

Changes in communication and swallowing of people with ALS – a follow-up study

Tanja Makkonen, Anna-Maija Korpijaakko-Huuhka
Tampere University Hospital, Finland

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Introduction

Adults with Amyotrophic Lateral Sclerosis (ALS) are a special group of clients in speech and language therapy because of the special features of their disease. ALS is generally considered a relentlessly and rapidly progressive disease, which destroys motor neurons in both brain and spinal cord (Somer, 2006). Its etiology is unknown, and there is no cure for it. A lot of medical research has been published about ALS, but only a few logopedic studies are found about communication and swallowing problems due to ALS. However, bulbar symptoms affecting both speech and swallowing functions are common in patients with ALS (Duffy, 1995; Yorkston *et al.*, 1995; Freed, 2000; Somer, 2006).

The goal of speech therapy for the adult with ALS is to make it possible for him/her to communicate independently – either using natural speech or with augmentative and alternative communication means – as well as to help the patient in keeping the nutritional level as good as possible (Yorkston *et al.*, 1995; Mathy *et al.*, 2000). This is very challenging because of the often very rapid changes of speech and swallowing functions typical of ALS. Information about the rate of symptom progression is critical in treatment planning. The deterioration proceeds individually, and little is known about factors predicting its course. Thus, we need more information about changes in speech and swallowing functions during the deterioration process.

In this presentation, we will picture a research scheme for a follow-up study of 20–30 patients with ALS. We will concentrate on communication and only mention other problems, such as swallowing difficulties. In order to predict the progression of symptoms and to plan the intervention adequately, it is most important to collect systematic data using valid and reliable measures. These will also be presented and discussed.

Speech and communication problems of patients with ALS

ALS affects both lower and upper motor neurons (Somer, 2006). The typical symptoms of lower motor neuron lesion are fasciculation, muscle atrophy and weakness, while upper motor neuron lesions typically result in hyperactive reflexes, and slow and spastic movements. Because ALS represents a mixed upper and lower motor neuron disease, the most common motor speech problem of patients with ALS is mixed flaccid-spastic dysarthria (Duffy, 1995; Yorkston *et al.*, 1995; Freed, 2000). Patients with lower motor neuron involvement will demonstrate flaccid dysarthria, and those with predominantly upper motor neuron involvement will show spastic dysarthria. At the early stages of the disease, the type of dysarthria may be either flaccid or spastic, but as the disease progresses, patients with ALS demonstrate the typical flaccid-spastic mixed dysarthria. Dysarthria is one of the first symptoms of ALS in about 25–30 of the patients (Yorkston *et al.*, 1993; Duffy, 1995; Freed, 2000).

Dysarthria influences all levels of speech production: phonation, respiration, articulation, resonance and prosody. The deteriorations proceed individually (e.g. Mulligan *et al.*, 1994), and the variation in the quality of speech problems is large in adults with ALS even at the time the diagnosis is confirmed (Yorkston *et al.*, 1993). In a case reported by Watts and Vanryckeghem (2001), dysarthria progressed rapidly and resulted in the decrease of sentence intelligibility from 98 % to 10 % during the six months follow-up. Mulligan *et al.* (1994) have reported word intelligibility decrease from 95 % to 88 % during a six months period.

ALS-patients with dysarthria show slower diadochokinetic rates than non-dysarthric ALS-patient (Mulligan *et al.*, 1994). Changes in the accuracy of oral movements and the slowing of production rates tend to precede the decrease of speech intelligibility (Yorkston *et al.*, 1993). However, even when speech rate (pauses included in total time of speaking) declines, speech may remain understandable as long as articulation rate (speech time without pauses) is not affected; the slowing down of articulatory movements and the increased number and duration of pauses result in unavoidable decrease of speech intelligibility (Nishio, 2000).

Communicative effectiveness varies across social situations also in patients with ALS. It is not surprising that patients with ALS and their family members evaluate that the speakers with ALS are more effective in quiet environments and least effective in a noisy environment, when speaking before a group and when speaking for a long period of time (Ball *et al.*, 2004b).

As the ALS progresses, dysarthria becomes more and more severe (e.g. Kent *et al.*, 1991; Watts & Vanryckeghem, 2001), and finally speech will no more serve as a functional tool of communication. Thus, sooner or later, patients with ALS become dependent on augmentative and alternative means of communication (AAC) (Yorkston *et al.*, 1993). Without an access to AAC means the patient with ALS is left without the possibility to communicate efficiently and to be treated as a communication partner. This may lead to reduced possibilities to participate in social interactions. However, people with ALS still need to take part in conversation, ask questions and express their wishes.

Speech therapy for patients with ALS

There is no cure for ALS, and often the speech function deteriorates very quickly, especially in the bulbar type of ALS (Watts & Vanryckeghem, 2001). Typically, there is not much time for the speech therapist to find out proper ways of communication that match the communicative needs of the client. Thus, it is important to start the intervention as early as possible.

Speech therapy aims at helping adults with ALS to maintain their active communicator role. Speech therapy should focus on the maintenance of functional communication rather than on trying to reduce the speech impairment (Yorkston *et al.*, 1995). Because ALS is a progressive disease without cure, it is unrealistic to expect that traditional articulatory or voice exercises can halt the progression of dysarthria. However, in clinical work, we have noticed that motor speech and voice exercises may temporally improve speech intelligibility and phonation time in some patients who are willing to train their speech on daily basis.

