Palliative Care Rounds

Locked-In Syndrome: Case Report and Discussion of Decisional Capacity

Samuel Maiser, MD, Ashish Kabir, MD, David Sabsevitz, PhD, and Wendy Peltier, MD

Palliative Care Center (S.M., W.P.) and Department of Neurology (A.K., D.S., W.P.), Medical College of Wisconsin, Milwaukee, Wisconsin; and Departments of Neurology and Medicine (S.M.), Hennepin County Medical Center, Minneapolis, Minnesota, USA

Abstract

Locked-in syndrome (LIS) is a rare neurologic disorder rendering an individual quadriplegic and anarthric with preserved self-awareness and normal if not near-normal cognition. A lesion to the ventral pons causes the classic form of LIS, and patients can typically interact with their environment with eye/eyelid movements. LIS patients may live for years with preserved quality of life (QoL) and cognitive function, but with severe disability. However, medical providers and family often underestimate the patient's QoL, and choose less aggressive care. Prompt assessment of decisionality in LIS patients is challenging, but it must be done to allow these patients to participate in their care. We present the case of a 54-year-old man with LIS. The medical team recommended comfort measures, but the family advocated involving the patient in goals of care discussions. The patient was determined to be decisional during the acute hospitalization, and he elected for life-prolonging care. This case emphasizes the importance of unbiased shared decision making, but also the importance of utilizing a practical framework to assess the decision-making capacity in these patients. We provide a suggested approach to determining decision-making capacity in similar cases or conditions. J Pain Symptom Manage 2016;51:789–793. © *2016 American Academy of Hospice and Palliative Medicine. Published by Elsevier Inc. All rights reserved.*

Key Words

Stroke, decision-making capacity, ethics, surrogate, neuropsychology

Introduction

The term locked-in syndrome (LIS) was first used in 1966 to describe patients in a "false coma from supranuclear motor deefferentation."[1] These patients are alert, aware of their environment but cannot speak or move their limbs. Fortunately, LIS is a rare condition that is secondary to the disruption of the motor tracts in the ventral brainstem, and at least 60% are caused by a stroke.[2] The American Congress of Rehabilitation Medicine defined LIS as neurologic impairment characterized by sustained eye opening, preserved cognitive function, severe hypophonia or aphonia, quadriparesis or quadriplegia, and ability to communicate through vertical or lateral eye movement or blinking the upper eyelid.[3] LIS patients are unique when compared to other severe stroke syndromes because of the preserved cognitive. The disability associated with LIS is most similar to upper cervical spine injury and advanced amyotrophic lateral sclerosis.

Given the severity of the neurologic impairment, clinicians are frequently confronted with questions regarding long-term morbidity and mortality. The data regarding the mortality of LIS patients are confusing. One study revealed that 87% of LIS patients die within the first four months.[4] Other studies have shown long-term survival with a 10- and 20-year survival of 83% and 40%, respectively.[5–7] Advances in medical technology, such as mechanical ventilation, allow prolongation of life in these patients.[8] Available resources and cultural differences likely contribute to the discrepancy. The French Association for Locked-In Syndrome database ($n = 250$) showed that 65% of patients eventually removed their tracheostomy tube, and 58% eventually removed their

Address correspondence to: Samuel Maiser, MD, Department of Neurology, Hennepin County Medical Center, 701 Park Avenue, Minneapolis, MN 55415, USA. E-mail: sam.maiser@gmail.com

Accepted for publication: October 28, 2015.

