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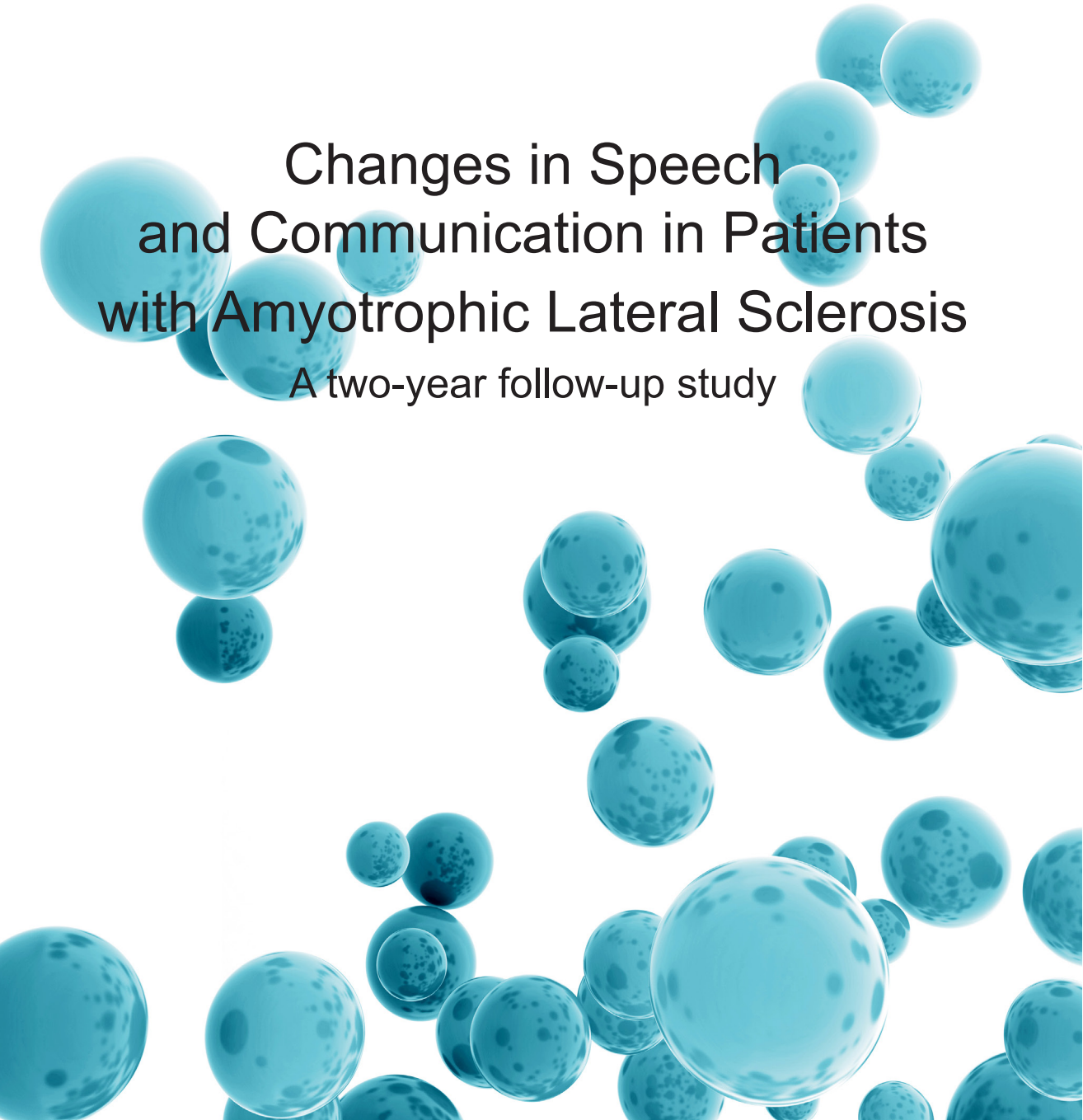
Changes in Speech and Communication in Patients with Amyotrophic Lateral Sclerosis

A two-year follow-up study



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ACADEMIC DISSERTATION
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To communication of all kinds

ABSTRACT

Motor speech disorders are common in patients with amyotrophic lateral sclerosis (ALS). The majority of ALS related research has concentrated on speech and communication symptoms and their progression only in patients with confirmed diagnosis. However, many individuals with ALS already have severe motor speech disorders and might have lost the ability to speak before any definite diagnosis is made. In clinical work, speech therapy interventions should be optimally-timed based on the patients' communicative needs to ensure that they have the opportunity to communicate effectively.

The focus of this thesis was to explore lower cranial nerve symptoms, speech disorders and communicative effectiveness, first at baseline which was set at the first visit to a speech-language pathologist (SLP) and then over a period of two years to identify the changes occurring in communication as defined by the components impairment, activity and participation in the International Classification of Functioning, Disability and Health (ICF). A secondary purpose was to identify whether the certainty level of diagnosis or the initial type of ALS (bulbar- or spinal-onset) affected the aforementioned aspects of speech and communication.

Altogether 30 consecutive patients underwent thorough speech and communication examinations during the follow-up consisting of 157 clinical assessments by SLP and 148 self-evaluations of communication effectiveness. The clinical assessment included an evaluation of motor speech functions, speed of speech movements, intelligibility and the severity of the speech disorder. Additionally, the patients' functional capacity to communicate, the need for augmentative and alternative communication (AAC) and the amount and type of used communication strategies were evaluated. The patients' self-rated communication effectiveness both with natural speech and the AAC means used. Both qualitative and quantitative methods were used to analyze the data.

ALS patients displayed varying degrees of speech deterioration and utilized differing communication means in their first visit to the SLP, but impairments and activity limitations were more severe in those patients without a definite diagnosis. In addition, the delay from bulbar symptom onset to the definite ALS diagnosis was longer and the speech therapy intervention was initiated later in these patients

without a definite diagnosis. In the majority of these patients, the initial symptoms had appeared in the bulbar area.

As expected, speech and communication abilities as well as self-rated effectiveness of natural speech declined in nearly all patients as the disease progressed. The severity of speech disorders deteriorated to a poor level on average within 18 months from the appearance of the first bulbar symptom in 60% of the patients, while 40% of patients maintained the ability to speak adequately. A total of 57% of the patients communicated with at least one low- or high-technology AAC strategy either to support or substitute for their impaired speech as the disease progresses. In over 60% of the patients functional capacity of communication deteriorated, evidencing the need for AAC in the first place or a change in the ability to utilize these AAC strategies. On the whole, bulbar-onset patients experienced more severe speech and communications difficulties than spinal-onset patients during the whole follow-up.

The use of multiple AAC means improved the effectiveness of their communication. Self-rated effectiveness of communication was better in these patients who used alternative communication strategies in comparison with those who communicated primarily with natural speech in the last year of the study period.

In clinical work, it is important to take the initial type of ALS into consideration as bulbar-onset patients are more susceptible to delays in diagnosis, speech therapy assessment and the provision of AAC services. Furthermore, the manifestation of the first bulbar symptoms predicted a loss of adequate speech more accurately than diagnostic certainty or the time of the first speech therapy evaluation. Regular speech therapy interventions initiated very soon after the appearance of symptoms can help to provide patients, their families and other health care professionals with detailed information about bulbar symptoms and speech and communication changes, and can ensure that patients have prompt access to optimal AAC strategies on time. In order to maintain the patients' communicative effectiveness, their individual abilities and preferred social contexts should also be taken into account.

TIIVISTELMÄ

Amyotrofiseen lateraaliskleroosiin (ALS) liittyy usein motorisia puhehäiriöitä. Useissa tutkimuksissa puheen ja kommunikoinnin häiriötä sekä niiden muutoksia on tutkittu potilailla, joiden ALS diagnoosi on jo varmistunut. Monilla ALS-potilailla on selkeitä puhemotoriikan vaikeuksia ennen diagnoosin varmistumista osan menettäessä puhekyvyn diagnosointiprosessin aikana. Kliinisessä työssä tulisi tavoitella oikea-aikaisia puheterapiainterventioita, jotta potilaiden kyky kommunikoida tehokkaasti ja viestintätarpeiden mukaisesti pystyttäisiin varmistamaan.

Tämän väitöskirjatutkimuksen tavoitteena oli tutkia alimpien aivohermojen oireita, puheentuoton vaikeutta sekä kommunikoinnin tehokkuutta lähtötilanteessa, joksi asetettiin ensimmäinen puheterapia-arvio sekä seurata kommunikoinnin muutoksia kahden vuoden ajan kansainvälisen toimintakykyluokituksen (International Classification of Functioning, Disability and Health (ICF)) toimintakyky ja rajoitteet osa-alueen näkökulmasta. Lisäksi tavoitteena oli selvittää vaikuttaako diagnoosin varmuusaste tai sairauden alkamismuoto (bulbaari- tai spinaalialkuinen) puheentuottoon ja kommunikointiin.

Tutkimukseen osallistui peräkkäisotannalla 30 ALS-potilasta, joilla oli vähintään yksi bulbaarioire. Aineisto koostuu 157 puheterapeutin tekemästä puheen ja kommunikoinnin arviosta sekä 148 kommunikoinnin tehokkuuden itsearvioinnista. Puheterapia-arvioinnit sisälsivät puheen motorisen tuoton, puhenopeuden, puheen ymmärrettävyyden sekä motorisen puhehäiriön vaikeusasteen arvioinnit. Lisäksi kartoitettiin toiminnallinen kyky kommunikoida, tarve kommunikointia tukeville ja korvaaville (AAC) keinoille sekä käytössä olevat AAC-keinot. Potilaat itsearvioivat kommunikoinnin tehokkuuden sekä puheella että mahdollisilla AAC-keinoilla. Aineiston analysoinnissa käytettiin sekä määrällisiä että laadullisia menetelmiä.

Alkuarviossa tutkittavilla esiintyi eriasteisia motorisen puheentuoton oireita, kolmella potilaalla oli jo käytössä AAC-keino heikentyvän puheen tukena. Puheentuoton ja kommunikoinnin häiriöt olivat vaikeampiasteisia potilailla, joiden ALS-diagnoosi ei ollut vielä varmentunut lähtötilanteessa. Näillä potilailla myös aika bulbaarioireiden ilmenemisestä puheterapiaintervention toteutumiseen sekä

diagnostiikan varmistumiseen oli pitempi. Suurimmalla osalla näistä potilasta sairaus oli alkanut bulbaarioireilla.

Puheen ja kommunikoinnin vaikeudet lisääntyivät sekä puheen tehokkuus heikentyi oletetusti sairauden edetessä lähes kaikilla potilailla. Motorinen puhehäiriö muuttui vaikea-asteiseksi 18 kk:n kuluessa bulbaarioireiden alkamisesta 60%:lla potilaista, kun taas 40%:lla potilaista säilyi kyky kommunikoida adekvaatisti puheella. Vähintään yhtä manuaalista tai tekniikkaa hyödyntävää AAC-keinoa käytti 57%:a tutkittavista. Yli 60%:lla tutkittavista toiminnallinen kyky kommunikoida heikentyi seurannan aikana. Toiminnallisen kommunikointikyvyn muutos ilmeni joko AAC-keinojen tarpeena tai heikentyvänä kykynä hyödyntää erilaisia AAC-keinoja. Potilailla, joiden sairaus alkoi bulbaarioireilla esiintyi vaikeampia puheen ja kommunikoinnin häiriöitä kuin niillä potilailla, joiden sairaus alkoi spinaalioireilla.

Monipuolinen kommunikoinnin apuvälineiden käyttö lisäsi kommunikoinnin tehokkuutta. AAC-välineitä hyödyntävät potilaat arvioivat kommunikointinsa tehokkaammaksi kuin ensisijaisesti puheella kommunikoivat potilaat.

Kliinisessä työssä on tärkeää huomioida ALS-taudin alkamismuoto. Niillä potilailla, joiden sairaus alkaa bulbaarioireilla, diagnoosin varmistuminen, ensimmäisen puheterapia-arvion toteutuminen ja kommunikoinnin apuvälinepalveluprosessin aloittaminen kestää pidempään bulbaarioireiden alkamisajankohtaan verraten. Bulbaarioireiden alkamisajankohta ennustaa puhekyvyn menettämisen todennäköisesti täsmällisemmin kuin diagnoosin varmistumisajankohta tai ensimmäisen puheterapiaintervention toteutumis-ajankohta. Säännöllisten puheterapia-interventioiden toteutuminen pian bulbaarioireiden ilmenemisen jälkeen antaa potilaille, heidän läheisilleen ja terveydenhuollon ammattihenkilöille tarkkaa tietoa bulbaarioireista sekä puhekyvyn ja kommunikoinnin muutoksesta sekä mahdollistaa oikea-aikaisen apuvälinepalveluprosessin. Potilaan toimintakyky sekä hänelle tärkeät sosiaaliset kommunikointitilanteet tulee huomioida ylläpidettäessä potilaan kommunikoinnin tehokkuutta.

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LIST OF ORIGINAL PUBLICATIONS

The original publications will be referred to in the text by Roman numerals I- IV. Author's contribution I, III & IV: The author was responsible for the data collection and analyses as well as writing the manuscript. II: The author was responsible for the data collection, analyses together with the fourth author and was responsible for writing the manuscript.

I Tanja Makkonen, Anna-Maija Korpijaakko-Huuhka, Hanna Ruottinen, Riitta Puhto, Kirsi Hollo, Aarne Ylinen & Johanna Palmio. Oral motor functions, speech and communication before a definitive diagnosis of amyotrophic lateral sclerosis. Journal of Communication Disorders, 2016; 61:97–105.

http://doi.org/10.1016/j.jcomdis.2016.04.

II Tanja Makkonen, Hanna Ruottinen, Riitta Puhto, Mika Helminen & Johanna Palmio. Speech deterioration in amyotrophic lateral sclerosis (ALS) after manifestation of bulbar symptoms. International Journal of Language & Communication Disorders, 2018; 53:385-392.

https://doi.org/10.1111/1460-6984.12357.

III Tanja Makkonen, Hanna Ruottinen, Anna-Maija Korpijaakko-Huuhka & Johanna Palmio. Variation in communication strategies in amyotrophic lateral sclerosis during a two-year follow-up. Speech, Language and Hearing, 2017; 21:123-130.

https://doi.org/10.1080/2050571X.2017.1362719

IV Tanja Makkonen, Hanna Ruottinen & Johanna Palmio. Self-reported communication effectiveness of persons with amyotrophic lateral sclerosis. Submitted/Under review.

ABBREVIATIONS

AAC	Augmentative and alternative communication
ALS	Amyotrophic lateral sclerosis
ALSFRS	The Amyotrophic Lateral Sclerosis Functional Rating Scale
ALSSS	The Amyotrophic Lateral Sclerosis Severity Scale
AMR	Alternating motion rate
CN	Cranial nerve
DDK	Diadochokinesis
CETI	The Communication Effectiveness Index
CETI-M	The Modified Communication Effectiveness Index
CNS	Central nervous system
EBP	Evidence based practice
EFNS	European Federation of Neurological Societies
ENMG	Electroneuromyography
FCC	Functional capacity of communication
FTD	Frontotemporal dementia
ICC	Intraclass correlation
ICF	International Classification of Functioning, Disability and Health
LME	Linear mixed-effect
LMN	Lower motor neuron
MPT	Maximum phonation time
MRI	Magnetic resonance imaging
NIV	Non-invasive ventilation
PEG	Percutaneous gastrostomy
PPA	Primary progressive aphasia
Q1	The first quartile
Q3	The third quartile
SD	Standard deviation
SLP	Speech-language pathologist
SMR	Sequential motion rate

SGD	Speech-generating device
TTSD	Text-to-speech device
UMN	Upper motor neuron
VAS	Visual analogy scale
WAV	Waveform Audio File Format

1 INTRODUCTION

Amyotrophic lateral sclerosis (ALS) is an incurable neurodegenerative disease affecting primarily motor neurons in the brain and spinal cord (Kiernan et al., 2011). This progressive motor deficit causes dysfunctions in movement and overall physical performance and in some patients also in cognition and behavior (van Es et al., 2017). Classical ALS leads to death within 3-4 years after the symptom-onset (Couratier et al., 2016; van Es et al., 2017). The recommended treatment is based on the management of symptoms with care being provided in multidisciplinary special clinics (EFNS Task Force on Diagnosis and Management of Amyotrophic Lateral Sclerosis, 2012;). Currently the only disease modifying medication in use is able to extend the patients' survival by 3-6 months (Miller et al., 2007; Bensimon et al., 1994; Lacomblez et al., 1996).

The most common communication difficulties in ALS results from dysarthria, which occurs in 80-95% of ALS patients (Beukelman et al., 2011; Creer et al., 2016; Tomik & Guilloff, 2010). The progressive speech disorder may impact on the patients' quality of life and have destructive psychological and social outcomes (Felgoise et al., 2015; Simmons, 2005).

Evidence based practice (EBP) consists of three basic principles: 1) research evidence, 2) clinical expertise and 3) well-informed patient's opinions (Dollaghan 2004; Rosenberg & Donald, 1995). Materialization of EBP requires functional communication. During the disease, ALS patients need to make many decisions related to care, treatment and aid services. It is important to support and promote a person's possibilities to discuss with professionals and family members about these difficult decisions. Person-centered care, where the person is an active partner in his/her care and treatment is not possible without functional communication (Ekman et al., 2011; Mead & Bower 2000).

At present, there has been little research published about speech and communication changes prior to a definite ALS diagnosis, with only occasional follow-up studies on speech and communication deterioration (Hanson et al., 2011). More information is needed in order to comprehend the quality and changes in bulbar symptoms, as well as the deterioration in speech and communication if we

are to promote ALS patients' communicative autonomy and help the patients and their caregivers to navigate the disease pathway.

The purpose of this study was to evaluate cranial nerve symptoms, speech disorders and communicative effectiveness at different levels of diagnostic certainty as well as investigating the kinds of changes that will occur in patients' speech, their ability to communicate and the communication strategies that they have applied and also to assess the effectiveness of the patients' communication strategies during a two-year follow-up.

2 REVIEW OF THE LITERATURE

2.1 Amyotrophic lateral sclerosis

Amyotrophic lateral sclerosis (ALS) is the most common motor neuron disease, it is also known as Charcot's disease after the French doctor who discovered the condition in 1869 or Lou Gehrig's disease after the American baseball player who was diagnosed with it in the 1930s (Goetz, 2000). ALS is a progressive multisystem degeneration affecting both upper motor neurons (UMN) and lower motor neurons (LMN) in the cerebral cortex, brainstem and spinal cord, causing muscle weakness and atrophy (van Es et al., 2017). It is believed that pathological changes probably begin long before the manifestation of clinical symptoms as in other neurodegenerative disorders and therefore a specific time of disease onset is difficult to determine (Eisen et al., 2014). The disease usually first appears locally, but symptoms eventually spread to encompass all voluntary muscles, including the respiratory muscles. The main independent risk factors are male gender, increasing age and a family history of ALS (Alonso et al., 2009; Haverkamp et al., 1995; Scott et al., 2009). The incidence of the disease is 2.16/100 000 person years in Europe (Lagroscino et al., 2010). The worldwide incidence is 1.90/100 000 with a prevalence of 4.48/100 000 person years (Chiò et al., 2013). In the whole of Finland, the prevalence was 3.56/100 000 in 1973 (Jokelainen 1977); in Middle-Finland Central Hospital District between 1976 and 1981 it was 6.4/100 000 (Murros & Fogelholm, 1983). Between 1963 and 1995, the ALS mortality rate in Finland increased from 0.91 to 2.75/100 000 (Jokelainen 1976; Maasilta et al., 2001).

The majority of the people diagnosed with ALS are classified as having the sporadic disease and only about 5-10% have familial ALS, although there are reports of higher frequencies i.e.16-20% in a population-based register in Ireland (Byrne et al., 2011; Byrne et al., 2013). The etiology of the disease is unknown, but variants in about 50 genes have been associated with ALS (Boylan 2015). C9orf72 is the gene most commonly mutated in ALS (Dejesus-Herdandez et al., 2011; Renton et al., 2011). The prevalence of the C90rf72 is high in Finland: it has been identified in 21.1% of the patients with sporadic ALS and 46.4% with familial ALS in the country (Majounie et al., 2012; Renton et al., 2011). It has been even speculated that the

C9orf72 repeat expansion first occurred in the Finnish population and spread with the Vikings to other parts of Europe (Pliner et al., 2014). Other globally common gene mutations in ALS in global terms are SOD1, TARDBP and FUS (Zou Z-Y et al., 2017). In Finland, the most common gene mutations are SOD1 and C90rf72, and these two mutations together explain nearly 90% of the cases of familial ALS in Finland (Renton et al., 2011).

Earlier ALS was thought to affect mostly the lower and upper motor neurons, but in the past decade behavioral changes and the wider involvement of the central nervous system (CNS) have been increasingly recognized among the patients. Both cognitive and behavioral declines are encountered in as many as every second individual with ALS and about 14% of patients develop FTD (Miller et al., 2009; Murphy et al., 2016; Phukan et al., 2007). In particular, a mutation in gene C9orf72 has been associated with FTD among ALS patients (Dejesus-Herdandez et al., 2011; Renton et al., 2011). The following other clinical phenotypes have been associated with C9orf72 expansions in small numbers of patients i.e. increased incidence of parkinsonism in ALS patients, olivopontocerebellar degeneration, Huntington disease-like syndrome, corticobasal syndrome and Alzheimer's disease (Cooper-Knock et al., 2014).

Riluzole is the only disease-modifying medication for patients with ALS, extending the survival of the patients by 3–6 months (Bensimon et al., 1994; Lacomblez et al., 1996; Miller et al., 2007). However, a recent retrospective study reported that the benefit of riluzole seemed to be more evident in the late stage of the ALS, and the drug did not seem to prolong the functional life time (Fang et al., 2018). Both non-invasive ventilation (NIV) and percutaneous gastrostomy (PEG) prolong survival in ALS (Burkhardt et al., 2017) and the American Academy of Neurology and also the European Federation of Neurological Societies (EFNS) guidelines recommend them as treatment options in ALS (EFNS Task Force on Diagnosis and Management of Amyotrophic Lateral Sclerosis, 2012; Miller et al., 2009a). The good practice management of ALS involves the treatment of a wide variety of symptoms such as weakness and disability, dysphagia, dyspnea and poor cough, pain, dysarthria, cognitive changes, sialorrhoea, thickened saliva, emotional lability, depression and anxiety, sleep disturbance and constipation (Andersen et al., 2007). Multidisciplinary care in special clinics may extend survival, decrease medical complications and improve the quality of life (EFNS Task Force on Diagnosis and Management of Amyotrophic Lateral Sclerosis, 2012, Miller et al. 2009b).