The unavoidable changes in speech may be compensated for by using different strategies (Yorkston *et al.*, 1993). These include behavioral intervention and environmental education, such as decreasing the speaking rate or maintaining a slow rate, repeating oneself, limiting the complexity and length of messages, and reducing the loudness of speech or avoiding noisy communication settings. When the intelligibility of natural speech is starting to decline, speech must be combined with augmentative approaches to communication.

Frequent monitoring of speech and early introduction of alternative and augmentative communication means are essential in ensuring that decisions about the AAC-related technology for people with ALS are based on communication preferences rather than critical need (Ball *et al.*, 2004a). Early intervention, including information about communication in general and about the sophisticated solutions of AAC technology, enhances right timing in the process of adopting new means of communication.

In selecting the most functional means of AAC, it is important to take into account some neuropsychological and physical premises. Mild cognitive changes (Bak & Hodges, 2001) or dementia proper (Brooks, 1999; Bak & Hodges, 2001; Pascuzzi, 2002) are rare co-morbidities with ALS, but the general cognitive status of the patient needs to be evaluated in decision making. Physical skills, such as the ability to walk, and occupational capabilities, such as preserved hand mobility, have an effect on the type of AAC technology the patient can use (Yorkston *et al.*, 1993). While someone needs light portable AAC means, the other may need more complicated technology

that will be used in a wheelchair or even in bed. The process of estimating the patient's needs and adjusting the technical options continues till late stages of the disease.

Aims of the study

The clinical work as speech and language therapists has motivated us to study communication and swallowing problems of adults with ALS. As the deteriorations proceed individually, it's difficult to know how rapidly communication and swallowing become impaired. In order to be able to plan the speech and language therapy process adequately, it is most important to know if there are some specific factors predicting the course of symptoms in these patients.

The first purpose of this study is to get more information about how communication and swallowing problems proceed in adults with ALS. The second purpose is to find out what kind of AAC means adults with ALS can use, and if these means are efficient. In addition, this study aims to describe changes in swallowing functions, the need to modify food and liquid viscosity, and other ways of maintaining adequate nutritional intake. The final purpose of this study is to find out specific factors that may predict the progression pattern – or patterns – of communication difficulty and swallowing disorder.

The research questions of this follow-up study are as follows:

1. *How do the symptoms of cranial nerve lesions progress in patients with ALS?*
2. *How does the ability to communicate change in patients with ALS?*
3. *How do the swallowing problems progress in patients with ALS?*
4. *What factors predict the changes in communication and swallowing functions?*

Methods

Participants

Participants in this study will be 20—30 adult patients with the diagnosis of probable ALS or ALS definitive. Patients with other diseases that affect speech, language, swallowing or hearing are excluded from this study. Participants should be native Finnish speakers.

Data collection

The research data will accumulate during clinical management of patients with ALS at the Department of Neurology and rehabilitation of the Tampere University Hospital. The follow-up starts at the first appointment with the speech therapists in the neurological department, and the participants will visit the department with the frequency of every 3-4 months for patients with bulbar symptoms and every 3-6 months for patients with spinal symptoms. The follow-up for each patient will last about two years.

Several variables are to be measured in this study. As there is no standard method for the assessment of speech and communication skills in patients with ALS, we chose to look at the symptom progression from four different angles: physiological, speech production, speech intelligibility and communication.

Physiological changes are followed by using oral motor tasks and measuring maximum phonation time. The oral motor tasks assess the functions of tongue (cranial nerve XII), soft palate (cn X and IX), lips (cn VII) and jaw (cn V) on a four-point scale (normal function, mild disorder, moderate disorder, and severe disorder). Maximum phonation time (in seconds) is measured from audiotape recorded while the patient is producing /a/ in an easy manner for as long as possible.

Speech samples will also be audio-recorded at each visit for monitoring speech production and intelligibility. The tasks will include story telling based on cartoon-frames (see Korpijaakko-Huuhka & Aulanko, 1994), sentence and word production (adopted from Speech Examination; Keller, 1990; the Finnish version by Werner *et al.*, 1994) and diadochokinetic tasks. Speech and articulation rates (syllables per minute) are measured from the story telling and sentence production tasks. For the measurement of the diadochokinetic rate (syllables and syllable sequences in 5

seconds) the patient is instructed to repetitively produce, in a single breath, the Finnish monosyllables /pa/, /ta/ and /ka/ as well as syllable sequences /pataka/ and /tapaka/ as quickly and accurately as possible. In addition, the severity of the speech disorder is assessed using the ALS Severity Scale of Speech (Yorkston *et al.*, 1993).

Speech intelligibility will be perceptually evaluated by a group of listeners. Spontaneous speech (story-telling) of the participants with ALS will be played for the listeners, who evaluate its general intelligibility with a severity scale. The listeners will also hear single sentences and words read by the patients with ALS; the percentage of correctly perceived word will serve as the second estimate of speech intelligibility.

Communicative ability will be evaluated at each visit by classifying the need and usage of means of augmentative communication (Yorkston *et al.*, 1993). Modified CETI (Ball *et al.*, 2004b) will be used to determine the communicative effectiveness of each of the communication tools used in different social situations.

The schedule of the study

The methods of data collection and analyses are going to be specified during the summer 2007.

After that, the Ethics committee of the Tampere University Hospital will be asked for a statement to launch the study.

Data collection is beginning in autumn 2007. Since about 10 new patients with ALS per year are expected to participate in the study, the data is expected to have accumulated for analyses till the end of the year 2011. The first results will report cranial nerve symptoms, communication and swallowing problems of participants at the time of diagnosis. This data is expected to be ready for analyses at end of the year 2009.

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