ARTICULATORY COMPENSATION IN AMYOTROPHIC LATERAL SCLEROSIS: TONGUE AND JAW IN SPEECH

by

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Abstract

This study investigated range, maximum speed, and duration of tongue and jaw movements in Amyotrophic Lateral Sclerosis (ALS; \( n=26 \)) and healthy controls \( (n=16) \). The study objectives were to examine tongue and jaw movements and their interactions at varying stages of bulbar impairment. The patient group was classified based on the severity of bulbar impairment, via the measure of speaking rate. Kinematic measures were obtained from a sentence produced at individual’s comfortable speaking rate and loudness. With ALS, the jaw movements decreased in maximum speed at a later stage of disease compared to the tongue. A positive correlation between range of tongue and jaw movements was observed at an early stage of disease. This correlation was lost at a later stage. Changes in jaw movements may be a compensatory response to tongue impairment. The findings of this study contribute to the understanding of disease progression and speech preservation in ALS.
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INTRODUCTION

ALS as a disease of the motor system

Amyotrophic Lateral Sclerosis (ALS) is a rapidly progressive, fatal neurological disease that affects upper motor neurons (UMN; motor cortical) and lower motor neurons (LMN; brainstem and spinal) (Gubbay et al., 1985; Mulder, 1980; Tandan & Bradley, 1985) that are responsible for controlling voluntary muscles. The disease is the most common of these neuron diseases, which are characterized by the gradual degeneration and death of motor neurons. Approximately 3000 Canadians over 18 years of age currently live with ALS (Hudson, Davenport, & Hader, 1986). The incidence rate is estimated to be 2/100,000 people per year (Hudson, Davenport, & Hader, 1986). Epidemiological studies have observed an equal male-to-female incidence ratio (Tollefsen, Midgren, Bakke, & Fondenes, 2010). ALS can strike at all ages but is most commonly diagnosed in middle and late adulthood with a mean age of onset of 65 years (Chio, 2000). The cause of this disease is unknown and there is no known effective cure.

Patients vary in the locus of disease onset, presentation at diagnosis, and rate of progression (Brooks, 1996). Of all affected individuals, ALS debuts as spinal onset in approximately two thirds and as a bulbar onset in one-third (Haverkamp, Appel, & Appel, 1995; Traynor et al., 1999). The typical spinal onset presents as asymmetric limb weakness and atrophy of a hand or foot. Bulbar involvement typically includes swallowing and speaking difficulties, and is associated with weakened respiratory muscles (Lyall, Donaldson, Polkey, Leigh, & Moxham, 2001; Elleker & Cosio, 1986). About 5% of patients present with respiratory weakness without significant limb or bulbar symptoms (Wijesekera & Leigh, 2009).

ALS is a terminal disease associated with short survival. Eighty percent of people with ALS die within two to five years of diagnosis. Ten percent of those affected may live ten years or longer. The most common cause of death is respiratory failure (Lyall, Donaldson, Polkey, Leigh, & Moxham, 2001). The vast majority of studies have found that age is a strong prognostic factor in ALS; a shorter survival time is associated with a higher age at symptom onset (Caroscio, Calhoun, & Yahr, 1984; Preux et al., 1996). Bulbar onset disease is also an independent
prognostic factor, indicating that bulbar involvement at any stage of the illness significantly shortens survival (Czaplinski, Yen, Simpson, & Appel, 2006; Desport et al., 1999; Traynor et al., 2003; Thijs et al., 2000).

**Bulbar ALS**

Bulbar ALS is perhaps the most devastating form of ALS as it affects the vital functions of airway and nutritional management, as well as the function of communication (Mulder, Bushek, Spring, Karnes, & Dyke, 1983). Although only 30% of patients initially present with bulbar signs and symptoms, approximately 85% of all patients show bulbar disease as ALS progresses (Armon & Moses, 1998; del Aguila, Longstreth, McGuire, Koepsell, & Van Belle, 2003; Millul et al., 2005; Tomik & Guiloff, 2010). Progressive bulbar symptoms are among the most significant contributors to the reduction in quality of life of patients with ALS (Bourke et al., 2006; Lyall, Donaldson, Polkey, Leigh, & Moxham, 2001). Unfortunately, little is known about the natural history of bulbar deterioration. Clinically, a neurologist currently assesses bulbar symptoms with subjective, perceptual methods. For example, tongue strength is assessed by having the patient press the tongue against a finger through the cheek (Kuhnlein et al., 2008) and by listening to slowing of the rate in the dydochokinetic (syllable) repetition task. Additionally, patient self-report plays a central role in the diagnosis of bulbar disease. These methods for clinical assessment of bulbar symptoms lead to late diagnosis. As a result, subgrouping of patients and monitoring disease progression remains challenging. Subgrouping of patients is crucial for the selection of a study sample, particularly with respect to recruitment into clinical trials (Friedman, Lawrence, Furberg, & Demets, 2010). Thus, there is a critical need for the identification of objective markers of bulbar deterioration that would aid in early detection and monitoring disease progression (Turner et al., 2009).

**Clinical Manifestations of Bulbar ALS**

The UMN involvement of the corticobulbar tract in ALS causes supranuclear symptoms, which are also known as pseudobulbar palsy. The clinical characteristics of pseudobulbar palsy are spasticity of the bulbar muscles including the muscles of the jaw, face, soft palate, pharynx, larynx and tongue, as well as emotional lability, and a brisk jaw jerk reflex (Steele, Richardson,
In contrast, degeneration of LMN results in a true bulbar palsy with flaccidity, muscular weakness and eventually atrophy, and fasciculations as primary clinical symptoms. As a result of these neuromuscular changes, patients with bulbar impairment experience difficulty in speech and swallowing. Approximately 95% and 85% of patients diagnosed with bulbar ALS present with dysarthria and dysphagia, respectively (Carpenter, McDonald, & Howard, 1978; Chen & Garrett, 2005; Borasio & Voltz, 1997).

Dysarthria in ALS

Dysarthria is a collective name for a group of speech disorders resulting from disturbances in muscular control over the speech mechanism (Darley, Aronson, and Brown, 1975). It designates problems in oral communication due to paralysis, weakness, or incoordination of the speech musculature. Due to the effects of both upper and lower motor neuron changes, the speech of individuals with ALS is classified as mixed dysarthria with spastic and flaccid characteristics (Darley, Aronson, & Brown, 1969a; 1969b; Duffy, 2012). The features of spastic dysarthria include low pitch, reduced stress, and strained voice quality. Audible inspiration and increased nasality are common indicators of flaccid dysarthria (McGuirt & Blalock, 1980; Aronson, Ramig, Winholtz, & Silber, 1992; Kent, Walker, Weiner, & Miller, 1998; Watts & Vanryckeghem, 2001). With disease progression and increased muscle wasting and atrophy, flaccid symptoms predominate. Although these perceptual features can be identified during assessment and disease monitoring, the reliability of perceptual assessment has been often questioned in speech literature due to its subjective nature (Kent, 1996). Zyski and Weisiger (1987) used recorded samples of dysarthria to determine the inter-rater reliability for classification of types of dysarthria in three groups of listeners with varying clinical experience. The authors found that the perceptual assessment of dysarthria showed low inter-rater reliability. A following study by Zeplin and Kent (1996) replicated the rating studies and also reported low reliability. Thus, the need for objective measures has been stated (Kent, 1996; Bunton & Weismer, 2002).
Speech Intelligibility and Speaking Rate in Bulbar ALS