2.1.1 Clinical presentation

ALS is a heterogenic syndrome and phenotypes occur in the so-called motor neuron disease – FTD continuum. Classical ALS with both LMN and UMN signs is one extreme on the spectrum with FTD representing the other extreme. Classical ALS is the most common phenotype (about 70%) whereas about 5-15% patients display the ALS-FTD phenotype meeting the diagnostic criteria of both ALS and FTD.

Common UMN signs involve muscle weakness, primitive reflexes, pseudobulbar affect, hyperreflexia, hypertonia and spasticity, while LMN manifestations include muscle weakness, fasciculation, atrophy, hyporeflexia and weakness. In about two thirds of the ALS patients, the disease starts with spinal symptoms and in the remain third with bulbar symptoms. The initial sign of the spinal-onset ALS is typically a progressive muscle weakness in the hand, arm, leg or foot. Dysarthria is the most common clinical characteristic in bulbar-onset disease, while dysphagia usually develops later or sometimes simultaneously. (van Es et al., 2017)

FTD usually manifests in two subtypes: behavioral/dysexecutive frontotemporal dementia or primary progressive aphasia (PPA) (Bang et al., 2015). Common clinical features in behavioural-variant FTD are behavioral or social symptoms (like disinhibition, inappropriate or offensive behavior, excessive jocularity, exaggerated emotional displays, loss of empathy, selfishness, personal neglect), emotional symptoms (like apathy, depression) and repetitive or compulsive behaviours (Bang et al., 2015). Primary progressive aphasia is usually divided into three variants: nonfluent, semantic and logopenic (Gorno-Tempini et al., 2011). The core features in the nonfluent variant are agrammatism and effortful, halting speech with apraxia; in the semantic variant, impaired confrontation naming and impaired single-word comprehension and in the locopenic variant, there is impaired single-word retrieval in spontaneous speech and naming and impaired sentence repetition (Gorno-Tempini et al., 2011). More behavioral than primary progressive aphasia variant have been seen among ALS patients (van Es et al., 2017).

Although the majority of clinically apparent communication difficulties in ALS result from dysarthria, both language impairment and cognition changes are possible and evidently impair communication (EFNS Task Force on Diagnosis and Management of Amyotrophic Lateral Sclerosis, 2012; Fried-Oken et al., 2015; Taylor et al., 2013). Language deficits in spelling, grammatical and syntactic processing, and word retrieval have been reported in ALS (Pinto-Grau et al., 2018). Executive dysfunctions, mild linguistic impairment and their combination are present also among non-demented patients with ALS (Taylor et al., 2013).

2.1.2 Diagnostic processes

There are no definitive diagnostic tests or biomarkers for ALS and therefore the diagnosis has to be based on clinical criteria (Brooks et al., 2000). Patients with signs of UMN or LMN degeneration might be suspected of having ALS, but neurological, electrophysiological, neuroimaging and laboratory examination are needed to ascertain these findings and to exclude other diseases. An appropriate neurological examination verifies the clinical findings of UMN and LMN degeneration and their progression (Brooks et al., 2000). Electrophysiological examinations should reveal the LMN degeneration and exclude other disorders. Neuroimaging examinations (like magnetic resonance imaging (MRI)) exclude other disease processes. Laboratory tests can identity possible ALS-related syndromes and determine whether clinical judgement and neuropathological examinations are required to confirm or exclude findings related to sporadic ALS, ALS-related syndromes and ALS-variants. Clinical and electrophysiological examinations may need to be repeated at least six months apart in order to detect the progression of the symptoms(Brooks et al., 2000).

The widely recognized revised El Escorial diagnostic criteria (Brooks et al., 2000) utilize a combination of LMN and UMN symptoms to establish levels of diagnostic certainty: possible, probable and definite ALS. Awaji criteria are used when undertaking neurophysiological studies (de Carvalho et al., 2008). According to the revised El Escorial criteria, a definitive ALS diagnosis requires evidence of LMN degeneration by clinical, electrophysiological or neuropathological examination; evidence of UMN degeneration in the clinical examination; a progressive spread of symptoms or signs within anatomical regions or to other regions as well as no evidence of other disease processes that might explain these symptoms and signs. (Brooks et al., 2000; de Carvalho et al., 2008). The diagnosis is challenging because of the heterogeneity of the clinical presentations and the varying speed of symptom progression (van Es et al., 2017; Kiernan et al., 2011). A recent study from Ireland reported that the patients are usually first attended by general practitioners, who then refer the patients mainly to neurologists or to ear, nose and throat specialists, orthopedic physicians or to physiotherapy (Galvin et al., 2017). The main mimics of motor neuron disease that can hinder the diagnostic processes, are benign fasciculations, multifocal motor neuropathy with conduction block, neuralgic amyotrophy, Kennedy's syndrome (spinobulbar muscular atrophy), motor-predominant chronic inflammatory demyelinating polyradiculoneuropathy, inclusion body myositis, hereditary spastic paraparesis, primary progressive multiple

sclerosis and mixed cervical myeloradiculopathy (Turner & Talbot, 2013). Often the diagnostic process from symptom onset to a definitive ALS diagnosis takes a year or more (Galvin et al., 2017; Paganoni et al., 2014; Turner et al., 2010; Williams et al., 2013) and older age, sporadic disease and limb onset can further delay diagnosis (Paganoni et al., 2014).

2.1.3 Treatment processes

A multidisciplinary specialist team should be available for individuals affected by ALS to optimized diagnostic and symptom management services. Optimally the multidisciplinary team should comprise specialists like neurologist, respiratory physician, gastroenterologist, rehabilitation medicine physician, social counsellor, occupational therapist, speech-language pathologist (SLP), respiratory therapist, specialized nurse, physical therapist, dietitian, psychologist, dentist and palliative care physician. Patients should be followed up every 2-3 months and regular contacts with the patient and relatives between visits are recommended. A less frequent follow-up protocol may be appropriate if the disease is progressing slowly whereas a more frequent review is valid in both the months following diagnosis and in the later stages of disease. The hospital-based multidisciplinary team, the palliative care team, the primary healthcare and community services should enjoy effective mutual communication and co-ordination channels. (EFNS Task Force on Diagnosis and Management of Amyotrophic Lateral Sclerosis, 2012)

2.1.4 Disease progression and survival

Classical ALS is a rapidly and invariably progressing disease, leading to death due to respiratory failure within 3-4 years after symptom onset (Couratier et al., 2016; van Es et al., 2017). In some atypical forms of ALS, the natural progression is slower and 10-20 % of patients may survive for ten years or more (Chiò et al., 2009). Some patients choose to increases survival time with invasive mechanical ventilation (IMV), but the use of NIV is recommended in preference to IMV for patients with respiratory insufficiency (EFNS Task Force on Diagnosis and Management of Amyotrophic Lateral Sclerosis, 2012). IMV increases the family and carer burden (including 24-h nursing requirement) and it is possible that a 'locked-in' stage will develop in the patient treated with IMV.

The progression of the disease symptoms usually spreads within an anatomical region, for example from the hand to the upper arm, and also to other regions, such as from the leg to the arm (Ravits & La Sprada, 2009). Most commonly symptoms spread from one arm to the contralateral arm, from one leg to the contralateral leg or the bulbar region to the arm, but also non-contiguous symptoms development such as from a leg to the bulbar area or an arm to the contralateral leg is not uncommon (Walhout et al., 2018). Among patients with initial bulbar symptoms 34.4% experienced leg symptoms and 46.7% arm symptoms after one year and 51.4% had leg symptoms and 62.1% arm symptoms after two years (Walhout et al., 2018). Bulbar symptom involvement was detected in 30.9% of patients with arm-onset ALS and in 21.3% of those with leg-onset after two years (Walhout et al., 2018).

Different clinical features, treatments, genetics, environmental and life style factors all affect the survival time (van Es et al., 2017). The clinical features associated with short survival are bulbar-onset ALS and overall bulbar dysfunction, poor nutritional status, respiratory-onset ALS, executive dysfunction and comorbid FTD, neck flexor weakness and old age at diagnosis (Chiò et al., 2009; van Es et al., 2017; Williams et al., 2013). Also expansion in C9orf72 and environmental and life style status such as low socioeconomic status and smoking have been associated with a poor prognosis (van Es et al., 2017). NIV and PEG (Burkhardt et al., 2017) and riluzole treatment (Bensimon et al., 1994; Lacomblez et al., 1996; Miller et al., 2007) prolong survival in ALS. A moderate exercise routine and certain clinical features have been associated with longer survival i.e. flail arm variant, LMN-predominant disease, UMN-predominant disease, long time to diagnosis and young age at diagnosis (van Es et al., 2017). In addition, multidisciplinary clinical care associated with a long survival (van Es et al., 2017).

2.2 Motor speech disorders in ALS

Dysarthria is the initial symptom of ALS in about one third of the patients and these initial symptoms have a tendency to remain as the most prominent symptom (van Es et al., 2017; Galvin et al., 2017; Yorkston et al., 1993). Motor speech difficulties occur in 80-95% of patients irrespective of the initial type of ALS at some point of the disease progression, weakening the functionality of natural speech and indicating the need for augmentative and alternative communication (AAC) methods (Beukelman et al., 2011; Creer et al., 2016; Tomik & Guilloff, 2010).

During the early stages of the disease, the type of dysarthria may be either flaccid or spastic. The bulbar LMN involvement will lead to a flaccid dysarthria whereas a bulbar UMN involvement is associated with spastic dysarthria. Patients with classical ALS will typically demonstrate a flaccid-spastic mixed dysarthria as the disease progresses. When the mixed dysarthria appears, either type may be predominant. (Duffy 2013)

In their classical reports, Darley, Aronson and Brown (1969 a, b) described the most deviant speech characteristics of dysarthria in ALS (Table 1). Slow rate, short phrases, distorted vowels and imprecise consonants were more common in the ALS groups in a comparison with another neurologic group (Darley et al., 1969 a, b). In addition to the LMN and UMN aspect, the motor speech productions might also be approached through four speech subsystems: respiratory, phonatory, articulatory and resonatory (Green et al., 2013).

Table 1. Typical flaccid, spastic and combined dysarthria characteristics according to Darley, Aronson and Brown (1989 a,b)

Typical flaccid dysarthria characteristics	Typical spastic dysarthria characteristics	Combined dysarthria characteristics associated with ALS
hypernasality, breathiness, imprecise articulation, audible inspiration/stridor	harshness, distorted vowels, articulatory-resonatory incompetence, imprecise consonants, prosodic insufficiency	prolonged intervals, prolonged phonemes inappropriate silences

Patients with ALS have perceived the potential loss of speech as one of the worst aspects of the disease and named it as the most problematic symptoms of all of the bulbar symptoms (eating, speaking, excessive saliva, and mucus) (Hecht et al., 2002; Raheja et al., 2016). The loss of effective natural speech may exert devastating psychological and social outcomes and it is likely to have a detrimental impact on the overall quality of life in ALS (Felgoise et al., 2015; Simmons, 2005).

2.2.1 Neuromuscular basis of dysarthria

ALS destroys cranial nerves, especially the lower ones, weakening the motor function of tongue, jaw, lips, velum and laryngeal muscles (Hillel et al., 1989; Kuruvilla et al., 2012; Langmore & Lehman 1994; Rong et al., 2016). Dysfunctions of the oral motor muscles such as decreased strength and movement, reduced speed of tongue movement and decreased spatiotemporal coupling between the different regions of the tongue during word production have been reported in many studies (Hillel et al., 1989; Langmore & Lehman, 1994; Kuruvilla et al., 2012; Yunusova et al., 2008; Yunusova et al., 2012). The progressive loss of bulbar muscle strength, control and movement and the increased duration of movements lead to more and more deteriorations in motor speech functions (Shellikeri et al., 2016; Yunusova et al., 2016). Numerous studies have demonstrated that bulbar motor dysfunction occurs prior to any perceived changes in speech intelligibility (DePaul & Brooks 1993; Kent et al., 1990; Mefferd et al., 2012; Nishio & Niimi 2000; Yorkston, et al., 1993). Measures of speech motor performance, especially the speed of tongue movement, may be well suited for early detection of ALS and also for monitoring the disease progress (Green et al., 2013).

In their classical work, Hillel and Miller (1987) have presented the typical physical findings of bulbar ALS based on their experience with over 100 bulbar ALS patients over a period of 10 years. They divided the findings into four groups according to the common temporal progression of the cranial nerve (CN) symptoms (Table 2).

Table 2. Temporal progression of typical cranial nerve symptoms.

	1.group	2. group	3. group	4. group
affected muscles	tongue (CN XII) lips (CN VII)	palatal (CN V, X and XI), masticular and buccal (CN V) pharyngeal constrictors (CN X)	buccal/orbital/frontal (CN VII), sternocleidomastoid/trapezius (CN XI) vocal cords (CN X)	extraocular (CN III, IV and VI)

CN= cranial nerve

Vocal fold dysfunction and changes in voice quality have been encountered among patients with ALS, and also abnormality in acoustic analysis (such as evaluating frequency range and phonatory stability) related to the laryngeal dysfunction have been found before clinically observed voice symptoms or dysarthria (Chen & Garret, 2005; Silbergleit et al., 1997; Tomik et al., 2007: Tomik & Guillof, 2010). The Maximum phonation time (MPT) is a common clinical task evaluating the laryngeal efficiency and respiratory capacity associated with dysarthria (Kent, 2009). Laryngeal efficiency and respiratory capacity are a part of motor speech production, but MPT has not been found to be sensitive in differentiating dysarthric ALS patients from their non-dysarthric counterparts (Mulligan et al., 1994).

2.2.2 Speech and articulation rate

The presence of bulbar symptoms is associated with a progressive reduction in the speech and articulation rates (Green et al., 2013; Turner & Weismer, 1993; Yorkston et al., 1993). A reduced speech rate is also often one of the first symptoms of dysarthria (Kent et al., 1991; Mulligan et al., 1994; Watts & Vanryckeghem, 2001). Not only the speech motor deficit but also respiratory insufficiency and cognitive/language impairment may affect the speech and articulation rate. An increase in the number and duration of pauses and a reduced variability of pauses may also be attributable to respiratory insufficiency (Lee et al., 1993; Yunusova et al., 2016). FTD patients, with either behavioral or non-fluent primary progressive aphasia, have been shown to have an impaired speech rate and FTD patients with non-fluent primary progressive aphasia also to demonstrate articulatory deficits (Yunusova et al., 2016).

The speech rate is a general measure of speech production and it involves both the articulation rate and the pause time. The speech rate has been reported to be slower in bulbar-onset patients than in their spinal-onset counterparts after the ALS diagnosis (Ball et al., 2002). A slower than normal speech rate has been found to precede the decline in speech intelligibility (Ball et al., 2002; Green et al., 2013; Kent et al., 1991; Nishio and Niimi 2000; Rong et al., 2016; Turner and Weismer 1993). It has been claimed that the reduce speech rate is a highly sensitive indicator for detecting ALS, but speech rates alone provide limited insights into the nature of the underlying impairment (Yunusova et al., 2016).

The articulation rate (speech rate excluding pauses) is primarily a measure of speech motor function and the assessment of articulation rate has diagnostic value in detecting speech motor changes (Yunusova et al., 2016). The articulation rate was reported to be impaired in ALS patients with bulbar symptoms and as well as in those with the combination of bulbar and respiratory symptoms (Yunusova et al., 2016). By measuring the articulation rate, it is possible to detect early-onset bulbar symptoms and this can assist in monitoring disease progression (Allison et al., 2017; Green et al., 2013; Yunusova et al., 2016).

Both syllable and syllable sequences rate i.e. the diadochokinesis (DDK), speech rate and articulation rate (speech rate excluding pauses) have been common ways used to evaluate motor speech functions among ALS (Ball et al., 2002; Langmore & Lehman, 1994; Nishio & Niimi, 2000; Turner & Weismer, 1993). Dysarthric patients with ALS have been observed to have a slower syllable sequence repetition rate than patients without any perceivable dysarthria (Mulligan et al., 1994). Further, non-dysarthric patients with ALS have been reported to have slower consonant-vowel syllable repetition than normal speakers (Langmore & Lehman, 1994). Speech intelligibility may remain good even when the maximum repetition rate declines (Nishio & Niimi, 2000).

2.2.3 Intelligibility

Progressive mixed dysarthria leads to the loss of intelligible speech. The bulbar-onset patients were observed to have reduced speech intelligibility compared to the spinal-onset participants after ALS diagnosis (Ball et al., 2002). Even a slightly deteriorated speech intelligibility may weaken participation in verbal communication in real life social situations (Ball et al., 2004b) and indicates a loss of adequate speech (Ball et al., 2004b; Green et al., 2013; Nishio & Niimi 2000; Rong et al., 2013; Yorkston et al., 1993).

The rate of decline in intelligibility varies from one individual to the next. Sentence intelligibility of 66 ALS patients was reported to decrease from 95% to 75% in over a year (Rong et al., 2016). Intelligibility probably remains good even if the noticeable signs of dysarthria occur in the early stages of the disease but it declines rapidly during the later stages of the disease (Ball et al., 2001; Green et al., 2013; Nishio and Niimi 2000; Rong et al., 2016; Yorkston et al., 1993).

Decreased speech intelligibility is a social handicap and is indicative of a need for communication aids and AAC strategies in order to allow the patient to

communicate effectively (Yorkston et al., 1993). The loss of speech intelligibility is a very important issue in the clinical care of ALS. For example, the decisions regarding palliative care and AAC processes have been found to be the most effective when made prior to the loss of speech intelligibility (Ball et al., 2002)

2.2.4 Severity of speech disorder

The ALS severity scale of speech (ALSSS of speech=speech scale) is one entity in a four-scale combination, where the other three scales contains 10 levels of upper and lower extremity functions and swallowing (Hillel et al., 1989; Yorkston et al., 2004). The speech scale is widely used to determine the severity of dysarthria and to assist clinicians in deciding the appropriate timing of interventions. Speech intelligibility is the primary focus of the speech scale. It contains 10 levels of speech function (score 10 = normal speech). According to the study of Yorkston et al. (1993) the speech rate and oral DDK rate declined when speech changes became noticeable to listeners, i.e. the speech score declined from 9 to 8. Speech intelligibility at a sentence level remains good if the speech scores are between 10-7 but intelligibility has declined with scores 6 and below. The most important change in speech intelligibility takes place between scores 5 to 4, this represents a point when augmentative communication means are first needed to help with natural speech communication.

The Amyotrophic Lateral Sclerosis Functional Rating Scale (ALSFRS) and its revised version supplemented with respiratory functions (ALSFRS-R) have been used both in clinical trials and in clinical practice to measure physical function when carrying out an assessment of a patient's daily activities (ALS CNTF Treatment Study Phase I–II Group et al., 1996; Cederbaum & Stambler 1997; Cederbaum et al., 1999). ALSFRS-R contains 12 items, three in all four domains that encompass gross motor functions, fine motor functions, bulbar functions and respiratory functions (Cederbaum et al., 1999). The three bulbar function items are speech, salivation and swallowing. Each item is further divided into five categories with the speech categories including 4=normal speech process, 3= detectable speech disturbance, 2= intelligible with repeating, 1= speech combined with non-vocal communication and 0= loss of useful speech.

2.2.5 Progression in speech disorders

Once bulbar symptoms appear and speech is affected, speech production declines unavoidably but not automatically in a linear fashion; it might change rapidly, but not necessarily steadily (DePaul & Kent, 2000; Mulligan et al., 1994; Nishio & Niimi, 2000; Watts & Vanryckeghem, 2001). In 49 bulbar-onset ALS patients median interval from bulbar symptom onset to anarthria was 18 months (range 0-77) (Turner et al., 2010).

Overall, as the disease progresses with time, dysarthria becomes more severe and the need for communication aids increases (Beukelman et al., 2011; Creer et al., 2016; Turner et al., 2010; Yorkston et al., 1993). The majority of the follow-up studies on the progression of dysarthria have focused on the time period after the definite diagnosis of ALS was made (Hanson et al., 2011) and therefore there is still only limited knowledge about early motor speech disorders in bulbar-onset patients.

2.3 Speech therapy in ALS

Speech therapy assessment in patients with ALS is based on common motor speech and language evaluation as in other etiologies although it should take into account the special features of the disease. There are no standardized diagnostic procedures for assessing the bulbar dysfunction associated with ALS (Green et. al., 2013, Tomik & Guillof, 2010). The widely used features in clinical speech therapy assessments are presented in Table 3. The evaluation should notice early signs of bulbar dysfunction and be precise enough to detect the progression of symptoms.

Multiple evaluation tools are needed; these should be based on experts' subjective judgements and also objective assessment strategies (Green et al., 2013). When attempting to optimize an appropriate speech therapy intervention, it is recommended that assessments of regular speech and language function should be conducted every 3–6 months by a trained SLP (EFNS Task Force on Diagnosis and Management of Amyotrophic Lateral Sclerosis, 2012).

Table 3. Generally used features of clinical speech therapy assessment for ALS (based on Ball et al., 2004b; Green et al., 2013; Yorkston et al., 2004 and clinical expertise)

Motor speech examination		
	Respiratory	Cough force Pauses needed in speech MPT
	Phonatory	MPT Voice quality & loudness
	Articulatory	Tongue Lips Jaw (muscles strength, spread and speed, fasciculation & atrophia) Accuracy and speed of articulation Syllable and syllable sequences DDK rate
	Resonance	Velum (muscular strength, spread and speed) Hypernasality Nasal emission
Speech production		
	Speaking rate	Speaking and/or articulation rate from reading or speaking
	Intelligibility	Word, sentence, speech
Speech severity		For example speech subscale from ALSSS or ALSFRS-R
Communication effectiveness		For example CETI-M
Language impairment		Aphasia tests
Functional capacity of communication		Adequacy of speech and lower and upper extremities
Need for AAC		Conclusion of assessment and progression of symptoms

AAC= Augmentative and Alternative Communication; ALSFRS-R= The Amyotrophic Lateral Sclerosis Functional Rating Scale; ALSSS=The Amyotrophic Lateral Sclerosis Severity Scale; CETI-M= The Modifies Communication Effectiveness Index; DDK= diadochokinesis; MPT= Maximum phonation time

There is a consensus among experts that the AAC process should be started when speech is still understandable (Ball et al., 2004a; Hanson et al., 2011). The focus of the speech therapy intervention is to maintain the ability to communicate by educating patients and their family and caregivers about speech and communication dysfunctions, teaching compensatory strategies for speech production and the exploitation of AAC services (Yorkston, Miller & Strand 2004). The use of appropriate AAC systems should be individualized and relevant training and support provided as required (EFNS Task Force on Diagnosis and Management of Amyotrophic Lateral Sclerosis, 2012).

