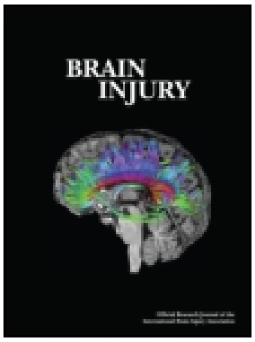

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Beyond the gaze: Communicating in chronic locked-in syndrome

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Abstract

Objective: Locked-in syndrome (LIS) usually follows a brainstem stroke and is characterized by paralysis of all voluntary muscles (except eyes' movements or blinking) and lack of speech with preserved consciousness. Several tools have been developed to promote communication with these patients. The aim of the study was to evaluate the current status regarding communication in a cohort of LIS patients.
Design: A survey was conducted in collaboration with the French Association of Locked-in syndrome (ALIS).
Subjects and methods: Two hundred and four patients, members of ALIS, were invited to fill in a questionnaire on communication issues and clinical evolution (recovery of verbal language and movements, presence of visual and/or auditory deficits).
Results: Eighty-eight responses were processed. All respondents (35% female, mean age = 52 ± 12 years, mean time in LIS = 10 ± 6 years) reported using a yes/no communication code using mainly eyes' movements and 62% used assisting technology; 49% could communicate through verbal language and 73% have recovered some functional movements within the years.
Conclusion: The results highlight the possibility to recover non-eye dependent communication, speech production and some functional movement in the majority of chronic LIS patients.

Keywords

Assistive technologies, brain–computer interface, rehabilitation

History

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Introduction

Locked-in syndrome (LIS) is a neurological condition of quadriplegia and anarthria associated with ventral pons infarction. The American Congress of Rehabilitation Medicine defined the LIS as: (i) the presence of sustained eye opening, (ii) the preservation of cognitive skills, (iii) a severe hoarseness or hypophonia, (iv) quadriplegia or quadriparesis and (v) a primary mode of communication using eye movements or blinking [1]. Based on the severity of motor deficits, three varieties of LIS have been described: the 'classical LIS', which corresponds to the original definition; the 'incomplete LIS', in which residual functional movements in addition to eye movements are present and the 'total LIS', where patients show total immobility, including eye movements [2].

However, other no-motor deficits associated with the pontine lesion have been described in patients with LIS: for example, the dissociation between mental manipulation of body parts (altered) and objects (preserved), possibly due to the involvement of the motor system in mental simulation of action [3]. It has also been described an impairment in recognizing negative emotions on the faces due to the alteration of voluntary facial mimicry in the LIS patients [4] and the presence of pathological crying and laughing due to direct alteration of the effectors pontine nuclei or to the ponto-cerebellar connections, leading to a dissociation between emotional responses and their subjacent context [5]. Finally, changes in the resting electroencephalogram have been found with decreased cortical sources of alpha rhythm and increased cortical sources of delta rhythm, which is possibly associated with the alteration of the mechanism of 'reciprocal inhibition' of these rhythms by the pontine lesion [6].

At the beginning, patients with LIS are confronted with an extreme difficulty to communicate with other means than eye movements or blinking [7, 8]. This makes the diagnosis of LIS particularly challenging [9, 10]. Advances in critical care medicine have promoted the survival rate of these patients and their eventual return to home. This has led to two challenges. First, there is a need to conduct a differential diagnosis, with high reliability and in a time span as short as possible. One study has shown that the average time elapsed between onset and diagnosis in a cohort of 44 LIS patients was ~2.5 months [11]. Differentiating the vegetative state (VS) from the minimally conscious state (MCS) is also a difficult task. A study has reported on the presence of signs of

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consciousness in 37–43% of patients with diagnosis of vegetative state [12]. A more recent study showed that 41% of patients considered in VS were re-classified in MCS after standardized neurobehavioural assessment [13]. There could also be some difficulty on differentiating the vegetative state from the LIS at the acute phase because the former can be a transitional state between the coma and the recovering of consciousness [14]. The case of a patient with total LIS whose diagnosis could only be done through the use of electrophysiological techniques (event-related potentials) that showed significant differences in the amplitude of the evaluated component has been reported [15]. Finally, there is the dramatic case of a patient who was thought to be in a vegetative state for a total of 7 years before the diagnosis of LIS was made by a speech therapist [16]. These examples illustrate both how difficult it can be to perform the diagnosis and the importance of having adequate tools to do so.

