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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., 2002; Ball, 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

signs of bulbar deterioration at the first SLP visit; (4) no other diseases affecting speech, language or swallowing; and (5) diagnosed with definitive or probable ALS during the study. Of the 17 participants not meeting the inclusion criteria, ten had another diagnosed disease or condition affecting speech, language or swallowing like stroke or Alzheimer's disease, three were excluded due to death before the diagnosis of probable or definitive ALS, and four declined to participate. The level of diagnostic certainty and the initial type of ALS (bulbar, spinal or mixed) were determined by an experienced neurologist (the third author). This study is a part of a larger ongoing project at Tampere University Hospital aiming to predict the progression of the disease within a two-year follow-up. The study protocol was approved by the Ethics Committee of Pirkanmaa Hospital District. All participants gave their written informed consent to participate in the study.

The final group of 30 participants was divided into two according to the level of diagnostic certainty. The first group consisted of 14 participants, five male and nine female, diagnosed with probable or definitive ALS during the first speech assessment (G1; the diagnosed ALS group). The remaining 16 participants, five male and 11 female, with suspected or possible ALS, were assigned to another group (G2; the possible ALS group). All participants in G2 were diagnosed with definitive or probable ALS within two years.

Table 1 presents the participants data. No statistical differences between the groups were found according to gender (Fisher's exact test, $p = 1.0$) or age (Mann–Whitney U test, $p = 0.383$). The most typical initial type of ALS was spinal in G1 and bulbar in G2. This difference was statistically significant (Fisher's exact test, $p = 0.014$). The time between the first visit to a neurologist until the time of diagnosis of probable or definitive ALS varied, being significantly longer in G2 ($M = 12.8$; range 3–38 months) than in G1 ($M = 2.5$; range 0–9 months) (Mann–Whitney U test, $p = 0.000$). The initial speech and communication examination was conducted approximately one month earlier in G2 ($M = 4.4$; range 0–12 months) than in G1 ($M = 3.5$; range 0–30 months), although not statistically significant (Mann–Whitney U test, $p = 0.052$). The participants in G2 had noticed their first bulbar symptoms approximately 5.8 months (range –19 to 20) before the first visit to the

Table 1
Participants data.

Subject code	Sex	Age	Diagnostic certainty of ALS at first speech evaluation	Initial symptoms of ALS	Time interval (months) from the first neurological evaluation to			ALSSS
					definitive/probable dg	first speech evaluation	first bulbar symptoms	
1	M	54	definitive	spinal	4	12	5	8
2	F	52	definitive	spinal	3	8	8	8
3	F	78	definitive	spinal	9	9	9	10
4	F	71	probable	bulbar	0	0	–11	6
5	F	68	definitive	spinal	2	4	4	9
6	F	65	definitive	bulbar	0	1	–9	3
7	F	73	definitive	spinal	1	1	–3	7
8	F	65	definitive	spinal	5	6	4	8
9	M	49	definitive	spinal	4	4	7	10
10	F	71	definitive	spinal	2	4	4	9
11	M	72	probable	mixed	1	3	0	9
12	M	67	probable	spinal	2	3	–2	8
13	M	31	probable	spinal	0	5	3	9
14	F	83	probable	spinal	2	2	2	10
15	F	57	suspected	bulbar	38	4	–12	7
16	F	76	suspected	bulbar	10	0	–17	4
17	M	41	possible	spinal	8	2	–12	9
18	F	59	suspected	bulbar	4	1	–19	5
19	F	68	suspected	spinal	12	5	3	8
20	F	61	suspected	mixed	14	0	–6	7
21	M	58	possible	spinal	11	6	6	8
22	M	51	possible	mixed	3	2	–3	6
23	M	73	suspected	bulbar	13	0	–7	7
24	F	72	possible	mixed	6	1	–11	5
25	F	59	suspected	bulbar	18	1	–3	5
26	M	56	suspected	bulbar	12	0	–7	7
27	F	44	possible	bulbar	6	4	–5	5
28	F	74	suspected	bulbar	13	0	–10	4
29	F	62	possible	bulbar	6	0	–10	6
30	F	59	possible	spinal	31	30	20	6

Note 1: ALS, amyotrophic lateral sclerosis; F, female; M, male; dg, diagnosis. ALSSS = ALS severity scale of speech (Hillel et al., 1989; Yorkston et al., 2004).