Speech intelligibility and speaking rate has been described as system-level measurements of bulbar decline as they are influenced by all physiological subsystems including the respiratory, phonatory, articulatory, and resonatory subsystems (Green et al., 2013). Speech intelligibility is defined as the measure of the degree to which a person’s speech can be understood by a listener. Speech intelligibility of dysarthric talkers has been evaluated in several studies using tests of single-words (Yorkston & Beukelman, 1981; Kent, Weismer, Kent, & Rosenbек, 1989; Mulligan et al., 1994), and sentences (Yorkston & Beukelman, 1978; 1981; Hammen & Yorkston, 1996; Weismer, Yunusova, & Westbury, 2003) and has been used as a measure of the severity of dysarthria. As a result of speech changes, individuals with ALS exhibit a loss of speech intelligibility requiring interventions focused on augmentative alternative means of communication (Ball, Beukelman, & Pattee, 2002; DePaul and Kent, 2000; Kent et al., 1991; Mulligan et al., 1994). Currently, the monitoring of speech deficits is in the center of clinical management of patients with bulbar ALS. However, speech intelligibility is far from being optimal for this purpose as numerous studies have indicated that bulbar motor dysfunction occurs prior to perceived changes in speech intelligibility (Ball, Beukelman, & Pattee, 2002, DePaul & Brooks, 1993; Kent et al., 1990; Mefferd, Green & Pattee, 2012; Nishio & Niimi, 2000; Yorkston, Strand, Miller, Hillel, & Smith, 1993; Mefferd, Nichols, Pakiz, & Rock, 2007; Kent, 2000). Therefore, although speech intelligibility is the behavioural standard of communication and important to be monitored as disease progresses, its measurement is not optimal for quantification of changes in speech musculature, particularly early in the disease.

Several investigators have advocated for monitoring speaking rate as a more appropriate and useful measure of bulbar disease (Yorkston, Strand, Miller, Hillel, & Smith, 1993). Slow speaking rate is a hallmark characteristic of dysarthria in ALS (Duffy, 2012; Yorkston, Strand, Miller, Hillel, & Smith, 1993) and its decline is seen before that of speech intelligibility (Ball, Willis, Beukelman, & Pattee, 2001). A normal rate of speech has been established for sentence reading tasks, with a range of 160-230 WPM (words per minute; Turner, Tjaden, & Weismer, 1995). In the ALS population, a speaking rate of 120 WPM or higher is associated with highly intelligible speech (Yorkston, Strand, Miller, Hillel, & Smith, 1993; Ball, Willis, Beukelman, & Pattee, 2001). A reduction in speaking rate below approximately 120 WPM is a hallmark in the
disease progression. At this point, a rapid deterioration in speech intelligibility typically occurs (Yorkston, Strand, Miller, Hillel, & Smith, 1993; Ball, Willis, Beukelman, & Pattee, 2001; Yunusova, Green, & Mefferd, 2009). This finding was reported crosssectionally in a retrospective study of more than a hundred clinical cases (Yorkston, Strand, Miller, Hillel, & Smith, 1993; Ball, Willis, Beukelman, & Pattee, 2001) and longitudinally (Yunusova, Green, & Mefferd, 2009), suggesting that speaking rate may be a valid measure to predict upcoming changes in intelligibility. In addition to detecting changes earlier than intelligibility, speaking rate decreases in a linear fashion as bulbar ALS progresses (Yorkston, Strand, Miller, Hillel, & Smith, 1993; Yunusova et al., 2010). This suggests that speaking rate might be more sensitive to bulbar disease progression than speech intelligibility.

One of the major problems with using system level measures, including speaking rate, however is that these measures are not sensitive to subsystem impairment. A decline in speaking rate is a result of multiple possible sources of impairment across various motor-speech subsystems (e.g., respiratory, phonatory, articulatory, and resonatory; Green et al., 2013). Preliminary findings suggest that subsystem variables appear to be more sensitive to disease progression than system level variables (Yunusova, Green, Wang, Pattee, & Zinman, 2011). Thus, a greater understanding of bulbar impairment within subsystems is needed in order to develop sensitive outcome measures, as well as to predict changes in speech with disease progression.

Articulatory Speech Subsystem: Pathophysiological Studies

The articulatory subsystem consists of the oral articulators (i.e., the tongue, lips and jaw). Among other speech subsystems, changes within the articulatory subsystem have been most consistently associated with changes in speech intelligibility (Kent et al., 1992; Yunusova, Weismer, Westbury, & Lindstrom, 2008; Yunusova et al., 2012). The articulatory subsystem has primarily been investigated acoustically. Articulatory events are usually indexed acoustically by measures of formant frequencies, which are defined as the bands of frequency that determine the phonetic quality of a vowel or a consonant. The slope of the second formant (F2) extracted from onset and offset of vowels has been investigated in dysarthria. Kent and colleagues (1989) performed acoustic analyses of speech in ALS and noted that the F2 slope was often reduced in ALS compared to healthy controls (Kent, Weismer, Kent, & Rosenbek, 1989). This reduction
was interpreted as a relative slowness in changing the vocal tract configuration in patients with neurological conditions (Stevens, 2000). Reductions in F2 slope have been associated to a great degree with the loss of speech intelligibility (Kent, Weismer, Kent, Vorperian, & Duffy, 1999; Kent, Weismer, Kent, & Rosenbek, 1989; Yunusova et al., 2012; Kim, Weismer, Kent, & Duffy). It was shown that in men with ALS, the average F2 slope was correlated with overall speech intelligibility scores (Kent et al., 1990). In addition, it was reported that the longitudinal decline in overall speech intelligibility paralleled a consistent decline in F2 slopes (Kent et al., 1991). However, one major limitation of assessing the articulatory subsystem acoustically is that acoustic signal represents the actions of the entire articulatory subsystem. Consequently, disease-related effects on individual articulators are not well reflected acoustically. In order to understand the contribution of individual articulators to intelligible speech, structures within the articulatory subsystem need to be investigated individually.

When studied individually, articulatory organs reveal non-uniform effects in ALS. An early neuropathological study examining the brainstem in 53 cases with ALS (Lawyer, & Netsky, 1953) reported more degeneration of hypoglossal nerve fibers, those that innervate tongue muscles, in comparison to trigeminal or facial nerve fibers, those that innervate the jaw and lips, respectively. Their results suggested that the tongue may be affected to a greater extent than the jaw and the lips in the disease. Similarly, in a chart review of 441 cases of ALS, Carpenter and colleagues (1978) found that most patients (72%) had clinical symptoms and signs of tongue weakness and only a small group (31%) showed signs of jaw weakness, supporting the disproportionate involvement of hypoglossal motor neurons as compared to trigeminal motor neurons.

The documentation of non-uniform impairment has also been investigated in physiological measures of force generation in bulbar musculature. Maximum strength studies have been used to quantify weakness in the orofacial motor system using specialized strain gauge manometry (DePaul, Abbs, Caliguiri, Gracco, & Brooks, 1988; 1993; Shaker, Cook, Dodds, & Hogan, 1988; Nagao, Kitaoka, Kawano, Komoda, & Ichikawa, 2002; Dworkin & Aronson, 1986; Langmore & Lehman, 1994) and bulb pressure sensors (i.e. the Iowa Oral Performance Instrument; IOPI; Robin, Goel, Somodi, & Luschei, 1992; Solomon, Robin, & Luschei, 2000; Crow & Ship, 1996). Muscular weakness was evaluated using isometric maximum voluntary contractions (MVC)
using these instruments. Early reports consistently demonstrated greater impairment relative to healthy controls in the tongue compared to the jaw and lower lip, even among ALS patients without bulbar signs or symptoms (Dworkin & Hartman, 1979; Dworkin, 1980; Dworkin, Aronson, & Mulder, 1980; DePaul et al., 1988; 1993; Langmore, & Lehman, 1994).