When consciousness has been detected, there is a second challenge to be faced: the need to provide LIS patients with appropriate means of communication, which will allow them to efficiently interact with their environment. A recent study has shown that quality-of-life (QoL) in patients with LIS is associated with the recovery of verbal communication and the possibility to carrying out recreational activities with a true integration to the community life [17]. This highlights the importance of developing adapted techniques to achieve these goals.

Since the publication of the book 'The Diving Bell and the Butterfly', written by Bauby [18] with the help of the flickering of his left eyelid and an alphabetic communication code, much progress has been made in the development of more advanced technology allowing patients with LIS to interact and communicate. Currently, a large variety of assistive technology (AT) for communication is available [19]. The simplest communication codes ('low tech' technologies) present the alphabet arranged on a board, with the patient signalling the target letter by eye movements/flickering or any other movement (e.g. head movement) or with a pointer. The vowels/consonants code, for example, in its simplest version, consists in vowels and consonants separated in two groups (vowels 'aeiouy' and consonants 'bcdfghjkl . . .') listed by the speaker (Table I). The patient validates using a pre-defined sign, first the group and next the letter from the group. A faster version is obtained by the subdivision of the consonants in three groups or by arranging the order of the letters according to the frequency of occurrence in the used language. For example, for the French language, the 'EJARIN' alphabet is used, with the speaker spelling the letters one by one according to the frequency of use in the spoken French language (the same code with a slight variation in the order of the letters, 'ESARIN', is used for writing). Table II shows examples of these different communication codes for English and French languages.

Among the 'high-tech' assistive technologies (AT), the speech-generating devices have a wide use and, in the case of the LIS patients, they consist of an alphanumeric, phonetic or pictographic keyboard that can be operated on either by pressing a key (with the finger, a stylus, an optical pointer or a joystick) or by scrolling successive boxes of the keyboard. The device can emit a voice which can either be pre-recorded by someone from the entourage (digitized voice) or obtained by a synthetic voice [19]. The different units vary in terms of autonomy, size, possibility of adaptation to a printer and other characteristics such as the cost, which can be highly relevant. The advantages of these devices are mainly linked to its easy transportation (low weight and some can be adjusted to a wheelchair or near the bed) and other functions that have been adapted in some models such as sending emails, text messages or switches to control the environment. Personal computer with adapted components (screen, mouse and keyboard) are another type of 'high-tech' AT which has significantly increased the possibilities of communication in these patients, not only with their immediate environment, but with the outside world through the use of the internet. More recently, brain–computer interface (BCI) has allowed the most severely motor-disabled patients to communicate through their brain activity (for a review see Naci et al. [20]).

This paper presents results of the annual ALIS' surveys, concerning the media used by patients with LIS to communicate. The aim of the study was to evaluate the current status regarding the communication in this patient population and to determine the extent to which these patients have access and use assisting technologies. This study also investigated factors that may facilitate communication (recovery of movement or any kind of language) as well as potential obstacles (as visual or hearing impairment). In light of these results, a brief account was given of the evolution of the AT for the LIS patients, the existing devices and the future perspectives in this field.

Materials and methods

In collaboration with the French association for LIS (ALIS), 204 LIS patients, members of ALIS, were invited by letter to fill in a structured questionnaire, aided by their proxies. The response to the questionnaire and return to the ALIS organizers were taken as consent to participate. The study was

Table I. Vowel and consonants eye-communication method.

