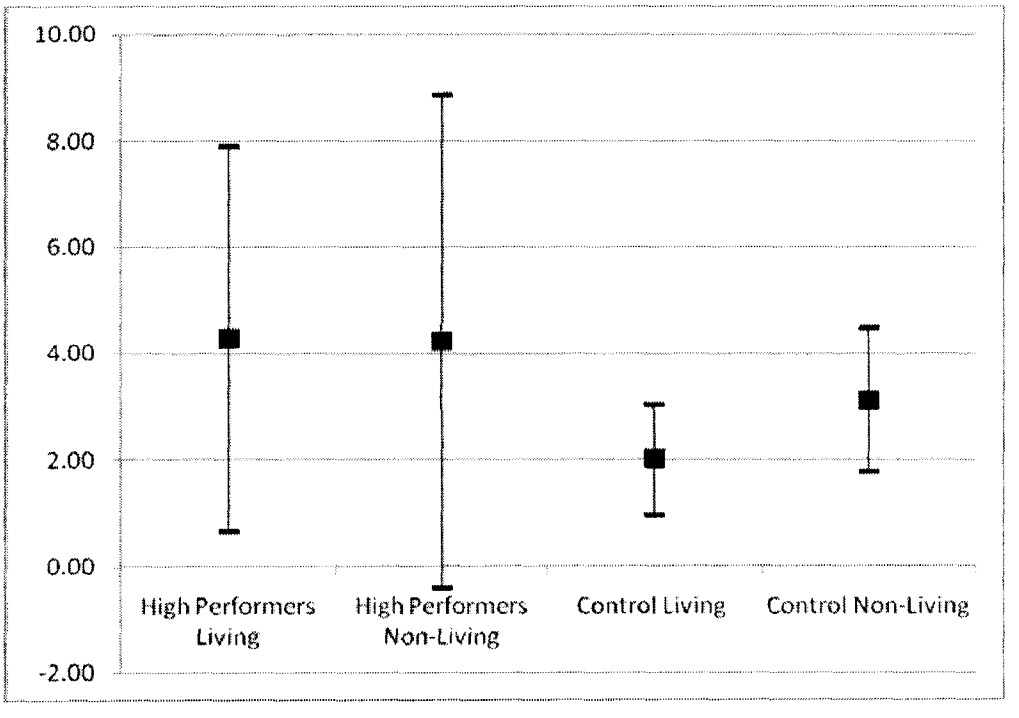

Figure 14. 95% confidence intervals for the percentages of errors produced for the living and non-living categories for the high performers and the control group at Time 1.

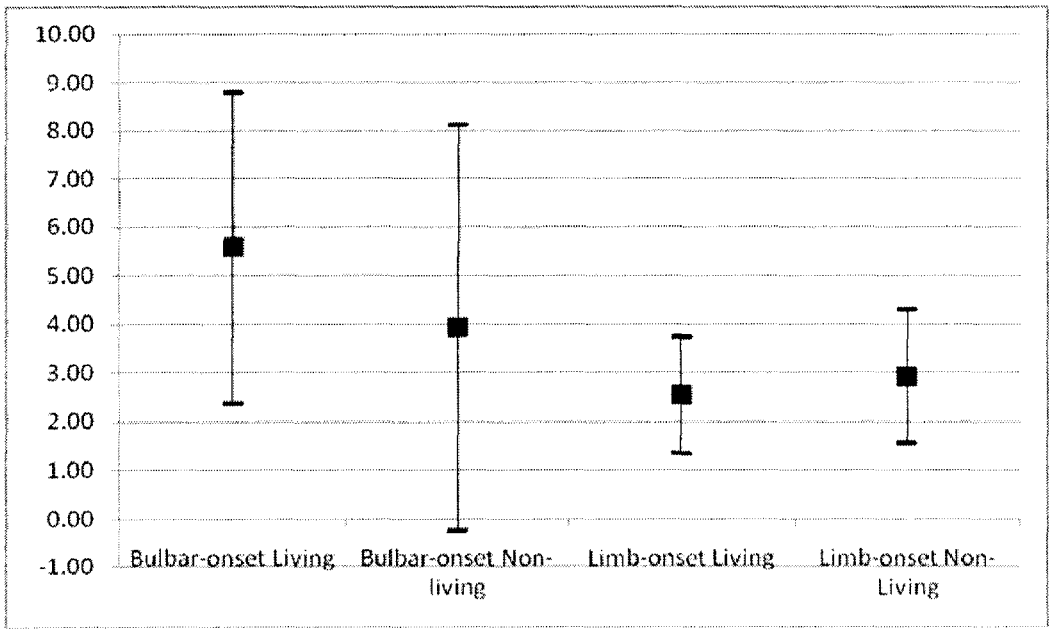

Figure 15. 95% confidence intervals for the percentages of errors produced for the living and non-living categories for the bulbar-onset and limb-onset ALS groups.

did not differ within either of the two groups. The types of errors produced at Time 1 and Time 2 did not differ between the two groups.

Discussion

The purpose of this study was fourfold. The first was to examine category fluency performances in ALS participants to determine if category specific impairments exist for living or non-living items. The second was to determine whether category fluency performances of individuals with ALS change over time. Thirdly, the performance of individuals with bulbar-onset ALS on category fluency tasks was compared to the performances of those with limb-onset ALS. Finally, the nature of the errors produced on category fluency tasks by the participants with ALS was investigated and compared to controls.

