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Communication Impairment in ALS Patients Assessment and Treatment

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1. Introduction

Amyotrophic lateral sclerosis (**ALS**), also called *Lou Gehrig's disease*, is a rapidly progressive neuromuscular disease that attacks the neurons responsible for controlling voluntary muscles. It belongs to a group of disorders known as *Motor neuron diseases* (*MND*): all these syndromes share a common molecular and cellular pathology comprising degeneration of motor neurons (MNs) in cortex, brainstem and/or spinal cord, and the presence of characteristic ubiquitin and TDP-43-immunoreactive intraneuronal inclusions.
ALS prevalence in Western countries ranges from 2.7 to 7.4 per 100,000 (Worms, 2001). In 90 to 95 percent of all patients with ALS (PALS), the disease occurs sporadically (*sporadic ALS*, *sALS*); in 5 to 10 percent there is a family history of ALS (*familial ALS*, *fALS*). Most people developing ALS are between the ages of 40 and 70 years (Haverkamp et al., 1995). The disease is 20% more common in men than in women, although more recent data suggest that the gender ratio may be approaching equality (Logroscino et al., 2008).
The cause of ALS is not known, and it is not clear why ALS strikes some people and not others, but both genetic (Ticozzi et al., 2011) and environmental factors (Callaghan et al., 2011; Calvo et al., 2010; Ferrante et al., 1997) may play a role.
In PALS, both the brain upper MNs (UMNs) and the brainstem or spinal cord lower MNs (LMNs) degenerate or die: unable to function, the muscles gradually weaken, waste away, and twitch, leading to a wide range of disabilities. Patients lose their strength and the ability to move their body, but usually maintain control of eye muscles.
Approximately 70% of PALS have a *spinal form* of the disease: they present with symptoms which may start either distally or proximally in the upper or lower limbs. Some patients see the effects of the disease on a hand or arm, as they experience difficulty with simple tasks requiring manual dexterity, such as buttoning a shirt, writing, or turning a key in a lock; in other cases, symptoms initially affect one of the legs, and patients experience awkwardness when walking or running, or they notice that they are tripping or stumbling more often.
Patients with *bulbar-onset* ALS usually present with dysarthria leading to slow slurred speech or a nasal quality; they may also develop dysphagia for solid or liquids after noticing speech problems; almost all patients with bulbar symptoms complain of sialorrhoea with excessive drooling due to difficulty of swallowing saliva and UMN-type facial weakness, which affects the lower part of the face, causing difficulty with lip seal and blowing cheeks.

The gag reflex is preserved and often brisk, whereas the soft palate may be weak; patients show wasting and fasciculations of the tongue which moves slowly, also due to muscle hypertonia. The other cranial nerves remain intact, although in late stages of the disease patients may very rarely develop a supranuclear gaze palsy or oculomotor palsy (Kobayashi et al., 1999; Okuda et al., 1992).

2. Communication issues in ALS patients

Five functional domains have to be taken into account when communication is concerned: a) motivation to interact; b) cognitive skills (particularly - but not exclusively - those related to language); c) visual and auditory capacities; d) ability to utter sounds and words; and e) writing skills. As a matter of fact, affective disorders, anxiety and emotional discomfort, as well as cognitive impairment or sensory deficits, can compromise communication processes, by interfering directly and/or indirectly with speech and writing performances.

However, ALS does not usually either affect a person's ability to see, smell, taste, hear, or recognize touch; nor impair mind or intelligence - although a small percentage of patients may experience problems with memory or decision-making (Raaphorst et al., 2010; Ringholz et al., 2005), and there is growing evidence that some may even develop a form of dementia (Guedj et al., 2007). On the other hand, since the disease usually does not affect cognitive abilities, PALS are aware of their progressive loss of function and may become anxious and depressed (Patten et al., 2007).

The vast majority of PALS experience a motor speech disorder as the disease progresses: since ALS involves both UMNs and LMNs, it results in *mixed dysarthria* of the *flaccid-spastic* type characterized by effortful, slow productions with short phrases, inappropriate pauses, imprecise consonants, hypernasality, strain-strangled voice, as well as decreased pitch and loudness range (Duffy, 1995; Tomik & Guiloff, 2010).