	Groups of consonants		
Vowels	C1	C2	C3
A	B	J	R
E	C	K	S
I	D	L	T
O	F	M	V
U	G	N	W
Y	H	P	X
		Q	Z

Table II. Alphabetic code in the traditional order and then adapted to the frequency of appearance of letters in English and French.

Order	Letter of the alphabet
Traditional	A B C D E F G H I J K L M N O P Q R S T U V W X Y Z
English	E T A O I N S R H L D C U M F P G W Y B V K X J Q Z
French	E J A R I N S T U L O M D P C F B V H G Q Z Y X K W

approved by the ethic committee of the University of Liège and the scientific committee of ALIS. The questionnaire included socio-demographic (i.e. age, gender, educational level, place of living), clinical (i.e. aetiology and duration of LIS, motor recovery, presence of visual or auditory impairment) and communication questions (i.e. level of speech production, use of eye movements to communicate during the first weeks after the onset of LIS, current way to answer yes/no questions, current use of alphabetic communication codes and utilization of technical means to communicate).

Data were analysed with the program Statistica version 10 (www.statsoft.fr). Descriptive analyses were used: for the quantitative variables, this study reported mean $\pm$ SD and range. For qualitative variables, this study reported subject counts and percentages.

Results

Out of the 204 invited participants, 93 patients completed the questionnaire (response rate = 46%). Three subjects were excluded because they did not meet the diagnostic criteria for LIS at the beginning of the condition (i.e. exclusive communication through eye movements) and two were excluded due to missing data on the topic of communication. The final sample for the analysis included 88 LIS patients (Figure 1).

Demographic and clinical characteristics are shown in Table III. All were in a chronic LIS (>6 months after the insult, median = 9 years, range = 10 months–29 years), due in most cases to a brainstem vascular accident (70 out of 88). By the time of the survey, most of the responders (70%) lived at home. The majority (74%) had recovered some speech production: unintelligible sounds in 25% of cases, functional communication with use of words in 14% and with complete sentences in 35% of the patients; 73% of the group had recovered some degree of functional movements—mainly head movements (35%)—which was the second most used movement for yes/no communication. Finally, 66% presented visual impairment (mainly nystagmus) and 22% had auditory impairment.

Concerning basic communication (Table IV), at the moment of the survey, 50% used primarily eye movements to answer yes/no questions, either alone (25%) or in combination with head movements (16%), with sounds (6%) or with other gestures as fingers movements (3%). For open questions and more elaborated communication, 67% of the patients used a specific and pre-defined code for communication. The most frequently used was the vowels and consonants code (31%) followed by the ESARIN code (15%). Among the 29 patients (33%) who did not report the use of any specific code for communication, 25 (28%) could verbalize (two out of the 25 with single words and 23 with short sentences).

Fifty-five out of 88 patients (63%) used 'high-tech' AT for communication. Personal computers were the most frequently used (73%), accompanied by different adaptations (e.g. modified keyboard, mouse, contactor). This was followed by the use of speech synthesizers (24%). The overall of reported technical means is described in Table V.

Ninety per cent of the users of 'high-tech' AT (50 out of 55) reported to be satisfied with the used equipment. Only five patients expressed dissatisfaction: two considered the devices very slow, one very tiring, one said that the equipment did not give him sufficient autonomy to write complete text and one considered the technology not reliable.

Discussion

This study evaluated the current methods of communication and the clinical recovery in a cohort of patients with LIS. It was found that 92% were able to establish a functional communication with their environment beyond the initial simple yes/no eye movements. Some patients still used alphabetic codes and eyes or body movements in their daily living, even if they had advanced technical means of