2.4 Augmentative and alternative communication

The American Speech-Language-Hearing Association (ASHA) has classified AAC as follows: "Augmentative and alternative communication (AAC) is an area of clinical practice that addresses the needs of individuals with significant and complex communication disorders characterized by impairments in speech-language production and/or comprehension, including spoken and written modes of communication. AAC uses a variety of techniques and tools, including picture communication boards, line drawings, speech-generating devices (SGDs), tangible objects, manual signs, gestures, and finger spelling, to help the individual express thoughts, wants and needs, feelings, and ideas." (ASHA, 2018). With the help of AAC, a person with a speech dysfunction should be able to maintain the ability to communicate and this can improve or at least stabilize his/her quality of life (Körner et al., 2013; Londral et al., 2015). In some cases, the use of AAC extends the ability to work (Ball et al., 2004a; Ball et al., 2007a; McNaughton et al., 2001). It has been estimated that the majority (72%) of the persons with ALS gain benefit from AAC as the disease progresses (Creer et al., 2016). A speech-language pathologist (SLP) plays a key role in helping patients and their loved ones to appreciate that alternative communication strategies may be needed to support and substitute for natural speech (Simmons, 2005).

AAC is classified into three categories: (1) high-technology (2) low-technology and (3) non-technology strategies (Beukelman et al., 2007). High-technology AAC strategies refer to speech-generating devices (SGDs) that produce speech output either in the form of prerecorded messages or with text-to-speech programs. SGDs can be designed solely for AAC or they can be integrated with other computer

functions and applications (Fried-Oken et al., 2015). Many communication devices may be used with minimal hand function for example there are head or eye-controlled keyboards or they can be manipulated by different switches. Low-technology AAC strategies include communication methods such as alphabet boards, printed words and sentences, handwriting or drawings. Often both low- and high-technology communication strategies are based on writing ability (for example handwriting, keyboard access, or alphabetic board) or ready-for-use messages (like words, common phrases, pictures, voice messages). Examples of communication with writing are presented in Figure 1. There are also AAC strategies that do not require any technology are for example gestures, vocalization and eye movements or blinks. Technological advances, especially high-technological solutions, are evolving continuously, but there is still a place for less sophisticated low-technology solution.

It is not unusual that an ALS patient will communicate with a combination of AAC-strategies belonging to all three AAC categories (Ball et al., 2007b). Many patients can still use AAC technology during the last weeks of their lives, but there is an increasing tendency to switch to low- or no-technology communication strategies in the late stages of the disease (Ball et al., 2007a). Sometimes issues related to the individual (e.g., desire to use his/her own speech), device (e.g., complex learning requirements) or professional input (e.g., lack of training) affect the patient's decision not to exploit the available AAC strategies (Baxter et al., 2012; Murphy, 2004).

Language impairment may cause the need for AAC or make to use of different AAC strategies more difficult and this needs to be taken into account in the AAC process. Some of the most widely used AAC strategies are based on writing skills, but these might not be options for persons with severe language impairment. For example, cognitive problems might complicate the ability to utilize some forms of AAC and severe cognitive deteriorations might be the reasons behind the patient's difficulties in using technology AAC devices (Geronimo et al., 2016). It is possible that some patients with ALS and cognitive problems cannot utilize any

Figure 1. Examples of communication by writing using the low- or high-tech AAC

communication strategies at all during the final stages of ALS (Brownlee & Bruening, 2012).

In clinical practice there are several alternative means to support or substitute for diminishing natural speech and to combine different communicative methods optimally: 1) a person might primarily communicate with natural speech, but will support communication with one or more AAC means in certain social contexts; 2) a person might primarily adopt AAC methods to communicate, but also use his/hers diminishing speech in certain situations; 3) a person might communicate completely by AAC methods, when speech is no longer functional in any situation; 4) deteriorated speech is functional only in a few communication situations and the speech might be unclear and 5) speech and low- or high-technology solutions are no longer functional. Finally, communication may be based on more or less ambiguous gestures, facial expressions and/or vocalization, which might nonetheless be a valid way to communicate with familiar persons.

2.4.1 Functional capacity of communication

Functional capacity of communication (FCC) is a combination of functional performance of speech and the use of the upper and lower extremities (Hillel et al., 1989; Yorkston et al., 2004; Yorkston et al., 1993). All of these three aspects should be taken into account when evaluating communicative options and selecting the most appropriate AAC means (Yorkston et al., 2004). Persons with poor speech can adequately communicate by supplementing their speech with handwriting and/or typing if hand functions persist. When both speech and hand functions are impaired, SGDs with minimal hand, leg or head movement (such as special keyboards, buttons and joysticks) or even activated only by eye movements (such as eye-tracking communication devices) are needed. If a patient's mobility is good, easy portability is an important aspect of the AAC devices. When mobility declines, AAC strategies should be usable when the patients are wheelchair-bound or bedridden. With poor hand functions and/or with poor mobility, a patient might need assistance in preparing the AAC devices for use or for using SGDs. There appear to be differences in hand function in different forms of ALS; spinal-onset patients have been found to use more partner-dependent AAC strategies than individuals with bulbar-onset (Mathy et al., 2000). Changes in FCC during the natural course of the ALS must be considered when planning an AAC intervention (Fried-Oken et al., 2015).

2.4.2 Communication aids services in Finland

Several Finnish law regulate complemented by quality recommendations form the basis of the aid services (Hurnasti et al., 2010). Communication aids are one part of the medical rehabilitation aid services and these are funded by public special-health care in Finland. The assistive Technology Unit of Tampere University Hospital has the responsibility for organizing technical communication aid services throughout the Pirkanmaa Hospital district, the area where the participants in this study were recruited. The multidisciplinary team of AAC specialists (SLPs, occupational therapist and technical advisor) is responsible for the provision of AAC intervention services. In the supply of medical rehabilitation aids, the Pirkanmaa Hospital district adhere to the national criteria (Pirkanmaa Hospital district, 2017). If the criteria are fulfilled, then the patients is given the aids for loan on a free-of-charge basis. Tampere University Hospital district owns the AAC devices and is responsible for device maintenance and usage follow-up.

2.5 Effectiveness of communication

Declining skills in motor speech have been associated with restrictions of communicative participations among ALS patients (Yorkston et al., 2017). The communicative effectiveness index (CETI) was developed to measure the effectiveness of functional communication. The original CETI was developed for patients with aphasia after a stroke and it contains 16 assessments of social situations in everyday life (Lomas et al., 1989) Each social situation is scored on a 100-mm visual analogue scale (VAS) to measure the effectiveness of a person´s current communication abilities. The CETI was found to be both a reliable and valid indicator for functional communication (Lomas et al., 1989), to be responsive to aphasia rehabilitation (Aftonomos et al., 2001; Bakheit et al., 2005) and also to be suitable for various other languages (Pedersen et al., 2001).

The modified CETI (the CETI-M), is an adaptation of the original CETI made for ALS persons and it focuses on ten social communication situations (Ball et al., 2004b; Yorkston et al., 1999). Patients with ALS and their communication partners participated in the modification process.

The effectiveness of communication as measured with the CETI-M was found to decrease in ALS patients. Communication in demanding social situations that require clear speech strategies such as using a louder voice, precise articulation and

a well-controlled speech rate were less effective than communication in easy daily situations. Effective communication in demanding social situations might become difficult even with a minor decrease in speech intelligibility. There were similar ratings of communication effectiveness made by patients and listeners. (Ball et al., 2004b)

The self-rated communication effectiveness was found to decrease over time as measured by the CETI-M. Low self-perceived communication effectiveness was found to correlate with a lower quality of life and more severe bulbar impairment. (Londral et al., 2015)

2.6 International Classification of Functioning, Disability and Health

The International Classification of Functioning, Disability and Health (ICF) is the World Health Organization's classification of health and health related domains. The ICF organizes information into two parts which again have two components: 1) Functioning and Disability: Body function and Body structure, and Activities and Participation and 2) Contextual factors: Environmental factors and Personal factors. Functioning and Disability with its two components were utilized as a general framework in this study. The interaction between components of Functioning and Disability and health conditions are described in Figure 2. (ICF, 2001)

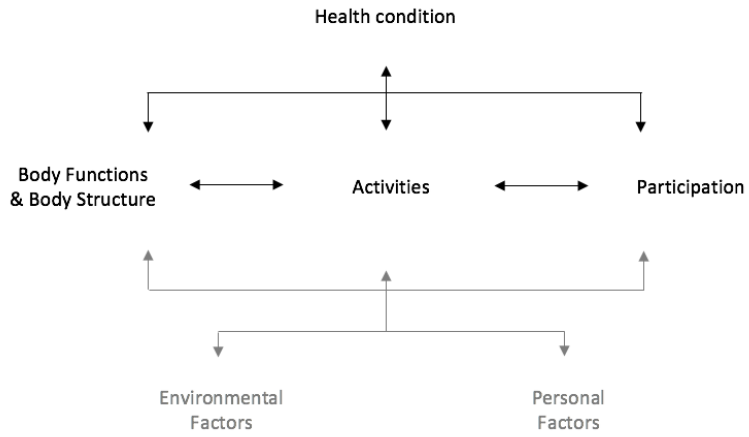

Figure 2. Interaction between ICF components (ICF, 2001). The component part of this thesis is marked in black.

3 AIMS OF THE STUDY

The purpose of the study was to analyze how speech and communication in patients with amyotrophic lateral sclerosis (ALS) change during a two-year follow-up. The more specific aims were:

1. to evaluate cranial nerve symptoms, speech disorders and communicative effectiveness at different levels of diagnostic certainty (I)

2. to determine how long natural speech will remain functional and what changes will occur in speech (II)

3. to study the changes in the ability to communicate and the potential communication strategies used by the patients (III)

4. to investigate how the self-rated effectiveness of communication changes (IV)

4 PARTICIPANTS AND METHODS

4.1 Patients

This study was conducted in the Tampere University Hospital. From August 2007 to December 2009, 47 consecutive patients with suspected or diagnosed ALS were referred to the Department of Neurology and Rehabilitation to the SLP because of one or more signs of bulbar deterioration. The recruitment process of the patients is described in Figure 3. Thirty of the patients provided their written informed consent for participation and met the inclusion criteria: 1) native speakers 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) no other diseases affecting speech, language or swallowing; and 4) diagnosed with definitive or probable ALS according to the revised el Escorial criteria (Brooks et al., 2000) during the study. All diagnoses were made by an experienced neurologist. All patients had normal hearing and adequate vision without or with eye-glasses. Five patients had clinically obvious cognitive difficulties although no formal cognitive assessment was not conducted as a part of the study. This research project and the informed consent forms were approved by the Ethics Committee of Pirkanmaa Hospital District (R07111).

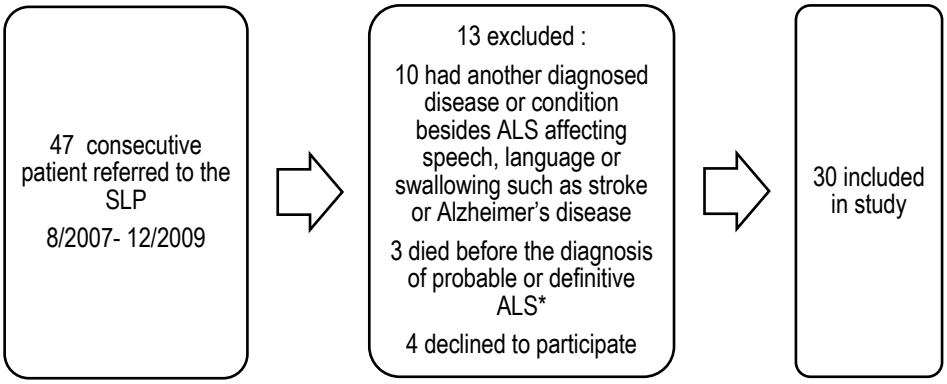

Figure 3. Recruitment process of the patients

ALS=amyotrophic lateral sclerosis; SLP= speech-language pathologist. * All of these three had bulbar-onset symptoms; 2 were male, 1 female; age 67-78.

The characteristics of the patients are presented in Table 4. Of the patients, 20 were female and 10 were male. The mean age was 62.8 (31.1-83.0) years at baseline. The initial type of ALS was bulbar in 11 patients, spinal in 15 patients and mixed in four patients. In studies II and III, four of the patients who had been originally defined as mixed-onset were re-assigned, two into the bulbar-onset and two into the spinal-onset group based on their major initial symptoms. On average, the speech therapy evaluation was performed 5.2 times (1-9) for each patient, and the first SLP visit was on average 4.1 (-34 – 8) months before the final ALS diagnosis was reached (probable or definitive ALS). The mean time from bulbar symptom manifestation to the first SLP visit was 6.5 months (-3 – 22).

Table 4. Characteristics of the participants

	gender	age at baseline	initial type of ALS	bulbar symptom to SLP[1]	neurologist to SLP[2]	speech therapy evaluations in moths (*=exitus)								
						0	3	6	9	12	15	18	21	24
1	f	57	bulbar	16	4	x	x	x	x	x		x		x
2	f	76	bulbar	17	0	x	x	x	x			x	*	
3	m	41	spinal	13	2	x	x		x	x	x	x	*	
4	m	54	spinal	7	12	x	x	x	x		x	x	x	*
5	f	52	spinal	0	8	x	x	x	x		x		x	x
6	f	59	bulbar	15	1	x	x	x	x	x	x	x	*	
7	f	68	spinal	2	5	x	x	x	x	x	*			
8	f	78	spinal	0	9	x	*							
9	f	60	mixed	6	0	x	x	x	x	x	x	x	x	x
10	m	58	spinal	0	6	x		x		x		x		x
11	m	51	mixed	5	2	x	x	*						
12	f	71	bulbar	11	0	x	x	x	x	*				
13	f	68	spinal	0	4	x		x	x	x	x	*		
14	m	73	bulbar	7	0	x	x		x	x	x	*		
15	f	72	mixed	12	1	x	x	x	x	*				
16	f	59	bulbar	4	1	x	x	x	x		x	x	x	x
17	m	56	bulbar	7	0	x		x	x	x	x	x	x	x
18	f	44	bulbar	8	4	x	x	x	x	*				
19	f	65	bulbar	10	1	x		x	*					
20	f	73	spinal	4	1	x	x	x	*					
21	f	65	spinal	2	6	x	x	x	x	x	*			
22	m	49	spinal	-3	4	x	x		x	x	x	x		x
23	f	74	bulbar	10	0	x		x	x	x	*			
24	f	71	spinal	0	4	x	x	x	x	*				
25	m	72	mixed	3	3	x	x	x	*					
26	f	62	bulbar	22	0	x	x	x	x	x		x	x	*
27	m	67	spinal	6	3	x	x	x	*					
28	m	31	spinal	2	5	x		x		x	x	x	x	x
29	f	82	spinal	0	2	x	x	x		x	x	x	x	*
30	f	59	spinal	10	30	x		x		x		x	x	x

ALS= amyotrophic lateral sclerosis; SLP= speech and language pathologist.
[1]= Interval from bulbar symptom onset to the first SLP visit in months; [2]= Interval from the first neurology visit to the first SLP visit[2]

4.2 Data collection

The objective of the study was to monitor each patient for two years. Owing to the natural course of the disease, 21(70%) patients died due to ALS-related respiratory insufficiency during the follow-up: 10 during the first year (3 bulbar, 4 spinal and 3 mixed onset) and 11 during the second year (5 bulbar and 6 spinal onset) (Figure 5.). Nine patients (30%) survived the whole two years; five spinal-onset and four bulbar-onset. The mean follow-up time was 15.3 (0-24) months. None of the patients had undergone invasive ventilation with tracheostomy.

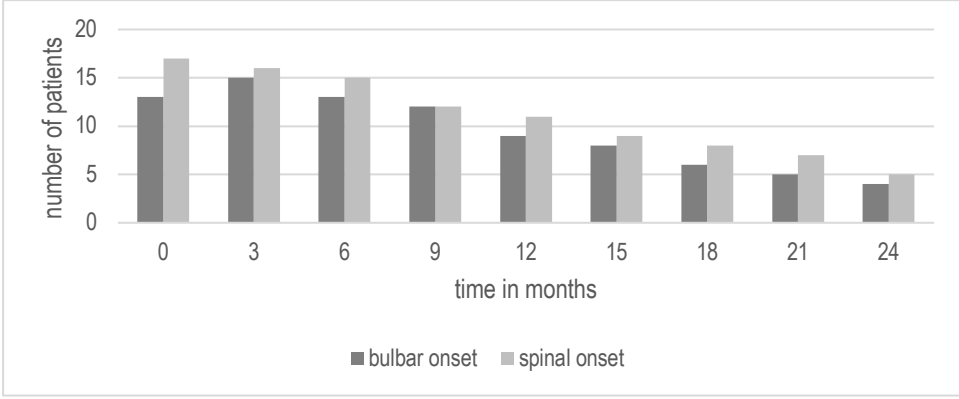

Figure 4. Survival of the participants

The data was collected in a comprehensive speech therapy evaluation at the neurology clinic during clinical visits by an experienced licensed SLP at baseline and on average every at three month intervals according to the patient's clinical needs. The data of speech and communication evaluation was organized into three-month intervals (+/- one month) to monitor the changes with time. If there were two speech therapy evaluations during the same interval (as occurred in 11 cases) the latter assessment was included in the study. The number of speech and communication evaluations and the number of self-rated communication effectiveness score evaluated at each follow-up point are presented in Table 5. A total of 157 clinical speech and communication evaluations including an assessment of functional capacity of communication (FCC) and communication strategies were included in the study.

Each SLP visit contained speech and swallowing evaluation and guidance on speech and communication, AAC and swallowing to patients and their carers (if present). The duration of speech therapy evaluation varied between 0.5-2 hours based on the clinical extensity of the current clinical needs. The evaluation entailed also a swallowing assessment which is not included in the present study.

The author conducted all of the speech therapy evaluations herself. The interjudge reliability of the oral motor and DDK task was made by two licensed SLPs: the author and one other SLP. The interjudge reliability of the speech rate assessment was made by the author and an SLP student. The intelligibility of semi-spontaneous speech from the narrative cartoon task was evaluated perceptually by seven licensed and experienced SLPs (none of which was the author).

Table 5. Number of SLP evaluations and self-ratings

Time in months	0	3	6	9	12	15	18	21	24	total
Number of SLP evaluations	30	22	25	20	16	13	13	9	9	157
Number of CETI-M self-ratings	29	22	25	19	13	10	12	9	9	148

CETI-M= The Modified Communication Effectiveness Index; SLP= speech-language pathologist

A total of 148 self-evaluations of communication effectiveness were performed. Nine self-evaluations could not be utilized: two lacked information and seven were missed because three patients were unable to complete the self-ratings at visits between 9-18 months, these three patients could no longer communicate reliably by speech or with any of the possible AAC means due to insufficient cognition.

4.3 Methods

The methods used in this study were chosen based on the following criteria: 1) clinically valid and commonly used methods to evaluate motor speech disorders, especially in patients with ALS; 2) methods including parameters capable of monitoring both the early and longitudinal changes in motor speech production and

3) methods including tools to measure each component of ICF Functioning and Disability: body function and structure, activity and participation. A summary of all methods is presented in Table 6 categorized according to the ICF components of Functioning and Disability (impairment of body function and structure, activity limitations and participation restrictions). The timeline of the sub-studies is presented in Figure 5.

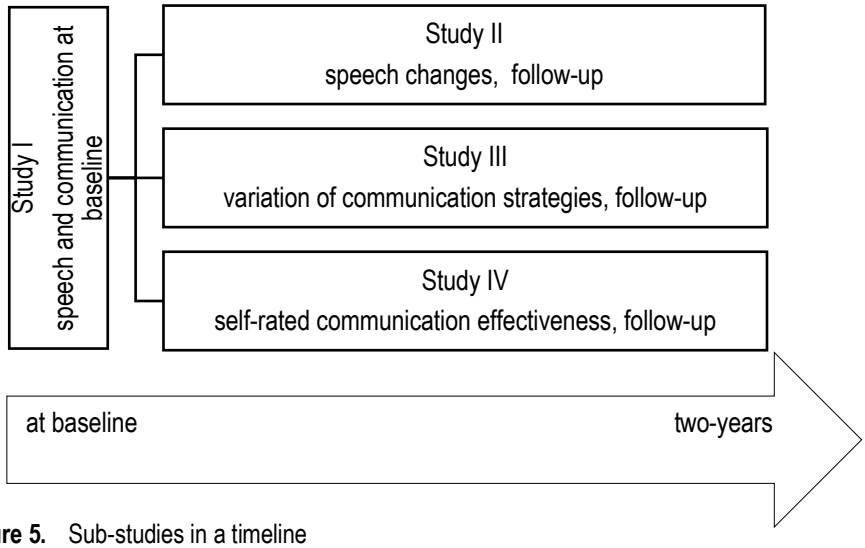

Figure 5. Sub-studies in a timeline

Table 6. Summary of the methods

ICF perspectives	Target	Tasks	Score/evaluation	Used in study
Impairment	cranial nerve function in dysarthria	oral motor tasks	0-42 (0-3 of each 14 items; 3= normal performance)	I
	laryngeal efficiency, respiratory capacity	MPT	the best of 3 attempt in seconds	I
	breath support for speech	count to 1-20	amount of numbers in one exhalation	I
	speech production	AMR in 5 secs :/ka/	syllables/1 second	I
		SMR in 5 secs: /pataka/	syllables/1 second	I
		count to 1-20	total time used to complete the task	I
		story-telling based on a nine-frame cartoon	1) speech rate: total number of syllable per total narration; syllables/1 second	1) I
			2) articulation rate: speech rate when pauses are excluded; syllables/1 second	2) II
Activity limitations	severity of speech disorder	ALS Severity Scale of Speech	1-10 evaluated both by patients and researcher (10= normal speech)	I, II
	intelligibility of semi-spontaneous speech	story-telling based on a nine-frame cartoon	100 mm VAS (0-100) perceptually by seven professional listeners (100=fully intelligible speech)	II
	communication strategies	according to the participants own description,	1) listed communication strategies to used	1) I, III
		conform during the the clinical visits by investigator	2) listed order of communication strategies as primary, secondary…	2) I, III
	Functional capacity of communication	Combination of ALS Severity Scale of speech, upper and lower extremity function	Classified into six different categories	III
Participation restriction	effectiveness of communication	The Modified Communication Effectiveness Index	The mean rating of the evaluated social contexts (1-7 of each context, max 7=very effective)	I, IV

ALS= amyotrophic lateral sclerosis; AMR=alternating motion rate; MPT=maximum phonation time; SMR= sequential motion rate; VAS= visual analogue scale.

