J Head Trauma Rehabil
Vol. 18, No. 5, pp. 435–444

Impairment, Activity, Participation, Life Satisfaction, and Survival in Persons With Locked-In Syndrome for Over a Decade

Follow-Up on a Previously Reported Cohort

Jennifer E. Doble, MD; Andrew J. Haig, MD; Christopher Anderson, DO; Richard Katz, MD

Objective: To determine the long-term outcome of patients with stable locked-in syndrome. **Setting:** The community. **Design:** Retrospective phone survey. This study was further follow-up on a previously reported cohort. **Participants:** Twenty-nine persons with locked-in syndrome were included in an initial cohort 11 years prior to the current study. Records or contact with family showed that 16 were deceased. Telephone interviews were made with 1 living patient and the caregivers of 11 others. Public records documented survival of 1 nonrespondent. **Outcome measures:** Survival, codified responses regarding functional activities, social activities, and satisfaction with life. **Results:** Five-, 10-, and 20-year survival were 83%, 83%, and 40%, respectively. Ten subjects had not been hospitalized in the previous year. Eight lived with family. Little change in impairment occurred, but care was simplified. Improvements in communication related to technology, including computer and Internet access. Eleven left home at least monthly. Caregivers reported seven expressed satisfaction with life; five were occasionally depressed. No deaths could be attributed to euthanasia and no survivor had a "no code" status. One patient wished to die, seven had never considered euthanasia, six had considered and rejected it. **Conclusions:** Persons with initially stable locked-in syndrome can have prolonged survival, can live in the community if there is enough support, and have some measure of quality of life. **Key words:** *brainstem, locked-in syndrome, quality of life, tetraplegia.*

PERSONS with profound neurologic disability, including persistent vegetative state, locked-in syndrome (LIS), and high tetraplegia have been the subject of substantial medical and ethical debate. There are concerns about quality of life.[1] Others debate the right to decide the patient's fate regarding end of life, health care, and social and financial decisions.[2,3] The allocation of resources to maintain their lives is a high-stakes game.[4] The debates have proceeded with little actual information on survival or quality of life, and with little input from the persons who have these disabilities.

Jean-Dominique Baube, the French editor and author, has put a human face on the

From the Associates in Physical Medicine and Rehabilitation, PC, Ann Arbor, MI (Dr Doble); The University of Michigan Departments of Physical Medicine and Rehabilitation and Surgery, Ann Arbor, MI (Dr Haig); Rehabilitation Institute, Kansas City, MO (Dr Anderson); St Louis, MO (Dr Katz).

We acknowledge Sally Thomas, Checkpoint Investigators, Inc, Phoenix, AZ, for assistance in finding subjects.

Corresponding author: Andrew J. Haig, MD, The Spine Program, The University of Michigan, 325 E Eisenhower, Ann Arbor, MI 48108 (e-mail: andyhaig@umich.edu.)

disabilities involved. His book, *The Diving Bell and the Butterfly*,[5] was written through eye-blink Morse code signals after he suffered a brainstem stroke. In the book he uses great artistry to describe not only his physical condition, but also the fantasies, humor, and despair of a real person going through this process. Baube died on the day that his book was released to the public, and the proceeds have gone to form the Association du Locked-in Syndrome (ALIS), whose website, www.club-internet.fr/alis/main.html, and French language newsletter are core sources of information for people with LIS and their families. The continued brilliant productivity of Steven Hawking, the physicist who has been unable to move due to a neuromuscular illness, has sensitized the public to the possibility that people with severe paralysis can be productive members of society.