In the early stages when dysarthria is mild, either spasticity or flaccidity is predominant: initial symptoms typically do not interfere with speech intelligibility and may be limited to a reduction in speaking rate, a change in phonatory quality, or imprecise articulation (Ball et al., 2004_a; Nishio & Niimi, 2000; Yorkston et al., 1993; Yunusova et al., 2010).

Features of *spasmodic dysphonia* (or *focal laryngeal dystonia*) may also occur in PALS, sometimes as the initial clinical symptom (Roth et al., 1996). Typically, laryngeal structure is normal in appearance. When *corticobulbar* involvement prevails (*spastic* forms), there is often a pattern of hyperadduction of the vocal mechanism, and when *bulbar* involvement dominates (*flaccid* forms), there is usually a pattern of hypoadduction.

As disease progresses and dysarthria becomes severe, profound weakness resulting in reduced movement of the speech musculature and severe hypophonia become increasingly common (Yunusova et al., 2010).

Perceptual and acoustic features of dysarthria in ALS have been well studied (Tomik & Guiloff, 2010): the decrease in rate is often associated with increased pause time and enhanced segment durations, particularly for vowel sounds (Green et al., 2004; Tjaden & Turner, 2000; Turner & Weismer, 1993); spectral vowel and consonant properties (e.g., formant frequencies, transition extents and slopes) are also affected, with vowels becoming more centralized and the consonant frequency spectrum less distinct (Kent et al., 1989, 1992; Tjaden & Turner, 1997; Turner et al., 1995; Weismer et al., 1988, 1992, 2001).

Such acoustic findings have been presumed to be due to the disease-related reduction and slowing of articulatory movements (Weismer et al., 1992). Articulatory findings, although

limited, support such an interpretation. An early study on articulatory kinematics in two PALS showed slowed articulatory movements, reduced displacement of the tongue and lip, together with exaggerated displacements of the jaw during diadokokinetic tasks (Hirose et al., 1982). A more recent study of articulatory movements in a group of 9 PALS reported an impairment of articulatory speed during vowels (Yunusova et al., 2008): aberrant displacements were found to be word- and vowel-dependent and were more consistently present in movements of the tongue than in those of other articulators, and occasionally in the jaw; the jaw displacements were smaller than normal in words requiring larger articulator movements (e.g., consonant plus low vowel), but were larger than normal in words that only required relatively small jaw movements (e.g., consonant plus high vowel), suggesting difficulty in scaling of the vowel-related movements.

Whereas initially, along a gradual slowing of *speaking rate*, *speech intelligibility* remains relatively high, it decreases overtime, when dysarthria becomes more and more apparent to PALS themselves and their listeners. Yorkston and co-workers (1993) suggested that speech intelligibility may vary across dysarthric patients depending on the subsystems that are preserved (e.g., relatively less impaired respiratory-phonatory subsystem and the jaw might be associated with better speech intelligibility); the rate of disease progression; and the patient's cognitive status.

Because a person's ability to communicate orally is typically assessed based on speech intelligibility, anticipating the decline in intelligibility in a sensitive way is critical for timely clinical management of bulbar PALS. In this regard, longitudinal studies are, indeed, necessary when the goal of research is to identify early predictors of future changes; additionally, longitudinal studies are advantageous when dealing with heterogeneous populations, as in the case of PALS, since each patient can serve as his own control. Investigations of such a type documented the decline in speech intelligibility and speaking rate (Kent et al., 1992; Mulligan et al, 1994; Nishio & Niimi, 2000; Yorkston et al., 1993); and some studies have also identified several acoustic-based speech markers of disease progression (Mulligan et al., 1994; Ramig et al., 1990).

In their retrospective study of more than a hundred clinical cases, Yorkston and co-workers (1993) reported that speaking rate was a reliable predictor of speech intelligibility decline, by observing a rapid deterioration in speech intelligibility shortly after a decline in speaking rate to 100-120 words per minute. Such a finding was replicated by Ball and co-workers (2002) in a large group of patients with bulbar symptoms of different severity: the authors suggested that speaking rate decline to 100-120 words per minute should serve as a clinical indicator for beginning to support communication by assistive technology.