Note 2: In column 'first bulbar symptoms' the minus refers to months before the first visit to a neurologist

neurologist, while in G1 it was on average 1.5 months (range –11 to 9) after the first visit (Mann-Whitney *U* test, $p=0.006$, see Note 2 under Table 1).

2.2. Speech and communication assessment

The 30 participants underwent a thorough clinical speech and communication examination by an experienced licensed SLP (the first author). The speech tasks were recorded with a headset microphone and saved as WAV files directly with Sound Forge software.

The neuromuscular basis of dysarthria was examined with motor tasks to reveal any cranial nerve associative dysfunction (Appendix A). Using these tasks, the strength, range and direction of the movements of tongue, soft palate, lips and jaw were assessed on a four-point scale (3 = normal function, 2 = mild, 1 = moderate and 0 = severe disorder). Evidence for fasciculation and atrophy of the tongue was observed during the tasks and at rest on a similar four-point scale. The maximum total score of 42 points (for tongue 18, soft palate 3, lips 9, and jaw 12) indicated normal function of all tasks. Laryngeal efficiency and respiratory capacity were assessed with a maximum phonation task (Kent, 2009). The best of three attempts was reported in the evaluation. To evaluate breath support for phonation, the participants were asked to count quickly from one to twenty as clearly as possible. The quantity of numbers produced during one exhalation was counted.

To estimate the speed of speech movements, the total time used to complete the counting task (see above) was measured in seconds. In diadochokinetic (DDK) tasks, the participants were instructed to repetitively produce the Finnish consonant-vowel monosyllable/ka/ and also the trisyllabic sequence/pataka/as quickly and accurately as possible for five seconds. The DDK rate was reported as syllables per second. To test the reliability of the scoring, two licensed SLPs assessed the oral motor functions and DDK tasks of six of the participants. The interjudge reliability between them was high both for oral motor tasks (ICC = 0.918) and DDK tasks (ICC = 0.984). Finally, to estimate the speed of articulatory movements in spontaneous speech, the rate was measured from a spoken narrative in syllables per total narration time and reported as syllables per second. The participants were asked to generate a story based on a nine-frame cartoon (Korpjaakko-Huuhka & Aulanko, 1994) and stories were transcribed orthographically for speech rate analysis. For two speakers, the speech rate could not be counted because the syllables of their speech were not detectable. To test the reliability of the speech rate assessment, an SLP student and an experienced licensed SLP counted the speech rate of six persons during the story-generation task. The interjudge reliability between the SLP and the SLP student was again high (ICC = 0.998).

The severity of the speech disorder was evaluated using the ALS Severity Scale of Speech (ALSSS) (Hillel et al., 1989; Yorkston et al., 2004) by the SLP and each participant. The rating was based on cumulative data during the speech assessment by the SLP, while the participants were advised to choose the best option from a grading list on the severity scale at the end of the speech assessment (1 = loss of useful speech; nonvocal, 2 = vocalizes for emotional expression, 3 = limits speech to one-word response, 4 = speech plus augmentative communication, 5 = frequent repeating required, 6 = repeats messages on occasion, 7 = obvious speech abnormalities, 8 = perceived speech changes, 9 = nominal speech abnormality, 10 = normal speech). As the interjudge reliability between the SLP and participants was high (intraclass correlation; ICC = 0.968), SLP scores were utilized in the analyses.

The communication modes used by the participants were listed according to self-report. Participants estimated the frequency of use of each mode (e.g. speaking, writing by hand, using a device) on a 3-point scale (always, often and seldom). If they reported more than one mode, the participants ranked the primary and the secondary mode of communication.

The effectiveness of communication in ten different social contexts was defined by the participants using the modified Communication Effectiveness Index (CETI-M) by Ball et al. (2004). Each context of communication was assessed separately on a seven-point scale (1 = not at all effective to 7 = very effective). The total score of CETI-M was computed as the mean rating in communication effectiveness of the evaluated social contexts.