Especially for persons with LIS, there have been quantum leaps in technologies that have potential to redefine quality of life. Augmentative communication devices have increased in sophistication and decreased in cost. Coupled with a computer and a connection, these devices can allow a person who is unable to move or talk to communicate with individuals around the world. With new switching mechanisms, computers can now efficiently transfer a miniscule biosignal into large and complex movements for environmental control and mobility. As a result of these changes, one can no longer make assumptions regarding the association between the impairment and the quality of life of persons with LIS. Albrect's contention that disability and quality of life are not directly related, in part because of use of technology, makes sense for this population.[6]

Locked-in-syndrome, generally caused by an insult to the ventral pons, is perhaps the most disabling condition in which a person retains consciousness. In 1966, Plum and Posner[7] first introduced the term "locked-in syndrome" into the medical literature, referring to the constellation of quadriplegia, lower cranial nerve paralysis, and mutism, with preservation of consciousness, vertical gaze, and upper eyelid movement. Haig et al[8] redefined LIS as quadriplegia and anarthria, with preservation of consciousness. Anarthria is the neurologic inability to speak, as opposed to mutism, which can imply the unwillingness to speak.

More than 20 different etiologies of LIS have been described, with ventral pontine strokes the most common.[9,10] Trauma, hypotensive events, tumors, infection, and lesions of other areas of the brainstem are among other causes. Morbidity and mortality (60%) are thought to be highest within the first 6 months.[11] The early literature, biased in favor of cases with autopsy evidence, and with almost no reporting of long-term follow-up on survivors, led many to believe that long-term survival is rare, or that survivors included only persons who recovered to some extent.[9]

In May of 1985, this study was initiated with a review of medical records of all persons treated at The Rehabilitation Institute of Chicago, a major free-standing rehabilitation hospital, during the previous 3-year period with a diagnosis of brainstem stroke or LIS. The inclusion criteria were consciousness, anarthria, quadriplegia, and persistence of the locked-in state for more than 1 year after onset. A cohort of 29 LIS patients who met the inclusion criteria was identified.

Clear evidence of consciousness was defined by mental status examination. Cobb[13] defined consciousness as the awareness of oneself and one's environment. Patients were required to respond consistently to questions about themselves and their environment on the admitting mental status examination. Patients were considered anarthric if they demonstrated an inability to produce voluntary speech sounds. They were not excluded if they occasionally demonstrated involuntary cries, which can occur in this patient population.[14]

Quadriplegia was defined as the inability to move any limb against gravity. Patients with minimal hand movements (eg, for activation of a small switch; not for object manipulation) were included as they had little functional advantage over patients who used facial muscles to trigger a switch. A 1-year time frame was selected to eliminate persons who died from their acute event or other premorbid illnesses and to eliminate persons who had early spontaneous recovery from LIS. It has been shown, for example, that the probability of dying after spinal cord injury is greater during the first year post injury.[15]

The initial study showed that 24 were still alive, with 1 subject alive 12.5 years after onset.[8] Significant recovery was noted in only a few patients. Progress was noted, however, in rehabilitation. Simplification of care with removal of tubes and changing of bowel, bladder, and eating regimens was shown years after onset. Seventeen of the cohort lived at home and 8 had never been hospitalized after the initial event.

A 5-year follow-up study used a life-table analysis to show a 5-year survival from onset of LIS of 81%.[12] Caregiver burden was noted to be substantial, but families typically continued to successfully care for their loved ones. During those interviews we were struck by the individual stories. The current 11-year follow-up was designed to probe into quality of life as well as the medical and functional issues. Here we present very long term survival, functional status, and quality-of-life data on this population.

METHODS

Permission for the follow-up study was granted by the institutional review boards of both the University of Michigan and the Rehabilitation Institute of Chicago (Northwestern University). Subjects were traced through previous research records and medical records. In some cases, contact with the subject or the family was made only after substantial searching of public documents by a private investigator. Twenty-nine subjects, 19 men and 10 women, fulfilled the study criteria. The mean age of subjects at onset of LIS was 33.6 years (median 33.0, range 1–70 years). The most common etiology of LIS was cerebrovascular disease (48%) followed by trauma (34%) and hypotension (10%), with individual cases of viral encephalitis and vaccine reaction. It is important to note that these diagnoses were made in the early 1980s with the diagnostic imaging tools available at that time.

