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Subgroups of Ataxic Dysarthria

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Abstract

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The role of the cerebellum in speech production is related to coordination and timing. When the cerebellar circuit is disrupted, the resulting speech disturbance is known as ataxic dysarthria. Features of this disruption include irregular articulatory breakdowns, distorted vowels, excess and equal stress, prolonged phonemes, and excess loudness variation. However, the manner in which these attributes present themselves is varied across the population. There is some suggestion in the literature that subgroups of ataxic dysarthria may exist, but these subgroups have never been developed or defined. The purpose of this pilot study was to determine a feasible, perceptual method for identifying and differentiating these proposed subgroups. To this end, a theoretical framework was created from existing literature which clustered speech symptoms according to notions of instability or inflexibility. Ten experienced speech-language pathologists listened to samples of speakers with ataxic dysarthria and completed rating forms of the speech features. Results suggested that five speakers fit the pattern of instability, one speaker aligned with inflexibility, and 4 speakers had a mixed presentation. The potential emergence of subgroups is promising and has implications for clinical and research practices. Additional research is warranted.

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Introduction

The exquisite design of the cerebellum challenges us to look beyond its relatively small size, about 10% of the volume of the brain. When we consider that more than half of all neurons of the brain reside in the cerebellum, or "little brain", we begin to get a glimpse into its complexity (Barlow, 2002). While our focus on the cerebellum often relates to its role in motor control and regulation, it should be noted that cerebellar networks also contribute to functions such as cognition, emotional regulation, auditory verbal imagery (Ackermann, Mathiak, & Riecker, 2007) and motor learning (Schulz, Dingwall, & Ludlow, 1999).

The cerebellum connects to the contralateral thalamus and cerebral hemispheres regulating movements on the ipsilateral side of the body (Barlow, 2002). The cerebellar anatomy consists of two primary components: the flocculonodular lobe and the body, which can be further divided into the vermis and lateral hemispheres, each of which is comprised of an anterior and a posterior lobe (Barlow, 2002; Duffy, 2013). Each of the peduncles (inferior, middle and superior) has either afferent, efferent or both kinds of fibers which communicate with different structures of the thalamus, pons and medulla (Duffy, 2013). Given the vast number of afferent fibers in the cerebellar circuit, about 40 afferent fibers for every efferent fiber, the cerebellum plays a critical role in processing sensory information in order to coordinate movement in a changing external environment (Duffy, 2013).

When the motor pathways of the cerebellum are disrupted, gait disturbances, nystagmus, tremor, and incoordination characterized by dysmetria, decomposition of movement and dysdiadochokinesia may result (Duffy, 2013). Typical etiologies for these disturbances are degenerative diseases (i.e. idiopathic cerebellar degeneration, spinocerebellar ataxia, multiple system atrophy, olivopontocerebellar atrophy, and Friedreich's ataxia); vascular disorders affecting the superior cerebellar artery (SCA), anterior-inferior cerebellar artery (AICA) and/or the posterior-inferior cerebellar artery (PICA); demyelinating disease (i.e., multiple sclerosis); and tumors (Duffy, 2013).

Ataxic Dysarthria

The role of the cerebellum in speech production is related to coordination and timing, including the regulation of prosody through the management of rate and tempo (Duffy, 2013). When the cerebellar circuit is disrupted, the resulting disturbances to speech are referred to as ataxic dysarthria.

Typical presentation includes irregular articulatory breakdowns, distorted vowels, excess and equal stress, prolonged phonemes, and excess loudness variation (Duffy, 2013; Kluin, et al., 1988). Other associated symptoms may include harsh voice quality, prolonged intervals, monopitch, monoloudness, slow rate, and voice tremor (Duffy, 2013), as well as transient breathy voice, transient altered nasality and audible inspirations (Folker, et al., 2012; Kent, Kent, Rosenbek, Vorperian, & Weismer, 1997). The characteristic of reduced rate is perhaps more precisely described as variable rather than simply slow; excessively slow rate is often more characteristic of spastic dysarthria, a frequent co-occurring disorder (Schalling, Hammarberg, & Hartelius, 2008). The perceptual characteristics of these features are typically assessed with perceptual measures, such as sequential and alternating motion rates (SMR/AMRs) and observation of speech rate, prosody and articulation during spontaneous speech. Acoustic analyses may also be employed, and include measurements of fundamental frequency (F0) and amplitude (Ackermann & Hertrich, 1994; Duffy, 2013; Schalling, Hammarberg, & Hartelius, 2008). Results of these assessments often reveal variable imprecision of consonants, a reduced and variable speech rate, reduced and variable AMR and SMRs, longer and variable syllable and pause durations, alternating loudness, fluctuating pitch, inconsistent nasality, and vocal instability, consistent with a 3 Hz tremor (Ackermann & Hertrich, 1994; Boutsen, Bakker, & Duffy, 1997; Kluin, et al., 1988; Schalling, Hammarberg, & Hartelius, 2008).

Originally identified by Darley, Aronson and Brown (1969), these speech attributes were grouped into clusters of *articulatory inaccuracy*, *prosodic excess* and *phonatory-prosodic insufficiency*. The clusters were defined using perceptual parameters and were the benchmark for diagnosing ataxic dysarthria (Brown, Darley, & Aronson, 1970). These clusters were intended to help characterize ataxic dysarthria and to distinguish it from other motor speech disorders. However, the varied presentation of ataxic dysarthria leads to challenges for differential diagnosis. Moreover, the heterogeneous presentation of individuals classified as having ataxic dysarthria brings into question whether it is a single entity. A better understanding of the speech profiles of this diverse population would facilitate differential diagnosis, advance the understanding and treatment of ataxic dysarthria, and identify disease progression as it pertains to speech changes (Ackermann & Hertrich, 1994; Boutsen, Duffy, Dimassi, & Christman, 2011; Portnoy & Aronson, 1982).

At this time, there are no original studies exploring the perceptual components of these purported subgroups. It is thus the purpose of the current study to identify the theoretical and

experimental evidence for subgroups of ataxic dysarthria, and to determine a feasible, perceptual method for differentiating these potential subgroups. To this end, a brief review of cerebellar lesion studies will first be provided, followed by a discussion of a theoretical framework that may help to explain divergence in the presentation of ataxic dysarthria.

Lesion loci and corresponding speech disturbances

Some studies have suggested that lesions to different areas of the cerebellum are responsible for the varied speech production seen in people with ataxic dysarthria. For example, Ackermann and colleagues (1992) purport that the presence of dysarthria is dependent on lesions to the left cerebellar hemisphere and the paravermal regions, suggesting a laterality component to the cerebellum's role in speech motor control and overall speech rate modulation (Boutsen & Christman, 2002; Vandana, 2007). Ackermann et al. (2007) identified ischemic infarctions of the SCA and/or PICA in particular as being associated with ataxic dysarthria. Further, bilateral lesions often resulted in more severe deficits of articulation; in addition, when there was involvement of the dentate nucleus, both phonatory and articulatory deficits resulted (Ackermann, Vogel, Petersen, & Poremba, 1992). A review by Spencer and Slocomb (2007) noted the role of the right cerebellar hemisphere as crucial to articulatory planning. Boutsen and Christman (2002) suggest this hemisphere modulates syllabic durations (Vandana, 2007). Moreover, right paravermal superior regions of the cerebellum were linked to ongoing control of the articulators (Bohland & Guenther, 2006; Vandana, 2007) and to adaptive timing mechanisms (Guenther, Ghosh, & Tourville, 2006) whereas left superior paravermal regions are associated with phonatory control (Vandana, 2007).

Though considered the most salient feature of ataxic dysarthria, excess and equal stress characteristics have not been linked to a specific locus in the cerebellar system (Ackermann, Vogel, Petersen, & Poremba, 1992). However, it has been suggested that this feature of "scanning" speech may be more related to the severity of the dysarthria, with emergence of this feature more commonly seen in moderate to severe forms of the dysarthria (Ackermann, Vogel, Petersen, & Poremba, 1992; Ackermann & Hertrich, 1994; Ackermann, Mathiak, & Riecker, 2007; Kent, Netsell, & Abbs, 1979).

A theoretical framework

One theory has emerged which suggests that features of ataxic dysarthria may be linked to patterns of *instability* or to patterns of *inflexibility* (Duffy, 2013). Duffy (2013) proposed these two

groups based on distinctive features documented in the research literature and in clinical practice. The principle of instability would be associated with unusually increased variability. For example, some individuals with ataxic dysarthria may have bursts of loudness and an overall inability to regulate breathing with speaking. In contrast, the principle of inflexibility would be associated with speech symptoms that are abnormally invariant and equalized. One example might be scanning speech, where equal emphasis is given to each syllable as it is produced. Understanding which characteristics are reflective of instability versus inflexibility may allow us to build a clinically applicable basis for subtypes.

Instability

Research in support of instability in ataxic dysarthria has targeted variation of rate, articulation, intensity, pitch and respiration. Most of the investigations have focused on acoustic analyses, but provide support for the perceptual correlates, as described below.

Inconsistent rate in ataxic dysarthria is a distinguishing characteristic with variation found in duration of syllables, vowels and consonants, as well as pause duration and voice onset time (VOT) (Duffy, 2013). Acoustic analyses have revealed inconsistent reductions in syllable duration, as well as varied length of syllables (Kent, Netsell, & Abbs, 1979). Further, acoustic analyses have supported intermittent occurrences of longer VOT and varied longer formant transitions (Duffy, 2013; Kent, Netsell, & Abbs, 1979), though findings differ depending on length of the target word (Kent et al., 1997; Schulz, Dingwall, & Ludlow, 1999). These acoustic measurements correlate perceptually to variable and inconsistent rate of speech (Duffy, 2013; Hartelius, Runmarker, Andersen, & Nord, 2000; Kent, et al., 2000; Kent, Netsell, & Abbs, 1979). This variation is most pronounced in maximum performance tasks, such as the production of syllables in a speech AMR/SMR task (Duffy, 2013). Further, it is this variable rate that helps to distinguish ataxic dysarthria from spastic dysarthria, which presents as more marked and consistent slowed rate (Portnoy & Aronson, 1982).

Articulatory variability is thought to be one of the essential features of ataxic dysarthria (Hartelius, Runmarker, Andersen, & Nord, 2000). This feature may be attributed to incoordination resulting in slow, inaccurate, and poorly timed articulatory movements (Duffy, 2013; Netsell & Kent, 1976) . Acoustic analyses by Ackermann and Hertrich (1994) suggest that the variable rate, discussed above, affects the ability of an individual to achieve an articulatory gesture, possibly attributed to a delay in initiation. Speakers may also undershoot or overshoot an articulatory target (Ackermann,

Vogel, Petersen, & Poremba, 1992; Netsell & Kent, 1976). Perceptually, this results in the same articulatory target, i.e., phoneme, being produced adequately in one instance and distorted, or possibly omitted, in another (Ackermann, Vogel, Petersen, & Poremba, 1992). However, the amount of articulatory variation observed is highly dependent on the task (Boutsen, Bakker, & Duffy, 1997; Duffy, 2013). For example, there is greater articulatory variation when comparing syllable repetition to connected speech (Ackermann & Hertrich, 1994; Boutsen, Bakker, & Duffy, 1997). Variation is most easily observed, and often clinically measured, in speech AMR/SMR tasks, where speed and precision is taxed (Ackermann & Hertrich, 1994; Duffy, 2013).

<u>Prosodic disruption</u> may exist in part from difficulties with modulating articulation and rate (Hartelius, Runmarker, Andersen, & Nord, 2000; Odell, McNeil, Rosenbek, & Hunter, 1991). Perceptually, prosody is primarily characterized by variations in intensity, pitch, stress, and rhythm. Features of dysprosody that are consistent with an "instability" mechanism would include abrupt and/or jerky control of intensity, a resulting variable fundamental frequency, poorly controlled stress, and dysrhythmia (Boutsen, Bakker, & Duffy, 1997; Boutsen, Duffy, Dimassi, & Christman, 2011; Hartelius, Runmarker, Andersen, & Nord, 2000; Ackermann, Mathiak, & Riecker, 2007). With respect to intensity, acoustic studies support excess loudness variation as a deviant characteristic suggesting incoordination of the subsystems of respiration and phonation (Duffy, 2013; Kent & Netsell, 1975; Kluin, et al., 1988; Murdoch & Theodoros, 1998). Alternating intensity behaviors may also stem from hypotonia, often seen with bilateral cerebellar damage and disruption to brainstem projections (Kluin, et al., 1988; Vandana & Manjul, 2006). Regarding variable fundamental frequency, descriptions are common of exaggerated and/or uncontrolled pitch excursions, which contribute to the labels of "drunken" and "slurred" speech (Netsell & Kent, 1976; Schalling, Hammarberg, & Hartelius, 2008). Further, the speaker may have difficulty voluntarily changing pitch (Lowit & Kuschmann, 2012). Research suggests that instability is also evidenced by difficulty signaling stress (Lowit, Kuschmann, MacLeod, Schaeffler, & Mennen, 2010), which is a salient perceptual feature of ataxic dysarthria (Odell, McNeil, Rosenbek, & Hunter, 1991). Uncontrolled stress can result in incorrect stress placement, in both words and sentences, as well as unnecessary pause insertions, and problematic durational control of the stressed syllable (Lowit, Kuschmann, MacLeod, Schaeffler, & Mennen, 2010). Irregular rhythm, or timing, is perhaps most evident during speech AMRs and SMRs, and has been supported with acoustic measures quantifying this element (Portnoy & Aronson, 1982). In connected speech tasks, dysrhythmia is often

measured by examining vowel length, which has been shown to be variable and shortened in speakers with ataxic dysarthria (Lowit, et al., 2010).