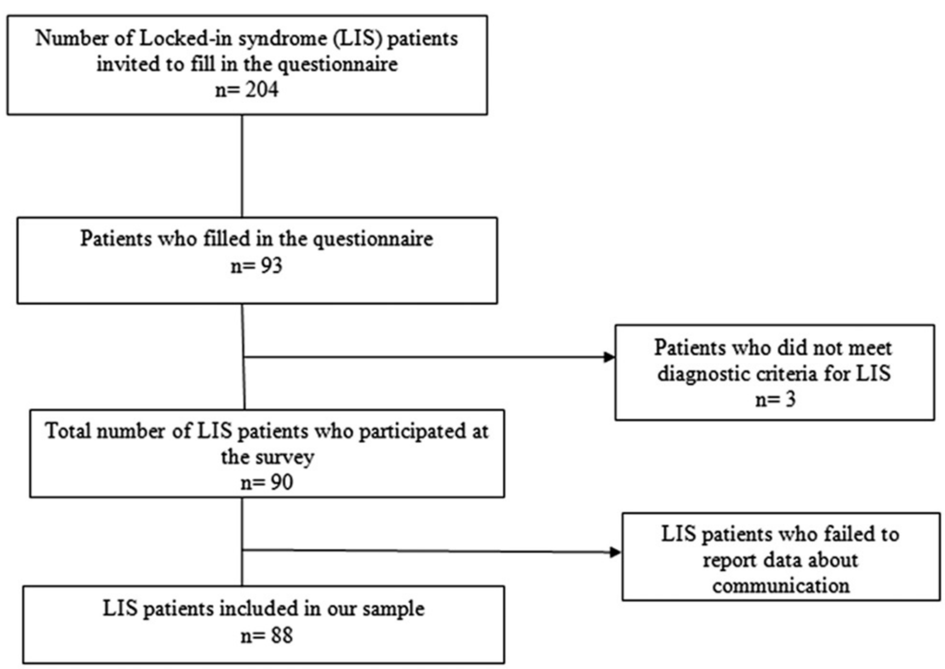

Figure 1. Participation in the survey.

Table III. Socio-demographic and clinical characteristics of the patients ($n = 88$).

Age mean in years ± SD (range)	52 ± 12 (16–74)
Gender (*n*, %)	
Women	31 (35%)
Men	57 (65%)
Mean age at the LIS onset in years ± SD (range)	42 ± 14 (14–73)
Mean time (years) in LIS (range)	10 ± 6 (10 months–29 years)
Aetiology (*n*, %)	
Ischaemic stroke	56 (63%)
Haemorrhagic stroke	14 (16%)
Traumatic brain injury	7 (8%)
Other	11 (13%)
Meningoencephalitis	1
Post-ablation occipital tumour	1
Neurodegenerative disease	1
No specified	8
Place of living	
Home	62 (70%)
Institution	26 (30%)
Educational level	
University or College	29 (33%)
High school or lower	59 (67%)
Recovery of movements	
None	14 (16%)
Non-functional	10 (11%)
Functional	64 (73%)
Recovery of speech production	
None	23 (26%)
Sound	22 (25%)
Words	12 (14%)
Sentences	31 (35%)
Visual impairment	
No	30 (34%)
Yes*	58 (66%)
Nystagmus	30 (34%)
Diplopia	25 (28%)
Decrease in visual acuity	24 (27%)
Decrease in visual field	24 (27%)
Others	6 (7%)
Auditory impairment**	
No	46 (72%)
Yes	14 (22%)
Missing data	4 (6%)

*Half of this group presented two or more visual troubles.
**Evaluated only in 64 subjects.

Table IV. Characteristics of non-technical communication in the studied sample.

	n (%)
Current yes/no communication	
Using eyes movements	
Alone	22 (25%)
With head movements	14 (16%)
With voice/sounds	5 (6%)
With other combinations	3 (3%)
Using head movements	
Alone	13 (15%)
With voice/sounds	2 (2%)
With other combination (except eyes)	2 (2%)
Using voice/sounds	23 (26%)
Using others movements	4 (5%)
Current system of communication	
None	29 (33%)
Vowels and consonants	27 (31%)
ESARIN alphabet	15 (17%)
Traditional alphabet	10 (11%)
Not specified	7 (8%)

Table V. Most frequent technical means used for communication ($n = 55$).

