J Rehab Med 2001; 33: 235–239

AUGMENTATIVE AND ALTERNATIVE COMMUNICATION METHODS IN LOCKED-IN SYNDROME

Sinikka Söderholm, Monica Meinander and Hannu Alaranta

From the Käpylä Rehabilitation Centre, National Association of the Disabled in Finland, Helsinki, Finland

Locked-in syndrome is a neurological condition due to a brain disease or an injury affecting the brain stem. The symptoms are tetraplegia, double-sided facial paresis, anarthria/dysarthrophonia, dysphagia and reactive involuntary laughing and crying. Vertical eye movements are the only commonly remaining voluntary motor function. Although the linguistic abilities as well as intellectual and emotional functions as a whole remain intact, all the motor abilities of self expression are lost. Seventeen chronic locked-in syndrome patients referred to Käpylä Rehabilitation Centre between 1979–2000 are reported. The multidisciplinary rehabilitation team developed an individual alternative communication method for all patients and trained them to use it by minor movements of e.g. thumb, chin or head. An alternative communication method enabled most of the patients to interact with other people using practical as well as theoretical thinking and decision making.

Key words: brain stem, locked-in syndrome, tetraplegia, communication, computer, anarthria.

J Rehab Med 2001; 33: 235–239

Correspondence address: Sinikka Söderholm, Speech-Language Pathologist, Käpylä Rehabilitation Centre, National Association of the Disabled in Finland, Koskelantie 22, FIN-00610 Helsinki, Finland. E-mail: sinikka.soderholm@invalidiliitto.fi

(Accepted March 13, 2001)

It had been agreed that the old man should express his approbation by closing his eyes, his refusal by winking them several times, and if he had some desire or feeling to express, he raised them to heaven.

(Dumas, 1844–45)

INTRODUCTION

The first descriptions of locked-in syndrome (LIS) appeared in 19th century fiction. Alexandre Dumas introduced in his novel *The Count of Monte Cristo* (1) Monsignor Noirtier de Villefort who, as a result of cerebrovascular disease, was only able to communicate by blinking his eyes. His helper pointed at words in a dictionary and the old Monsignor indicated with his eyes the words he wanted. Émile Zola (original work 1868) wrote in his novel *Thérèse Raquin* (2) about a paralysed woman who had "language only in her eyes and others had to guess what she wanted". A contemporary French journalist Jean Dominique Bauby tells about his own experiencies of LIS. He "dictated" with his eyelid the book entitled *The Diving Bell & the Butterfly* (3). "Paralysed from head to toe, the patient, with his mind intact, is imprisoned inside his own body and unable to speak or move."

LIS is a condition caused by extensive damage in the brain stem. The patient's limbs, body muscles, face and the muscles controlling speech and swallowing are paralysed. Symptoms also include reactive involuntary crying and laughing, which are exaggerated in comparison with the emotional state. In most cases the patient can move his/her eyes, but optical disturbances, e.g. double vision, limited range of vision or difficulties in focusing are possible. Linguistic ability, memory, intellectual, cognitive and emotional functions remain intact. The patient is conscious and aware of his/her surroundings, but the abilities for self expression are lacking (4–7). The patient may be depressed which is natural, especially at the onset of the illness. There may also be changes in alertness and a tendency to tiredness.

The most common cause for LIS is a cerebrovascular disease in the vertebrobasilar area (8, 9). Other causes may be traumatic brain injuries affecting the brain stem or the cerebellum, tumours and the sequela of viral infections (4, 10). At the onset of the disease the mortality rate is high and the majority of deaths take place during the first few months (11). Total recovery is rare but slight improvement is possible. Generally the locked-in patient's condition stabilizes during the first year after onset. The crucial point is that the patient's life expectancy may be several years (4, 6, 12).

LIS has been known in medicine for decades, but the sophisticated means enabling the patients to express themselves have been uncommon. Advanced knowledge of the syndrome and computer-based communication methods have greatly improved some patients' quality of life (13). Communication devices and computer-based communication methods have only been in use a few years. However, the awareness of these sophisticated instruments is still rather low both among the population at large and health-care professionals.