Respiratory incoordination has been characterized by irregularities in chest wall movement, visible during tasks of sustained phonation and syllable repetition, as well as incoordinated inspirations and expirations at syntactically inappropriate places (Duffy, 2013; Kent R. D., et al., 2000; Yorkston, Beukelman, Strand, & Hakel, 2010). Incoordination may yield air wastage and is often manifested by releasing the air supply prior to initiating an utterance (Yorkston, et al., 2010). Respiratory incoordination impairments may manifest early in cerebellar pathology and remain fairly static (Schalling, Hammarberg, & Hartelius, 2008). Paradoxical breath patterning has also been described for individuals with Freidreich's ataxia, but the unstable breath pattern was task-dependent (Folker, et al., 2012).

Thus, for the purposes of this study, *instability* will be defined as varied and inconsisent rate, articulation and prosody (i.e., intensity, pitch, stress, and rhythm) with respiratory incoordination. With this in mind, the manner in which these elements vary will define the subgroup of instability. Given the findings of previous research, the expectation is that when articulation is variable, there will be greater prosodic variation with co-occuring variations in intensity, pitch, stress and rhythm (Boutsen, Bakker, & Duffy, 1997; Hartelius, Runmarker, Andersen, & Nord, 2000; Duffy, 2013).

Inflexibility

The converse to Duffy's (2013) notion of instability is the principle of inflexibility. Speech elements that fit with the concept of inflexibility are characterized as less variable, with more equalized and isochronous production, e.g., scanning speech (Duffy, 2013; Hartelius, Runmarker, Andersen, & Nord, 2000). Research suggests reduced variability for some speakers with ataxic dysarthria in the following domains: rate, prosody and articulation. Also noted are consistently impaired respiration and phonation in terms of reduced breath support and harsh voice quality (Murdoch & Theodoros, 1998).

In some individuals with ataxic dysarthria, reduced rate is characterized by less variation in syllable duration and spacing between the syllables (Ackermann, Mathiak, & Riecker, 2007; Boutsen, Bakker, & Duffy, 1997; Duffy, 2013). There is an equalization pattern across the syllables such that syllables are lengthened and acoustically a longer VOT is measured (Kent, Netsell, & Abbs, 1979). Kent and colleagues (1997) describe this equalization pattern as inserting pauses between words as well as

syllables. Consequently, there is an inability to increase rate (Schulz, Dingwall, & Ludlow, 1999). It is unknown at this time if the reduced rate partially stems from a compensatory strategy to increase intelligibility or is solely a result of temporal dysregulation from cerebellar disease (Ackermann, Mathiak, & Riecker, 2007; Duffy, 2013; Folker, et al., 2012; Hartelius, et al., 2000; Kent R. D., et al., 1997; Kent, et al., 2000; Yorkston & Beukelman, 1981). Nevertheless, the reduction in rate is a salient, perceptual feature that it is often used in diagnosis of ataxic dysarthria (Folker, et al., 2012).

Dysprosody in the form of less prosodic variation has also been noted in some speakers with ataxic dysarthria (Duffy, 2013; Hartelius, et al., 2000). This lack of variation is evidenced in dimensions of intensity, pitch and stress. Specifically, a lack of intensity variation has been noted (Duffy, 2013). Acoustic studies support this perception of monoloudness by demonstrating reduced intensity fluctations in sustained phonation tasks (Boutsen, Duffy, Dimassi, & Christman, 2011). Simultaneous reduced pitch excursions are also evidenced (Duffy, 2013; Kent & Netsell, 1975; Murdoch & Theodoros, 1998). Typical pitch fluctations are diminished in this group as demonstrated by acoustic measures showing less fluctuation and flat F0 contours, often described as monotony (Ackermann & Hertrich, 1994; Hartelius, et al., 2000; Kent, Netsell, & Abbs, 1979; Paslawski, Duffy, & Vernino, 2005). In addition, a reduction in the duration of stressed syllables such that they are matched to unstressed syllables create an equalized, metered pattern (Hartelius, et al., 2000; Ziegler & Wessel, 1996). Therefore, isochrony results, because the individual is unable to adequately place stress on syllables (Hartelius, et al., 2000) indicating a tendency toward uniform syllable durations. Kent and colleagues (1979) ascribe this finding to the reciprocal lengthening of unstressed vowels, lax vowels and consonant clusters. The rhythm that results is syllable timed and metered prosody, sometimes called “scanning” speech, which is considered a prominent feature in some speakers with ataxic dysartria. As such, it can be effective in differential diagnosis from other dysarthrias, such as spastic dysarthria, which often co-occurs (Henrich, Lowit, Schalling, & Mennen, 2006; Liss, et al., 2009; Portnoy & Aronson, 1982). This prosodic pattern creates rhythm abnormalities particularly on linguistically significant syllables (Liss, et al., 2009; Yorkston & Beukelman, 1981). Further, impairments of respiration may contribute to this syllable by syllable rhythmic pattern from “pulsed” alveolar air pressure release possibly employed as a compensatory strategy (Yorkston, et al., 2010, page 153). These combined elements of monoloudness and monopitch may be attributed to overall hypotonia which limits muscle contraction and range of motion (Duffy, 2013; Kent and Netsell, 1975; Murdoch & Theodoros, 1998).

Relatively consistent articulatory imprecision has been described for some speakers with ataxic dysarthria (Ziegler & Wessel, 1996) and documented acoustically (Ackermann & Hertrich, 1994; Duffy, 2013; Ziegler & Wessel, 1996). Notably, vowel distortions tend to be more prominent than errored consonantal targets (Odell, McNeil, Rosenbek, & Hunter, 1991). Consonantal errors often result from undershoot or overshoot, leading to imprecision (Ackermann, Vogel, Petersen, & Poremba, 1992; Netsell & Kent, 1976; Odell, McNeil, Rosenbek, & Hunter, 1991; Paslawski, Duffy, & Vernino, 2005). A proposed explanation for this errored pattern is the impact of hypotonicity limiting tongue and lip movements such that an articulatory target is not achieved (Ackermann & Hertrich, 1994; Duffy, 2013; Kent, Netsell, & Abbs, 1979; Murdoch & Theodoros, 1998). Further, a reduction in the movement so it is slower and smaller constrains the extent and speed, yielding imprecision and distortion (Schulz, Dingwall, & Ludlow, 1999).

Ultimately, what *inflexibility* refers to is the tendency of speakers with ataxic dysarthria to speak with less variation resulting in a staccato speech rhythm (Ackermann & Hertrich, 1994; Duffy, 2013; Hartelius, Runmarker, Andersen, & Nord, 2000; Kent, Netsell, & Abbs, 1979; Murdoch & Theodoros, 1998). Moreoever, a reduction in rate is expected and restrictions exist on prosody, such that it is monoloud, monopitch and isochronous. Articulation is consistently errored and marked by distorted vowels (Duffy, 2013; Hartelius, Runmarker, Andersen, & Nord, 2000; Kent, Netsell, & Abbs, 1979; Ziegler & Wessel, 1996). Many of these characteristics are possibly linked to hypotonicity. Table 1 summarizes the salient features associated with the hypothesized frameworks of inflexibity and instability as they pertain to ataxic dysarthria.

Combined inflexibility and instability

It is possible that individuals will share features across the two subgroups of inflexibility and instability. Indeed, the combination of subgroups may be most common (Duffy, 2013, personal communication). While there is no literature to directly substantiate this notion, indirect support stems from the clinical presentation of some individuals with ataxic dysarthria. That is, elements of instability, such as poorly modulated prosody, may intermingle with the attributes of inflexibility, such as a relatively consistent disruption to articulation. Given the heterogeneity already discussed, as well as the many etiologies which lead to ataxic dysarthria, this proposed third group may explain the contradictory perceptual characteristics which seem to co-occur in some individuals (Duffy, 2013).

Table 1. Summary of salient speech features that are consistent with patterns of instability versus inflexibility

	Instability	**Inflexibility**
Respiration	• Incoordination of breathing with speaking • Paradoxical breath patterning	• Consistently reduced breath support
Phonation	(see pitch description under Prosody)	
Articulation	• Variable; possible undershoot and overshoot	• Relatively consistent articulatory imprecision
Resonance	• Intermittent instances of hypernasality and hyponasality	
Prosody	• Inconsistent rate with variable syllable length • Uncontrolled loudness variation • Variable pitch • Variable stress patterns • Variable rhythm (especially in AMRs/SMRs)	• Decreased rate with equal pattern across syllables • Monoloudness • Monopitch • Isochronous, metered stress patterns • Steady rhythm (especially in AMRs/SMRs)

Note. AMRs = alternating motion rates, SMRs = sequential motion rates (Duffy, 2013).

Why does variation exist?

Unknown at this time is why there is variation in individuals with cerebellar impairment regardless of whether the person is in the acute or chronic stage of the disease (Ackermann, Mathiak, & Riecker, 2007; Kent, Netsell, & Abbs, 1979; Duffy, 2013). Is the variation simply due to a difference in lesion loci? Given the array of possible lesion sites with potential impact on speech in the cerebellar circuit, this may seem an obvious conclusion. However, as previously discussed, lesions to the same area of the cerebellum can result in different impairments (Spencer & Slocomb, 2007). At this time, there is a lack of systematic information conclusively linking lesion site to dysarthria. Moreover, lesion location may not be the only variable impacting the diversity of characteristics observed. Ackermann and Hertrich (1994) suggest that some variation in symptoms may be attributed to the overall severity of the dysarthria. It has also been suggested that variable presentation of the dysarthria tends to increase with progressive disorders suggesting inflexibility may precede instability if a severity continuum is employed for these potential subgroups (Kent, Netsell, & Abbs, 1979; Schalling, Hammarberg, & Hartelius, 2008). In direct contrast, other researchers have suggested scanning speech, from the inflexibility framework, reflects a more severe presentation of ataxic dysarthria (Ackermann &

Hertrich, 1994; Ackermann, Vogel, Petersen, & Poremba, 1992). However, these trends are not applicable to all etiologies as disease progression is variable across cases (Schalling, Hammarberg, & Hartelius, 2008). Therefore, even individuals with the same etiology may manifest different impairments. For example, individuals with spinocerebellar ataxia may show greater disease progression in the brainstem before the cerebellum or vice versa (Schalling, Hammarberg, & Hartelius, 2008). Thus, symptomology would reflect which areas of the cerebellar network are most impacted.

Tasks measuring features of ataxic dysarthria

Studies examining features of ataxic dysarthria consistently use tasks of diadochokinesis, sustained phonation and connected speech. Often these tasks were measured with both acoustic and perceptual analyses. What follows is a discussion of the salient perceptual features associated with these standard assessment tasks.

Tasks of diadochokinesis are frequently used and often take the form of alternating motion rates (AMRs, e.g., “pa-pa-pa”) and sequential motion rates (SMRs, e.g., “pa-ta-ka”). These tasks are typically performed at maximum rates (Ackermann, et al., 1992; Duffy, 2013). Diadochokinetics can reveal articulatory imprecision and variation, including control of voicing, as well as rate and rhythm disturbances (Ackermann, et al., 1992; Duffy, 2013).

Tasks of sustained phonation typically reveal inabilities to sustain loudness and a steady pitch (Boutsen, et al., 2011; Duffy, 2013). Measures of vocal quality and stability, as well as inferences of respiratory support, may also be attained (Ackermann & Hertrich, 1994; Yorkston et al., 2010). These tasks may additionally include pitch and loudness glides in order to measure the speaker’s ability to make volitional fluctuations (Lowit, et al., 2010).

