Dysarthria in amyotrophic lateral sclerosis: A review

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Abstract
Dysarthria is a motor disorder of speech characterized by abnormalities of the articulation and intelligibility of speech. Phonation and the rate of facial movements may also be affected. Understanding the nature and course of dysarthria in amyotrophic lateral sclerosis (ALS) is important because loss of communication prevents patients from participating in many activities, may lead to social isolation, and reduces the quality of life. The goal of management of dysarthria in ALS patients is to optimize communication effectiveness for as long as possible.

The information about dysarthria in ALS is dispersed in physiological, pathological, speech therapy, otorhinolaringological and neurological publications. This review summarizes the current state of knowledge on the clinical features, differential diagnosis, pathophysiology, investigations and management of dysarthria in ALS patients. There is a need to compare the different methods used to assess dysarthria and for controlled clinical trials to assess therapeutic strategies.

Key words: ALS, MND, dysarthria, bulbar palsy

Introduction
Dysarthria occurs in more than 80% of ALS patients and may cause major disability, earlier in those with bulbar onset (1,2) who may become anarthric after a few months (3). Careful neurological examination, including cranial nerves, and monitoring the rate of progression in affected muscles are important (4). Loss of communication prevents ALS patients from participating in a number activities and leads to social isolation (4,5). Dysarthria significantly reduces the quality of life of ALS patients (6,7).

There is little research on dysarthria in ALS. There are no published controlled trials in adults to support or refute the effectiveness of pharmacological approaches or of speech and language therapy for dysarthria in ALS or following non-progressive brain damage (8). Most of the data presented here are from Class IV studies (uncontrolled trials, case series, case reports, expert opinion) (9).

This review examines the clinical features, differential diagnosis, pathophysiology, investigations and management of dysarthria in ALS patients.

Methods
The English literature was electronically searched using MEDLINE-OVID (January 1966 to date); MEDLINE-ProQuest; MEDLINE-EIFL; EMBASE-OVID (January 1990 to date), the Cochrane Library Central Register of Controlled Trials (CENTRAL), World Federation of Neurology ALS website (searching for articles with SCIRUS), and American Speech Language Hearing Association website page of reviews of published research (ASHA online reviews). In the first search the key words – ‘ALS’ and ‘motor neuron disease’ – were included for relevant subtopics. The next searches included ‘bulbar palsy’, ‘dysarthria’ as well as ‘communication’, ‘pathophysiology’, perceptual and acoustic properties, and management (‘assessment’ and ‘treatment’). AAN criteria for types of evidence in clinical trials (9) were used to judge articles on management or treatment.

Clinical features of dysarthria in ALS
Symptoms of dysarthria may not be evident until about 80% of motor neurons are lost (10). The time between the onset of speech symptoms and the diagnosis may range from 33 months prior to...
diagnosis to 60 months after diagnosis (11). Twenty-five to thirty percent of ALS patients have dysarthria as a first or predominant sign in the early stage of the disease. Dysarthria as an initial symptom is eight times more frequent than dysphagia in ALS (12). It affects up to 70% of patients with limb onset at a later stage.

Neurological disease affecting different structures can result in different forms of dysarthria: lower motor neuron (LMN) (flaccid), upper motor neuron (UMN) (spastic), UMN and LMN (mixed), cerebellar (ataxic), extrapyramidal (hypokinetic, hyperkinetic) (13,14).

Figure 1 summarizes the type, pathological anatomy and clinical signs of the dysarthrias.

There has not been a systematic re-evaluation of these clinico-anatomical correlations, even with the availability of neuroimaging techniques (15).

Muscle wasting and weakness with proportional slowness of movements is characteristic of LMN involvement, while marked slowness of movement with variable weakness and no wasting are features of UMN dysarthria (14).

ALS patients usually have a mixed dysarthria (spastic-flaccid). It is characterized by defective articulation, slow laborious speech, imprecise consonant production, marked hypernasality with nasal emission of air during speech and harshness. A strained/strangled voice (spastic dysphonia) and disruption of prosody (16) may also be present. Decreased respiratory function leads to a weak (low volume) voice, also referred to as inappropriate vocal loudness for conversational utterances (10). Abnormal vowel production, which may result in monopitch voice, short phrases, distorted vowels, monoloudness and ‘breathy’ voice quality are also seen (17).