4.3.1 Motor speech production

Non-verbal motor functions of tongue, soft palate, lips and jaw were assessed as markers of cranial nerve dysfunction in the bulbar region (I). The strength, range and direction of the movements were observed during the non-verbal tasks as well as the evidence for fasciculation and atrophy of the tongue in motion and at rest. The evaluation of oral motor functions was composed to the present study by the author and is presented in Table 7. Each 14 tasks were evaluated on a four-point scale (3 = normal function; 0 = severe disorder) and the maximum total score was 42 points. The interjudge reliability between two licensed SPLs was high for oral motor tasks (intraclass correlation (ICC) = 0.918) in a sample of six patients (I). Maximum phonation time (MPT) was evaluated as a marker of laryngeal efficiency and respiratory capacity (Kent, 2009) reporting the best of three attempts in seconds. The patients were asked to count from one to twenty as quickly and as clearly as possible. The total number produced during one exhalation was used to evaluate breath support for phonation and the total time used to complete the counting 1-20 was measured in seconds to estimate the speed of speech movements.

To estimate the speed of articulatory movements both syllable and syllable sequence diadochokinesis (DDK) (I), speech rate (I) and articulation rate (II) were evaluated. In the DDK tasks, the patients were instructed to produce repetitively the Finnish syllable /ka/ and trisyllabic sequence /pataka/ as quickly and as accurately as possible for five seconds. The DDK rate was reported as syllables per second. The interjudge reliability between two licensed SLPs was high for DDK tasks (ICC = 0.984) in a sample of six patients (I). To evaluate the speech and articulation rate, the patients were asked to produce semi-spontaneous speech by generating a story based on a wordless nine-frame cartoon strip (Korpijaakko-Huuhka & Aulanko, 1994). The speech rate was reported as syllables per second per total narration time. The articulation rate was reported excluding either silent or filled (e.g. "um", "er") pauses of 200 msec or longer (Nishio & Niimi, 2000; Turner & Weismer1993) from the speech time. The interjudge reliability of the speech rate assessment between an SLP and an SLP student was high (ICC = 0.998) in six samples (I).

The semi-spontaneous stories were recorded with a headset microphone and saved as WAV files directly with Sound Forge software. Stories were transcribed orthographically for speech and articulation rate analysis and with syllable boundaries being verified acoustically using the PRAAT-program. In Study I, one patient who could not communicate with natural speech was excluded both from the syllable sequence and speech rate analysis. If the patient's speech was so impaired that

syllable boundaries were impossible to detect, the articulation rate was scored as 0 (II).

Table 7. Evaluation of oral motor functions (I)

Task	Functional level (3=mornal-0=severe impaired)	CN
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	XII
Base of tongue 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	X & 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= weakened strength of tongue either in protrusion or during lateral movement 1= decreased strength of tongue movements 0= minimal or no strength of tongue	XII
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 (/a:/) 1= incomplete or asymmetrical movement of soft palate 0= minimal velar movement/ no movement	X, IX & V
Lip rounding (puckers)	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 (Cheek 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	VII
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	V
Sub-scores	tongue: /18 velum: /3 lips: /9 mandible: /12 Total: /42	

4.3.2 Communicative activity

The functional change of speech was evaluated using the ALS Severity Scale of Speech (the speech scale) by the SLP and each patient (Hillel et al., 1989; Yorkston et al., 2004) (I, II, IV). The rating was based on cumulative data during the speech assessment by the SLP, while the patients were instructed to choose the best option from a ten-point speech scale to represent their current opinion about their speech impairment at the end of the speech assessment.

As the interjudge reliability between the SLP and patients was high (ICC = 0.968) in a sample of 30 evaluations, SLP scores were utilized in the analyses (I). The scores on a 10-point speech scale are as follows: score 10= normal speech; 9 = nominal speech abnormality; 8 = perceived speech changes; 7 = obvious speech abnormalities; 6=repeats messages on occasion; 5=frequent repeating required; 4=speech plus augmentative communication; 3 = limits speech to one- word responses; 2 = vocalizes for emotional expression and 1 = loss of useful speech. Speech scores 10 to 5 signify adequate speech, meaning that some deterioration in motor speech production might be observed, but speech still remains as a functional means of communication. A score 4 or less represents poor speech and AAC methods are needed to supplement or replace natural speech.

Functional capacity of communication (FCC) is a combination of three different ten-point ALS Severity Scales of speech and upper and lower extremities (Hillel et al., 1989; Yorkston et al., 2004; Yorkston et al., 1993) (III). FCC classifies functional performance into six different categories according to the adequacy of speech, hand function and mobility (Table 8.). Speech and hand functions were classified as poor if the scale score was 4 (needed AAC; needed assistance in self-care) or less, and mobility if the scale score was 6 (walked with a mechanical device) or less.

Table 8. Functional capacity of communication (FCC) categories

1	adequate speech and adequate hand function
2	adequate speech and poor hand function
3	poor speech, adequate hand function and adequate mobility
4	poor speech, adequate hand function and poor mobility
5	poor speech, poor hand function and good mobility
6	poor speech, poor hand function and poor mobility

The intelligibility of semi-spontaneous speech from the narrative cartoon task was evaluated perceptually by seven experienced SLPs (II, IV). A visual Analogue Scale (VAS) was used: value 0 (0mm) equals non-intelligible and a value of 100 (100mm) fully intelligible speech. The intelligibility was reported as the mean VAS value of seven listeners. The interjudge reliability between the listeners was good (ICC=.896) and the median across-judge difference was 5.5 (Q1=1 and Q3=20.75, min= 0 and max= 84) (III). The listeners listened to the speech samples in a random order. All seven listeners had the opportunity to listen to each sample as many times as required to be confident about their rating. A total of 126 speech samples, in which the patients were able to produce at least some narration, were presented to the listeners. If a patient was unable to produce any speech at all, as occurred in 31 cases, the intelligibility was scored as 0.

The means of communication used by the patients were listed according to their own report and the use of communication methods was verified in a clinical SLP evaluation (I, III, IV). If the patient reported using more than one communication strategy, the patients were asked to name the primary communication means. Communication aids needed due to only poor hand function were not included in this study. Communication methods were categorized into natural speech or into two AAC strategies: high-technology and low-technology. Further, high-technology devices were categorized into SGDs for AAC purposes only (SGDs) and into computer-based devices (computer-based SGDs) when all computer functions and applications were available. AAC strategies that involved no-technology were not included in this study.

4.3.3 Communicative participation

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

The social contexts in the CETI-M consisted of: 1) having a conversation with familiar persons in a quiet place, 2) having a conversation with strangers in a quiet place, 3) having a conversation with a familiar person over the phone, 4) speaking

with young children, 5) speaking with strangers over the phone, 6) speaking while traveling in a car, 7) speaking at a distance, 8) speaking in a noisy environment, 9) speaking before a group and 10) having a lengthy conversation (>1 h).

The social contexts 1-5 represent interactions in easy daily communication situations whereas contexts 6-10 are interactions in more demanding social situations requiring clear speaking strategies such as using a louder voice, precise articulation and a well-controlled speech rate. The first social context (having a conversation with familiar persons in a quiet place) was used as an example of an easy daily communication situation in this study; it was evaluated as the easiest one by both persons with ALS and their listeners (Ball et al., 2004b). Instead, the social context 8 (having a conversation in a noisy environment) was one of the most difficult social contexts described in the study of Ball et al. (2004b) and it was, therefore, used as an example of a demanding communication situation requiring clear speech strategies.

The patients self-rated the effectiveness of communication with all of the communication strategies (speech/low- or high-technology AAC) that they used. The mean value of the best given evaluation of each social context in the CETI-M was included regardless of which communication strategy was being used by the AAC-users. Thus, among AAC-users, the mean CETI-M with the best communication strategy might contain 1-4 different strategies (speech/ low- or high-technology AAC). The same protocol was used in the example situations of easy and demanding (social context 1 and 8) communication situations.

Only those social situations which are significant in a patients present life were self-rated, not situations that were irrelevant for them (for example, speaking with young children, if the patients did not have any contact with young children), because we wanted to observe possible communicative participation restrictions in real life situations. Further, the change in the number of self-rated social contexts relevant for a patient's real life during the follow-up was also observed as a marker of both for possible activity limitations and communicative restrictions.

4.3.4 Statistical methods

Both descriptive and statistical methods were used. The mean values, ranges and standard deviations (SD), quartiles and percentiles were used as descriptive statistics (I-IV). The intraclass correlation (ICC) was used for assessing interjudge reliability (I). Since the sample sizes were small, nonparametric tests were chosen for the group comparison. Mann- Whitney U test was used for continuous and interval variables

and Fisher's exact test was used for categorical variables (Studies I-IV). These statistical analyses were performed using SPSS (version 23; SPSS Inc.) All reported p-values are based on two-tailed tests and the statistical significance level was set at $p < 0.05$.

The data was unbalanced, i.e. there was a varying number of measurements for individual patients and also the severity of symptoms and communication abilities differed between patients. Therefore, a linear mixed-effect (LME) model with the symptom or score value as a dependent variable was fitted using the function LME in R (Software environment for statistical computing and graphics, version 3.3.0, The R Foundation for Statistical Computing) (II). Time, age at the first evaluation, gender and initial type of ALS were used as independent variables. Additionally, the interaction effect of time and the initial type of ALS were analyzed. The likelihood ratio test was used to compare models. A random intercept was used together with independent random errors. P-value of <.05 was considered statistically significant. A statistician performed the LME analysis in R. The author is responsible for the other analyses.

5 RESULTS

5.1 Clinical data

The study group (30 patients) was divided into groups based on the targeting being applied in the different sub-studies. In Study I, patients were divided into two groups based on the level of diagnostic certainty (possible vs. diagnosed ALS) so that it was possible to evaluate the influence of that certainty level on oral motor function, speech and communication at the beginning of the speech therapy process. The possible ALS-group includes patients with suspected and possible ALS and diagnosed ALS-group includes patients with probable and definitive ALS at baseline. In studies II and III, the groups were sub-divided according to the initial symptom at onset (bulbar vs. spinal) in order to investigate if the initial type of ALS would affect the deterioration of speech and the use of communication means with time. In Study IV, the patients were divided into two groups based on their primary communication means (speakers vs. AAC-users) to investigate how the use of deteriorating speech and other supplementing or substituting communication means would influences the effectiveness of the patients' communication. Patients data classified into the groups examined in the sub-studies is presented in Table 9. No statistical differences between the groups were found according to gender, age of follow-up times in any of the Studies I-IV. The most typical initial type of ALS was bulbar in the possible-group and spinal in the diagnosed ALS-group in Study I (Fisher Exact test, p=.014).

The patients in the possible ALS-group had noticed their first bulbar symptoms significantly earlier (approximately 5.8 months before the first visit to the neurologist) than the patients in the diagnosed ALS-group (approximately 1.5 months after the first to the neurologist) (Mann-Whitney U test, p = .006). Patients in the possible ALS-group had a significantly longer time between the first visit to a neurologist and the time of diagnosis of probable or definitive ALS in comparison with the diagnosed ALS-group (Mann-Whitney U test, p=.000) (I).

Table 9. Patient data in the separate studies

	Study I		Study II & III		Study IV	
	Possible ALS	Diagnosed ALS	Bulbar-onset	Spinal-onset	AAC-users	Speakers
Total (n)	16	14	13	17	14	16
Gender (F/M)	11/5	9/5	11/2	9/8	12/2	8/8
Initial type of ALS						
Spinal	4	11	0	17	4	11
Bulbar	9	2	13	0	8	3
Mixed	3	1			2	2
Age*, in years [mean±SD (min-max)]	61.1±10.4 (41.4–76.7)	64.6±13.6 (31.1–83.0)	64.3±9.4 (44.3–76.6)	61.6±13.7 (31.1–83.0)	62.8±10.5 (41.4-76.7)	62.7±13.4 (31.1–83.0)
Follow-up time, in months [mean±SD (min-max)]			16.7±7.1 (5–25)	14.3±8.6 (0–24)	16.0±7.0 (5–25)	14.8±8.8 (0–24)

AAC= augmentative and alternative communication; ALS=amyotrophic lateral sclerosis; F= female; M=male; SD=standard deviation.*age at baseline.

The times from the first noticeable bulbar symptoms to both the first SLP visit (Mann-Whitney U test, p=.001) and the final ALS diagnosis (Mann-Whitney U test, p=.001) were significantly longer in the bulbar group than in the spinal group (II). The first SLP visit occurred significantly earlier before a probable or definite ALS diagnosis in the bulbar-onset than in the spinal-onset patients (Mann-Whitney U test, p=.001)(II). The time from the first neurology visit to the first SLP visit was longer in the spinal-onset than in the bulbar-onset patients (Mann-Whitney U test, p=.000).

5.2 Motor speech disorder and communication before a definitive ALS diagnosis (I)

Patients in the possible ALS-group had greater decreased soft palate function and slower syllable, number and speech production than patients in the diagnosed group at the baseline. There were statistically significant differences between the diagnosed and possible ALS-groups in the soft palate sub-score in their oral motor assessment, time to count 1–20, DDK tasks /ka/ and /pataka/ and also in the speech rate (Mann-Whitney U test, $p < 0.05$). There were no statistical differences between these

groups in total score of oral motor tasks, breath support for speech or laryngeal efficiency and respiratory capacity.

Based on the mean speech score (ALS severity scale of speech), at baseline the speech disorders were significantly more severe in the possible than in the diagnosed ALS group (Mann-Whitney U test, p=.002). Patients in the possible ALS group speech required repeated messages on occasion (mean speech score 6.19±1.47), whereas those patients in the diagnosed ALS group had perceivable speech changes (mean speech score 8.14±1.88).

At baseline, 90% (27/30) of the patients communicating with speech and not exploiting any other communication. Two patients occasionally used handwriting to support their diminished speech. One patient communicated primarily with handwriting and occasionally by one-word speech responses.

5.3 Changes in speech and communication during a two-year follow-up

5.3.1 Articulation rate (II)

Patients with bulbar-onset ALS had a slower articulation rate than spinal-onset patients at baseline and this difference remained during the follow-up period despite the fact that the articulation rate became slower in both groups (LME model, p<.001) (Figure 6.). Time and initial type of ALS were significant factors in determining the articulation rate LME model, but age or gender were not statistically significant.

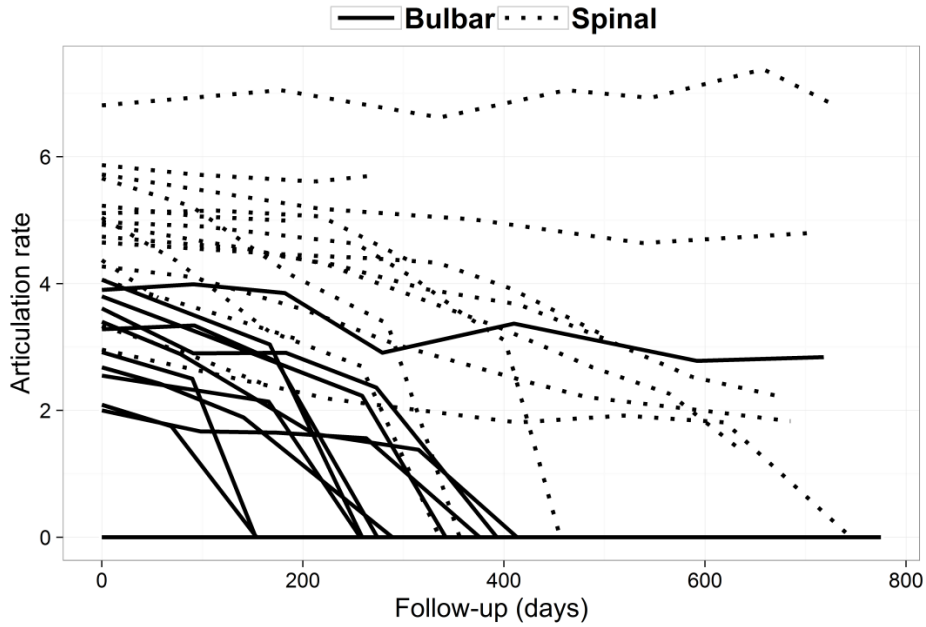

Figure 6. Changes in articulation rates in patients in the bulbar and spinal groups

5.3.2 Severity of speech disorder (II)

Time and initial type of ALS were significant factors in determining the speech score LME model ($p = < .001$) and patients with spinal-onset ALS had higher speech scores than patients with the bulbar-onset disease (Figure 7). This difference remained during the study despite the fact that the speech scores declined in both groups. Age or gender were not significant determinants of the speech severity LME model.

At baseline, speech was poor (a speech score of 4 or less) in three bulbar-onset patients (I). After the first-year follow-up, only one out of the 8 remaining bulbar-onset patients was able to speak adequately. The first spinal-onset patient lost an adequate speech capability just before the 9 month visit.

In total, speech deteriorated to poor in 18/30 patients during the study (13/13 bulbar-onset and 5/17 spinal onset). The average time from the appearance of bulbar symptoms to the loss of adequate speech was 18.4 (range 4-60) months in those 18 patients. Two spinal-onset and four bulbar-onset patients (out of those 18) survived the whole two-year follow-up time. Three spinal-onset patients out of the nine remaining patients were able to speak adequately (speech score 5 or more) at the end

of two-year follow-up. In those 12 out of the original 17 spinal-onset patients, who were lost during follow-up, the most common speech score was 6 (range 1-8) at last visit.

The mean speech score was lower (Mann-Whitney U test, p=.001) in AAC-users (mean speech score 5.9 ±1.7) than speakers (mean speech score 8.1±1.5) at baseline (IV).

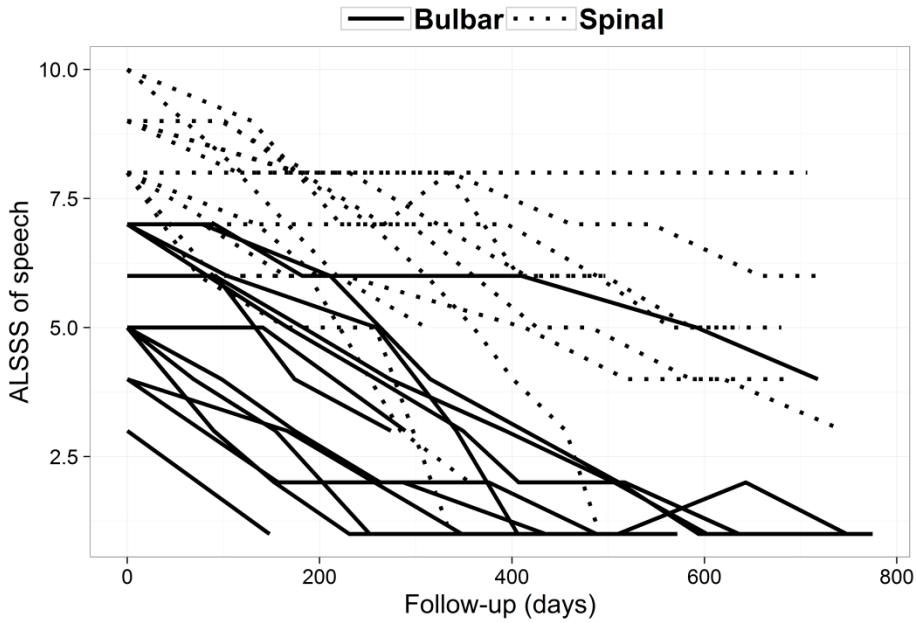

Figure 7. Changes in speech score of patients in bulbar and spinal group

5.3.3 Intelligibility (II)

Time (p = < .001), initial type of ALS (p = .0043) as well as an interaction effect between time and initial type of ALS (p = .0047) were significant factors in speech intelligibility LME models. Instead, age or gender were not significant in intelligibility LME model. Overall, speech intelligibility was better in patients with spinal-onset ALS than in patients with bulbar-onset disease and this difference remained during the study despite the fact that intelligibility decreased in both groups (Figure 8).

Speech intelligibility was more extensively deteriorated (Mann-Whitney U-test p=.009) in AAC-users (mean intelligibility 85.3± 30.5) compared to speakers (98.3 ± 5.9) at baseline (IV).

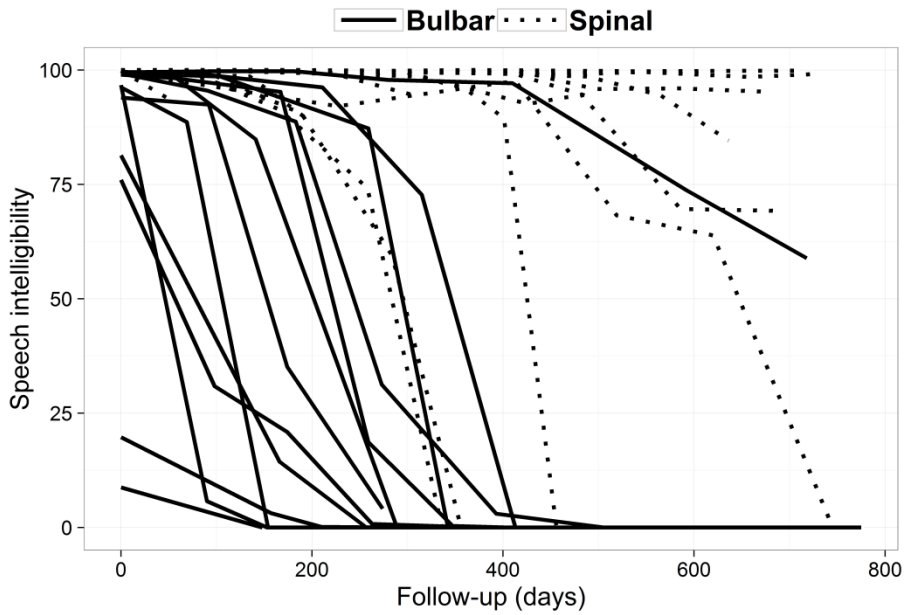

Figure 8. Changes in speech intelligibility of patients in bulbar and spinal group

5.3.4 Variations in communication strategies (III)

The need and use of different communication means varied from one individual to the next. The variations in communication means are presented in Figure 9. in the bulbar-onset group and in Figure 10. in the spinal-onset group. Twelve bulbar-onset and five spinal-onset patients supported or substituted speech with at least one low- or high- technology AAC strategy. Patients with initial bulbar symptoms primarily favored low-technology communication strategies such as handwriting to support their deteriorated speech but later also exploited high-technology solutions. The variations in the used communication means were 1-7 among bulbar-onset and 1-5 among spinal-onset patients during the study. Patients might use as many as four different communication means at the same time in order to ensure functional communication.

