

Augmentative and alternative communication improves quality of life in the early stages of amyotrophic lateral sclerosis

Giuseppa Maresca, PsyD
Federica Pranio, PsyD
Antonino Naro, MD, PhD
Rosaria De Luca, MSc
Maria Grazia Maggio, PsyD
Ileana Scarcella, PsyD
Carmela De Domenico, PsyD
Placido Bramanti, MD
Valeria Conti Nibali, MSc
Simona Portaro, MD, PhD
Rocco Salvatore Calabrò, MD, PhD

IRCCS Centro Neurolesi, "Bonino Pulejo", Messina, Italy

Correspondence to: Rocco Salvatore Calabrò
E-mail: salbro77@tiscali.it

Summary

This study aimed to evaluate the efficacy of low-tech augmentative and alternative communication (AAC) aids in improving quality of life (QoL) and mood disorders, as well as the psychosocial impact of assistive devices, in 10 patients affected by amyotrophic lateral sclerosis (ALS) in the early stage of the disease, when speech difficulties appear. The AAC-training (AAC-T) study lasted around six months and comprised two phases of three months each: AAC-intervention (AAC-I) followed by AAC-familiarization (AAC-F). All the patients were assessed at the beginning (T0) and at the end (T1) of AAC-I, as well as at the end of AAC-F (T2). During the AAC-I phase, we applied a three-phase AAC intervention model to evaluate intelligibility of language, participation patterns, communication needs and adaptation to the AAC tools. All the patients showed a gradual and significant improvement, especially in acceptance of the AAC aids, mood and QoL. Moreover, a reduction of caregiver burden was noted. Our study has demonstrated the utility of the AAC aids also in the early stages of the disease in patients with ALS presenting with dysarthria. In our opinion, AAC-T may act as a bridge between the initial and later phases of the disease, when patients need to use high-tech aids, including an eye-tracking communication device. In conclusion, in this study we highlighted how early AAC-T in patients with ALS may be a valuable tool to demonstrate how specific strategies and low-technology aids can improve QoL of these patients and their caregivers, also decreasing stress and depression.

KEY WORDS: *assessment, communication, electronic aids to daily living, neuromuscular diseases.*

Introduction

Amyotrophic lateral sclerosis (ALS) is a progressive neurodegenerative multisystem disorder, characterized by the loss of cortical, brain stem and spinal motor neurons (Calvo et al., 2014), but usually sparing cognitive, sensory, sexual and sphincter functions (Yoshizawa et al., 2014). The main ALS presentations are the limb-onset form (70%), which is the most typical, and the bulbar-onset form (25%), which is characterized by speech and swallowing difficulties. Although the progression of speech disorders may vary from person to person, individuals with bulbar involvement have early dysarthria (which in the most severe forms evolves into anarthria); furthermore, the functions involved in the swallowing process can rapidly worsen (Makkonen et al., 2018).

Instead, people with spinal involvement present serious motor problems; on the other hand, their language skills can be preserved in the early stage of the disease, but usually worsen with its progression (Van Es et al., 2017). A proportion of ALS patients, ranging from 1 to 40%, may exhibit mild cognitive impairment with the clinical features of frontotemporal dementia (Consonni et al., 2013; Phukan et al., 2012), i.e. behavioral abnormalities and executive dysfunctions.

Considering that ALS is a rapidly worsening disease, taking care of the caregiver burden is also essential in order to maintain the physiological equilibrium within the family, allowing it to cope better with the difficulties related to the disease in all its phases (Foley et al., 2014). In fact, the patient's growing need for assistance exposes the caregiver and all the family members to pressure, conflict and considerable stress (Adelman et al., 2004). Studies reveal that caregivers, when subjected to considerable and prolonged stress, easily develop anxiety and depression and are more likely to get sick, partly because of the number of hours spent caring (Pagnini et al., 2011; Lillo et al., 2012; Palmieri et al., 2012). For this reason, in order to ensure the wellbeing of both patients and caregivers, it is very important to support and help the latter (Burke, 2017). Involving the caregiver in the multidisciplinary team taking care of the patient with ALS is mandatory in order to familiarize the caregiver with the disease, and thus support the care of the patient at home. In fact, the caregiver's feelings of helplessness and frustration are reduced when he/she is better able to understand the needs and emotions of the patient, and the characteristics of the disease (Burke, 2015). Moreover, in this way, a caregiver can be helped to confront the difficulties related to the communication barrier created by the disease.

Given the important social function of language, the

speech disorder in ALS may be considered one of its most disabling symptoms, and a major problem. To address it, augmentative and alternative communication (AAC) training may be a valuable tool, worth considering even in the early stages of the disease. Indeed, early AAC training (AAC-T), even using low-technology aids, is necessary when speech difficulties appear, in order to support communication and functional needs of daily life, preserve dignity and autonomy, and prevent loss of social motivation.