2.3. Statistical analysis

The mean values, ranges and standard deviations (SD) were used as descriptive statistics. Since the parameters were not normally distributed and the sample sizes were small, nonparametric tests were chosen. In the group comparison, the Mann-Whitney *U* test was used for continuous and interval variables and Fisher's exact test was used for categorical variables. The statistical significance level was set at $p < 0.05$. All reported *p*-values are based on two-tailed tests.

Table 2

Oral motor functions and speech production in groups G1 and G2 (N = 30).

	group G1					group G2					Mann-Whitney <i>U</i> test p-value
	n	mean	range	SD	median	n	mean	range	SD	median	
Tongue	14	13.0	1–18	5.0	14.5	16	11.5	1–17	3.9	11.0	0.136
Soft palate	14	2.7	0–3	0.8	3.0	16	2.1	1–3	0.9	2.0	0.015 ⁺
Lip	14	7.6	3–9	1.9	8.5	16	7.1	5–9	1.4	7.0	0.209
Jaw	14	10.4	4–12	2.3	11.5	16	10.7	8–12	1.5	11.5	0.982
Oral motor total	14	33.8	8–42	9.4	36.5	16	31.4	16–41	6.5	31.5	0.123
Max phonation time in second	14	15.6	5–31	7.5	16.0	16	17.5	6–32	6.3	17.5	0.465
Number/one exhalation	14	16.1	3–20	5.0	17.5	16	12.0	1–20	6.6	11.5	0.100
Time to count 1–20 in second	14	11.0	6–26	5.0	10.0	16	21.3	8–47	12.0	17.0	0.002 ⁺
/ka/	14	4.5	1.4–7.4	1.6	4.8	16	3.0	1.4–6.2	1.3	3.1	0.008 ⁺
/pataka/	13	4.7	2.4–7.2	1.4	4.8	16	3.4	1.2–6	1.4	3.3	0.025 ⁺
Speech rate	13	3.4	2.2–5.3	0.9	3.3	15	2.5	1.2–4.5	0.9	2.5	0.029 ⁺

G1, probable or definitive ALS group; G2, suspected or possible ALS group.

⁺ p < 0.05.

3. Results

3.1. Impaired oral motor functions and speech production

The maximum score of oral motor assessment was 42 points (i.e. normal function) and in all 14 tasks there were participants in both groups who scored below normal functional level 3. There were no statistically significant differences in the total score of oral motor assessment between the groups (Mann-Whitney *U* test, $p = 0.123$, Table 2). The statistical difference of sub-scores (tongue, soft palate, lips and jaw) between the groups was significant only for soft palate functions (Mann-Whitney *U* test, $p = 0.015$).

Furthermore, the speech production of the participants in G2 was significantly slower than in G1 in all four tasks: time to count 1–20, /ta/, /pataka/ and speech rate (Mann-Whitney *U* test, $p < 0.05$, Table 2). All participants were able to perform the /ka/ task and the counting speech task. The /pataka/ task was overwhelming for one person in G1. Speech rate could not be counted for one participant in each group.

3.2. Activity limitations in communication

The mean score of the ALSSS for G2 was 6.19 (range 4–9, SD 1.471), while for G1 it was 8.14 (range 3–10, SD 1.875). The speech disorders based on the ALSSS of G2 were significantly more severe than G1 (Mann-Whitney *U* test, $p = 0.002$; Table 1).

Most of the persons with ALS (27/30) communicated with speech and used no other modes of communication. One participant in G1 communicated mainly by writing, and occasionally by using one-word oral responses. Two

Table 3

Self-evaluated effectiveness of speech communication using CETI-M.

