International Journal of Progressive Sciences and Technologies (IJPSAT)
ISSN: 2509-0119.

https://ijpsat.org/
Vol. 49 No. 2 March 2025, pp. 593-604

Trapped But Aware: Understanding Locked-In Syndrome And Its Treatments

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Abstract: Locked-In Syndrome (LIS) is a rare neurological disorder with quadriplegia and anarthria accompanied by the preservation of cognitive function and vertical eye movement. It results from injury to the ventral pons, most commonly by stroke, trauma, or demyelinating disease. LIS is very challenging to diagnose, and advanced neuroimaging techniques like fMRI, EEG, and CT scanning are required to differentiate it from coma and vegetative states. The quality of life is relatively good for most LIS patients, especially with regard to the availability of assistive communication devices and psychological counseling, despite the motor disabilities being catastrophic. The management of LIS also includes neurorehabilitation, which consists of physical therapy aimed at avoiding complications such as muscle atrophy and deep venous thrombosis, combined with mental fortitude, communication, and cognitive enhancement therapies. Emerging technologies, including brain-computer interfaces and neural implants, show immense potential for boosting autonomy. Ethical considerations involving patient autonomy and end-of-life decisions are still complex, with the majority of LIS patients initially expressing a wish for hastened death but later accepting their situation. Advances in neuroimaging, electrophysiology, assistive technologies, and multidisciplinary rehabilitation have significantly improved the outcomes of LIS patients. Societal myths and ethical issues continue to impact clinical care. Individualized rehabilitation strategies and further incorporation of neuroprosthetic solutions are areas where future research must be focused to encourage patient independence and quality of life.

Keywords: Locked-in Syndrome, quadriplegia, neuroimaging, electrophysiology, brain-computer interface, quality of life, neurorehabilitation, patient autonomy, assistive technology, ethical considerations.

1. INTRODUCTION

Locked-in syndrome (LiS) is a rare yet profoundly disabling neurological impairment, characterized by quadriplegia and anarthria with preserved vertical eye and eyelid movements and retained cognitive abilities. The term locked-in syndrome (LIS) was coined in 1966 by Plum and Posner in their classic text "Diagnosis of Stupor and Coma"[5] .locked-in syndrome is caused by an insult to the ventral pons and caudal midbrain by infarct, hemorrhage, trauma and demyelinating disorders.

The mean age of onset for all cases of the locked-in syndrome has generally ranged from 30-50 years ,with a slight male predominance.

Locked-in syndrome has three main types. The classical form is defined by Quadriplegia and anarthria with preserved consciousness and the ability to perform vertical eye movements, blink . Incomplete is same as classic but with remnants of voluntary movement other than vertical eye movement. Total is characterized by complete immobility ,loss of eye movement and inability to communicate, with preserved consciousness.

The pathophysiology of locked in syndrome is attributed to the damage of the corticospinal and corticobulbar tracts which run along the ventral part of the pons and the medulla oblongata and will subsequently be affected by a ventral brainstem lesion.

Damage to the corticospinal tract impairs the voluntary motor movements resulting in quadriplegia whereas damage to the corticobulbar tract compromise the voluntary, non-ocular movements of muscles innervated by the caudal cranial nerves. Anarthria is due to bilateral facio-glosso- pharyngo-laryngeal paralysis, and damage of the corticobulbar fibers which also causes dysphagia and limits the use of facial expression in communication. The vertical gaze is usually unaffected because of the nucleus' location in the midbrain's rostral portion. The reticular activating system, which lies dorsally and responsible for wakefulness is spared by ventral lesions.

The pontine respiratory group located in the pons tegmentum could be affected in large lesions leading to signs of apnea and respiratory distress. The leading cause of Death in patients with LIS is atelectasis and pneumonia due to aspiration and impaired cough reflex. Depression is another long-term complication of patients with LIS ,hence psychological and psychiatric support and treatment is essential. Immobility also predisposes to pulmonary embolism therefore, chest physiotherapy like deep breathing exercises, frequent position changes, postural drainage and suctioning is recommended in LIS patients.

