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# Oral motor functions, speech and communication before a definitive diagnosis of amyotrophic lateral sclerosis

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ABSTRACT

The aim of this study was to explore the cranial nerve symptoms, speech disorders and communicative effectiveness of Finnish patients with diagnosed or possible amyotrophic lateral sclerosis (ALS) at their first assessment by a speech-language pathologist. The group studied consisted of 30 participants who had clinical signs of bulbar deterioration at the beginning of the study. They underwent a thorough clinical speech and communication examination. The cranial nerve symptoms and ability to communicate were compared in 14 participants with probable or definitive ALS and in 16 participants with suspected or possible ALS. The initial type of ALS was also assessed.

More deterioration in soft palate function was found in participants with possible ALS than with diagnosed ALS. Likewise, a slower speech rate combined with more severe dysarthria was observed in possible ALS. In both groups, there was some deterioration in communicative effectiveness. In the possible ALS group the diagnostic delay was longer and speech therapy intervention actualized later.

The participants with ALS showed multidimensional decline in communication at their first visit to the speech-language pathologist, but impairments and activity limitations were more severe in suspected or possible ALS. The majority of persons with bulbar-onset ALS in this study were in the latter diagnostic group. This suggests that they are more susceptible to delayed diagnosis and delayed speech therapy assessment. It is important to start speech therapy intervention during the diagnostic processes particularly if the person already shows bulbar symptoms.



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## 1. Introduction

Amyotrophic lateral sclerosis (ALS) is generally considered a relentlessly and rapidly progressing neurological disease that destroys motor neurons in the cerebral cortex, brainstem and spinal cord (Kiernan et al., 2011). The diagnosis of ALS is challenging and based on clinical criteria (Brooks, Miller, Swash, Munsat, & World Federation of Neurology Research Group on Motor Neuron Diseases, 2000; Kiernan et al., 2011). The widely recognized El Escorial criteria utilize a combination of upper and lower motor neuron symptoms to establish levels of diagnostic certainty (Brooks et al., 2000). In one third of the persons with ALS, the disease starts with bulbar symptoms as the deterioration of the corticobulbar tract affects the innervation of the muscles responsible for speech functions (Yorkston, Strand, Miller, Hillel, & Smith, 1993). In about 70% of the cases, the first symptoms appear in the limbs (spinal-onset disease), and in some the disease starts with initial trunk or respiratory involvement.

Typically, it takes a long time before a definitive diagnosis of ALS is reached (Williams, Fitzhenry, Grant, Martyn & Kerr, 2013). The median delay between the onset of symptoms and definitive diagnosis is about 14 months, and the diagnostic delay is especially common for bulbar-onset (Kiernan et al., 2011; Turner et al., 2010). The bulbar-onset disease is associated with a worse prognosis than spinal-onset (Williams et al., 2013), and bulbar function plays a major role in determining the outcome at any stage of the illness (Chio et al., 2009). As the disease progresses, a person with ALS typically demonstrates a flaccid-spastic mixed dysarthria (Duffy, 2013).

Speech and communication research on ALS has so far focused on people with a definitive ALS diagnosis. Dysfunctions such as decreased strength and movement of the velopharynx, larynx, lips, tongue and jaw have been reported (Hanson, Yorkston, & Britton, 2011; Hillel et al., 1989; Langmore & Lehman, 1994; Yunusova, Weismer, Westburry & Lindstrom, 2008) as well as reduced speed of tongue movements and decreased spatiotemporal coupling between different regions of the tongue during word production (Kuruvilla et al., 2012). Thus, lowered articulatory speed may be the earliest sign of motor neuron degeneration in the tongue (Yunusova et al., 2012).

Speech rate is considered important in assessing speech changes in ALS (Ball et al., 2002Ball, Beukelman, & Pattee, 2002; Hanson et al., 2011; Yorkston et al., 1993) because a decreased speech rate is often one of the first symptoms of dysarthria (Kent et al., 1991; Mulligan et al., 1994; Watts & Vanryckeghem, 2001). Slower than normal speech rates have been reported in people with ALS when intelligibility of speech has still been high (Nishio & Niimi, 2000; Turner & Weismer, 1993). Changes in motor functions, such as rapid articulatory movements, seem to precede lowered speech intelligibility in ALS (Hanson et al., 2011). Thus, an individually paced gradual loss of intelligible speech is the ultimate result of progressive, mixed spastic and flaccid dysarthria accompanying ALS (Duffy, 2013; Hanson et al., 2011; Yorkston, Miller, & Strand, 2004). Lowered speech intelligibility is a social handicap, indicating a need for augmentative and alternative communication (AAC) intervention (Yorkston et al., 1993).

As dysarthria becomes more severe and speech intelligibility declines, people with ALS may need AAC modes to communicate effectively. However, communicative effectiveness varies across social situations (Ball, Beukelman, & Pattee, 2004). Communication is more effectively perceived in quiet environments with a familiar person, whereas it is least effective in noisy environments, speaking in a group and in long conversations. The availability of varying modes of communication to be used in different contexts may improve the quality of life and mood of dysarthric people, and the different modes of communication should be provided early in the course of the disease (Körner et al., 2013).

Until now, published data on cranial nerve symptoms, speech and communication changes in ALS have focused on confirmed diagnosis of ALS (e.g. Ball, Willis, Beukelman, & Pattee, 2001; Ball et al., 2004; Langmore & Lehman, 1994; Mulligan et al., 1994; Watts & Vanryckeghem, 2001). A number of research reports lack data on the diagnostic accuracy of ALS. There is no reported research on the changes in speech and communication before ALS diagnosis is confirmed. In clinical work, however, we have noticed that many individuals show bulbar symptoms and speech changes prior to the diagnosis of probable or definitive ALS. The aim of the present prospective study is to compare the speech-related cranial nerve deficits, speech production and communication changes in people with different levels of diagnostic certainties of ALS. We utilize the general framework of the International Classification of Functioning, Disability and Health (ICF, 2001), and focus on the differences between persons with definitive or probable ALS and those with suspected or possible ALS in terms of cranial nerve symptoms and speech production (body functions), severity of dysarthria and use of communication aids (activity), and the efficacy of communication (participation).

## 2. Methods

### 2.1. Participants

This study was conducted at the Neurology and Rehabilitation Department of Tampere University Hospital. From August 2007 to December 2009, 47 consecutive persons were referred to the speech-language pathologist (SLP). Thirty of them met the initial inclusion criteria: (1) native speaker of Finnish; (2) diagnosis of suspected, possible, probable or definitive ALS according to the revised el Escorial criteria (Brooks et al., 2000) at the first SLP visit; (3) one or more clinical