CETI-M contexts	Diagnosed group (G1)				Possible group (G2)				Mann-Whitney <i>U</i> test p-value
	n	mean	range	SD	n	mean	range	SD	
1	14	6.1	4–7	1.17	15	5.5	2–7	1.60	0.252
2	13	5.9	4–7	1.14	15	5.2	2–7	1.94	0.555
3	13	6.4	5–7	0.77	15	5.3	3–7	1.63	0.130
4	11	6.2	4–7	1.25	9	5.8	3–7	1.56	0.656
5	13	6.0	4–7	1.29	15	5.0	2–7	1.89	0.170
6	13	5.3	3–7	1.55	15	5.1	2–7	1.92	0.964
7	13	5.3	2–7	1.84	15	4.9	1–7	2.09	0.650
8	14	5.1	3–7	1.64	15	4.5	1–7	2.10	0.561
9	13	5.5	2–7	1.66	15	4.7	1–7	2.13	0.316
10	12	5.2	1–7	1.99	14	4.5	1–7	1.94	0.274

CETI-M contexts: 1. Familiar persons, in a quiet place; 2. Strangers, in a quiet place; 3. Familiar person on the phone; 4. Speaking with young children; 5. Strangers, on the phone; 6. Speaking while traveling in car; 7. Speaking at a distance; 8. In a noisy environment; 9. Speaking before a group and 10. Lengthy conversation (>1 h). Scores 1–7, 1 = not at all effective to 7 = very effective.

participants in G2 sometimes used writing to enhance communication. None of them used more than two modes of communication.

3.3. Communication participation restrictions

Twenty-nine of thirty participants self-evaluated the effectiveness of their speech communication and 18 of them were able to evaluate all social situations of the CETI-M. Eleven participants, five in G1 and six in G2, could not evaluate all proposed social contexts as some were irrelevant to their present life (usually one or two contexts, but seven contexts in the case of one participant). There were no statistically significant differences in the mean score of CETI-M in any of the contexts between the two groups (Mann-Whitney *U* test, $p > 0.05$, Table 3). Eleven participants self-rated their communication effectiveness as decreased (mean score of CETI-M < 5); five in G1 and six in G2. Three participants in G1 and one in G2 self-rated their speech communication as very effective ($= 7$) in all ten social situations. In both groups, decline in communicative effectiveness was reported in demanding social contexts (contexts 6–10 in CETI-M; Table 3).

4. Discussion

At their first visit to an SLP, the persons with possible ALS showed more severe impairments and activity limitations than persons with diagnosed ALS. Significant differences were found in soft palate function and speech rate (impairments of body function) and also in the severity of dysarthria based on the ALSSS (activity limitation). Over 30% of the participants rated their communication as non-effective (participation restriction). These results support our clinical experience that some people with ALS have significant speech disorders before they receive a definitive diagnosis. In this study, our findings cannot be explained by other conditions affecting speech because of both strict exclusion criteria and the fact that all diagnoses of suspected or possible ALS were later confirmed as probable or definitive ALS. However, most of the bulbar-onset ALS cases were included in G2, i.e. possible ALS, underlining the impact of diagnostic delay in this disease-onset group and delayed access to rehabilitation professionals including SLP.

The onset of the disease was bulbar in about one third (11/30) of our participants, reflecting the typical distribution of spinal versus bulbar types of onset (Kiernan et al., 2011). Female participants outnumbered males in both study groups. Women with bulbar-onset ALS reportedly have more severe speech problems and therefore may seek treatment earlier than men with both types of onset or women with spinal-onset (Yorkston et al., 1993). This may explain the majority of females in our study.

The two groups differed significantly in only one oral motor dimension, namely soft palate function. People with possible ALS showed more velopharyngeal dysfunction than did people with diagnosed ALS. Deterioration of velopharyngeal function causes nasality, one of the most affected phonetic features in ALS (Kent et al., 1990, 1992). Thus, although decline in tongue function is considered an important factor in incipient ALS, there may be a group of people with ALS whose first and predominant symptom is impaired soft palate function. The fact that most persons participating in this study had cranial nerve deficits was not unexpected because clinical signs of bulbar symptoms were set as inclusion criteria. In both groups, the most impaired function was tongue function. This finding is in accordance with literature reporting tongue dysfunction as a typical early symptom and an overall common symptom in ALS (Hanson et al., 2011; Hillel & Miller, 1987; Kent et al., 1992; Langmore & Lehman, 1994; Yunusova et al., 2008).