Motor speech disorders

A typical LIS patient is unable to speak because of anarthria (4, 6). The muscles producing and controlling speech are either partially or totally paralysed. The patient also has difficulties in coordinating breathing and voice. To aid the breathing the patient has a tracheostomy at the onset of the disease. There are only few facial movements left, normally vertical eye movements, which however combine with synergic elevation of the

 ISSN 1650–1977

Table I. *Communication devices, access methods, aims, and frequency of use of 17 locked-in patients*

Patient/ sex	Onset (year)	Methods, technical devices, assistive software	Access methods, operated from	Aim/frequency of use
1/M	1996	Alphabet board, sounds, words, text communicator with voice output, PC-computer with keyboard software, word prediction, a music composing programme, fax, e-mail and Internet, printer, modem, synthetic speech	Infrared headmouse operated by head movements, single hand switch operated by right hand wrist extension, powered wheelchair	Communication, reading daily news, shopping via Internet, e-mail, composing music, writing short stories, telephone conversations, games/daily use
2/F	1994	PC-computer with scanning keyboard software, fax, e-mail, Internet, printer, modem	Single pneumatic switch operated by biting, wheelchair	Communication, reading daily news, e-mail, office work relating to her personal assistants and family/daily use
3/M	1992	Auditive alphabet feedback, PC-computer with external keyboard, later Macintosh computer with scanning keyboard + auditive feedback, synthetic speech, printer	Single switch operated by thumb, bed, wheelchair	Communication and writings in the beginning/no computer use for some years due to weak physical condition and depression
4/M	1998	PC-computer with auditive scanning keyboard software, fax, e-mail, Internet, printer, modem	Single switch activated by hand movement, wheelchair	Communication, practising writing in order to finish his university studies/daily use depending on physical condition
5/F	1994	Alphabet board, auditive alphabet feedback, Macintosh computer with scanning keyboard software + auditive feedback, synthetic speech	Single switch operated by thumb movement, bed, wheelchair	Communication and correspondence/daily use
6/M	1986	Text communicator with voice output, PC-computer, keyboard software, word prediction, fax, e-mail, Internet, printer, modem	Headpointer, joystick mouse operated by chin movement, single hand switch operated by right hand wrist extension, powered wheelchair	Communication, studies, Internet use in many ways/daily use
7/F	1995	Alphabet board, laptop computer with keyboard software, word prediction, fax, e-mail, Internet printer, modem	Infrared headmouse with automatic mouse click, wheelchair	Communication, daily use of fax and e-mail, language studies, shopping, games/daily use
8/M	1989	Dysarthric speech, PC-computer, small curved keyboard, fax, e-mail, Internet, printer, modem	Mouthstick, powered wheelchair	Typing work/daily use
9/F	1996	Alphabet board, Macintosh computer with auditive scanning keyboard software, printer	Single switch operated by finger flexion, wheelchair	Practising communication/a few times per week
10/F[1]	1979	Alphabet board, electronic scanning device, PC-computer with Morse software, printer	Single switch, bed, wheelchair	Communication and correspondence
11/M	1984	Text communicator, PC-computer, regular keyboard and mouse, fax, e-mail, Internet, printer, modem	Middle finger of left hand, powered wheelchair	E-mail and daily use of all kinds of Internet possibilities
12/F	1980	Auditive alphabet feedback, electronic scanning device (Zygo 100) connected to a typewriter, computer assessment recommendations made 2/2000	Single switch operated by left hand, bed, wheelchair	Daily use of Zygo 100, waiting for computer delivery
13/M	1992	Alphabet board, speech when using a cannula, PC-computer, keyboard software with word prediction, fax, e-mail, Internet, printer, modem	Infrared headmouse, single switch operated by shoulder movement, powered wheelchair	Communication, corresponding with Internet friends, helping children with homework/daily use
14/F[2]	1980	Alphabet board, text communicator	Pointer fastened to patient's left hand, bed, wheelchair	No information available
15/M	1985	Dysarthric speech, PC-computer, keyboard software, word prediction, e-mail, Internet printer, modem	Infrared headmouse, single switch activated by left thumb, powered wheelchair	No computer available in present living accommodation
16/M	1999	Alphabet board, auditive alphabet feedback, trial period with Macintosh computer with auditive scanning keyboard software	Single switch operated by head movements, wheelchair	Waiting for new rehabilitation period when computer assessment will continue
17/M	1985	Alphabet board, auditive alphabet feedback, laptop computer with scanning keyboard software, printer	Single switch operated by head movement	Practising to use computer communication (computer delivered 7/2000)

M = male, F = female.