While discrete speaking tasks are helpful for targeting specific aspects of speech, the coordination across speech subsystems is best seen in connected speech tasks, often through a conversation, a picture description, or a story retell (Ackermann, et al., 1992; Duffy, 2013; Hartelius, et al., 2000; Kent, et al., 1997; Lowit & Kuschmann, 2012). Connected speech samples can provide a wealth of information, including intonation and stress patterns (Lowit & Kuschmann, 2012), paradoxical and/or incoordinated breathing patterns (Duffy, 2013; Folker, et al., 2012; Yorkston K. M., et al., 2010), patterns of articulatory imprecision (Ackermann & Hertrich, 1994; Duffy, 2013; Kent, et al., 1979;), changes to rate (Ackermann, et al., 2007; Boutsen, et al., 1997; Duffy, 2013) and modulation of various

aspects of speech, such as loudness, pitch and resonance (Duffy, 2013; Hartelius, et al., 2000; Kent & Netsell, 1975; Kent, et al., 1979; Murdoch & Theodoros, 1998). Sentence and passage reading have also been employed to measure rhythm and intonation patterns as a highly structured task (Cummins, Lowit, & van Brenk, 2013; Folker, et al., 2012; Liss, et al., 2009; Netsell & Kent, 1976; Schalling, et al., 2008).

Using the three tasks of diadochokinetics, sustained phonation and connected speech, a sketch of perceptual features can be constructed based on the theoretical model of instability and inflexibility. What follows in Table 2 are the expected perceptual qualities which should be most evident from each task given the previously described acoustic and perceptual features documented from the literature for each putative subgroup.

Table 2. Proposed perceptual characteristics for the theoretical subgroups of instability and inflexibility across speech tasks.

Task	Instability	Inflexibility
Diadochokinesis	• Variable articulation/voicing • Inconsistent rate • Variable rhythm	• Relatively consistent articulatory imprecision • Decreased rate • Equalized pattern across syllables
Sustained Phonation	• Variable loudness • Variable pitch • Incoordinated respiratory control	• Reduced respiratory support
Connected Speech	• Variable articulation/voicing • Variable rate • Variable loudness • Variable pitch • Variable stress patterns • Variable rhythm • Incoordinated respiration/ paradoxical breath patterning	• Shorter utterances possibly with frequent inhalations • Speaking on residual air • Relatively consistent articulatory imprecision • Decreased rate • Monopitch/monoloudness • Isochrony - metered stress pattern, steady rhythm • Equalized syllables

The “gold standard” of clinical diagnosis for motor speech disorders is using the auditory perceptual characteristics of speech and recognizing predictable patterns of speech disturbances (Duffy, 2013). Yet, the heterogeneity of symptoms related to ataxic dysarthria leads clinicians and researchers to combine speakers with very different speech profiles. Therefore, delineating patterns or subgroups

within the larger group of individuals with ataxic dysarthria may lead to more precise clinical diagnoses and improved subject selection for research. Further, the identification of subgroups might inform theoretical and clinical models of cerebellar functioning and disease. It is thus the purpose of the current study to determine a feasible, perceptual method for differentiating these putative subgroups. Specifically, we tested the hypothesis that patterns of speech characteristics will emerge that align with the proposed symptoms of inflexibility and/or instability.

Methods

Participants

Listeners

For this study, 10 experienced speech language pathologists with more than 5 years of clinical experience (beyond the clinical fellowship) with speakers with dysarthria were selected (Bunton, et al., 2007). It has been suggested that experienced listeners tend to show higher intra- and inter-rater reliability than inexperienced listeners (Zeplin & Kent, 1996). All listeners were native English speakers with no self-reported history of speech or language difficulties. Clinicians practiced in a variety of settings, with highest frequency in acute care, inpatient rehabilitation and long-term care settings; home care and academic settings were also represented in the clinical experience. Clinicians had an average of 16.3 years of experience, with a range of 5 – 40 years, with patients with dysarthria. Listeners consisted of 3 males and 7 females. All listeners passed a hearing screening at 35 dB for frequencies 1000, 2000 and 4000 Hz (American Speech-Language-Hearing Association, 1997), and a vision screening by correctly reading the 20/30 line of a Snellen chart while sitting 2.3 feet away; vision correction, i.e., glasses, were permitted.

Speaker Samples

For this feasibility study, audio files were compiled from Aronson (1993), Duffy (2013), Freed (2000) and Spencer (2004) patient samples used in educational programs as exemplary presentations of ataxic dysarthria. Information on the gender and etiology of each sample is provided in Table 3. Only selected portions of the samples were presented according to the following criteria:

(1) **Sustained phonation:** Samples of sustained phonation were provided if available, which was the case for 8/10 speaker samples. Samples ranged from 8-21 seconds.

(2) AMRs/SMRs: AMRs and/or SMRs were available for all speakers. AMRs only were available for 3/10 speakers, while both AMRs and SMRs were available for 7/10 speakers. Samples ranged from 17-79 seconds. Longer samples included instruction from the investigator on the elicitation of the AMR and/or SMR target. The average of the samples was 34.1 seconds.

(3) **Connected speech:** Samples of connected speech were available for all speakers. In the absence of conversational speech, a reading sample was used, which was the case for only 1/10 speakers. The goal was to provide a sample of 30 seconds of uninterrupted speech. For the available samples of conversational speech, the longest uninterrupted portion was identified and the sampling began 7-10 seconds into the running speech (at a linguistically appropriate location) to maintain sampling objectivity across subjects. In one instance (speaker 4), in order approximate the 30-second goal across speakers, a portion of the reading sample was added to a short conversational clip. Finally, given these were prerecorded samples, some samples consisted of a dyad between the speaker and a clinician. In such cases, the running speech sample was longer in order to assure a 30-second speech sample was pieced together, as was the case for speakers 2, 9 and 10. Final samples ranged from 23-46 seconds, with an average of 32.6 seconds.

Table 3 outlines the combination of clips that were presented to the listeners for each speaker. Note samples 9 and 10 are considerably longer due to the length of time for AMRs which included instruction and modeling of the task by the clinician. In sample 9, the clinician requested that the speaker continue AMRs/SMRs through the duration of a single breath making the sample longer than others. Overall, speaker samples ranged from 52 – 140 seconds, with an average of 76.5 seconds.

Task

The rating sheet provided to participants is reflected in Table 4. Features and corresponding options were derived from the literature and are the basis for the rating sheet. For purposes of subgroup identification, characteristics thought to represent the notion of instability are indicated by red font, while symptoms hypothesized to occur with inflexibility are indicated by green font. These color codes were not provided to the listeners.

Each of the listeners was seated at a desk and presented with audio speech samples over computer speakers (Realtek High Definition Audio version 6.0.1.5931), with each sample adjusted to a comfortable listening level. The investigator was seated next to the listener and manipulated the files; the computer screen was angled toward the investigator and away from the listener. Therefore, the listener had no visual information for the speaker. Videos housed on the Duffy (2012) website were setup prior to the listener's arrival. Each speaker and corresponding task was preset, so the listeners were presented with the same audio samples. If the listener required repetition, the investigator used headphones to isolate the same auditory sample, removed the headphones, and played the sample aloud over the computer speakers for the listener. Samples on compact disc, Aronson (1993), Freed (2000), Spencer (2004) were set based on the counter available through Windows 7 Media Player. For repetitions, the counter was always returned to the same position. For each sample, the listener was oriented to the task on the rating sheet and asked to select speech characteristics which best described the feature they were observing (see Table 4). For example, the listener was directed to "Sustained Phonation" and instructed only to rate loudness and pitch as indicated by the sustained phonation task. Listeners were allowed a maximum of five repetitions of a sample.

Speaker samples, as well as speaking tasks, were presented to listeners in a randomized order to avoid bias in subject rating. Two samples per listener were repeated as an indicator of intra-rater reliability. The repetitions were also randomized and always presented after the initial 10 samples. A summary of sample presentations may be found in Appendix B.

Table 3. Summary of samples of speakers with ataxic dysarthria presented for the listeners.

Sample	Source	Sex	Etiology	Sustained phonation	AMRs/ SMRs	Reading	Running speech	Total time of sample
1	Duffy Slide 136	Male	Cerebellar tumor	8 sec	AMRs- 24 sec SMRs- 6 sec		32 sec	70 sec
2	Duffy Slide 171	Female	Cerebellar brainstem ataxia, possibly autoimmune	9 sec	AMRs – 21 sec SMRs – 6 sec		36 sec	72 sec
3	Duffy Slide 159	Female	Cerebellum/cerebellar control circuit, cerebellitis of uncertain etiology		AMRs – 25 sec		30 sec	55 sec
4	Duffy Slide 128	Female	Moderate cerebellar atrophy, etiology undetermined		AMRs – 29 sec	10 sec	13 sec	52 sec
5	Duffy Slide 140	Female	Cerebellar ataxia, not otherwise specified	9 sec	AMRs – 22 sec SMRs – 7 sec		24 sec	62 sec
6	Duffy Slide 149	Male	Vertebrobasilar strokes (cerebellum and pons)	21 sec	AMRs – 15 sec SMRs – 4 sec		34 sec	74 sec
7	Duffy Slide 152	Male	TBI and cerebral fat emboli	17 sec	AMRs – 15 sec SMRs – 4 sec		36 sec	72 sec
8	Aronson 16	Female	Cerebellar degenerative disease	12 sec	AMRs – 26 sec	23 sec		61 sec
9	Freed	Male	Cerebellar degeneration	15 sec	AMRs- 67 sec SMRs- 12 sec		46 sec	140 sec
10	Spencer (Dys-04-6: ddk/phon) (Dys-04-5: conn sp)	Female	Friedreich's Ataxia	7 sec	AMRs- 41 sec SMRs- 17 sec		42 sec	107 sec

Table 4. Rating sheet for perceptual features according to task with color coding to reflect hypothetical subgroups of Instability (red) and Inflexibility (green).

Listener #		Sample #		
DDKs				
Articulation	☐ Variable errors	☐ Consistent errors		☐ Normal
Voicing	☐ Variable errors	☐ Consistent errors		☐ Normal
Rate	☐ Variable	☐ Consistently slow		☐ Normal
Rhythm	☐ Variable			☐ Normal
Sustained Phonation				
Loudness	☐ Variable	☐ Consistently too loud	☐ Consistently too quiet	☐ Normal
Pitch	☐ Variable			☐ Normal
Connected Speech				
Articulation	☐ Variable errors	☐ Consistent errors		☐ Normal
Stress	☐ Irregular placement	☐ Equalized across syllables		☐ Normal
Rate	☐ Variable	☐ Consistently slow		☐ Normal
Pitch	☐ Variable	☐ Monopitch		☐ Normal
Respiration	☐ Incoordination of breath with speech	☐ Speech on residual air	☐ Short phrases	☐ Normal
Loudness	☐ Uncontrolled	☐ Consistently too loud	☐ Consistently too quiet	☐ Normal

Note: Color coding will be removed for listener task.

Analyses

Parametric and non-parametric statistics were not an option for this pilot/feasibility study, given the small sample size, the manner in which the listeners rated the features, and the inconsistent number of options available to rate for each feature. Instead, descriptive statistics, including means and 95% confidence intervals were employed.

Reliability

Point-to-point agreement was chosen as the metric for intra-rater reliability. Traditional analyses of reliability (e.g., intra-class correlation coefficients) were not possible given the categorical nature of the data and the uneven distribution of variables across features and speaking tasks. Point-to-point reliability is a conservative approach and was calculated by matching each rated variable from time 1 and time 2. When a listener rated a given feature identically from time 1 to time 2, one point was given. Number of points was divided by number of ratings given, and a percentage was derived. Table 6 summarizes the point-to-point agreement. Reliability averages ranged from 58.4% to 82.3%. Intra-rater agreement did not appear to be influenced by subgroup profile (identification of the repeated samples can be found in Appendix B).

Table 5. Point-to-point intra-rater reliability.

	Repeated sample 1	Repeated sample 2	Average Reliability
Listener 1	10/13 (76.9%)	10/13 (76.9%)	76.9%
Listener 2	8/10 (80%)	8/12 (66.7%)	72.7%
Listener 3	9/12 (75%)	7/12 (58.3%)	66.7%
Listener 4	8/10 (80%)	6/12 (50%)	65%
Listener 5	9/12 (75%)	5/12 (41.7%)	58.4%
Listener 6	9/13 (69.2%)	7/13 (53.8%)	61.5%
Listener 7	7/11 (63.6%)	11/13 (84.6%)	74.1%
Listener 8	8/10 (80%)	11/13 (84.6%)	82.3%
Listener 9	9/12 (75%)	8/12 (66.7%)	70.9%
Listener 10	8/12 (66.7%)	6/12 (50%)	58.4%

Results

Average ratings across the ten speech-language pathologists can be found in Table 5. Only AMRs/SMRs and connected speech conditions were considered; sustained phonation was removed from the analyses for two reasons. First, the sustained phonation task was not available for all 10 samples. Second, the options on the rating sheet may have been too restrictive, as further elaborated in the Discussion.