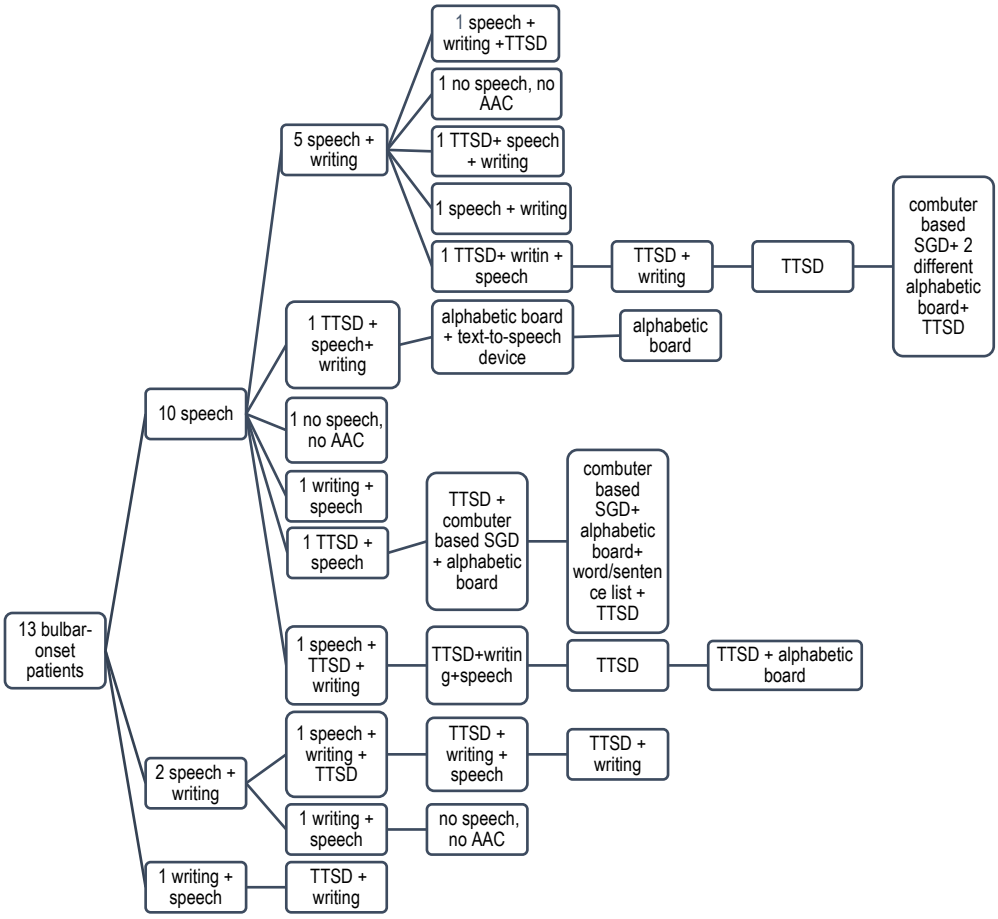

Figure 9. Variation and modification in the communication means in bulbar-onset patients during the study period. The number of participants using each communication mean is indicated in the box.

AAC=augmentative and alternative communication; SGD= speech-generating devices; TTSD= text-to-speech devices; Writing= handwriting

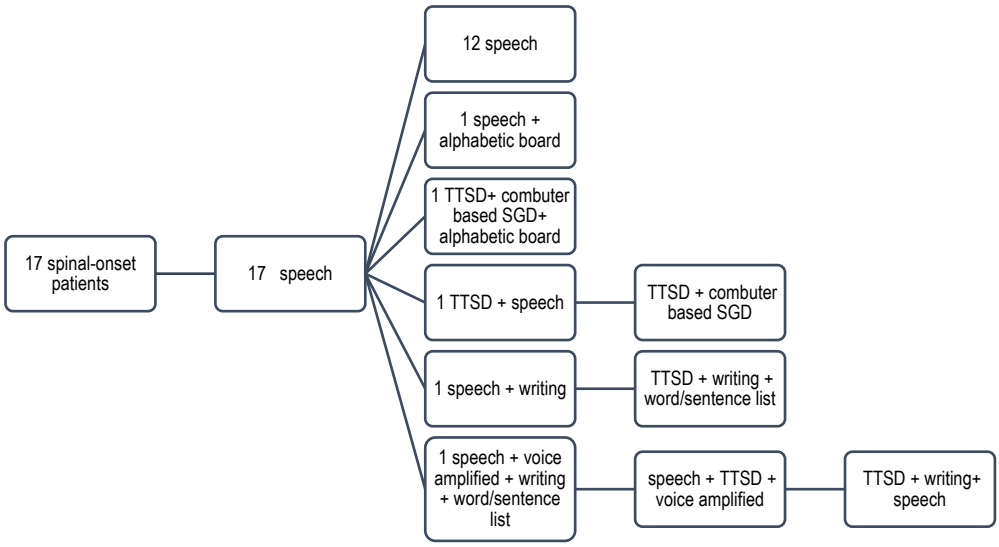

Figure 10. Variation and modification in the communication means in spinal-onset patients during the study period. The number of participants using each mean is indicated in the box.

SGD= speech-generating devices; TTSD= text-to-speech devices; Writing= handwriting

5.3.5 Functional capacity of communication (III)

At the baseline, 90% of the patients (27/30) communicated with natural speech and were classified into one of the two good speech FCC categories. A total of 63.3% of patients (19/30) with both adequate speech and adequate hand function were classified into FCC category 1, while 26.7 % of patients (8/30) with adequate speech but poor hand function were classified into FCC category 2. Three bulbar-onset patients had already poor speech with good hand function and mobility at baseline and therefore classified into category 3.

During the follow-up, the FFC classification changed in 92% (12/13) of the bulbar-onset patients, while only one patient's classification remained in category 3. At the end, all bulbar-onset patients FCC had declined to one of the four poor-speech categories: 3 (n=4, poor speech, adequate hand function and mobility), 4 (n=1, poor speech, adequate hand function, poor mobility), 5 (n=2, poor speech and hand function, good mobility) or 6 (n=6, poor speech, hand function and mobility).

Among patients with initial spinal-onset disease, speech remained adequate in 71% (12/17) and therefore their FCC classification was either 1 or 2 also at last visit. However, in 29% (5/17) of spinal-onset patients, the FCC category changed from the adequate speech categories to poor speech and poor mobility categories with adequate (n=1) or poor hand function (n=4). It is noticeable that none of the spinal-onset patients were categorized into either category 3 or 5 at any phase of the study.

5.4 Changes in effectiveness of communication (IV)

Already at the baseline, a significant proportion, about 37% (11/29) of patients reported that the effectiveness of their natural speech was decreased (mean score of CETI-M<5) (I). Mean CETI-M was 5.17. At same time, 17% (5/29) of patients self-rated their natural speech as very effective (mean CETI-M=7). The self-rated effectiveness of natural speech declined steadily with time. The changes in the effectiveness of natural speech in the two groups (both patients who communicated predominantly with natural speech and also in patients who started to communicate primarily by exploiting some low- or high-technology AAC means during the study) are demonstrated in Figure 11.

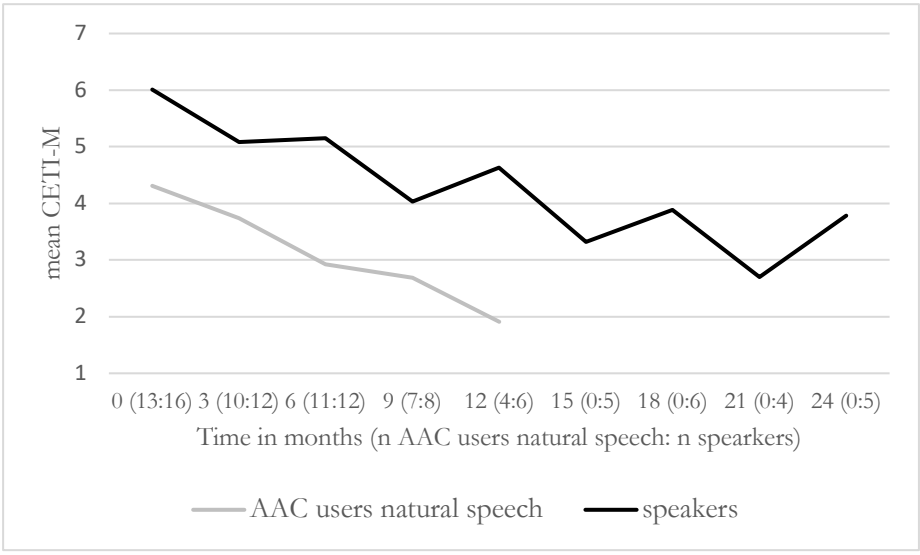

Figure 11. Changes in effectiveness of natural speech

The AAC users self-rated their communication effectiveness as being higher with AAC means than when communicating with natural speech alone. Communicating with multiple communication strategies such as utilizing low- or high-technology AAC means and using diminished natural speech when possible, made it possible to achieve more effective communication than using natural speech alone. In fact, the speakers self-rated their effectiveness of communication in a noisy environment (a demanding communication situation) as worse than the corresponding rating given by AAC users did.

The need for AAC means impacted on the amount of self-evaluated social contexts. The mean number of social contexts as self-evaluated among speakers remained at the same level (mean 9.69 at baseline, 9.8 at 12 months and 9.6 at 24 months), whereas the self-evaluated number of social contexts decreased (mean 9.15 at baseline, 8.5 at 12 months and 6.25 at 24 months) in the AAC-users during the study possibly indicating that they felt that they were experiencing a reduction in this parameter.

6 DISCUSSION

The purpose of this thesis was to investigate the longitudinal changes in speech, communication strategies and their effectiveness after the manifestation of bulbar symptoms in ALS patients. Impairment of motor speech production, activity limitations in communication and restrictions in participation in communicatively different situations progressed with time; more so in bulbar-onset than spinal-onset patients. With various AAC strategies, it is possible to maintain the effectiveness of communication in social situations important to the person's life. The majority of spinal-onset patients maintain an ability to communicate with speech, although some patients lost both the ability to speak and the ability to exploit AAC means. We chose to examine the motor speech production and communication from the perspective of the ICF Functioning and Disability components (ICF, 2001). The relationship between the sub-studies and ICF component are presented in Figure 12.

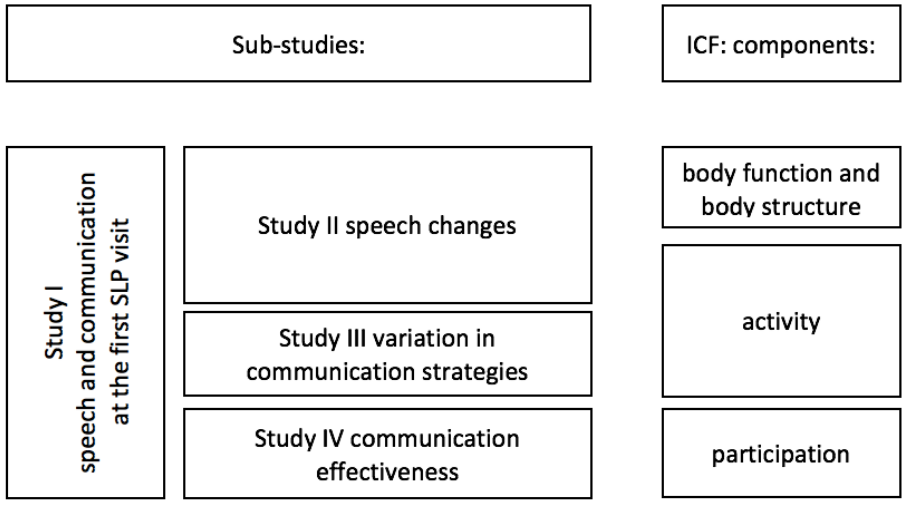

Figure 12. The relationship between the sub-studies and ICF-domains.

6.1 Speech and communication changes

Patients with bulbar-onset disease had a longer interval between onset of bulbar symptoms and the first SLP visit as compared to the spinal-onset patients. Spinal-onset patients were referred to an SLP sooner, because they were already being monitored and treated in a multi-disciplinary neurological unit due to their other symptoms. As bulbar symptoms have a negative impact on survival (Chi et al., 2009; Zoccolella et al., 2008), SLP intervention and also scientific studies, should be initiated as soon as possible after their manifestation.

6.1.1 Impairment of body function and structure

The variation of oral motor dysfunctions were wide at baseline; from totally lost tongue and soft palate function with clearly decrease of lip and jaw function to the normal oral motor function. The tongue functions were the most severely impaired bulbar functions, but nasality is the first and the most predominant symptom in some patients with ALS in the present study. This finding is in line with numerous studies reporting tongue dysfunction as a typical early symptom and an overall common symptom in ALS (Hillel & Miller, 1987; Kent et al., 1992; Kuruvilla et al., 2012; Langmore & Lehman, 1994; Rong et al., 2016; Yunusova et al., 2008; Yunusova et al., 2012). Impaired soft palate function causes nasality, which is one of the most affected phonetic features in ALS (Kent et al., 1990; Kent et al., 1992).

Speech production was slower in patients with possible ALS than in patients with ALS diagnosed at baseline. The DDK rates were slower than the performance reported in English-speaking healthy adults (Kent et al., 1987; Kent, 2009) or healthy elderly Finnish-speaking adults (Nevala 2014), reflecting a reduced speed of articulatory movements in all ALS patients already at baseline. The semi-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). However, the mean speech rate in diagnosed ALS group was at a level similar to that of healthy Finnish-speaking adults.

The articulation rate declined in time in nearly all participants; this was worse in bulbar-onset patients both at baseline and during the follow-up. All but one patient's articulation rate decrease to zero, in contrast in the spinal-onset group four out of 17 patients experienced a similar decline. Because the interaction effect between time and initial type of ALS was not significant for the articulation rate, this suggests that

the reduction of the speech rate progresses similarly, regardless of the initial type of ALS.

All these speech rate tasks examine the rapidity of speech production from somewhat different aspects of this phenomenon. Syllable and syllable sequences DDK provide primary information about the speed and regularity of rapid, repetitive articulation movements. Respiratory impairment is better assessed in the longer tasks such as the time used for counting from 1 to 20 than by the shorter 5 second DDK tasks but severe respiratory insufficiency is also detectable in the short speech task performances. The articulation rate (speech rate minus pauses) is thought to represent the speed of articulation movements and to be sensitive to the changes of motor speech functions, whereas instead the speech rate is thought to represent also other functions including cognitive and respiratory capacity (Green et al., 2013; Yunusova et al., 2016).

Overall the patients in the possible ALS group performed more poorly in impairment-level tasks in comparison with the diagnosed group. The fact that speech rates may deteriorate significantly before the confirmation of ALS diagnosis highlights the importance of conducting an early speech assessment. Regular monitoring of speech rates is considered an essential part in the timely introduction of the AAC process (Hanson et al., 2011).

6.1.2 Activity limitations in communication

The patients with possible ALS showed significantly more severe dysarthria than participants in the group already diagnosed at baseline. The result is explained by a large number of bulbar-onset patients in the possible ALS group. All impairments in bulbar function do not necessarily impact the severity of speech disorders and three spinal-onset patients had a speech score of 10 at the baseline even though they had at least one bulbar symptom.

Time and the initial type of ALS were the most important factors to predict the decline in the speech score and speech intelligibility. Both of these parameters declined in nearly all participants, as was expected based on previous studies and clinical experience (Ball et al., 2002; Green et al., 2013; Hillel et al., 1989; Kent et al., 1991; Nishio and Niimi 2000; Rong et al., 2016, Turner and Weismer 1993; Watts and Vanryckeghem 2001). Bulbar-onset ALS patients displayed more severe speech disorders and poorer intelligibility at baseline than their spinal-onset counterparts. This difference persisted throughout the study period being compatible with earlier

results in the sense that patients with initial bulbar symptoms tend to have more severe motor speech disorders in comparison with those with initial spinal symptoms (Turner et al., 2010, Yorkston et al., 1993).

Furthermore, during the follow-up, interaction effect between time and initial type of ALS was a significant factor contributing to intelligibility, suggesting that intelligibility declines differently in bulbar- and spinal-onset patients. Speech intelligibility declined more in bulbar-onset patients during the follow-up: all except one bulbar-onset patient had unintelligible speech at the last visit in contrast to only four out of the 17 spinal-onset patients. Over half of the spinal-onset patients had rather good intelligibility (>90/100) at the last visit (see figure 8.). One reason why intelligibility may decline differently than the articulation rate and speech severity is that the intelligibility measurements lacking sensitivity in the early stages of speech disorders. It has been claimed that intelligibility declines rapidly and progresses to the complete loss of adequate speech in a rather short time during the later stages of the ALS (Ball et al., 2001; Green et al., 2013; Nishio and Niimi 2000; Rong et al., 2016, Yorkston et al., 1993). The present study has found similar results, even though the language of our participants and the methods used for evaluating intelligibility were different; nonetheless some caution is necessary when making comparisons with the earlier studies. We evaluated intelligibility from semi-spontaneous speech in the cartoon narrative task while in the previous studies, intelligibility has been assessed by single word or sentence reading tasks.

The progression speed from the first bulbar symptoms into poor speech function (=speech score 4 or less) was similar in both bulbar- and spinal-onset patients although there was a discrepancy in the numbers of patients who lost functionally adequate speech. Speech become impaired to a poor level in all (100%) bulbar-onset patient in contrary to only about the 30% of spinal-onset patients. This study confirms to the results of the previous studies where dysarthria progressed from mild to severe in the same 18 month-time-frame in bulbar-onset patients (Turner et al., 2010) and that a speech change occurred earlier in bulbar-onset patients in comparison to their spinal-onset counterparts (Yorkston et al., 1993). Unlike in the earlier studies, we focused on the starting point for the bulbar symptoms rather than the confirmation of diagnosis which made it possible to estimate that the timeframe of speech deterioration after the appearance of the bulbar symptoms, regardless of the initial type of ALS.

6.1.2.1 Means of communication

According to our data and clinical expertise maintaining the ability to communicate with AAC means is a demanding and continuing process, which is not always timely recognized in a timely manner. The time-frame of organizing the communication aid services being is on average seven months longer in the spinal- than that of bulbar-onset patients. The bulbar-onset patients exhibited more severe speech symptoms at the first SLP visit and therefore they needed to exploit AAC services sooner than spinal-onset patients who had been referred to the SLP with milder bulbar and speech symptoms.

The proportion of AAC users is smaller in our study than in earlier reports which have stated that most of the ALS patients could not communicate adequately with natural speech or lost their ability to speak as their disease progressed (Ball et al., 2007b; Beukelman et al., 2011). However, 57% of bulbar-onset patients have been reported to develop anarthria (Turner et al., 2010). In our data, speech became nonfunctional in 60 % of the participants and 57% of the participants were using at least one low- or high technology AAC strategy. Again, the proportion of AAC users is also smaller compared to the 72% of ALS patients who have previously been shown to benefit from AAC strategies (Creer et al., 2016). In our data nearly 30% did not need any AAC means (speech score 6-8) at the last visit. The dissimilarity of the results may be due to the differences in the distribution of the initial types of ALS in the studied populations and the use of invasive ventilation. More than half of the patients in the current data had spinal-onset ALS, and 77% of them retained their ability to speak (speech score 5 or more) even during the late stages of their lives. Nevertheless, the present results are in line with previous reports when only bulbar-onset patients are considered. Additionally, very few ALS patients in Finland generally have (and none in this study had) supported invasive ventilation with tracheostomy. Invasive ventilation support has been recognized to affect both the AAC technology which will be chosen and the duration of its use (Ball et al., 2007a).

The bulbar-onset patients favored both low- and high-technology solutions, while patients in the spinal-onset disease showed a preference for only high-technology devices. The most common AAC mean used by bulbar-onset patients was handwriting or a text-to-speech device, whereas spinal-onset patients most frequently used text-to-speech devices. The variation may be explained by differences in their functional capacities especially with regard to their upper extremity capabilities (Mathy et al., 2000).

The accessibly of technological AAC solutions or economic reasons did not explain the choice of AAC strategies. All patients in the current study received equal guidance regarding the possibilities of AAC use and equal possibilities to borrow, free-of-charge, all appropriate low- and high- technology AAC solutions.

Natural speech is a powerful means of communication even when impaired and some people with ALS prefer to communicate primarily with speech throughout the progression of their disease. Our findings are in line with the result that some ALS patients choose to use their own speech for as long as possible, even when it is unintelligible to others (Murphy, 2004). On the other hand, gestures and facial expressions (=non-technology communication) serve as functional ways to communicate and support diminishing speech capabilities. The use this kind of non-technology communication is difficult to evaluate or measure and therefore it was not included in the present study. Our data does not offer any explanation why some people choose to communicate with unintelligible speech.

It is difficult to substitute for natural speech. Often multiple AAC means were needed to support or substitute for the deteriorating speech to maintain effective communication in different social situations. Some of the patients needed up to four different communication strategies simultaneously and utilize as many as seven different AAC strategies during the study, because the FCC changed as the disease progressed. One explanation for the high number of secondary communication strategies is our clinic's policy which states that the SLP shall provide guidance also in the use of at least one low-technology AAC means when patients start to learn to apply some of the high-technology AAC strategies. This is to ensure the patients' abilities to communicate in conditions where high technology is not suitable or accessible, for example when a device is out of order or in case of a power failure.

6.1.2.2 Functional capacity of communication

The change in the functional capacity of communication (FCC) influences the need and use of different AAC forms (Fried-Oken et al., 2015; Yorkston et al., 2004). In the present study, the category of FCC classification changed in 63% of the participants as the disease progressed and again, there were differences between the groups based on the initial symptoms: FCC classification changed in almost all 92% of the bulbar-onset patients but only in 41% of spinal-onset patients. The majority of bulbar-onset patients first lost the ability to speak adequately and then their hand function or overall mobility or both deteriorated. Most spinal-onset patients,

however, already had poor mobility with or without poor hand function when their speech became non-functional. This is in agreement with the natural course of ALS and, in fact, the initial symptoms tend to predominate throughout the disease (Yorkston et al., 1993). It should be noted, that poor speech with good mobility categories occurred in only a few bulbar-onset patients. Category 5 (=poor speech, poor hand function and good mobility) was very rare and occurred only in two different bulbar-onset patients after their speech had become unintelligible and hand functions had decreased. Therefore it is extremely challenging to ensure independent possibilities to communicate. As the FCC changes during the progression of the disease, a regular assessment of speech and communication is needed (EFNS Task Force on Diagnosis and Management of Amyotrophic Lateral Sclerosis, 2012; Hanson et al., 2011) and suitable AAC means should be individualized and modified throughout the disease progression (Fried-Oken et al., 2015).

6.1.3 Participation restriction

The effectiveness of speech had already declined in about 30% of patients at baseline and in all patients the effectiveness of natural speech inevitably decreased. Previously there have been mainly reports about non-effective communication in those ALS patients who communicate exclusive by speech (Ball et al., 2004b; Beukelman et al., 2011).

However, the effectiveness of communication may even increase or at least remain at the same level when patients start to communicate with AAC strategies since these can support or substitute for their impaired natural speech. Our results emphasize the significance of multiple communication strategies with regard to the functional participation in a variety of social situations. Londral et al. (2015) also reported that patients enjoyed more effective communication with communication devices in comparison with no devices.