Patients do not recover completely , therefore Multidisciplinary rehabilitation is the cornerstone of the rehabilitation of LIS. The reviewed studies indicate a positive trend of effect of exercise on physical recovery of people with LIS after stroke including the improvement of muscle strength, tone, walking ability, and activity in daily living

2. OBJECTIVE

The purpose of this study is to detect pathophysiology, diagnosis and management of lock-in syndrome (LIS). It wants to analyze the role of neuroimaging and electrophysiological devices in the diagnosis of LIS, assesses the impact of LIS on the quality of life and psychological welfare of LIS, and examines the current neurorehabilitation strategies. Additionally, it reviews moral ideas about the patient autonomy and the decisions of the end of life, highlighting progress in subsidiary technologies that improve communication and daily functioning for LIS patients.

3. MATERIALS AND METHODS

35 articles have been analyzed for the literature review. Keywords such as Locked-in Syndrome, quadriplegia, neuroimaging, electrophysiology, brain-computer interface, quality of life, neurorehabilitation, patient autonomy, assistive technology, ethical considerations were used for the literature searches in databases such as Google Scholar and PubMed.

4. RESULTS

Literature review confirms that LIS is mainly caused by damage to ventral pons, which causes the fourth and unseen, conserving cognitive function. Neuroimaging techniques such as FMRI, EEG and CT scans are important for accurate diagnosis and discrimination from conditions such as coma or vegetable state. Despite severe physical disadvantages, many LIS patients report satisfactory quality of patients, especially with effective communication strategies and psychological support. Neurorehabilitation, including physical therapy, cognitive training and support communication technologies such as brain-computer interfaces, improves the patient's freedom significantly. The patients are complex about the decisions of the patient autonomy and the end of life, the studies show that the initial crisis often gives way to adaptation and improvement over time. Progress in rehabilitation and assistant technologies continues to increase results for LIS patients.

5. DISCUSSION

5.1 Neuroimaging and Electrophysiological Tools for Evaluation of LIS

Neuroimaging observes the structural and functional changes of the brain, becoming revolutionary for the diagnosis of locked-in syndrome. It can be used to see how the brain responds to treatment and identify potential biomarkers for neurological disorders. With the advancement of technology over the years, the quality of data obtained for research has greatly increased, providing more accurate diagnoses.

- **Role of fMRI and EEG as Electrophysiological Tools**

  Commonly used electrophysiologic techniques are functional magnetic resonance imaging (fMRI) and electroencephalography (EEG). Both methods are non-invasive making them advantageous and allow for repeated usage.

    - fMRI produces images of the brain using magnetic fields and radio waves. Innovations through technology have allowed increased resolutions with a more comprehensive understanding of the brain. Patients with locked-in syndrome have distinct patterns of brain activity and engage in cognitive processes compared to those with unconscious states.
    - EEG monitors neuronal activity and helps to reduce misdiagnosis of locked-in syndrome with other conditions such as coma. It can distinguish between patients who are conscious and unconscious whereas in Locked-in syndrome, consciousness is preserved. EEG findings appear either normal or slow in patients with locked-in syndrome. Physical examination must be done along with EEG to monitor whether the symptoms progress or regress over time.

- **Additional techniques for evaluation of Locked-in syndrome**
    - Other essential tools are MRI and CT which can help show extensive lesions in the brainstem as in locked-in syndrome the ventral portion of the lower pons and medulla are affected.
    - MRI uses diffusion-weight images and is undoubtedly the best when it comes to visualizing the brainstem, and if contraindicated or if hemorrhage is suspected, a CT scan is performed. Both CT and MRI visualize changes that occur in the brainstem as it is important for many functions of the body which are often affected in locked-in syndrome.
    - Additionally, photic stimulation can be done. This involves using light as a stimulus to observe brain activity. In Locked-in syndrome, photic stimulation would show a brain response that could occur at any time to the patient during the process.

- **A further dive into the evaluation of Locked-in syndrome using clinical approaches**
    - Assessment of locked-in syndrome can be diagnosed based on the symptoms presented. Examining the history of the patient from family members can help clinicians to identify how the symptoms have changed or progressed over time and to differentiate it from other similar diagnoses.

