

Locked-in Syndrome: Causes and Treatment

[See online here](#)

Locked-in patients are not restricted in their higher neuronal functions when locked into a body that has become incapable of movement. While the former had to face miserable living circumstances and were moved to nursing homes, they can now be re-integrated into life with sophisticated therapies and techniques.



Definition of Locked-in Syndrome

Locked-in syndrome (pseudocoma) describes patients who are awake and conscious but selectively de-efferented. They cannot produce speech or limb movements. Acute ventral pontine lesions, such as hemorrhages or infarctions, are the most common cause.

People with such lesions remain comatose for some time requiring artificial respiration, and then gradually wake up but never return to the normal state of speech and limb movement. They are in a chronic vegetative state.

The corticospinal and bulbar tracts in the ventral part of the pons are interrupted. The disease is characterized by **spastic tetraplegia** and cerebral nerve disruption (loss of **pharynx**, **larynx**, chewing, and facial musculature) and, thus, the inability to speak. Horizontal eye movements are not possible.

On the other hand, consciousness, language comprehension, vertical eye movements, eyelid closure, and breathing regulation are preserved. **Sensitivity** and **algesia** (pain

perception) are usually normal.

Background of Locked-in Syndrome

Locked-in syndrome (LIS) is probably as old as humanity itself. Alexandre Dumas mentioned this phenomenon in his novel *The Count of Monte Cristo*. Therefore, it was often called the Monte Cristo syndrome. Clinically, the syndrome was first described in 1966 by Plum and Posner.

Etiology and Pathophysiology of Locked-in Syndrome

The clinical picture is often misjudged due to a bilateral lesion of the **ventral pons**, usually by occlusion of the **basilar artery**, or by a **central pontine myelinolysis**. Also, locked-in syndrome may be due to brainstem bleeding or contusions, tumors, or **brainstem nephroliths**. In amyotrophic lateral sclerosis (ALS), a state corresponding to locked-in syndrome can occur in the terminal stages.

The long tracts (**corticospinal** and **corticonuclear tract**), the abducens nucleus, and the **paramedian pontine reticular formation** (PPRF) are affected by the lesion. The rostral interstitial nucleus of the **medial longitudinal fasciculus** is responsible for the vertical eye movements, which are still possible.

Symptoms of Locked-in Syndrome

Initially, the patients are often in a coma, with **vigilance** rapidly improving in cases of **ischemia**. The awake, receptive, and cognitively intact patients can see and hear their environment. However, they demonstrate **spastic tetraplegia** and are unable to speak, swallow, or show facial expressions. **Horizontal view paresis** is present on both sides, and blinking and corneal reflexes are impaired.

Vertical eye movement and, in part, blinking of the eye are preserved, both of which can be used for communication.

Recurrent **features of LIS** are as follows:

- Tetraplegia/tetraparesis
- Paralysis of almost all motor cranial nerves
- Necessity of artificial respiration (acute phase)
- Alertness of the patient
- Perception of the environment

In order to clarify the last two points, **electrophysiologic methods** such as evoked potentials to somatosensory and auditory stimuli are useful (somatosensory evoked potential (SEP) and auditory evoked potential (AEP)).

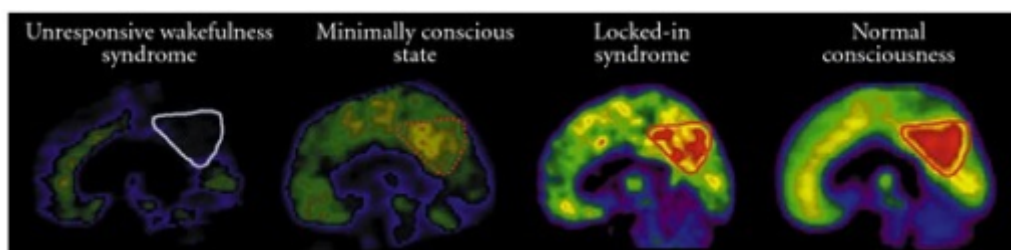


Image: Spontaneous brain activity in VS/UWS, MCS, locked-in syndrome, and health, as seen with PET. A triangle is drawn around the precuneus, an area whose spontaneous metabolic intensity is indicative of the level of consciousness. By Guldenmund, P., Stender, J., Heine, L., & Laureys, S. in *Crit Care Res Pract* (2012). License: [CC BY 3.0](#).