The caregiver (person identified as primarily responsible for daily care) was contacted by telephone. A structured interview asked for information on medical complications after discharge, current functional status, and data related to patient care. Specifically, questions assessed (1) date of onset and etiology, (2) frequency and cause of hospitalizations within the previous year, (3) most difficult problems for the subject and caregiver, (4) present placement (home, nursing home, hospital), (5) ability to verbally communicate, (6) limb movement, (7) method of feeding, (8) bowel and bladder management, (9) use of a communication device, (10) sleep patterns, (11) mobility status, (12) vocational and avocational activities, (13) evidence of depression, (14) amount and type of ancillary care, (15) amount and type of therapy (eg, physical, occupational, or speech-language therapy), (16) ability to leave the patient alone during the day or night, and (17) presence of tubes (indwelling catheter or gastrostomy or tracheostomy). In addition, we collected qualitative data including mood, ability to enjoy various activities, family involvement, community activities, perception of independence, and considerations of euthanasia and resuscitation wishes. With the exception of the one patient who was able to respond independently, these findings were the opinion of caregivers, not the patients. The updated rehabilitation hospital charts and previous research records of the 29 subjects were reviewed for demographic, medical, and functional data. Standard statistical parameters were calculated for the entire cohort, and a life-table analysis was performed on survival data. An approximate formula for the standard error of the survival curve was used, as proposed by Greenwood.[16]

Table 1. The number of subjects alive and dead at each study period. Note that subjects may have entered the study after a varying number of years of survival

Time frame	Alive, n (%)	Deceased	Not available/lost
Study initiation	24 (88)	3	2
5 years	20 (69)	6	3
11 years	13 (45)	15	1

RESULTS

Survival ranged from 2.02 years to more than 25 years. Table 1 shows the survival and follow-up data at each survey interval. Twenty-seven patients in the original cohort had follow-up information (whether dead or alive) at 5-year follow-up. Two who qualified could not be found at that time, but they were located through subsequent investigation. At 11-year follow-up, three subjects could not be located initially, but with extensive searching, all but one subject was found. Public information on that missing subject indicated that the subject still had a home address and was registered with Social Security. Fourteen patients were still living at the last follow-up. Phone contact was made directly with 1 subject (via computer) and with 12 caregivers.

Figure 1 shows the survival rates for the patients, demonstrating a 5- and 10-year survival rate of 83%, and a 20-year survival rate of 40%. Survivors trended to be younger at onset than those who died (survivor mean 28.2 ± 13.8 years, deceased subjects 38.1 ± 17.1 years, $t = .103$, $P = .08$). Women comprised 23% of survivors and 47% of deceased subjects, not statistically different (chi-square .095, $P = .75$).

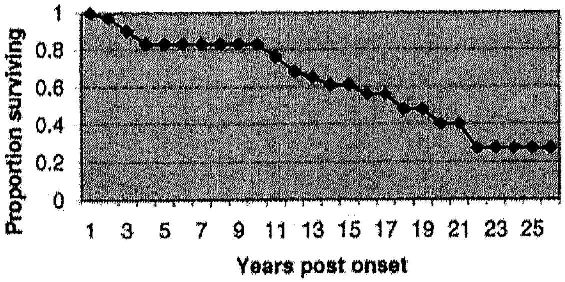

Fig 1. Survival curve for chronic locked-in syndrome.

Causes of death for the 15 deceased subjects included heart disease in 4; multisystem failure in 2; and 1 each from pneumonia, subsequent cerebrovascular infarct, respiratory failure, internal bleeding, and aspiration/asphyxiation. The cause of death was not found for four subjects. Although all families of the deceased indicated that there was consideration of euthanasia at some point, there was no indication that any of the deaths was the result of euthanasia.

Ten subjects had not been hospitalized within the previous year. The other three subjects were admitted to a hospital on seven occasions. Discharge diagnoses included renal stones (1), deep venous thrombosis (1), decubitus ulcer (2), pneumonia (1), urinary tract infection (1), and cervical cancer (1).

Physical impairments remained substantial, as Table 2 demonstrates. Only a few subjects evolved to the point where they could manipulate an object. None of the survivors could speak in sentences, and most could not speak a single word consistently.

Substantial functional gains were made despite the lack of neurologic change. Six survivors were involved in formal therapy at follow-up, ranging from 5 hours of physical therapy, occupational therapy, and speech-language pathology treatment per week to 1 hour of speech therapy per week. Five were receiving ongoing speech therapy, four had physical therapy, and three had occupational therapy.

