

Long-Term Survival, Prognosis, and Life-Care Planning for 29 Patients With Chronic Locked-In Syndrome

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ABSTRACT. 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;73:403-8.

• We present a life-table analysis of a cohort of 29 locked-in syndrome (LIS) patients followed for a minimum of five years, and we report on the status of the chronic LIS patient. Twenty-nine LIS patients who remained locked-in for more than one year were identified. Inpatient charts were reviewed for demographic, medical, and functional data. Telephone followup was obtained to examine medical complications after discharge, survival, neurologic recovery, care issues, and permanent disposition. A life-table analysis was performed on survival data. Cerebrovascular disease was the most common cause of LIS. Survival ranged from 2.02 to 18.15 years. Twenty of the 26 patients available for five-year followup survived; hence, five-year survival was 81%. An alternative method of communication and emotional stress for the patient's caregiver was the key issue in patient care. Most patients were cared for in their own homes. Although minimal late neurologic recovery occurs in chronic LIS, survival may, nonetheless, be prolonged with adequate supportive care. Modern computerized technology offers LIS patients the ability to interact with their environment. This information may assist physicians in making ethical and long-term care decisions with the patient rather than for the patient with LIS.

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KEY WORDS: Mortality; Pons; Quadriplegia (locked-in syndrome); Rehabilitation

It is difficult to think of a physical disability more severe than the inability to speak and move the extremities. In 1966, Plum and Posner¹ first introduced the term "locked-in syndrome" (LIS) to refer to the constellation of quadriplegia, lower cranial nerve paralysis, and mutism, with preservation of only vertical gaze and upper eyelid movement. However, LIS was vividly and accurately described 120 years earlier in Alexandre Dumas's novel *The Count of Monte Cristo*. Herein a character, Monsieur Noirtier de Villefort, was depicted as "a corpse with living eyes." Mr. Noirtier had been in this state for more than six years, and he could only communicate by blinking his eyes.² Dumas highlights the condition of LIS where, unlike the persistent vegetative state, consciousness remains intact. The patient is locked inside his body, able to perceive his environment but almost unable to interact with it.

Previously, LIS was mainly a postmortem diagnosis, as revealed in recent reviews of the literature by us³ and others.⁴ Other than isolated case reports,⁵⁻¹⁰ there was no systematic study of the recovery, morbidity, and mortality of patients with LIS, although systematic studies had been

carried out for severely disabled patients in the persistent vegetative state.^{11,12} In 1987, we reported on the first large cohort of LIS patients and discussed their mean survival, recovery, medical complications, and functional difficulties.¹³

The surprising finding of our previous report was that LIS patients can demonstrate dramatically prolonged survival times. Twenty-four members of our cohort lived with LIS as long as 12½ years. These new data emphasize the need for renewed ethical and medicolegal examination of the needs of the LIS patient. As asked in a Hastings Center report, "Who speaks for the patient with LIS?"¹⁴ With the initial handicap of communicating only through eyeblink, who can decide whether the patient is competent to consent to or to refuse treatment? Fred¹⁵ recounted his dilemma with this situation when his 80-year-old mother became locked-in. In concert with the attending physician, the decision was made, without the consent of the patient, to "have her senses dulled" and provide supportive care only. She died shortly thereafter, with a temperature of 109F.¹⁵ Stumpf¹⁶ argued that "human life is to be preserved as long as there is consciousness and cognitive function in contrast to a vegetative state or a condition of neocortical death."

Ironically, the devices to allow the LIS patient and other patients with severe motoric impairment to "speak" are readily available. The preeminent physicist Stephen Hawking, author of *A Brief History of Time*—which topped the *New York Times* best-seller list for nonfiction—is able to communicate solely through the use of a computerized voice synthesizer. He selects words presented serially on a computer screen; the words are then stored and later presented as a synthesized and coherent message. Similar sys-

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tems, activated through head or eye-gaze control of an infra-red beam, are available for the patient with high quadriplegia or LIS. Unfortunately, many of the patients who could benefit from such technology find it unavailable due to high cost or myriad complexities of third-party payers.¹⁷

In this report, we present a life-table analysis of an enlarged cohort after an additional, prospective five-year followup, and we extend our observations concerning the status of the patient in chronic LIS. It is our hope that the prognostic and life-care data presented here can be used to plan for the needs and cost of caring for the patient with chronic LIS.¹⁸

METHODS

In May 1985, inpatient and outpatient medical records of all persons treated for the previous three-year period, with a diagnosis of brainstem stroke or LIS, were collected for review from a major free-standing rehabilitation hospital. In addition, a request was made to identify and refer these patients to all attending and resident physicians. A cohort of 29 LIS patients who met our inclusionary criteria was identified.