Factors influencing the degree of difficulties in social situations might be different between speakers and AAC-users. Communication in demanding contexts, such as having a conversation in a noisy environment or having a lengthy conversation, might be more effective when using AAC strategies than with natural speech. For example, with speech devices, it is easy to have a loud voice simply by increasing the equipment's volume setting. Furthermore writing with a predictive text application or using a set of pre-recorded phrases might not be as difficult as trying to have a conversation when speech is badly impaired.

Though the communication effectiveness remained the same or even increased, the number of social contexts relevant to a person's real life decreased over time, especially in those who primarily communicated with AAC means. One possible explanation might be the narrowing of their social life as the disease progressed because the patients had a decreasing functional capacity and an increasing need for aid and care. Another explanation might be that persons who used AAC means focus on social situations that are important to them while ignoring the less significant occasions. It is also worth mentioning that despite the fact that overall functional abilities decline as ALS progresses, the patients' self-perceived quality of life does not necessary deteriorate (Jacobsson Larsson et al., 2017).

6.2 Methodological considerations

The strengths of this study are the inclusion of patients as a consecutive series with their prospective follow-up from the first SLP evaluation in a multidisciplinary neurological clinic. Strict exclusion criteria and the confirmation of all patients' diagnoses by an experienced neurologist strengthen the conclusion that speech and communication of the patients truly changed due to ALS.

A larger sample size would have improved the generalizability of the findings. However, it is challenging to collect a clinically valid follow-up data from patients with ALS because of the progressive and fatal course of the disease. Patients were inevitably lost during the long follow-up. In the present study, the patients were lost regularly; 10 patients during the first year and 11 during the second year. Three patients were lost before a probable or definitive ALS diagnosis was made and therefore their data was not included in this study.

In about one third (11/30) of the patients, the initial type of ALS was bulbar reflecting the typical distribution of spinal versus bulbar types of onset (Kiernan et al., 2011). Female participants outnumbered males (20 vs 10), especially among the bulbar-onset patients. ALS is known to be more common in men than in women (van Es et al., 2017; Kiernan et al., 2011). Women with bulbar-onset ALS are reported to experience more pronounced speech problems and they may, therefore seek treatment earlier than women with spinal-onset disease or men with any type of ALS (Yorkston et al., 1993). This may be one explanation for the female patients being referred to an SLP comparatively early and consequently also for the predominance of females sample in this study.

Neither age nor gender were significant in relation to the declines of articulation rate, speech intelligibility or the severity of speech disorders in this study. In earlier reports, male gender and increasing age have been found to be among the independent risk factors for ALS (Alonso et al., 2009; Haverkamp et al., 1995; Scott et al., 2009). Female gender and higher age have been related to a more severe bulbar dysfunction and to a greater probability of dysarthria progressing into anarthria (Turner et al., 2010; Yorkston et al., 1993).

This is a mixed-method study utilizing both qualitative and quantitative methods. LME models were used to analyze longitudinal data because there was a varying number of measurements (data was unbalanced) for individual patients and because the severity of symptoms and communication abilities differed between patients. The methods used in present study are commonly used with dysarthria assessments, in general, especially in ALS (Ball et al., 2004b; Duffy, 2013; Kent, 2009; Yorkston et al., 1993). The measurement tools were chosen to assess both the early and the longitudinal changes in motor speech production. There is a floor effect, where the performance cannot decrease any further; this was, achieved in variables related to speech (for example, in the articulation rate) when the patient lost the ability to speak. In order to increase the reliability of subjective measurements (for example, the oral motor assessment, ALSSS of speech, intelligibility) the interjudge reliability was estimated. One potential bias is the patients' self-evaluation about the effectiveness of communication. Furthermore, persons with ALS and their loved ones have been found to evaluate similarly the effectiveness of communication and social communication difficulties (Ball et al., 2004b; Fisher et al., 2017).

The CETI-M is limited for measuring participation and its restriction. CETI-M makes it possible to examine the quality of participation from the aspect of communication effectiveness, but from a wider perspective, for example, it is difficult to evaluate the extent of participation in work and leisure activities and the amount of communicative participation. The ICF components of environmental and personal factors, which were not included this study, also affect the activity and participations capabilities especially in users of AAC.

There is also some potential bias in the patients' recall of the timepoint when bulbar symptoms appeared. As a relatively small number of spinal-onset patients (5/17) lost their ability to speak, this might be reflected in some of the statistical findings such as the time that the patient started to need AAC. The progression of dysarthria was similar in those five spinal onset patients compared to bulbar-onset patients. One limitation is the lack of knowledge of the time-point when spinal symptoms appeared.

One limitation is the lack of a formal cognitive assessment. It should be noted however that we measured articulation rate without pauses to detect changes in the speed of articulatory muscles. The articulatory rate is not as sensitive to the effect of possible cognitive deteriorations as the speech rate (Green et al., 2013; Yunusova et al., 2016). In addition, the speech intelligibility and the speech and articulation rate were measured from semi-spontaneous narrative speech which did not demand reading skills from the participants.

6.3 Clinical implications and future directions

There seem to be two important factors on which to focus when organizing a speech therapy intervention and communication aid processes. Both the timing of the intervention and the initial type of ALS are significant. In clinical work, it would be preferable to focus on the appearance of the first bulbar symptoms when planning a timely SLP intervention. After the manifestation of bulbar symptoms, the loss of functional speech will occur within 18 months on average, as observed in those 18 patients whose speech decreased to a poor level in this study. SLP intervention will support the person's communicative independence even before the definite diagnosis.

Patients with bulbar symptoms should be referred to an experienced SLP who is a part of a multidisciplinary ALS-team as soon as possible to undergo an appropriate diagnostic evaluation and to receive information and guidance about their bulbar symptoms, speech disorders as well as information about future procedures. It is beneficial for patients with bulbar symptoms to be referred to an SLP even before seeing a neurologist. A thorough and well documented evaluation by an ALS specializing SLP may also further the diagnostic processes. The importance of specialized SLP expertise and regular speech therapy assessments are emphasized during the time before a certain diagnosis is made and in the months immediately after the ALS diagnosis has been determined.

It is most likely that virtually all patients with bulbar-onset ALS will lose the ability to speak while part of the spinal-onset patients will probably still be able communicate with natural speech even in the very late stages of the disease. Furthermore, not all ALS patients will need to exploit AAC means. The articulation rate and the level of speech disorder severity seem to decline before intelligibility, which is an important issue to note and it is recommended to focus on monitoring

the change in these aspects of motor speech production. It is recommended that regular speech and communication assessments should be organized for the patients with bulbar symptoms and there should be a focus on counseling patients with ALS and their family members about the possibilities of AAC means at a sufficiently early time to help them maintain the ability to communicate functionally and effectively for as long as possible.

The initial type of ALS should also be taken into consideration when undertaking the communication aid process in order to choose the most functional AAC means. Patients with bulbar-onset ALS typically have good hand function at the time when they first need AAC options, and they are able to master a , wide variety of different AAC solutions. Nevertheless, the overall functional capacity of these patients is likely to change over the course of the disease, demanding a regular assessment and the readiness to react quickly should there be a need to change the AAC mean. In comparison, spinal-onset patients most probably already use a walker or a wheelchair and their hand function has most likely deteriorated at the time when AAC means became necessary. At this point it will be necessary to consider AAC devices accessible with minimal functional capacity.

In the future, more research should be conducted especially on the aspects of communicative capacity, performance and effectiveness of using multiple communication means. It would be useful to study the possibilities of supporting communicative participations by including also both environmental and personal factors.

It is unclear why some patients with ALS prefer to communicate with nonfunctional speech and this issue will need further investigation: the problem might become clearer with respect to cognitive impairments due to the association with FTD. It would also be interesting to study whether genetics might explain the differences in symptom progression, communicative tendencies and receptivity AAC means. Cognitive impairments probably impact on the patient's willingness to accept and utilize different AAC strategies; if the impairment is sufficiently severe then it may nullify their possibilities to utilize certain AAC means (Brownlee & Bruening, 2012; Geronimo et al., 2016). Thus, a formal cognitive assessment should also be conducted when evaluating speech and communication changes in future studies.

With respect to AAC guidance and education, SLPs and other health care professionals should acknowledge that the patient with ALS usually will need several different strategies if he/she is to communicate effectively in all the social situations important in their lives; they should be offered the opportunity to practice with

various AAC options. During an education situation it, might be worthwhile to highlight the fact that based on the ALS patients' self-ratings, it is more likely that the use of multiple AAC means can help the patient to maintain or even improve the effectiveness of communication in preference over confining him/herself to diminished speech

Although ALS entails a characteristic deterioration of motor speech abilities and the probable need for AAC strategies, it would be beneficial for clinicians to recognize the heterogeneity of motor speech disorders and communication capacity from the first SLP visit onwards. There should be a clear aim towards individualized guidance, intervention planning and provision of AAC services. However, occasionally it is necessary to recognize and appreciate that functional communication strategies no longer exist due to reasons such as cognitive dysfunction or the person's own decision.

ALS is a heterogeneous syndrome and also the speech and communication difficulties in ALS appear in very complex combinations. A multidisciplinary approach to the speech and communication problems will be required also in the future. A successful evaluation of early bulbar symptoms requires professional logopedic expertise and an active multidisciplinary team specialized to ALS; this can facilitate of the diagnostic process and help in monitoring the progression of motor speech symptoms and functional capacity of communication as well as also realizing the possibilities for maintaining communicative effectiveness with AAC means.

7 SUMMARY AND CONCLUSIONS

1) ALS patients, especially those with bulbar-onset disease, might suffer a significant speech and communication deterioration even before a definite diagnosis has been made.

   An individual with bulbar symptoms should be subjected to a speech therapy evaluation as soon as possible regardless of the certainty level of ALS diagnosis. This makes possible optimizing speech therapy interventions and the provision of AAC services and also a thorough speech therapy evaluation can produce important information about bulbar symptoms, motor speech production and communication which might facilitate diagnostic processes.

2) There are decreases in the articulation rate and motor speech production before there is any decline in intelligibility. In this thesis project, all of the bulbar-onset patients lost their ability to speak so they could be understood by others, while in the speech capabilities of about 70% of spinal-onset patients remained adequate. Speech remained functional on average for 18 months after the appearance of the first bulbar symptoms in those 18 patients who lost the ability to speak.

   Attention should be paid to the initial symptoms of ALS and also assessing when the first bulbar symptoms appeared. In the speech therapy evaluations, observing changes in the speech and articulation rates and the severity of dysarthria are important factors in predicting the loss of intelligibility of speech and the need for AAC services.

3) Multiple communication strategies are needed to compensate for impaired speech production and the changes in the functional capacity of communication should be noted during disease progression. Patients with spinal-onset disease favor high-tech communication strategies while bulbar-onset patients show a preference for both low- and high-technology AAC means.

   The various AAC strategies should be demonstrated and offered to the patients to allow him/her to maintain the ability to communicate in all important possible social contexts; there is also a need to be able to react quickly to any

changes in functional capacity which can impair the patient's ability to communicate with others.

4) The effectiveness of communication may remain or even improve if the patient is able to utilize multiple communication strategies.

Guidance and appropriate training of the different AAC means are required to support the ALS patients' ability to stabilize their communicative effectiveness as their natural speech deteriorates.

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10 ORIGINAL PUBLICATIONS

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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 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 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.

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; Yorkstonet al., 2004).
Note 2: In column 'first bulbar symptoms' the minus refers to months before the first visit to a neurologist

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 (Korpijaakko-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.

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).

Table 2
Oral motor functions and speech production in groups G1 and G2 (N = 30).

	group G1					group G2					Mann-Whitney *U* test
	n	mean	range	SD	median	n	mean	range	SD	median	p-value
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.
* $p < 0.05$.

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 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

Table 3
Self-evaluated effectiveness of speech communication using CETI-M.

CETI-M contexts	Diagnosed group (G1)				Possible group (G2)				Mann-Whitney *U* 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.

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

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Research Report

Speech deterioration in amyotrophic lateral sclerosis (ALS) after manifestation of bulbar symptoms

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Abstract

Background: The symptoms and their progression in amyotrophic lateral sclerosis (ALS) are typically studied after the diagnosis has been confirmed. However, many people with ALS already have severe dysarthria and loss of adequate speech at the time of diagnosis. Speech-and-language therapy interventions should be targeted timely based on communicative need in ALS.
Aims: To investigate how long natural speech will remain functional and to identify the changes in the speech of persons with ALS.
Methods & Procedures: Altogether 30 consecutive participants were studied and divided into two groups based on the initial type of ALS, bulbar or spinal. Their speech disorder was evaluated on severity, articulation rate and intelligibility during the 2-year follow-up.
Outcome & Results: The ability to speak deteriorated to poor and necessitated augmentative and alternative communication (AAC) methods with 60% of the participants. Their speech remained adequate on average for 18 months from the first bulbar symptom. Severity, articulation rate and intelligibility declined with nearly all participants during the study. To begin with speech deteriorated more in the bulbar group than in the spinal group and the difference remained during the whole follow-up with some exceptions.
Conclusions & Implications: The onset of bulbar symptoms indicated the time to loss of speech better than when assessed from ALS diagnosis or the first speech therapy evaluation. In clinical work, it is important to take the initial type of ALS into consideration when determining the urgency of AAC measures as people with bulbar-onset ALS are more susceptible to delayed evaluation and AAC intervention.

Keywords: amyotrophic lateral sclerosis (ALS), bulbar symptoms, motor speech disorder.

What this paper adds

What is already known on the subject

The speech of people with ALS can range from normal to a need for augmentative communication at the time of diagnosis. The interval from diagnosis may not be the best indicator to predict the rate of speech deterioration.

What this paper adds to existing knowledge

On average, 60% of the participants lost their ability to speak 18 months from the first bulbar symptom. It would be preferable to focus more on the time when the first bulbar symptoms appear than when the ALS diagnosis is confirmed to predict the rate of speech deterioration.

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What are the potential or actual clinical implications of this work?
More detailed knowledge on the progression of speech deterioration helps the SLTs to plan timely speech-therapy and communication-aid processes and to counsel people with ALS, their family members and healthcare professionals.

Introduction

Amyotrophic lateral sclerosis (ALS) is a progressive neurodegenerative disorder that destroys motor neurons in the cerebral cortex, brainstem and spinal cord (Kiernan *et al.* 2011). The disease affects both upper and lower motor neurons. Upper motor neuron involvement leads to spasticity, weakness and brisk deep tendon reflexes, while lower motor neuron manifestations include fasciculations, wasting and weakness. The first symptoms can appear anywhere along the motor tract. In classical ALS, muscle weakness spreads inevitably to encompass all voluntary muscles including respiratory muscles, leading to death within 2–5 years after the onset of symptoms (median survival time of 3 years) (Couratier *et al.* 2016). In approximately one-third of the persons with ALS, the disease starts with bulbar symptoms, and these people tend to have more severe type of motor speech disorder than those with initial spinal symptoms (Turner *et al.* 2010; Yorkston *et al.* 1993). Bulbar-onset disease and bulbar dysfunction overall is associated with poorer prognosis (Chio *et al.* 2009, Williams *et al.* 2013). Progressive loss of muscle control and strength, reduction in movement and increased duration of movements in the bulbar area lead to a gradual loss of speech function (Shellikeri *et al.* 2016,Yunusova *et al.* 2016).

Typically, the diagnostic process takes a long time from the onset of symptoms to a definitive diagnosis of ALS (Williams *et al.* 2013). The median delay from the initial symptoms to diagnosis is about 14 months and especially common in bulbar-onset disease (Kiernan *et al.* 2011; Turner *et al.* 2010). People with ALS demonstrate various speech symptoms at the time of diagnosis, ranging from normal speech to the use of augmentative communication (Yorkston *et al.* 1993). Particularly those with bulbar-onset ALS have obvious speech deterioration before a definitive diagnosis and the beginning of speech-therapy intervention (Makkonen *et al.* 2016). The speaking rate and intelligibility was observed to be lower in bulbar-onset than spinal-onset participants after the diagnosis (Ball *et al.* 2002).

Speech difficulties occur in 80–95% of people with ALS at some point during the progression of the disease, which weakens the functionality of natural speech and indicates the need for augmentative and alternative communication (AAC) methods (Beukelman *et al.* 2011, Creer *et al.* 2016). The loss of effective communication may result in difficult psychological and social problems (Simmons 2005) and also deterioration in quality of life (Felgoise *et al.* 2015). In fact, persons with ALS often consider the potential loss of speech as one of the worst aspects of the disease (Hecht *et al.* 2002).

There is limited follow-up research on the progression of dysarthria in ALS, and most follow-up studies have the time of diagnosis as a starting point (Hanson *et al.* 2011). In Dworkin and Hartman's (1979) case study dysarthria progressed from mild to severe in about 1.5 years and, similarly, anarthria appeared after 17 months in 49 bulbar-onset ALS patients in another longitudinal study by Turner *et al.* (2010). Sentence intelligibility declined from 96% to 10% in 7 months in Watts and Vanryckeghem's (2001) case study, and single-word intelligibility declined from high to low between 6 and 12 months in another multi-case study (Nishio and Niimi 2000). Average sentence intelligibility dropped from 95% to 75% in over a year (median of 455 days) during a follow-up of 66 ALS patients (Rong *et al.* 2016). Slower motor-speech performance has been shown to precede the decline in intelligibility with persons suffering from ALS (Ball *et al.* 2002, Green *et al.* 2013, Kent *et al.* 1991, Nishio and Niimi 2000, Rong *et al.* 2016, Turner and Weismer 1993). Prior work has suggested that instrumental-based measuring of speech, such as that of articulation rate, can detect early-onset bulbar symptoms of ALS and can be used for monitoring disease progression (Allison *et al.* 2017, Green *et al.* 2013, Yunusova *et al.* 2016).

In clinical work, more information about speech deterioration would be highly appreciated by people with ALS, their family members and healthcare professionals. One of the most common questions patients with ALS have asked during clinical visits is how long will they be able to speak. The main objective of this study was to assess how long natural speech will meet the patients' communication needs studied at three different time points: time since (1) the first bulbar symptoms, (2) the diagnosis of ALS and (3) the first speech and language therapist (SLT) evaluation, and to study the differences between the initial type, bulbar or spinal, of ALS. Another objective has been to identify the changes and the effects of different variables on severity of speech disorders, intelligibility and articulation rate.

Methods

Forty-seven consecutive participants were referred to the SLT in the Department of Neurology and Rehabilitation from August 2007 to December 2009 because of

Table 1. Demographic and clinical features of participants with amyotrophic lateral sclerosis (ALS)

Feature	All ($N = 30$)	Bulbar group ($n = 13$)	Spinal group ($n = 17$)	Significance between groups
Mean age at first SLT evaluation (minimum–maximum)	62.8 (31.1–83.0)	64.3 (44.3–76.6)	61.6 (31.1–83.0)	$p = 0.536$
Gender (male:female)	10:20	2:11	8:9	$p = 0.119$
Mean follow-up time, months (minimum–maximum)	15.3 (0–25)	16.7 (5–25)	14.3 (0–24)	$p = 0.385$
Mean time from bulbar symptoms to diagnosis, months (minimum–maximum)	10.6 (–5 to 50)	20.5 (8–50)	3.1 (–5 to 19)	$p = 0.001$
Mean time from bulbar symptoms to first SLT evaluation, months (minimum–maximum)	6.5 (–3 to 22)	11.2 (4–22)	3.0 (–3 to 13)	$p = 0.001$
Mean time from diagnosis to first SLT evaluation, months (minimum–maximum)	–4.1 (–34 to 8)	–9.4 (–34 to 1)	–0.1 (–7 to 8)	$p = 0.001$
Mean time from bulbar symptoms to last SLT evaluation, months (minimum–maximum)	21.87 (0–44)	27.8 (15–44)	17.3 (0–34)	$p = 0.007$

Note: SLT, speech and language therapist.

suspected or diagnosed ALS and one or more clinical signs of bulbar symptoms. Of these 47 participants, 30 met inclusion criteria: (1) native Finnish speaker; and (2) no other diseases affecting speech, language or swallowing. Of the 17 excluded patients, 10 had another diagnosed disease or condition affecting speech, language or swallowing such as stroke or Alzheimer's disease, three were excluded because they died before the diagnosis of probable or definitive ALS, and four declined to participate. All participants had normal hearing and adequate vision without or with eyeglasses. Clinically obvious cognitive dysfunction due to ALS was evident in five participants (four bulbar and one spinal-onset) as the disease progressed. All diagnoses were confirmed by a neurologist as probable or definitive ALS according to revised el Escorial criteria (Brooks *et al.* 2000) during the data collection by August 2011. 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 30 participants were divided into two groups based on the initial type of ALS: bulbar ($n = 13$) and spinal ($n = 17$). In four participants the initial type of ALS was originally determined as mixed, but as in two of them the major initial symptoms were bulbar and in the other two spinal, they were classified into these groups. The demographic and clinical features of the participants are presented in table 1. Females were more numerous in the bulbar group (11 females, two males), but no statistical differences in gender (Fisher's exact test) or age (Mann–Whitney *U*-test) between the groups were found. The participants were asked to determine the date (in months) when they first noticed bulbar symptoms (i.e., speech or voice changes, difficulty with tongue movement, nasal voice). Verification of onset of symptoms was requested from family or accompanying persons if possible. The time of adequate speech was counted in months from (1) bulbar symptoms, (2) diagnosis of probable or definitive ALS and (3) the first SLT evaluation. Time from the first bulbar symptom to the last SLT evaluation was counted in months.

The changes in various aspects of speech were recorded at the first SLT visit and on average every 3 months according to the participants' clinical needs. The objective of the study was to monitor each participant for 2 years. Due to the nature of the disease, 21 participants died due to respiratory insufficiency during the follow-up (table 2). Nine participants survived the whole 2 years: five in the spinal group and four in the bulbar group.

The functional change of speech was evaluated using the ALS Severity Scale of Speech (Hillel *et al.* 1989, Yorkston *et al.* 2004). On this 10-point scale a speech score of 10 represents normal speech and speech score of 1 indicates loss of useful speech limited to occasional vocalizations. Speech scores 5–10 represent adequate speech, meaning that natural speech remains a functional means of communication despite possible deterioration in motor speech production. A score of 4 or less signifies poor speech and the need for AAC to supplement or replace natural speech.

To estimate the speed of articulatory movements in spontaneous speech, the participants were asked to generate a story based on a wordless cartoon strip. Articulation rate was measured as a spoken narrative in syllables per total narration time excluding silent or filled (e.g., 'um', 'er') pauses of 200 ms or longer (Nishio and Niimi 2000, Turner and Weismer 1993) and reported as syllables per second. The stories were recorded with

Table 2. Cumulative mortality

	Cumulative mortality per cent every 3 months							
	3	6	9	12	15	18	21	24
Bulbar ($n = 13$)	0	0	7.7	30.8	38.5	53.8	61.5	69.2
Spinal ($n = 17$)	5.9	11.8	29.4	35.3	47.1	52.9	58.8	70.6
All ($N = 30$)	3.3	6.7	20.0	33.3	43.3	53.3	60.0	70.0

a headset microphone, saved as WAV files directly with Sound Forge software and transcribed orthographically for articulation rate analysis. The PRAAT program was used to verify acoustically the syllable boundary. If the participant was unable to speak clearly enough for syllable boundaries to be detected, the articulation rate was scored as 0.

