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Mortality and Complications of the Locked-in Syndrome

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ABSTRACT. Haig AJ, Katz RT, Sahgal V: Mortality and complications of the locked-in syndrome. Arch Phys Med Rehabil 68:24–27, 1987.

• The locked-in syndrome is a severe disability consisting of quadriplegia and anarthria with preserved consciousness. No large series of cases have been reported and very few cases of long-term survival have been described. We present a follow-up of 27 patients "locked-in" for more than one year. Twenty-four were still alive up to 12.5 years after onset, with a mean survival of 4.9 years. Significant recovery was noted in only a few patients. Seventeen patients lived at home at the time of study. Eight were never hospitalized after the initial event. Gastrostomy and tracheostomy tubes and indwelling catheters were eventually removed from many patients. Electronic devices were used by ten patients to facilitate communication. We conclude that rehabilitation and medical care must be planned carefully, given the length of survival shown in this group.

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

It is difficult to think of a physical impairment more severe than the inability to speak and move one's extremities. The locked-in syndrome (LIS), usually caused by ventral pontine lesions, has been defined as quadriplegia and anarthria with preserved consciousness.[7]

Before the 1960s LIS was not recognized as distinct from conditions such as akinetic mutism or persistent vegetative state, in which the patient is not conscious.[11] While much of the literature discusses etiology and clinical findings, few articles address morbidity, mortality, or functional outcome. Some authors have concluded that LIS is almost universally fatal.[5,10]

A review of 117 cases of LIS published over the last 30 years showed that 69 (59%) were postmortem reports.[6] Many of the articles did not comment on change in functional status. Of the 37 cases in which recovery was mentioned, 14 remained locked-in, 13 recovered to the point where they were no longer considered locked-in, and ten had full or near-full recovery. Length of follow-up was highly variable. Only two persons were reported to have survived in the locked-in state for more than two years.[3,5]

It is difficult to extrapolate useful information on mortality from the literature because of the variability in follow-up and the bias inherent in many of the articles which required postmortem pathologic material.

With electronic communication devices such as computers, printers, and synthetic voice machines triggered by sensitive switches, electromyographic devices, and eye gaze sensors, there is a potential for improved quality of life for LIS patients.

Since the cost of medical care is a key factor in the decisions of responsible health care providers, it is imperative that the morbidity and mortality of LIS be evaluated. This will enhance our ability to plan intelligently for the care of these patients.

A meaningful study of mortality in LIS must take into consideration the length of time subjects stay locked-in. A case of LIS has been reported which lasted only a few minutes, with good recovery.[9] The mortality of patients who make early significant recovery may be related more to the degree and speed of their recovery than to the state of quadriplegia and anarthria. Including short survival cases would reflect mortality from complicating factors such as multiple trauma and respiratory arrest. Analysis of either of these conditions would detract from the question of how to plan the rehabilitation of long-term LIS patients.

We present here an analysis and follow-up of 27 patients locked-in for more than one year. Their mortality, morbidity, and functional status are discussed.

METHOD

The medical records of all persons admitted to the Rehabilitation Institute of Chicago from 1982 through 1985 with a diagnosis of brainstem stroke or LIS were selected for review. Staff physicians, nurses, and therapists were interviewed to identify additional patients who might be appropriate. Patients meeting the following criteria were included in the study:

(1) Consciousness, as defined by Cobb[4] is awareness of self and environment. Patients considered for the study had a mental status examination performed by either the admitting physician or a staff psychologist. Only patients able to respond consistently to questions about themselves and their environment were included.

(2) Anarthria was interpreted as lack of voluntary speech sounds. Patients with LIS may make involuntary cries.[1] Persons who could consistently and voluntarily phonate were excluded.

(3) Quadriplegia was defined as inability to move any limb against gravity. Patients with minimal hand movements which permitted activation of a small switch were included, as they have no functional advantage over persons who use facial muscles to trigger a switch.

(4) The study was limited to patients who remained locked-in for more than one year after onset. Patients first evaluated more than one year after onset were included if they met the above criteria at evaluation. Patients who were seen before the

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Table 1: Etiology of LIS

	N	%
Trauma	8	29
Pontine ischemic infarct	6	22
Hypotension	3	11
Suspected pontine infarct	2	7
Nonbrainstem hemorrhage	2	7
Pontine hemorrhagic infarct	1	4
Midbrain infarct	1	4
Multiple infarctions	1	4
Post-traumatic infarct	1	4
Viral encephalitis	1	4
Pertussus vaccine reaction	1	4

end of the first year were included only if subsequent evaluations revealed that they met the criteria after one year. One patient who met the criteria initially but died before the end of the first year was excluded.

The inpatient charts of all patients fulfilling the criteria were reviewed for demographic, medical, and rehabilitation data. Outpatient records were screened for medical problems. The caretaker of each LIS patient was contacted by telephone to supply information on medical complications after discharge and current functional status.

