Locked-in Syndrome — Causes and Treatment

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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 the 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, as well as breathing regulation, are preserved. Sensitivity and algesia (pain perception) are usually normal.

**Background of the 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 referred to as the Monte Cristo syndrome. Clinically, the syndrome was first described in 1966 by Plum and Posner.

**Etiology and Pathophysiology of the 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, the locked-in syndrome may be due to brain stem bleeding or contusions, tumors, or brain stem nephroliths. In the context of amyotrophic lateral sclerosis (ALS), a state corresponding to the 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 the Locked-in Syndrome**

Initially, the patients are frequently in a coma, with vigilance rapidly improving in the case of ischemia. The awake, receptive, and cognitively not impaired 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, which can be used for communication.

Recurrent features of LIS are:

- 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 2 points, electrophysiological methods such as evoked potentials to somatosensory and auditory stimuli are useful (somatosensory evoked potential SEP and auditory evoked potential AEP).
Differential diagnosis of the Locked-in Syndrome

Symptoms like LIS are present in the following conditions:

**Guillain-Barré syndrome:** 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 brain stem reflexes are preserved. The horizontal view function is intact.

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

Therapy of the Locked-in Syndrome

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

**Acute hospital care**

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

**Supportive management**

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

- 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 locked-in patients

There is no definitive treatment for a locked-in state, and thus, a way of communication must be devised. The first step in dealing with a locked-in patient is to find a well-functioning code to communicate. In most cases, an **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, the lid closure was used as a communication path. 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 suffered 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 patterns (CSPs) 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](image)

History and forecast

The mortality rate of patients with locked-in syndrome was still over 90% in the 70s but has dropped to less than 50% due to advances in intensive care medicine.

Often, the phase of the ‘Locked-in syndrome’ is a transitory stage (**transient locked-in syndrome**). Through persistent and intensive rehabilitation measures, good results can be achieved, and by means of many 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, the condition of LIS was considered a severe one. Those who were condemned to passivity could do nothing more than wait and watch what was going on with them. 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 agonizing' 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 on euthanasia should not be taken too early. Of 59 respondents, only 4 persons had wanted euthanasia.

**References**


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