Table 2. Physical impairment in LIS survivors

	Onset	Study Initiation	5 years	11 years
Available patients	27	27	18	13
Object manipulation				
None	16	11	3	3
Nonfunctional	10	6	0	2
Trigger switch	1	6	8	4
Point or type	0	4	1	2
Lift object	0	0	3	2
Oral communication				
No sounds	21	10	5	2
Involuntary cries	4	7	8	6
Rare words	2	6	4	2
Single words	0	1	1	3
Sentences	0	1	0	0

Medical device use had declined. Table 3 indicates that there was a decreased use of tracheostomies and feeding tubes over time, and that most survivors eventually were able to enjoy some oral food. Most no longer had indwelling urinary catheters. With autonomic functions intact, most were able to signal for a urinal. The functional category "voluntary with assist" for bowel and bladder management reflects patients who signal to void or defecate. Condom catheters or diapers were still used in many cases, but for caregiver convenience more than because of intrinsic bladder failure. Similarly, despite the ability to feel and somewhat control bowel movements, a number of patients used a bowel

Table 3. Activity in LIS survivors

	Onset	Study Initiation	5 years	11 years
Available patients	27	27	18	13
Breathing				
Tracheostomy	17	7	3	2
Nutrition				
Nothing by mouth	18	7	4	1
Occasional treats	4	6	4	3
Completely oral	5	14	10	9
Gastrostomy in place	24	19	10	6
Urination				
Foley catheter	9	5	5	2
Condom cath/diaper	15	13	6	6
Voluntary w/assist	3	6	7	5
Independent	0	0	0	0
Bowel movement				
Incontinent	0	0	0	0
Bowel program	27	23	14	10
Voluntary w/assist	0	1	4	3
Independent	0	0	0	0

program, typically consisting of a suppository every other day, in order to maintain continence.

Most survivors (8) lived with families, 3 were in extended care facilities, and 1 each in a state-run school and a hospital with private nursing. Eleven of the 13 survivors leave home more than once a month. Caregiver burden was substantial. Four caregivers felt they could leave their family member alone briefly in the house, but none would leave more than 2 hours. Eight did not feel that they could leave their relative alone in a room at night. Two survivors were legally competent. The legal guardian was a mother for seven, spouse for four, and the state for one.

Most patients continued to remain active through eye and facial movements. Four used computers consistently and two accessed the Internet. Three each preferred letter boards and facial movements for communication. Two used limb movements to signal with a letter board. Only 1 used vocalizations in communication.

Avocational and vocational activities listed by these persons include the following: TV, radio, music, books on tape, visiting with family, visit vacation home, e-mail, telephone, teaching, movies, shows, the beach, bars, school, and vocational training. One subject, an attorney, uses Morse code eye blinks with a caregiver to interpret them so that he could provide legal opinions and keep up with colleagues through fax and e-mail. Another helps teach math and spelling to third graders using a mouth stick to trigger an electronic voice device. A third reported greatly enjoying spending summer months in the north and winter months on the Gulf coast. A fourth sent a newspaper clipping describing him as the strongest fan of his local college team. It included a picture of this individual surrounded by team flags, shirts, and posters.

One personal newsletter is most illustrative: "___is busy every day. In addition to all [therapy] mentioned, he has to make time for letter writing, relating, drinking (1.5 l/day), eating, sleeping, and keeping up with all the latest news and politics. ___is not just a 'body' being moved from here to there. He has moods, ideas, communications. [the caregiver] I'm here as a sort of 'fifth wheel.' Every year I am needed less and probably am obsolete even now. But that's OK. It's really what we've been working toward for 20 years."

Subjectively, telephone interviewers were impressed with the real and substantial social interactions. Family members or caregivers were eager to discuss the lives of the patients beyond the structured questions. From the context as well as the content of the conversations (eg, offering to mail pictures, stories, etc.) it was apparent to the interviewers that the patients were actively involved in family and personal decisions, and that their presence was valued at home.

