



## Review on Locked-in Syndrome

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### ABSTRACT

The locked-in syndrome (pseudocoma) defines people who are awake and conscious but selectively deafferented, i.e., have no means of creating speech, limb, or facial movements. It is most commonly caused by acute ventral pontine lesions. People with such brainstem lesions frequently remain unconscious for days or weeks, requiring artificial respiration, before eventually awakening but remaining paralysed and mute, seemingly resembling patients in a vegetative state or akinetic mutism. It has been demonstrated that more than half of the time, the family, rather than the physician, is the first to notice that the patient is aware... Clinicians must be familiar with the disease because there is a high risk of incorrect diagnosis, such as coma or vegetative state, following a lengthy unconscious state. It is frequently relatives or caregivers that first notice the aware state and remark that the patient may communicate through his eyes. There has recently been an upsurge in public awareness of this rare ailment, and more cases have been reported.

**Keywords:** Locked-in Syndrome (LiS), Electroencephalogram (EEG), amyotrophic lateral sclerosis (ALS), Patterson and Grabios' analysis

### Introduction<sup>1-11</sup>

The phrase "Locked in Syndrome" was coined by Plum and Posner in 1966 to describe a neurological disease known as ventral pons infarction. Quadriplegia, inferior cranial nerve paralysis, and aphonic support with just an upward glance and eyelid mobility serve as metaphors for the condition. Because consciousness is still intact, the patient is able to talk appropriately and coherently through eye blinking. Patients suffering with locked in syndrome are imperceptibly trapped inside the human body, aware of their surroundings but with limited communication abilities.

Once consciousness was restored, LiS was categorized as quadriplegia and anarthria in 1986. LiS is caused by all-four-limb palsy, anarthria, and lower nerve loss caused by a brain stem lesion. Anarthria is caused by bilateral paralysis of the laryngeal facio-glossopharyngo. The "locked-in" state is described by various words in the literature. "Locked-in syndrome," "de-efferented state," and "pseudocoma" are all identical phrases. "Ventral brainstem syndrome ventral pontine syndrome Cerebro medulla spinal disconnection "Mortality can be decreased with early rehabilitation and more effective medical care".

Even when LIS patients have serious physical impairments and are medically stable, their life expectancy can be enhanced as a result.

### LIS<sup>12</sup>

Locked-in syndrome (LiS) has three main types, or forms, including:

- The classic form: If you have this type of LiS, you are completely immobile (loss of voluntary movement), but you can still blink and move your eyes vertically (up and down). You still retain your regular cognitive functions. You may hear as well.
- The incomplete form: This sort of LiS is identical to the classical version except that specific parts of your body can have movement and feeling functions.
- Complete body paralysis and lack of eye movement are symptoms of the total immobility variant of LiS, but cognitive function is unaffected. An electroencephalogram (EEG), a test that records brain waves, can be used to examine cortical function to determine whether a patient with this kind still maintains cognitive (thinking and reasoning) function.

### Etiology<sup>13,14</sup>

Most frequently, damage to the pons, a specific region of the brainstem, results in locked-in syndrome. Important neural connections between the cerebrum, spinal cord, and cerebellum are found in the pons. All the motor fibres that travel from the brain's grey matter through the spinal cord to the body's muscles are cut off in locked-in syndrome, and there is also damage to the brainstem regions crucial for speech and facial control. Trauma is a less frequent cause of damage to the pons than tissue loss due to lack of blood supply (infarct) or bleeding (haemorrhage). A blood clot (thrombosis) or stroke are two examples of the many diverse disorders that can result in an infarct. The loss of the myelin sheathing that surrounds nerve cells (myelinolysis),

inflammation of the nerves (polymyositis), infections in specific regions of the brain, tumours, inflammation of the nerves, and certain diseases like amyotrophic lateral sclerosis are additional conditions that can result in locked-in syndrome (ALS).

Any lesion affecting the ventral pons and midbrain, including vascular lesions, tumours, infections, traumas, and demyelinating illnesses, might result in locked-in syndrome.

## Epidemiology <sup>14</sup>

The average age of onset for all cases of locked-in syndrome has typically ranged from 30 to 50 years. The syndrome has a little male prevalence. Men and women in the ALIS study who obtained LiS from vascular and nonvascular sources were similarly affected: 51.2% of men and 48.1% of women were diagnosed. The age of onset in this study ranged from 22 to 77 years old, with a mean of 46.79 years. Other retrospective studies found similar results, with mean ages ranging from 33 to 45 years for both vascular and nonvascular causes and, in general, more males affected. Many patients with LiS due to a vascular cause have concomitant diseases such as hypertension, major artery atherosclerosis, and diabetes.

## Diagnosis

Differential diagnosis is intended to comprehend symptoms which may match the acute quadriplegia and trauma-related mutism posed by the patient. The LiSt of diagnoses contain

- Mental health conditions. Coma, unresponsive wakefulness, and state of limited awareness are the three DOC classifications. Perception is characterised by both alertness and knowledge. While awareness needs higher order processing that integrates both sensory input and motor information, alertness is predicated on the healthy operation of the reticular nerves, thalamus, and cortex.
- Injury to the upper cervical spinal cord. An upper cervical SCI was contained because to the paralysis presentation. It was determined that the injury should be above level C5 when the motor test begins because both of the main muscles LiSted by the International Standards for Neurological Classifications of Spinal Cord Injury Association (ISNCSCI) within the standard motor test were rated at zero out of five on a bilateral basis.
- Acetic Mutism With a point of alertness and intact ocular tracking, AM is a condition marked by a diminished neurological drive and a decline in nearly all motor activities, including face expression, movements, and voice output.