The inclusionary criteria were consciousness, anarthria, quadriplegia, and persistence of the locked-in state for more than one year after onset. Clear evidence of consciousness was defined by mental status examination. Cobb¹⁹ 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.

We chose anarthria, rather than mutism, as a criterion mutism can imply a *refusal* to speak; whereas, anarthria more correctly implies an inability to articulate due to damage to the central or peripheral nervous system.²⁰ Patients were considered anarthric if they demonstrated an inability to produce voluntary speech sounds. Patients were not excluded if they occasionally demonstrated involuntary cries, which can occur in this patient population.²¹

Quadriplegia was defined as the inability to move any limb against gravity. Patients with minimal hand movements that permitted activation of a small switch (such as on a communication device, computer, or environmental control unit) were included as they had no functional advantage over patients who use facial muscles to trigger a switch.

Twenty-nine patients who had been locked-in for more than one year were identified. A one-year time frame was selected as a reasonable criterion to assure that spontaneous recovery had plateaued, and that we would be able to assess long-term vs short-term morbidity and mortality. It has been shown, for example, that the probability of dying after spinal cord injury is greatest during the first year postinjury; patients who pass this milestone demonstrate significant longevity.²² It is expected that LIS patients could fare similarly.

The inpatient charts of all patients fulfilling these criteria were reviewed for demographic, medical, and functional data. Outpatient records were screened for medical complications, and the caretaker (person primarily responsible for

the care of each LIS patient) was contacted by telephone to supply information on medical complications after discharge, neurologic and functional status, and data related to patient care. Follow-up telephone contact was first made in May 1985 and, most recently, in May 1990.

Telephone contact followed the format of a written questionnaire. Data collected included the following: (1) date of onset, (2) frequency and cause of hospitalizations within the previous year, (3) most difficult problems for the subject and caretaker, (4) present placement (eg, home or nursing home or hospital), (5) ability to verbally communicate, (6) limb movement, (7) method of feeding, (8) bladder and bowel 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 or occupational or speech), (16) ability to leave the patient alone during the day or night, and (17) presence of tubes (indwelling catheter or gastrostomy or tracheostomy).

Standard statistical parameters were calculated for the entire cohort, and a life-table analysis was performed on survival data.²³ An approximate formula for the standard error of the survival curve was used, as proposed by Greenwood.²⁴

RESULTS

Twenty-nine patients, 19 men and ten women, fulfilled the study criteria. The most common etiology of LIS (see table 1) were cerebrovascular disease (15 patients or 52%, combining various stroke types) followed by trauma (nine patients or 31%). Unusual etiologies included viral encephalitis and reaction to pertussis vaccine.

Survival in our cohort ranged from 2.02 to 18.15 years (table 2). Six patients had died by the 1990 followup. Causes of death included heart disease (3), pneumonia (1), recurrent cerebrovascular disease (1), and "multisystem failure" (1). The mean age of patients at onset of LIS was 33.6 years (median = 33.0, range = 1 to 70, SD = 16.1). The mean age of patients who died was 50.4 years (median = 45.0, range = 34.3 to 73.0, SD = 15.4).

Twenty-six patients were available for five-year prospective followup; of the 26, 20 survived. Ten subjects had onset

Table 1: Etiology of LIS in 29 Patients

Etiology	n	%
Trauma	9	31
Pontine ischemic infarction	7	24
Hypotension	3	10
Suspected pontine infarction	2	7
Nonbrainstem hemorrhage	2	7
Pontine hemorrhagic infarct	1	3
Midbrain infarct	1	3
Multiple infarctions	1	3
Posttraumatic infarct	1	3
Viral encephalitis	1	3
Pertussis vaccine reaction	1	3

Table 2: Length of Survival After LIS

Patient	Age at Onset (Years)	Survival (Years)	Comments*
Alive			
1	39	1.03	Lost to F/U
2	35	1.48	Lost to F/U
3	45	4.96	Lost to F/U
4	43	6.28	
5	34	6.41	
6	29	6.55	
7	33	7.20	
8	51	7.69	
9	33	7.75	
10	33	8.21	
11	10	8.40	
12	18	8.61	
13	28	8.73	
14	27	8.93	
15	18	11.00	
16	49	11.44	
17	24	11.65	
18	64	11.96	
19	23	13.86	
20	1	14.61	
21	24	15.88	
22	19	16.76	
23	20	18.15	
Dead			
1	52	2.02	Pneumonia
2	31	2.49	Heart failure
3	45	2.54	Multiple causes
4	70	3.05	MI
5	58	3.57	Recurrent CVA
6	19	15.32	MI

* F/U = follow up.

