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 the Locked-in Syndrome

Locked-in syndrome (pseudo coma) describes patients who are awake and conscious but selectively deafferented. They have no means of producing 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 like patients in a chronic vegetative state.

The state interrupts corticospinal and bulbar tracts in the ventral part of the pons. Spastic tetraplegia, cerebral nerve disruption (loss of pharynx, larynx, chewing and facial musculature) and thus also the inability to speak. Horizontal eye movements
are not possible.

On the other hand, consciousness and language comprehension, vertical eye movements and 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 syndromes (LISs) are probably as old as humanity itself. Already, Alexandre Dumas mentions this phenomenon in his book “The Count of Monte Christo”; therefore, it was often referred to as Monte-Christo syndrome. Clinically, the syndrome was first described in 1966 by Plum and Posner.

Etiology and pathophysiology of the Locked-in Syndrome

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

Affected by the lesion are the long tracts (tractus corticospinalis and corticonuclearis), the abducens nucleus and the paramedian pontine formation reticularis (PPRF), while, in the rostral interstitial nucleus of the fasciculus longitudinalis medialis, occurring vertical eye movements are still possible.

Symptoms of the Locked-in Syndrome

How the Locked-in syndrome manifests itself

Initially, the patients are frequently in a coma, with the vigilance rapidly improving in the case of ischemia. The awake, receptive and cognitively not impaired patient can see and hear his environment. However, he shows spastic tetraplegia, is unable to speak, swallow, or show facial expressions. There is a horizontal view paresis on both sides, blinking and corneal reflex is canceled.

On the extremities, there are strecksynergysms, which can be triggered by the frequently obtained sensitivity.

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

Recurrent features of locked-in syndromes are:

- Tetraplegia/tetraparalysis
- Paralysis of almost all motor cranial nerves
- The necessity of artificial respiration (acute phase)
- The alertness of the patient
- Perception of the environment

In order to clarify the last two points, electrophysiological methods, such as evoked
potentials to somatosensible and acoustic stimuli, can help out (SEPs and AEPs).

**Differential diagnoses of the Locked-in Syndrome**

**Similar symptoms to Locked-in Syndrome**

**Guillain-Barré syndrome:** Autoimmune reaction to peripheral nervous tissue; when motor nerve neurons are involved, similar symptoms can occur. However, there is additional involvement of the autonomic nervous system and thus respiratory difficulty and altered vital signs.

**Apallic syndrome:** (Syn. Coma vigile, persistent vegetative status, so-called “wake coma”). The apallic patient is awake, but does not consciously perceive his environment and himself. The eyes are open, but no visual fixation is possible, no cognition takes place.

**Akinetic Mutism:** There is no motor response to strong pain stimuli; caudal brain stem reflexes are preserved. The horizontal view function is intact.

The distinction is important in order to meet the locked-In patient, who clearly perceives his environment, is able to feel pain, and can be questioned for treatment and therapy.

**Therapy of the 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).

**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 an improvement in the state of health should take place, weaning of the ventilation can finally take place.

**Supportive management**

This is the mainstay method of management for patients with locked-in syndrome and it entails:

- Preventing systemic complications of immobilization such as infections and
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 coding in order to communicate. In most cases, an **eye movement is used as a code**. A movement upwards means YES, down NO.

In the well-known case of “Elle” editor Jean-Dominique Bauby, the lid closure was used as a communication path. Bauby was able to dictate his book *Butterfly and Diver Bell* by means of a series of letters that have 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 a lock-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 has 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 – PLoS ONE (2015). License: [CC BY 4.0](https://creativecommons.org/licenses/by/4.0/).
History and forecast

The mortality rate of patients with locked-in syndrome was still over 90% in the 1970s but has fallen 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 here, 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.

An emotional perspective

For a long time, the condition of a LIS was considered to be 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 have questioned the LIS sufferers themselves. 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 considered 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. A decision on euthanasia should not be taken too early. Of 59 respondents, only four persons had wanted euthanasia.

References


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