Moreover, since ALS progresses so rapidly in many subjects, an important goal of clinical management is to anticipate functional changes in patients' performance in order to teach new communication strategies and compensatory skills before the patient's ability to learn these skills is impacted by the severity of their condition. Recently, Yunusova and co-workers (2010) in a longitudinal study on 3 PALS tested the feasibility of using kinematic measures as early predictors of intelligibility decline, trying to understand the relationship between physiologic changes in speech movements and clinical measures of speech performance (such as speaking rate and speech intelligibility). Lip and jaw movements were quantified with respect to their size, speed, and duration.

Results showed that, differently from oral strength measures, changes in lip and jaw movements were related to ALS progression: in two out of 3 PALS, the changes in measures

of path distance and speed anticipated the drop in speech intelligibility by approximately 3 months, whereas speaking rate decline was more gradual; and increases in movement duration overtime closely mimicked the pattern of speech intelligibility decline. Overall, the kinematic measures seemed to be sensitive to disease progression: they might therefore be useful clinical markers for initiation of compensatory interventions.

Parallelly to decline in speech intelligibility, *communication effectiveness* is reduced at first in adverse speaking situations, such as noisy crowds, and then in all situations. Ball and co-workers (2004_b) reported that perceptions of communication effectiveness for PALS were quite similar to those of their frequent listeners (spouse or family member) across 10 different social situations: a range of communication effectiveness was reported depending upon the adversity of specific social situations.

Significant dysarthria can lead to frustration on the part of the patient when others are unwilling to spend the time to carefully listen. Friends and healthcare workers may not listen to the patient; there is a temptation to anticipate answers and finish sentences for the patient.

Fatally, at some point in their disease progression, 80 to 95% of PALS are unable to meet their daily communication needs using their natural speech, and finally most become unable to speak at all.

Moreover, upper limb paralysis prevents them from using hands in writing (directly or through computer-linked keyboards or communication devices).

Ultimately, in the so-called "locked-in" cases, a diffuse somatic immobility takes them away any possibility to interact with the world, except by using eye movements - even more unfortunately lost in those PALS classified as having a "super locked-in" syndrome, who may rely only upon their brain electrical waves as a communication tool processed through complex *brain-computer interface* (*BCI*) devices.

3. Functional assessment

Many assessments have been proposed for patient's follow-up in order to analyze the state of motor function and their consequences on activities of everyday life (Couratier et al., 2006). Clinimetric scales must be validated and relatively simple to use, and generate ordinate results allowing statistical analysis: global scales - *Norris Scale* (Norris et al., 1974), *Appel ALS Rating Scale* (Appel et al., 1987), *ALS Severity Scale* (Hillel et al., 1989), and *ALS Functional Rating Scale* (*ALSFRS*) (Cedarbaum & Stambler, 1997) - can be employed to evaluate disability progression.

By using, for instance, ALSFRS - or its revised version, $ALSFRS_R$ (Cedarbaum et al., 1999) - , communication impairment can be assessed through scores on speech function together with those related to handwriting, since people communicate by speaking and/or writing: scores < 2 in both speech and handwriting items correspond to a substantial inability to communicate.

Dysarthric speech can be evaluated through the *Frenchay Dysarthria Assessment* (Enderby & Palmer, 2008) originally developed by Pamela Enderby in 1983, which represents a well-established clinical tool to quantitatively evaluate the organs involved in speech and provides a measurement of intelligibility.

Complete kit includes examiner's manual, 25 rating forms, and intelligibility cards: patient is rated on a number of simple performance tasks related to speech function.