In mixed dysarthrias there is both UMN and LMN involvement in the bulbar region (4). The flaccid or bulbar type has predominance of LMN bulbar signs (tongue, palatal and facial weakness and wasting, poor or absent palatal elevation and tongue movements, and poor or absent gag and facial and jaw reflexes). In spastic dysarthria, or pseudobulbar type, UMN signs predominate (slow tongue movements, tongue, palatal and facial weakness, poor voluntary palatal elevation and brisk gag, facial and jaw reflexes), and there may be other features of the pseudobulbar syndrome, such as emotional lability, brisk palmovestibular reflexes, pout, corneomandibular reflexes (18) as well as dysphagia. The relative contribution of flaccidity and spasticity in the impairment of speech intelligibility varies across individuals (4,19).

Dysarthria in ALS can be rapidly progressive (4). The bulbar (LMN) ALS patients are generally more

![Figure 1. Type, pathological anatomy and clinical signs of the dysarthrias (adapted and modified from references 13,14).](image-url)
severely affected than the corticobulbar (UMN) or spinal ALS patients.

Otorhinolaryngology (ENT) assessments

Early dysarthria or dysphonia, frequent initial symptoms in bulbar ALS, are often seen by ENT surgeons first (20). Careful laryngological examination of the motion of the vocal cords as well as fibrestroboscopy analysis may help diagnosis (4,21,22). Direct and indirect laryngeal observations in ALS include aperiodic vocal fold vibration, paradoxical adduction patterns, hyperadduction of the ventricular folds (when corticobulbar involvement predominates), hypoadduction of the vocal folds (when LMN bulbar involvement predominates) as well as phonatory disruptions (4,16,21,23-26).

Speech analysis

Slow speech rate is a prominent characteristic of the dysarthria in ALS (27-30). Longer stop closures and vowel duration (31), as well as reduced vowel space area, were demonstrated in ALS persons compared to healthy persons (32). In an examination of the relationship between vowel space area and speech intelligibility, smaller vowel space areas were found in ALS patients, compared to neurologically intact speakers. The vowel space was found to account for 45% of the variance in speech intelligibility (32). Early manifestations of speech and bulbar dysfunction are altered voice quality (dysphonia), speaking rate and communication effectiveness (27).

Assessment of dysarthria in ALS

Clinical

Dysarthria is first assessed in clinical practice by listening to speech when taking a history, also called ‘perceptual’ assessment (13,14). The next step is the neurological examination of bulbar function and classifying the dysarthria as UMN, LMN, or mixed with or without predominance or UMN or LMN features. ENT examination of the vocal cords may or may not be required. The ALS Severity Scale – Speech (33) and ALSFRS-R (34), Appel scale (35), Norris score (36) and Charing Cross quantitative and qualitative scales (37,38) are clinically simple and useful ways of grading the severity of the dysarthria and its evolution during follow-up. For timed tests it is important to consider learning effects in establishing baselines for follow-up assessments (39–41). There are no comparative studies between these scales to guide the clinician on which one to choose.

Clinical trials

A consensus statement from the WFN Research Committee on Motor Neuron Diseases on guidelines for clinical trials in ALS acknowledged that precise measurements of bulbar function needed further development but deemed as validated bulbar functional rating scales (as the ones mentioned above), Frenchay Scale, Hillel Scale, Norris ALS Scale Bulbar Subscale and timed speech tests (time required to repeat a pre-established number of syllables) (42). It was felt then that phonetic feature analysis, and orofacial strength measurement with bulbar force transducers, were acceptable for small, single-centre studies.

Speech analysis

There are few validated methods to assess the nature and changes of speech and few systematic comparative studies of such methods. Most have been small research studies (Class IV, (9)) without established clinical applications.

The speech characteristics of dysarthria due to ALS have been studied by perceptual (30,32,43–47), physiological (electromyographic (43)), kinematic (43,48) and acoustic (30,49–57) analyses.