Research Question 1

Research question 1 addressed the question of whether individuals with ALS exhibit category-specific impairments for living or non-living items. Eight category fluency measures (4 living and 4 non-living) were obtained from participants with ALS and controls. Based on the data obtained from the study participants, generative naming of living and non-living items was not significantly different among individuals with ALS at any time period. Furthermore, individuals with ALS were not statistically different than controls in their verbal category fluency measures regardless of living/non-living distinction. Participants from the ALS and control groups did not differ significantly on the total number of words generated across all categories. This is inconsistent with the literature which shows that both verbal and category fluency measures are impaired in ALS

(Schreiber et al., 2005; Strong et al., 1999). The only category in which participants with ALS performed worse than controls was household items. However, this category was very difficult to score. There were few clear distinctions as to what constitutes a household item, despite repeated attempts to obtain a formal definition from Hodges et al., (1992) who first used this category in their naming studies. Therefore, the differences for the household category should be interpreted with caution.

The ALS group did differ from the controls on measures of intelligibility with the ALS group receiving higher scores (i.e., more unintelligible). However, the mean intelligibility scores for the participants with ALS still fell within the mild to moderately unintelligible range for each time period. Furthermore, intelligibility did not affect generative naming scores as participants with ALS produced only 10 unintelligible items at Time 2 out of a total of 2227 items. At Times 1, 3, and 4, no unintelligible items were produced. Additionally, there was no statistically significant correlation between intelligibility and total number of items generated for the ALS and control participants.

There also was a difference between participants with ALS and controls on the measure of rate with the ALS group receiving lower scores (i.e. slower speaking rate) at Times 1, 2, and 3. This is expected as one of the main speech characteristics of the mixed dysarthria associated with ALS is a slow speaking rate (Darley et al., 1975). However, the mean rate score for the ALS group at Time 1 was 34.4, at Time 2 it was 34.8, and at Time 3 it was 33.0. These scores are still much closer to normal (i.e., 50) than they are to a very slow speaking rate

(i.e., 0). Additionally, there was no statistically significant correlation between speaking rate and the total number of items generated by the ALS and control participants suggesting that speaking rate did not affect verbal category fluency performance. Furthermore, even though there were group differences in rate, there were no group differences in performance on any of the category measures.

The disagreement between the current findings on the performances of participants with ALS on the verbal category fluency tasks and those previously reported in the literature could be explained by the small sample size and the high variability in the ALS participants' performances. The exact prevalence of cognitive impairment in ALS is unknown but is estimated to be as high as 30% to 35.6% in two large studies (Massman et. al, 1996; Ringholz et. al, 2005). Massman and colleagues (1996) found that the remaining 64.4% of their participants with ALS did not show notable neuropsychological deficits. In addition to the 30% of participants with ALS and cognitive impairment found in the study by Ringholz and colleagues (2005) 20% of the remaining participants had impairments severe enough to be considered dementia. Language impairment identified by deficits in confrontation naming was found in the participants with impairment but to a lesser degree. The remaining 50% of participants performed normally on neuropsychological and language tasks. Furthermore, in a study examining the language performance of 9 participants with ALS, Cobble (1998) identified only a sub-group of 3 participants (30%) who obtained low scores on language measures. Therefore, it is possible that with

the large variability among the participants with ALS within the current study, any low scores were masked by participants who did not have cognitive or language impairment and who obtained high scores on the category fluency tasks. At Time 1, there were groups found within the data including high performers and low performers. It is possible that the scores produced by the high performing participants with ALS masked those with the low scores and therefore no statistical difference was found between the ALS group and controls. It also is possible that none of the participants with ALS in this study had cognitive or language impairment. Further analyses are in progress on the cognitive data obtained from neuropsychological testing in the larger study on which this current one is based.

At Time 1, the low performers included a small subset of approximately 20% (n = 3) of the ALS participants who did have scores on the verbal category fluency tasks that were significantly lower than the control group. It is possible that these participants are performing poorly as a result of language impairment. This is consistent with the findings in the literature on ALS. Rakowicz and Hodges (1998) identified 3 subgroups in their study of 18 participants with ALS based on results of language and neuropsychological tests. Three participants with ALS suffered from dementia and had impaired language, 2 were described as having word finding difficulties and anomia and thirteen performed within normal limits. However, within the group of participants who performed within normal limits, 2 of the ALS participants had deficits in category fluency. Although, the one standard deviation cut-off has been used in exploratory studies

it should be acknowledged that these individuals may just be the low performers within a normal distribution. Future studies may wish to consider a more stringent cut-off.

Another explanation for the current findings is that the ALS participants in the sub-group who produced a lower number of items did so because they had fewer years of formal education. Two of the 3 participants in the lower performing sub-group were more than one standard deviation below the mean of the ALS group in number of years of education. However, 1 of the participants within the lower performing sub-group had 15 years of formal education and still met the criteria for ALS group 1. Additionally, 3 control participants had 10 years of formal education and performed well within 1 standard deviation of the mean for the total number of words generated across all categories for the control participants.