RESULTS

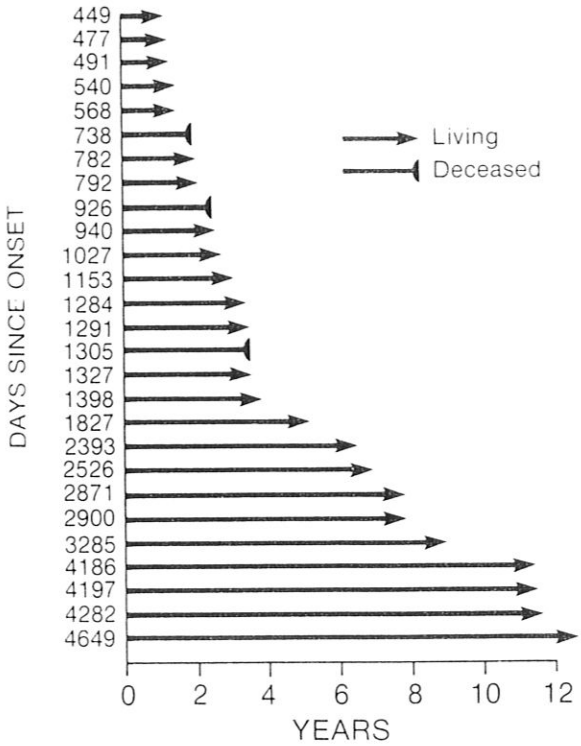
Twenty-eight patients, 18 men and 9 women, fulfilled the study criteria. One patient could not be located for follow-up and was dropped from the study. Average age at onset was 32 years (SD 14.9 years) with a range of 0.5 to 64 years. The etiology of LIS is listed in table 1. The subset of patients whose LIS was caused by cerebrovascular accident was slightly older than the mean, averaging 39 years.

Length of acute hospitalization could be determined in 19 cases. The mean was 159 days, with a range of 30 to 406 days. Fourteen patients lived in the Chicago metropolitan area before the onset of LIS; four lived within 150 kilometers of the city. The nine remaining patients were referred by out-of-town sources. Survival after onset of LIS ranged from 1.2 years to 12.8 years with a mean of 4.9 years (fig).

Three men selected for the follow-up study had died before follow-up. Their LIS was caused by cerebrovascular accidents (pontine ischemic, midbrain ischemic, and multiple infarctions). Death was due to multiple causes in one, to pneumonia in another, and to a subsequent cerebrovascular accident in the third. Length of survival was 2.5, 2.0, and 3.6 years, respectively. The first patient lived at home, the second in a nursing home, and the third in an acute-care hospital for much of the time before death. At time of onset of LIS all were older than the average (45, 52, and 58), and had indwelling urinary catheters and gastrostomy tubes; two had tracheostomies. Caretakers reported that the patients still met the inclusion criteria of this study within a month of death.

Reports of hospitalizations were obtained for 23 patients. Eight had not been hospitalized since their initial event, six had been hospitalized only once, and five had been admitted more than once, including one patient who was admitted eight times. Seven of the 23 had been admitted to an intensive care unit after the initial hospitalization.

Caretakers were asked to identify the following specific causes



Survival of 27 chronically locked-in patients.

of morbidity (regardless of whether the patient had been hospitalized for the problem): urinary tract infections, pneumonias, pressure sores, urinary tract stones, gastrointestinal bleeding, and deep venous thrombosis. Urinary tract infections were the most common complication, affecting 56% of the patients (table 2).

Gastrostomy tubes were used by 23 patients on admission, and by 18 on follow-up. Indwelling catheters were used by 18 patients on admission and by 11 on follow-up. Tracheostomy tubes were used by 17 patients on admission and by 7 on follow-up. No statistically significant relationship was found between medical complications and the presence of the tubes or catheters.

Outcome status was evaluated in five areas: mental status, communication, motor function, living arrangements, and daily care.

Comprehensive evaluation of cognition was difficult because of the patients' limited ability to interact with the environment. A few patients were tested formally beyond a mental status examination at admission, but such testing was time-consuming and was sometimes considered unreliable. Nine-

Table 2: Medical Problems After Discharge from Hospital

	None		1		> 1		No Info
	N	(%)	N	(%)	N	(%)	N*
Urinary tract infection	10	(43)	13	(54)	1	(4)	3
Pneumonia	14	(64)	8	(36)	1	(4)	4
Pressure sores	16	(73)	7	(32)	0	(0)	4
Urinary tract stones	17	(77)	5	(23)	1	(4)	4
Gastrointestinal bleed	18	(81)	4	(18)	1	(4)	4
Deep venous thrombosis	20	(91)	3	(14)	0	(0)	4

*"No information": excluded from percentages.

Table 3: Oral Communication on Admission and Follow-Up

	Admission	Follow-up
No sounds	21	10
Involuntary cries	4	7
Rare words	2	6
Consistent single words	0	1
Sentences	0	2
No information	0	1

teen patients (68%) were not found to have decreased mental status during rehabilitation. Eight (29%) were able to respond to questions consistently and accurately, but demonstrated cognitive deficiencies in learning and retention in either formal tests or therapy activities. Three of the eight suffered LIS secondary to trauma, two had had nonpontine intracerebral hemorrhages, and three had LIS as a result of pontine ischemic infarct, hypotension, or multiple infarcts. The cognitive performance of one patient was not well documented, but he was clearly able to respond to questions.