According to the reporting sources, seven survivors were satisfied with life in general, five were occasionally depressed, and the caregiver of one subject felt she could not determine the mood. Seven had never considered or discussed euthanasia, six had considered it in the past, but not currently, and one subject wished to die. None of the survivors had a "do not resuscitate" order.

DISCUSSION

Published in 1820, Alexander Dumas's novel, *The Count of Monte Cristo*, included the first known description of locked-in syndrome.[17] His character, Monsieur Noirtier de Villefort, had an apoplectic event, and was depicted as a "corpse with living eyes." The daughter established a communication system by interpreting his eye blinks as "yes or no." One hundred and fifty years later the medical literature first hinted that Dumas's character might be more than simply a metaphorical comparison to his Count "locked in" a dungeon.[18] Only a few years ago did the modern medical literature acknowledge that long-term survival was possible.[9]

The current study examined the largest and longest surviving population of persons with LIS. Given the small amount of information on severe disability, the current study may have implications for information on persons with high tetraplegia, persistent vegetative state, and other coma-like states.[9,24,25]

Findings need to be interpreted in light of a number of issues in this study. First of all, the information in most cases is based on the report of a caregiver. While survival figures are certainly accurate as reported, caregivers might over- or underreport functional abilities in comparison to a clinical examination. Caregivers' report of quality of life or the patients' feelings about death may be substantially different from the actual feelings of the patient. It would have been impractical to have a clinician visit each patient, as they are scattered across the country.

It is possible that persons admitted to the Rehabilitation Institute of Chicago in the 1980s were different from the population of persons with LIS at large. They may have been younger, or have had more resources and social support than persons who were sent to smaller community rehabilitation facilities or nursing homes. Only 14 of 29 were from the Chicago area, for instance. In order to reflect the population for whom life care plans are made, we selected only persons who had survived for more than a year and who remained locked-in after the year. Persons who are locked-in for a shorter period of time are probably more likely to either recover or die of comorbidities.

The primary finding is that people with LIS survive. The 10-year survival rate of 83% indicates that early medical interventions should not be temporizing in nature. Since the current study includes persons detected up to 14 years after onset of symptoms, the actual survival rate is probably less than 83%. Certainly some patients die, but others appear to be medically stable—most of the current survivors have not been hospitalized within the year. Survival data are important for a number of purposes. Medical and family decisions regarding resuscitation status, discharge location, and personal finances require some estimate of survival. Accurate actuarial estimates can save insurance companies and litigants substantial money while protecting the covered individual.[19]

Perhaps more surprising to those who do not deal with these patients on a daily basis is their social participation. With the help of family and friends, these people lead meaningful lives. It is unfortunate that the subject population was too geographically spread out for the investigators to personally and privately ask important ethical questions. Certainly there may be discrepancies between the wishes of persons with high tetraplegia and their caregivers. Hall et al used more direct input—telephone surveys, personal interviews and only occasional mail surveys—with long term survivors of C1-C4 spinal cord injuries, and found that 95% were "glad to be alive"[24] As best as the current methodology allows, the data demonstrate that almost all of the patients chose to continue living despite severe limitations.

While the right of individuals to withdraw from treatment is not questioned, Hall's work and the current study call into question the assumption among some health care providers and policy makers that severe disability is intolerable. This prejudice is not inconsequential. Biased clinicians provide less aggressive medical treatment and/or influence family and friends (in ways not appropriate to the situation).

The medical rehabilitation of persons with LIS is complex and atypical. With medical stability comes the opportunity for more freedom in basic care. Partial independence in bowel and bladder management can be tried as soon as communication is established. Oral feeding can be successful for many—even if just for the entertainment value of tasting favorite foods. Whether this progress is related to slow neurologic improvement or increased experience and risk taking of the caregivers is speculation, but it is clear that care can be simplified over time; tracheostomy and gastrostomy tubes may be removed in many cases.