Strength measures in the bulbar system have their limitations, however. They have been deemed unsuitable for clinical trials due to their large variability even between healthy individuals (Robin et al., 2000), making identification of a strength deficit a very challenging task. Strength measures are also difficult to obtain without the accompaniment of contamination by other muscle groups (Cook & Soutter-Glass, 1987; deBoer, Boukes, & Sterk, 1982; Dworkin & Aronson, 1986). For example, MVC of the tongue may be contaminated by a jaw effort as it is difficult to eliminate the co-contraction of jaw musculature in a tongue task (i.e., tongue elevation; Solomon, 2004). However, the biggest disadvantage of these measures is their lack of association to speech changes (Weismer, 2006). Tongue strength has been shown to be a poor predictor of speech proficiency (Dworkin, 1980). Although considered a valid index of muscle force generating capacity, isometric MVC is a static task and indirect measure of motor unit integrity and thus, may not quantify tasks requiring dynamic continuous muscle function, such as speech. Further studies investigating the structures within the articulatory subsystem in a speech context are needed.

**Kinematic Studies in ALS**

As mentioned previously, perceptual and acoustic characteristics of ALS speech deficits have been relatively well studied (Darley, Aronson, & Brown, 1975; Hirose, Kiritani, & Sawashima, 1982; Hirose, Kiritani, Ushijima & Sawashima, 1978; Kent et al., 1989, 1990, 1992; Tjaden & Turner, 1997; Turner & Tjaden, 2000; Turner & Weismer, 1993; Weismer et al., 2001, 1988, 1992; Duffy, 2005). However, these measures can be clinically meaningful only if linked to articulatory events so that foci of impairment and treatment can be determined. Kinematic studies, compared to acoustic studies, allow a speech subsystem approach to the assessment of bulbar impairment. Particularly, they can aid in the understanding of articulatory subsystem motor involvement. Thus far, articulatory kinematic studies in ALS are limited. The lack of kinematic studies has partly been due to the logistical difficulties posed by the inaccessibility of
the bulbar system to direct observation (e.g., the tongue is almost fully hidden in the mouth). Another contributing factor to the limited research has been the lack of technology enabling the measurement of the articulatory movements. However, recent development and improvements in electromagnetic tracking systems (e.g., WAVE Speech System, NDI, Canada; EMA, Carstens Medizinelectronik, Germany) render this line of research possible.

Speech is the product of highly coordinated movements of the tongue, lips, and jaw. Testing of articulatory involvement can be achieved through measures of movement. Multiple features of motor performance can be evaluated from speech movement recordings including movement size, speed and working spaces (Berry, 2011) of individual articulators, as well as the temporal and spatial coordination of oral articulators (Green, Moore, Higashikawa, & Steeve, 2000). Thus far, only a handful of studies have examined speech kinematics in ALS. Existing studies investigating the tongue observed smaller and slower speech movements in individuals with ALS. An early case study by Kent, Netsell, and Bauer (1975) investigated jaw, lips, and tongue movements during syllables using a cineradiography technique in 4 talkers and observed reduced movement size for all articulators. Subsequent case studies used the x-ray microbeam (Westbury, 1994) and suggested a reduction in movement size, as well as speed, for tongue movements during fastest rate of syllable repetition (Hirose, 1982) and word productions (Kuruvilla, Green, Yunusova, & Hanford, 2012; Yunusova et al., 2008; 2012). The limited literature on tongue kinematics warrants the need for further investigation on the natural history of changes in tongue movements in ALS.

Although very few, most kinematic studies in ALS have examined disease-related effects on jaw movements. Articulatory speeds of jaw movements were observed to be impaired during vowels (Yunusova et al., 2008), as well as with alternating motion rate (AMR) tasks (Mefferd, Green, & Pattee, 2012; Kent, Netsell, and Bauer, 1975). However, other studies reported contradictory findings demonstrating exaggerated displacements of the jaw during fast rate of speech (Hirose et al., 1982; Mefferd, Green, & Pattee, 2012) and word productions (Yunusova, Weismer, Westbury, & Lindstrom, 2008). A larger than normal jaw movement has been interpreted as compensatory in response to a significantly more affected tongue (Yunusova, Weismer, Westbury, & Lindstrom, 2008; Mefferd. Green, & Pattee, 2012). Indeed, case studies observed that tongue movements of talkers with ALS showed a greater dependency on jaw movements to
acquire spatial targets during speech in comparison to healthy talkers (Hirose et al., 1982; Mefferd, Green, & Pattee, 2012). Kinematics of both the tongue and jaw, however, have rarely been studied together within the same talker, except for one study (see Hirose, Kiritani, & Sawashima, 1982). In order to gain a better understanding of the compensatory behaviours between articulators, both tongue and jaw need to be examined together.

In summary, the few studies that have investigated articulatory behavior of the tongue and jaw showed that, relative to healthy controls, individuals with ALS exhibited slower and smaller tongue movements during speech and syllable repetitions tasks (Hirose et al., 1982a, 1982b; Yunusova, Weismer, Westbury, & Lindstrom, 2008). They exhibited larger jaw movements during word productions (Yunusova et al., 2008) and syllable repetition tasks (Mefferd, Green, & Pattee, 2012). Existing kinematic studies had a very small sample size (n < 8), didn’t account for bulbar disease severity, and rarely investigated the tongue and jaw together. Also, most studies have investigated alternating motion rate (AMR) or speech-like tasks (Mefferd, Green, & Pattee, 2012; Hirose et al., 1982a; 1982b; Kent et al., 1975). Speech movements at the sentence level have never been investigated, particularly in the context of their usefulness in the diagnostic process.

In this study, we aimed to examine changes in kinematic measures for the tongue and jaw in a group of talkers with ALS who ranged in severity of bulbar disease. We compared their performance to that of age-matched healthy controls. The controls acted as baseline normative data because sentence-level analyses for this age group have not been previously established in a healthy population. The overall objective of this study was to investigate how changes in tongue and jaw movements relate to system level changes in bulbar ALS (e.g. speaking rate and intelligibility). This thesis had two specific objectives. The first objective was to determine tongue and jaw kinematics during speech at varying stages of bulbar impairment. Based on existing literature, we hypothesized that speech movements would differ across stages of bulbar impairment. At more severe stages of disease, tongue movements will decrease in speed and size while jaw movements will increase in speed and size. The second objective was to examine the interactions between tongue and jaw speech movements at varying stages of bulbar impairment. Based on the nature of differential impairment in ALS, we hypothesized that there would be differences in correlations between tongue and jaw speech movements at certain stages of bulbar
impairment. In particular, we hypothesized a negative correlation between tongue and jaw movement measures at a mild stage of disease, thus serving as a potential indicator of compensatory interactions between articulators.
METHODS

Participants

Participants were recruited from the ALS/MND Clinic at the Sunnybrook Health Sciences Center, University of Toronto for this cross-sectional study. Participants were selected from a larger pool \(n = 143\) of patients undergoing a 5-year longitudinal study of bulbar deterioration in ALS. All participants were diagnosed with possible, probable or definite ALS as defined by the El Escorial Criteria from the World Federation of Neurology (Brooks, Miller, Swash, & Munsat, 2000) by a neurologist. All participants exhibited bulbar involvement in at least one region of the speech system (e.g. voice, soft palate, tongue, and/or face). Participants were native speakers of English. All participants passed a hearing screening and were screened for cognitive dysfunction using The Montreal Cognitive Assessment (MoCA; Nasreddine et al., 2005). Participants had no history of significant health, cognitive, or sensory problems or a history of other neurologic conditions (e.g. stroke). Participants were excluded if they reported taking any medications known to affect speech production (see Forshew & Bromberg, 2003).