The Participation Model is a systematic process model for performing AAC assessment and for planning interventions for people with complex communication needs (CCN) (Beukelman and Mirenda, 2012). This model evaluates the quality of participation, considering four areas: participation schemes, barriers to participation, effectiveness of previous strategies to improve participation, and potential new strategies. The barriers to participation are of two types: barriers to communication opportunities and barriers to communication access. The former occur when the person with CCN (in this case the patient with ALS) cannot participate because his/her own facilitator (i.e. communication partner) does not have the skills necessary to support his/her efforts to communicate, whilst the latter type are due to impaired abilities of the person with CCN, or to limits of his/her communication system (Beukelman and Yorkston, 1989; Blackstein-Adler, 2004). At the initial stage of ALS, most patients use mobile phones, laptops and personal computers, whereas multimodal AAC systems, i.e. involving gestures, communication boards, speech-generating devices, writing, symbols and photographs, may better meet their communication needs (Ray, 2015; Ball et al., 2004). Patients with ALS and their caregivers usually have difficulty accepting the patient's speech difficulties; as a result, the former will go on striving, with difficulty, to communicate, and the latter to understand, and they will fail to seek easier solutions (Galvin et al., 2017). Instead, early introduction of AAC strategies and devices would be appropriate, and would also help to sustain quality of life (QoL) and prevent depression in patients with ALS (Ray, 2015; Ball et al., 2004). Nonetheless, patients are reluctant to use communication aids in the early stages of ALS, as using AAC tends, initially, to increase their feelings of frustration; furthermore, given that it forces them to acknowledge their communication limits, it can lead to depressed mood and anxiety (Beukelman et al., 2011).

Thus, psychological support, aimed at helping patients to accept the progression of the disease and appreciate the benefits of AAC, is mandatory in order to adequately manage language problems and CCN, and teach patients, in advance, how to use the high-tech communication aids, e.g. eye-tracking communication devices and brain computer interfaces, that will be increasingly useful as their ALS advances.

The most appropriate AAC system for the patient should be identified, according to the type of ALS (Doyle and Phillips, 2001).

The aim of this study was to evaluate the efficacy of low-tech AAC aids in improving QoL and mood disorders in patients affected by ALS in the early stages of the disease. Another aim is to assess the psychosocial impact of assistive devices on patients and the influence of these tools on the caregiver burden.

Materials and methods

Study population

Ten patients with ALS (5M – 5F; mean \pm SD age: 71.5 \pm 8.5 years; mean \pm SD education: 8.1 \pm 2.2 years) and their primary caregiver (4M – 6F; mean \pm SD age: 53.2 \pm 10.2 years; mean \pm SD education: 12.7 \pm 3.6 years), consecutively admitted to the Rare Diseases Unit of the IRCCS Centro Neurolesi “Bonino-Pulejo”, Messina, Italy, from March to December 2017, were enrolled in the study. Table I provides more details on the patients and caregivers.

The inclusion criteria were: i) age range 18-80 years; ii) diagnosis of ALS according to the revised El Escorial criteria (Brooks et al., 2000; De Carvalho et al., 2008); iii) Mini-Mental State Examination score $>$ 24.

The exclusion criteria were: i) the presence of concomitant frontotemporal dementia; ii) an alteration in cognitive functioning as shown by a Frontal Assessment Battery (Dubois et al., 2000) score of under 8.5, and a Brief Psychiatric Rating Scale (Ventura et al., 1993) score of over 39; iii) a response other than “Detectable speech disturbance” or “Intelligible with repeating” on the “Speech” item of the ALS Functional Rating Scale – Revised (Brooks et al., 1996); iv) concomitant severe medical illness.

All the patients and their caregivers were adequately informed about the study and they signed an informed consent to participate in it. The Ethics Committee of the IRCCS Centro Neurolesi “Bonino-Pulejo” approved this study.

Assessment tools

The whole study lasted around six months and comprised two phases of three months each: AAC-intervention (AAC-I) followed by AAC-familiarization (AAC-F). All the patients were assessed by means of neurological, neuropsychological and neurobehavioral tests at the beginning (T0) and at the end (T1) of AAC-I, as well as at the end of AAC-F (T2).

The assessment included the Edinburgh Cognitive and Behavioral ALS Screen (ECAS) (Poletti et al., 2016) to evaluate emotional and behavioral involvement; the Caregiver Burden Inventory (CBI) (Novak and Guest, 1989) to evaluate the different dimensions of the caregiver burden; the Hamilton Rating Scale for Anxiety (HRS-A) (Hamilton, 1959) and the Beck Depression Inventory-II (BDI-II) (Dozois et al., 1998) to self-assess severity of depression; the 36-Item Short Form Survey Instrument (SF-36) (Apolone and Mosconi, 1998) to evaluate patient health status, and the Coping Orientation to Problems Experienced (COPE) tool (Carver et al., 1989) to evaluate coping strategies. In order to assess the effects of the aids on functional independence, psychological well-being and ALS patients' QoL (at T1 and T2), we administered the Psychosocial Impact of Assistive Devices Scale (PIADS) (Day, 1996).

Table II provides more details on the assessment tools. Moreover, in the initial phase, the AAC specialist administered, to each patient, the Participation Inventory (Blackstein-Adler, 2004), i.e. a form for detecting patterns of communicative participation (Fig.1), and an Observation Board for Functional Communication Skills (Fig. 2) to evaluate different communication skills displayed by the patient during a session.