Differential Diagnosis of Locked-in Syndrome

Symptoms similar to LIS are present in the following conditions:

Guillain-Barré syndrome: An autoimmune reaction to peripheral nervous tissue is present. When motor nerve neurons are involved, similar symptoms to LIS can occur. However, additional involvement of the autonomic nervous system and, thus, respiratory difficulty and altered vital signs, are present.

Apallic syndrome (coma vigil, persistent vegetative state, so-called ‘wake coma’): Apallic patients are awake but do not consciously perceive their environment and themselves. Their eyes are open, but visual fixation is not possible, and cognition does not take place.

Akinetic mutism: There is no motor response to strong pain stimuli; however, caudal brainstem reflexes are preserved. The horizontal view function is intact.

The distinction is important to diagnose patients with locked-in syndrome. These patients can perceive their environment, feel pain, and can be questioned for treatment and therapy.

Therapy of Locked-in Syndrome

In addition to the causative treatment (e.g., **brainstem ischemia**), communication aids are used at an early stage (e.g., computer-assisted communication aids).

Acute hospital care

In the acute phase of a stroke, most patients are dependent on ventilation. The patients cannot swallow, so a **tracheostomy** is necessary to protect the patient from **aspiration**. If there is an improvement in the patient’s health, the patient can be weaned off ventilation.

Supportive management

Supportive management is the mainstay treatment method for patients with locked-in syndrome, and it entails the following:

- Preventing systemic complications of immobilization such as infections and thrombotic disorders
- Nutritional care

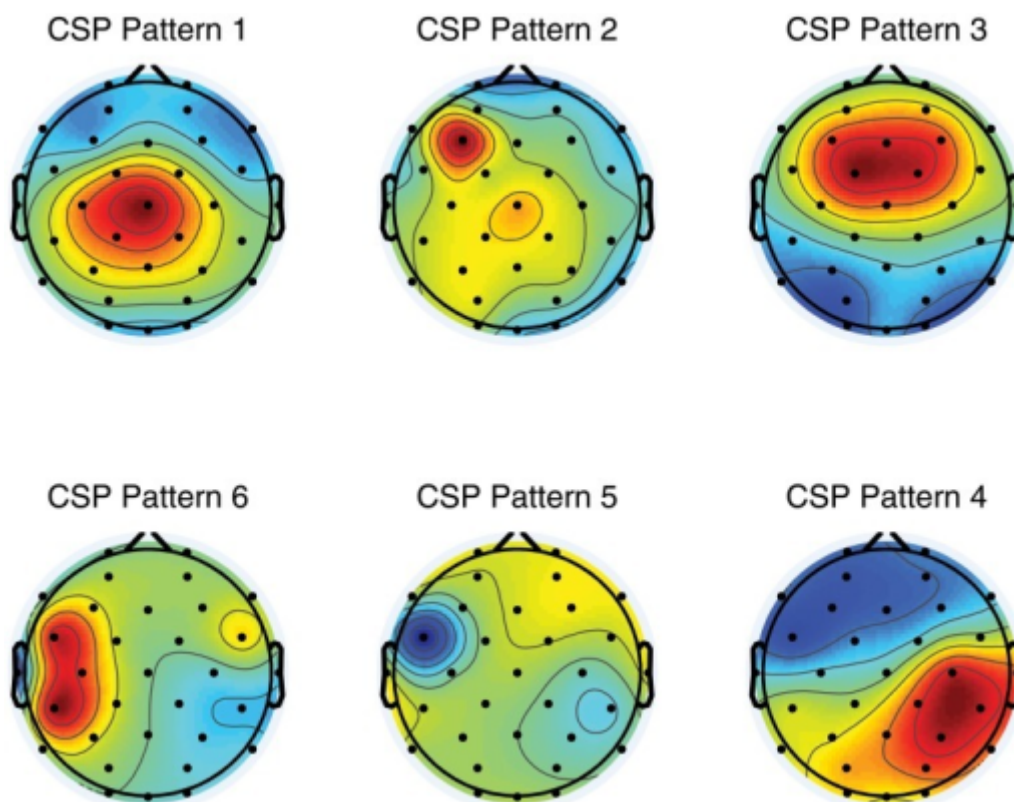
- Preventing pressure ulcers
- Physical therapy to avoid limb contractures