Perceptual studies. One such method is the Frenchay Dysarthria Assessment Scale (58). Early phonetic disturbances of speech were detected in dysarthric as well as in non-dysarthric highly intelligible ALS subjects and were most marked in the dysarthric cases (45). Another quantitative study using the Dysarthria Profile Tests (47) (closest to Frenchay Dysarthria Assessment), showed that adequate characterization of the dysarthria in ALS could be achieved by perceptual assessments of articulation, phonation, reflexes and prosody and that respiratory and phonation tests detected abnormalities in non-dysarthric ALS (46). There are no comparative studies between these methods to guide the clinician

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Table I. Longitudinal assessments of dysarthria in ALS patients.
Dysarthria in ALS

Dysarthria is well known to be important to intelligibility. Formants are primarily determined by the position of the tongue. The first formant (F1) is called F1; the second F2 is the next highest. These two frequencies are used to determine vowel sounds. The formant with the lowest frequency content is used to distinguish between vowels. A formant is a peak in an acoustic frequency spectrum that results from the resonant frequencies of the vocal tracts. Distinguishing between vowels can be quantitatively demonstrated by the Sentence Intelligibility Test (SIT) (59).

Acoustic studies. Early changes have also been documented in the speech of highly intelligible individuals with ALS using acoustic methods (3,32,44,45,51–55,60). However, acoustic impairments are variable across studies and individual patients.

Physiological and acoustic data point to specific speech correlates of neural degeneration, e.g. reduction of the F2 (second formant)* slope in acoustics and slow force generation in physiology (50). ALS patients had a significantly slower speaking rate than neurologically normal subjects (29).

A study (57) of the relationship between speech in ALS and acoustic differences for vowels in content and function words showed that vowel space area for content and function words was smaller for ALS than for controls, and suggested that the magnitude of temporal differences for vowels in content and function words was a better predictor of impaired speech than the magnitude of spectral differences for vowels (57).

The relationship between intelligibility and the acoustic parameters of speech has been measured by single word intelligibility, F2 formant trajectories (extent, duration and rate) as well as diadochokinetic rate in ALS patients and has shown decreased performance in dysarthric patients compared to non-dysarthric patients at baseline (56).

Acoustic analyses of the voice in ALS have revealed also deviant fundamental frequency (Fo), amplitude and frequency, perturbation (e.g. shimmer, jitter), voice range, vocal quality and phonatory instability (54).

Acoustic analysis may detect early involvement of the orofacial and laryngeal system (51,54), but its clinical usefulness has not been established.

Different dysarthria profiles were described in bulbar and spinal onset ALS patients using a computer-acoustic method and analysing the most affected vowels. Abnormal acoustic parameters of the voice were also demonstrated in ALS subjects with perceptually normal vocal quality on sustained phonation (54). Both studies suggest that acoustic analysis can detect abnormalities in speech before they become perceptually apparent.

Table I summarizes currently used assessment tools for dysarthria.

Differential diagnosis of dysarthria in ALS

The evaluation and classification of dysarthria and dysphonia are part of the standard neurological examination.

Dysarthria versus language pathology

Impairment of communication in ALS can also be due to language changes. ALS patients with cognitive impairment, such as frontal lobe type dementia, may have reduced verbal output preceding or following a bulbar syndrome with dysarthria, and often leading to complete mutism within a few months, reduced spelling ability, word finding difficulty, non-fluent aphasia, impaired comprehension and changes in spoken and written language (52,61–64). However, aphasic symptoms can be found independently of dementia in ALS patients (65) and overt dementia and aphasic syndrome may precede the onset of upper and lower motor neuron signs in bulbar regions and limbs (66).

Dysarthria in other neurological diseases

For experienced neurologists the spastic–flaccid dysarthria and the other clinical features of ALS are easy to recognize (see Figure 1).

Other spastic or UMN and mixed UMN and LMN dysarthrias

Primary lateral sclerosis may produce a pure spastic dysarthria evolving much later to an ALS picture with LMN signs. Cerebrovascular disease is a frequent cause of pseudobulbar palsy with or without a spastic dysarthria. Foramen magnum pathology, such as Arnold-Chiari or basilar invagination, brainstem intrinsic or posterior fossa tumours with brainstem compression and syringobulbia may produce UMN, LMN or mixed dysarthrias. Vasculitic disorders, as in connective tissue diseases, may behave similarly. Brainstem encephalitides of various aetiologies may also lead to variable types of dysarthria.