Twenty-six participants (19 males and 7 females) diagnosed with ALS were included in the study. Only participants whose kinematic data was collected using the NDI Wave Speech System were included \(n=32\). The patients for the current study were selected based on the completeness of the dataset – both tongue and jaw data had to be collected in the speech task for the patient to be included in this analysis \(n = 26\). Complete sessions with lowest intelligibility and speaking rate scores were preferred.

The control group comprised of 6 males and 10 females recruited from the University of Toronto. Participants had no history of significant health, cognitive, or sensory problems or a history of other neurologic conditions. Participants were excluded if they reported taking any medications known to affect speech production (i.e. Nasonex).

All talkers were native speakers of English. The study was approved by the REB of the Sunnybrook Research Institute and University of Toronto; all participants signed informed consent according to the requirements of the Declaration of Helsinki.
Speech Sample

The data were obtained as part of a larger protocol, where functions of each subsystem were evaluated in the following order: (1) Laryngeal tasks (2) Respiratory tasks (3) Velopharyngeal tasks, and (4) Articulatory tasks. The protocol took under 30 minutes for healthy speakers and could have lasted up to an hour for more impaired talkers. The order of tasks was preserved between talkers and sessions in order to ensure the completeness of the protocol longitudinally.

The recorded task consisted of a sentence, *Buy bobby a Puppy*, read at a comfortable reading rate and loudness and repeated 10 times. This sentence was chosen in order to elicit large jaw movements and complex tongue movements (i.e. the diphthong ‘ai’ in ‘Buy’) (Kleinow & Smith, 2000; Smith & Zalaznik, 2004; Kleinow, Smith, & Ramig, 2001; McHenry, 2003; Yunusova, Green, Wang, Pattee, & Zinman, 2011).

Instrumentation

Articulatory movements of the tongue and jaw were collected with an electromagnetic tracking device (the Wave Speech system; NDI, Canada), which records the position and rotation of small sensors that are attached to the tongue and jaw with an accuracy of < 0.5mm (Berry, 2011). The system tracks articulatory movements during speech at a sampling rate of 400 Hz and uses a combination of 5 and 6-degree-of-freedom (5DOF and 6DOF) sensors to record motions in a calibrated volume (30 x 30 x 30 cm). Jaw movements were obtained by attaching a small 5DOF sensor to the gums under the bottom incisors. Tongue movements were obtained by attaching one small 5DOF sensor to the tongue blade, approximately 20mm from the tongue tip. Placement of tongue sensors was determined by measuring the distance from tongue tip to sensor with the use of a desk ruler. The system and tongue sensor position is presented in Figure 1.
Articulatory movements were corrected for head movements during acquisition (see Berry, 2011). Post-acquisition, the data were transferred into SMASH (Green, Wang, & Wilson, 2013), a MATLAB-based custom speech analysis program, where signals were manually checked for tracking errors and low-pass filtered at 15 Hz using a zero-phase shift forward and reverse digital filter.

In order to study tongue movements independent of the jaw (T-J), the jaw (J) movements were subtracted from the tongue movements using a linear subtraction method (LSM; McClean, 2000; Hertrich & Ackermann, 2000; Westbury, Lindstrom, & McClean, 2002) post acquisition by subtracting jaw coordinates from the tongue coordinate system along the horizontal, vertical, and lateral axes (Westbury, Lindstrom, & McClean, 2002).

Acoustic signals were recorded simultaneously with kinematic signals directly onto a hard drive of a computer at the sampling rate of 22 KHz and 16-bit resolution. A high quality lapel microphone (Countryman B3P4FF05B) was positioned approximately 15 cm from the mouth during the recordings.
Upper and lower lip aperture (UL-LL) time history was used for parsing movement traces as shown in Figure 2. Vertical lines mark the point of minimal UL – LL distance prior to the acoustic onset and following the last Consonant-Vowel-Consonant syllable of the utterance.

**Figure 2.** Upper and lower lip aperture (UL-LL) time history was used for identification of the onset and offset of movement traces in the sentence *Buy Bobby a Puppy.*

**Kinematic Measures**

Kinematic measures were selected based on prior studies demonstrating their sensitivity to bulbar dysfunction in ALS (Kent, Netsell, and Bauer, 1975; Hirose et al., 1982, Yunusova et al, 2008, 2010; 2012; Mefferd, Green, & Pattee, 2012; Kuruvilla, Green, Yunusova, & Hanford, 2012). The following measures were derived from J and T-J:

1. **Range (mm) of movement**, a measure representative of movement size, was calculated as the difference between the maximum and minimum values of the 3D Euclidean distance movement history of the sentence.
2. Maximum speed (mm/s) of movement was calculated as the maximum value of the absolute values of the first derivative of the 3D Euclidean movement history.

3. Duration (sec) was calculated as the difference between the offset and onset time of the sentence defined kinematically based on the lip aperture signal (see Figure 2).

Speech Intelligibility and Speaking Rate

Speech intelligibility and speaking rate were obtained for each speaker and session using the Sentence Intelligibility Test (SIT; Beukelman, Yorkston, Haken & Dorsey, 2007). These measures were essential because they are current clinical “gold standards” for characterizing bulbar speech performance. They provide an indication of the functional status of the speech production system as a whole and quantify the severity of speech impairment. The SIT software generated a random list of 11 sentences of increasing length (from 5 to 15 words). Participants were asked to read the list of these sentences at the beginning of each recording session. Each participant had a unique list. A single naive listener, who was unfamiliar to the participants’ speech and stimuli, transcribed the sentences orthographically and measured sentences durations. The listener was inexperienced with respect to knowledge of dysarthria. This method has been a standard procedure for intelligibility assessment and published previously (Green, Beukelman, & Ball, 2004; Green et al., 2013; Yunusova et al., 2009; 2010). The SIT software automatically calculated speech intelligibility, expressed as the percent of total words transcribed correctly by the listener. Speaking rate (words per minute) was also calculated by this software, using information about sentence onset and offset provided by the listener. This method of establishing speech intelligibility and speaking rate measures has been commonly used in ALS research (Ball, Beukelman, & Pattee, 2004; Ball et al., 2001; Kent, Weismer, Kent, & Rosenbek, 1989; Yorkston & Beukelman, 1980; Yunusova et al., 2010; 2011).

Patient Classification by Severity

Participants with ALS were categorized into three groups based on their speaking rates and speech intelligibility: ALS 1, normal speaking rate (>160 wpm) and intact intelligibility (>98%); ALS 2, reduced speaking rate (120-160 wpm) and intact intelligibility (>98%); and ALS 3, slow
speaking rate (<120 wpm) and impaired intelligibility (<92%). The cutoff of 160 wpm was chosen because healthy talkers exhibit a speaking rate of greater than 160 wpm during sentence reading tasks (Turner, Tjaden, & Weismer, 1995; Yorkston et al., 1993). The cutoff of 120 wpm was chosen because it has been previously identified as the point in disease progression when speech intelligibility begins to decline rapidly (Yorkston et al., 1993). The ALS 3 (severe) group was composed of 2 talkers only; however, since the main focus of kinematic analyses was to detect early changes in speech, the analyses continued with this small N.