Personal computer	40/55 (73%)
With virtual keyboards	16/55 (29%)
With adapted keyboards	2/55 (4%)
With adapted contactors	
To the chin	7/55 (13%)
To the fingers	3/55 (5%)
To the head	1/55 (2%)
To the temple	1/55 (2%)
To glasses	1/55 (2%)
Not specified	8/55 (15%)
With adapted mouse	4/55 (7%)
Speech synthesizer	13/55 (4%)
Eye-gaze system	3/55 (5%)
Laser pointers	2/55 (4%)
Cell phone	2/55 (4%)

communication (64%). This study corroborates the study of Snoeys et al. [21] in which six out of eight studied patients using 'high-tech' AT still employed eye coded communication. Although there is a high percentage of patients in the cohort with visual impairment (66%), eye movements (alone or combined with other gestures) continue to be the most used form of non-technical communication means (50%). This could be related to the localization of the lesion (ventral pons), which allows in most patients preservation of vertical eye movements and blinking. It has been shown that it is possible to train a LIS patient to use eye blinks for augmentative and alternative communication [22]. In this group, it is possible that the continuous use through the years of this gesture has done it more effectively as a communication mean. Nevertheless, any other minimal interpretable gesture done in a consistent way by the patient can—and should be—used as a means to communicate with them (for example, one of the patients reported in the survey the use of his left foot for the yes/no communication).

Sixty-three per cent (55 out of 88) use 'high-tech' AT as principal means for communication. For the patients who have access to these technical methods, the personal computer was used by two thirds, making it the most used communication device. The use of these systems in these patients with severe disabilities—including visual impairments—is only possible thanks to the many adaptations of the different system components (screen, mouse, keyboard, etc.) allowing the adaptation to such disabilities [19]. Among the multiple options, these systems allow modifications of the characteristics (size, colour) of what is seen on the screen to permit better visualization. Similarly, the look of the mouse pointer can be changed on the screen to make it more visible and the selection system can also be changed to make it more accurate with less movements. Also, the keyboard can be displayed on the screen (virtual board) and texts can be auto-completed to allow writing words or short sentences by just typing the first few letters. There is also the important option of the speech synthesizers, which provides return hearing of what the patient types, this system being used by 24% of the users of high-tech technical means. The overall report of these means in the sample is described in Table V.

The high percentage of satisfaction (90%) expressed with this equipment (the high-tech AT) seems to confirm the

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Table VI. Clinical and communication characteristics reported in different studies of LIS patients.

Reference	*n*	Gender (M/F)	Mean age at the onset (years)	Aetiology	Movements recovery (%)	Sounds/ speech production (%)	Visual impairment (%)	Auditory impairment (%)	No-technical communication (%)	Use of technical communication (%)
Hawkes [32]	7	4/3	39	Vascular	71	nr	nr	nr	86	nr
Pattterson and Grabois [28]	139	85/52	52	Vascular	76[a]	nr	nr	Abnormal BAEP reported in 7/10 patients	nr	nr
Haig et al. [29]	27	18/9	32	TBI	59[b]	57[f]	nr	nr	86	36
Katz et al. [30]	29	19/10	33	TBI	67[c]	72[g]	nr	nr	44	39
Richard et al. [27]	11	9/2	45	Vascular	100[d]	36	45[j]	nr	100[l]	36
Leon-Carrion et al. [11]	44	23/21	47	Vascular	nr	78	14	nr	65.8	nr
Casanova et al. [26]	14	9/5	45	Vascular	79	28	100[k]	nr	100	42
Doble et al. [31]	29	19/10	34	Vascular	77	85[h]	nr	nr	38	31[m]
Bruno et al. [10]	5	2/3	15	Vascular	60	40[i]	nr	20	100	60
Snoeys et al. [21]	8	4/4	41	Vascular	100[e]	75	nr	nr	100	100
This paper	88	57/31	42	Vascular	85	75	67	22	100	63