Other flaccid or LMN dysarthrias

Diseases affecting the lower brainstem motor neurons such as Kennedy Syndrome, lower cranial nerves, such as Guillain-Barré syndrome and chronic inflammatory demyelinating polynuropathy, neuromuscular junction such as myasthenia gravis

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* Acoustic cues in speech are: fundamental frequency, vowel formants, noise bursts, transitions. A formant is a peak in an acoustic frequency spectrum that results from the resonant frequencies of the vocal tracts. Distinguishing between vowels can be quantitatively demonstrated by the frequency content of the vowel sounds. The formant with the lowest frequency is called F1; the second F2 is the next highest. These two formants are primarily determined by the position of the tongue. The second formant (F2) is well known to be important to intelligibility.
(MG) and muscle, such as polymyositis or muscular dystrophies (e.g. myotonic and oculopharyngeal muscular dystrophies), may mimic the pure LMN bulbar palsy with flaccid dysarthria that can be seen in ALS. Retropharyngeal and laryngeal tumours may also lead to a flaccid dysarthria or dysphonia. It is the remainder of the neurological examination or ENT assessment, or features like abnormal fatiguability in MG, that allows a distinction.

**Other dysarthrias**

A discussion of the differential diagnosis with the extrapyramidal and ataxic dysarthrias is beyond the scope of this review.

**Pathophysiology of dysarthria in ALS**

*Definition of dysarthria*

Dysarthria may be defined as a group of speech disorders resulting from disturbances in muscular control over the speech production mechanism (13,14). The production of speech sounds depends on several highly integrated factors: 1) respiration, 2) phonation, 3) resonation, 4) articulation, and 5) neurological integration (15).

*Anatomical pathways controlling speech (67)*

The muscles controlling articulation, mastication and deglutition are innervated by the trigeminal (jaw movements), facial (face and lip movements), glossopharyngeal (stylopharyngeal muscle), vagus (palate elevation, vocal cords, laryngeal movements and pharyngeal constriction), and hypoglossal (tongue) nerves. The motor trigeminal and facial nuclei are in the pons. The ambiguous (IX, X) and hypoglossal (XII) nuclei are in the medulla. Nerve fibres to the tensor palate influence soft palate position and tone that are important in adjusting the internal shape of the upper oropharynx. The IX nerve is mainly sensory, supplying only the stylopharyngeal muscle which elevates the pharynx during deglutition and speech. It plays no major role in speech; its motor cells are in the dorsal part of the nucleus ambiguous. The rostral and caudal parts of this nucleus (X) provide innervation to the adductor and abductor muscles of the larynx and to the muscles of the pharynx and soft palate, respectively. This topographical organization accounts for the clinically known presentations of ALS with dysphonia and no palatal palsy or with palatal palsy and no dysphonia, mimicking similar selective involvement of anterior horn nuclei for specific muscles in the limbs, with no root or nerve distribution of weakness. The XII nerve innervates the nine paired muscles of the tongue involved in speech, swallowing and chewing (19).

**Pathological changes in the pathways controlling speech in ALS**

The nuclei of the above cranial nerves are under the control of specific cortical, subcortical, cerebellar and brainstem centres, especially by the primary motor cortex, where speech is initiated. Cortical control is effected via the corticobulbar tracts. The corticobulbar pathways innervate cranial motor nuclei bilaterally (with the exception of the lower facial nucleus which is innervated contralaterally) and terminate on motor neurons within brainstem motor nuclei and in segmental interneurons (68).

The lesion(s) in ALS can be located in the primary motor cortex and/or descending corticobulbar tracts (upper motor neurons and/or their axons), cranial nerve motor nuclei (V, VII, IX, X, XII) in the pons and medulla oblongata (lower motor neurons and their axons). Clinically, in ALS, degeneration of motor neurons in the cortical areas and corticobulbar tracts (UMN) results in pseudobulbar palsy (spastic bulbar palsy) while predominant degeneration of motor neurons of the lower brainstem nuclei and their axons (LMN) results in a 'pure' (flaccid) bulbar palsy with denervation of muscles of face, oropharynx, larynx and tongue.