MI = myocardial infarction. CVA = cerebrovascular accident.

of LIS ten or more years before the 1990 followup; nine of the ten subjects survived. Life-table analysis revealed that 81.5% of our cohort of LIS patients who survived the first year would be expected to survive five- and ten-year followups (cumulative survival) (table 3) The survival curve (with standard error) of the cumulative survival is displayed in the figure. Of the 20 patients known to be alive at the 1990 followup, contact with caretakers was available for 18.

Medical Complications Since Discharge from Hospital

Thirteen patients had not been hospitalized within the last year. The remaining five patients were admitted to a hospital on 12 occasions. Discharge diagnoses included renal stones (1), bladder infections (1), hospitalization of primary caretaker (one subject on three occasions), pneumonia (1), electrolyte imbalance (1), feeding tube complications (1), and deep-venous thrombosis (one subject on two occasions). Medical complications after discharge from initial hospitalization included urinary tract infection, pneumonia, pressure sores, urinary tract stones, gastrointestinal bleed, deep-venous thrombosis, skin breakdown, difficulties with dentition, fractures, and gastroesophageal re-

flux. Three patients continued to have tracheostomies in place.

Neurologic Followup

Neurologic status as followup was assessed as a function of verbal communication skills, extremity movements, and feeding. Thirteen subjects improved ten in one parameter, and three in two parameters; one subject was unchanged. Two subjects had subtle loss of function, which may not represent neurologic decline; one subject who had minimal thumb movement stopped using his thumb functionally, and another took a lesser percentage of food by mouth. One subject had a true neurologic decline, with deterioration in all three areas.

Communication. No subjects could speak in full sentences. One could utter single words consistently, four could do so inconsistently, eight expressed involuntary cries, and five subjects could make no sounds at all. Five improved and one regressed.

Extremity movements. Three subjects had no voluntary movements of their extremities, eight had enough voluntary movement to trigger a switch, one could point to a letter board, and three were able to lift objects against gravity. Nine of the 18 patients continued to show improvement since the 1985 followup, and two deteriorated.

Feeding. Ten of the subjects had feeding gastrostomies. Of these ten, four took nothing by mouth, four occasionally ate foods by mouth, and two were primarily fed by mouth. Of the remaining eight subjects, two had unrestricted consistencies (ie, thin fluids, thick fluids, solids); whereas, six were restricted. Two subjects improved in feeding function, and two subjects worsened.

Data on sleep patterns were available for only 17 subjects. Ten subjects slept more than five but less than eight hours per night, and seven subjects slept eight to 12 hours.

Functional Issues

Bladder. Five patients had indwelling urinary catheters (four of the five were women), and none used intermittent catheterization. Six men used external catheters, and seven needed physical assistance to void voluntarily. There had been no significant changes in bladder management during the five years. Fourteen subjects used a regular bowel program with a suppository or other method, and four were continent but required physical assistance to evacuate.

Mode of communication. Three patients had no formal method of communication. One subject used his voice for functional communication, three subjects used facial movements to encode yes and no responses, four subjects used facial movements in conjunction with a letter board, and seven subjects had a computer with printer or voice synthesizer. Two subjects who had not changed since the 1985 followup now harnessed a computer or letter board to improve communication. Two had abandoned devices as they obtained the ability to gesture. One subject improved neurologically, enabling the use of a computer.

Mobility. Two subjects never left their beds. 13 were pushed in a manual wheelchair, two used electric wheel-

Table 3: Life-Table Analysis

Interval	Alive at Interval Onset	Deaths During Interval	Withdrawn During Interval	Cumulative Survival (CS)	Standard Error of CS	Mortality
0-1	29	0	0	1.0000	.0000	
1-2	29	0	2	1.0000	.0000	
2-3	27	3	0	.8889	.0604	
3-4	24	2	0	.8149	.0748	
4-5	22	0	1	.8149	.0748	.1851
5-6	21	0	0	.8149	.0748	
6-7	21	0	3	.8149	.0748	
7-8	18	0	3	.8149	.0748	
8-9	15	0	5	.8149	.0748	
9-10	10	0	0	.8149	.0748	.1851
10-11	10	0	0	.8149	.0748	
11-12	10	0	4	.8149	.0748	
12-13	6	0	0	.8149	.0748	
13-14	6	0	1	.8149	.0748	
14-15	5	0	1	.8149	.0748	.1851
15-16	4	1	1	.5821	.2039	
16-17	2	0	1	.5821	.2039	
17-18	1	0	0	.5821	.2039	
18-19	1	0	1	.5821	.2039	

chairs, and one was independent in the use of a manual wheelchair. One patient improved, and three declined in functional mobility since the 1985 followup.