Percentages are referred to percentage of patients. [a]percentage on 47 survivals; [b]in 38% not functional and none could execute movements against gravity; [c]reported only as 'limb movements'; [d]63% had only 'minimal recovery'; [e]in six out of eight some upper limbs movements and in five out of eight not functional lower limbs movements; [f]44% reported as 'involuntary cries'; [g]44% reported as 'involuntary cries'; [h]46% reported as 'involuntary cries'; [i]also reported pathological laughing and crying; [j]paresis of lateral gaze in three patients and nystagmus in two patients; [k]at the onset gaze alteration, improved in 64% of cases; [l]since the onset of the rehabilitation through eyes' movements; [m]data from the last year of follow-up on 13 survivals.

Abbreviations: nr, not reported; BAEP, brainstem auditory evoked potentials; TBI, traumatic brain injury.

appropriate adaptation of these technologies for motor-disabled people. However, besides the improvement of the already existing devices, brain–computer interface development is growing to enhance the available tools for communication and diagnostic purposes [23, 24] and even have been already used for recreational activities such as painting in severally disabled patients [25].

A recovery of functional movements was identified in 72% of the cohort, which is in line with other studies, also showing a high percentage of improvement in chronic LIS [21, 26, 27], as shown in Table VI. These findings highlight the importance of rehabilitation and physical therapy. Casanova et al. [26] reported 79% of motor recovery according to the classification of Patterson and Grabois [28] after an intensive and early rehabilitative programme. In light of the findings, a re-definition is suggested of functional movements to the possibility of using minimal movements for communication and/or environmental control systems, enabling autonomy in the presence of a very severe motor handicap. Most of these patients (63%) were able to use a minimal movement to operate sophisticated systems, allowing them to interact with their environment. This was appropriately described by one of the patients in the questionnaire:

> I have suffered a LIS resulting in a complete paralysis, with difficulty of swallowing and impossibility to speak. I can only make some small head movements that allow me to run a small laser beam on a special board attached to a frame of glasses.

Through small movements, this patient is able to handle his own web page and to interact with the external world over the internet. This also highlights the importance of recovery of any kind of movements, not just the limbs.

In this sample, 74% could produce sounds, which is in line with the study of Leon-Carrion et al. [11], who reported 78%. In this cohort there is a high percentage (49%) of sounds production, including understandable words or sentences enabling for functional verbal communication. This contrasts with the high percentage of 'involuntary cries' as only vocalization reported in others studies [29–31] and might be related to the lack of an effective speech therapy. The study of Casanova et al. [26] shows that, despite an intensive and early programme of speech therapy, only 29% of patients recovered verbal communication. It could be speculated that prolonged speech therapy sessions would lead to a better functional outcome.

It must be stressed that this sample may not be representative of most patients with LIS, as the response rate was only 43%. Thus, the results in terms of language, movement recovery and use of AT for communication may reflect selective bias (reflecting the group of patients in better conditions and with greater opportunities and capabilities to respond to the questionnaire). However, it is also likely that there is a real possibility of functional recovery enabling improved quality-of-life, as high rates of recovery of movement, speech and the use of technologies to communicate have been previously described in other studies [21, 26–28, 31, 32]. As suggested by Schjolberg and Sunnerhagen [33], rehabilitation of patients with LIS has an impact on their quality-of-life, as one of the major goals is to establish a reliable and efficient communication. Bruno et al. [17] showed that, in a cohort of LIS patients, non-recovery of speech production was associated with unhappiness and Pistoia et al. [34] have reported on the improvement of the mental well-being in a group of LIS patients after the pharmacological treatment of an eye-movement disorder (opsoclonus-myoclonus), enabling them to communicate through eye movements. These findings seem to confirm the importance of the recovery of communication for these patients.

In conclusion, despite the devastating neurological deficit in patients with LIS, combining the most modern technology and the slowly recovering mobility, some patients seem to be able to recover important autonomy by establishing extended communication beyond the yes/no responses to closed questions. More research is still needed to improve and expand these methods and to adapt them to the patients.

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Declaration of interest

The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the paper.

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