Table I - Demographic and clinical characteristics of the patients and their caregivers.

	Male	Female	All
PATIENTS			
Age (58-87 years)	5 (50%) 73.4 ± 6.3	5 (50%) 69.6 ± 10.8	10 (100%) 71.5 ± 8.5
Education			
Elementary	1 (20.0%)	1 (20.0%)	2 (20%)
Middle	3 (60.0%)	4 (80.0%)	7 (70.0%)
High	1 (20.0%)	-	1 (10.0%)
University	-	-	-
Type			
Bulbar	1 (20%)	1 (20%)	2 (20%)
Spinal/Limb	3 (60%)	1 (20%)	4 (40%)
Mixed	1 (20%)	3 (60%)	4 (40%)
Duration of illness (36-4 months)	21.6 ± 10.5	16.4 ± 6.8	19 ± 8.8

Mean ± standard deviation was used to describe continuous variables; proportions (numbers and percentages) were used to describe categorical variables.

	Male	Female	All
CAREGIVERS			
Age (44-67 years)	4 (40%) 51.5 ± 15.4	6 (60%) 54.3 ± 15.2	10 (100%) 53.2 ± 8.5
Education			
Elementary	-	-	-
Middle	1 (25.0%)	2 (33.3%)	3 (30.0%)
High	2 (50.0%)	2 (33.3%)	4 (40.0%)
University	1 (25.0%)	2 (33.3%)	3 (30.0%)
Relationship to patient			
Son/daughter	3 (75.0%)	3 (50.0%)	6 (60.0%)
Spouse/partner	1 (25.0%)	2 (33.3%)	3 (30.0%)
Other	-	1 (16.6%)	1 (10.0%)

Mean ± standard deviation was used to describe continuous variables; proportions (numbers and percentages) were used to describe categorical variables.

Procedures

The AAC-I lasted three months (T0-T1) and it was conducted by a skilled speech therapist and neuropsychologist, who used the three-phase (baseline, intermediate, final) version (Ball et al., 2004) of the Participation Model (Beukelman and Yorkston, 1989; Ball et al., 2004; Beukelman et al., 2011). The purpose of the baseline phase was to: i) monitor the performance of language in a systematic way; ii) perform a timely AAC assessment; and iii) set up the AAC-I, before the patient showed severely reduced intelligibility of the word. In the intermediate phase we: i) evaluated patterns of participation, communication needs, and personal interests; ii) assessed present and future motor, cognitive, linguistic and sensory abilities through a clinical assessment; iii) evaluated the presence of social and personal support; iv) refined the low-tech AAC strategies and aids, in order to obtain those best suited to the needs of the ALS patient. In the final phase, we provided the patients and caregivers with effective communication options, in order to cater for changes in communication needs and abilities with disease progression (Beukelman and Yorkston, 1989; Ball et al., 2004).

At the end of AAC-I, we provided the patients with three paper communication tables, laminated and bound as a notepad: i) an alphanumeric table, at which to point in order to compose sentences quickly; ii) a table with preformed sentences, established during individual interviews with the patients; iii) a table of picture communication symbols, to be used for the identification and classification of pain (Fig. 3).

We opted to use symbols because visual-symbolic supports are an immediate and simpler means of expressing an abstract concept. Consideration was given to the number, size and arrangement of the items in the tables (i.e. letters/numbers/symbols), taking into account the patients' upper limb movement ability and selection skills. In each table, customized to the single patient, there were series of symbols representing specific categories within a conceptual pyramid of human need, which is a model for categorizing people's needs (David et al., 2009; Collier, 2000). The categories were Physical, Functional, Social (or Behavioral) and Emotional. The "Physical" category includes personal care, health, daily routine, food and drink; the "Functional" category includes free time and daily activities; the "Social" cate-

Table II - Assessment tools.