Psychosocial Issues

Residence. Eleven of the 18 survivors for whom follow-up data were available resided in their own homes, two were permanent hospital residents, and five were in nursing homes. Since the 1985 follow-up, one person was discharged from hospital to home, and two went from home to nursing home.

Personal care. Of the 11 subjects who were at home, six had more than 40 hours per week of hired assistance (licensed practicing nurse or aide), two had part-time help (four to 15 hours per week), and three had no outside help. Five subjects had variable respite help from volunteers. Nine of the 18 subjects continued to receive therapy (physical or occupational or speech) for three to 14 hours per week. Of the 11 subjects at home, only three families felt they could leave the patient unattended while they were outside the home. Caretaker problems included communi-

cation with the patient, health of the caretaker, emotional stress and frustration, and reliability of hired caregivers from outside the home. Caretakers rated the subject's mood as "frequently depressed" for two, "occasionally depressed" for eight, and "seldom depressed" for seven.

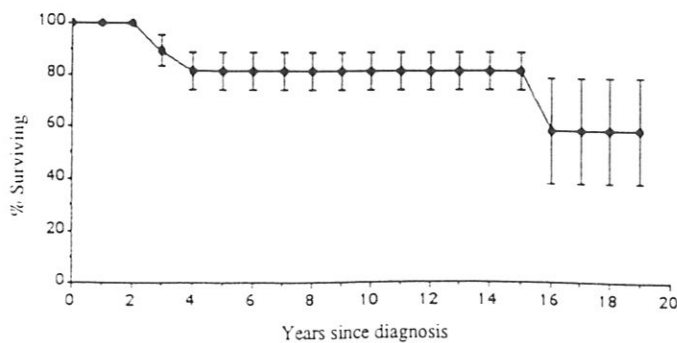
Vocational and avocational pursuits. Fourteen of the 18 subjects left home at least once per month (range = 1 to 30, median = 4). Two of the subjects were gainfully employed on a part-time basis; one volunteered as a teacher, and one assisted in the operation of a small business. The teacher had experienced significant neurologic improvement. Only one of the subjects improved to a level where he was independent in avocational activities involving mobility. Six subjects could participate in interactive bedside activities, and eleven primarily enjoyed passive activities such as listening to music or taking rides in their wheelchairs.

DISCUSSION

How are we to care for the LIS patient? This paper proves definitively that patients can survive for extended periods while remaining in the locked-in state. Only by studying their survival rate and daily condition are we able to address the issue of competency and the long-term needs of these patients.

Survival and Mortality

It has been suggested that long-term survival in the LIS patient is rare.²⁵ Retrospective analysis of mortality suggests that mortality is high in acute LIS (67% for vascular cases and 41% for nonvascular cases), with 87% of the deaths occurring within the first four months.⁴ However, the biased and retrospective nature of this literature makes it difficult to draw conclusions. Our data clearly suggest that once a patient has medically stabilized in LIS for more than one year, the life span can be prolonged. This is in clear contrast to patients in the persistent vegetative state,



Survival curve for chronic LIS.

even though LIS patients are physically as disabled as persistent vegetative state patients. Higashi and associates¹¹ and Higashi and colleagues¹² followed a large cohort of such patients, and they described a 65% mortality rate during the first three years and 73% mortality within the first five years. Only 10% recovered partially from the vegetative state.^{11,12}

There are some statistical limitations for investigating survival in our cohort. Specifically, many of the patients had been locked-in for many years before initial identification in 1982 and subsequent followup. To optimally study the survival rate of patients with LIS, patients should be identified at the time of onset and followed until their death. Nonetheless, our cohort does display the following four requirements outlined by Colton²³ for a life-table analysis: (1) a clear and well-defined starting point (onset of LIS); (2) a clear and well-defined endpoint (death); (3) patients are observed at different times and, at study termination, have been observed for different lengths of time; and (4) at study termination, the endpoints for some patients are unknown.

