

# Review on Locked-in Syndrome

Nolin Johncy<sup>1</sup>, A. R. Shabaraya<sup>2</sup>

<sup>1</sup>Pharm D, <sup>2</sup>Principal and Director,  
Srinivas College of Pharmacy, Mangalore, India

Corresponding Author: Nolin Johncy

## ABSTRACT

The expression "Locked in Syndrome" was established by Plum and Posner in 1966 which adverts to a neurological condition identified with ventral pons infarction. The disorder is symbolized by quadriplegia, inferior paralysis of the cranial nerve and aphonic sustaining only the upward look and motion of eyelid. Patients with locked in syndrome is unerringly sealed within the human form, conscious of their surroundings but with a confined capacity to communicate with it. Cerebrovascular malady is the most driving etiology of LIS. Infarction of the ventral pontine following hindrance of basilar arteries is usually the result. Another significant and recurrent instigation of LIS is brain injury due to trauma. Conveying of messages for LIS persons involves using alternative broadcasting, such as eye blinks or eye movements for a yes-no code or communication boards with alphabets or marks suggested by eye gestures. The initial treatment is on the preservation of an airway and adequate oxygenation. Insurers can be helped to prepare for the expense of treating these seriously impaired patients in the long term by a greater understanding of the outcome for LIS patients. Progressed study is desired to effectively elucidate the advantages of low-level patients in-patient rehabilitation and health care providers need to identify the value of factual diagnosis for the treatment of development plans, giving preference to a team approach that has the ability to manage patients with poor response.

**Keywords:** Locked in syndrome, quadriplegia, infarction, oxygenation

## 1. INTRODUCTION

The expression "Locked in Syndrome" was established by Plum and Posner in 1966 which adverts to a neurological condition identified with ventral pons infarction. The disorder is symbolized by quadriplegia, inferior paralysis of the cranial nerve, and aphonic sustaining only the upward look and motion of eyelid. Consciousness continues to remain scatheless and therefore the patient is in a position to use eye blinking to speak clearly and intelligently. Patients with locked in syndrome is unerringly sealed within the human form, conscious of their surroundings but with a confined capacity to communicate with it<sup>1</sup>.

LIS was relabeled as quadriplegia and anarthria in 1986, with restoration of consciousness<sup>2</sup>. A brain stem lesion causes palsy of all four limbs, anarthria, and lower nerve damage leading to LIS<sup>3</sup>. Anarthria is caused by a bilateral paralysis of the laryngeal facio-glossopharyngo<sup>4</sup>. Other terms referring to the "locked-in" state are found in the literature."De-efferented state"<sup>5</sup> and "pseudocoma"<sup>6</sup> are synonyms for "locked-in syndrome." "Ventral pontine syndrome"<sup>7</sup> cerebro medullo spinal disconnection<sup>8</sup>, ventral pontine state<sup>9</sup>, "ventral brainstem syndrome"<sup>10</sup>

Early rehabilitation and more effective medical assistance can reduce mortality. A medically stable LIS patients' expectancy of life can be consequently increased, even though the patient is chronically and seriously physically impaired<sup>11</sup>.

## 2. CLASSIFICATION

There are three main categories of LIS:

2.1 Classic: Presence of vertical eye movement, palsy of fore limbs and hind limbs and anarthria with preserved cognizance.

2.2 Incomplete: Slight intentional movement other than eyelid and eye gestures.

2.3 Total: Total inactivity and ineptitude to convey messages or connect with full consciousness<sup>12</sup>.

## 3. ETIOLOGY

The most driving etiology of LIS is cerebrovascular malady (52%): Infarction of the ventral pontine following hindrance of basilar arteries is usually the result. Another significant and recurrent instigation of LIS<sup>13</sup> is brain injury (31%) due to trauma. The lesion was also confirmed to be the product of tumour, central pontine myelinolysis, misuse of drugs, pontine abscess, brainstem encephalitis, post infective polyneuropathy and air embolism.

**Table 1: Sources and mechanisms of locked-in syndrome**

SOURCE	MECHANISM
Ischaemic	An occlusion of basilar vessels, hypotensive or hypoxic events
Haemorrhage	Hemorrhage that originates inside or infiltrates into the pons
Traumatic	Direct cerebral contusion
Tumour	Infiltration of the ventral pons that is primary or secondary
Metabolic	Myelinolysis of central pontine
Demyelination	Multiple sclerosis to the ventral pons
Infectious	Abscess infiltrating the ventral pons, encephalitis in the brain stem