Intelligibility can be measured also through another test developed by Yorkston and co-workers: the *Assessment of Intelligibility of Dysarthric Speech* (Yorkston et al., 1984), a tool for

quantifying single-word intelligibility, sentence intelligibility, and speaking rate of adult and adolescent speakers with dysarthria. Standard protocols containing speaker tasks, recording techniques, and listener response formats are employed to obtain a variety of intelligibility and communication efficiency measures.
Yorkston and co-workers (1993) initially suggested that PALS speaking rate reduction precedes decreases in intelligibility; Ball and co-workers (2001, 2002) reported that speaking rate on the *Speech Intelligibility Test - Sentence Subtest* (Yorkston et al., 2007) is a relatively good predictor of PALS intelligibility deterioration. This computerized test supports the efficient measurement of speaking rate in clinical settings; it helps patients and their families monitor changes over time, and reinforces their understanding of speaking rate and intelligibility. Using this test, speaking rate can also be accurately monitored over the telephone if a patient lives at a distance, or is unable to travel (Ball et al., 2005_a): it should be noted, anyway, that speech intelligibility could not be objectively assessed over the telephone, as a clinical measure of understandability.
The *vocal impairment* can be difficult to assess because the voice disorder in dysarthria often occurs along with other impairments affecting articulation, resonance, and respiration: an effective assessment tool is the *Multi-Dimensional Voice Program*, a multi-parameter acoustic analysis (Kent et al., 2003).

4. Treatment

Differently from an acute, self-limited disease with expected recovery, the choice of appropriate therapeutic options for PALS raises more difficult concerns, since one must take into account many personal and ethical considerations. Several decisions by PALS and their families regarding treatment hinge on their concept of the quality of life that will result from such treatments.
At the present time, ALS therapy can be organized under the following multiple modalities: a **pathogenetic treatment** – to counteract MN degeneration; and a **symptomatic treatment** – to reduce impairments in motor abilities including those involved in communication. The appropriate implementation of each one of these types of therapy reflects the difficulties that we now have to face in ALS treatment. Supportive care is best provided by multidisciplinary teams of health care professionals, such as physicians; physical, occupational, and speech therapists; nutritionists; social workers; and home care and hospice nurses (Bede et al., 2011): working with patients and caregivers, these teams can design an individualized plan of medical and physical therapy and provide special equipment aimed at keeping patients as "functional" as possible.
Taking now into account such *a symptomatic approach*, two kinds of therapeutic strategies have to be implemented, those using drugs and those employing assistive/rehabilitative methods and techniques, aids and devices.

4.1 Pharmacological strategies

Physicians can prescribe medications to ameliorate fatigue, ease muscle cramps, control spasticity, and reduce excess saliva and phlegm; drugs also are available to help patients with pain, depression, anxiety, and sleep disturbances (Bede et al., 2011; Gordon, 2011; Guidubaldi et al., 2011; Guy et al., 2011; Miller et al., 1999; $2009_{a,b}$).
It is almost obvious that a patient experiencing less fatigue, pain, anxiety and depression, and controlling better saliva and spasticity, also apart from specific speech and writing

motor deficits (which, indeed, are improved by reduced sialorrhoea and muscle hypertonia), will be able to successfully manage his/her language impairment, being more committed to communicate and keep social contacts.

4.2 Non pharmacological strategies

The primary goal of an effective assistive rehabilitation for PALS is the management of disabilities, symptoms and complications arising from the progressive weakness of limb, trunk, and bulbar muscles. Further goals include keeping the patient functioning as independently as possible, and maintaining quality of life even into the terminal stage (Francis et al., 1999).

The rehabilitation program varies depending on whether the patient has a long clinical course or rapid progression of the disease: in the former case, PALS become able to compensate remarkably well for the motor unit loss and are able to continue with their daily activity for several years (Chen et al., 2008). The success of the rehabilitation approach depends on the active participation of the patient who should be a full partner in the therapeutic team even during the advanced stages of the disease. It may be difficult for the physician to discuss such a fatal illness: however, a direct approach allows the patient to deal most effectively with the disease and its physical limitations. This also helps in decisions about the intensity of the therapeutic effort (Bede et al., 2011; Gordon, 2011).

The family and other caregivers should be encouraged to participate in the patient's early rehabilitation program: the family role will then likely increase as weakness progresses, requirements for assistive devices change, and new problems arise in the management of activities of daily living.