Reported causes of death in LIS are predominantly respiratory (including pneumonia, respiratory arrest, and PE), followed by recurrent cerebrovascular event.^{3,4} Other, less common causes included cardiac complications, sepsis, GI hemorrhage, disseminated intravascular coagulation, and pontine abscess. Twenty-three of 83 patients had unknown cause of death in one retrospective review.⁴ The lack of postmortem follow-up in previous series and in our own limits the accuracy of determining cause of death.

Neurologic Recovery

Bauer and coworkers²⁶ have suggested a tripartite classification for LIS: (1) classical (complying with Plum and Posner's¹ original criteria), incomplete (demonstrating movements other than in the upper eyelid and vertical eye movements), and total (demonstrating no movement). Furthermore, patients may remain in the locked-in state transiently or chronically. In this report, we present 29 cases of LIS that fall into each of Bauer and coworkers' three categories. All of these subjects remained locked-in for more than one year—beyond the usual time when spontaneous recovery for central nervous system injury occurs.

Our results indicate that late neurologic recovery does occur in chronic LIS, but the amount of recovery is relatively slight. Patterson and Grabois⁴ graded neurologic return as no recovery, minimal, moderate, full, and no neurologic deficit. Only one of our patients made a moderate recovery (independence in some but not all daily activities); whereas, the others made no recovery or had minimal return of voluntary motor function, leaving them still totally dependent for their care. Our data agree with the retrospective study of Patterson and Grabois⁴ who found that lack of significant recovery within the first six months was strongly predictive of chronic and persistent LIS.

The ability to communicate orally never improved to a highly functional level; thus, it was of critical importance that most of the survivors improved enough neurologically to be able to activate a switch for a communication device or computer. De Graaf and Rybnikar²⁷ reported how even

subtle motor control—in their case, mandibular movements—can be used to activate an electronic switch. Several of the patients in our cohort developed some voluntary control of previously involuntary cries, and they were able to use them for communication. This allowed caretakers to leave the patient's room occasionally, and it allowed family members who were primary caregivers to get a reasonable amount of sleep at night. Another of the patients could initiate laughter to draw the attention of his caretaker. Most patients did not regain enough swallowing function to safely ingest a satisfactory caloric intake, and they continued to require feeding gastrostomies for at least part of their nutrition.

Sleep studies in LIS have revealed a disorganization in non-REM sleep, with an absence or reduction of sleep stages 3 and 4. Overall, sleep was reduced, with a range of 1.25 to 6.5 hours.⁴ This was not the case in our cohort of patients, all of whom slept more than five hours per night, and seven of 17 slept more than eight hours. Our subjects did not offer sleep-related complaints.

Functional and Psychosocial Status

Bowel and bladder management changed little in most patients, mirroring the needs of other severely disabled populations such as persons with spinal cord injury.²⁸ Nursing care needs, including respite care, must be carefully structured in a financial life-care plan if these patients are managed at home. Most families did not feel they could leave the LIS person at home alone at any time; the psychologic toll on the primary caretaker of these patients cannot be overestimated.

Most of the patients were extremely isolated by their lack of mobility and their inability to pursue vocational and avocational interests. It could be anticipated that such seclusion would represent a severe psychologic stress for these individuals. However, our depression scale, albeit crude in comparison with previously described scales in able-bodied populations,^{29,30} did not suggest that most of our subjects were severely depressed. This agrees with the limited observations of Cappa and colleagues.⁵ The decline in functional mobility in three of our patients most likely reflected the reluctance of caretakers to transfer immobile patients and propel them in a manual wheelchair rather than a true neurologic decline.

CONCLUSION

Medical and rehabilitation personnel who work with LIS patients need a better understanding of long-term outcome. Ethical decisions must be made in light of morbidity and survival data as presented in this report. Improvement in medical care and biomedical electronics have created realistic goals in communication, feeding, and mobility; these include computers, printers, and synthetic voice machines triggered by sensitive switches, electromyographic devices, and infrared eye-gaze sensors.

A better understanding of the outcome for LIS patients allows insurers to make long-term plans for the costs of sustaining these severely disabled patients. Steps in the for-

mulation of a long-term plan include (1) determining the extent and sequelae of the physical and cognitive impairment; (2) estimating prognosis for recovery; (3) estimating the need and benefit of further medical and rehabilitative interventions; (4) calculating the costs of future personal needs (eg, wheelchairs, orthopedic equipment, home furnishings and modifications, medical supplies, and recreational equipment); and (5) financial adjustment of estimated costs for future interest rates and inflation.^{18,31} We hope the information provided in this report will help physicians make ethical long-term care decisions with rather than for the patient with LIS.

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