With simplification of care, most of the patients can be discharged to live with family. The effort and support necessary to keep them independent in the community is substantial and is a major focus of the 5-year follow-up report on this population.[12] Compared with institutionalization, home care with family is much less costly, but it requires creative solutions not often covered by traditional insurance. Architectural changes and special equipment may not be covered by the insurance, but when they are paid, the person's ability to stay at home is greatly enhanced. Caregiver burden is substantial and stressful. A wise home discharge plan typically includes respite care, oversight by a professional, and emotional counseling for family members to continue their work in the long term. The cost savings of deinstitutionalization and the social benefit for the patient are substantial, but these are not possible without physical, financial, and emotional support for the families and flexibility on the part of payers.

Oral communication improvements are best attributed to neurologic change. Most patients are still essentially nonverbal. During tasks where computer connections are impractical, such as bathing and dressing, patients use simple eye gaze or facial communication systems, ranging from "eyes up means yes" to Wu's A-E-I-O-U system.[20]

During the 1980s when the cohort underwent rehabilitation, there was very little access to computers, and the Internet did not exist for private individuals. It is not surprising that only four of them use computers for communication, and that only two are Internet connected. One of these four was able to complete the telephone interview by himself using a computer and voice synthesizer.

Computers can drastically change the lives of people with LIS. Instead of passively responding to the requests of others, the patients can initiate conversations and initiate new topics. They can prepare detailed messages for caregivers who do not have time for lengthy guessing games. On the Internet, persons with LIS are arguably no more handicapped than their caregivers. Except in chat rooms, the time frame for responding to an e-mail is long enough to accommodate slow typing of a person with LIS. The rise in the Internet as a preferred means of communication for much of the able-bodied world, and the integration of Internet and traditional telephone services is a kind of "universal design." One can predict substantial new uses for people with LIS.

Experts in rehabilitation engineering and speech-language pathology devise various patient-computer interfaces such as infrared eye movement sensors.[21-23] Voice simulation programs can provide them with an oral personality. Unfortunately the cost of the computer interface is often substantial, and insurance companies that routinely pay for prosthetic limbs seldom approve prosthetic voices. The inequity might be convenient to the payers, who presume that the person using an artificial communication system will die in a short time frame. The current population shows that the payer's obligation is more long term.

The population chosen for this study—persons who remained locked-in for more than a year—comprise what some might consider a worst case scenario. But these persons had the fortune or resources to be admitted to a world-class rehabilitation facility. They underwent expert rehabilitation including medical stabilization, simplification of nursing care, appropriate techniques in activities of daily living, efficacious assistive and mobility devices, detailed education and counseling, and coordinated discharge planning. It is probable that older, more medically unstable, poorer, less socially supported, or less resourceful persons did not receive such high-quality care.

The World Health Organization's revised "International Classification of Impairment, Disease, and Handicap"[25] defines these aspects of health as quite separate—disease, impairment, activity, participation, and life satisfaction. The LIS cohort demonstrates quite concretely that disease and impairment do not necessarily equate with quality of life.

The results of this study may run contrary to the feelings of many health care providers. Superficially involved for the short term when the patient is at his or her worst, fragile in our own struggles with quality of life, clinicians often comfortably assume that these persons will die anyway, or would choose to die if they only knew what the clinicians knew. As a result, debates about cost, quality of life, euthanasia, and so forth often go on with prejudice—and without the input of the patient.[26] Clinicians may not appreciate that quality of life often equates with social, rather than physical interaction, and that the will to live is strong. It is not surprising that many people with disabilities feel their physicians will be too quick to help them with euthanasia.[2] The data in this study suggest that medical treatment for persons with LIS should be as aggressive as it would be for other people with diseases with potential survival of a decade or more.

For survivors, this research offers realistic hope for reasonable quality of life. The current study, along with the previous 2 studies on the same population, offers them and their families a roadmap toward improved function through rehabilitation. LIS is uncommon enough that many clinicians do not know how to approach rehabilitation. This population of survivors provides clinicians not familiar with LIS a series of specific functional and medical goals that can be accomplished and a big picture view that helps them plan toward improved quality of life.

CONCLUSION

Once they become medically stable, some persons with LIS survive for decades. Contrary to the perceptions of some health care providers, who have not experienced such a severe disability, they typically have a wish to live. They return to home and develop meaningful roles in their families and communities. In the future, access to improved computer voice prosthetics and the Internet will further enhance their quality of life.