TEST/SCALE	DOMAIN	DESCRIPTION
Edinburgh Cognitive and Behavioral ALS Screen (ECAS)	Cognition and behavior	The ECAS is a short screen for cognitive/behavioural alterations in patients with ALS. In total ECAS consists of 15 individual tasks correlated with certain cognitive functions which are divided into 5 different subdomains (language, verbal fluency, executive, memory and visuospatial). These subdomains comprise ALS-specific functions (language, verbal fluency and further executive) and non-specific functions (memory and visuospatial) for which scores can be identified, respectively. The ECAS-total score (cut-off=105) is the sum of the scores of all 15 individual tasks. It is a global measure for the evaluation of cognitive impairments in ALS patients.
Hamilton Anxiety Rating Scale (HRS-A)	Anxiety	The HRS-A is a rating scale developed to measure the severity of anxiety symptoms; the scale consists of 14 items, each defined by a series of symptoms, and measures both psychic and somatic anxiety. Each item is scored for severity on a scale of 0 (not present) to 4 (severe), giving a total score range of 0-56, where <17 indicates mild, 18-24 mild to moderate, and 25-30 moderate to severe.
Beck Depression Inventory – II (BDI-II)	Depression	The BDI-II is a 21-question multiple-choice self-report inventory for measuring the severity of depression. It is composed of items relating to symptoms of depression such as hopelessness and irritability, cognitions such as guilt or feelings of being punished, as well as physical symptoms such as fatigue, weight loss, and lack of interest in sex. BDI-II items are rated on a 4-point scale ranging from 0 to 3 based on the severity of each item. A raw score of 0-13 indicates minimal depression; 14-19 mild depression; 20-28 moderate depression; 29-63 (maximum total score) severe depression.
Psychosocial Impact of Assistive Devices Scale (PIADS)	Psychosocial	The PIADS is a 26-item, self-report questionnaire used to assess the effects of an assistive device on functional independence, well-being, and quality of life. The three subscales of the PIADS are Competence, Adaptability and Self-esteem. Competence (12 items) measures feelings of competence and efficacy. Adaptability (6 items) indicates the degree of willingness to try out new things and take risks. Self-esteem (8 items) indicates feelings of emotional health and happiness. Scores can range from -3 (maximum negative impact) through zero (no perceived impact) to +3 (maximum positive impact).
36-Item Short Form Survey Instrument (SF-36)	Health status	The SF-36 is a set of generic, coherent, and easily administered quality-of-life measures. It consists of eight scaled scores (vitality, physical functioning, bodily pain, general health perceptions, physical role functioning, emotional role functioning, social role functioning and mental health), which are the weighted sums of the questions in their section. Each scale is directly transformed into a 0-100 scale on the assumption that each question carries equal weight. The lower the score the more disability. The higher the score the less disability i.e., a score of zero is equivalent to maximum disability and a score of 100 is equivalent to no disability.
The Brief Coping Orientation to Problems Experienced (COPE)	Coping strategies	The Brief COPE inventory is the measure usually used to identify the nature of coping strategies implemented by individuals and it explores 14 coping strategies. The scale consists of 14 subscales with two items per subscale measuring: Self-distraction, Active coping, Denial, Substance use, Use of emotional support, Use of instrumental support, Behavioral disengagement, Venting, Positive reframing, Planning, Humor, Acceptance, Religion and Self-blame. For each subscale the score can range from 2 to 8 with higher scores indicating greater use of a specific coping strategy.
Caregiver Burden Inventory (CBI)	Care load	The CBI is a scale used to quantify burdens in different aspects of a caregiver's life, and the influence of patients' and caregivers' characteristics on its different dimensions. CBI comprises 24 closed questions divided into five dimensions of caregiver burden: time-dependence, developmental, physical, social and emotional. Each item is given a score of between 0 (not at all descriptive) and 4 (very descriptive), where higher scores indicate greater caregiver burden; there are no cut-off points for classifying burden.

to be continued

Table II - Continue.

Participation Inventory	The Participation Inventory is a record of the communicative participation of the person with complex communication needs (CCN). This Inventory evaluates the quality of participation in four areas: participation schemes, barriers to participation, effectiveness of previous strategies to improve participation and potential new strategies. Furthermore, the level of independence of the person with CCN is observed and recorded.
Communication skills	
Communication Observation Board for Functional Communication Skills	The Observation Board for Functional Communication Skills is an observational grid used to evaluate different abilities of a patient with ALS during a session: paying attention to and requesting the attention of the therapist; joint attention; expressing "yes/no" and "more/stop"; taking turns in an activity; shifting attention at the request of the therapist during an activity; requesting objects; ability to start/maintain/end a conversation.

Directions:

1. List the *primary and secondary* activities in which peers without disabilities are expected to participate.
2. Select one or more peers without disabilities who are typical in terms of their ability to achieve the expected standards. After observing one of the peers in each activity listed, indicate the *level of peer participation* achieved by entering a *P* in the appropriate category for each activity.
3. After observing the target individual in each activity, indicate the *level of participation* achieved by entering a *T* in the appropriate category for each activity.
4. In the Discrepancy column, indicate *yes* if a participation gap exists for the target individual compared to peers and *no* if a participation gap does not exist.
5. Based on your observations and impressions, indicate if the barrier to participation appears to be related to *opportunity barriers, access barriers, or both*.

Activity	Level of peer participation					Discrepancy		Type(s) of barrier(s)	
	Independent	Independent with set-up	Verbal assistance	Physical assistance	Unable to participate	Yes	No	Opportunity	Access
	1. _____								
2. _____									
3. _____									
4. _____									
5. _____									
6. _____									

Figure 1 - The Participation Inventory (Blackstein-Adler, 2004).

gory includes the social networks of the person with complex communication needs, meaning family, friends and caregivers.

Finally, the "Emotional" category covers all the patient's feelings. All the ALS patients were questioned to allow us to insert phrases for each category, according to their own communication needs.

In the AAC-F phase (T1-T2), lasting three months, we used the communication tables in the context of highly motivating activities for the patient (i.e. listening to music, watching and commenting on movies or video clips, playing cards or board games, etc.), so that they might accept them more easily.

In addition, using them over time, the patients were able to

construct a sort of mental map of how to use and combine letters/numbers/symbols during the communicative act (Trudeau et al., 2007; Schlosser, 1997).

Statistical analysis

Statistical analyses were carried out using Statview software (version 5; SAS Institute Inc., Cary, NC). The Shapiro-Wilk statistic was used to test the normality of the distribution of the variables (all non-normally distributed, $p < 0.05$).