Yet, another explanation could be that ALS group 1 produced a lower number of items due to dysarthria or effortful speech. The group was significantly slower than controls as demonstrated by the rate measures. However, one of the participants that met the requirements for ALS group 1 obtained a rate score of 42.3 which is very close to a normal speaking rate (i.e., 50). Additionally, the group of high performers also were significantly slower than the control participants. However, these differences in rate did not affect their performance because they produced a greater number of items in the verbal category fluency tasks than the control participants. Furthermore, there was no statistically significant correlation found between rate and total number of items

generated.

The lower performing sub-group of ALS participants (i.e., ALS group 1) did not demonstrate differences in scores for living or non-living items. Furthermore, they did not perform more poorly on one category or another. Their scores were uniform across all categories. However, this is a very small sample and results can not be generalized to the population of individuals with ALS as a whole. A larger sample could increase the statistical power. Additionally, a larger sample might reveal sub-groups within the ALS participants based on their performance with a greater number of participants within each sub-group. This could provide more information about the nature of category fluency deficits within the subgroups.

Furthermore, a different method of interpreting the data could provide more insight into the category fluency performances of these 3 low performing individuals as well as the ALS group as a whole. It is well known that word frequency and age of acquisition influence word retrieval. Items that are acquired earlier in life and which are higher in frequency are named faster than those that are acquired later in life and that are lower in frequency (Ellis & Morrison, 1998). Furthermore, studies examining naming impairments in participants with Alzheimer's disease and semantic dementia (SD) have shown greater impairments naming items that are acquired later in life (Holmes, Fitch and Ellis, 2006; Lambon Ralph, Graham, Ellis and Hodges, 1998). Lambon Ralph and colleagues (1998) found that spoken frequency affected naming in their participants with SD. Furthermore, in a study of 20 participants with Alzheimer's

disease, 8 participants with semantic dementia and 12 participants with primary progressive aphasia, Marczinski and Kertesz (2005) found that all three groups differed from control groups in mean word frequency on letter and category fluency tasks with the control group producing the lower frequency exemplars. Additionally, in a study of 92 participants with Alzheimer's disease, Forbes-McKay, Ellis, Shanks, Venneri (2005) reported that the words generated by the AD participants on semantic fluency tasks were shorter, were acquired earlier, were of higher frequency, and were more typical of the semantic category. They also found that age of acquisition was better at predicting AD participants from controls than the number of words generated. Using this type of analysis on the current data could provide more information about the category fluency performances of the ALS group in comparison to controls. Furthermore, this type of analysis and interpretation might reveal a greater impairment for living or non-living categories within the group. Further future research is warranted.

Research Question 2

Research question 2 addressed how category fluency performances of participants with ALS change over time in comparison to controls. There was no difference in the rate of change of performances on generative naming tasks for the ALS participants in comparison to the control participants for either the living or non-living categories. Additionally, no difference was found in the rate of change of performances for any one of the sub-categories. This finding is consistent with the literature as previous researchers have found either slow progression of language and cognitive impairments or no progression at all

(Abrahams et al., 2005; Kilani et al., 2004; Schrieber et al., 2005; Strong et al., 1999). In an 18 month longitudinal study of 52 participants with ALS, Schreiber and colleagues (2005) found verbal fluency to be impaired at initial testing with no significant change over the course of the study. Similarly, Abrahams and colleagues found no decline in verbal fluency performance over a 6 month period in a study of 20 participants with ALS who did not exhibit dementia. In a study of 19 participants with ALS conducted by Robinson et al. (2006), a sub-group of 7 participants with ALS showed progression of impaired cognitive performance over 6 months; however, verbal fluency was not among the neuropsychological tests performed. Therefore, it is possible that verbal category fluency performance does not decline with disease progression.

There are other possible explanations for the findings of the current study. Firstly, the small sample size and the high variability in performance, especially among the ALS participants, could contribute to the lack of difference in rate of change in performance between the two groups. At Time 1 different sub-groups of participants with ALS were found based on their performance. There was a group of high performers and a group of low performers. Due to the difference in baseline functioning of the participants in the current study, it would be expected that the different groups would change differently over time. Secondly, the time post onset differed across individuals at baseline testing with the time post onset ranging from 9 to 79 months. This large variability in time post onset might make it difficult to detect patterns of change within the group as it is possible that some individuals' performances changed before initial testing even began.

Research Question 3

Research question 3 addressed whether there were differences in category naming of living or non-living items between the bulbar-onset and limb-onset ALS groups. Question 3 also addressed how the groups' performances change over time.

Differences in Category Naming

Participants with bulbar-onset ALS were not statistically different than those with limb-onset ALS at Time 1 or Time 2 on any of the category fluency measures. This is inconsistent with some findings in the literature showing that individuals with bulbar-onset ALS are more impaired on neuropsychological tasks including word generation (Schreiber et al., 2005; Abrahams et al., 1997; Strong et al., 1999). Interestingly, several researchers have not found a correlation between cognitive impairment and site of onset (Massman et al., 1996; Rippon et al., 2006). In a study of 40 participants with ALS, Rippon et al. (2006) concluded that bulbar dysfunction did not affect test performance. Their study included 9 participants who met criteria for dementia and 3 who had cognitive impairment but no dementia. Of the nine participants with dementia only 1 had bulbar onset ALS. Of the 5 other participants with bulbar-onset, 1 had mild cognitive impairment and 4 had no cognitive impairment. Therefore, one explanation for the findings of the current study is that performance on category fluency tasks is not affected by site of onset.