4.2.1 Treatment of impaired communication

Loss of effective communication prevents patients from participating in many activities; may lead them to social isolation; and reduces their quality of life: the goal of clinical management of dysarthric PALS is to optimize communication effectiveness for as long as possible. Communication solutions, which may include no-technology, low-technology and high-technology options will be discussed, as well as the importance of psychosocial issues and the factors influencing the use of these systems.

Dysarthric PALS may benefit from working with a *speech therapist*: these health professionals can teach patients adaptive strategies, such as techniques to help them speak louder and more clearly. In early disease stages, patients can be taught to emphasize certain syllables and slow their speech patterns so that others can understand them better: lip and tongue exercises can sometimes help the patient to enunciate words more clearly on a regular basis.

A recent review on ALS communication research (Hanson et al., 2011) concluded that, due to ALS pathophysiology and the intrinsic degenerative nature of disease, speech treatment strategies designed to increase strength or mobility of the oral musculature are not recommended for PALS. Patients or their caregivers, on the contrary, often request oral exercises to improve strength and mobility for speech, as strengthening exercises seem intuitively to them as a way to increase performance: however, such exercise programs should be discouraged, and PALS should be informed that the speaking that they do each day provides a sufficient amount of speech mechanism activity and exercise.

Speech intervention should focus on learning to conserve energy for priority speaking tasks and to rest often to reduce fatigue, instead of increasing effort with speech exercises. PALS

speakers should learn to avoid adverse speaking/listening situations by muting the television, inviting people to speak with them in a quiet place rather than in a crowded room, and using voice amplification when speaking in noisy environments to reduce the effort required (Ball et al., 2007; Yorkston et al., 2010).

On the basis of a retrospective study on 25 dysarthric PALS treated with a palatal lift and/or augmentation prosthesis, the use of such devices should be regarded as effective in improving speech: 84% of patients treated with a palatal lift reported reduction of hypernasality (76% benefiting, at least moderately, for 6 months), and 60 % of those treated with a combination of palatal lift and augmentation prosthesis demonstrated improvement in articulation (Esposito et al., 2000).

Writing may be used as a substitute for speech, and devices as simple as paper and pencil, alphabet cards, portable typewriter, and letter boards may be utilized by patients with adequate hand function. Becoming speech more and more difficult to understand, many PALS supplement their speech by identifying the first letter of each word on an alphabet board (*alphabet supplementation*), or by identifying the topic on a communication board (*topic supplementation*).

As ALS progresses, speech therapists can help patients develop ways for responding to yes-or-no questions with their eyes or by other nonverbal means, and can recommend aids such as speech synthesizers and computer-based communication systems: these methods and devices help patients communicate when they can no longer speak or produce vocal sounds.

The technological revolution has expanded communication options for PALS who cannot rely on natural speech and writing. The assistive technologies are categorized as *Augmentative and Alternative Communication* (*AAC*) devices. Four critical features need to be considered within clinical and research domains: language representation, output mode, motor access, and microprocessor units.

Language representation has got remarkable attention for speaking rate enhancement. Whereas most PALS spell and rely on typing as a form of input, they can never approach speech production rates: often the slowness of AAC devices reduces their utility. Nowadays devices are being designed that integrate natural language processing and prediction algorithms for word, utterance and even conversational level units as one tries to approach natural speaking rates.

The **output mode** has seen advances for the storage of digitized voice as well as qualitative improvements to synthetic speech.

Voice banking is often considered as an early treatment option: PALS with intact motor speech skills store their spoken sounds, words, sentences for future use in customized communication devices. The personalized voice and messages can be used along with standard text-to-speech output to retain the PALS' voice signature: engineering efforts to customize synthetic speech to the user's own voice through minimal speech sampling are going on. Scientists and technicians are pursuing the gold standard for a device: bad speech in and good speech out, with attention being paid to recognition of dysarthric speech and production of personalized voices.

Motor access problems are being addressed with visual evoked potentials, detection of alpha and theta waves, and eye gaze recognizers so that head, shoulders, knees and toes are no longer needed: most devices now offer a range of access methods, starting with keyboards, touch screens, a head mouse, and Morse code.