Amyotrophic Lateral Sclerosis Functional Rating Scale-Revised

The clinical manifestation of overall and bulbar dysfunction was assessed for each speaker and session using the Amyotrophic Lateral Sclerosis Functional Rating Scale-Revised (ALSFRS-R; total and bulbar subscores; Cedarbaum et al., 1999). The ALSFRS-R is a validated rating instrument of global function in patients with ALS (Cedarbaum et al., 1999). It includes three questions related to speech, swallowing and salivation function with a maximum score of 12, indicating intact bulbar function. Values below 12 indicate bulbar impairment in one, or all, bulbar functions. A change in ALSFRS-R score and its ratio are highly correlated with survival time (Kaufmann et al., 2005; Gordon & Cheung, 2006) and sensitive to disease progression (Kimura et al., 2006; Kollewe et al, 2008).

Statistical Analyses

All statistical analyses were conducted using IBM SPSS Statistics Version 20. Analyses were performed on across-repetition averages for each speaker in each group. Speaking rate and all kinematic variables were normally distributed for healthy controls and participants with ALS, as determined by the Shapiro-Wilk test. The assumption of homogeneity of variance was met for all kinematic variables, as determined by Levene’s Test of Equality of Variance. For objective 1, group (healthy, ALS1, ALS2, ALS3) differences were tested for each J and T-J kinematic using a One-way ANOVA at an alpha level of .05. Post-hoc pairwise comparisons were conducted using Tukey’s HSD. Tukey’s HSD compares the difference between each pair of means with appropriate adjustment to \( p \)-values for multiple testing; Tukey’s method uses the highest and lowest sample differences as the determining aspect between all other pairs of populations.
(Mendehall et al., 2009). For objective 2, interactions between T-J and K were assessed using Pearson’s Correlation Coefficients ($r$) for each kinematic measure.

In order to compare the LSM decoupling method to a standard method of tongue isolation in speech literature, range and maximum speed of T-J movements were statistically compared to independent tongue movements obtained via the bite block method (see Gay, Lindblom and Lubker, 1981; Solomon & Munson, 2004; Flege, 1989; Riley, 2013; Mefferd, Green, & Pattee, 2012). One-way ANOVAs were conducted for both methods and results were compared. Pearson’s Correlation Coefficients were conducted for comparisons of the two methods for range and maximum speed of tongue movements.
RESULTS

Participant Characteristics

Participant characteristics such as age, sex, speech characteristics and global function scores (i.e. ALSFRS-R; Cedarbaum et al., 1999) are summarized in Table 1. There were no significant differences in age between talkers with ALS and healthy controls ($t (36) = -4.08, p = .068$).

Table 1. Participant characteristics for talkers with ALS and healthy talkers (Control); group means and standard deviations (in parentheses) are shown. WPM= words per minute.

<table>
<thead>
<tr>
<th></th>
<th>Sex, n</th>
<th>Age, years</th>
<th>ALSFRS-R total score</th>
<th>ALSFRS-R bulbar subscore</th>
<th>Speaking Rate, wpm;</th>
<th>Sentence Intelligibility, %</th>
<th>Site of Onset (spinal, bulbar)</th>
</tr>
</thead>
<tbody>
<tr>
<td>ALS</td>
<td>M(19)</td>
<td>61.4</td>
<td>33.50</td>
<td>10.50</td>
<td>167.77</td>
<td>97.07</td>
<td>13,2</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>(7.8)</td>
<td>(8.46)</td>
<td>(7.58)</td>
<td>(39.80)</td>
<td></td>
</tr>
<tr>
<td></td>
<td>F(7)</td>
<td>59.5</td>
<td>32.28</td>
<td>9.45</td>
<td>162.68</td>
<td>99.39</td>
<td>5,1</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>(5.0)</td>
<td>(7.02)</td>
<td>(3.04)</td>
<td>(24.24)</td>
<td></td>
</tr>
<tr>
<td>Control</td>
<td>M(6)</td>
<td>69.2</td>
<td>-</td>
<td>-</td>
<td>153.65</td>
<td>97.79</td>
<td>-</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>(1.6)</td>
<td></td>
<td>(8.32)</td>
<td>(2.99)</td>
<td></td>
</tr>
<tr>
<td></td>
<td>F(10)</td>
<td>69.7</td>
<td>-</td>
<td>-</td>
<td>154.97</td>
<td>99.14</td>
<td>-</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>(3.8)</td>
<td></td>
<td>(16.22)</td>
<td>(0.94)</td>
<td></td>
</tr>
</tbody>
</table>

Speech Intelligibility, Speaking Rate and ALSFRS-R

Patient distributions by subgroup are shown in Table 2. The ALS group showed intelligibility scores between 74.45% and 100% (mean=97.63, SD=5.33) and speaking rate scores between 74.32 WPM and 230.70 WPM (mean=166.55, SD=36.27). Patients in ALS1 and ALS2 groups showed intelligibility within normal limits (>98%). The speaking rate significantly decreased by
25% between these subgroups, \( t(21) = 6.99, \ p = .001 \). Patients in the ALS 3 \((n=2)\) group showed significantly reduced speech intelligibility and speaking rate (see Table 2). Correlation analysis between ALSFRS-R bulbar subscores, the clinical standard to detect bulbar impairment, and speaking rates showed that these measures were significantly correlated, \( r (42) = .545, \ p = .001 \).

Table 2. Participant characteristics for ALS subgroups; normal speaking rate (ALS1; >160 wpm), reduced speaking rate (ALS2; 120-160 wpm), and slow speaking rate (ALS3; <120 wpm). Means and standard deviations (in parentheses) are reported for each severity subgroup.

<table>
<thead>
<tr>
<th>Patient Subgroup</th>
<th>( n )</th>
<th>Age (mean,SD)</th>
<th>ALSFRS-R total score (mean,SD)</th>
<th>ALSFRS-R bulbar subscore (mean,SD)</th>
<th>Speaking Rate (wpm; mean,SD)</th>
<th>Sentence Intelligibility (%; mean,SD)</th>
</tr>
</thead>
<tbody>
<tr>
<td>ALS1</td>
<td>15</td>
<td>60.3 (8.5)</td>
<td>33.45 (6.49)</td>
<td>11.20 (1.28)</td>
<td>192.18 (18.15)</td>
<td>99.28 (0.73)</td>
</tr>
<tr>
<td>ALS2</td>
<td>9</td>
<td>62.8(6.7)</td>
<td>31.72 (10.39)</td>
<td>9.27 (1.95)</td>
<td>143.51 (12.72)</td>
<td>98.77 (3.49)</td>
</tr>
<tr>
<td>ALS3</td>
<td>2</td>
<td>59.0(1.4)</td>
<td>33.92 (8.25)</td>
<td>9.07 (1.44)</td>
<td>90.81 (23.32)</td>
<td>85.41 (15.49)</td>
</tr>
</tbody>
</table>

**Group Differences for Kinematic Measures**

Objective one of the study was to determine the effect of varying stages of bulbar impairment on tongue and jaw movements during speech. Jaw measures were subjected to one-way analyses of variance with four levels of group (controls, ALS1, ALS2, ALS3). Group effect for jaw range was non-significant, suggesting that size of jaw movements do not differ at different stages of disease. Group effect for maximum speed of jaw movements was non-significant as well. However, descriptively jaw movements showed a decrease with changes in severity with the largest drop between groups ALS2 to ALS3 (see Figure 3a).