Interestingly, there was no difference in intelligibility between the two groups at Time 1. Additionally, at both Time 1 and Time 2 there were no

differences in speaking rate between the two groups. This is unexpected as dysarthria is more likely with bulbar involvement as bulbar palsy affects the muscles of articulation (Mitsumoto, 1998). However, it is possible that some participants with limb-onset ALS developed bulbar symptoms throughout the progression of the disease. At Time 1 the limb-onset group had a longer time post onset than the bulbar-onset group. The range of the time post onset for the limb-onset participants was 9 to 79 months (M = 28.73 $\pm$ 22.02) and the range of time post onset for the bulbar-onset participants was 11 to 28 months (M = 18.4 $\pm$ 8.12). At Time 2, although there was a difference in intelligibility between the two groups there was no difference in rate. Furthermore, the limb-onset group had a longer time post onset than the bulbar-onset group at Time 2. The range of time post onset for the limb-onset group was 15 to 84 (M = 42 $\pm$ 28.64) and the range of time post onset for the bulbar-onset group was 17 to 32 (M = 21.75 $\pm$ 7.32). Therefore, with the longer time post onset it is possible some of the participants with limb-onset ALS developed bulbar involvement. Furthermore, the extent of bulbar involvement for each participant was not determined in the current study. Therefore, it is possible that the lack of differences in performances on the verbal category fluency measures could be due to possible bulbar involvement within the some of the limb-onset ALS participants.

Another explanation for the findings of this study is the small sample size. With such a small sample size, it is possible that none of the patients within the bulbar-onset group (n = 5) were exhibiting language and cognitive impairments. Furthermore, the high variability in the current study could be contributing to the

lack of difference between the two groups. There were 2 participants with bulbar-onset ALS in the high performing group, therefore, it is possible that the scores of these participants were masking any scores of those exhibiting language and cognitive impairments.

Change in Category Naming over Time

The rate of change of performances on verbal category fluency tasks did not differ between participants with bulbar-onset ALS and those with limb-onset. This is inconsistent with the literature that shows individuals with bulbar-onset ALS demonstrate a greater impairment in verbal fluency over time (Schreiber et al., 2005; Strong et al., 1999). Once again, it is possible that the small sample size had an impact on the results and was responsible for the lack of difference between the two groups. A small sample size together with potentially low power and small effect size would make it difficult to detect differences between groups. Only 4 participants with bulbar-onset remained in the study for more than 1 time period and only 1 participant remained in the study for more than 6 months. Therefore, it was not possible to evaluate performances towards the end stages of the disease. Another explanation is that there is no difference between the change in performance over time for the two groups.

Research Question 4

Research question 4 addressed the nature of errors on verbal category fluency tasks among participants with ALS versus control participants. The participants with ALS did not produce a greater percentage of errors than controls in either the living or non-living categories. Furthermore, they did not

produce a greater percentage of errors in any one category or in the total of all categories. This is an unexpected result. I had hypothesized that the ALS group would produce more errors than controls if they had language and cognitive impairments.

There were no group differences between the types of errors produced between the ALS and control groups. However, more participants with ALS produced semantically related errors than control participants at Time 1. This is consistent with the findings of studies performed by both Rakowicz and Hodges (1998) and Strong et al. (1996). In both studies participants with ALS produced semantic paraphasias on confrontation picture naming tasks. It is possible that these individuals in the current study have language impairments in which their semantic system is affected. At Times 1, 2, and 3 more participants with ALS were producing errors other than repetitions compared to controls thus providing further support for the possibility that language is affected in these individuals. Percentage of repetitions did not differ among the two groups. This is an unexpected result as studies have documented impaired attention and self-monitoring in both ALS and FTD participants (Abe et al., 1997; Hanagasi et al, 2002; Talbot et al., 1995, Neary et al., 1998).

Low Performers

The low performers group comprised 3 participants with ALS who performed one standard deviation below the mean on the total of all categories measure at Time 1. Due to the large variability within the ALS group, subgroups were formed to determine if the low performers performed significantly worse

than the control group on category fluency tasks. The ALS group 1 produced fewer errors in several sub-categories which included household items, vehicles, water creatures and musical instruments. This was unexpected because if this group had language and cognitive impairment it would be expected that they would produce a greater percentage of errors. One possible explanation for the low scores on the category fluency tasks along with the few errors is that these participants with ALS were using cognitive strategies during testing. This group of participants may have felt pressure to do well on the tests. It is possible they had a fear of producing incorrect items and therefore monitored their performance closely ensuring few errors. As a result, they produced fewer exemplars for each category. However, the instructions by the examiner were to produce as many items as they could with no indication that errors would be penalized, therefore, this might not have been the case.