A Wilcoxon test was used to estimate the differences between baseline, post-treatment, and follow-up. The level of statistical significance was set at $p < 0.05$ (two-tailed).

Observation Board for Functional Communication Skills						
		PATIENT			SETTING	
		MODALITY	FREQUENCY	CONTENT	ACCESS BARRIERS	SETTING PARTNER
1	ATTENTION TO COMMUNICATION PARTNER					
2	ATTENTION REQUEST					
3	REFUSAL OF COLLABORATION					
4	JOINT ATTENTION					
5	TURN ALTERNATION					
6	EXPRESSION OF "YES" "NO"					
7	EXPRESSION OF "MORE" "STOP"					
8	EXPRESSION OF COMMENTS					
9	ABILITY TO GENERALIZE					
10	ABILITY TO CONTEXT					
11	SELECTION					
12	SPONTANEOUS REQUEST OF HELP					
13	COMMUNICATIVE INITIATIVE					
14	HOW START CONVERSATION					
15	HOW PARTICIPATE / ENCLOSE CONVERSATION					
16	NON-VERBAL COMMUNICATION					
17	VERBAL COMMUNICATION					

Figure 2 - Observation Board for Functional Communication Skills

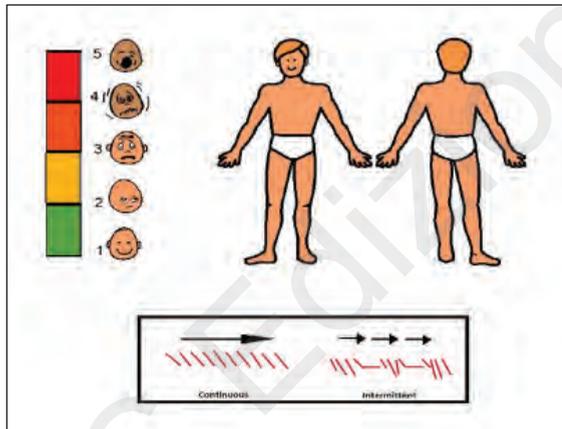


Figure 3 - Sample of AAC table. Example of an AAC table using PCS symbols for identification and rating of pain. The pain intensity scale consists of facial expressions denoting different levels of pain. A whole human figure, front and backs, serves to identify the area of the pain. There are also symbols for indicating the intensity of the pain (continuous or intermittent).

Results

The AAC-T was well tolerated, and all the enrolled patients completed the study.

The Wilcoxon test showed significant improvements in nearly all the variables we investigated (Table III). Evaluation of the scores recorded in AAC-I (T0 vs T1) revealed

significant improvements in anxiety (HRS-A), in two COPE subscales — i.e. “Self-distraction” and “Active coping” — and in some of the SF-36 subscales, i.e. “Energy/vitality”, “Physical functioning”, “Change in health”, “Social functioning” and “Mental health” (all p-level ≤ 0.01).

Moreover, the patients recorded significant improvements in cognitive performances (ECAS Total and Non-Specific) (all p-level ≤ 0.02) and the caregivers showed a significant reduction in psychological, social and emotional burden, as shown by the relative CBI subscales (all p-level ≤ 0.01). As regards the scores recorded at the end of AAC-F as compared to the end of AAC-I (T2 vs T1), significant improvements were also noted in depression (BDI), some COPE subscales — i.e. “denial” and “use of emotional and instrumental support” — and in some of the SF-36 subscales, i.e. “role limitation-physical” and “role limitation-mental” (all p-level ≤ 0.04). All the PIADS sub-items also showed significant improvements (all p-level ≤ 0.004), while the “objective burden” of caregivers decreased significantly (p-level ≤ 0.03).

Finally, the AAC-T (T2 vs T0) showed further improvements, particularly with regard to cognitive performances (ECAS), mood (BDI and HRS-A) and QoL (SF-36) (all p-level ≤ 0.04), and improved adaptive coping strategies (COPE) (all p-level ≤ 0.03).

Discussion

To the best of our knowledge, this is the first attempt to use AAC as a means of improving the management of ALS patients from the early stages of the disease, when speech difficulties appear. ALS is a progressive multisystem neurodegenerative disorder, characterized by loss of cortical, brain stem and spinal motor neurons. Given that speech and swallowing disorders are among its most common symptoms, especially in the bulbar form, and considering the negative burden of speech difficulties, interventions aimed at improving communication are mandatory, even in the early stages of the disease.

Our study showed the efficacy of AAC-T in the management of communication disorders in ALS from the early stages of the disease, as well as demonstrating that AAC low-tech aids had a positive impact on patients’ QoL and mood. Indeed, our results revealed that all the ALS patients undergoing AAC-T showed improvements in the several areas we investigated. In particular, the data highlighted significant improvements in mood, global cognitive functioning, and use of adaptive and functional coping strategies, as well as positive effects of assistive devices on functional independence, well-being and QoL.