The intelligibility of semi-spontaneous speech in the cartoon narrative task was evaluated perceptually by seven experienced SLTs on a 100 mm visual analogy scale (VAS). A VAS value of 0 (0 mm) equals non-intelligible and 100 (100 mm) fully intelligible speech. The mean VAS was reported in the evaluation. All seven SLT listeners rated every speech sample entirely in random order. The listeners had the possibility to listen to the samples as many times as required to be confident about their rating. The inter-judge reliability between the listeners was good (intraclass correlation (ICC) = 0.896). The median across-judge difference was 5.5 ($Q_1 = 1$ and $Q_3 = 20.75$, minimum = 0 and maximum = 84). If a person with ALS was unable to produce any speech in the narrative task, the intelligibility of spontaneous speech was scored as 0.

In the group comparison of the participants' demographic and clinical features at the first visit, the non-parametric Fisher's exact test was used for categorical variable (gender) and the Mann–Whitney *U*-test was used for continuous and interval variables. These statistical analyses were performed using SPSS (version 23; SPSS Inc.).

The severity of symptoms and communication abilities differed between patients and the data was also unbalanced, i.e., there was a varying number of measurements for individual patients. Therefore, linear mixed-effects (LME) models with a symptom or score value as a dependent variable were fitted using function LME in R (Software Environment for Statistical Computing and Graphics, version 3.3.0; The R Foundation for Statistical Computing). Time, age at the first evaluation, gender and initial type of ALS were used as independent variables. Additionally, the interaction effect of time and initial type of ALS was analysed. A likelihood ratio test was used to compare models. A random intercept was used together with independent random errors. A p-value < 0.05 was considered to be significant.

A total of 157 communication assessments were performed in this study. LME model analysis of the data was carried out with actual information on evaluation dates for all 30 participants. The data collected on a speech scale were organized into 3-month intervals ($\pm$ 1 month) to describe the changes in time. If there were two assessments during the same interval (as in 11 cases), the latter was included in the study.

Results

Speech deteriorated to poor, necessitating the use of AAC methods (a score of 4 or less) in 60% of the participants (18/30) during the study. The speech score was 4 or less in all bulbar-onset participants at the last evaluation. Speech was functional at the first visit with all spinal-onset participants and with 29% (5/17) of them speech deteriorated to poor by the time of the last evaluation. With those 18 participants whose speech score decreased to 4 or less, speech remained adequate on average for 18 months from the first bulbar symptoms and no statistical difference between the two groups was found (table 3).

Three bulbar-onset participants had no functional speech at the beginning of the follow-up, and after the first year only one participant was able to speak adequately. In contrast, the first participants in the spinal group to lose functional speech were identified after 9 months of follow-up. At the end of the 2-year follow-up, three spinal-onset ALS participants—out of the nine remaining—were still able to speak sufficiently.

Time and initial type of ALS were significant factors ($p = < 0.001$) in all LME models (articulation rate, intelligibility and speech score) (table 4). Overall, participants with spinal-onset ALS performed better in all these tasks than persons with bulbar-onset disease (figures 1a–c). Age or gender were not significant in LME model. Interaction effect between the time and initial type of ALS was significant only for speech intelligibility ($p = 0.0043$) (table 4).

Discussion

This study has focused on the progression of motor speech symptoms in persons with ALS. The main finding was that the ability to speak was lost in 60% of

Table 3. Time of decrease of speech to score 4 or less

Initial type of ALS	Mean time to poor speech, months (range) from: Bulbar symptoms	ALS dg	First SLT evaluation
Bulbar ($n = 13$)	18.0 (6–40)	–2.5 (–15 to 8)	–6.8 (0–24)
Spinal ($n = 5$)	19.6 (12–27)	11.4 (3–25)	14.2 (10–20)
All ($n = 18$)	18.4 (6–40)	1.3 (–15 to 25)	8.9 (0–24)
Significance between groups	$p = 0.503$	$p = 0.003$	$p = 0.014$

Note: SLT, speech–language therapist.

Table 4. Linear mixed-effects (LME) model estimates

		Estimates without interaction effect			Estimates with interaction effect between time and initial type of ALS		
		Beta	SE	*p*-value	Beta	SE	*p*-value
Articulation rate	Intercept	3.49	1.12	**0.0023**	3.52	1.12	**0.0021**
	Time	–0.0050	0.00075	**< 0.001**	–0.0054	0.0011	**< 0.001**
	Gender (male)	0.77	0.43	0.084	0.78	0.43	0.083
	Age	–0.018	0.016	0.29	–0.018	0.016	0.30
	Initial type of ALS (spinal)	2.27	0.37	**< 0.001**	2.18	0.41	**< 0.001**
	Time∗spinal				0.00075	0.0015	0.62
Intelligibility	Intercept	70.76	23.51	**0.0031**	74.57	23.50	**0.0019**
	Time	–0.11	0.019	**< 0.001**	–0.16	0.023	**< 0.001**
	Gender (male)	8.06	8.98	0.38	8.58	9.01	0.35
	Age	–0.063	0.34	0.86	–0.016	0.35	0.96
	Initial type of ALS (spinal)	40.42	7.79	**< 0.001**	27.35	8.85	**0.0047**
	Time∗spinal				0.095	0.03	**0.0043**
Speech score (ALSSS of speech)	Intercept	4.36	1.45	**0.0032**	4.39	1.46	**0.0031**
	Time	–0.0081	0.00090	**< 0.001**	–0.0085	0.0013	**< 0.001**
	Gender (male)	0.91	0.56	0.12	0.91	0.56	0.12
	Age	0.012	0.021	0.57	0.012	0.021	0.57
	Initial type of ALS (spinal)	2.89	0.49	**< 0.001**	2.83	0.51	**< 0.001**
	Time∗spinal				0.00070	0.0018	0.70

Note: ALSSS = ALS severity scale; 1–10.

the participants during the 2-year follow-up. In these persons, speech remained adequate and communication functional on average for 18 months after the appearance of the first bulbar symptoms regardless of the initial symptom type, whether bulbar or spinal. A participant's speech varied from fully functional speech to the need for AAC measures when assessed at the time of ALS diagnosis or the first speech therapy evaluation. The beginning-point of bulbar symptoms reported by the participant appeared to indicate the duration of adequate speech function better than the time point of ALS diagnosis or of the first SLT evaluation.

In the present study, dysarthria progressed from mild to severe in a similar timeframe as in the previous studies in bulbar-onset participants (Dworkin and Hartman 1979, Turner *et al.* 2010). In their retrospective database study, Yorkston *et al.* (1993) reviewed speech deterioration in 44 ALS participants post-diagnosis and found variable changes in speech both in bulbar- and spinal-onset participants. These changes occurred earlier in bulbar-onset participants. Our study confirms the earlier results. In the present prospective study we used similar methods such as speech score, speech intelligibility and articulation rate, but unlike in the previous studies we focused on the starting point of bulbar symptoms rather than confirmation of diagnosis.

While all bulbar-onset participants lost their ability to speak during the follow-up, only five (29%) in the spinal group had non-functional speech at the end of the follow-up. Speech performance at the first SLT visit was more decreased in the bulbar-onset than in the spinal-onset participants (figures 1a–c). The monitoring time from the first bulbar symptoms to the last SLT visit (either at the end of the 2-year follow-up or before the patient passed away) was on average 10.5 months shorter in the spinal-onset group (table 1). It is possible that in some spinal-onset patients the disease progresses to encompass respiratory muscles and is fatal before the major decline in speech appears. This indicates that even though the progression of motor speech symptoms is a

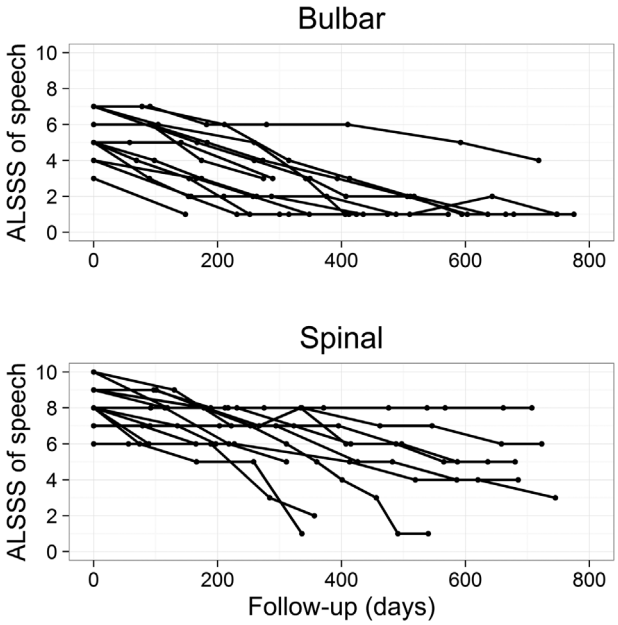

Figure 1a. Decline in speech scores of individuals in the bulbar (13) and spinal (17) groups.

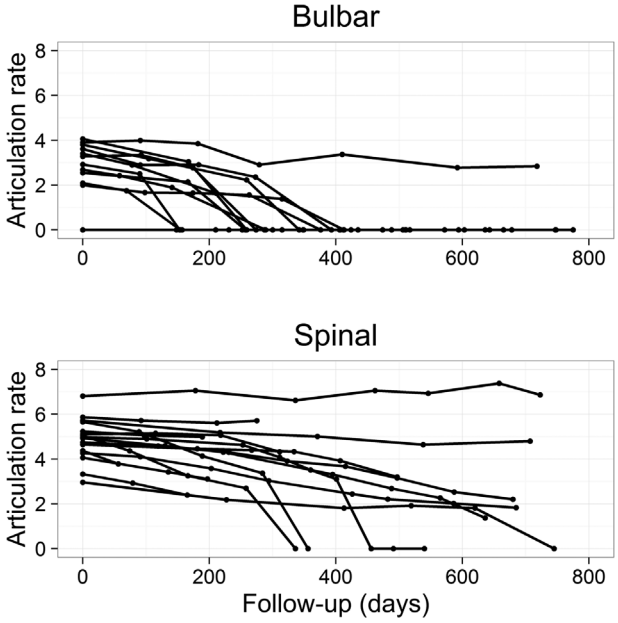

Figure 1b. Decline in articulation rate of individuals in the bulbar ($n = 13$) and spinal ($n = 17$) group.

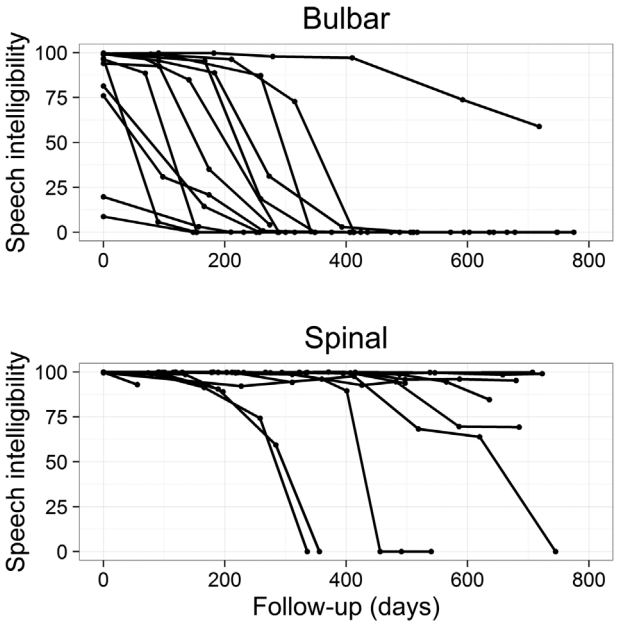

Figure 1c. Decline in intelligibility of narrative speech of individuals in the bulbar (13) and spinal (17) groups.

significant part of ALS, not all participants need AAC during the course of the disease.

The information on how fast speech will worsen is important in clinical work as it helps the SLT to plan timely speech therapy and communication aid processes and to counsel people with ALS and family members. The diagnosis of ALS is based on clinical criteria and an accurate diagnosis is typically delayed from the appearance of first symptoms (Williams *et al.* 2013, Kiernan *et al.* 2011, Turner *et al.* 2010). According to the present data the people with bulbar-onset ALS lost their functional speech on average 7 months after the first speech therapy intervention, rendering a mere 7 months to organize individually appropriate AAC. People with spinal-onset ALS are referred to the speech therapist sooner than bulbar-onset patients because they are usually already being regularly monitored for their other symptoms. Thus, there is substantially more time (on average 14 months) to organize communication aid services for them.

Speech score, speech intelligibility and articulation rate declined in nearly all participants as expected (Ball *et al.* 2002, Green *et al.* 2013, Hillel *et al.* 1989, Kent *et al.* 1991, Nishio and Niimi 2000, Rong *et al.* 2016, Turner and Weismer 1993, Watts and Vanryckeghem 2001). Time and onset type of ALS were the most significant factors contributing to the motor speech functions. Our results are compatible with earlier results in that people with initial bulbar symptoms tend to have more severe motor speech disorders than those with initial spinal symptoms (Turner *et al.* 2010, Yorkston *et al.* 1993).

Neither age nor gender were significant in relation to the decline in speech functions. In earlier studies, female gender and higher age were associated with more severe bulbar dysfunction and with greater probability of progression to anarthria (Turner *et al.* 2010, Yorkston *et al.* 1993). Females were more numerous in the bulbar group. However, no reliable conclusions can be made based on gender in our study.

The interaction effect between time and initial type of ALS was not significant for speech score or articulation rate analysed by LME, suggesting that most speech symptoms progress similarly, once started, irrespective of the initial type of ALS. Speech intelligibility declined more in bulbar-onset participants during the follow-up, as shown in 1c, although the level of speech intelligibility was lower at the first SLT visit. A possible explanation is that nearly all bulbar-onset participants had unintelligible speech at the last evaluation against only four in the spinal-onset group. Another explanation might be that intelligibility decreases at a slow rate and intelligibility measurements, therefore, lack sensitivity in the early stages of speech disorders. During the later stages of the disease intelligibility declines rapidly and progresses to the loss of adequate speech in quite a short time (Ball *et al.* 2001, Green *et al.* 2013, Nishio and Niimi 2000, Rong *et al.* 2016, Yorkston *et al.* 1993). The progression of speech intelligibility declining after the first manifestation of bulbar symptoms has been reported in a limited number of studies (Nishio and Niimi 2000, Rong *et al.* 2016, Watts and Vanryckeghem 2001). Our results are similar, even though the language of our participants and the methods used for rating intelligibility were different, making comparisons with earlier studies difficult. We rated intelligibility from semi-spontaneous speech in the cartoon narrative task while in the earlier studies intelligibility has been rated from single word or sentence reading tasks.

Collecting clinically valid follow-up data from participants with ALS is challenging, owing to the progressive and fatal nature of the disease. Participants are unavoidably lost during a long follow-up, and their number varies between different observation points. Therefore, LME models were used to analyse longitudinal data in this study. The selection of the measurement tools is also challenging as there is a need to reach both the early and the longitudinal changes in motor speech production. The severity scale of speech is not sensitive enough to detect the early changes in motor speech function (Allison *et al.*, 2017), but the scale still indicates a need for AAC intervention (Yorkston *et al.*, 1993). Even a minor decline in intelligibility again indicates loss of adequate speech (Ball *et al.* 2001, Green *et al.* 2013, Nishio and Niimi 2000, Rong *et al.* 2016, Yorkston *et al.* 1993), and can no longer be measured due to the loss of spoken communication. Articulation rate is more sensitive for early changes (Allison *et al.* 2017, Green *et al.* 2013, Yunusova *et al.* 2016), the syllable boundaries need to be extractable from spoken words to be measured and, therefore, become unmeasurable when speech is no longer functional. As in this study the challenging choice of measures may result in floor effect, because the variables related to speech cannot be assessed.

We observed speech deterioration prospectively from the first speech assessment, which is the earliest point in our healthcare system where communication ability is evaluated. We collected information on speech and communication changes in people with symptoms of ALS but with yet uncertain diagnosis. To ensure the changes in speech and communication were due to ALS, only those participants whose ALS diagnosis was confirmed as probable or definitive during the data collection were included in the final analyses.

One of the limitations of this study is the low number of participants. In the spinal group, a relatively small number of participants (five) lost their ability to speak adequately. This might reflect on some statistical findings such as the time to need for AAC. Other limitations were the bias in gender, the potential bias in the participant's recall of bulbar symptom-onset point and lack of formal cognitive assessment. Speech performance in the five participants with signs of cognitive dysfunction did not differ from the other participants. Intelligibility and articulation rate were determined from semi-spontaneous speech in the cartoon narrative task that does not require reading skills. The professional listeners were instructed to rate intelligibility solely based on the aspects of motor performance, as articulation rate without pauses measures bulbar motor symptoms well without the effect of possible cognitive or respiratory dysfunction (Green *et al.* 2013, Yunusova *et al.* 2016). A formal screening or assessment of cognitive abilities should be performed in future studies.

Conclusions

Motor speech function declined inevitablyin most of the participants and they lost adequate speech on average in 18 months from the first bulbar symptoms. In clinical work when assessing the need for AAC procedures, it would be preferable to focus more on when the first bulbar symptoms appear than when the ALS diagnosis was confirmed or the first SLT evaluation was undertaken. The motor speech function of persons with bulbar-onset ALS is probably more deteriorated than that of those with spinal-onset at the first speech therapy assessment. This indicates a rapid need for speech evaluation and AAC services.

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Variation in communication strategies in amyotrophic lateral sclerosis during a two-year follow-up

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ABSTRACT

We report on how functional capacity of communication and the use of augmentative and alternative communication (AAC) technology changed in persons with amyotrophic lateral sclerosis (ALS) and progressive dysarthria during a period of two years.

The initial sample comprised 30 participants, nine of them survived the whole two years. The data was gathered in 157 communication assessments by a speech-language pathologist. Functional capacity of communication was evaluated with combined severity scales of speech and upper and lower limb function. Communication strategies used were gathered based on the participants' own reports, and their use was confirmed during the clinical visits.

A total of 57% of the participants either supported or substituted their natural speech with high- or low- technology strategies. The majority of them reported that they used high-technology rather than low-technology AAC as the main communication means after speech had deteriorated. The classification of functional capacity of communication changed in 63 % of participants, more in bulbar than spinal-onset ALS as the disease progressed.

Natural speech is a strong means of communication and not effortlessly replaceable. However, AAC means, such as handwriting, text-to-speech devices and computer-based devices, are

essential for persons with ALS as their speech deteriorates. Several AAC strategies to augment deteriorated speech and replace natural speech are often needed. Functional capacity of communication and its individual changes along the disease progression must be taken into account during the continuing communication aid processes.

Keywords: communication, augmentative and alternative communication, AAC, amyotrophic lateral sclerosis, ALS, functional capacity of communication, bulbar symptoms, follow-up study

INTRODUCTION

Amyotrophic lateral sclerosis (ALS) is a progressive neurodegenerative disease which affects motor functions (Kiernan et al., 2011). In most cases the disease starts with a combination of upper and lower motor neuron symptoms in the limb, and in about one third of patients the disease starts with bulbar symptoms affecting speech function (Kiernan et al., 2011; Duffy, 2013). Lower motor neuron involvement leads to fasciculations and muscle weakness and wasting, while upper motor neuron manifestations include spasticity, weakness, and brisk deep tendon reflexes. Therefore, ALS patients will typically demonstrate a flaccid-spastic mixed dysarthria (Duffy, 2013). Progressive loss of motor speech function results in progressive loss of muscle strength and control, increased duration of movements and reduction in bulbar muscle movements (Shellikeri et al. 2016, Yunusova et al. 2016). A rapid change in speech production is possible (Nishio & Niimi, 2000; Turner et al., 2010; Watts & Vanryckeghem, 2001). As ALS progresses the ability to speak declines in nearly all patients irrespective of the initial type of the disease (Tomik & Guiloff, 2010). It is reported that dysarthria occurs in 80-95% of persons with ALS at some point during the disease (Beukelman, Fager, &

Nordness, 2011). Progressive dysarthria weakens the ability to meet daily communication needs using natural speech.

Progressive loss of intelligible speech is the ultimate result of mixed dysarthria (Duffy, 2013; Hanson, Yorkston & Britton 2011). The loss of effective communication may have devastating psychological and social consequences and debilitating impact on overall quality of life in ALS (Simmons, 2005; Felgoise, Zaccheo, Duff, & Simmons, 2015). When the intelligibility of natural speech decreases, speech should be combined with alternative or augmentative approaches to communication (AAC) (Hanson et al., 2011). It has been presented that 72% of persons with ALS could benefit from AAC (Creer, Enderby, Judge, & John, 2016). The speech-language pathologist (SLP) plays a key role in helping patients and their caregivers to realize that other forms of communication may be needed to substitute speech (Simmons, 2005). With the help of AAC a person with ALS and dysarthria will be able to maintain the ability to communicate and improve or at least stabilize his/her quality of life (Körner et al., 2013; Londral, Pinto, Pinto, Azevedo, & De Carvalho, 2015) and also in some cases to maintain the possibility of working longer (McNaughton, Light, & Groszyk, 2001; Ball et al., 2007). Thus, a regular assessment (i.e. every 3–6 months) of speech and language function by a trained SLP is recommended for ALS patients (Andersen et al., 2012).

AAC strategies can be classified into three categories: 1) high-technology 2) low-technology and 3) non-technology strategies (Beukelman, Yorkston, & Garret, 2007). Speech-generating devices (SGDs) consist of high-technology systems that produce speech output either in the form of prerecorded messages or with text-to-speech programs. SGDs may be used with keyboards or different switches even with minimal motor function, for example by head or eye control. SGDs can be solely for AAC or integrated with other computer functions and applications (Fried-Oken, Mooney, & Peters, 2015). Low-technology strategies refer to communication methods like alphabet boards, printed words, writing, drawings or picture boards. Examples of AAC strategies that do not require technology are gestures, vocalization and eye movements or blinks. Persons with

ALS will typically communicate with a combination of non-technology, low-technology and high-technology solutions (Ball, Beukelman, & Bardach, 2007b). Often low- and high-technology solutions based on writing ability (for example hand writing, keyboard access, or alphabetic board).