The errors that the participants in ALS group 1 did make were primarily within the living sub-categories. If these participants do indeed have language and cognitive impairments, as demonstrated by analyses that are part of the larger group study, then the results of the current study suggest that the living categories are more vulnerable to impairment than the non-living categories in this group. This is consistent with the findings of Rakowicz and Hodges (1998). One participant with ALS/aphasia within their study showed more difficulty naming living creatures than man-made objects on a confrontation naming task. However, the conjecture that the participants in ALS group 1 have a problem naming items in the living sub-categories is inconsistent with a study of

individuals with primary lateral sclerosis (PLS) conducted by Yee (2004). Yee (2004) found participants with PLS performed more poorly on verbal fluency tasks in the non-living category. However, her findings were not statistically significant. In addition, her findings pertained to the number of items generated and not to the number of errors produced.

The error types produced by ALS group 1 consisted of only repetitions and semantically related errors with the majority of errors being repetitions. This is the type of error that would be expected with frontal lobe impairment as deficits in attention are common (Neary et al., 1998). However, this group did not produce significantly more repetitions than the other participants with ALS or the controls.

Bulbar-onset and Limb-onset ALS

There were no differences between the percentages of errors between the two groups at either of the first two time periods. Furthermore, there was no difference between the types of errors produced by the two groups. It was expected that the bulbar-onset ALS group would have produced more errors as a result of greater language and cognitive impairment. However, this was not the case. In the current study, site of onset did not appear to affect the amount or types of errors produced.

Limitations of the Study

There were several limitations to the current study. Firstly, there was a small sample size and large variability, particularly among the participants with ALS. These two factors make it difficult to detect differences between the two diagnostic groups or between the two ALS sub-groups (bulbar-onset vs. limb-

onset). Secondly, there was a large drop out of participants at each time period. It is difficult to perform a longitudinal study with individuals with ALS because of the rapid progression of the disease leading to rapid increases in physical limitations and death. Thirdly, one of the sub-categories (i.e., household items) was very difficult to score because there were few clear definitions as to what items belong to this category. Fourthly, the time post onset differed greatly among the participants with ALS. This makes it difficult to explore the change in performance over time for the ALS group. It also makes it difficult to detect differences between limb-onset and bulbar-onset groups as the participants with limb-onset ALS can develop bulbar symptoms over time.

Future Directions

This is the first study to examine in detail the nature of naming impairments over time on category fluency tasks among individuals with ALS. Further investigation of category fluency impairments is warranted.

The first recommendation for future research is to perform a study with a larger sample of participants with ALS and controls. A larger sample may help to reduce the variability within the ALS group and could provide a better representation of their true performance. Additionally, a larger sample size could reveal clearer distinctions between sub-groups of ALS participants based on their performance. Obtaining a large sample of ALS participants is difficult due to the prevalence of the disease. Furthermore it is difficult to conduct a longitudinal study due to high attrition rates as a result of the rapid progression of physical symptoms and death. This could be overcome by performing a multi-centre

study with the same language and cognitive testing procedure. A link between the group of ALS researchers in the United Kingdom, Dr. Laura Goldstein and colleagues, and the group of ALS researchers in London, Ontario, Dr. Michael Strong and colleagues, is currently be discussed. Therefore, a multi-centre study is a future possibility.

The second recommendation is to conduct a study which includes only those individuals with ALS with documented language and cognitive impairments. These data would help provide more information about the nature of the verbal category fluency deficits that exist in some individuals with ALS. This is beneficial in providing more information about the similarities or differences between the cognitive impairment found in ALS and other types of dementia, such as Alzheimer's disease.

The third recommendation is to incorporate a variety of methods of analyses for the verbal category fluency data. For example, investigators in future studies could include measures such as frequency and age of acquisition of items generated. Analyzing the frequency and age of acquisition of items produced could provide more information about the nature of category fluency performances of individuals with ALS.

The fourth recommendation is to perform a study with a larger battery of language tasks. In a study by Rakowicz and Hodges (1998) 1 participant with ALS showed a category specific deficit for living things compared to manmade objects on both confrontation naming and picture word matching tasks. Furthermore, in a study conducted by Strong and colleagues (1996) participants

with ALS demonstrated impairments in single word comprehension. Therefore, expanding the naming test battery to include these types of tasks with representations from the different sub-categories could provide more insight into any possible category specific deficits.

The fifth recommendation is to perform a study to determine the test-retest reliability of the category fluency tasks used by Hodges et al. (1992). This information is valuable to ensure that any changes in performances over time are due to a decline in language and not to an unreliable test.

The sixth recommendation is to perform correlational analyses between the category fluency performances and the CT perfusion results of the larger study on which this current study is based. These analyses could provide insight into the relationship between category fluency performances and brain functioning. This is important because it would provide further information as to whether categorical information is processed differently in the brain. Furthermore, if there are differences it would provide insight into which areas may be involved in the processing of different categories. This information could help to support or refute one or more of the current theories.

The seventh and final recommendation is to compare the verbal category fluency data with the language data from the larger study to determine if there are parallels in language performance among the ALS participants. For example, data from the Action Naming Test and the confrontation naming task in the BDAE, used in the larger study, could be compared to the data from the generative naming tasks in the current study to determine if there are any

patterns in naming performance. This would provide more insight into the nature of language and naming impairments in ALS.