[1] Patient died 14 years after the onset.

[2] The facts are based on the documents of Käpylä Rehabilitation Centre.

upper eyelids when looking upwards (4). The strength and range of the movement of lips and tongue are weakened as well as the flexibility of the soft palate. The patient often has pathological suck/bite reflexes. Due to the lack of coordination of breathing and voice a voluntary voice output is not possible although there usually is no paralysis of the vocal cords. The voice comes as a reflex with compulsive/involuntary crying or laughing.

Communication

As LIS patients' linguistic and intellectual capabilities are intact it is primarily important to provide them with compensatory means of communication as soon as possible to enable them to express their basic needs. Communication is based on vertical eye movements which usually remain intact (4–6). By eye movements the patient can give either positive or negative answers to questions. Communication using the alphabet is also based on eye movements. The alphabet is either read out or pointed from a board to the patient and he/she uses eye movements to choose the letters needed for formulating words and sentences. In the beginning the method based on read-out alphabet can be very slow and requires keen concentration, continuous patience and good memory of both the patient and the discussion partner. Communication is easier when both the vertical and horizontal eye movements are good. In those cases the aid can be a transparent plastic alphabet board which helps the patient to formulate messages by moving his/her eyes from one letter to the other. This method is a small step towards more normal interaction as the transparent board also allows the discussion partners to have eye contact with each other.

METHODS AND SUBJECTS

Method of computer-based communication

This article aims to discuss LIS patients' augmentative skills and alternative communication methods based on the clinical follow-up of 17 subjects.

The main problems in designing a computer-based communication method are due to the lack of voluntary muscle activity and the difficulties in visual focusing (4, 6). The use of a computer-based communication method also demands good memory and alertness as well as good linguistic ability. Effective utilization of computers is only possible when the communication method is planned by a multi-disciplinary rehabilitation team with thorough knowledge of the subject (14). Choosing a suitable computer-based method is a time-consuming process. It also requires cooperation of device engineers and the rehabilitation team.

First a speech and language pathologist examines the functions of the patient's face muscles and those needed for producing speech as well as the movements of the chin. In cooperation with physio- and occupational therapists the patient's sitting position will be adjusted to ensure the best possible control of head, limbs and body. In spite of the patient's reduced movements and spasticity of the limbs, the team aims at finding a voluntary reliable and easily controlled muscle movement, which can be used to operate the computer. Computers can be operated by the slightest movement of a finger or the wrist, by frowning the forehead, a movement of the chin or by biting. If the patient has controlled head movements the use of an ultrasound or infrared head mouse may also be possible (15). Choosing the right switches, finding their correct placement and implementing the training of the patient constitute the most important, time-consuming and demanding part of developing a personal computer-based communication method.

With assistive software a computer can be activated even by one single switch. The regular keyboard can be replaced by a keyboard programme which brings the picture of the keyboard onto the computer screen and enables the patient to use it with a mouse or a scanning keyboard programme and a single switch. Writing can be made easier by word prediction programmes. Some patients can support their communication by speech synthesizer programmes which convert written text into speech. If the patient has problems with eyesight or focusing, a keyboard programme combined with voice feedback and scanning may make the computer more user friendly.

Subjects

Seventeen LIS patients (7 women and 10 men) were examined and rehabilitated in Käpylä Rehabilitation Centre, Helsinki, Finland, with regular follow-ups between 1979 and 2000 (Table I). The diagnosis of LIS has been made by the neurologist. At the onset of the disease the youngest patient was 17 and the oldest 57 years of age. The mean age at the onset of the disease was for the men 42.0 years and for the women 35.1 years. Fifteen patients had cerebrovascular disease, one traumatic brain stem injury and one the sequela of a virus infection (Guillain-Barré).