The ability to vocalize either involuntarily or voluntarily improved in 16 patients (table 3). Some caretakers noted that reliable involuntary cries caused by pain or emotional upset allowed them to leave the patient's bedside. Three patients consistently uttered words, but were difficult to understand.

Nonverbal communication was used by all but three patients at follow-up. Two patients were able to point to letters—one with head movements, the other with upper extremity movements. All had learned a system of eye or facial movements interpreted by the observer as "yes" or "no." Nine also learned to use a letter board,[13] which permitted them to spell to a trained observer. Ten of the patients had electronic equipment which allowed them to communicate with untrained observers.

None of the patients achieved recovery of motor control sufficient to move an extremity against gravity. Some did make minor improvements (table 4), which could be harnessed. Electric wheelchair use was possible for seven patients who used hand, head, or tongue movements.

Eighteen (66%) of the patients were at home, six (22%) were in nursing homes, two (7%) were permanent residents of acute-care hospitals, and one (4%) was undergoing rehabilitation at our institution.

The exact amount of time spent caring for each patient could not be easily documented, but caretakers were able to state the amount of time they were willing to leave patients alone. Four patients could be left alone in their rooms for eight to 12 hours at night; and ten were able to be unattended for up to four hours. In addition, six were believed safe if left alone in a house for more than 30 minutes. Four caretakers did not answer the question.

The primary caretaker for 13 of the 17 patients living at home was a family member. Three had nursing help for eight to 12 hours a day; five others had an aide for one to eight hours a day. Of the four non-family primary caretakers, three were nurses and one was an aide. Eight patients had regular physical therapy sessions, six had occupational therapy, and six were seen by speech pathologists.

Table 4: Motor Recovery on Admission and Follow-Up*

Movement	Admission	Follow-up
None	16	11
Nonfunctional	10	6
Can trigger switch	1	6
Can point or type	0	4

*None of the patients could voluntarily lift an extremity against gravity.

Table 5: Method of Feeding on Admission and Follow-Up

	Admission	Follow-up
Gastrostomy in place:		
Nothing by mouth	18	7
"Treats" by mouth	4	6
Primarily oral feeding	2	6
	24 (88%)	18 (66%)
No gastrostomy:		
Limits on oral feeding	2	1
Unlimited oral feeding	1	7
	3 (11%)	8 (30%)

Methods of bowel and bladder control were also studied. Four of nine patients who had indwelling catheters early in their course of treatment were weaned from them. Ten men used external collecting devices. Six patients signalled for a urinal when needed. Two remained incontinent of urine. Only one patient signalled for a bedpan before spontaneous bowel movement. Most others used a routine daily bowel program, usually requiring suppositories.

Table 5 shows changes in the feeding methods. All but seven of the patients were able to take at least some food by mouth at follow-up. Often oral intake depended on involuntary mouth movements.[9] Six of the eighteen gastrostomy tubes were still in place only to ensure hydration.

DISCUSSION

Medical and rehabilitation personnel who work with LIS patients need to have some expectation of morbidity and mortality to plan proper care. Legal[2] and ethical[12] debates hinge on these factors. Neither morbidity nor mortality has been systematically addressed in the past. A review of 117 cases from the literature[6] revealed that 67% of the patients were dead, and that 75% of 44 reported causes of death were pulmonary. In contrast, this study records only 11% mortality in 27 patients with an average of 4.9 years follow-up. Pulmonary problems did occur; eight patients had pneumonias, one of whom died. Fourteen patients had one or fewer hospitalizations after the initial onset of LIS.

The relationship of gastrostomy, tracheostomy, and indwelling urinary catheters to morbidity in LIS has not been discussed in the literature. In this study most of the patients survived for years despite the presence of these devices. The removal of these tubes allowed oral feeding, continent voiding, and easier respiratory care in some patients.

Recovery from LIS has been reported[8] but not systematically studied. In this group of patients studied after the first year, minimal but significant recovery of motor or speech ability was noted in some patients.

Functional abilities were noted in the two documented cases of long-standing LIS, but neither used electronic communication devices. At least ten patients presented here used these devices.

The cost of care as measured by manpower has never been commented upon in the literature. Most patients discussed here

had 24-hour-a-day care, but for 17 the care was provided at home. A family member was most often the primary caregiver. There were patients who had more extensive care: five were in nursing homes, and one lived permanently in an acute-care hospital. Four of the patients who were at home received most of their care from a nurse.

This study clearly demonstrates prolonged periods of survival among LIS patients admitted to a rehabilitation facility. We present the longest surviving case on record, 12½ years, as well as many other stable LIS patients. Many of these patients were successfully cared for in their home setting, usually at the patient's request, at dramatic savings to medical reimbursement agencies. Medical complications and hospital readmissions have not been obstructions to continued home care.

Careful analysis of LIS patients' future medical and rehabilitation needs must be undertaken in consideration of these findings of extended survival. A cooperative study is needed to pool our knowledge about the medical care of the LIS patient.

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