Summary and Conclusions

Overall, the results from this study indicate that the ALS group did not differ from controls on either living or non-living verbal category fluency tasks. Furthermore, the two groups did not differ in the changes in their performance over time. There were a greater number of participants with ALS who produced errors other than repetitions, suggesting language might be affected in these participants. Furthermore, there was a small subgroup of participants with ALS who performed significantly worse than controls on all category fluency measures but who did not show a specific deficit for either living or non-living categories. However, this group did produce more errors on living categories than non-living categories. It is possible that this group demonstrates language impairment with living categories more vulnerable to impairment. This finding is important relative to clinical considerations because using category fluency tasks for living items could be more sensitive in detecting language impairments than using category fluency tasks for non-living items.

Results also indicated there was no difference found between the bulbar-onset and limb-onset ALS groups in the numbers of items generated or in the types of errors produced for category fluency tasks for either living or non-living categories.

This was a preliminary study of the nature of the verbal category deficits found in ALS. Further studies with a larger sample size and a more extensive

battery of tests using a variety of methods of naming analyses could provide more insight into the nature of the naming and language deficits in ALS and how they might change over time.

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Appendix A

Summary of Demographic Information for ALS and Control Participants at Time 1

	Participants																	
Measures	1	2	3	4	5	6	7	8	9	10	11	12	13	14	15	16	*M*	*SD*
	ALS																	
Sex	F	M	F	M	M	M	F	M	F	F	M	F	M	F	M	M		
Age (yrs)	40	51	59	47	42	53	56	34	61	58	60	48	56	63	48	68	52.8	9.15
Ed (yrs)	17	15	12	15	14	22	16	12	16	18	10	17	13	-	17	10	14.9	3.22
Site at Onset	B	LL	LL	UL	UL	B	LL	UL	UL	UL	B	B	B	UL	LL	UL		
Familial (F)/ Sporadic (S)	S	S	F	S	S	F	S	S	S	S	S	S	S	S	S	S		
Control	*Control*																	
Sex	M	F	M	F	M	M	M	M	F	M	F	F						
Age (yrs)	34	53	58	45	63	54	59	57	54	53	62	48					53.3	8.03
Ed (yrs)	16	10	10	14	14	13	14	16	14	11.5	18	10					13.8	2.67

Note. Dashes indicate the data were not obtained for the participant. B = Bulbar. LL = Lower Limb. UL = Upper Limb

Appendix B

Summary of Demographic Information for ALS and Control Participants at Time 2

	Participants																	
Measures	1	2	3	4	5	6	7	8	9	10	11	12	13	14	15	16	*M*	*SD*
								ALS										
Sex		M		M	M	M	F	M		F	M	F	M					
Age (yrs)		52		48	43	53	57	35		58	60	48	57				51.1	7.78
Ed (yrs)		15		15	14	22	16	12		18	10	17	13				15.2	3.36
Site at Onset		LL		UL	UL	B	LL	UL		UL	B	B	B					
Familial (F)/ Sporadic (S)		S		S	S	F	S	S		S	S	S	S					
								Control										
Sex		F			M	M		M	F	M	F							
Age (yrs)		54			63	55		58	54	54	62						57.1	3.93
Ed (yrs)		10			14	13		16	14	11.5	18						13.8	2.48

Note. B = Bulbar. LL = Lower Limb. UL = Upper Limb

Appendix C

Summary of Demographic Information for ALS and control participants at Time 3

	Participants																	
Measures	1	2	3	4	5	6	7	8	9	10	11	12	13	14	15	16	*M*	*SD*
								ALS										
Sex		M			M		F	M		F	M							
Age (yrs)		52			43		58	35		59	61						51	9.47
Ed (yrs)		15			14		16	12		18	10						14.6	2.82
Site at Onset		LL			UL		LL	UL		UL	B							
Familial (F)/ Sporadic (S)		S			S		S	S		S	S							
								Control										
Sex		F			M	M		M	F	M								
Age (yrs)		54			64	55		58	55	54							56.7	3.88
Ed (yrs)		10			14	13		16	14	11.5							13.1	2.11

Note. B = Bulbar. LL = Lower Limb. UL = Upper Limb

Appendix D

Summary of Demographic Information for ALS and Control Participants at Time 4

	Participants																	
Measures	1	2	3	4	5	6	7	8	9	10	11	12	13	14	15	16	*M*	*SD*
								ALS										
Sex					M		F			F	M							
Age (yrs)					44		59			59	61						55.8	7.89
Ed (yrs)					14		16			18	10						14.5	3.42
Site at Onset					UL		LL			UL	B							
Familial (F)/ Sporadic (S)					N		N			N	N							
								Control										
Sex					M			M	F									
Age (yrs)					64			59	55								59.3	4.51
Ed (yrs)					14			16	14								14.7	1.15

Note. B = Bulbar. LL = Lower Limb. UL = Upper Limb

Appendix E

Summary of Demographic Information for ALS and Control Participants for Time 5

	Participants																	
Measures	1	2	3	4	5	6	7	8	9	10	11	12	13	14	15	16	*M*	*SD*
					ALS													
Sex					M						M							
Age (yrs)					44						62						52	12.7
Ed (yrs)					14						10						12	2.83
Site at Onset					UL						B							
Familial (F)/ Sporadic (S)					N						N							
					Control													
Sex								F										
Age (yrs)								55										
Ed (yrs)								14										

Note. B = Bulbar. LL = Lower Limb. UL = Upper Limb

Appendix F

Assessment of Speech Intelligibility

(Cooper & Orange, 2008)

Listener's Name: _____ Age:____ Randomized List: ____ Date:_____

Instructions

Please rank each of the speech samples you will hear using the scale provided below. The left side of the scale represents "completely intelligible" (normal) speech, while the right side of the scale represents "completely unintelligible" (profoundly disordered) speech. The scale provided also indicates reductions in the level of intelligibility change according the following categories of intelligibility deficit:

MI = *Mildly Unintelligible* ***MO*** = Moderately *Unintelligible* ***SE*** = *Severely Unintelligible*

Please make your rating for each sample in its entirety with consideration of the entire scale. You may mark the scale at any point along its length that you believe best corresponds to your judgment of the speech sample relative to your perceived level of intelligibility it represents. The speech sample that you will hear may vary in length and content; however, please make your judgments of intelligibility independent of these variations across speech samples.