For each speaking condition, the maximum number of items that align with a potential subgroup are provided in the shaded rows. For example, for the connected speech condition, there were 7 possible choices on the rating form (see Table 4) that were considered to be reflective of

“variability”, 6 possible choices that were associated with “consistency”, and 6 options to rate a speech feature as “normal”. The means in Table 5 reflect the average ratings across listeners for each speaker and the 95% confidence intervals. Individual data per speaker sample and listener, and calculations of descriptive statistics, can be found in Appendix C. Patterns begin to emerge when comparing means across the categories of variability and consistency. For example, speaker sample 1 has substantially higher means in both speaking conditions for the variability category. Conversely, sample 2 shows a higher mean in the consistency category, but only for the connected speech speaking condition.

Table 6. Mean number of items selected for possible features with 95% confidence intervals across all listeners for each speaker sample.

	AMRs/SMRs			**Connected Speech**		
	Variable (max=4)	**Consistent (max=3)**	**Normal (max=4)**	**Variable (max=7)**	**Consistent (max=6)**	**Normal (max=6)**
Speaker 1	2.6 ± 0.84	0.8 ± 0.57	0.6 ± 0.43	3.1 ± 0.90	1.1 ± 0.68	1.9 ± 0.68
Speaker 2	1.3 ± 0.78	1.4 ± 0.67	1.3 ± 0.51	1.2 ± 0.76	4.1 ± 1.03	0.6 ± 0.52
Speaker 3	2.6 + 0.63	0.89 ± 0.48	0.56 ± 0.33	3.56 ± 1.12	1.0 ± 0.44	1.44 ± 0.88
Speaker 4	3.22 ± 0.60	0.56 ± 0.45	0.22 ± 0.27	2.78 ± 1.19	0.78 ± 0.68	2.44 ± 1.08
Speaker 5	2.22 ± 0.52	1.22 ± 0.27	0.56 ± 0.33	3.0 ± 0.98	2.33 ± 0.98	0.78 ± 0.60
Speaker 6	3.22 ± 0.52	0.56 ± 0.33	0.22 ± 0.27	3.33 ± 1.16	3.22 ± 1.27	0.11 ± 0.21
Speaker 7	3.67 ± 0.44	0 ± 0	0.33 ± 0.44	2.89 ± 1.05	0.80 ± 0.37	2.22 ± 1.02
Speaker 8	2.78 ± 0.74	0.89 ± 0.48	0.33 ± 0.44	3.67 ± 1.12	1.56 ± 0.94	0.89 ± 0.65
Speaker 9	2 ± 0.69	1.33 ± 0.54	0.67 ± 0.54	1.22 ± 0.52	4.44 ± 0.33	0.56 ± 0.45
Speaker 10	1.78 ± 0.52	1.67 ± 0.54	0.56 ± 0.45	2.67 ± 0.93	2.44 ± 0.88	1.0 ± 0.88

Membership in a given subgroup was based on the following criterion: samples needed greater than a 20% difference between inflexibility/instability in a given speaking task, with the second speaking task demonstrating a minimum of a 10% difference in the same direction. The percent of features chosen for a given profile (i.e., instability, inflexibility, normal), is shown in Figures 1-10 as well as Appendix C.

Instability

Patterns of instability are visually depicted in Figures 1-5. The instability or “variability” subgroup had five speakers who fit the proposed profile. Frequency counts of listener ratings per subgroup can be found in Appendix D (AMR/SMR task) and Appendix E (connected speech task).

Figure 1-5. Percent of features chosen in any given rating category, per task, across the 10 listeners for speakers fitting the "instability" profile.

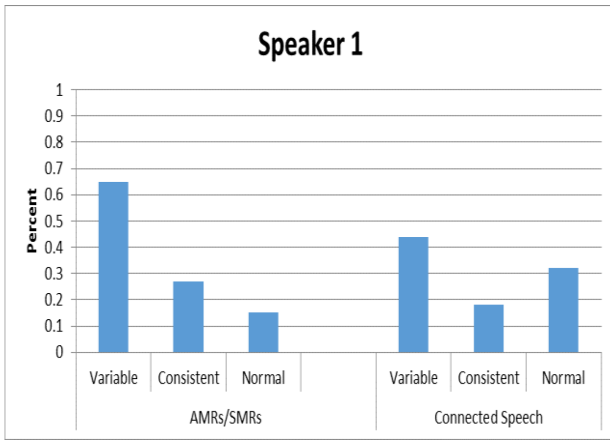


Figure 1

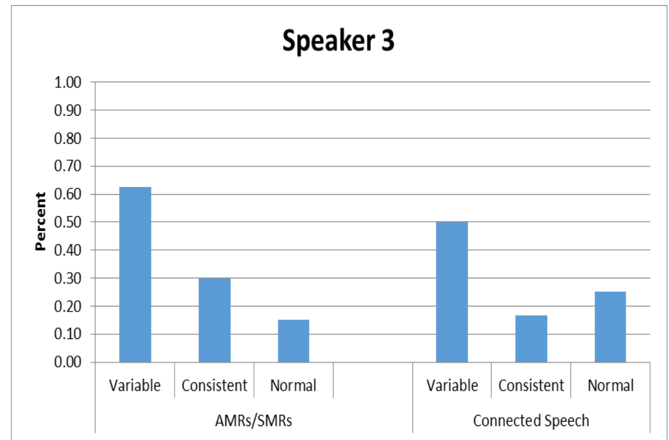


Figure 2

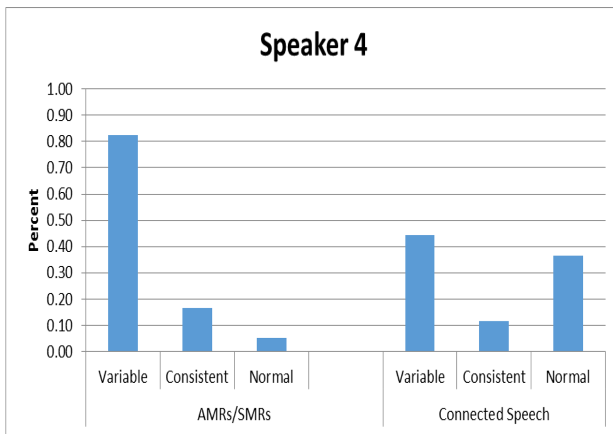


Figure 3

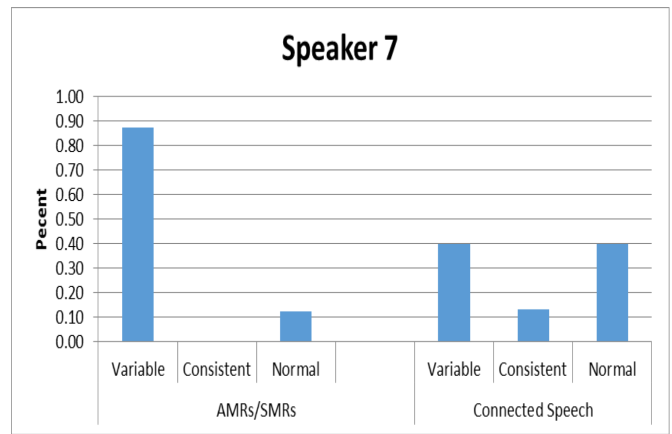


Figure 4

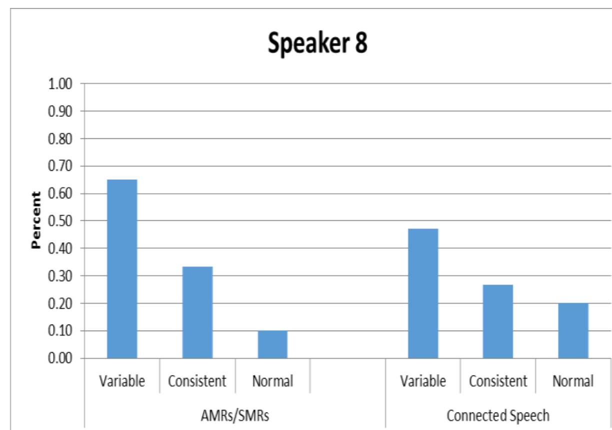


Figure 5

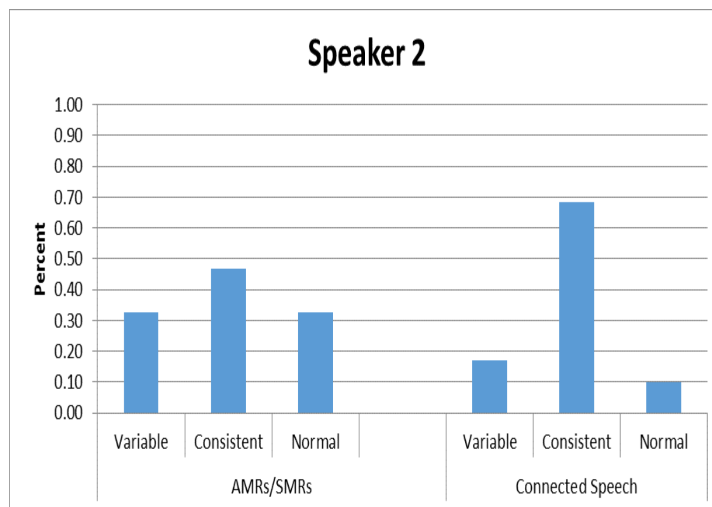
The AMR/SMR task appears to be more sensitive to the percept of variability than the connected speech task. As AMRs/SMRS are a maximum performance task that requires motor control and steadiness, deviations to steadiness are readily apparent. Closer examination of the individual rating forms (see Appendix D) revealed disruptions in rhythm as the most salient feature with 92% of listeners selecting variable rhythm for these five samples. The feature of variable articulation within the AMRs/SMRs task was selected by 74% of listeners.

During the connected speech task, 74% of raters selected articulation as being variable (see Appendix E). Stress was the next salient feature to be rated as variable by a majority of listeners; 60% heard variability in these five samples. Conversely, loudness was rated as variable by the least number of listeners; only 32% felt that loudness varied unusually during connected speech, while 56% of listeners felt that loudness was normal.

Inflexibility

Patterns of inflexibility or "consistency" were apparent only for speaker 2. These rating trends are reflected below in Figure 6.

Figure 6. Percent of features chosen in any given rating category, per task, across the 10 listeners for the speaker fitting the "inflexibility" profile.



The task of connected speech appeared more sensitive to the percept of consistency than the AMR/SMR task. Examination of the individual rating forms suggested that features associated with prosodic disruption were most salient to the listeners. Specifically, slow rate was noted by 9 of 10

listeners, excess loudness by 8 of 10 listeners, and equalized stress patterns and monopitch were selected by 7 of 10 raters.

Mixed features

Samples not clearly meeting criteria of inflexibility or instability were classified as “mixed”. Rating trends are reflected below in Figures 7 -10. For several of these speakers, one speaking task favored one subgroup, but the second task did not meet the 10% criterion. Samples 5, 6 and 9 illustrate this pattern; one speaking task was compelling for a particular subgroup, but the second task showed a negligible difference between subgroup categories. Specifically, samples 5 and 6 were rated as predominantly variable for the AMR/SMR tasks but, during conversation, listeners were equally split between features representing variability and consistency. Sample 9, on the other hand, demonstrated consistency during the conversational speaking task, yet this consistency was not detected by a majority of listeners during the AMR/SMR task. Speaker 10 showed minimal to no difference between variability and consistency for both speaking tasks.

Figures 7 – 10. Percent of features chosen in any given rating category, per task, across the 10 listeners for speakers fitting the "mixed" profile.