- **Identifying overlaps**
    - Spinal cord injury can also be a possible cause of motor paralysis. However, only motor and sensory functions below the level of injury are affected.
    - Akinetic mutism is characterized by decreased motor functions with preserved reflexes in the brainstem. Lesions are present in the frontal lobe and this is responsible for voluntary movements and speech. It can be differentiated on MRI as the lesion sites for both locked-in syndrome and akinetic mutism are different.

    - In Guillain-Barré Syndrome, immobility of the extremities are present and could involve the cranial nerves. Differentiating factors include a negative Babinsky's reflex and areflexia in Guillain-Barré Syndrome.
    - Persistent vegetative state, just like locked-in syndrome has absent motor movements. It can be distinguished by the lack of awareness of the environment or interactions which is characteristic of a vegetative state.

5.2 Impact of LIS on Quality of Life and Psychological Well-being

Locked-in syndrome as many people describe it is "conscious but paralyzed and voiceless". So how does one adjust and ensure a good quality of life? Communication is key! Adapting strategies focusing primarily on communication such as seeking out is greatly beneficial. As consciousness is intact and eye movements are present, one blink for yes and two blinks for no has been established as one of the methods of communication. Having strong emotional support from family and friends would make psychological adjustment easier, proving to be one of the greatest impacts on quality of life. Most of the time, family members notice that the patient is more aware than physicians do. Despite motor paralysis, many patients with locked-in syndrome do lead a satisfactory life.

- **Assessment of Quality of Life in Locked-in Syndrome Using Questionnaires**

  Standardized questionnaires including their emotional and physiologic adjustments were used to measure the quality of life. Questionnaires were answered by blink responses or by a visual keyboard.

    - Assessment questionnaires such as Anamnestic Comparative Self-Assessment (ACSA) was used on a scale of -5 being the least to +5 the most favourable. A score above 0 indicates a satisfactory quality of life.
    - The Schedule for the Evaluation of Individual Quality of Life-Direct Weighting (SEIQoL-DW) was used to assess different fields that are important to the patient and their overall satisfaction of each field, with a scale of 100% and 50 percent being the minimum satisfaction.
    - ALS Depression Inventory-12 (ADI-12) was used and assessment was based on a scale of 1 (fully agree) to 4 (fully disagree). A score above 28 would indicate clinical depression. Therapy for depression or anxiety should be part of health care and must be accessible if needed.
    - Very few number of patients wished for a haste death. They had to answer 20 questions binarily by choosing either correct or wrong and were assessed by Schedule of Attitudes toward Hastened Death (SAHD).

- **Adapting despite restrictions: The well-being paradox**

  Although physical limitations, patients with Locked-in syndrome found a way to adapt known as the "well-being paradox". A person's physical state doesn't always mirror their state of mind. Psychological condition is merely independent and is more than capable of getting better despite physical well-being. The longer locked-in syndrome persists the more psychological well-being improves. This correlation conveyed that patients with depression and anxiety symptoms subside over time.

    - **Enhancing the Quality of Life through Healthcare**
        - Rehabilitation and involvement of a multidisciplinary team consisting of a nurse, physiotherapist, and communication team would greatly improve living conditions. Most of the reported concerns from patients are inadequate rehabilitation, improper treatment, and lack of support from close ones.
        - Effective relationship between the doctor and patient is vital for communication especially for listening to what the patient needs, respecting their choices in treatment, and refraining from precedence.

- ○ Primary factors leading to unsatisfactory doctor-to-patient relationships are affiliated with improper communication. This includes not respecting the patient's freedom of choice, lack of empathy, and poor follow-up.

5.3 Neurorehabilitation Approaches: Physical and Cognitive Therapy

The main focus of Neurorehabilitation for Locked-In Syndrome (LIS) patients is to maximize their independence and quality of life and help strengthening their communication through motor and central processing activities. Unfortunately, spontaneous treatment and recovery in LIS patients is minimal and nearly impossible, but it is possible to enhance neuroplasticity and advance existing motor control through targeted rehabilitation programs. Managing LIS patients requires a multidisciplinary team of therapists that includes, a physiotherapist, occupational therapist, and cognitive trainer.