Speech rates were significantly slower for people with possible than with diagnosed ALS. Mean /ka/ syllable repetition rate in people with possible ALS was nearly 1.5 syllables/second lower than normal values reported in older English-speaking controls (Kent, 2009). The spontaneous speech of people with possible ALS was on average one syllable/second slower than that of healthy Finnish-speaking adults performing the same story-generation task (Moore, 1991). The fact that speech rates may decline significantly before the confirmation of ALS diagnosis underlines the importance of early speech assessment. Establishing the baseline and regular monitoring of speaking rates is considered essential in the timing of AAC assessment and intervention (Hanson et al., 2011).

In addition, apart from small sub-groups, the participants with possible ALS ($n = 16$) showed significantly more severe dysarthria than participants in the diagnosed group ($n = 14$). Most importantly, when looking at the second aspect of activity limitations, three of the 30 participants could no longer communicate using speech alone at their first visit to an SLP; two of them had not yet probable/definite ALS diagnosis. Regrettably, information concerning AAC had not reached these participants, who were no longer able to communicate via speech alone.

When looking at communication in the context of social participation, only four participants self-rated their communication with speech as very effective in all ten social contexts of CETI-M, whereas one third of the participants (11/29) reported that their effectiveness of speech communication had been decreased (mean CETI-M value < 5). These 11 participants reported that their communication effectiveness had already been decreased in social contexts of normal daily living at the time of their first speech therapy intervention. In both groups communication with speech was hardest in demanding contexts such as in noisy environment and in long conversations and best perceived with a familiar person in a

quiet environment or over the phone and when communicating with young children. These findings are compatible with the earlier report by Ball et al. (2004).

The differences between the two groups in this study most probably result from differences in the initial onset symptoms. For most participants in the diagnosed group G1, the disease had started with spinal symptoms (11/14). In group G2 with possible ALS the onset was more heterogeneous, although the most typical initial type of ALS was bulbar (9/16). As the majority of persons with bulbar-onset ALS were in G2, our result confirms the notion that people with bulbar-onset are more likely to experience a delayed diagnosis. This is particularly problematic as speech in bulbar-onset ALS is also more impacted early on than in spinal-onset ALS. The mean time from the first perceived bulbar symptoms to the first speech assessment was over three times longer in the possible ALS group than in the diagnosed ALS group. Participants in the diagnosed G1 group were referred to SLP assessment sooner after the first bulbar symptoms than those in group G2. One explanation is that participants in the diagnosed group were already being treated by a multi-disciplinary team and they had most likely been informed of speech difficulties related to ALS. The time between the first visit to the neurologist and the diagnosis was, on average, five times longer in the possible group than in the diagnosed group. This underscores the protracted nature of the diagnostic process among people with bulbar-onset ALS. As bulbar symptoms have been reported to negatively affect survival (Chio et al., 2009; Zoccolella et al., 2008), and as the disease progresses while the person waits for final diagnosis, we consider it unreasonable to delay intervention and scientific study until ALS diagnosis is probable or definitive.

Assessment by an SLP produces multidimensional and important information on speech production processes, severity, and type of speech disorders. This specialist is an important member of the treatment team for ALS. Multidisciplinary specialists' care appears to improve both quality of life and life expectancy (Jenkins, Hollinger, & McDermott, 2014; Nageshwaran, Davies, Rafi, & Radunović, 2014). The methods used in this study are consistent with dysarthria assessment in general and specifically for the evaluation of people with ALS (Ball et al., 2004; Duffy, 2013; Kent, 2009; Yorkston et al., 1993), and they may facilitate the diagnostic process. Furthermore, although our sample of participants was relatively small (N = 30) these results support the notion that communication assessment is an important intervention during the diagnostic process if the person shows clinical signs of bulbar involvement.

5. Conclusion

The participants in this study with possible ALS who ultimately were diagnosed with this disease experienced more speech and communication deterioration than those with confirmed ALS. Challenges associated with both diagnostic uncertainty and the waiting time for the beginning of speech therapy intervention are particularly relevant to persons with bulbar-onset ALS. SLP assessment supports the person's communicative independence, and provides potential early research data on speech and communication changes, when bulbar symptoms are recognized early and even before the definite diagnosis.

Conflict of interest

The first author received financial support from the University of Tampere. The third author has received funding for a congress and speakers' fees from Abbvie, Lundbeck, Medtronic and Orion Pharma. The other authors have no conflicts of interest. There are no non-financial relationships to disclose.