Example: _____
MI MO SE

1. _____
MI MO SE

2. _____
MI MO SE

3. _____
MI MO SE

4. _____
MI MO SE

Appendix G

Assessment of Speech Rate

(Cooper & Orange, 2008)

Listener's Name: _____ Age:____ Randomized List: ____ Date:_____

Instructions

Please rate each of the speech samples you will hear using the scale provided below. The scale provided is designed to address "speaking rate". The left side of the scale represents "very slow" speech, while the right side of the scale represents "very fast" speech. The midpoint of the scale should be interpreted to represent a "normal" speaking rate.

Please make your rating for each sample in its entirety with consideration of the entire scale. You may mark the scale at any point along its length that you believe best corresponds to your judgment of the speech sample relative to your perceived judgment of speaking rate. Please make your judgments of speech rate independently, and do not allow other samples, overall intelligibility, language content, accent, etc. to influence your ratings.

Example: _____

Very Slow **Very Fast**

1. _____

Very Slow **Very Fast**

2. _____

Very Slow **Very Fast**

3. _____

Very Slow **Very Fast**

4. _____

Very Slow **Very Fast**

Appendix H

Category fluency scores for ALS Participants at Time 1

Category	1	2	3	4	5	6	7	8	9	10	11	12	13	14	15	16
Animals	20	29	22	13	12	25	27	22	11	17	15	30	23	28	23	9
Household items	34	37	17	11	18	24	30	25	19	20	7	27	20	22	22	17
Birds	15	25	14	10	14	23	20	18	16	10	6	22	14	24	16	11
Vehicles	17	8	9	9	12	19	18	14	8	9	11	17	8	20	17	7
Water Creatures	19	25	6	11	5	16	12	13	9	8	7	17	12	19	19	9
Musical Instruments	20	22	18	11	15	22	16	9	13	14	4	18	11	24	12	10
Dogs	17	28	12	8	11	17	14	17	9	11	3	22	10	15	13	10
Tools	11	23	8	10	9	20	15	16	5	8	6	17	14	8	13	10
Living items	71	107	54	42	42	81	73	70	45	46	31	91	59	86	71	39
Non-living items	82	90	52	41	54	85	79	64	45	51	28	79	53	74	64	44
Total all categories	153	197	106	83	96	166	152	134	90	97	59	170	112	160	135	83

Appendix I

Category fluency scores for ALS Participants at Time 2

Category	2	4	5	6	7	8	10	11	12	13
Animals	30	19	14	19	26	21	13	12	25	20
Household items	25	11	16	23	28	18	20	13	27	16
Birds	23	9	13	20	20	15	11	7	16	11
Vehicles	23	9	8	14	24	15	7	9	16	12
Water Creatures	20	11	8	21	22	19	12	6	17	11
Musical Instruments	19	12	11	16	20	11	14	7	20	14
Dogs	21	11	12	15	19	17	12	6	17	8
Tools	19	11	9	13	15	17	11	5	11	14
Living items	94	50	47	75	87	72	48	31	75	50
Non-living items	86	43	44	66	87	61	52	34	74	56
Total all categories	180	93	91	141	174	133	100	65	149	106

Appendix J

Category fluency scores for Controls at Time 1

Category	1	2	3	4	5	6	7	8	9	10	11	12
Animals	21	24	15	20	20	25	20	18	18	17	23	7
Household items	27	20	21	21	31	19	26	18	25	16	21	24
Birds	18	15	18	12	18	13	19	11	10	9	22	18
Vehicles	10	6	9	9	14	16	18	18	12	11	13	11
Water Creatures	16	13	14	12	16	14	15	9	9	8	16	16
Musical Instruments	20	12	14	12	14	8	14	15	12	7	13	18
Dogs	18	16	13	7	10	9	12	13	11	10	12	17
Tools	17	15	13	9	17	19	16	14	13	12	8	14
Living items	73	68	60	51	64	61	66	51	48	44	73	58
Non-living items	74	53	57	51	76	62	74	65	62	46	55	67
Total all categories	147	121	117	102	140	123	140	116	110	90	128	125

Appendix K

Category fluency scores for Controls at Time 2

Category	2	7	8	10	11	12	13
Animals	23	22	28	19	16	20	27
Household items	20	30	28	24	24	29	27
Birds	20	17	14	17	14	13	19
Vehicles	14	15	15	16	13	15	14
Water creatures	11	16	15	17	14	8	17
Musical instruments	11	13	7	17	14	10	15
Dogs	13	13	10	16	12	12	14
Tools	11	14	18	18	10	11	9
Living items	67	68	67	69	56	53	77
Non-living items	56	72	68	75	61	65	65
Total all categories	123	140	135	144	117	118	142