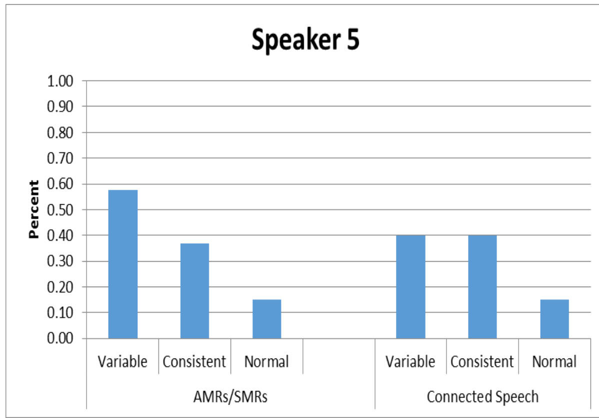


Figure 7

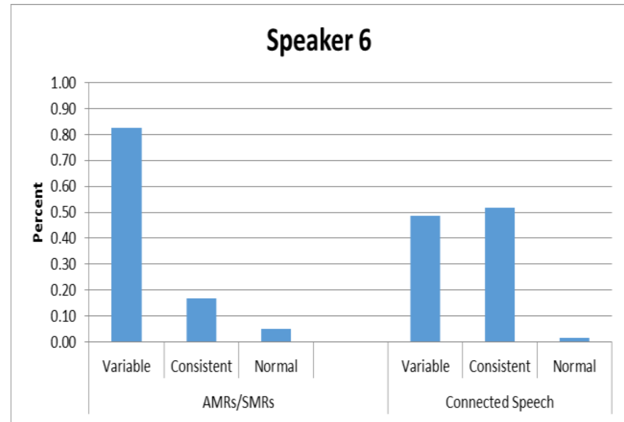


Figure 8

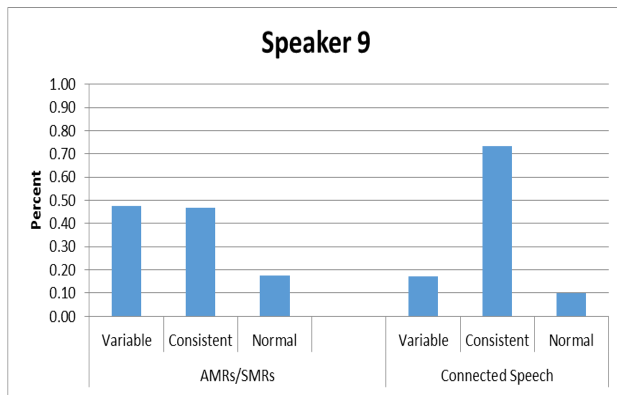


Figure 9

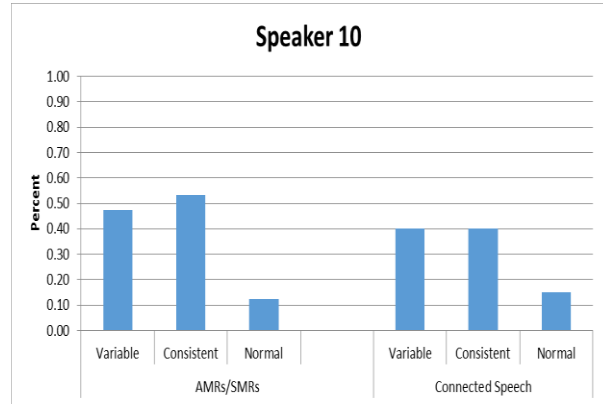


Figure 10

Severity

Aside from speaker diagnosis, there was little information available to generally characterize the speakers. To add to the subject description, perceptual ratings of severity were conducted by the two authors (AF and KS). Using the Mayo Clinic rating scale (Duffy, 2012), where 0 is normal; 1 is mild; 2 is moderate; 3 is marked, and 4 is severely deviant, samples were rated by the investigator and classified through consensus agreement. Results are summarized in Table 7. Most of the samples, 6/10, were classified as moderate; 1/10 was considered mild and 3/10 were marked.

Table 5. Summary of severity classification for speaker samples.

Sample	Investigator 1	Investigator 2	Classification	Subgroup
1	2	2	Moderate	Instability
2	2	2	Moderate	Inflexibility
3	1/2	2	Moderate	Instability
4	2	2	Moderate	Instability
5	2	2	Moderate	Mixed
6	3	3	Marked	Mixed
7	3	3	Marked	Instability
8	2	2	Moderate	Instability
9	3	3	Marked	Mixed
10	2/1	1	Mild	Mixed

Discussion

The goal of this study was to determine if subgroups of ataxic dysarthria exist. While there is limited mention in the literature that ataxic dysarthria may potentially be divided into subgroups, no one has outlined the possible characteristics of these subgroups. Using a framework of motor instability and inflexibility as proposed by Duffy (2012), the hypothesized subgroups were developed. Additionally, a pilot study was conducted to determine if experienced speech-language pathologists could perceptually differentiate these putative subgroups. Specifically, we examined the hypothesis that patterns of speech characteristics would emerge that align with the proposed symptoms of inflexibility and/or instability.

Instability

Speaker samples 1, 3, 4, 7 and 8 demonstrated qualities rated by expert listeners as "variable", thus fitting the proposed subgroup of instability. Both the connected speech task and the AMR/SMR task were able to capture the speech variability. However, AMRs/SMRs appeared most sensitive, as features of articulation and rhythm were perceived to be more variable in this task than during connected speech. By their very nature, AMRs/SMRs require steadiness, rhythmicity, and speed, and would therefore showcase arrhythmic, irregular performance. The focus on a discrete phoneme (e.g., ta-ta-ta) also likely facilitated the detection of irregular breakdowns of articulation. Upon review of the literature, it was felt that rate would also be a salient indicator of instability in this population. While the majority of listeners (66%) did indeed detect rate abnormalities across the five speakers in this subgroup, it was not as prominent a perceptual feature as rhythm abnormalities (92% of listeners) or irregular articulation (74% of listeners). Least often detected during the AMR/SMR task was instability of voicing.

In the connected speech task, the listeners were most often noting instability of articulation (74% of listeners) and stress placement (60% of listeners) for this subgroup. They less often judged rate, pitch, loudness, and respiration as being variable or irregular. While irregularities of rate, pitch, and loudness are frequently reported in acoustic studies of ataxic dysarthria (Boutsen, et al., 1997; Boutsen, et al., 2011; Hartelius, et al., 2000; Ackermann, et al., 2007), it appears these features are not as apparent as disruptions to articulation and stress placement. Variations in loudness, in particular, were least often detected by the listeners (32%). Respiratory incoordination is another perceptual feature hypothesized to be closely associated with a profile of instability. Yet this characteristic was infrequently identified by the listeners for the instability subgroup. It is possible that the speakers simply did not manifest this characteristic within the brief sample of connected speech that was provided. Furthermore, it appears the listeners were equally split on the rating options for respiration: respiratory incoordination (34%), short phrases (22%), speaking on residual air (25%), and normal performance (36%). This pattern suggests that the listeners were often identifying *some* abnormality of breathing during speech, but were perceiving the respiratory characteristics differently.

Interestingly, of the ten speaker samples, five fit within the instability profile (versus only one speaker fitting the inflexibility profile). As many of the features proposed for the instability profile reflect aspects of coordination and timing (e.g., impaired rhythm, disrupted stress placement, timing of

articulation), it may be that this subgroup underscores the importance of the cerebellar circuit in timing. The essential role of the cerebellum in timing has been well documented via lesion studies, neuroimaging evidence, and perceptual investigations (Spencer & Ivry, 2013). Irregular rhythm, or timing, is ofetn most evident during speech AMRs and SMRs, and has been supported with acoustic measures quantifying this element (Portnoy & Aronson, 1982).

For the five speakers in the instability subgroup, there did not appear to be a pattern regarding severity or etiology. Four of the speakers were rated as “moderate” and one was considered “marked” in severity. Of course, patterns of severity will be better explored in a larger sample size (Boutsen, Bakker, & Duffy, 1997). Very limited information was provided regarding etiology of the speakers. However, with the exception of speaker 1 (cerebellar tumor), the four other speakers had more diffuse impairment rather than focal. Given the limited and restricted sample of speakers available for the present study, it is premature to speculate further about the relationship of instability to etiology or lesion site.

Inflexibility

Speaker sample 2 was the only speaker to demonstrate qualities of excessive “consistency” or inflexibility. The connected speech task appeared most sensitive to the consistent errors. Most notably, these disruptions were best detected on prosodic elements of rate and rhythm.

Results from prior research of ataxic dysarthria suggest the profile of inflexibility may be one of less variation resulting in staccato speech rhythm (Ackermann & Hertrich, 1994; Duffy, 2013; Hartelius, Runmarker, Andersen, & Nord, 2000), equalized stress, reduced rate, monoloudness, monopitch, and consistently errored articulation (Duffy, 2013; Hartelius, Runmarker, Andersen, & Nord, 2000; Kent, Netsell, & Abbs, 1979; Ziegler & Wessel, 1996). Results from this study suggest that prosodic disruption, characterized by a slow rate, excess loudness, equalized stress patterns and monopitch, were the most salient features for the speaker that fit this subgroup. The connected speech task provided the greatest opportunity to observe these characteristics and to differentiate the inflexibility profile.

The listener ratings of speaker 2 are consistent with the literature on prosodic patterns in ataxic dysarthria. Specifically, the equalization pattern of inserting abnormal pauses between words and

syllables is often associated with a slow rate of speech (Kent, Netsell, & Abbs, 1979; Schulz, Dingwall, & Ludlow, 1999). Speaker 2 was recognized by 70% of the listeners as having an equalized stress pattern during connected speech, and by 90% of the listeners as having a slow, consistent rate of speech.

Many of the perceptual features hypothesized to align with a subgroup of inflexibility (e.g., monopitch, monoloudness, consistently distorted vowels, isochronous speech, etc.) were thought to have a possible relationship to hypotonicity. It is impossible to judge the tone of speaker 2 relative to the other speakers (though there was an overall reduction in her facial expression in the video [not viewed by the listeners]); however, it would be interesting for future studies to consider the relationship of this motor feature to the putative subgroups.

Mixed

Speaker samples 5, 6, 9 and 10 did not meet the criteria for membership in the instability or inflexibility subgroups. These speakers often showed a tendency toward a subgroup in one speaking task but not the other. Importantly, no speaker was rated as variable for one speaking task and consistent for the other. This pattern (or lack thereof) lends some credence to the descriptors on the rating form to align with one subgroup and to the ability of the listeners to distinguish the salient features.

A mixed subgroup was proposed to be the most likely presentation of ataxic dysarthria (Duffy, 2013, personal communication) and it emerged as the second most common pattern. It had been suggested that ratings would reflect elements of instability intermingled with attributes of inflexibility (Duffy, 2013). This pattern was seen on numerous occasions where the listeners were equally split in their ratings of variability and consistency for the AMRs/SMRs (speakers 9 and 10) and for connected speech (speakers 5 and 6). As three of the four speakers trended toward a subgroup in one of the speaking tasks, it remains unknown whether a longer sample or a refined rating scale would alter their subgroup membership.

Speaker 10 was the only speaker with split ratings in *both* speaking tasks. Her AMRs/SMRs were characterized by slow rate, variable rhythm and mixed variable-consistent articulatory errors. Similarly, connected speech primarily revealed a slow rate, uncoordinated respiration, and variable articulation. Speaker 10 is unique in that she was the only speaker with a mild severity rating and the only known speaker with spinocerebellar degeneration (i.e., Friedreich's Ataxia). It is uncertain if or how these

characteristics may have contributed to the perception that speaker 10 manifests both instability and inflexibility. As Friedreich's ataxia alters the functioning of the spinal and peripheral nerves, leading to issues with tone, it will be interesting for future studies to delineate the possible influence of tone on ataxic dysarthria subgroup membership.

Sustained Phonation

Sustained phonation ratings were removed from the analyses. As indicated earlier, only 8 of the 10 speaker samples contained a recording of sustained phonation. Additionally, as this was a feasibility study, further refinement may be needed for the rating sheet to better define the variable and/or consistent features that are perceptually salient within a sustained phonation task. Specifically, characteristics which yield better descriptions of consistency may need to be added, given that one of the features only allowed a rating aligned with variability. While a sustained vowel may not best demonstrate a pathology when compared to a more complex task such as connected speech (Bele, 2005), it may still shed light on respiratory-phonatory integrity and elements of stability in the motor system.

Further, for "pitch", only a binary judgment could be made, i.e., variable or normal. Having only these two options may not have accurately reflected a rater's preference. In fact, several listeners commented that though a sample did not fit the "variable" selection, the pitch should not be categorized as "normal". Acoustic studies suggest that the task of sustained phonation should be particularly useful in the demonstration of instability (Ackermann & Hertrich, 1994). However, as was suggested in Lowit et al. (2010), features of instability and inflexibility may be more evident in tasks of pitch and loudness glides rather than simple sustained phonation.