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Appendix A.

EVALUATION OF ORAL MOTOR FUNCTIONS:

Tanja Makkonen, 2007

Patient's name: _____ Identity number: _____

Date of examination: _____ dg: _____ Researcher: _____

Cranial nerve	Task	Functional level (3=normal functions, 2=mild disorder, 1=moderate disorder, 0=severe disorder)	result
XII	Tongue protrusion	3=fluent repeated tongue protrusion with good range of motion 2=laborious tongue protrusion out from the mouth 1=laborious tongue protrusion to the lips 0=minimal tongue protrusion / no protrusion	
XII	Tip of tongue movement	3=fluent repeated tip of tongue movement from upper lip to lower lip with good range of motion 2=laborious tip of tongue movement to the upper lip, decreased vertical range of motion 1=tip of tongue movement only up to the alveolar ridge 0=no tip of tongue movement above the horizontal level	
XII	Lateral tongue movement	3=fluent lateral tongue movement from molar to molar 2=good lateral tongue movement from cheek to cheek 1=lateral tongue movement from one corner of the lips to the other 0=minimal lateral tongue movement / no movement	
X, XII	Tongue base movement	3=fluent tongue base movement up to the palate 2=laborious tongue base movement up to the palate 1=incomplete tongue base movement 0=minimal tongue base movement / no movement	
XII	Fasciculation and atrophy of tongue	3=no fasciculation or atrophy 2=fasciculation, no atrophy 1=both fasciculation and lateral atrophy 0=atrophied tongue	
XII	Tongue strength against resistance	3=good strength both in tongue protrusion and lateral movement 2=diminished strength of tongue either in protrusion or during lateral movement 1=diminished strength of tongue movements 0=minimal or no strength of tongue	
V, X, XI	Soft palate closure	3=good velar closure during repeated /ah/s, and sustained closure during vowel prolongation (/a:/) 2=good velar closure during single /ah/s or closure not sustained during vowel prolongation 1=incomplete or asymmetrical movement of soft palate 0=minimal velar movement / no movement	
VII	Lip rounding (pucker)	3=good lip rounding 2=laborious or asymmetrical lip rounding 1=incomplete lip rounding 0=minimal lip rounding / no movement	
VII	lip spreading (smile)	3=wide and symmetrical smile 2=laborious or asymmetrical smile 1=incomplete smile 0=minimal or no smile	
VII	Lip strength (cheeks puff out)	3=good puff out of the cheeks and sustained lip seal with intraoral pressure for over 15 seconds 2=puff out of the cheeks for no more than 10 seconds 1=puff out of the cheeks for only a few seconds 0=no puff out because of minimal or no lip seal	
V	Vertical mandible movement	3=fluent repeated mouth opening and closing with good range of motion 2=laborious mouth opening or skewed vertical movement 1=incomplete mouth opening 0=minimal mouth opening / no movement	
V	Lateral mandible movement	3=fluent lateral movement of the mandible with good range of motion 2=laborious lateral movement of the mandible with good range of motion 1=incomplete range of lateral movement of the mandible 0=minimal lateral movement / no lateral movement	
V	Anterior mandible movement	3=fluent anterior movement of mandible with good range of motion 2=laborious anterior movement of the mandible with good range of motion 1=incomplete range of anterior movement 0=minimal anterior movement / no anterior movement	
V	Mandible strength against resistance	3 =good vertical movement during mouth opening and good vertical bite (palpate masseter) 2=weakened strength of mandible during mouth opening or vertical bite 1=decreased strength of mandible 0=minimal or no strength of mandible	
	Sensory problems:	Yes (abnormal sensation) No (no problem of sensation)	
Scores		tongue:	/18
		soft palate:	/ 3
		lips:	/ 9
		mandible:	/ 12
		Total:	/42