Physical Therapy: Enhancing Motor Function and Preventing Complications

Despite disabling motor impairments, a well planned physiotherapy strategy plays an important role in LIS patients rehabilitation, where passive and active-assisted exercises help in preventing joint contractures, the atrophy of muscles, and secondary complications such as deep venous thrombosis (DVT) and pressure ulcers.

The structured physiotherapy strategies include:

- Passive Range of Motion (ROM) Exercises: It maintains the musculoskeletal integrity and prevent contractures.
- Functional Electrical Stimulation (FES): It helps in improving motor responses and stimulating paralyzed muscles to prevent muscle wasting.
- Task-Specific Training: Enhances neuroplasticity by reinforcing residual movements, generally with the utilization of exoskeletons and robotics.

Cognitive Therapy: Supporting Mental Resilience and Neuroplasticity

Cognitive rehabilitation is meant to enhance mental processing, emotional state, and communication abilities among LIS patients.

This involves:

- Brain-Computer Interface (BCI) Training: Enables direct neural control of assistive devices to support patient independence.
- Virtual Reality (VR) and Motor Imagery Training: Utilizes neurofeedback pathways to enhance motor mechanisms and promote neural plasticity.
- Psychological Support and Neurocognitive Exercises: These trainings treats depression, anxiety, and cognitive activation, which are essential for long-term recovery.

Integrating Rehabilitation with Assistive Communication Technologies

A coordinated neurorehabilitation approach should be complemented with assistive communication technologies, such as eye-trackers and speech-adaptive speech synthesizers, to encourage independence and social engagement (Hochberg et al., 2012) Therapies that emerged recently, such as neural implants and brain-machine interfaces, present promising areas to improve motor recovery and voluntary motor control restoration.

Physical and cognitive rehabilitation strategies play a critical and crucial role to control LIS through the promotion of both physiologic recovery and psychosocial adjustment. Although there are significant advances have been made, continued investigation into personalized rehabilitation regimens and novel neurotechnology is required to enhance patient outcomes and long-term quality of life.

5.4 Multidisciplinary Management of Locked-In Syndrome: Acute Care, Rehabilitation, and Communication Strategies

In a disease with as many detrimental effects as in Locked-in Syndrome, a poor prognosis is inevitable. However, although patients with LIS have a marked decrease in quality of life and are heavily dependent on caregivers and family for even simple daily tasks, advancement in technologies has made possible the implementation of Brain Computer Interfaces (BCIs) and neural implants that assist with communication and speech therapy.

Treatment of Locked-In Syndrome is broadly classified into two main categories: immediate and long-term management. The current acceptable golden-time of assistance is within one month of onset of morbid symptoms - if identified and started on correctional therapy, these patients have improved functional outcomes and significantly reduced mortality rates. Studies suggest that if the patient survives the first year of onset of symptoms (where 86% of deaths occur in first four-five months), five-year survival of these patients can increase up to 87%, and survival after ten years of onset of LIS is deemed to be 80%.

Pharmacological and non-invasive management of Locked-In Syndrome:

Locked-In Syndrome requires optimal management of time and an in-depth understanding of possible afflictions to provide emergency and immediate care. Acute stabilization and treatment of the underlying causes, such as pulmonary complications like atelectasis and pneumonia due to aspiration and impaired cough (the most common cause of death in LIS patients) is essential. A critical component of acute care and first line management of LIS patients includes securing the airways and providing sufficient oxygenation. Chest physiotherapy like deep breathing exercises, frequent position changes, postural drainage and suctioning is recommended. In patients with onset of LIs after ischemic stroke of arteries, re-canalization through intravenous or intra-arterial means within 48 hours of onset of symptoms is essential for survival.