In ALS patients a mixed bulbar palsy and a mixed dysarthria type are usually seen, which consist of varying flaccid and spastic components. The initial complaints of dysarthria in ALS patients include the inability to shout or sing, a weakened voice, and difficulty with enunciation. Because of reduced dexterity, repetitive movements of the lips, tongue and pharynx become slow. Slurred and difficult speech may suggest tongue, jaw, or lip (articulatory regions) weakness, spasticity or both. Incompetence of the velopharyngeal port allows air in the mouth to leak into the nose during enunciation, which results in a nasal tone. The pharyngeal dysfunction caused by ALS differs among patients. Often, the palatopharyngeal isthmus fails to close in speech, but closes adequately in swallowing, causing hypernasality and nasal emission without nasal aspiration or dysphagia (5). This discrepancy is due to greater displacement of the palate in swallowing than in speech. Errors of oral articulation are combined with, or compounded by, this nasal escape of air and abnormalities of nasal and pharyngeal resonance (20). Hoarseness associated with low volume suggests vocal cord, and possibly respiratory muscle, weakness. LMN facial weakness is usually not an initial symptom of the disease but a later, generally constant, feature.
Pathology, function and imaging studies of cranial motor nerve involvement in ALS

Systematic pathological data on cranial motor nerve nuclei are sparse in ALS, and good quantitative analyses are lacking. There is evidence that the hypoglossal motor neurons are the earliest and most severely affected, while facial and trigeminal motor neurons are less commonly involved initially (69). The motor nucleus of the Vth nerve is usually the least frequently affected (69). Tongue (including bending movements for lingual consonants) and larynx were more impaired and earlier than the facial and mandibular movements (48,69,70). Subtle clinical changes in the XII, X, VII and Vth cranial nerves can be detected without any apparent functional speech change (51,69–71). The above sequence of pathological and clinical changes in cranial nerve involvement was confirmed by quantitative measurements of lip, mandible, and tongue function in bulbar, and also spinal, groups of ALS patients with no detectable dysarthria (48,71).

Qualitative measurement of the strength of the tongue and of the range and velocity of its movement are routinely used during the neurological examination of ALS patients but quantitative measurements can also be performed (36,37,72). Tongue dysfunction in ALS includes reduced range and velocity of movement (36–38,73), reduction in strength (73), smaller vowel space areas (32) and flattening of vowel formant (especially F2 trajectories) (49,50,74).

ALS tongues were 30% smaller, more rectangular, and situated more posteriorly and ventrally in the oral cavity on magnetic resonance imaging (75). There was an abnormal loss of the radial and curvilinear bands of the intrinsic tongue muscles. Significant impairment of lingual strength is seen in ALS patients (43,73), greater than the weakness of the jaw and lower lip, even among those without bulbar signs and/or symptoms (71). Three functional regions were defined for myometric studies of the orofacial structures in ALS dysarthria – articulator (lips, tongue, mandible), velopharyngeal ( palate, pharynx), and phonatory (larynx); there are abnormalities of lingual, velopharyngeal and laryngeal articulation in ALS (19,43,44,71). The severity of dysarthria correlated better with repeated contraction rate than with strength, suggesting that severe dysarthria may be largely due to slow orofacial and tongue movements until substantial muscle strength has been lost (48). This probably applies to the UMN and mixed, but not to the LMN or flaccid, dysarthria, of ALS.

Management of dysarthria in ALS (Figure 2)

The management of dysarthria in the ALS clinic starts with a neurological diagnosis of ALS and of the type of dysarthria. The otolaryngologist assessment may be helpful (20). Neurological assessments, clinical scales and SLT examinations should be performed periodically. Voice quality, speaking rate and communication effectiveness measured perceptually are one of the bases for making the appropriate decision about future speech support.

The EFNS-ALS guidelines (76) suggest as good practice points assessment of communication (every three to six months) and the use of appropriate communication support systems.

The goal of management of dysarthria in ALS is to optimize the intelligibility of speech for as long as possible and to concentrate not only on the disabled person, but also on partner-to-partner communication (76). The timing for assessments, interventions, and the methods of intervention, should be tailored to each patient. Effective evaluation and management of dysarthria in ALS patients may be limited by the availability and accessibility of neurological and SLT services, as well as by age, sex, psychological and psychiatric factors (cognitive status, motivation, personality, psychiatric illness), language function, physical function, hearing status (11,77) and socio-economic circumstances.