Tongue measures were also subjected to one-way analyses of variance with four levels of group (controls, ALS1, ALS2, ALS3). Group effect for tongue range was non-significant, suggesting that size of tongue movements do not differ at different stages of disease. Group effect for
maximum speed of tongue movements was non-significant. However, descriptively tongue movements showed a decrease with disease progression, with the largest drop from ALS1 to ALS2 (see Figure 3b).

**Figure 3.** The mean values and standard deviations for maximum speed (mm/s) of a) jaw and b) tongue movements in healthy controls ($n = 16$), ALS1 ($n = 15$), ALS2 ($n = 9$), and ALS3 ($n = 3$). Jaw movements show maximum decrease from ALS2 to ALS3, while tongue movements show maximum decrease from ALS1 to ALS2.

Figure 4 shows changes in movement durations at different stages of ALS. A significant main effect of group on duration of tongue and jaw movements was observed, $F(3,32) = 11.35$, $p < .001$. A post hoc Tukey test showed that ALS3 had significantly larger durations than control, ALS1, and ALS2 subgroups.
Figure 4. The mean values and standard deviations for duration (s) of jaw and tongue movements in healthy controls, ALS1, ALS2, and ALS3. Square brackets signify significant pairwise differences.

Tongue-Jaw Interactions

Objective 2 of the study was to examine the interactions between tongue and jaw speech movements at varying stages of bulbar impairment. Pearson product-moment correlation coefficients ($r$) were computed to assess the relationships between the tongue and jaw for movement range and maximum speed at varying stages of bulbar impairment. ALS3 was omitted from these analyses due to the small sample size of the group. The graphical (see Figure 5) and statistical results revealed a significant negative correlation between tongue and jaw range in healthy talkers, ($r$ (13) = -.482, $p = .049$). ALS1 showed a strong positive correlation between tongue and jaw for range of movement, $r$ (12) = .766, $p = .001$. The correlation between range of movements for tongue and jaw in ALS2 was non-significant. The correlations between maximum speed of movements for tongue and jaw in all groups were non-significant (see Table 3).
Figure 5. Associations between the tongue and jaw for range (mm) of movements in healthy talkers, ALS1, and ALS2. Pearson product-moment correlation coefficients ($r$) and $p$ values are shown.

Table 3. Pearson product-moment correlation coefficients ($r$) and $p$-values for tongue and jaw range (mm) and maximum speed (mm/s) by subgroup; healthy controls, normal speaking rate (ALS1; >160 wpm), and reduced speaking rate (ALS2; 120-160 wpm). * = significant at $p < .05$, ** = significant at $p < .001$.

<table>
<thead>
<tr>
<th>Kinematic Variables</th>
<th>Group</th>
<th>$r$</th>
<th>$p$</th>
</tr>
</thead>
<tbody>
<tr>
<td>Range (mm)</td>
<td>Control</td>
<td>-.482*</td>
<td>.049</td>
</tr>
<tr>
<td></td>
<td>ALS1</td>
<td>.766**</td>
<td>.001</td>
</tr>
<tr>
<td></td>
<td>ALS2</td>
<td>-.106</td>
<td>.787</td>
</tr>
<tr>
<td>Maximum Speed (mm/s)</td>
<td>Control</td>
<td>-.402</td>
<td>.137</td>
</tr>
<tr>
<td></td>
<td>ALS1</td>
<td>.295</td>
<td>.329</td>
</tr>
<tr>
<td></td>
<td>ALS2</td>
<td>-.024</td>
<td>.959</td>
</tr>
</tbody>
</table>
Comparing LSM to the Bite Block Method of Jaw Subtraction

Significant group differences were not observed for tongue range or maximum speed in either of the two decoupling methods.

Independent tongue movements using the LSM method and the bite block method were significantly correlated for range and maximum speed of movements, \( r = .799, p = .003 \), \( r = .955, p = .001 \), respectively.
DISCUSSION

Summary

The primary objectives of the study were to determine the effects of varying stages of bulbar impairment, as identified by differences in speaking rate, on tongue and jaw movements and their interactions. Movements of the jaw and tongue independent of the jaw during speech were studied cross-sectionally in a group of talkers with bulbar ALS. We found that, with disease progression, tongue and jaw movements during a sentence production did not significantly change in size. However, tongue movements showed a tendency to decrease in maximum speed with the maximum decrease at an early stage of disease, when speaking rate was high and intelligibility was still intact. Meanwhile, jaw movements appeared to decrease in maximum speed with a maximum decrease at a severe stage of disease, when speaking rates and intelligibility were severely impaired. Duration of articulatory movements increased only at the stage of disease with impaired intelligibility. Tongue and jaw movement range showed a significant positive correlation at a mild stage of disease, when intelligibility and speaking rates were not yet affected. This correlation was not seen at a later stage of disease. Findings are interpreted below with respect to the issue of task dependency in the diagnostic process and the compensatory role of the jaw in the preservation of speech function in ALS.

Natural History of Changes in Tongue and Jaw Movements in ALS

Previous research showed that the tongue and jaw kinematics were affected in ALS (Hirose et al., 1982; Kent, Netsell, and Bauer, 1975; Kuruvilla et al., 2012; Yunusova et al., 2008; 2010; 2012). Specifically, tongue movements during syllable and segment production were reduced in size (Hirose et al., 1982; Kent, Netsell, and Bauer, 1975; Weismer, Yunusova, & Westbury, 2003; Kuruvilla, Green, Yunusova, & Hanford, 2012; Yunusova et al., 2008; 2012) and speed (Hirose et al., 1982; Weismer, Yunusova, & Westbury, 2003; Kuruvilla, Green, Yunusova, & Hanford, 2012; Yunusova et al., 2012) in patients with ALS compared to healthy controls. Studies also identified a reduction in size (Kent, Netsell, & Bauer, 1975) and speed (Mefferd, Green, & Pattee, 2012; Hirose et al., 1982) of jaw movements compared to healthy controls in similar tasks. However, other studies investigating jaw kinematics in ALS identified an increase
in movement size and speed compared to healthy controls (Mefferd, Green, & Pattee, 2012; Hirose et al., 1982; Yunusova, Weismer, Westbury, & Lindstrom, 2008) and longitudinally (Yunusova et al., 2010).

In our study, the range of tongue and jaw movements did not differ compared to healthy controls or at different stages of disease. Our findings are not consistent with the results of previous research reporting reduced tongue movement sizes compared to healthy individuals. This discrepancy may be due the differences in the severity of disease in patients studied in different studies. The majority of published works focused on reporting results for individuals with speech intelligibility deficits (Yunusova et al., 2008; Hirose et al., 1982; Kuruvilla, Green, Yunusova, & Hanford, 2012). In this study, we focused on earlier stages of disease progression, when intelligibility was minimally affected. Individuals with ALS may only exhibit smaller tongue movements during speech at severe stages of speech impairment.

Maximum speed of tongue and jaw movements during speaking did not show significant differences with increasing disease severity. However, tongue movements tended to decrease in maximum speed by approximately 30% in the group characterized by the initial slowing of the rate (ALS2), compared to the essentially normal performance group (ALS1). Meanwhile, maximum speed of jaw movements remained high and decreased only in ALS3 group, when speaking rate reached below 120 wpm and speech intelligibility declined. In support of these findings, longitudinal data by Yunusova and colleagues (2010) found that jaw movements increased in speed in three individuals at a stage immediately prior to a loss of intelligibility.