Reliability

There are many possible influences on intra-rater reliability, including the severity of the samples, the novelty of the rating system, and years of experience. In terms of severity, the speaker samples were limited in the range of severity levels reflected. The majority of speaker samples (60%) were considered moderate in severity. This severity rating is known to be less reliable in perceptual analyses; there tends to be more agreement for the severe and mild ends of the spectrum versus ratings

of moderate samples (Bunton, Kent, Duffy, Rosenbek, & Kent, 2007; Law, et al., 2012; Sofranko & Prosek, 2012). In addition, there is increased reliability when the parameters and rating scale are known to the rater (Eadie, Sroka, Wright, & Merati, 2011; Law, et al., 2012). As this was a feasibility study, the rating form was novel to all participants, and there was no training given to the listeners. However, the features included on the rating form are considered standard in a thorough motor speech assessment (Duffy, 2012). A brief practice or familiarization period with the perceptual task may have improved listener reliability. With respect to years of experience, research from perceptual studies suggests more experience contributes to the rater's template of the range of abnormality (Bunton, Kent, Duffy, Rosenbek, & Kent, 2007; Eadie, et al., 2010; Law, et al., 2012). Therefore, it was thought that experience may have influenced the rater's ability to identify the abnormal profiles. However, in this study, there was no influence of years of experience relative to reliability (r = 0.15).

Overall, the audio samples played for the listeners were brief, and the listeners were asked to rate a complex, interactive set of speech processes on a limited audio sample. Bunton, Kent, Duffy, Rosenbek, and Kent (2007) investigated listener agreement in dysarthria and found the range of reliability to be 58.6% – 90.8% using a known scale (the 0-4 Mayo Clinic Rating Scale). Kearns and Simmons (1988) found a similar range (60% - 90%) of intra-rater reliability for their perceptual study of dysarthria. As this was a novel rating form, the range of reliability for this study, 58.4% to 82.3%, is comparable in range to other perceptual studies of dysarthria. Bunton et al (2007) does discuss that clinicians are trained to identify, but not necessarily qualify, deviant perceptual patterns which may account for the lower end of the reliability range. Therefore, a high level of listener agreement on a particular scale may not be crucial to clinical practice and may be a reflection of the tool used (Bunton et al, 2007). Finally, Sheard et al. (1991) suggested that different dimensions, such as irregular articulation, are poorly defined, or the presence of an option, such as normal, may cause confusion, resulting in lower agreement when perceptually rating features of ataxic dysarthria.

Clinical Applications

As this was a pilot study, immediate and direct clinical applications are not expected. However, the theoretical and acoustic support for potential subgroups of ataxic dysarthria appears to have salient perceptual correlates. Ataxic dysarthria is heterogeneous, and its characteristics represent a broad spectrum of potential disturbances to the motor speech system. Results of this pilot study provide initial support the presence of instability and mixed subgroups. Further research with a larger sample

size is necessary for classification of the subgroup of inflexibility and confirmation of the other subgroups. Identification of these subgroups using perceptually salient characteristics is relevant to clinical practice and clinical research. Speech-language pathologists may ultimately benefit from a better understanding of the potential heterogeneity in the presentation of ataxic dysarthria. Thus, while two speakers may sound quite dissimilar, a correct diagnosis of ataxic dysarthria may be more readily provided with knowledge of the potential subgroups and their prominent speech characteristics. Improved understanding of the underlying deficit (i.e., instability versus inflexibility) may also potentially influence treatment planning. For example, the treatment approach for someone fitting the inflexibility profile may preferentially focus on activities that bring variability to speech, such as contrastive stress or flexible breath groups. Conversely, the treatment approach for someone fitting the instability profile may target activities to bring stability to speech, such as attenuated use of pitch/loudness for emphasis or regulated breath groups. Clinical research also could be improved by an enhanced description of participants with ataxic dysarthria. At present, all speakers are combined into one group which is likely leading to murky, if not misleading, results.

Limitations and Future Directions

This study was limited by several factors. First, the ten speakers used as exemplary samples of ataxic dysarthria were prerecorded for educational purposes. Therefore, the investigator of this study had no control over the tasks available and how they were elicited and performed for the recording. Also, limited information was known about each speaker. Secondly, the samples could not be spliced to provide a uniform and consistent excerpt without distortion to the sample. Repetitions required some time for the investigator to manipulate the audio file. Third, sustained phonation was not available for all samples, and none of the samples had examples of pitch and loudness glides to measure control of voicing which may have aided creating subgroups. Fourth, while many considerations were made to create a rating form which would capture the proposed elements of each subgroup, further modification and refinement is warranted to best identify the salient features. Finally, for the connected speech task, some listeners chose features from both inflexibility and instability when rating respiration. This happened on 8 occasions and affected 5 speaker samples. Removal of the respiration ratings did not alter subgroup classification and thus were left in the descriptive statistics.

Future development of the rating form should involve consideration of how the respiratory features are presented and whether they indisputably align with each subgroup. Moreover, the form

should include additional ratings for the task of sustained phonation to capture proposed features of inflexibility and a rating for voice quality.

Overall, it is clear that pronounced heterogeneity exists among individuals with ataxic dyarthria. This heterogeneity needs to be better understood to improve clinical diagnosis/treatment, research design and outcomes, as well as models of cerebellar contributions to speech. Ideally, large samples of speakers with well-characterized ataxic dysarthria, across the spectrum of severity, will be studied with an eye to potential subgroups. The convergence of such studies across speech-pathology, neurology, and neuroimaging would potentially bring exciting and meaningful change to the lives of those with ataxia.

Appendix A. Summary of available speech samples of speakers with ataxic dysarthria.

Sample	Sustained phonation	AMRs/ SMRs	Reading	Running speech	Total time available
1	8 sec	AMRs- 24 sec SMRs- 6 sec	33 sec	55 sec	126 sec
2	9 sec	AMRs – 21 sec SMRs – 6 sec	33 sec	62 sec	131 sec
3		AMRs – 25 sec	24 sec	122 sec	171 sec
4		AMRs – 29 sec	14 sec	20 sec	63 sec
5	9 sec	AMRs – 22 sec SMRs – 7 sec	42 sec	32 sec	112 sec
6	21 sec	AMRs – 15 sec SMRs – 4 sec		54 sec	94 sec
7	17 sec	AMRs – 15 sec SMRs – 4 sec		84 sec	120 sec
8	12 sec	AMRs – 26 sec	23 sec		61 sec
9	15 sec	AMRs- 67 sec SMRs- 12 sec		99 sec	193 sec
10	7 sec	AMRs- 41 sec SMRs- 17 sec		86 sec	151 sec

Appendix B. Order of presentation of speaker samples for each listener and repeated samples for intra-rater reliability.

Listener											Reliability	
1	8	9	2	1	10	5	4	3	7	6	7	1
2	4	1	2	6	7	5	10	8	9	3	5	4
3	5	4	2	6	7	3	1	8	10	9	1	5
4	4	8	10	1	7	9	2	3	5	6	3	8
5	1	4	10	3	7	6	8	2	9	5	2	10
6	2	8	9	7	1	5	4	3	6	10	9	7
7	5	3	2	1	4	8	7	9	10	6	3	9
8	7	5	2	3	4	10	6	9	1	8	4	6
9	4	7	10	5	6	8	3	2	1	9	8	6
10	2	6	10	5	1	4	7	3	9	8	10	2

Appendix C. Descriptive statistics for each speaker sample per 10 listeners.

Sample 1.

	AMRs/SMRs			Phonation			Connected Speech		
	Variable	**Consistent**	**Normal**	**Variable**	**Consistent**	**Normal**	**Variable**	**Consistent**	**Normal**
	(max=4)	**(max=3)**	**(max=4)**	**(max=2)**	**(max=1)**	**(max=2)**	**(max=7)**	**(max=6)**	**(max=6)**
Listener 1	4	0	0	1	1	0	2	3	2
Listener 2	3	0	1	2	0	0	4	0	2
Listener 3	3	1	0	1	1	0	4	0	2
Listener 4	1	2	1	1	0	1	2	2	2
Listener 5	0	2	2	1	0	1	1	1	4
Listener 6	2	2	0	2	0	0	3	2	1
Listener 7	4	0	0	2	0	0	6	0	0
Listener 8	3	0	1	2	0	0	3	0	3
Listener 9	2	1	1	1	1	0	4	1	1
Listener 10	4	0	0	1	0	1	2	2	2
%	0.65	0.27	0.15				0.44	0.18	0.32
Average	**2.6**	**0.8**	**0.6**				**3.1**	**1.1**	**1.9**
SD	1.35	0.92	0.70				1.45	1.10	1.10
95% CI	**0.84**	**0.57**	**0.43**				**0.90**	**0.68**	**0.68**
Upper bound	3.44	1.37	1.03				4.00	1.78	2.58
Lower bound	1.76	0.23	0.17	`			2.20	0.42	1.22

Note. % reflects number of selected characteristics in a single category out of the total available for a task; SD = standard deviation; 95% CI =Confidence Interval.

Appendix C (continued).

Sample 2

	AMRs/SMRs			**Phonation**			**Connected Speech**		
	Variable (max=4)	**Consistent (max=3)**	**Normal (max=4)**	**Variable (max=2)**	**Consistent (max=1)**	**Normal (max=2)**	**Variable (max=7)**	**Consistent (max=6)**	**Normal (max=6)**
Listener 1	2	1	1	1	2	0	1	5	0
Listener 2	4	0	0	2	0	0	1	3	2
Listener 3	2	2	0	1	0	1	4	2	0
Listener 4	1	1	2	1	0	1	0	6	0
Listener 5	0	2	2	1	1	0	1	4	1
Listener 6	1	1	2	1	1	0	2	4	0
Listener 7	0	3	1	1	1	0	0	6	0
Listener 8	1	1	2	1	1	0	1	5	0
Listener 9	2	0	2	0	0	2	2	1	2
Listener 10	0	3	1	1	0	1	0	5	1
%	0.33	0.47	0.33				0.17	0.68	0.10
Average	**1.3**	**1.4**	**1.3**				**1.2**	**4.1**	**0.6**
SD	1.25	1.07	0.82				1.23	1.66	0.84
95% CI	**0.78**	**0.67**	**0.51**				**0.76**	**1.03**	**0.52**
Upper bound	2.08	2.07	1.81				1.96	5.13	1.12
Lower bound	0.52	0.73	0.79				0.44	3.07	0.08

Appendix C. (continued).

Sample 3

	AMRs/SMRs			Phonation			Connected Speech		
	Variable (max=4)	Consistent (max=3)	Normal (max=4)	Variable (max=2)	Consistent (max=1)	Normal (max=2)	Variable (max=7)	Consistent (max=6)	Normal (max=6)
Listener 1	3	1	0				6	0	0
Listener 2	3	0	1				3	1	2
Listener 3	2	2	0				5	1	0
Listener 4	2	1	1				3	1	2
Listener 5	4	0	0				2	1	3
Listener 6	1	2	1				4	2	0
Listener 7	4	0	0				5	0	1
Listener 8	2	1	1				0	2	4
Listener 9	2	1	1				4	1	1
Listener 10	2	1	1				3	1	2
%	0.625	0.3	0.15				0.50	0.17	0.25
Average	**2.56**	**0.89**	**0.56**				**3.56**	**1.00**	**1.44**
SD	1.01	0.78	0.53				1.81	0.71	1.42
95% CI	**0.63**	**0.48**	**0.33**				**1.12**	**0.44**	**0.88**
Upper bound	3.18	1.37	0.88				4.68	1.44	2.33
Lower bound	1.93	0.40	0.23				2.43	0.56	0.56

Appendix C. (continued).

Sample 4

	AMRs/SMRs			Phonation			Connected Speech		
	Variable (max=4)	Consistent (max=3)	Normal (max=4)	Variable (max=2)	Consistent (max=1)	Normal (max=2)	Variable (max=7)	Consistent (max=6)	Normal (max=6)
Listener 1	4	0	0				4	0	3
Listener 2	4	0	0				4	0	2
Listener 3	3	1	0				3	2	1
Listener 4	2	1	1				0	3	3
Listener 5	4	0	0				0	1	5
Listener 6	2	2	0				4	1	0
Listener 7	4	0	0				4	0	2
Listener 8	2	1	1				1	0	5
Listener 9	4	0	0				5	0	1
Listener 10	4	0	0				6	0	0
%	0.83	0.17	0.05				0.44	0.12	0.37
Average	**3.22**	**0.56**	**0.22**				**2.78**	**0.78**	**2.44**
SD	0.97	0.73	0.44				1.92	1.09	1.74
95% CI	**0.60**	**0.45**	**0.27**				**1.19**	**0.68**	**1.08**
Upper bound	3.82	1.01	0.50				3.97	1.46	3.52
Lower bound	2.62	0.11	-0.05				1.59	0.10	1.37

Appendix C. (continued).