Moreover, caregivers’ CBI scores significantly improved, especially on the psychological and emotional subscales. Bongioanni (2012) found that early use of AAC aids (including low-tech tools) is a useful means of improving communication, addressing needs, supporting functional activities of daily life, preserving dignity and autonomy, and preventing loss of motivation. It has also been shown that, during the early stages of ALS, patients struggle to accept the changes brought by the disease and, despite presenting dysarthria, continue to try to use their residual language (Fried-Oken, 2015), which leads to distress, depressed mood, a sense of inadequacy, low self-esteem and consequently impairment of

Table III - Median values with first and third quartile at each time point.

		T0	T1	T2	T0-T1	T1-T2	T0-T2
ECAS	Total	66 44 - 82	73 48 - 91	77 56 - 98	0.02	0.1	0.007
	Specific	54 38 - 65	57 39 - 74	59 43 - 73	0.1	0.1	0.04
	Non-specific	13 9 - 16	15 11 - 19	18 13 - 22	0.002	0.1	0.002
HRS-A		9 6 - 11	7 5 - 8	6 4 - 7	0.007	0.2	0.002
BDI-II		14 10 - 17	12 8 - 16	10 7 - 13	0.2	0.003	0.008
SF36	PF	16 10 - 20	19 13 - 24	34 25 - 42	0.04	0.02	0.009
	RLP	20 15 - 24	24 17 - 30	43 30 - 53	0.5	<0.001	0.004
	P	52 34 - 68	64 47 - 80	71 46 - 88	0.1	0.3	0.04
	CiH	23 17 - 29	32 24 - 41	43 29 - 53	0.006	0.005	0.001
	EV	29 21 - 36	44 30 - 57	52 35 - 64	0.003	0.1	0.001
	SC	64 46 - 78	77 58 - 95	85 56 - 106	0.02	0.1	0.006
	RLM	50 32 - 64	66 44 - 84	80 58 - 98	0.1	0.04	0.001
CBI	MH	53 36 - 67	62 43 - 78	9 6 - 11	0.006	<0.001	<0.001
	OB	8 5 - 10	7 5 - 9	8 6 - 10	0.1	0.02	0.5
	PB	6 5 - 8	5 4 - 7	7 5 - 9	0.001	0.03	0.3
	PhB	6 4 - 8	6 4 - 8	7 5 - 9	0.1	0.4	0.5
	SB	4 3 - 6	4 3 - 5	6 4 - 8	0.01	0.1	0.1
COPE	EB	4 3 - 5	3 2 - 4	6 4 - 7	0.008	0.03	0.3
	SD	5 3 - 6	4 3 - 5	4 2 - 4	0.02	0.3	0.03
	AC	5 3 - 7	6 4 - 8	4 3 - 5	0.01	0.01	0.1
	D	4 2 - 5	3 2 - 4	2 1 - 3	0.2	0.005	0.008
	SU	2 1 - 2	2 1 - 3	5 4 - 6	0.9	<0.001	<0.001
	UES	6 4 - 7	6 4 - 8	4 3 - 5	0.3	0.001	0.003
	UIS	4 3 - 5	4 3 - 5	5 4 - 7	0.8	0.02	0.02
	PB	5 3 - 6	5 3 - 6	5 4 - 6	0.9	0.8	0.9
	V	5 3 - 6	5 4 - 7	5 4 - 7	0.5	0.8	0.3
	PR	5 3 - 5	5 3 - 6	5 4 - 7	0.3	0.2	0.1
	P	5 4 - 7	6 4 - 7	6 4 - 8	0.1	0.3	0.01
	H	4 3 - 5	4 3 - 5	4 2 - 5	0.8	0.6	0.5
	A	6 4 - 7	6 4 - 7	6 4 - 8	0.6	0.6	0.5
	R	4 2 - 4	4 2 - 4	4 3 - 5	0.9	0.9	0.9
SB	4 3 - 6	4 3 - 5	4 3 - 5	0.7	0.7	0.6	
PIADS	Competence		0.6 0.4 - 0.8	0.7 0.5 - 0.9		0.004	
	Adaptability		0.9 0.7 - 1.1	1.1 0.8 - 1.4		0.002	
	Self-esteem		1.0 0.7 - 1.3	1.3 0.9 - 1.7		0.000	

Abbreviations: ECAS=Edinburgh Cognitive and Behavioral ALS Screen; HRS-A= Hamilton Rating Scale for Anxiety; BDI-II=Beck Depression Inventory-II; SF-36= 36-Item Short Form Survey - Subscales: Physical Function (PF); Role Limitation - Physical (RLP); Pain (P); Change in Health (CiH); Energy/Vitality (EV); Social Function (SC); Role Limitation - Mental (RLM); Mental Health (MH); CBI= Caregiver Burden Inventory - Subscales: Objective Burden (OB); Psychological Burden (PB); Physical Burden (PhB); Social Burden (SB); Emotional Burden (EB); COPE= Coping Orientation To Problems Experienced - Subscales: Self-distraction (SD); Active Coping (AC); Denial (D); Substance Use (SU); Use of Emotional Support (UES); Use of Instrumental Support (UIS); Behavioral Disengagement (BD); Venting (V); Positive Reframing (PR); Planning (P); Humor (H); Acceptance (A); Religion (R); Self-blame (SB); PIADS=The Psychosocial Impact of Assistive Devices Scale.

their and their caregivers' QoL (Gauthier et al., 2007). Thus, in line with previous data, we think that providing ALS patients with an alternative method of communicating and expressing needs, opinions and thoughts from an early stage can facilitate their acceptance of AAC aids and help improve their interpersonal relationships, mood, perceived QoL, and capacity to adapt, as well as their self-esteem and ability to communicate their needs and opinions (Corallo et al., 2017).