Diagnosis considered		Key finding to reject	
Disorder of consciousness	↓	Evaluation of patient knowledge and ability to communicate with the eye	×
Upper cervical spinal cord injury	↓	Observation of normal, still respiration and supraspinal muscle weakness	×
Akinetic mutism	↓	Few automatic defensive extension / equilibrium responses and no pain relief	×
Locked in syndrome		Suspect lead diagnosis by dismissal of all potential diagnoses	✓

**Table No.1: Differential interpretation: acute quadriplegia and onset of mutism**

## Treatment <sup>15</sup>

Locked-in syndrome has no specific treatment. The primary form of treatment for locked-in syndrome is supportive care. The following are examples of supportive care:

- Support for breathing;
- Good nutrition;
- Prevention of immobilization-related problems include blood clot development, lung infections, and urinary tract infections;
- Prevention of pressure ulcers;
- Physical treatment to prevent contractures
- Speech therapy to aid in the development of eye blinks and/or vertical eye movements for communication
- Potential computer terminal control based on the patient's eye movements

Rarely, the patient's condition may become better with treatment of the underlying cause, such as reducing a tumour or quickly treating a medical overdose.

### Development of communication devices<sup>14</sup>

Rehabilitation engineers and speech language therapists are working to advance patient-computer interfaces such as infrared eye movement sensors and computer voice prosthesis. People with locked-in syndrome now have more freedom because of computers, which allow them to start conversations, write inquiries or other messages, and use the internet. Patients who are unable to speak may be able to communicate widely when augmentative communication tools are coupled to a computer. The straightforward, inexpensive, and transportable alphabet board must support sophisticated technologies. If a patient regains distal movement, they can point at the letters; if not, a family member or caregiver can help them use the board. A well-used communication book containing daily activities, news events, visits, and programmed changes can help keep everyone up to date.

A	B	C	D	End of word	
E	F	G	H	End of sentence	
I	J	K	L	M	N
O	P	Q	R	S	T
U	V	W	X	Y	Z

**Fig 1: AEIOU alphabet board. The assistant calls out the colors and the patient signals the required colour by an upward eye movement. The assistant then sequentially calls out the letters on that line. The chosen letters are written down to formulate a sentence, question, or statement.**

### Prognosis<sup>16</sup>

Depending on the underlying cause of the locked-in condition, mortality and recovery are widely different. In the acute context, the death rate for locked-in syndrome is typically significant. The estimated 5-year and 10-year death rates are 84% and 31%, respectively, according to retrospective research and analysis models. An overall death rate of 60% was found by Patterson and Grabois' analysis of 139 LIS patients. Compared to nonvascular cases, which had a mortality rate of 41% (14 of 34 patients), vascular cases had a mortality rate of 67% (70 of 105 patients). The motor ability of many survivors in this investigation had only slightly improved. Based on how well they had recovered a few months after the commencement of their disease, Patterson and Grabois divided their patients into five groups:

No improvement: patients who showed no improvement in their motor skills.

Patients with the very minimum of recovery showed some voluntary motor return but were completely reliant on others for care.

Patients with a moderate recovery are able to do some but not all activities of daily living due to a noticeable return of motor function.

Patients who, with very minor neurological abnormalities, were able to undertake all activities of daily living were said to have made a full recovery.

Patients who stated they had no neurological impairment did not

The study showed that nonvascular cases recovered to full capacity more frequently. Out of the 34 patients who had a nonvascular condition, 18 showed some improvement in their motor skills. In addition, full recovery was achieved in 12 out of the 18 recovery cases in the first six months.

Only 29 of the 105 total vascular patients had experienced some sort of healing since their illness had started, though. Only two cases—13 of 16—of the patients who survived after the first four months showed evidence of full to partial functional improvement.

### Quality of life<sup>14</sup>

Many patients decide to move back in with their families, supposedly allowing for more frequent social interactions. The patient's desire to live may improve after returning home. On the other hand, the family is forced to bear a long-term physical and psychological cost. Community care is frequently sparse and the caregivers are not well supported due to a lack of financing. Only two instances of patients going back to work were identified. The first was a lawyer who gave legal advice by blinking Morse code, and the second was a teacher of math and spelling who used a mouth stick to activate an electronic speech device.

In the only review of quality of life, the authors discovered that a group of seven patients with locked-in syndrome had a Spitzer quality of life index score that was lower than that of cancer patients but higher than that of patients who were terminally ill. Of the five patients who answered the general health questionnaire, three were depressed, but they all said they would want antibiotics if they developed pneumonia. After 11 years, the longest-living group of patients studied had a survival rate of 54%, 46% had previously considered euthanasia, and none had a "not for resuscitation" order. The discovery that people with locked-in syndrome who are nonetheless severely incapacitated rarely seek to end their lives refutes the common belief that such patients would have been better off dead.

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## Conclusion<sup>16</sup>

Patients with LIS syndrome typically have a poor prognosis, remain locked in, and only partially regain function. Scores on QoL stay poor throughout time, particularly when it comes to the motor element. Long-term complications for LIS sufferers also include depression. The prognosis for children with LIS is generally better than for adults.

The cornerstone of LIS rehabilitation is multidisciplinary rehabilitation. Rapid action is essential for the long-term prognosis in the acute setting. Aggressive verbal, physical, respiratory, and swallowing measures must be taken in the subacute situation. Long-term therapy includes preventing comorbidities and managing the quality of life. Independence regaining becomes more challenging for LIS patients over time. Some of the long-term therapies used in the patients' long-term care include speech therapy, respiratory therapy, visual stimulation, and music therapy.

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