There is no cure for progressive dysarthria in ALS. Some symptomatic and compensatory strategies may temporarily improve the patient’s communication and have an impact on quality of life. The patient may move from oral communication to written communication, to using an augmentative communication device, or via another person (78). Neurologists will usually consider such supporting strategies when patients feel that they need help to communicate better, and this is often obvious on examination. In clinical practice, some patients choose to communicate to the few close persons who can understand their severely dysarthric speech, or to use the cheapest communication aid (i.e. writing on a piece of paper, alphabet chart). There are also patients who do not use the communication support provided and prefer to remain mute. Their wishes should be respected.

Pharmacological management

It is limited and with only Class IV evidence.

Measures to reduce spasticity. Sometimes patients with spastic dysarthria are temporarily helped by ice placed over the larynx or sucked, or antispastic drugs such as baclofen (79) (10 mg t.d.s. increasing gradually according to response) or tizanidine (2 mg t.d.s. increasing gradually according to response). Botulinum toxin type A has been reported as effective in spastic dysarthria (80) and spasmodic dysphonia (81–84).
**LMN dysarthria.** Pyridostigmine (30 mg t.d.s. or q.d.s. increasing gradually according to response) may help temporarily in some patients, perhaps because of the known uncertainty of transmission in terminal degenerating and regenerating axons, partly responsible for abnormal fatiguability and for positive decremental repetitive stimulation studies and abnormal jitter on single fibre EMG recordings in ALS (85,86).

**Excess salivation.** This may be helped temporarily, sometimes, with oral (10 or up to 20 mg t.d.s. or q.d.s.) or transdermal (1 mg every 72 h) hyoscine, atropine tablets (0.6 mg) or drops, glycopyrrolate (0.6–1.2 mg/24 h) or amitriptyline (10 mg/day or more) (class IV – (ALS guidelines (76)). Injections of botulinum toxin type A into the salivary glands have also been reported as useful in ALS (87–89). However, acute deterioration of bulbar function after botulinum toxin treatment for sialorrhoea in ALS was recently described (90).

**Speech and language management strategies**

There is no hard evidence (only Class IV) regarding speech and language management strategies in ALS patients. It has been suggested that perceptual identification of ‘critical periods’ of progressive dysarthria and timely intervention can be effective (77), but it is common experience that although effective communication may be improved temporarily by a number of strategies, the dysarthria itself continues to deteriorate. There is no consensus as to when, and based on which kind of procedure, to start strategies in dysarthric ALS patients.

**Strategies for coping.** In just detectable speech impairment (speech is made worse by fatigue or stress), simple things such as minimizing the noise in the environment, reducing the distance from the listener, are helpful (77). In mild dysarthria, ALS patients may compensate by a number of speaking strategies – slowing the speech rate, speaking face to face, substituting articulation manoeuvres such as: alternative words, spelling, repetition, overarticulating consonants, or even using key words or monosyllabic speech (77,78). Concentration on speaking only and energy conservation may prolong the time of successful communication (19,77,78).

**Speech therapy (logopedic) training.** Speech therapy training may be useful in patients with relatively slow progression of dysarthria (1,19,91) but there is no such evidence in ALS cases.

Lip and tongue exercises may sometimes help the patient to enunciate words more clearly. There are no credible data on strengthening exercises of the orofacial muscles in ALS patients and a number of neurologists discourage this practice (92). Energy conservation is a key component in managing bulbar function in ALS (19,78).

**Speech interventions are summarized in Figure 3.**

- **Palatal lift and palatal augmentation prostheses**

A palatal lift may temporarily improve resonance by displacing the weak soft palate to the level of normal palatal elevation and reduce hypernasality and hypophonia (19,93). A palatal augmentation prosthesis may temporarily improve articulation by lowering the palate, improving the production of the
lingual consonant sounds (19). There is no good evidence as to the effectiveness of these prostheses, or of their period of usefulness if effective (93–97). Palatal lift would not be contemplated in patients with spastic dysarthria. In most centres they are not, or only rarely, used. In the few cases where we have seen them used they have been of little temporary, or no, help.

Other communication strategies in ALS

Communication strategies in ALS patients should concentrate not only on the disabled person, but should consider social closeness and personal partner–to–partner communication as well (78,95). As the dysarthria progresses, conversation strategies (based on partner interpretation, understanding and confirmation, context, topic cues) and alternative communications, such as gestures, positioning, facial expression and eye contact (non-verbal strategies) can be used (78). The communication strategies have to be adopted by both communication partners, patients and their listeners, which may improve the patient’s quality of life (78). Important relationships among speech intelligibility and communication effectiveness between speakers and their listeners have been highlighted (78,98).