Speaker 5

	AMRs/SMRs			Phonation			Connected Speech		
	Variable (max=4)	Consistent (max=3)	Normal (max=4)	Variable (max=2)	Consistent (max=1)	Normal (max=2)	Variable (max=7)	Consistent (max=6)	Normal (max=6)
Listener 1	3	1	0	1	1	0	3	4	0
Listener 2	3	1	0	1	1	0	3	3	0
Listener 3	2	1	1	1	0	1	2	1	3
Listener 4	2	1	1	2	0	0	3	2	1
Listener 5	1	2	1	1	0	1	3	2	1
Listener 6	3	1	0	2	0	0	3	3	0
Listener 7	3	1	0	0	0	2	4	1	1
Listener 8	1	2	1	1	0	1	0	5	1
Listener 9	2	1	1	2	0	0	6	0	0
Listener 10	3	0	1	1	0	1	1	3	2
%	0.58	0.37	0.15				0.40	0.40	0.15
Average	**2.22**	**1.22**	**0.56**				**3.00**	**2.33**	**0.78**
SD	0.83	0.44	0.53				1.58	1.58	0.97
95% CI	**0.52**	**0.27**	**0.33**				**0.98**	**0.98**	**0.60**
Upper bound	2.74	1.50	0.88				3.98	3.31	1.38
Lower bound	1.71	0.95	0.23				2.02	1.35	0.18

Appendix C. (continued).

Speaker 6

	AMRs/SMRs			Phonation			Connected Speech		
	Variable (max=4)	Consistent (max=3)	Normal (max=4)	Variable (max=2)	Consistent (max=1)	Normal (max=2)	Variable (max=7)	Consistent (max=6)	Normal (max=6)
Listener 1	3	1	0	1	1	0	3	4	0
Listener 2	2	1	1	2	0	0	3	2	1
Listener 3	3	1	0	2	0	0	7	0	0
Listener 4	2	1	1	1	1	0	2	4	0
Listener 5	4	0	0	2	0	0	2	5	0
Listener 6	3	1	0	1	1	0	3	4	0
Listener 7	4	0	0	1	1	0	2	5	0
Listener 8	4	0	0	1	1	0	2	5	0
Listener 9	4	0	0	1	1	0	6	0	0
Listener 10	4	0	0	1	1	0	4	2	0
%	0.83	0.17	0.05				0.49	0.52	0.02
Average	**3.22**	**0.56**	**0.22**				**3.33**	**3.22**	**0.11**
SD	0.83	0.53	0.44				1.87	2.05	0.33
95% CI	**0.52**	**0.33**	**0.27**				**1.16**	**1.27**	**0.21**
Upper bound	3.74	0.88	0.50				4.49	4.49	0.32
Lower bound	2.71	0.23	-0.05				2.17	1.95	-0.10

Appendix C. (continued).

Sample 7

	AMRs/SMRs			Phonation			Connected Speech		
	Variable (max=4)	Consistent (max=3)	Normal (max=4)	Variable (max=2)	Consistent (max=1)	Normal (max=2)	Variable (max=7)	Consistent (max=6)	Normal (max=6)
Listener 1	4	0	0	1	0	1	1	1	4
Listener 2	4	0	0	1	0	1	3	1	2
Listener 3	3	0	1	0	1	1	2	1	3
Listener 4	2	0	2	2	0	0	5	1	0
Listener 5	4	0	0	1	0	1	1	0	5
Listener 6	4	0	0	1	0	1	4	1	1
Listener 7	4	0	0	1	0	1	4	1	1
Listener 8	4	0	0	0	0	2	1	2	3
Listener 9	4	0	0	1	0	1	5	0	1
Listener 10	2	0	2	1	0	1	2	0	4
%	0.88	0.00	0.13				0.40	0.13	0.40
Average	**3.67**	**0.00**	**0.33**				**2.89**	**0.89**	**2.22**
SD	0.71	0.00	0.71				1.69	0.60	1.64
95% CI	**0.44**	**0.00**	**0.44**				**1.05**	**0.37**	**1.02**
Upper bound	4.10	0.00	0.77				3.94	1.26	3.24
Lower bound	3.23	0.00	-0.10				1.84	0.52	1.20

Appendix C. (continued).

Speaker 8

	AMRs/SMRs			Phonation			Connected Speech		
	Variable (max=4)	**Consistent (max=3)**	**Normal (max=4)**	**Variable (max=2)**	**Consistent (max=1)**	**Normal (max=2)**	**Variable (max=7)**	**Consistent (max=6)**	**Normal (max=6)**
Listener 1	3	1	0	1	0	1	3	1	3
Listener 2	3	0	1	1	0	1	4	1	1
Listener 3	2	2	0	2	0	0	6	0	1
Listener 4	0	2	2	2	0	0	2	4	0
Listener 5	3	1	0	1	0	1	2	2	2
Listener 6	3	1	0	1	1	0	4	1	1
Listener 7	4	0	0	1	1	0	6	0	0
Listener 8	3	1	0	1	0	1	1	4	0
Listener 9	4	0	0	1	1	0	5	1	0
Listener 10	1	2	1	1	0	1	0	2	4
%	0.65	0.33	0.10				0.47	0.27	0.20
Average	**2.78**	**0.89**	**0.33**				**3.67**	**1.56**	**0.89**
SD	1.20	0.78	0.71				1.80	1.51	1.05
95% CI	**0.74**	**0.48**	**0.44**				**1.12**	**0.94**	**0.65**
Upper bound	3.52	1.37	0.77				4.78	2.49	1.54
Lower bound	2.03	0.40	-0.10				2.55	0.62	0.24

Appendix C. (continued).

Sample 9

	AMRs/SMRs			Phonation			Connected Speech		
	Variable (max=4)	Consistent (max=3)	Normal (max=4)	Variable (max=2)	Consistent (max=1)	Normal (max=2)	Variable (max=7)	Consistent (max=6)	Normal (max=6)
Listener 1	3	1	0	1	0	1	2	5	0
Listener 2	4	0	0	1	0	1	1	4	1
Listener 3	2	2	0	0	0	2	2	4	0
Listener 4	1	1	2	1	0	1	0	5	1
Listener 5	1	2	1	2	0	0	0	4	2
Listener 6	1	3	0	2	0	0	2	4	0
Listener 7	3	1	0	2	0	0	2	5	0
Listener 8	2	1	1	2	0	0	1	5	0
Listener 9	1	1	2	1	0	1	1	4	1
Listener 10	1	2	1	1	0	1	1	4	1
%	0.48	0.47	0.18				0.17	0.73	0.10
Average	**2.00**	**1.33**	**0.67**				**1.22**	**4.44**	**0.56**
SD	1.12	0.87	0.87				0.83	0.53	0.73
95% CI	**0.69**	**0.54**	**0.54**				**0.52**	**0.33**	**0.45**
Upper bound	2.69	1.87	1.20				1.74	4.77	1.01
Lower bound	1.31	0.80	0.13				0.71	4.12	0.11

Appendix C. (continued).

Speaker 10

	AMRs/SMRs			Phonation			Connected Speech		
	Variable (max=4)	**Consistent (max=3)**	**Normal (max=4)**	**Variable (max=2)**	**Consistent (max=1)**	**Normal (max=2)**	**Variable (max=7)**	**Consistent (max=6)**	**Normal (max=6)**
Listener 1	3	1	0	1	1	0	3	4	0
Listener 2	3	1	0	1	1	0	2	3	1
Listener 3	1	3	0	1	1	0	2	3	2
Listener 4	1	2	1	1	0	1	2	4	0
Listener 5	1	1	2	1	0	1	1	1	4
Listener 6	2	1	1	1	1	0	5	1	0
Listener 7	2	1	1	1	0	1	3	1	2
Listener 8	1	3	0	1	1	0	1	4	0
Listener 9	2	2	0	2	0	0	5	1	0
Listener 10	3	1	0	2	0	0	4	2	0
%	0.48	0.53	0.13				0.40	0.40	0.15
Average	**1.78**	**1.67**	**0.56**				**2.67**	**2.44**	**1.00**
SD	0.83	0.87	0.73				1.50	1.42	1.41
95% CI	**0.52**	**0.54**	**0.45**				**0.93**	**0.88**	**0.88**
Upper bound	2.29	2.20	1.01				3.60	3.33	1.88
Lower bound	1.26	1.13	0.11				1.74	1.56	0.12

Appendix D. Frequency counts of listener ratings per subgroup for AMR/SMR task.

	Articulation			**Voicing**			**Rate**			**Rhythm**		**Total**
	Variable	Consistent	Normal	Variable	Consistent	Normal	Variable	Consistent	Normal	Variable	Normal	
Sample 1	6	4		5	1	4	6	3	1	9	1	40
Sample 2	4	4	2	2	3	5	3	7		4	6	40
Sample 3	7	2	1	5	1	4	4	6		9	1	40
Sample 4	8	2		6	2	2	9	1		10		40
Sample 5	7	3		5		5	2	8		9	1	40
Sample 6	10			7	1	2	6	4		10		40
Sample 7	9		1	8		2	9		1	9	1	40
Sample 8	7	3		6	1	3	5	5		9	1	40
Sample 9	4	5	1	4	2	4	2	8		9	1	40
Sample 10	4	4	2	4	4	2	2	8		9	1	40
Instability												
Sample 1	6	4		5	1	4	6	3	1	9	1	40
Sample 3	7	2	1	5	1	4	4	6		9	1	40
Sample 4	8	2		6	2	2	9	1		10		40
Sample 7	9		1	8		2	9		1	9	1	40
Sample 8	7	3		6	1	3	5	5		9	1	40
Average	*7.4*	*2.75*	*1*	*6*	*1.25*	*3*	*6.6*	*3.75*	*1*	*9.2*	*1*	
Inflexibility												
Sample 2	4	4	2	2	3	5	3	7		4	6	40
Mixed												
Sample 5	7	3		5		5	2	8		9	1	40
Sample 6	10			7	1	2	6	4		10		40
Sample 9	4	5	1	4	2	4	2	8		9	1	40
Sample 10	4	4	2	4	4	2	2	8		9	1	40
Average	*6.25*	*4*	*1.5*	*5*	*2.33*	*3.25*	*3*	*7*		*9.25*	*1*	

Appendix E. Frequency counts of listener ratings per subgroup for connected speech task.

	Articulation			Stress			Rate			Pitch			Respiration				Loudness				
	Variable	Consistent	Normal	Irregular	Equalized	Normal	Variable	Slow	Normal	Variable	Monopitch	Normal	Incoordination	Residual air	Short phrases	Normal	Uncontrolled	Too loud	Too quiet	Normal	Total
Sample 1	9	1		6		4	4	5	1	4	1	5	3	2	3	3	3		1	6	61
Sample 2	5	4	1	3	7		1	9		3	7				6	3		8		2	59
Sample 3	7	1	2	8	1	1	4	5	1	6	2	2	3	2		5	5	1		4	60
Sample 4	6	3	1	6	1	3	7	1	2	4	1	5	2	4	1	4	3			7	61
Sample 5	8	2		5	5		3	7		2	6	2	3	1	2	5	6	1	1	2	61
Sample 6	9	1		4	6		3	7		3	7		6	6	3		2	7		1	65
Sample 7	9	1		6	1	3	6		4	3	2	5	1		4	5	3			7	60
Sample 8	6	4		4	4	2	5	1	3	6	2	2	8	2	1	1	2	1	3	4	61
Sample 9	5	5			10			10			10			6	4	2	1	5		4	62
Sample 10	6	4		5	4	1	1	9		3	4	3	7	1	2	1	5		1	4	61
Instability																					
Sample 1	9	1		6		4	4	5	1	4	1	5	3	2	3	3	3		1	6	61
Sample 3	7	1	2	8	1	1	4	5	1	6	2	2	3	2		5	5	1		4	60
Sample 4	6	3	1	6	1	3	7	1	2	4	1	5	2	4	1	4	3			7	61
Sample 7	9	1		6	1	3	6		4	3	2	5	1		4	5	3			7	60
Sample 8	6	4		4	4	2	5	1	3	6	2	2	8	2	1	1	2	1	3	4	61
Average	*7.4*	*2*	*1.5*	*6*	*1.75*	*2.6*	*5.2*	*3*	*2.2*	*4.6*	*1.6*	*3.8*	*3.4*	*2.5*	*2.25*	*3.6*	*3.2*	*1*	*2*	*5.6*	
Inflexibility																					
Sample 2	5	4	1	3	7		1	9		3	7				6	3		8		2	59
Mixed																					
Sample 5	8	2		5	5		3	7		2	6	2	3	1	2	5	6	1	1	2	61
Sample 6	9	1		4	6		3	7		3	7		6	6	3		2	7		1	65
Sample 9	5	5			10			10			10			6	4	2	1	5		4	62
Sample 10	6	4		5	4	1	1	9		3	4	3	7	1	2	1	5		1	4	61
Average	*7.00*	*3.00*		*4.67*	*6.25*	*1.00*	*2.33*	*8.25*		*2.67*	*6.75*	*2.50*	*5.33*	*3.50*	*2.75*	*2.67*	*3.50*	*4.33*	*1.00*	*2.75*	

Appendix F. Script for instructing listeners.