Another important aspect investigated in our work is the burden of ALS on caregivers.

The caregiver and all the members of the family are exposed to pressure and severe stress due to the af-

ected individual's growing care needs (Adelman et al., 2004; Pagnini et al., 2011; Lillo et al., 2012; Palmieri et al., 2012). In fact, it is very important to support and help them, too, so as to reduce their feelings of impotence and frustration. In this regard, it should be noted that active and collaborative caregivers have lower stress levels (Burke et al., 2017).

Furthermore, at both the beginning and the end of AAC-F, we assessed, using the PIADS scale, the degree of satisfaction with the AAC aids. We found that the low-tech AAC aids had a good psychosocial impact on patients with ALS in all the investigated sub-scales: competence, adaptability and self-esteem. The AAC ap-

proach offered each patient a channel of communication, allowing them to express their needs and feelings to family and caregivers.

Also, this may have helped the caregivers to confront the difficulties related to the communication barrier created by the disease.

Although AAC aids have already been shown to improve QoL and reduce communication disability in late-stage ALS patients with anarthria (Caligari et al., 2013), in this study, we demonstrated that they also show promising utility in the early stages of ALS in patients with mild to moderate dysarthria. In our opinion, the AAC-T phase may act as a bridge between the initial and later phases of the disease, when patients need to use high-tech aids, including an eye-tracking communication device.

The main limitation of the study is the small size of the sample and the lack of a control group. However, this is a pilot study, whose findings need to be confirmed by a randomized multicenter clinical trial.

In conclusion, in this study we highlighted how early AAC-T in patients with ALS may be a valuable tool. Indeed, this integration of medical and psychological evaluation with AAC-T provided an effective demonstration of how strategies and low-technology aids can improve the QoL of these individuals and their caregivers, also decreasing stress and depression.

Although our findings await confirmation in the near future, we believe that providing a clear diagnosis and prognosis and proposing an alternative communication method could favor a better management of ALS patients and reduce their caregivers' burden.

References

- Adelman EE, Albert SM, Rabkin JG, et al (2004). Disparities in perceptions of distress and burden in ALS patients and family caregivers. *Neurology* 62:1766-1770.
- Apolone G, Mosconi P (1998). The Italian SF-36 Health Survey: translation, validation and norming. *J Clin Epidemiol* 51:1025-1036.
- Ball LJ, Beukelman DR, Pattee GL (2004). Acceptance of augmentative and alternative communication technology by persons with amyotrophic lateral sclerosis. *Augment Altern Commun* 20:113-122.
- Beukelman D, Fager S, Nordness A (2011). Communication Support for People with ALS. *Neurol Res Inter* 2011:714693.
- Beukelman DR, Yorkston KM (1989). Augmentative and alternative communication application for persons with severe acquired communication disorders: An introduction. *Augmentative and Alternative Communication* 5:42-48.
- Beukelman DR, Mirenda P (2012). *Augmentative and Alternative Communication: Supporting Children and Adults with Complex Communication Needs – Fourth Edition*. Paul H. Brookes Publishing Co., Baltimore.
- Blackstein-Adler S (2004). Simple Technology for Participation. *Bridges Canada*. Retrieved October 17, 2011. From http://www.setbc.org/setbc/conf/docs/simple_technology_participation.html
- Bongioanni P (2012). Communication impairment in ALS patients assessment and treatment. In: Martin Maurer (Ed.) *Amyotrophic Lateral Sclerosis*, InTech: 665-682.
- Brooks BR, Miller RG, Swash M, et al (2000). El Escorial revisited: revised criteria for the diagnosis of amyotrophic lateral sclerosis. *Amyotroph Lateral Scler Other Motor Neuron Disord* 1:293-299.
- Brooks BR, Sanjak M, Ringel S, et al (1996). The amyotrophic lateral sclerosis functional rating scale-Assessment of activities of daily living in patients with amyotrophic lateral sclerosis. *Arch Neurol* 53(2): 141-147.
- Burke T, Elamin M, Galvin M, et al (2015). Caregiver burden in amyotrophic lateral sclerosis: a cross-sectional investigation of predictors. *J Neurol* 262: 1526-1532.
- Burke T, Galvin M, Pinto-Grau M, et al (2017). Caregivers of patients with amyotrophic lateral sclerosis: investigating quality of life, caregiver burden, service engagement, and patient survival. *J Neurol* 264: 898-904.
- Caligari M, Godi M, Guglielmetti S, et al (2013). Eye tracking communication devices in amyotrophic lateral sclerosis: Impact on disability and quality of life. *Amyotroph Lateral Scler Frontotemporal Degen* 14:546-552.
- Calvo AC, Manzano R, Mendonça DMF, et al (2014). Amyotrophic lateral sclerosis: a focus on disease progression. *Biomed Res Int* 2014:629630.
- Carver CS, Scheier MF, Weintraub JK (1989). Assessing coping strategies: a theoretically based approach. *J Pers Soc Psychol* 5:267-283.
- Collier B (2000). *See What We Say: Situational Vocabulary for Adults Who Use Augmentative and Alternative Communication*. Paul H. Brookes Publishing Co., Baltimore.
- Consonni M, Iannaccone S, Cerami C, et al (2013). The cognitive and behavioural profile of amyotrophic lateral sclerosis: application of the consensus criteria. *Behav Neurol* 27:143-153.
- Corallo F, Bonanno L, Buono VL, et al (2017). Augmentative and alternative communication effects on quality of life in patients with locked-in syndrome and their caregivers. *J Stroke Cerebrovasc Dis* 26:1929-1933.
- David R, Beukelman SF, Ball L, et al (2009). AAC for adults with acquired neurological conditions: A review. *Augment Altern Commun* 23:230-242.
- Day H (1996). Measuring the psychosocial impact of assistive devices: the PIADS. *Can J Rehabil* 9:159-168.
- De Carvalho M, Dengler R, Eisen A, et al (2008). Electrodiagnostic criteria for diagnosis of ALS. *Clin Neurophysiol* 119:497-503.
- Doyle M, Phillips B (2001). Trends in augmentative and alternative communication use by individuals with amyotrophic lateral sclerosis. *Augment Altern Commun* 17:167-178.
- Dozois DJ, Dobson KS, Ahnberg JL (1998). A psychometric evaluation of the Beck Depression Inventory–II. *Psychol Assess* 10:83.
- Dubois B, Slachevsky A, Litvan I, et al (2000). The FAB: a frontal assessment battery at bedside. *Neurology* 55:1621-1626.
- Foley G, Timonen V, Hardiman O (2014). Exerting control and adapting to loss in amyotrophic lateral sclerosis. *Soc Sci Med* 101:113-119.
- Fried-Oken M, Mooney A, Peters B (2015). Supporting