Another documented manifestation of LIS is PCL - pathological crying and laughter. This unprovoked emotional volatility, that is isolated and not in conjunction with any known mood disorders in the patients, has been attributed to and is a known effect of direct injury to the pontine brainstem. It results in lapses in initial attention span, communication ability and cognition. Although most patients do improve overtime, studies have reported diminishing or altered mental states, with weighty impairments like memory loss in about 18% of patients. This perplexing condition is also hypothesized to arise from insult to the pinto-cerebellar pathway, which is linked to emotional behavior to contextual stimulation. The use of antidepressants has little to no effect on PCL, and it must be maintained through use of cognitive-behavior approach. In line with the revised emotional states, hallucinations and depression are other less noted and often overlooked manifestations of LIS. If present in patients, it requires adequate management in the form of psychological/psychiatric support. Offering coping strategies to help patients better adapt to their new realities and surroundings or repeatedly informing them about the illusory nature of their perspectives, helps improve cognitive states.

Patients with LIS also greatly benefit from ophthalmological examinations, since visual deficits are common. Appropriate treatment to improve impaired visual function can help with comfort and communication. Patients suffering from opsoclonus-myoclonus syndrome (a condition in which the eyes seemingly randomly and involuntarily move rapidly in the horizontal, vertical, and diagonal directions), convenience from the use of gabapentin, which works by either enhancing pause neurons or by hindering the burst ones in the saccadic circuit. It yields rapid and long-lasting resolution of this syndrome, which is crucial, especially in patients relying on eye communication.

Extremity training and motor function improvement is one of the most challenging aspects of LIS. In patients with incomplete LIS, use of limb robotics, such as the Armeo®Spring program (Hocoma AG, Volketswil, Switzerland), has shown efficacy in increasing accuracy. Such occupational therapy, in combination with technological progression, has aided in enhancing grip strength and active ROM. Managements like TT (treadmill therapy) and repetitive sensor motor training beat the allegations of futility associated with Locked-In Syndrome, facilitating adverse effect-free physical reform and postural control. With the visual feedback provided by extremity neuroprosthesis, such as Bioness H200, functional recovery is now a realizable objective. In addition, intrathecal baclofen is now regularly used in settings of LIS, to treat muscle spasticity and improve motor recovery. Supplemental therapies thought to benefit LIS patients also include spinal cord stimulation, which can not only revive residual motor function and neuro-plasticity, especially in patients in whom onset of LIS is secondary to brainstem strokes, but also improve autonomic functions like bowel and bladder control, possibly affording a better quality of life to LIS patients. Treatment of LIS also depends heavily on the underlying causes of it - from central pontine myelinolysis, to Guillain-Barre syndrome or ALS, management varies tremendously and to varying degrees. While the initial care administered is essential similar in all cases, the following intensive measures to eradicate particular causes can be helpful:

- gradual correction of sodium abnormalities and close monitoring of respiratory status for central pontine myelinolysis LIS
- IVIG, plasmapheresis, and monitored airway ventilation for Guillain-Barre syndrome
- supportive measures in conjunction with long-term agents riluzole or edaravone to prolong survival in patients with ALS
- Meningitis and abscesses may be treated with broad-spectrum antibiotics

Communication in patients with Locked-In Syndrome

Once initial evaluation of the patient is complete, it is necessary to establish level of consciousness and understanding, and ability to follow commands and reliability of their yes/no responses. Communication assistance ranges from no-tech involvement to high-tech involvement in augmentative and alternative communication (AAC), entailing dependency on patient's eye coordination for communication to instalment of neural implants or brain-computer interface usage (to be discussed in the upcoming sub-topics).

- No-tech AAC relies on patients eye movements, body and/or sign language, extremity movement on command or facial gestures to establish effective communication.
- Low-tech AAC enlists the assistance of materials like pen-paper, letter boards or single message voice output devices to engage the patient in meaningful conversation. In patients with preserved ocular function, ETRAN Board can be used to substantiate communication.
- High-tech AAC involves the use of implanted devices and/or BCIs to encourage communication through utilization of neural signals in conjunction with eye-gaze devices and ocular movements. (Fig. 1)

Use of Brain-Computer Interfaces in Locked-In Syndrome:

BCIs exploit neural activity as opposed to muscular activity to aid with functional communication. Once raw neural signals have been isolated, BCIs identify and decode these signals to orchestrate coherent messages. These neural signals can be obtained via invasive and non-invasive means, as explored below.