References

- Ball, L. J., Willis, A., Beukelman, D. R., & Pattee, G. L. (2001). A protocol for identification of early bulbar signs in amyotrophic lateral sclerosis. *Journal of the Neurological Sciences*, 191, 43–53.
- Ball, L. J., Beukelman, D. R., & Pattee, G. L. (2002). Timing of speech deterioration in people with amyotrophic lateral sclerosis. *Journal of Medical Speech-Language Pathology*, 10, 231–235.
- Ball, L. J., Beukelman, D. R., & Pattee, G. L. (2004). Communication effectiveness of individuals with amyotrophic lateral sclerosis. *Journal of Communication Disorders*, 37, 197–215.
- Brooks, B. R., Miller, R. G., Swash, M., Munsat, T. L., & World Federation of Neurology Research Group on Motor Neuron Diseases (2000). El Escorial revisited: revised criteria for the diagnosis of amyotrophic lateral sclerosis. *Amyotrophic Lateral Sclerosis & Other Motor Neuron Disorders*, 1, 293–299.
- Chio, A., Logroscino, G., Hardiman, O., Swigler, R., Mitchell, D., Beghi, E., et al. (2009). Prognostic factors in ALS: a critical review. *Amyotrophic Lateral Sclerosis*, 10, 310–323.
- Duffy, J. R. (2013). *Motor speech disorders: substrates, differential diagnosis, and management*, 3rd ed. St. Louis: Elsevier Mosby.
- Hanson, E. K., Yorkston, K. M., & Britton, D. (2011). Dysarthria in amyotrophic lateral sclerosis: a systematic review of characteristics, speech treatment, and augmentative and alternative communication options. *Journal of Medical Speech-Language Pathology*, 19, 12–30.
- Hillel, A. D., & Miller, R. M. (1987). Management of bulbar symptoms in amyotrophic lateral sclerosis. *Advances in Experimental Medicine & Biology*, 209, 201–221.
- Hillel, A. D., Miller, R. M., Yorkston, K., McDonald, E., Norris, F. H., & Konikow, N. (1989). Amyotrophic lateral sclerosis severity scale. *Neuroepidemiology*, 8, 142–150.
- International classification of functioning, disability and health: ICF (2001). Geneva: World Health Organization.
- Jenkins, T. M., Hollinger, H., & McDermott, C. J. (2014). The Evidence for symptomatic treatments in amyotrophic lateral sclerosis. *Current Opinion in Neurology*, 27, 524–531.
- Körner, S., Siniawski, M., Kollwe, K., Rath, K. J., Krampf, K., Zapf, A., et al. (2013). Speech therapy and communication device: impact on quality of life and mood in patients with amyotrophic lateral sclerosis. *Amyotrophic Lateral Sclerosis and Frontotemporal Degeneration*, 14, 20–25.
- Kent, R. D. (2009). Perceptual sensorimotor speech examination for motor speech disorders. In M. R. McNeil (Ed.), *Clinical management of sensorimotor speech disorders* (pp. 19–29). 2nd ed. New York: Thieme.
- Kent, R. D., Kent, J. F., Weismer, G., Sufit, R. L., Rosenbek, J. C., Martin, R. E., et al. (1990). Impairment of speech intelligibility in men with amyotrophic lateral sclerosis. *Journal of Speech & Hearing Disorders*, 55, 721–728.
- Kent, R. D., Sufit, R. L., Rosenbek, J. C., Kent, J. F., Weismer, G., Martin, R. E., et al. (1991). Speech deterioration in amyotrophic lateral sclerosis: a case study. *Journal of Speech & Hearing Research*, 34, 1269–1275.
- Kent, J. F., Kent, R. D., Rosenbek, J. C., Weismer, G., Martin, R., Sufit, R., et al. (1992). Quantitative description of the dysarthria in women with amyotrophic lateral sclerosis. *Journal of Speech & Hearing Research*, 35, 723–733.
- Kiernan, M. C., Vucic, S., Cheah, B. C., Turner, M. R., Eisen, A., Hardiman, O., et al. (2011). Amyotrophic lateral sclerosis. *Lancet*, 377, 942–955.