0885-3924/$ - see front matter
http://dx.doi.org/10.1016/j.jpainsymman.2015.10.021

gastrostomy tube.[7] However, patients with LIS often have limited motor recovery of their extremities, even with long-term survival.[4,5]

Despite the presence of persistent neurologic disability and low health-related quality of life (QoL), global quality of life studies have shown that LIS patients have comparable QoL to healthy controls as well as chronically ill patients.[7,9] In addition, most studies have not found a correlation between the extent of physical impairment and depression,[7,10] although one study did.[9] It appears that both global QoL and depression may not be affected by the physical impairment in LIS.[7,10]

We present a case of LIS and review the importance of determining decision-making capacity in LIS, and we provide a suggested approach to determining decision-making capacity in LIS and similar conditions. Although cognitive functions are often preserved in LIS patients, medical providers easily assume they are not, given the patients limited responsiveness. Not realizing that LIS patients, a vulnerable population, are frequently decisional is both dangerous and has many ethical implications. This case report is informative and unique because we provide a practical framework for assessing decision-making capacity in patients with preserved cognitive function in the setting of profound motor weakness. Previous publications regarding decision making in severe stroke tend to focus non-LIS stroke syndromes that are accompanied by profound cognitive, attention, and language deficits.[11]

Case Description

Mr. S., a 54-year-old African-American man, was brought to the Emergency Department via ambulance after family found him agitated in his apartment. The patient was placed on noninvasive ventilation in the Emergency Department because of an irregular and rapid respiratory pattern. He had a history of drug abuse, and his urine toxicology screen was positive for cocaine. He was admitted to the intensive care unit (ICU) with the diagnosis of respiratory insufficiency secondary to drug intoxication/overdose.

In the ICU, the patient became increasingly agitated and unresponsive, requiring endotracheal intubation for airway protection. Within 48 hours, the patient became quadriplegic and anarthric, prompting neurologic investigations. An MRI/MRA scan of the brain revealed an acute ischemic infarction in the ventral pons with severe basilar artery stenosis. Although arousal varied, the patient was able to respond with vertical eye movements and, therefore, he was diagnosed with LIS.

Extensive discussion among medical providers and the family resulted in significant distress and uncertainty about goals of care. Mr. S. was one of eight children; he was not married, had no children, and did not have an advance directive. During the first family meeting in the ICU, the family was informed about the seriousness of the diagnosis and that, if Mr. S. survived, he would most likely require 24-hour nursing care. The family also was informed by the medical team that patients with LIS are often awake and cognitively intact. Therefore, the family wanted more time to allow the patient to be involved in the decision-making process.

While in the ICU, Mr. S. developed worsening decompensated heart failure and was treated for aspiration pneumonia. The family was counseled that he would likely die despite aggressive measures. It was recommended to the family to consider shifting the focus of care to comfort. The family continued to advocate for more time. Attempted and inconsistent communication with the patient resulted in confusion over the goals of care. A communication strategy of "look up for YES" and "look down for NO" was being used, but not always consistently followed. This worsened the distress among medical providers and family. The ICU team consulted palliative care to help with goals of care.

The family understood that Mr. S. had a very low chance of recovering independence, had a high chance of dying, and that having LIS is like "being a prisoner in his own body." The family was counseled on a comfort care plan versus a time-limited trial including tracheostomy/gastrostomy and rehabilitation. End points for a time-limited trial were discussed (i.e., escalation of care at the next acute medical crisis, duration of trial, reassessment, etc.), and worst- and best-case scenarios were provided. The family's distress of not having a designated surrogate, but knowing that the patient could potentially make his own decisions led to a time-limited trial of life prolongation. The family did elect to make the patient do-not-resuscitate at the recommendation of the medical providers.

In an effort to involve Mr. S. in his care, neuropsychology was consulted to determine decision-making capacity. The patient's fluctuating level of alertness was concerning for delirium likely from his aspiration pneumonia. Considerable time (45–60 minutes over multiple days) was spent with the patient establishing a reliable and consistent communication method to minimize the impact of delirium on decision-making capacity. A vertical eye movement up was determined reliable in responding in the affirmative to questions, and no upward movement was used for responding in the negative. All questions were presented both in the affirmative and negative to establish reliability (e.g., after explaining what a G-tube was, the patient was asked "Do you want a G-tube placed to help provide you nutrition?" but also later asked "Do you want to