With disease progression, articulatory movements showed an increase in duration. This is in accordance with existing literature that showed a lengthening of fricative durations (Tjaden & Turner, 2000) as well as individual vowel durations (Turner & Weismer, 1993) in ALS. Interestingly, significantly larger durations of movement was observed only in the severe group, characterized by a speaking rate of <120 wpm and impaired intelligibility. Significantly larger duration of movements was not observed at earlier stages of disease, even though speaking rate was decreasing. This suggests that other mechanisms, apart from an increase in movement durations, are contributing to a speaking rate reduction at early stages of disease. One such alternative may be that individuals with ALS decrease their speaking rate due to an increase in
pause durations and frequencies during speech production (Turner & Weismer, 1993; Green, Beukelman, & Ball, 2004). The calculation of speaking rates includes both articulation time and pause time (Turner & Weismer, 1993). However, during sentence repetitions, such as the speech task used in this study, pausing is not prominent (Rochester, 1973). Findings suggest that durations of movements was preserved at early stages of disease and only increased at a later stage of disease when intelligibility declined. This is to be expected as prior research observed that duration of movements is negatively correlated with intelligibility (Yunusova et al., 2012).

**Tongue and Jaw in Speech: Are Movements Compensatory?**

The differential degree of impairment among different structures of the speech system reported in the past (Lawyer & Netsky, 1953; Carpenter et al., 1978; Dworkin & Hartman, 1979; Dworkin, 1980; Dworkin, Aronson, & Mulder, 1980; DePaul et al., 1988; Langmore & Lehman, 1994) makes ALS an interesting condition to study for understanding the physiologic basis of speech impairment. Yet, the nature of differential impairment makes it difficult to determine the underpinnings of changes in speech movement characteristics, as they may reflect disease-related changes or compensatory responses (Hirose et al., 1982; Yunusova et al., 2010; Mefferd, Green, & Pattee, 2012). On one hand, the severity-dependent pattern of deterioration in tongue and jaw movements may occur due to the differential degree of impairment of articulators. For example, a delayed reduction in jaw maximum speed, compared to the tongue, may simply reflect a late onset of deterioration in the jaw. Alternatively, jaw movements may preserve size and maximum speed at early stages of disease as a compensatory response to tongue impairment.

Existing studies provide preliminary findings for both possible disease-related outcomes. Previous literature confirms a non-uniform rate of deterioration (DePaul & Brooks, 1993; DePaul & Abbs, 1987; Lawyer & Netsky, 1953) and shows that the tongue is affected earlier and to a greater extent compared to the jaw and lips. Differential impairment has been observed neuropathologically, reporting more degeneration of hypoglossal nerve fibers than trigeminal and facial nerve fibers (Lawyer & Netsky, 1953). The non-uniform rate of deterioration has also been observed in force generation studies with the tongue muscles exhibiting greater strength deficits than the jaw and the lips (DePaul & Abbs, 1987). However, kinematic speech data, although limited, suggest a compensatory outcome to differential impairment. Smaller and
slower than normal tongue movements have been reported (Kent, Netsell, & Bauer, 1975; Hirose, 1982; Kuruvilla, Green, Yunusova & Hanford, 2012; Yunusova et al., 2008; 2012). Meanwhile, a larger than normal jaw movement size has been observed in ALS, suggestive of a compensatory-type jaw response to the tongue impairment (Yunusova et al., 2008; Mefferd, Green, & Pattee, 2012). Kinematics studies in ALS are, however, in their infancy; tongue and jaw kinematics have always been investigated separately. Further insight into the distinction between disease-related outcomes requires that the tongue and jaw speech kinematics be investigated in the same group of talkers. The strength of this study is that it is the first to investigate movements of both articulators in a relatively large group of talkers. Findings suggest that the differential degree of impairment of the tongue and jaw may not reflect in changes of movement size, at least at a sentence level. However, an examination of tongue and jaw relationships at different stages of disease suggests compensatory interactions.

When examining the association between the tongue and jaw movement range, a significant moderate negative association was observed in the healthy talkers. This finding can be interpreted as indicating a reciprocal relationship between the two articulators in healthy speech, as observed in previous literature (Kuhnert, Ledl, Hoole, & Tillmann, 1991; Stone, 1995). Kuhnert and colleagues (1991) reported a lingual-mandibular pattern of coarticulation, particularly a vertical trade-off between the articulators. Similar observations of reciprocity between the tongue and jaw were observed in Dutch vowels by Stone (1995). In contrast, tongue and jaw movement range showed a significant positive correlation in the ALS1 group, consisting of individuals with intact intelligibility and rate. A positive relationship suggested that articulators were spatially coupled in contrast to the healthy talkers who showed reciprocity in the articulator interactions. However, there was no evidence of spatial coupling in ALS2 group composed of individuals showing a notable slowing in their speaking rate but still highly intelligible.

Traditionally, researchers have assumed that a compensatory interaction would render a negative correlation between articulators; if the tongue movement decreases in size, the jaw movement will increase in size as a compensatory response (Hirose et al., 1982; Mefferd, Green, & Pattee, 2012; Yunusova et al., 2008). However, this view assumes that all individuals compensate using the same strategy. ALS is a heterogeneous disease with varying symptom onsets and
presentations across individuals (Brooks, 1996). Thus, different patterns of deterioration among individuals might demand unique strategies for compensation, potentially involving structures that are not restricted to the articulatory subsystem. Variations in compensation strategies, therefore, would result in deviations from a normal tongue and jaw relationship, resulting in a lack of an association between tongue and jaw movements. Findings suggest compensatory strategies may be in effect at a stage in disease when speaking rate is impaired, but intelligibility is intact.

Slowing of the speaking rate is a hallmark characteristic of ALS (Duffy et al., 2005; Yorkston et al., 1993), however, the underlying mechanisms of slow rate have not yet been established. A decrease in speaking rate may, by itself, be compensatory in nature; patients in ALS2 may have been decreasing speaking rates in order to preserve intelligibility. Wieneke and colleagues (1987) suggested that by decreasing the rate of speech, individuals are able to reduce kinematic interactions, thereby making the task of speaking ‘easier’, yet successfully achieving their acoustic target. Consistent with this view is the finding that slow-rate movements appear to be less effortful than normal movements (Perkell et al., 1997). Further longitudinal work is required to tease out the compensatory versus physiological effect of disease on articulatory movements and speaking rate alike within individuals affected by ALS.