Alternative communication methods

When progressive dysarthria leads to severe or complete unintelligibility of speech, augmentative and alternative communication (AAC) systems are needed (77). Choosing the best system involves detailed evaluation of the individuals, their hand function, mobility; social and work environments, insurance coverage and finances, as well as their cognitive function (7). Starting AAC depends primarily on patients’ and carers’ choices, the individual’s intelligibility (33) and changes in speaking rate (11,27).

Light-tech devices. Examples are alphabet boards, individual picture communication charts, picture communication symbols, alerting systems (e.g. buzzers), telephone communication systems and portable writing systems.

Electronic high-tech devices. These are based on multipurpose augmentative computer communication systems that are commercially available (19,99). There are portable amplifiers that increase the volume of the patient’s voice for improving intelligibility of speech, digital recorders that play back prerecorded words and phrases on command, keyboard activated printout or sound-producing communicators, and dedicated voice synthesizers. Speech synthesis software is available for use in desktops or laptops (19). The usefulness of brain–computer interface (BCI) communication devices for individuals with advanced ALS has been reported recently (100). The methods of asynchronous communication (by email or messageboard forums) and ‘voice banking’ (recording phrases of the patient’s own voice) can also be used.

Expert advice is required for choosing the appropriate system for each patient. Caregivers may need to provide support for their use of speech
and communication management (19). Figure 4 summarizes the communication strategies.

Tracheostomy

It is known that tracheostomy impairs oral communication (101). A standard cuffed tube prevents the patient from talking, but fenestrated cuffed tubes may allow speech and still protect against aspiration (102). Speech is often maintained by using some adaptations to the tracheostomy tube, e.g. Passy-Muir valve (103). Loss of useful speech requires the use of alternative communication technology and ‘yes/no’ technique by use of eye pointing or eye gaze, eyebrow or finger movement, etc. For ventilated patients eye-gaze high-tech AAC devices may be used (3,19,77).

For patients of the ‘locked-in’ status, brain-computer interface (BCI) methods which use direct connections between brain and computer, are being developed based on electroencephalogram and evoked potentials (104–109). This kind of support is still at the experimental stage.

Communication strategies for ALS patients with cognitive impairment or frontal lobe type dementia (FLTD)

In advanced cases, patients will often communicate to their families through their behaviour and expressions of emotion. Caregivers must be flexible and adapt their verbal and non-verbal communication techniques according to each individual changing cognitive levels and needs.

There are no systematic or conclusive studies regarding support strategies in these ALS patients.

Neurophysiological examination and careful language assessment is used as a diagnostic procedure (110,111). Verbal fluency tests (VFT) have been shown to be useful in identifying cognitive deficits (112,113) in ALS patients with language changes. A word generation test (WGT) may be useful to screen patients in whom more detailed neuropsychological evaluations are needed to document frontal deficits (114).

Conclusion

The assessment of dysarthria and the neurological examination, including the type of bulbar syndrome, are essential for the diagnosis of, and the condition causing, dysarthria, and will inform subsequent decisions about investigation and management in the ALS clinic. There is currently no hard evidence to support particular assessment methods or management strategies for dysarthria in ALS. Based on clinical opinion, dysarthria in ALS should be assessed early and monitored regularly. Perceptual assessment of the intelligibility of speech by the patient, carers and professionals, remains the main criterion for decisions on communication support. Assessment methods in common use include also various qualitative and quantitative clinical scales of bulbar function and standardized dysarthria scales. A few pharmacological approaches can be tried but are of no proven value. Communication support should be individualized from the onset of dysarthria according to patient needs and wishes. Various speaking conversation and non-verbal strategies, low-tech and high-tech AAC systems should be offered at the appropriate time.
Dysarthria in ALS

For future research on communication in ALS, the EFNS-ALS guidelines (76) propose further studies for evaluating language dysfunction and its treatment. Controlled clinical trials to assess therapeutic strategies and comparisons of the different methods used to assess dysarthria and to help communication in ALS patients with cognitive impairment are needed.

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