- Korpiaakko-Huuhka, A., & Aulanko, R. (1994). Auditory and acoustic analysis of prosody in clinical evaluation of narrative speech. In R. Aulanko, & A.-M. Korpiaakko-Huuhka (Eds.), *Proceedings of the third congress of the International Clinical Phonetics and Linguistics Association* (pp. 91–98). Helsinki: Publications of the Department of Phonetics 39.
- Kuruvilla, M. S., Green, J. R., Yunusova, Y., Hanford, K., Smith, A., & Liss, J. (2012). Spatiotemporal coupling of the tongue in amyotrophic lateral sclerosis. *Journal of Speech, Language & Hearing Research*, 55, 1897–1909. [http://dx.doi.org/10.1044/1092-4388\(2012/11-0259\)](http://dx.doi.org/10.1044/1092-4388(2012/11-0259)).
- Langmore, S. E., & Lehman, M. E. (1994). Physiologic deficits in the orofacial system underlying dysarthria in amyotrophic lateral sclerosis. *Journal of Speech & Hearing Research*, 37, 28–37.
- Moore, K. (1991). Speech rate, phonation rate, and pauses in cartoon and sport narrations. In R. Aulanko, & M. Leiwo (Eds.), *Studies in logopedics and phonetics* 2 (pp. 135–143). University of Helsinki.
- Mulligan, M., Carpenter, J., Riddell, J., Delaney, M. K., Badger, G., Krusinski, P., et al. (1994). Intelligibility and the acoustic characteristics of speech in amyotrophic lateral sclerosis (ALS). *Journal of Speech & Hearing Research*, 37, 496–503.
- Nageshwaran, S., Davies, L. M., Rafi, I., & Radunović, A. (2014). Motor neuron disease. *The BMJ*, 349, g4052. <http://dx.doi.org/10.1136/bmj.g4052>.
- Nishio, M., & Niimi, S. (2000). Changes over time in dysarthric patients with amyotrophic lateral sclerosis (ALS): a study of changes in speaking rate and maximum repetition rate (MRR). *Clinical Linguistics & Phonetics*, 14, 485–497.
- Turner, G. S., & Weismer, G. (1993). Characteristics of speaking rate in the dysarthria associated with amyotrophic lateral sclerosis. *Journal of Speech & Hearing Research*, 36, 1134–1144.
- Turner, M. R., Scaber, J., Goodfellow, J. A., Lord, M. E., Marsden, R., & Talbot, K. (2010). The diagnostic pathway and prognosis in bulbar-onset amyotrophic lateral sclerosis. *Journal of the Neurological Sciences*, 294, 81–85.
- Watts, C. R., & Vanryckeghem, M. (2001). Laryngeal dysfunction in amyotrophic lateral sclerosis: a review and case report. *BMC Ear, Nose, and Throat Disorders*, 1, 1.
- Williams, J. R., Fitzhenry, D., Grant, L., & Martyn, D. (2013). Diagnosis pathway for patients with amyotrophic lateral sclerosis: retrospective analysis of the US Medicare longitudinal claims database. *BMC Neurology*, 13, 160. <http://dx.doi.org/10.1186/1471-2377-13-160>.
- Yorkston, K. M., Strand, E., Miller, R., Hillel, A., & Smith, K. (1993). Speech deterioration in amyotrophic lateral sclerosis: implications for the timing of intervention. *Journal of Medical Speech-Language Pathology*, 1, 35–46.
- Yorkston, K. M., Miller, R. M., & Strand, E. A. (2004). *Management of speech and swallowing disorders in degenerative diseases*, 2nd ed. Austin: Pro-Ed.
- Yunusova, Y., Weismer, G., Westbury, J. R., & Lindstrom, M. J. (2008). Articulatory movements during vowels in speakers with dysarthria and healthy controls. *Journal of Speech Language & Hearing Research*, 51, 596–611.
- Yunusova, Y., Green, J. R., Greenwood, L., Wang, J., Pattee, G. L., & Zinman, L. (2012). Tongue movements and their acoustic consequences in amyotrophic lateral sclerosis. *Folia Phoniatrica et Logopedica*, 64, 94–102.
- Zoccollella, S., Beghi, E., Palagano, G., Fraddosio, A., Guerra, V., Samarelli, V., et al. (2008). Analysis of survival and prognostic factors in amyotrophic lateral sclerosis: a population based study. *Journal of Neurology, Neurosurgery & Psychiatry*, 79, 33–37.