## 4. SIGNS ALONG WITH SYMPTOMS

Locked-in syndrome sometimes results from quadriplegia and also the inability to talk. Those with a locked-in condition interact with others by blinking or turning their eyes by coded messages that are not normally affected by the palsy. The symptoms of LIS can be compared to those of sleep paralysis. LIS patients are alert and aware about surroundings, without any disorientation of cognitive function. Proprioception is intermittently retained throughout their bodies. Movement of certain facial muscles are able in some patients. Some or all of the extraocular muscles can most often be relocated. Entities with the syndrome can't persevere a connection amidst breathing and speech. While the vocal cords are not paralyzed, this forbids them from making voluntary sounds.<sup>14</sup>

## 5. COMMUNICATION IN LOCKED IN SYNDROME

All the LIS people involved in this research were able to communicate; electronic transmission devices were used by more than 50% of patients, a yes-no code was expressed by remaining patients. The apportionment of patients who interacted using a yes-no code outlived to devaluate over time. A single criterion correlated with a markedly lower QoL was this last circumscribed communication mode. Conveying of messages for LIS persons involves using alternative broadcasting such as eye blinks or eye movements for a yes-no code or communication boards with alphabets or marks suggested by eye gestures. Electronic communication tools, as well as patient-CPU articulates such as infrared eye motion sensors and machine speech medical specialty, produce a releasing effect on individuals with LIS and allow them to have actual dialogs and use the web instead of responding obediently to others' summons<sup>15</sup>.

**Table 2: Types of modes of contact used in Locked-in syndrome**

Device	Use
Head mouse	Translates head gestures into the CPU mouse printer's direct proportional locomotives
Movement of the eye look	Tracks the position of the patient's attention to conduct
Infra red sensors	A virtual keyboard, switch or computer
EMG technology	Sensors of muscle fibres
Brain electrode automation	Invasive sensors to brain electrodes
EEG technology	Words translated by EEG activity

**6. 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 contains

6.1. Disorders of Consciousness. There are three DOC classifications including coma, unresponsive wakefulness, and state of limited cognizance. Alertness and knowledge both describe perception. Alertness is based on the normal functioning of the reticular nerves, thalamus, and cortex while awareness requires higher order processing which integrates both sensory data and motor information<sup>16</sup>.

6.2. Upper Cervical Spinal Cord Injury.

Because of the paralysis presentation an upper cervical SCI was enclosed. Both main muscles identified by the International Standards for Neurological Classifications of Spinal cord Injury Association (ISNCSI) within the standard motor test were rated at zero out of 5 on a bilateral basis, leading to the discernment that the injury should be above level C5 when the motor test starts<sup>17</sup>.

6.3. Akinetic Mutism. AM is a disorder represented by a reduced neurological drive with a decline in almost all motor functions along with face expression, movements and speech production, but with a point of alertness and intact optical tracking<sup>18</sup>.

**Table 3: Differential interpretation: acute quadriplegia and onset of mutism**

Diagnoses 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	✓

**7. TREATMENT**

Maintaining an airway and ensuring sufficient oxygenation is the prime consideration. Diminishing risk factors and reversible management of safety are important, thus avoiding immobility, dysphagia and incontinence complications. Pulmonary problems may be restricted by chest physiotherapy including deep breathing exercises, multiple changes in posture, postural drainage and suctioning. Lateral tarsorrhaphy or botulinum therapy can be used as treatment for corneal ulceration which is caused due to impaired eye closure. Communication will be prevented if full eye closure is done. Persistent crying can react to inhibitors of the selective reuptake of serotonin<sup>19</sup>.

that has the ability to manage patients with poor response. With high levels of misdiagnosis, limitations in the coverage of health care and financing of esteemed studies will make this difficult additionally undermining science<sup>20</sup>. Financial adjustment of estimated costs for future interest rates and inflation should also be considered in the future<sup>21</sup>.

**8. FUTURE AGENDAS**

To better define the advantages of low-level patients in-patient rehabilitation and health care providers continued research is need to recognize the importance of accurate diagnosis for the treatment of development plans, giving preference to a team approach

**9. CONCLUSION**

Health and recovery staffs working with LIS patients need to grasp the long-term result better. We must make ethical choices in the light of morbidity. Realistic goals in communications and mobility were provided by improvement in medical care and biomedical technology: these include computers, printers, and synthetic speech machines activated by responsive switches, electromyographic systems, and infrared eye-gape sensors. Insurers can be helped to prepare for the expense of treating these seriously impaired patients in the long term by a greater understanding of the outcome

for LIS patients. Steps in the formulation of a long-term plan include

- Determining the physical and cognitive disability spectrum and sequelae;
- estimating prognosis for recovery;
- Estimating the need for further medical and rehabilitative treatments and their benefits;
- Calculation of potential personal needs expenses (e.g. wheelchairs, orthopedic devices, home furnishings and upgrades, medical supplies and recreational facilities)

Overall the life expectancy has improved for LIS patients by factors like

- Effective recovery by professionals helps patients recover some control
- An effective communication network as an early goal should be set
- The involvement of the family in recovery that allows for the return home eventually
- Patients really don't want to die because of a debilitating illness

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