Appendix L

Category fluency scores for ALS and Control Participants at Time 3

	ALS						*Controls*					
Category	2	5	7	8	10	11	2	7	8	10	11	12
Animals	24	16	27	21	12	19	18	29	32	16	21	17
Household items	32	20	30	16	15	13	23	26	28	24	26	16
Birds	23	16	27	13	10	9	12	14	14	19	13	15
Vehicles	16	14	20	10	14	10	7	9	16	17	11	14
Water creatures	28	13	15	10	16	6	12	21	16	15	15	11
Musical instruments	25	17	17	10	14	8	11	16	8	19	16	8
Dogs	19	10	20	19	16	3	11	13	7	18	9	9
Tools	17	10	12	16	8	9	7	17	17	15	11	10
Living items	94	55	89	63	54	37	53	77	69	68	58	52
Non-living items	90	61	79	52	51	40	48	68	69	75	64	48
Total all categories	184	116	168	115	105	77	101	145	138	143	122	100

Appendix M

Category fluency scores for ALS and Control Participants at Time 4

	ALS				*Control*		
Category	5	7	10	11	7	10	11
Animals	7	30	13	17	24	13	20
Household items	16	31	19	16	22	27	23
Birds	16	23	8	10	20	20	10
Vehicles	11	22	11	7	17	16	13
Water creatures	8	18	12	8	20	13	14
Musical instruments	11	18	11	7	14	20	12
Dogs	13	11	11	4	17	12	10
Tools	9	22	8	7	17	16	10
Living items	44	82	44	39	81	58	54
Non-living items	47	93	49	37	70	79	58
Total all categories	91	175	93	76	151	137	112

Appendix N

Category fluency scores for ALS and Control Participants at Time 4

	ALS		*Control*
Category	5	11	11
Animals	22	21	19
Household items	13	14	20
Birds	16	12	9
Vehicles	8	8	13
Water creatures	9	6	16
Musical instruments	13	4	16
Dogs	12	5	9
Tools	10	7	8
Living items	59	44	53
Non-living items	44	33	57
Total all categories	103	77	110

Appendix O

Definitions of error types

Repetition: An item that was generated more than once within the same sub-category.

Phonemic/literal: An item that sounds similar to a word that belongs to the particular sub-category but one or more of the phonemes is substituted, removed or rearranged within the word (e.g. "mantanees" for "manatees" within the water creatures category).

Verbal: An item that is an actual word but is not related to the particular sub-category (e.g. "humans" listed as a household item).

Semantically related: An item that is related in meaning to the category but does not specifically belong to the category (e.g. "bats" listed as a bird).

Supraordinate semantically related: A supraordinate item that encompasses objects that would be considered a member of the particular sub-category (e.g. "spider" listed as a water creature).

Confabulations: An item that does not exist (e.g., "easy outs" listed as a tool).

Whole/part: An item that is a part of an object that belongs to the particular sub-category (e.g., "caboose" listed for a method of transportation)

Trade name: An item that is a brand name of an exemplar that belongs to a particular sub-category (e.g., "Lysol" listed as a household item)

Appendix P

Slope values for ALS and Control Participants

	ALS										*Control*						
Category	2	4	5	6	7	8	10	11	12	13	2	5	6	8	9	10	11
Animals	-.24	.17	.11	-.17	.56	-1.5	-.44	0.35	-0.2	-.33	-.29	.21	.28	-.43	.41	0	.25
Household items	-.07	0	-.37	-1.0	.53	-.20	-.24	.38	0	0.5	0.5	-.31	.17	.32	-.52	0	0.17
Birds	-.75	-1.0	.88	-.33	0.24	-.40	-.74	.66	-.17	-.33	-.09	.08	1.5	.30	-.32	.32	-.33
Vehicles	.07	0	-.18	-0.2	0.2	-.29	.24	-0.8	-1.0	.25	.03	.04	0	-.91	.63	.35	1.0
Water Creatures	.09	0	.24	0.2	0.1	-.07	.25	0	0	-1.0	-0.5	.41	1.0	.14	.48	0.5	1.0
Musical Instruments	.17	1.0	-.15	-.17	.17	0.5	-.67	0	0.5	.33	-1.5	.32	0	.58	.38	.21	0.5
Dogs	-.20	.33	.58	-0.5	-.07	.75	.12	.29	-0.2	-0.5	-.40	.42	-.43	-.02	-.88	-.21	0.5
Tools	-.32	1.0	1.67	-.14	0.17	0	-.22	.46	-.17	0	-.25	.22	-1.0	.17	-.76	-1.0	1.0
Living items	-.12	.13	.15	-.17	.10	-.16	.01	.28	-.06	-.11	-.11	.16	.23	.05	.14	.16	.25
Non-living items	0	0.5	-.08	-.05	.12	-.15	-.74	.16	-0.2	.11	-.15	-.31	.24	.20	-.39	.01	0.1
Total all categories	.08	0.1	.03	-.04	.09	-.08	-.05	.17	-.05	0.0	-.07	.23	.12	.06	-.05	.03	.07