Finally, **microprocessor units** are available in every shape and size to meet user needs, from palmtops to laptops made of magnesium-alloy shells, to software that can be downloaded from the Internet and accessed through any home computer.

AAC has a remarkable importance in dysarthric PALS management. When a person has a severe verbal communication impairment, AAC can meet the overall goals of palliative care: AAC can improve quality of life by optimizing function, assisting with decision making, and providing opportunities for personal growth.

Clinical decision-making related to communication is quite complex as screening, referral, assessment, acquisition of technology, and training must occur in a timely manner, so that when residual speech is no longer effective, AAC strategies are in place to support communication related to personal care, healthcare, social interaction, community involvement. Many reports of use and frequency for the purposes of staying connected and discussing important issues point out that AAC technology can assist the patient-caregiver dyad in maintaining previous relationships. The face-to-face spontaneous conversation mode is used most frequently, despite the slow rate of production, the lack of permanence, and the demands on conversational partners during message generation (Fried-Oken et al., 2006).

PALS, their family members, and, at times, their medical team, usually do not wish to consider an AAC decision until the deteriorating speech intelligibility limits the communication effectiveness: unfortunately, once intelligibility begins to decrease, speech performance often deteriorates so rapidly that there is little time to implement an appropriate AAC intervention. Indeed, appropriate timing of referral for AAC assessment and intervention continues to be a relevant clinical decision-making issue. The speaking rate should be clinically monitored so that the referral for an AAC intervention is initiated in a timely manner: Ball and co-workers (2001, 2002) recommend that patients be referred for AAC assessment when their speaking rates reach 125 words/min (normal value: 190 words/min) on the *Speech Intelligibility Test - Sentence Subtest*) (Yorkston et al., 2007). With sufficient education and preparation, PALS and their caregivers are ready to examine their AAC options timely: nevertheless, speech deterioration can be so rapid anyway that individuals can be left with limited communication options, if they are not really prepared to act in an opportune manner.

Due to the extended AAC use with deteriorating levels of physical control, it is imperative that recommended technology has adjustable access options to meet the range of motor capability as the disease progresses (s. above).

PALS should be fitted with AAC technology that supports multiple access methods, such as allowing them to transition from hand access to scanning and/or head/eye-tracking. Many AAC devices now incorporate a variety of access options so that the technology can continue to meet the needs of the user despite a decline in physical capability: the sensitivity of dynamic touch screens can be adjusted to allow for lighter touch; the improved sensitivity of head-tracking technology has allowed many patients to use this access method with minimal head/neck movement control.

Perhaps the most significant advancement in access technology has occurred with the widespread availability of *eye-tracking systems* to allow cursor control with eye movement to access high-technology AAC devices. As the disease progresses, many PALS require the use of eye-tracking for several reasons. Firstly, compared to other access methods (such as switch-activated scanning), eye-tracking is often reported to be the least fatiguing method

(Gibbons & Beneteau, 2010) and its technology requires relatively little effort (Calvo et al., 2008; Harris & Goren, 2009): eye gaze is natural, and eye muscles generally do not fatigue with use. Secondly, eye gaze may be the only volitional movement that the individual continues to exhibit over time, particularly in cases where invasive ventilation has been chosen (Ball et al., 2010).

BCI technology has generated considerable interest for people who are physically "locked-in", such as PALS in the late stages of the disease. BCI devices translate into computer commands volitional modulation of brain signals which can be recorded from the scalp using electroencephalography (EEG) or magnetoencephalography; from the dura mater or cortical surface using electrocorticography; or from neurons within the cortex.

A common signal for BCI is the P300 event-related potential, a positive deflection in the EEG over parietal cortex, that occurs approximately 300 ms after an "oddball" stimulus: a rare but meaningful stimulus among a series of frequently occurring stimuli. Since the P300 occurs among other ongoing EEG activity, several P300 responses must usually be averaged for the response to be recognized (Polich, 2007). Farwell and Donchin (1988) introduced the first P300-based BCI paradigm: computer presents a 6×6 matrix of letters and commands on-screen and participants attend to the item they wish to select; groups of matrix items are flashed randomly: only flashes of groups containing the attended item should elicit a P300. Items are grouped for flashing as rows and columns: hence, the so-called "*row-column paradigm*" (*RCP*). The computer identifies the attended item as the intersection of the row and column that elicited the largest P300.