A. Non-Invasive BCIs:

Non-invasive BCIs aim to track neural activity without the use of invasive procedures like implantations or surgery. They record brain function externally, mainly through electrodes placed on the scalp or sensors detecting brain signals. Common types of non-invasive BCIs include:

- Electroencephalography (EEG) - electrodes on scalp to measure brainwaves
- Magnetoencephalography (MEG) - detects magnetic fields produced by the brain
- Functional Near-Infrared Spectroscopy (fNIRS) - provides data by detecting changes in blood oxygen levels
- Functional Magnetic Resonance Imaging (fMRI) BCIs - records brain function and activity through changes in blood flow.

B. Invasive BCIs:

With the distinctive retainment of cognitive activity in patients with LIS, invasive BCIs present the opportunity for advancement in quality of life of these patients. These provide innovative solutions to bridge the gap between body and mind. Invasive BCIs employ similar practices as non-invasive means, only implanting these devices directly into the brain offers higher precision and direct links between brain signals and devices. Although they offer superior signal resolution and faster communication, the surgical nature of these BCIs makes them a riskier alternative.

- Intracortical BCIs - surgical implantation of electrodes directly into the cerebral cortex of the brain. Provides an unparalleled level of spatial and temporal resolution. This granular level of control opens up avenues for more sophisticated forms of communication, including intricate speech generation and the dexterous operation of robotic prosthetics closely mirroring natural limb movements. Modern day examples of this include Utah Array, Elon Musk’s Neuralink.
- Electrocorticography (ECoG) BCIs - electrodes placed on the brain’s surface - under the skull but above the cortex. Assists in motion function restoration.
- Deep Brain Stimulation (DBS) BCIs - used primarily to modulate neural activity rather than record it.

In summary, treatment and management of LIS patients is a multifaceted approach, and no one aspect of it can be compromised or prioritized over the other (except in acute cases that necessitate vitals management). In addition to all above listed means, researchers have also demonstrated the efficacy of physiotherapy and psychotherapy in long-term management of LIS. In their study on pain perception in patients with LIS, French researchers commented - ‘…a decrease in pain sensitivity was associated with decreases in brain activity in brain regions involved in emotional processing and executive functions, in conjunction with increases of brain activity in regions involved in pain processing’. To wield to the benefit of patients these complimentary effects of fluctuation in pain with perceptions of it; studies suggest the use of psychological maneuvers like hypnosis in pain management.

In theory, choosing such non-pharmacological means supplies patients with comfort, and leaves them better suited to employ all available communication tools without the undeniable side effects of heavy analgesics.

5.5 Ethical and Societal Implications in the Care of LIS Patients

Patient Autonomy and choice in LIS

With the treatment of LIS, one of the most difficult issues that can be faced is that of patient autonomy. Patients have very severe motor disabilities that prevent them from interacting with the outside world. They cannot speak or move freely, but they have cognitive ability. So this poses the question, how much autonomy do they actually have? Considering that they often lack the ability to express their desires directly. However, patient autonomy is based on the informed consent principle. Autonomy may be regained through some degree of communication through alternative methods. For example, eye tracking or augmentative and alternative communication technologies. Even if the patient is unable to speak for themselves, advanced directives or wills, if available, become important in ensuring that their wishes are respected.

End of Life ethics in LIS

One of the most morally challenging aspects in LIS are end-of-life decisions. Many people question whether life-sustaining procedures are necessary, considering that many LIS patients endure long term suffering. Their argument is that keeping them on life support, especially when the disease is irreversible only makes their suffering worse. This brings up the topics of assisted suicide and euthanasia. According to studies, initially, LIS patients indicate that they want an early death. However, as the time progresses and they get used to their condition, this desire lessens. Choosing between honoring a patient’s initial desires for assisted suicide or euthanasia or to give them time to adjust to their condition psychologically presents an ethical dillemma. Suggesting a moratorium on life-ending decisions might make sense considering that it takes time for patients to adapt to their new situation. This could lead to less premature decisions that happen during the adaptive phase. Additionally, there are cultural and legal implications that can affect these decisions. For example, in eastern cultures, it is shown that there is a tendency for more support towards life support rather than euthanasia. Healthcare professionals must consider all these variables while also honoring the country's legal systems and culture when making end-of-life decisions.