- communication for patients with neurodegenerative disease. *Neurorehabilitation* 37:69-87.
- Galvin M, Gaffney R, Corr B, et al (2017). From first symptoms to diagnosis of amyotrophic lateral sclerosis: perspectives of an Irish informal caregiver cohort—a thematic analysis. *BMJ Open* 7: e014985.
- Gauthier A, Vignola A, Calvo A, et al (2007). A longitudinal study on quality of life and depression in ALS patient-caregiver couples. *Neurology* 68:923-926.
- Hamilton M (1959). The assessment of anxiety states by rating. *Br J Med Psychol* 32:50-55.
- Lillo P, Mioshi E, Hodges JR (2012). Caregiver burden in amyotrophic lateral sclerosis is more dependent on patients' behavioral changes than physical disability: a comparative study. *BMC Neurol* 12:156.
- Makkonen T, Ruottinen H, Puhto R (2018). Speech deterioration in amyotrophic lateral sclerosis (ALS) after manifestation of bulbar symptoms. *Int J Lang Commun Disord* 53:385-392.
- Novak M, Guest C (1989). Caregiver Burden Inventory (CBI). *Gerontologist* 29:798-803.
- Pagnini F, Lunetta C, Rossi G, et al (2011). Existential well-being and spirituality of individuals with amyotrophic lateral sclerosis is related to psychological well-being of their caregivers. *Amyotroph Lateral Sclerosis* 12:105-108.
- Palmieri A, Kleinbub JR, Calvo V, et al (2012). Efficacy of hypnosis-based treatment in amyotrophic lateral sclerosis: a pilot study. *Front Psychol* 3:465.
- Phukan J, Elamin M, Bede P, et al (2012). The syndrome of cognitive impairment in amyotrophic lateral sclerosis: a population-based study. *J Neurol Neurosurg Psychiatry* 83:102-108.
- Poletti B, Solca F, Carelli L, et al (2016). The validation of the Italian Edinburgh cognitive and behavioural ALS screen (ECAS). *Amyotroph Lateral Scler Frontotemporal Degener* 17:489-498.
- Ray J (2015). Real-life challenges in using augmentative and alternative communication by persons with amyotrophic lateral sclerosis. *Commun Disord Q* 36:187-192.
- Schlosser RW (1997). Nomenclature of category levels in graphic symbols, part I: is a flower a flower a flower? *Augment Altern Commun* 13:4-13.
- Trudeau N, Sutton A, Dagenais E, et al (2007). Construction of graphic symbol utterances by children, teenagers, and adults: the effect of structure and task demands. *J Speech Lang Hear Res* 50:1314-1329.
- Van Es MA, Hardiman O, Chio A, et al (2017). Amyotrophic lateral sclerosis. *Lancet* 390:2084-2098.
- Ventura MA, Green MF, Shaner A, et al (1993). Training and quality assurance with the Brief Psychiatric Rating Scale: "the drift busters". *Int J Methods Psychiatr Res* 3:221-244.
- Yoshizawa K, Yasuda N, Fukuda M, et al (2014). Syntactic comprehension in patients with amyotrophic lateral sclerosis. *Behav Neurol* 2014:230578.