During the follow-up study the Käpylä team met 15 patients personally either in the Rehabilitation Centre, in local service flats or in their homes. One patient was unavailable for a personal meeting. One patient died 14 years after the onset of the disease. One of the 15 follow-up patients is cared for in a local hospital, eight patients live in local service flats. Six patients live at home and are cared for by their family and personal assistants. The longest follow-up has lasted for 21 years, the shortest for 5 months. All 15 follow-up patients need help in many daily routines. Seven patients are able to use powered wheelchairs with the help of individually designed steering systems, six patients use daily environmental control systems as well.

RESULTS

The summary of communication devices, access methods, aims, and frequency of use of 17 locked-in patients are shown in Table I.

The average rehabilitation period took 3–4 months. During their rehabilitation all follow-up patients have learned to use an individually designed alternative communication method or a computer-based communication method or both. During the follow-up the development of information technology has made it possible to find new user-friendly alternatives to access the computer. Nine of the follow-up patients use their computer daily as a communication method and keep up contacts through e-mail and use of the Internet, e.g. for shopping, finding new contacts and for reading daily papers. Four of the patients have also dysarthric speech, only two of them use speech as their main communication method.

In spite of a high education and a good ability to use the computer, the patient who is cared for in a hospital has given up the use of his computer due to his weak physical condition and severe depression (Table I, patient 3). One of the patients is adynamic and has never used written self expression and therefore the experience of using the computer is too strange for her (Table I, patient 9). One of the patients had an opportunity to come in for a computer-based communication assessment 15 years after the onset of his illness (Table I, patient 17). One patient is still waiting for his computer delivery (Table I, patient 15).

Case Reports

"One of the most successful cases". Patient 1 is a man aged 33

years. In spite of his very severe handicap and the continuous need for personal assistance in all daily activities he manages with the help of his computer to build his life in a very satisfactory way. He had developed LIS in November 1996 as a result of an infarction of the vertebrobasilar area. The acute hospital treatment included a magnetic resonance imaging (MRI) which revealed an extensive infarction in the pons region. The patient was referred to Käpylä Rehabilitation Centre in April 1997. He was tetraplegic and unable to speak. Other symptoms included strong involuntary laughing and crying. On arrival his tracheostomy was removed but he was still tube-fed. Eye movements were good, enabling him to give yes–no answers to questions. Other facial movements were very limited. As his first everyday communication method the patient used fluently a transparent plastic alphabet board. The reading out of the alphabet was used as an additional communication method during therapy sessions. The patient received intensive rehabilitation for 2 months. The nasogastric tube was removed in May 1997 and the function of his speech organs reactivated enabling him to express himself with some basic phrases. For faster communication he used the transparent alphabet board and his eyes. A multidisciplinary team designed for the patient a computer-based communication method with supplementary software. The patient uses an infrared mouse operated by head movements. He also manages to use a single switch with minor movements of his right wrist. Towards the end of his rehabilitation period the patient started practising moving around with a powered wheelchair, which he steered with his chin. His next rehabilitation period was in the autumn of 1997. By then his swallowing and communication were much improved. At this time the patient practised the use of an environmental control system. In addition to previously acquired communication skills he also learned to use a text/speech communicator with the help of his mouthstick. This also enabled him to use a loudspeaker telephone. After the rehabilitation the patient moved to a service flat. At the end of 1998 he exchanged his laptop computer for a table workstation. He has enlarged the use of his computer to composing music. He uses e-mail regularly, makes telephone calls, shops via the Internet and takes part in chat groups on the Internet. The patient comes in for regular follow-ups once a year, in between he has regular weekly physiotherapy and speech therapy. He has described his own life situation in the following words:

> *Locked-in syndrome is a monster combination of words, which literally captures a person "locked in" inside his own body. I cannot eat or drink nor speak properly, I cannot anymore do things which before came automatically. This illness has robbed me of my ability to move and to use my hands. I do not know how to cough, spit or gargle. I cannot control my emotions, my crying and laughing, however much I would like to. I often fail to make myself understood, because I can only communicate by moving my eyes on the alphabet board. Luckily I now have this computer, which has enabled me to write this text. My other connections with the outside world are practically non existent. My illness coincided with my wife's first pregnancy. Now we have a sweet little daughter. Perhaps my life has a meaning after all—as a father—a privilege nobody can take away from me.*

"A more complicated case". Patient 2, a 40-year-old woman and a mother of 4 children, has been chosen to show how a person with these severe communication problems and physical handicap can manage to take part in the everyday life of her family. She had developed LIS in November 1994. She had suffered from headaches of migraine type for years. Otherwise she had been very healthy. She had a sudden severe headache at work which soon caused the loss of consciousness. LIS was caused by an extensive left pons infarction and a dissection of the basilar artery which was confirmed by MRI. After a few days she developed pneumonia and was tracheostomized.

The patient came to Käpylä Rehabilitation Centre in September 1995. On arrival she was tetraplegic. Her face movements were reduced, she also had lateral gaze palsy and hemianopia and total anarthria. For her everyday communication she used eye movements to respond with yes–no answers to questions. The patient received intensive rehabilitation for 3 months. During the rehabilitation she developed her communication skills. Many trials and errors were made before a suitable switch was found. In the beginning, due to her vision problems, the only possibility was to use a computer with the help of auditory scanning software and a single switch controlled by head movements. The switch was placed in front of her chin, by moving her head upwards she triggered it to choose the letters she needed to build words and sentences. She returned home to live with her family and has the help of two personal assistants. Her husband has built her a new single switch into a clothes peg. The switch was placed in her mouth and was triggered by biting. Later her vision improved so much that she could use the scanning software without the auditory feedback. Her next rehabilitation period was in May 1996, when an environmental control system was designed for her. Her computer software was developed, the individually designed keyboard was expanded so she could control several new functions on her computer, e.g. working out the work schedules for her assistants. In November 1998 the Macintosh computer was exchanged for a PC. This possibility is now available as the applications used in the Macintosh are now compatible for PC computers. Now she is using e-mail and the Internet and her fax to keep in contact with friends and relations. She has again changed her method of accessing her computer and is now using a pneumatic switch which has been built into a baby's dummy. She still uses biting to trigger it. Over a period of 5 years she has changed her access method three times. She is still hoping to be able to use her right hand for controlling her computer because she has regained some voluntary movement of one of her fingers.

DISCUSSION

Locked-in patients are in a way "Cinderellas" of the medical world, quite a rare and partially forgotten group. The symptoms of LIS are often misunderstood. In international medical literature there are many case histories recorded. The groups are often small, consisting only of 1 or 2 patients (4, 12, 16, 17) or 6–7 patients (18–22). Patterson & Grabois (11) published a literary review of 139 patients, of whom only six belonged to their own study group. There is only one report, taking in a larger group of 29 patients (6). The Finnish follow-up study of 17 patients is large even by international standards. The catchment area of the centre is the whole country with 5.2 million people. However, the exact prevalence of LIS is not known in Finland.

The most common cause for LIS is a vascular disease. The reason for the injury in young people might be a dissection e.g. after ice hockey tackle or a whiplash injury. The prognosis is dependent on the extent and the location of the injury as well as personal characteristics of the patient. Learning to use the computer is not only dependent on physical capacity but requires also motivation and acceptance of the situation (23).

The rehabilitation of locked-in patients is a long process and requires close cooperation of a specialized group of experts. It is a particularly demanding process for the patient and his/her next of kin, requiring persistence, patience and optimistic belief in a better future. When a person has lost practically all capabilities for voluntary activity, it is of vital importance to do everything to improve his/her life, however small the steps may be. Interaction with one's environment is one of the basic needs and most important for improving the patient's quality of life.

Utilization of information technology in health care and rehabilitation requires from the rehabilitation team enthusiasm and constant learning and updating. There are many possibilities to choose from, but the expert training and know-how required for planning individual methods and progammes need continuous development.