Role of Speech in the Assessment of Disease Related Changes

There is increasing research and clinical emphasis on finding measures to diagnose changes in bulbar function as early as possible. The search for sensitive measures of bulbar disease identification and progression has led researchers to the investigation of multiple non-speech measures, including static isometric maximum voluntary contract (MVC), and peak rate of change of force (PRCF; Brooks, 1996). These measures have shown to be sensitive in the assessment of bulbar physiology even in mild ALS (Depaul et al., 1987; 1988; 1993) and predictive of survival (Weikamp et al., 2012). However, these measures have a disadvantage with respect to the predicting of speech proficiency (Dworkin et al., 1980; Weismer & Forrest, 1992; Langmore & Lehman, 1994).
Speech measures, on the other hand, have been consistently related to changes in speech intelligibility and speaking rate in ALS (Yunusova et al., 2012; Weismer et al., 2000; Turner, Tjaden, & Weismer, 1995; Kent et al., 1997). Specifically, studies have identified strong linkages between speech movements, acoustics, and perception, with changes in kinematic measures reflecting changes in speaking rate and intelligibility (Yunusova et al., 2012). However, this study showed that speech measures might not be ideal for detecting group differences for diagnostic purposes. The quality of the speech acoustic signal is considered the ultimate goal in speech production and this goal can be achieved through various combinations of movements (Perkell et al., 1993; Kelso et al., 1984; Stone, 1995; Savariaux, Perrier, Orliguet, 1995; Maeda, 1991). Individuals with motor speech deficits can exhibit varying compensation strategies using various structures between and within speech subsystems while still preserving the target acoustic signal. As a result, a speech task, such as a sentence production as used in this study, may induce large variability between individuals, and may also be insensitive to speech musculature deficits that may be masked by compensation. However, speech tasks are necessary if the goal is to examine inter-articulatory interactions. Results from this study support this notion; analysis of a sentence production was able to identify significant associations between articulators even though the sample size within severity groups was relatively small. This suggests that a speech task, although relatively insensitive to muscular impairment, is sensitive and necessary to understand the principles of coordination among articulators during speaking. Thus, a combined use of speech and non-speech tasks may be more beneficial in understanding the effect of disease on the physiology and functional outcomes in the bulbar system.

UMN- LMN Speech Characteristics

A comparison of speech movement characteristics in ALS to other motor neuron diseases, such as a predominantly upper motor neuron (i.e. Primary Lateral Sclerosis (PLS)) or lower motor neuron (i.e. Progressive bulbar palsy (PBP) and progressive muscular atrophy (PMA)) disorder, would yield useful diagnostic information. The distinction between the effects of UMN degeneration and LMN degeneration on speech movements would contribute to early diagnosis,
as well as to tracking changes associated with disease progression. However, this comparison is a
difficult task, primarily because speech kinematic research is in its infancy. A single existing
study has compared the perceptual differences in speech samples (i.e. speech intelligibility) as
well as differences on various neurological dimensions (i.e. difficulty breathing, tongue atrophy)
among motor neuron diseases (MND) (Carrow, Rivera, Mauldin, & Shamblin, 1974). The study
found that neurological symptoms of tongue atrophy, observed in the ALS and PBP groups, were
most strongly related to decreases in intelligibility (Carrow, Rivera, Mauldin, & Shamblin,
1974). Because tongue atrophy is a symptom primarily associated with LMN degeneration,
findings suggest that LMN degeneration may be associated with a loss in speech intelligibility. A
better understanding of speech characteristics associated with UMN and LMN dysfunction
requires further investigation among the subgroups of MND.

Limitations
Small N in the group with impaired intelligibility

The subset of data in the present study came from a longitudinal study of subsystem decline in
ALS. Longitudinal research is extremely challenging in this clinical population and attrition rates
are extremely high (nearly 50% at each session; Blain et al., 2007; Hecht et al., 2002; Block et
al., 1998; Suhy et al., 2002). The majority of patients drop out before reaching a severe stage of
the disease due to difficulties with task completion (i.e. fatigue, respiratory complications, rapid
loss of speech function). Thus, our most affected group (ALS3) has a very small N. However, the
main focus of the study was to investigate early changes in speech, which have rarely been
examined. The strength of this study is that it characterizes interactions of tongue and jaw
movements at an extremely early stage of speech impairment, when individuals do not present
with speech intelligibility deficits.

Decoupling tongue from jaw movements

In this study, tongue movements were decoupled from the jaw using a linear subtraction method
(LSM), originally proposed by Westbury and colleagues (2002). A potential limitation of the
above method relates to the fact that jaw motion in speech involves both rotation and translation
Because of the rotational component, error might be introduced of up to 2 mm (Henriques & Lieshout, 2013). However, the LSM method was compared to a decoupling method that incorporates jaw rotation (Westbury, 1994) on the basis of repeated measures in 3 individuals and found that any error associated with the LMS method was insufficient to have a substantive effect on interpretation on the results for single sentence analysis (McClean, 2000).

To examine the potential effect of the residual error on the result of this study, tongue movements were compared to the tongue movements obtained via the decoupling method with the bite block (Gay, Lindblom and Lubker, 1981; Solomon & Munson, 2004) in the same group of participants. A 5-mm bite block inserted between the molars on the right side of the mouth stabilized the jaw during the sentences. The overall pattern of deterioration for tongue movements across ALS groups was the same for both methods. Significant group differences were not observed for tongue range or maximum speed in either of the two decoupling techniques. Furthermore, the two methods were highly correlated for range and maximum speed of movements, suggesting that participants exhibit similar patterns of tongue movements irrespective of method. The development of a more mathematically complex method of jaw subtraction is currently being conducted.

The need to examine trajectory characteristics

The study did not find group differences in movement size and speed; however, it would be premature to believe that tongue and jaw kinematics at a sentence level do not change with disease progression. Statistically significant group differences were not found, possibly due to the nature of the measures; 3 dimensional movement paths were reduced into 1 dimensional distance measures, from which range and maximum speed were extracted. These measures would be insensitive to any changes that are directionally specific. Changes in speech movements may involve more complex characteristics that require detailed by-axis analysis. For example, an observational study by Hirose and colleagues (1982) reported an “upward and forward” displacement of tongue movements in 2 individuals with ALS. In a subsequent analysis of vowel trajectories, Yunusova and colleagues (2008) found that talkers with ALS differed in their tongue trajectory shapes, however size of movements did not differ from healthy talkers.
Based on these preliminary findings, it might be fair to suggest that complex changes in trajectory paths could be one of the features associated with speech impairment. A longitudinal study on a larger group of individuals investigating trajectory characteristics is currently being conducted.

Clinical Implications

Findings from this study might be useful in staging speech characteristics of the disease. A stage in disease, when speaking rate is impaired but intelligibility is still intact, seems to be when compensatory interactions among the tongue and jaw are in effect. Findings may also aid in the development of speech intervention strategies that focus on jaw articulation, as results are suggestive of a compensatory role of the jaw in speech preservation. However, the large variability in speech kinematics between individuals demands need for better understanding of compensation strategies, both within the articulatory subsystem (i.e. tongue and jaw) and between subsystems (i.e. velopharyngeal, articulatory, respiratory). By gaining a better understanding of compensatory articulation as a function of disease, intervention techniques may shift focus from augmentative and alternative communication (AAC) techniques to the development of strategies that directly focus on speech preservation.

Conclusions

The current study investigated both the tongue and jaw movement characteristics in a group of participants with ALS, focusing on the early stages of speech impairment. Kinematic data were discussed in relation to system level measures of speech (i.e. speaking rate and intelligibility). Participants with intact intelligibility but mildly impaired speaking rate may have exhibited compensatory interactions between the tongue and jaw. The current study contributed to the extensive discussion in the literature on motor control on the kinds of tasks (i.e. speech, speech-like, or non-speech tasks) needed for the assessment of speech impairment. Findings suggested that while speech tasks may not be ideal to detect movement deterioration, speech measures are crucial if interactions between articulators are to be addressed. The compensatory role of the jaw
in speech preservation at an early stage of disease can be further explored to the development of intervention strategies that focus on articulatory compensation.

The findings of this study need to be expanded in the future to the investigation of individual differences in movement characteristics, as well as to shorter segments of speech (i.e. vowels) and speech-like tasks (i.e. AMR), to further understand compensatory articulation in ALS.
REFERENCES