REFERENCES

1. Zimbelman J. Changing the present legal prohibitions on assisted suicide is a bad idea. *J Prof Nurs*. 1999;15(3):160–169.
2. Batavia AI. Disability and physician assisted suicide. *N Engl J Med*. 1997;336(23):1671–1673.
3. Gostin L. Editor's introduction: family privacy and persistent vegetative state. *Law, Med Health Care*. 1989;17(4):295–297.
4. Mattson-Prince J. A rational approach to long-term care: comparing the independent living model with agency-based care for persons with high spinal cord injuries. *Spinal Cord*. 1997;35(5):326–331.
5. Baube J-D. *The Diving Suit and the Butterfly*. New York, NY: AA Knopf; 1997.
6. Abresch RT, Seyden NK, Wineinger MA. Quality of life: issues for persons with neuromuscular diseases. *Phys Med Rehabil Clin N Am*. 1998;9(1):233–248.
7. Plum F, Posner JB. *The Diagnosis of Stupor and Coma*. Philadelphia, PA: FA Davis; 1966.
8. Haig AJ, Katz RT, Sahgal V. Mortality and complications in the locked-in syndrome. *Arch Phys Med Rehabil*. 1987;68:24–27.
9. Haig AJ, Katz RT, Sahgal V. Locked-in syndrome: a review of the literature. *Curr Concepts Rehabil Med*. 1986;3:12–16.
10. Patterson JR, Grabois M. Locked-in Syndrome: a review of 139 cases. *Stroke*. 1986;17(4):758–764.
11. Thadani VM, Rimm DL, Urquhart L, et al 'Locked-in syndrome' for 27 years following a viral illness: clinical and pathologic findings. *Neurology*. 1991;41(4):498–500.
12. 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.
13. Cobb S. *Foundations of Neuropsychiatry*. Baltimore, MD: Williams and Wilkins; 1948.
14. Bauer G, Pruggger M, Rumpl E. Stimulus evoked oral automatisms in locked-in syndrome. *Arch Neurol*. 1982;39:435–463.
15. Devivo MJ, Kartus PL, Stover SL, Rutt RD, Fine PR. Seven-year survival following spinal cord injury. *Arch Neurol*. 1987;44:872–875.
16. Greenwood M. *The Errors of Sampling the Survivorship Tables: Appendix I in Reports on Public Health and Statistical Subjects*. No 33. London, UK: His Majesty's Stationery Office; 1926.
17. Dumas A. *The Count of Monte Cristo*. New York: FP Collier and Sons; 1910.
18. Fogg M: Unconsciousness. *Lancet*. 1958;1:433–449.
19. *Block v General Signal Corp*, Illinois Circuit Court, Cook County, Docked 82L3713. 1986;17(4):758–764.
20. Wu Y, Voda JA. User-friendly communication board

for non-verbal, severely physically disabled individuals. *Arch Phys Med Rehabil.* 1985;66:827–828.
21. The Eyegaze Communication System. LC Technologies, Inc., 9455 Silver King Court, Fairfax, VA 22031–4713.
22. Cedrick. Santech Pty Ltd, PO Box 231, Brooklyn Park, South Australia 5032, Australia.
23. ETS Eyetracker. Applied Science Laboratories, 175 Middlesex Turnpike, Bedford MA 01730–1428.
24. Haig AJ. The persistent vegetative state. *N Engl J Med.* 1994;331(20):1380.
25. Hall KM, Knudsen ST, Wright J, Chaarlifue SW, Graves DE, Werner P. Follow-up study of individuals with high tetraplegia (C1-C4) 14 to 24 years post injury. *Arch Phys Med Rehabil.* 1999;80(11):1507–1513.
26. *ICIDH-2: International Classification of Impairments, Activities and Participation. A manual of Dimensions of Disablement and Health. Beta-1 draft for field trials.* Geneva, Switzerland: World Health Organization; 1997.
27. Steffen GE, Franklin C. Who speaks for the patient with locked-in syndrome? *Hastings Cent Rep.* 1985;15:13–15.