In addition to speech, upper and lower extremity function of the patient should be taken into account when evaluating communicative options and selecting appropriate AAC means (functional capacity of communication; Yorkston, Miller, & Strand, 2004). Even with poor speech a patient can communicate functionally by writing and/or typing if hand functions persist. When both functions, hand and speech, are lost, AAC solutions with minimal hand or leg movement (like buttons and joysticks) or even only eye or head movements are needed. In such cases, the patient might need assistance in using AAC strategies or in preparing the devices for use. People with spinal-onset ALS have been found to use more partner-dependent techniques than those with bulbar-onset ALS regarding the differences in upper extremity functions (Mathy, Yorkston, & Gutmann, 2000). If a person's mobility is good, easily portable AAC means may be utilized. When mobility declines, AAC means should be usable while sitting in a wheelchair or lying in bed. Changes in physical and communication needs during the natural course of the disease must also be considered when planning an AAC intervention (Fried-Oken et al., 2015). Sometimes issues related to the person (e.g. desire to use one's own speech), device (e.g. complex learning requirements) or professional input (e.g. lack of training) influence the patient's choice not to use available technological devices (Baxter, Enderby, Evans, & Judge, 2012; Murphy, 2004). It is possible that at the later stages of the disease, ALS patients cannot communicate at all (Brownlee & Bruening, 2012).

Patients and their families need information on potentials of various AAC strategies and timely AAC interventions. The aim of this study has been to characterize how the functional capacity to communicate and the usage of AAC strategies change in patients with ALS as the disease progresses. We will focus on how high- and low-technology communication strategies are used to support diminishing speech.

MATERIALS AND METHODS

Subjects

Forty-seven consecutive patients were referred to the SLP at Tampere University Hospital, Department of Neurology and Rehabilitation due to suspected or diagnosed ALS and one or more clinical signs of bulbar deterioration from August 2007 to December 2009. The diagnosis and the initial type of the disease (bulbar or spinal) were determined by an experienced neurologist (second author). Of these 47 referred patients 30 met inclusion criteria: 1) native Finnish speaker; 2) diagnosis of suspected, possible, probable or definitive ALS according to revised el Escorial criteria (Brooks, Miller, Swash, Munsat, & World Federation of Neurology Research Group of Motor Neuron Disease, 2000) at the first SLP visit; 3) no other diseases affecting speech, language or swallowing and 4) the final diagnosis of probable or definitive ALS reached during data collection by August 2011. The study protocol was approved by the Ethics Committee of Pirkanmaa Hospital District and all patients gave their written informed consent.

The Department of Neurology and Rehabilitation has a multidisciplinary team for ALS patients that includes an SLP in integrated practice and an AAC specialist (SLP) in the Assistive Technology Unit. The latter primarily provides AAC intervention services. All ALS patients have an equal possibility for AAC intervention funded by public special-health care.

The 30 participants were divided into two groups based on the initial type of ALS: bulbar (n=13) and spinal (n=17). In four participants the initial type of ALS was originally determined as mixed, but two of them were classified into the bulbar group and two of them into the spinal group based on the major initial symptoms.

Data collection

A total of 157 assessments of functional capacity of communication (FCC) and communication strategies were performed in this study. The study aimed at monitoring each participant for two years,

and they were assessed averagely every three months according to their clinical needs. Owing to the natural course of the disease 70% (n=21) of the participants died during the follow-up (Table 1). Nine participants (30%), four in the bulbar and five in the spinal group, survived the whole two years. The data was grouped into three-month (+/- one month) intervals based on the timing of the assessments. If there were two communication assessments during the same interval, as in 11 cases, the latter assessment was included in this study.

Table 1. Cumulative survival

	Cumulative survival (percentage) in 3 months interval							
Initial type of ALS	3	6	9	12	15	18	21	24
Bulbar (n=13)	100	100	92.3	69.2	61.5	46.2	38.5	30.8
Spinal (n=17)	94.1	88.2	70.6	64.7	52.9	47.1	41.2	29.4
All (N=30)	96.7	93.3	80.0	66.7	56.7	46.7	40.0	30.0

Methods

In this study we focused on functional aspects of communication and AAC methods needed with declining speech function. Changes in motor speech function have been reported earlier (Makkonen, Ruottinen, Puhto, Helminen, & Palmio, 2017 *submitted)*. FCC was classified into six different categories using ALS Severity Scale of speech and upper and lower extremities: 1) adequate speech and adequate hand function, 2) adequate speech and poor hand function, 3) poor speech, adequate hand function and adequate mobility, 4) poor speech, adequate hand function and poor mobility, 5) poor speech, poor hand function and good mobility and 6) poor speech, poor hand function and poor mobility (Hillel et al. 1989; Yorkston, Strand, Miller, Hillel, & Smith,1993; Yorkston et al., 2004). Speech function was poor if the participant needed or used means of AAC or had lost useful speech (speech scale score 4 or less). Hand function was poor if the participant needed assistance in self-care or was totally dependent on assistance (upper extremity scale score 4 or less). If the participant walked

with assistance or showed lowered capacity of leg movement, mobility was classified as poor (lower extremity scale score 6 or less). Rating was determined on speech and communication assessment and clinical observation made by the SLP at the outpatient clinic or neurological ward.

The communication methods used by the participants were listed according to their own report, and their use was confirmed during the clinical visits. The participants reported their methods by speaking and/or by using AAC means. If the participant reported that he/she used more than one communication strategy, the primary communication method was asked to be named or to be pointed by hand or gesture. AAC methods that were used only in actual communication were noted in the study, not those used for training. As we focused on speech changes, communication support needed only for poor hand function is not included. Communication methods were categorized into speech or two AAC strategies: high technology and low technology. High-technology devices were categorized into SGDs for AAC purposes only (SGDs) and into computer-based devices (computer-based SGDs) when all computer functions and applications were available. AAC strategies that involved no technology were not included.

RESULTS

Twenty of the participants were female and 10 male. The mean age was 62.8 (31.1-83.0) years (bulbar 64.3 and spinal 61.6). No statistical gender (Fisher exact test p=.119) or age (Mann-Whitney U-test p=.536) differences between the groups were found. The mean follow-up time was 15.3 months (bulbar 16.7 (5-25), spinal 14.3 (0-24), one participant with spinal-onset ALS had one evaluation) and no statistically significant difference between the groups was found (Mann-Whitney U-test p=.385).

Primary communication methods

At the beginning of the follow-up 29 (96.7%) participants could speak adequately while one participant with bulbar-onset ALS communicated mainly by handwriting. During the first nine months, the main communication method was speech but from six months on an increasing number of participants started to use other communication methods. After one year and till the end of the follow-up approximately half of the remaining participants communicated primarily with AAC.

Primary communication methods are described in Figure 1. At the first visit one participant in the bulbar group communicated with low-technology but no one with high-technology AAC. At 21 and 24 months' observation points high- and low-technology AAC means were used by equally many participants. At all other observation points the participants used more often high than low technology. At the end of the follow-up, five of the remaining nine participants communicated primarily with natural speech, two preferred low-technology strategies and another two high-technology AAC.

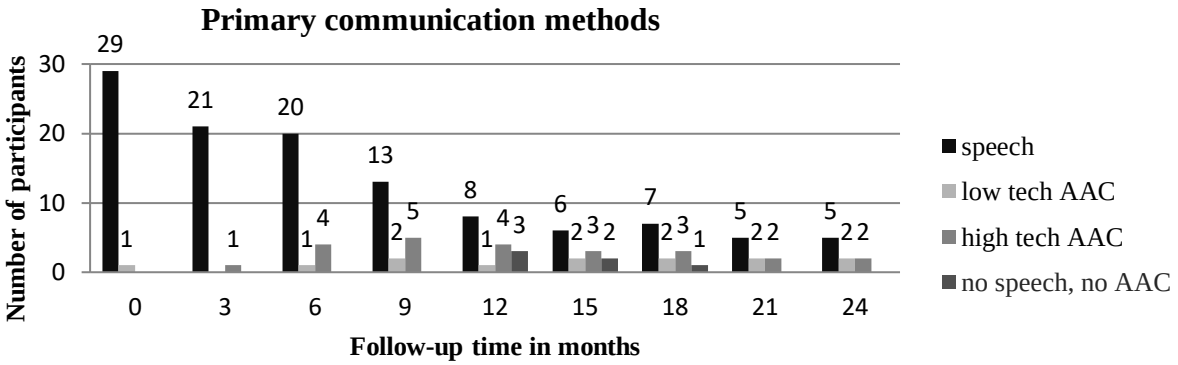

Figure 1. Primary communication methods used at three-month interval.

Changes in the primary communication strategies of all 30 participants during the study period are illustrated in Figure 2. A total of 14 participants (10 in the bulbar group, 4 in the spinal group) used low- or high-technology AAC to substitute speech at some point of the follow-up. A

further 14 participants (1 in the bulbar group, 13 in the spinal group) primarily used speech throughout the study period. Two participants in the bulbar group could not use any of the AAC means as primary communication strategy after a loss of effective speech. The participants in the bulbar group used five different primary communication strategies while in the spinal group they used only two.

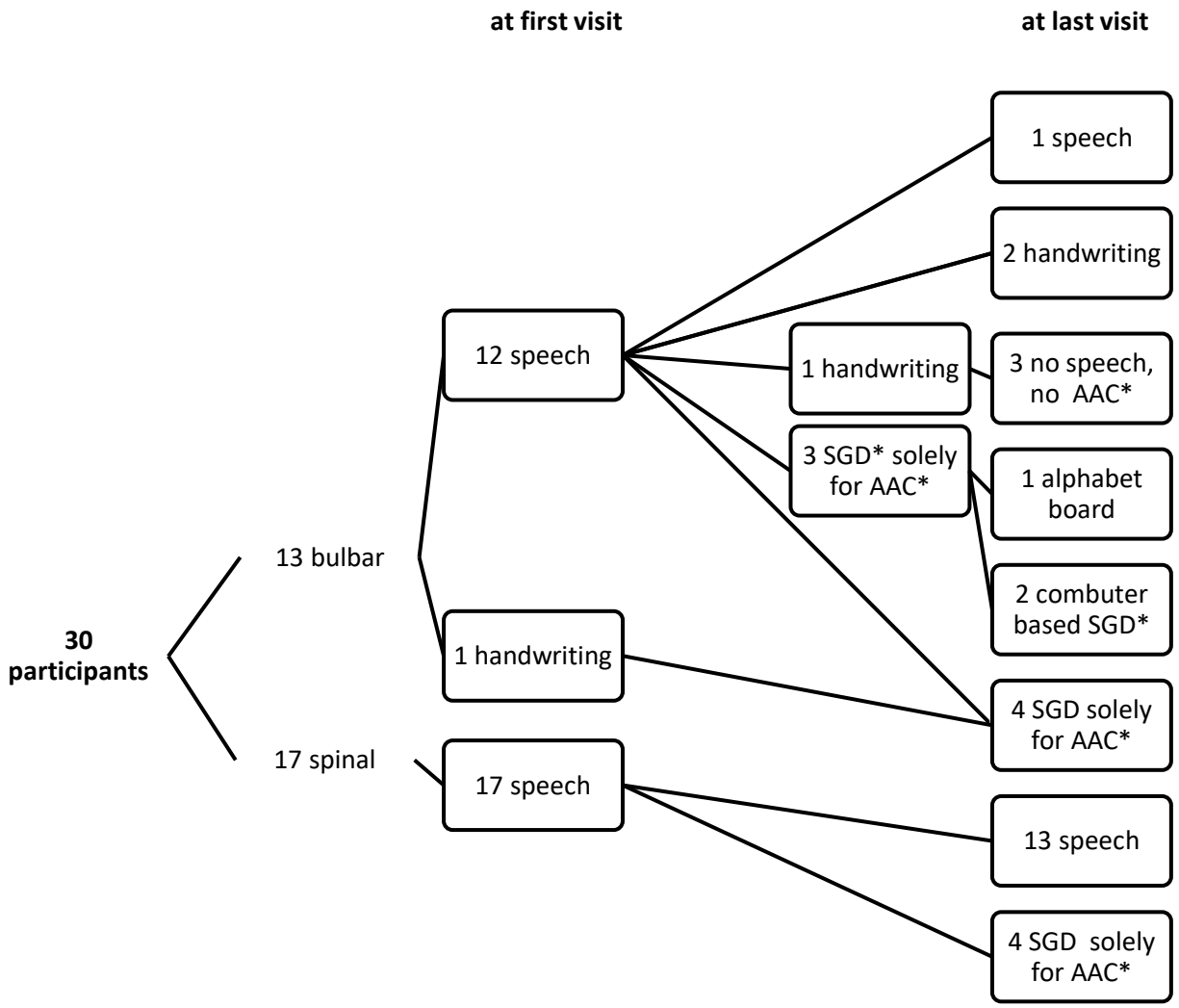

Figure 2. Modification of primarily communication strategy during the study period. The number of participants using each communication strategy is indicated in the box.

*AAC= augmentative and alternative communication; SGD= Speech-generating device

Three participants in the bulbar group lost both their ability to speak and their ability to use low- or high-technology AAC (Figure 2). Two of them communicated first by combining speech and handwriting (one primarily by speech, other primarily by handwriting), but after one year they lost both abilities. One participant did not use any AAC means, only decreased speech. All these three participants suffered from cognitive impairment affecting their everyday life.

Secondary communication strategies in addition to speech

Altogether 11 out of 30 participants augmented their natural speech with high- or low-technology communication strategies. Eight participants in the bulbar group compensated decreased speech with handwriting and /or SGD (3 handwriting, 4 text-to-speech device, 1 handwriting and text-to-speech device). Three participants in the spinal group supplemented natural speech with handwriting, with alphabetic board or various combinations of voice amplifier, handwriting, word/sentence list or SGD. Later, the secondary communication strategy became primary for eight participants (6 in the bulbar group, 2 in the spinal group).

Secondary communication strategies in addition to low- or high-technology communication

All seven participants in the bulbar group who primarily communicated with SGDs supported their communication with residual speech and/or handwriting. After both became impossible, two of them supported their communication with alphabetic board and/or computer based SGDs. Four participants in the spinal group operated with a combination of handwriting and word/sentence list, alphabetic board and computer based speech device, residual speech and computer-based speech device or handwriting and residual speech.

Four participants who communicated with handwriting at some point of the follow-up supported their communication with residual speech or with SGD. Those who communicated primarily with computer-based SGDs also used alphabetic board, other SGDs or word/sentence lists.

One participant used SGD secondarily to alphabetic board. Altogether 12 participants communicated with residual speech in addition to low- or high-technology strategies at some point of the study period.

Communication strategies used

Seventeen participants (12 in the bulbar, 5 in the spinal group) substituted speech with at least one low- or high-technology AAC strategy. Twelve participants in the bulbar group used one to four communication means at a time, and the number of different strategies used during the whole follow-up varied from two to seven. Five participants in the spinal group who needed AAC used two to four different AAC methods at a time, and the total number of communication strategies varied from two to five during the study period.

Functional capacity of communication

The category of FCC classification changed in 63 % of the participants as the disease progressed. The changes occurred in two phases (Table 2). During the first year, the mode of FCC changed from categories 1 (adequate speech, good hand function) or 2 (adequate speech, poor hand function) to category 6 (poor speech, poor hand function, poor mobility). At the beginning of the second year, the remaining participants' mode of functional capacity was category 2, and towards the end of the second year the mode changed to category 6. Poor speech categories 3 (poor speech, adequate hand function, good mobility) and 5 (poor speech, poor hand function and good mobility) were found only in the bulbar group, while categories 4 (poor speech, adequate hand function and poor mobility) and 6 were seen in both ALS groups.

Table 2. Changes in modes of functional capacity of communication (FCC) every three months

months	0	3	6	9	12	15	18	21	24
N	30	22	25	20	16	13	13	9	9
FCC* mode all	1	1	2	2	6	2	2&6	6	6
n bulbar	13	10	12	12	7	6	6	4	4
FCC* mode bulbar	1	1	3	3	3&6	3	6	6	6
n spinal	17	12	13	8	9	7	7	5	5
mode spinal	1	2	2	2	1&2&6	2	2	2	1

*FCC categories: 1 =adequate speech, good hand function; 2 =adequate speech, poor hand function; 3 =poor speech, adequate hand function, good mobility; 4 =poor speech, adequate hand function and poor mobility; 5 =poor speech, poor hand function and good mobility and 6=poor speech, poor hand function, poor mobility.

A change of FCC was more typical with bulbar-onset (12/13) than with spinal-onset ALS (7/17) (92% and 41%, respectively). In the bulbar group, FCC converted from good (1-2) to poorer categories (3-6) with ten participants. With two of them, the FCC changed from 3 to 5 (n=1) or 6 (n=1), and one participant's FCC (category 3) remained stable throughout the study period. In the spinal group, the FCC category changed from adequate (categories 1-2) to poor (categories 4-6) with 29% (5/17) of the participants. With most of the participants (59%) in the spinal group the FCC stayed in adequate speech categories, either 1 or 2.

DISCUSSION

The study focused on changes in both communication strategies and the functional capacity of communication (FCC). After six months of the first visit to a speech-language pathologist, over 50% of the participants needed different means of AAC to compensate for deteriorated speech and preferred high- to low-technology strategies. The participants increased their use of AAC methods as their natural speech continued to decline. FCC also decreased from good speech and good hand function to poor speech, poor mobility and poor hand function during the follow-up, but there were differences according to the initial type of ALS.

A total of 57% of the participants either supported or substituted their natural speech with high- or low- technology strategies during the study period. This is less than in earlier reported studies in which most of the ALS patients could not adequately communicate with natural speech (Beukelman et al., 2011) or became unable to speak at all (Ball et al., 2007b) as their disease progressed. This is also a smaller proportion compared to the 72% of participants who have been shown to benefit from AAC (Creer et al., 2016). Our results are, however, in line with previous reports regarding bulbar-onset ALS according to which most of the ALS patients with initial bulbar symptoms became unable to communicate with natural speech (Ball et al., 2007b; Beukelman et al., 2011). The difference in the results may be due to the finding that, in this prospective study, half of the participants had spinal-onset ALS, and most of them retained their ability to speak (FCC categories 1-2) even during the final months of their lives. Our results may also be explained by the source of the data as it is based on consecutive patients followed-up at the neurological unit. The situation might be different for example in an AAC unit where the patients are referred to with already poor speech and evident need of AAC. Furthermore, very few ALS patients in Finland generally have and none in this study had supported invasive ventilation with tracheostomy. Invasive ventilation support is known to affect both the AAC technology chosen and the duration of its use (Ball et al., 2007) and therefore might contribute to the results.

The participants in this study more commonly used high- than low- technology AAC when their speech was no more functional. There was a difference between the groups based on the initial type of ALS. The participants in the bulbar group used either low- or high-technology solution (the most common being handwriting or text-to-speech device) as the primary communication method while the spinal group used only high-technology devices (most frequently text-to-speech device). The variation may be explained by differences between the groups in relation to physical access to AAC technology, especially regarding upper extremity functions (Mathy et al., 2000). All participants in the current study had equal access to low- or high- technology AAC solutions after the

first SLP visit with a possibility to get an applicable AAC device that utilizes hand, feet, head or eye movement control. For example, eye-tracking communication devices were available as they have been found to be useful at the late stages of ALS with tetraplegia and anarthria (Caligari, Godi, Guglielmetti, Franchignoni, & Nardone, 2013). None of the participants in this study, however, used them as the primary AAC means.

Speech is a powerful means of communication even when deteriorated. Some people with ALS primarily communicate with poor speech, as did four of our participants, and often even residual speech remains a part of the communication method as in 12 cases in this study. This is in line with the finding that some ALS patients have a desire to use their own speech for as long as possible, even when it is unintelligible to others (Murphy, 2004). This finding suggests that in some situations and for some people even poor speech is quicker and more effective, i.e., a more functional way to communicate compared to using AAC means. On the other hand, non-technology communication, for example gestures and facial expression, is a valuable way to communicate and support deteriorating speech. As their use is difficult to measure they were not included in the present study. Although the continuing development of technical solutions may make it possible to use even minimal functional ability to support communication, technical solutions are not an answer to all communication difficulties and not to all people.

Natural speech is difficult to replace. It was noted that several strategies were needed to support deteriorated speech and to substitute it. When speech started to deteriorate handwriting and/or SGDs were common options. Handwriting is a suitable strategy in face-to-face situations for people with intact linguistic abilities and good upper limb control (Mathy at al., 2000), as was the case with most participants with bulbar-onset ALS in this study. When FCC changed as the disease progressed, some of the participants needed up to four different communication strategies simultaneously and up to seven separate strategies during the study to be able to maintain communication in different situations. The high number of secondary communication strategies is most probably due to our

clinic's AAC policy which says that when patients start using some of the high-technology strategies the SLP shall guide them also in the use of at least one low-technology strategy. This is to ensure the ability to communicate in conditions where high technology is not possible, for example when a device is out of commission or in case of power failure.

The majority of participants in the bulbar group first lost their ability to speak and then their hand function, mobility or both deteriorated. This is in line with the natural course of the disease, and in fact the initial symptoms tend to dominate throughout the disease (Yorkston et al., 1993). In contrast, most participants in the spinal group already had either poor mobility with or without poor hand function when their speech became unintelligible. As the disease progresses individually, regular assessment of speech and communication is needed (Andersen et al., 2012; Hanson et al., 2011) and appropriate communication support systems should be individualized and modified throughout disease progression (Fried-Oken et al., 2015).

Although ALS primarily affects motor skills including speech, language impairment and cognition changes are also noticeable in patients with ALS (Ball et al., 2007b, Fried-Oken et al. 2015; Taylor et al., 2013). Three of our participants who were not able to speak adequately during the second year of the follow-up could not use any of the low- or high-technology AAC solutions to communicate functionally. Although the cognitive functions of the participants were not formally assessed in this study, it was obvious that the three patients had such cognitive and language problems that their ability to write or type and/or use low- or high-technology AAC means was critically impaired. About 10-75% of persons with ALS have been reported to show cognitive impairment, 5-41% meeting criteria for dementia (Miller et al., 2009; Hanson et al., 2011; Phukan, Pender, & Hardiman, 2007). Cognitive problems may also be one of the reasons for non-acceptance of the available AAC (Hanson et al., 2011) or for difficulties in using high-technology devices (Geronimo, Simmons, & Schiff, 2016), which contributes to the increasing number of patients who cannot

communicate at all at the last stages of ALS (Brownlee and Bruening, 2012). The possibility of cognitive decline is important to consider when planning AAC strategies.

The strength of our study is the consecutive series of patients and prospective follow-up from the first SLP evaluation at a multidisciplinary neurological clinic. Our sample size was, however, small, which reduces the generalizability of our results. In addition, the effectiveness of communication is not explained by the amount or quality of different AAC strategies used, and this perspective needs more focus in future research. To conclude, AAC strategies are necessary for persons with ALS whose speech is deteriorating. However, maintaining the ability to communicate with high- or low-technology AAC means is a complex and continuing process. Individual changes of functional capacity to communicate are challenges to overcome when deteriorating speech needs to be augmented or natural speech replaced by other strategies. Typically, several AAC strategies are required in response to communication needs of persons with deteriorating speech due to ALS.

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