Ways to communicate with patients with locked-in syndrome

There is no definitive treatment for a locked-in state, and thus, a means of communication must be devised. The first step in dealing with a patient with locked-in syndrome is to find an effective communication code. In most cases, **eye movement is used as a code**: Upward movement means YES, and a downward movement means NO.

In the well-known case of *Elle* magazine editor Jean-Dominique Bauby, eyelid closure was used as a means of communication. Bauby was able to dictate his **book *The Diving Bell and the Butterfly*** using a series of letters that had been read to him. He winked at the letter he needed. Bauby himself described the opening and closing of the eyelid as a wing flap of a butterfly with which he could recapture the world.

Former creative director Georg Claus also experienced locked-in syndrome. He managed to move his head easily. He then invented a method by which he used glasses with a built-in laser pointer to point to letter and symbol panels. With this method, he created his own LIS Help website.



[Image](#): Improving the performance of an EEG-based motor imagery brain-computer interface using task-evoked changes in pupil diameter: Common Spatial Pattern Maps. The figure illustrates a set of common spatial pattern (CSP) filters of a single participant in the study. The CSPs are optimized for the discrimination of left-hand motor imagery from a control rest condition. By Rozado, D., Duenser, A., & Howell, B. License: [CC BY 4.0](#).

History and forecast

The mortality rate of patients with locked-in syndrome was still over 90% in the 1970s but has dropped to less than 50% due to advances in intensive care medicine.

Often, the phase of the LIS is a transitory stage (**transient locked-in syndrome**). Through persistent and intensive rehabilitation measures, good results can be achieved, and after several years of therapy, many affected individuals are again able to live independently.

However, there are also cases of chronic locked-in syndrome.

Emotional perspective

For a long time, LIS was considered severe. Those who were condemned to passivity could do nothing more than wait and watch what was going on. Accordingly, **Professor Werner Hacke**, Director of the Neurological Clinic Heidelberg, writes: "We, therefore, sedate patients...always deeply, in order not to let them experience the certainly tormenting inclination in their irreversible paralysis."

However, researchers at the University of Liège have further investigated the emotional state of patients and questioned them. What had previously been speculatively supposed to be "certainly tormenting" has not been confirmed:

Only 28% consider themselves unhappy. On the other hand, 72% of LIS patients consider themselves happy.

Further, questions concerned fears, pain, and suicidal tendencies.

According to this research group, the satisfaction of the patients depends on the quality of the care; on the other hand, the affected persons need to be familiarized with the new situation. The decision regarding euthanasia should not be made too early. Of 59 respondents, only 4 persons had wanted euthanasia.

References

Beineix, & J.-J. (1997). *Locked in syndrome*. London: BBC2.

Locked-in Syndrome. (n.d.). *SpringerReference*. doi:10.1007/springerreference_183269

Williams, O. (2010). *Stroke diaries: A guide for survivors and their families*. Oxford University Press, USA.

Young, G. B., & Wijdicks, E. F. (2008). *Disorders of consciousness*. Edinburgh: Elsevier.

Legal Note: Unless otherwise stated, all rights reserved by Lecturio GmbH. For further legal regulations see our [legal information page](#).