A computer can never fully compensate for natural speech and non-verbal emotional communication. Computer-based communication, however, offers the patient the possibility to interact with his/her environment, not only in practical matters but also in deeper theoretical discussion and personal decision making. Through his/her texts a patient can express himself/herself as a unique, feeling individual.

ACKNOWLEDGEMENT

The study has been financially supported by the Finnish Slot Machine Association.

REFERENCES

1. Dumas A. The Count of Monte Cristo. London: Wordworth Editions Limited; 1997.
2. Zola É. Thérèse Raquin (in Swedish). Stockholm: Prisma; 1983.
3. Bauby J-D. The diving bell & the butterfly. London: Fourth Estate; 1998.
4. Dollfus P, Milos PL, Chapuis A, Real P, Orenstein M, Soutter JW. The locked-in syndrome: a review and presentation of two chronic cases. Paraplegia 1990; 28: 5–16.
5. Isaka Y, Iiji O, Ashida K, Imaizumi M, Itoi Y. Cerebral blood flow and magnetic resonance imaging in locked-in syndrome. J Nucl Med 1993; 34: 291–293.
6. Katz RT, Haig AJ, Clark BB, DiPaola RJ. Long-term survival, prognosis, and life-care planning for 29 patients with chronic locked-in syndrome. Arch Phys Med Rehabil 1992; 73: 403–408.
7. Ohry A. The locked-in syndrome and related states. Paraplegia 1990; 28: 73–75.
8. Blunt SB, Boulton J, Wise R, Kennard C, Lewis PD. Locked-in syndrome in fulminant demyelating disease. J Neurol Neurosurg Psychiatr 1994; 57(4): 404–405.
9. Chia L-G. Locked-in syndrome with bilateral ventral midbrain infarcts. Neurology 1991; 41: 445–446.
10. Rowland LP. Clinical syndromes of the spinal cord and brain stem. In: Kandel E, Schwartz J, Jessell T, eds. Principles of neural science. Connecticut: East Norwalk; 3rd edition 1991. p. 729.
11. Patterson JR, Grabois M. Locked-in syndrome: a review of 139 cases. Stroke 1986; 17: 758–764.
12. Thadani VM, Rimm DL, Urquahart L, Fisher L, Williamson PD, Enriquez R, Kim JH, Levy LL. Locked-in syndrome for 27 years following a viral illness: clinical and pathological findings. Neurology 1991; 41: 498–500.
13. Rechlin TA. Communication system in cases of "locked-in"-syndrome. Int J Rehabil Res 1993; 16: 340–342.
14. Söderholm S, Meinander M, Kahela K, Alaranta H. Suun motoriikan häiriöt ja korvaavat kommunikaatiomenetelmät "locked-in"—oireyhtymässä (in Finnish). Duodecim 1998; 114: 879–886.
15. Anson D. Using the HeadMaster with Macintosh, Apple II, and MS-DOS computers. Am J Occup Ther 1991; 45: 889–897.
16. Ostrum AE. The locked-in syndrome—comments from a survivor. Brain Injury 1994; 8: 95–98.
17. Sepcic J, Sepic-Grahovac D, Strenja-Grubesic J, Antonelli L, Andrasevic D. Locked-in syndrome. Neurologia Croatica 1992; 41:157–161.
18. Anderson C, Dillon C, Burns R. Life-sustaining treatment and locked-in syndrome. Lancet 1993; 342: 867–868.
19. Hawkes CH. Locked-in syndrome: report of seven cases. Br J Med 1974; 4: 379–382.
20. Kovanen J, Sulkava R. Locked-in-oireyhtymä eli valetajuttomuus (in Finnish). Duodecim 1982; 98: 1079–1083.
21. Nordgren RE, Markesberry WR, Fakuda K, Reeves AG. Seven cases of cerebromedullospinal disconnection: the locked-in syndrome. Neurology 1971; 21: 1140–1148.
22. Reznik M. Neuropathology in seven cases of locked-in syndrome. J Neurol Sci 1983; 60: 67–78.
23. Hummelsheim H, Eickhof C. Repetitive sensorimotor training for arm and hand in a patient with locked-in syndrome. Scand J Rehabil Med 1999; 31: 250–256.