

# ETHICAL PERSPECTIVES IN NEUROLOGY

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The practice of neurology presents a series of ethical challenges for the clinician. These rarely have simple or straightforward solutions, but require careful consideration by the neurologist. This section of *CONTINUUM*, written by colleagues with particular interest in the area of bioethics, provides a case vignette that raises one or more ethical questions related to the subject area of this issue. The discussion that follows should help the reader understand and resolve the ethical dilemma.

## LOCKED-IN SYNDROME: ENDOVASCULAR STROKE THERAPY AND END-OF-LIFE DECISIONS

**NOTE:** The following is based on an actual case.

A 33-year-old man presented to an emergency department with shortness of breath and altered mental status. He was awake and following commands on arrival, but was nonverbal. His mental status declined, and he became unresponsive. An MRI of the brain showed acute bilateral pontine infarctions consistent with basilar artery thrombosis. He was transferred to a hospital with a neuro-intensive care unit (ICU) and neuroendovascular interventional capabilities. There, he was intubated. His examination was consistent with locked-in-syndrome (LIS) with quadriplegia, ophthalmoplegia, and anarthria. Immediately upon admission he had the ability to communicate using eye blinking. Attempts to treat the midbasilar artery occlusion with balloon angioplasty resulted in partial recanalization but no clinical improvement.

The family members were uncertain of the patient's wishes and whether he would like to be sustained on artificial life-support measures. He did not have a living will, and initially it was difficult to determine whether he was conscious. The family was ambivalent about the plan of care, and was unable to come to a decision. As a result, full support was maintained. The patient gradually became more alert and was able to communicate with eye blinking, but the family and some members of the ICU team were uncertain about his comprehension and ability to make decisions. This case generated much discussion among the health care team about futility of care, quality of life, power of attorney, and burden to the community.

## COMMENT

- The term *locked-in* was first described by Plum and Posner in 1966 to describe the clinical condition of quadriplegia, lower cranial nerve palsies, and anarthria with preserved consciousness. It is difficult to ascertain whether patients with LIS can understand or communicate complex concepts, which makes the assessment of

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decision-making capacity in LIS very difficult. If a patient is incorrectly determined to *have* decision-making capacity when it is actually lacking, then the eye-blinking pattern may wrongly be interpreted as informed communication. On the other hand, if a patient is incorrectly determined to *lack* decision-making capacity when it is actually present, then the patient's surrogate may make decisions that are either consistent or inconsistent with the patient's actual wishes. It is possible that the patient could be fully aware of the decisions, which would potentially add further emotional and psychological stress if the decision made is counter to the awake patient's wishes.

## DISCUSSION

- It is well known that patients with LIS can retain their cognitive abilities and recover enough function to be able to communicate. For example, the 1997 book *The Diving Bell and the Butterfly* is the memoir of Jean-Dominique Bauby, who developed LIS as a result of a stroke at age 43. Bauby "dictated" the memoir to an assistant by eye blinks and a communication board. In describing living in the LIS state, he eloquently recounts the onset of his stroke, "Half conscious, I feel extremely strange, as if I had swallowed an LSD tablet...Not for a second does it occur to me that I may be dying...Then I sink to coma" (Bauby, 1997).

Not all patients with LIS, however, regain full consciousness, and the rate of recovery of consciousness is hard to predict. Especially if the patient neither has an advance directive nor has discussed such a scenario with family or friends, this leaves the patient's family and the ICU team with the dilemma of deciding how long to continue ICU treatment while waiting for communication capacity to return. Furthermore, even if the ability to communicate is established, it is especially challenging to determine comprehension and decision-making capacity when the ability to communicate is limited to eye blinking or eye movements.