The RCP has been tested in various configurations to achieve efficient communication that is practical for in-home use (Krusienski et al., 2006; Lenhardt et al., 2008; Sellers et al., 2006); the paradigm itself has been modified (Guger et al., 2009; Hong et al., 2009; Martens et al., 2009; Salvaris & Sepulveda, 2009; Takano et al., 2009). Unfortunately, none of such alternative paradigms substantially improves P300-based BCI performance. The RCP remains subject to errors that slow communication, cause frustration and diminish attentional resources (Vaughan et al., 2006). Further RCP research could possibly help severely disabled BCI users, who desire speed, accuracy, and ease of use. Moreover, with the RCP, some people are not able to achieve accuracy high enough for practical BCI use (Sellers & Donchin, 2006).

In recognition of these issues, Townsend and co-workers (2010) sought to create an alternative stimulation paradigm that could be faster, more accurate and more reliable than the RCP: they designed the so-called "*checkerboard paradigm*" (*CBP*), using a standard 8×9 matrix of alphanumeric characters and keyboard commands. In the RCP, the 8 columns and 9 rows flash at random: in contrast, in the CBP, the standard matrix is virtually superimposed on a checkerboard which the subjects never actually see. The items in white cells of the standard matrix are segregated into a white 6×6 matrix and the items in the black cells are segregated into a black 6×6 matrix; before each sequence of flashes, the items randomly populate the white or black matrix, respectively. The end result is that the subjects see random groups of 6 items flashing (as opposed to rows and columns), because the virtual rows and columns flash. In other words, the standard matrix never changes: only the pattern of flashing items is changed. After all rows and columns in both matrices have flashed (i.e., 24 flashes, comprising one complete sequence), the program re-randomizes the positions of the items in each virtual matrix and the next sequence of flashes begins (Townsend et al., 2010). The CBP produced a significant increase in BCI performance and

user acceptability over the RCP, thus providing a substantially more effective BCI, which is so important for PALS management. Experimental data showed that, whereas average PALS performances were much lower than those of the healthy controls using the RCP, upon switching to the CBP, PALS performed only slightly lower than the healthy controls: patients improved their classification accuracy rates by an average of about 25 % after switching from the RCP to the CBP, whereas accuracy rates of control group improved only of 14 %, thus suggesting that the CBP improvements may be more pronounced for PALS than for healthy controls. In particular, for two patients, the improvement brought them into an accuracy range sufficient for effective BCI control, while previously their accuracy was not consistently enough for effective control.

Non-invasive BCI methods have been utilized more extensively than invasive methods for people with disabilities (Birbaumer & Cohen, 2007; Birbaumer et al., 2008; Gerven et al., 2009): unfortunately, whereas PALS and other patients in "locked-in" conditions have motivated research in this area, very few systems have been successfully used - such as that reported by Townsend and co-workers (2010). It has been postulated that some forms of cognitive impairment and changes in EEG signatures in late ALS stages may contribute to the lack of success using BCI technology (Iversen et al., 2008), as the technology was introduced after the participants had become "locked-in" (Gerven et al., 2009; Münte et al., 1998): really, the most successful application for communication has occurred in people at the beginning stages of ALS (Birbaumer et al., 1999; Birbaumer, 2006; Kubler et al., 2001).

Nowadays, AAC acceptance and use represent two areas of interest for physicians and scientists. Both involve PALS and their caregivers.

In the Ball et al.'s review (2004_b), those who rejected AAC had a co-occurring cognitive deficit or experienced a severe diseases, such as cancer, in addition to ALS.

Fried-Oken and co-workers (2006) reported very positive caregivers' attitudes toward AAC technology: those with greater AAC technology skills got greater rewards associated with caregiving.

In a follow-up study on 15 PALS, Ball and co-workers (2010) examined the acceptance, training, and extended use patterns of eye-tracking technology to support communication.