Quality of Life in LIS

A crucial factor in the treatment of LIS patients is quality of life (QoL). According to studies, many LIS patients report a quality of life that is comparable to the average population, despite their condition. However, it is seen that many caregivers and medical professionals believe that their QoL is far worse. This disparity in perception is explained by the “response shift” phenomenon. The “response shift” phenomenon is when individuals with long-term illnesses adjust to the difficulties of their condition overtime by readjusting their expectations. Some of the variables that affect the psychological wellbeing of LIS patients are length of illness, communication skills, and psychological adaptation over time. Long term LIS patients are more often adapted and their quality of life is seen to be not as compromised. A higher quality of life is seen in patients who regain some sort of communication capability. This could be done by partial speech recovery or AAC devices. However, patients who rely on nonverbal cues could encounter difficulties that have a detrimental impact on their emotional health. Because of these issues, it is important that QoL is evaluated in terms of mental health, communication skills and the patients long term psychological adaptation to their condition.

Societal Perception of LIS

There are many misconceptions that shape and affect the public’s knowledge of LIS. In contrast to the fact that many LIS patients do retain cognitive function, the general public believe that these patients are completely unconscious or unaware of their environment. This misconception may be increased by media representations of LIS, which only show the most severe cases. This feeds public false beliefs about the illness. The way that the public perceives LIS patients can greatly impact the treatment of these patients. For example, some healthcare providers may underestimate the psychological health of LOS patients because they already have a preconceived notion that their cognitive abilities are severely compromised. This can result in devaluation of patient autonomy and reduced quality of life. An important topic this brings up is end-of-life discussions. If the general public's perception of LIS patients is that they have reduced cognitive ability, caregivers may be more inclined to administer euthanasia when in reality it may be against the patient's wishes. To combat this, there needs to be more public awareness on the reality of these LIS patients and their cognitive skills.

The Ethics of Decision-Making in Early-Stage LIS

Many LIS patients initially experience psychological anguish, including suicidal ideation. Due to this, the decision making process in the early phases of LIS is very delicate. As they struggle with the abrupt start of their illness and the loss of motor function, many patients yearn for death during the initial stages. Regardless of this, the possibility of psychological adaptation must be considered. Care should be taken while making these initial decisions and allowing for a moratorium on decisions to end a patient's life should be considered until they have time to adjust. Transparent communication, accessible resources, social support care and different communication tools such as AAC devices can help aid in this process. Healthcare professionals should guarantee caregivers and family that decisions on a patient's life are founded on well informed and thoughtful preferences by allowing the patients enough time to adapt to their condition.

6. CONCLUSION

Locked in syndrome (LIS) is a rare and severe condition that causes quadriplegia together with anarthria while retaining both patient consciousness and cognitive skills. Despite the motor impairment, the patient is fully conscious and this presents unique challenges to medical professionals. The development of neuroimaging methods has enabled physicians to diagnose LIS and more importantly distinguish it from a coma or vegetative state diagnosis. Research shows that LIS patients experience initial emotional turmoil but ultimately end up in a relatively satisfactory mental condition. Neurorehabilitation combined with physical therapy and pharmacotherapy alongside non-invasive therapy methods enable patients to achieve maximum independence and quality of life. With the help of communication tools like AAC’s and BCI’s, patients with LIS can reconnect with their loved ones and express their needs. This brings a sense of normalcy to their lives and greatly improves their mental state. Ethical dialogues about LIS should include thoughtful evaluation and discussion of patient autonomy, end-of-life discussions and quality of life assessments. Patient autonomy should be guaranteed for patients with LIS as they do have cognitive ability and a means to communicate. End of life discussions should be done with the individual's mental state in mind. Considering the psychological adaptation overtime, there should be a waiting period before making any major decisions. Ultimately, LIS is not a death sentence, and it shouldn't be treated as such. With the many advancements and ongoing research, it is a certainty that patients with LIS can live and maintain a satisfactory life with fulfillment and dignity.

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DISCLOSURE

Acknowledgment

We thank the authors for their contribution to this project.

Ethical approval

Ethical approval was not required for this study

Declaration of patient consent

Patient’s consent was not required as there are no patients in this study.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.