There is evidence to suggest that the recovery of communication capacity takes months, which may mean that decisions to withdraw life-sustaining therapies should be delayed, especially if the goal is to allow the patient the opportunity to participate in the decision. Family members are the first to notice that the patient is able to communicate in over 50% of cases, with a mean time to recognition of the LIS (ie, recognition of consciousness) of 78 days (Leon-Carrion et al, 2002). Such a long wait, however, is likely to extend beyond the initial ICU admission, and arrangements would need to be made to reevaluate the patient's ability to communicate at regular intervals. Ideally this should be done by neurologists or rehabilitation specialists familiar with LIS and also should incorporate efforts by communication experts, possibly with facilitated communication devices and methods. Actual physician practices, however, may vary. A 1997 survey of 93 physicians treating patients with LIS found that 55% recommended intubating the patient in the presence of swallowing disturbances and imminent aspiration, 38% were in favor of stopping intensive care, 52% advocated not treating severe infections with antibiotics, 58% were in favor of discussing these problems in detail with the patient, and 87% advocated discussing them with relatives (Thiel et al, 1997).

Surrogate decision making is well known to be fraught with uncertainty. In the case of LIS, health care professionals and even family may assume that the quality of life of a patient with LIS is so poor and limited that it is not worth living. Thus,

knowledge of long-term outcomes is important. The 5-year and 10-year survival rates may be as high as 85% (Casanova et al, 2003; Doble et al, 2003). Interviews with the families and caretakers of long-term survivors of LIS suggest that these patients can develop meaningful roles in their families and communities, despite their severe impairment (Doble et al, 2003). The 1993 AAN position statement on the care of irreversibly paralyzed patients with retained consciousness and cognition recommends regularly reassessing patients' general medical condition, psychological state, and cognitive and communicative abilities to assure their ability to make sound decisions about sustaining or withholding life-support measures (American Academy of Neurology, 1993).

Not all physicians and nurses in the ICU setting may be willing to sustain a patient with possible LIS whose outcome requires prolonged ICU support, as they may consider such care futile. Health care professionals sometimes use their own views and biases of long-term outcome when advising families about early decisions, which is why familiarity with outcomes research is important. Nonetheless, there may still be health care professionals who feel they cannot participate in the care of such patients or who feel that such care is burdensome to society. Consultation from a hospital ethics committee can be helpful, as can consultation from a specialist familiar with outcomes in LIS. If a patient with LIS is determined to have both the capacity to communicate and make informed decisions, and if the patient wishes to have treatment continued, then there is little ethical foundation for health care professionals to refuse to participate in the patient's care. On the other hand, if the patient lacks capacity, or if surrogate decision makers have asked for care that the health care professionals believe is causing more burden than benefit to the patient, an option is to request not to be assigned to the patient's care, but in doing so, it is important that neither the patient nor the family be abandoned.

Family members carry a great burden and responsibilities when a patient has no advance directive and has not had prior discussions about goals of care. Furthermore, few patients would ever have contemplated an illness that would leave them quadriplegic, mute, wheelchair bound, and ventilator dependent. It is not surprising that conflicts can arise within the family over decisions to be made.

First and foremost for the ICU team caring for the patient and family is to acknowledge their burden and the uncertainty of the circumstances and to offer to help them make the best decisions they can. Considering the length of time necessary for patients with possible LIS to recover to the point that their ability to communicate is established, requests from the family for more time to observe the patient's response to therapy are probably reasonable. The ICU team can use this time to provide the family with an overview of the LIS and its long-term outcomes. As for establishing the emergence of the patient's communication ability, the family can be encouraged to establish an eye-blinking communication code with the patient. Once the communication code is established, it can then be determined whether the patient has decision-making capacity and (even more important) whether the patient wishes to make decisions. All autonomous patients have the option of deferring decision-making authority to someone else, and this option should be discussed and offered to patients with LIS who may find such complex communication so difficult that they would rather their surrogate make such decisions for them. Even if this is the case, it is important for the surrogate and the ICU team to inform the patient of the decisions they have made.

**CONCLUSION**

- In this case, after the patient became more alert and very brisk and consistent with his eye-blinking responses to the family's questions, the family gained confidence in his cognitive abilities and felt very strongly that he should make his own decisions. The patient wanted to be maintained on life-support measures. He is now thinking of writing his own memoir with encouragement from his family.

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