For 53% of the participants, eye-tracking technology was selected because eye movement was the only viable access option available. More than 90% of the participants reported successful implementation of the technology: the only one patient who was not able to successfully use eye-tracking technology had difficulty with eyelid control. The communicative functions served by eye-tracking devices were extensive: all of the participants used their device to support face-to-face communication, and other functions included group communication, phone, e-mail and internet. More than 40% of the participants also reported using the eye-tracking technology to support other computer-based functions (e.g., word processing, voice-related software programs).

Training and support are an essential component of AAC service delivery for PALS. The significant changes in movement abilities require that service providers not only be proactive in their AAC technology recommendations by fitting up technology options that can meet the changing physical needs over time, but also by supplying adequate training and support to ensure that PALS and their caregivers can successfully implement diachronically these access strategies. Reports of low AAC use often are related to descriptions of minimal training or follow-up (Murphy, 2004).

New advances in AAC technology may need a greater amount of training and intervention than other access options: for instance, implementation of eye-tracking systems often requires for successful technology use trouble-shooting in the form of physical or environmental compensations (Ball et al., 2010). Whereas AAC specialists are professionals who provide the AAC intervention services (such as assessment and initial instruction), AAC facilitators for PALS tend to be family members who typically provide ongoing support (including instruction of new communication partners and caregivers, programming new messages into the AAC device, maintaining the AAC system, and interacting with the technology manufacturer, if necessary) (Beukelman et al., 2008).
In a survey on 68 PALS using AAC technology Ball and co-workers (2005_b) studied the AAC facilitators: almost all of them were family members, the majority with nontechnical backgrounds. They reported to prefer hands-on and detailed step-by-step instruction; and to have received an appropriate training amount (slightly over 2 hours of instruction).

5. Conclusion

Multifunctional impairments of PALS result from a relentlessly progressive muscle weakness, leading ultimately to a widespread body paralysis. In the late disease stages, patients eventually find themselves in a "locked-in" state, totally unable to move neck, trunk and limbs; autonomously breath and feed; and speak, although most of them retain their cognitive skills, thereby assisting impotent at their dreadful somatic decay.
As human beings, and therefore "persons", namely "individuals within a network of relationships", PALS particularly suffer from communication impairment.
Today, much more than in the past, we are able to give interdisciplinary assistance to them, enhancing their possibilities of keeping in touch with their caregivers, friends and other persons, with the aim to maintain their quality of life as high as possible.
Further research is needed to better implement AAC devices and services (trying to optimize communication aids and interfaces, and increase our understanding of acceptance and use of AAC approaches); and to develop new intervention strategies and document their effectiveness.
Anyway, besides scientific and clinical achievements, we look forward to building up in the near future more empathic care strategies for PALS and their families, with respect to them in their dignity of suffering persons.

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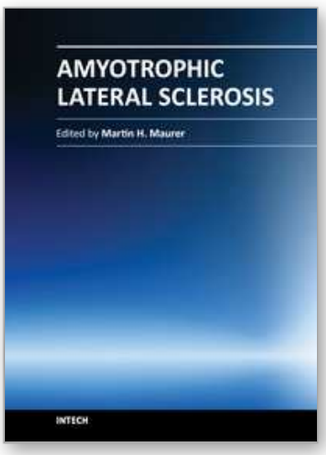

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Though considerable amount of research, both pre-clinical and clinical, has been conducted during recent years, Amyotrophic Lateral Sclerosis (ALS) remains one of the mysterious diseases of the 21st century. Great efforts have been made to develop pathophysiological models and to clarify the underlying pathology, and with novel instruments in genetics and transgenic techniques, the aim for finding a durable cure comes into scope. On the other hand, most pharmacological trials failed to show a benefit for ALS patients. In this book, the reader will find a compilation of state-of-the-art reviews about the etiology, epidemiology, and pathophysiology of ALS, the molecular basis of disease progression and clinical manifestations, the genetics familial ALS, as well as novel diagnostic criteria in the field of electrophysiology. An overview over all relevant pharmacological trials in ALS patients is also included, while the book concludes with a discussion on current advances and future trends in ALS research.

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