Investigator: Here are the rating forms you will be using for this study. I will play a sample of DDKs, sustained phonation and connected speech (pointing to each section). Please rate only the features found in each section. So, for example, for the DDKs, rate whether you are hearing variable or consistent or normal articulation, voicing, rate and rhythm (again pointing to each feature). I will always tell you what task you are going to be listening to before I start playing the sample. You can ask for the sample to be repeated up to 5 times. Please rate only what you are hearing in that sample. So, for example, you will be rating articulation in the DDKs and the connected speech. When I play the connected speech portion, rate the articulation you are hearing only in the connected speech portion. You may select two features for respiration and loudness in the connected speech task, if needed. Do you have any questions?

References

Ackermann, H., & Hertrich, I. (1994). Speech Rate and Rhythm in Cerebellar Dysarthria: An Acoustic Analysis of Syllabic Timing. *Folia Phoniatrica et Logopaedica, 46*(2), 70-78.

Ackermann, H., Hertich, I., & Hehr, T. (1995). Oral diadochokinesis in neurological dysarthrias. *Folia Phoniatrica et Logopaedica*, 15-23.

Ackermann, H., Mathiak, K., & Riecker, A. (2007). The contribution of the cerebellum to speech production and speech perception: Clinical and functional imaging data. *The Cerebellum*, 202-213.

Ackermann, H., Vogel, M., Petersen, D., & Poremba, M. (1992). Speech deficits in ischaemic cerebellar lesions. *Journal of Neurology, 239*(4), 223-227.

American Speech-Language-Hearing Association. (1997). *Guidelines for audiological screening.* Retrieved from asha.org: http://www.asha.org/policy/GL1997-00199.htm

Barlow, J. S. (2002). *The Cerebellum and Adaptive Control.* Cambridge: Cambridge University Press.

Bele, I. V. (2005). Reliability in Perceptual Analysis of Voice Quality. *Journal of Voice, 19*(4), 555-573.

Bohland, J. W., & Guenther, F. H. (2006). An fMRI investigation of syllable sequence production. *Neuroimage, 32*(2), 821-841.

Boutsen, F. R., Bakker, K., & Duffy, J. R. (1997). Subgroups in Ataxic Dysarthria. *Journal of Medical Speech-Language Pathology*, 27-36.

Boutsen, F., & Christman, S. (2002). Prosody in apraxia of speech. *Seminars in Speech and Language*, 245-256.

Boutsen, F., Duffy, J. R., Dimassi, H., & Christman, S. S. (2011). Long-Term Phonatory Instability in Ataxic Dysarthria. *Folia Phoniatrica et Logopaedica*, 216-220.

Brown, J. R., Darley, F. L., & Aronson, A. E. (1970). Ataxic Dysarthria. *International Journal of Neurology, 7*(2), 302-318.

Bunton, K., Kent, R. D., Duffy, J. R., Rosenbek, J. C., & Kent, J. F. (2007, December). Listener Agreement for Auditory-Perceptual Ratings of Dysarthria. *Journal of Speech, Language, and Hearing Research, 50*, 1481-1495. doi:1092-4388/07/5006-1481

Cummins, F., Lowit, A., & van Brenk, F. (2013). Quantitative Assessment of Interutterance Stability: Application to Dysarthria. *Journal of Speech, Language and Hearing Reseach*, 1-9.

Darley, F. L., Aronson, A. E., & Brown, J. R. (1969). Clusters of Deviant Speech Dimensions in the Dysarthrias. *Journal of Speech and Hearing Research*, 462-496.

Darley, F. L., Aronson, A. E., & Brown, J. R. (1969). Differential Diagnostic Patterns of Dysarthria. *Journal of Speech and Hearing Research, 12*, 246-269.

Duffy, J. R. (2013). *Motor Speech Disorders: Substrates, Differential Diagnosis, and Management* (Third ed.). St. Louis: Elsevier Mosby.

Eadie, T., Kapsner, M., Rosenzweig, J., Waugh, P., Hillel, A., & Merati, A. (2010). The Role of Experience on Judgments of Dysphonia. *The Journal of Voice, 24*(5), 564-573.

Eadie, T., Sroka, A., Wright, D. R., & Merati, A. (2011). Does Knowledge of Medical Diagnosis Bias Auditory-Perceptual Judgments of Dysphonia? *Journal of Voice, 25*(4), 420-429.

Folker, J. E., Murdoch, B. E., Rosen, K. M., Cahill, L. M., Delaytcki, M. B., Corben, L. A., & Vogel, A. P. (2012). Differentiating profiles of speech impairments in Friedreich's ataxia: a perceptual and instrumental approach. *International Journal of Language & Communication Disorders*, 65-76.

Guenther, F. H., Ghosh, S. S., & Tourville, J. A. (2006). Neural modeling and imaging of the cortical interactions underlying syllable production. *Brain Language, 96*(3), 280-301.

Hartelius, L., Runmarker, B., Andersen, O., & Nord, L. (2000). Temporal Speech Characteristics of Individuals with Multiple Sclerosis and Ataxic Dysarthria: 'Scanning Speech' Revisited. *Folia Phoniatrica et Logopaedica*, 228-238.

Henrich, J., Lowit, A., Schalling, E., & Mennen, I. (2006, December). Rhythmic Disturbance in Ataxic Dysarthria: A Comparison of Different Measures and Speech Tasks. *Journal of Medical Speech-Language Pathology, 14*(4), 291-296.

Hertrich, I., & Ackermann, H. (1999, April). Temporal and Spectral Aspects of Coarticulation in Ataxic Dysarthria: An Acoustic Analysis. *Journal of Speech, Language and Hearing Research, 42*, 367-381.

Kearns, K. P., & Simmons, N. N. (1988). Interobserver Reliability and Perceptual Ratings: More Than Meets the Ear. *Journal of Speech and Hearing Research, 31*, 131-136. doi:0022-4685/88/3101-0131

Kent, R. D., Kent, J. F., Duffy, J. R., Thomas, J. E., Weismer, G., & Stuntebeck, S. (2000, October). Ataxic Dysarthria. *Journal of Speech, Language and Hearing Research, 43*, 1275-1289. doi:1092-4388/00/4305-1275

Kent, R. D., Kent, J. F., Rosenbek, J. C., Vorperian, H. K., & Weismer, G. (1997). A Speaking Task Analysis of the Dysarthria in Cerebellar Disease. *Folia Phoniatrica et Logopaedica, 49*, 63-82. doi:1021-7762/97/0492-0063

Kent, R. D., Netsell, R., & Abbs, J. H. (1979, September). Acoustic Characteristics of Dysarthria Associated with Cerebellar Disease. *Journal of Speech and Hearing Research, 22*, 627-648.

Kent, R., & Netsell, R. (1975). A case study of an ataxic dysarthric: cineradiographic and spectrographic. *Journal of Speech and Hearing Disorders*, 115-134.

Kluin, K. J., Gilman, S., Markel, D. S., Koeppe, R. A., Rosenthal, G., & Junck, L. (1988). Speech disorders in olivopontocerebellar atrophy correlate with positron emission tomography findings. *Annals of Neurology, 23*(6), 547-554.

Law, T., Kim, J. H., Lee, K. Y., Tang, E. C., Lam, J. H., van Hasselt, A. C., & Tong, M. C. (2012). Comparison of Rater's Relaibility on Perceptual Evaluation of Different Types of Voice Sample. *Journal of Voice, 26*(5), 666.e13-666.e21.

Liss, J. M., White, L., Mattys, S. L., Lansford, K., Lotto, A. J., Spitzer, S. M., & Caviness, J. N. (2009). Quantifying Speech Rhythm Abnormalities in the Dysarthrias. *Journal of Speech, Language, and Hearing Research*, 1334-1352.

Lowit, A., & Kuschmann, A. (2012, October). Characterizing Intonation Deficit in Motor Speech Disorders: An Autosegmental-Metrical Analysis of Spontaneous Speech in Hypokinetic Dysarthria, Ataxic Dysarthria, and Foreign Accent Syndrome. *Journal of Speech, Language, and Hearing Research, 55*, 1472-1484. doi:10.1044/1092-4388

Lowit, A., Kuschmann, A., MacLeod, J. M., Schaeffler, F., & Mennen, I. (2010). Sentence Stress in Ataxic Dysarthria - A Perceptual and Acoustic Study . *Journal of Medical Speech-Language Pathology*, 77-82.

Murdoch, B. E., & Theodoros, D. G. (1998). *Dysarthria: A Physiological Approach to Assessment and Treatment.* (B. E. Murdoch, Ed.) Bolton: Stanley Thornes (Publishers) Ltd.

Netsell, R., & Kent, R. D. (1976). Paroxysmal Ataxic Dysarthria. *Journal of Speech and Hearing Disorders, XLI*, 93-109.

Odell, K., McNeil, M. R., Rosenbek, J. C., & Hunter, L. (1991, February). Perceptual characteristics of vowel and prosody production in apraxic, aphasic and dysarthric speakers. *Journal of Speech and Hearing Research, 34*, 67-80. doi:0022-4685/91/3401-0067

Paslawski, T., Duffy, J. R., & Vernino, S. (2005, August). Speech and Language Findings Associated with Paraneoplastic Cerebellar Degeneration. *American Journal of Speech-Language Pathology, 14*, 200-207.

Portnoy, R. A., & Aronson, A. E. (1982, August). Diadochokinetic syllable rate and regularity in normal and in spastic and ataxic dysarthric subjects. *Journal of Speech and Hearing Disorders, 47*, 324-328. doi:0022-4677/82/4703-0324

Schalling, E., Hammarberg, B., & Hartelius, L. (2008). A Longitudinal Study of Dysarthria in Spinocerebellar Ataxia (SCA): Aspects of Articulation, Prosody, and Voice. *Journal of Medical Speech-Language Pathology, 16*(2), 103-117.

Sheard, C., Adams, R. D., & Davis, P. J. (1991, April). Reliability and Agreement of Ratings of Ataxic Dysarthric Speech Samples with Varying Intelligibility. *Journal of Speech and Hearing Research, 34*, 285-293.

Schulz, G. M., Dingwall, W. O., & Ludlow, C. L. (1999, October). Speech and Oral Motor Learning in Individuals with Cerebellar Atrophy. *Journal of Speech, Language, and Hearing Research, 42*, 1157-1175. doi:1092-4388/99/4205-1157

Sofranko, J. L., & Prosek, R. A. (2012). The Effect of Experience on Classification of Voice Quality. *Journal of Voice, 26*(3), 299-303.

Spencer, M.C. & Ivry, R.B. (2013). Cerebellum and timing. *Handbook of the Cerebellum and Cerebellar Disorders*, pg. 1201-1219.

Spencer, K. A., & Slocomb, D. L. (2007). The neural basis of ataxic dysarthria. *The Cerebellum, 6*, 58-65. doi:10.1080/14734220601145459

Vandana, V. P. (2007). Reflections on Speech Motor Control Based on Phonatory and DDK Tasks in Dysarthric Subjects with Lesions in Different Cerebellar Loci. *Indian Journal of Applied Linguistics*, 63-75.

Vandana, V. P., & Manjul, R. (2006). Relational Speech Timing in Dysarthria Associated wtih Cerebellar Lesions in Different Loci: Word Context. *Indian Journal of Applied Linguistics*, 41-54.

Yorkston, K. M., & Beukelman, D. R. (1981, November). Ataxic Dysarthria: Treatment Sequences Based on Intelligibility and Prosodic Considerations. *Journal of Speech and Hearing Disorders, 46*, 398-404.

Yorkston, K. M., & Beukelman, D. R. (1984). Assessment of Intelligibility of Dysarthric Speech. Austin, TX: PRO-ED.

Yorkston, K. M., Beukelman, D. R., Strand, E. A., & Hakel, M. (2010). *Management of Motor Speech Disorders in Children and Adults.* Austin: PRO-ED Inc.

Zeplin, J., & Kent, R. D. (1996). Reliability of auditory perceptual scaling of dysarthria. In D. Robin, K. Yorkston, & D. R. Buekelman, *Disorders of Motor Speech: Recent advances in assessment, treatment, and clinical characterization.* Baltimore: Paul H. Brookes.

Ziegler, W., & Wessel, K. (1996). Speech timing in ataxic disorders: Sentence production and rapid repetitive articulation. *Neurology*, 208-214.