

**FAST FACTS AND CONCEPTS #303  
LOCKED-IN SYNDROME**

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**Background:** Locked-in Syndrome (LIS) is a rare neurologic condition characterized by quadriplegia and an inability to articulate speech, but with preserved self-awareness (1,2). It is easily misdiagnosed and once identified, a constellation of medical, psychological, social, and ethical issues may ensue (3,7).

**Pathophysiology and Prognosis:** LIS is caused by the disruption of motor tracts in the ventral brainstem. At least 60% of cases are caused by an acute stroke (4). LIS patients have a high risk of dying within the first several months; however, those that survive that period are likely to live 10 years or more (3, 5-7). They often have limited motor recovery of their extremities, but with long-term survival, many patients eventually have their tracheostomy and gastrostomy tubes safely removed (7). The severe neurologic disability results in a low health-related quality of life, yet, the global quality of life and rate of depression may be no different than healthy controls (7-9). There are no particular symptoms associated with LIS other than those expected from immobility. The prevalence of bodily pain is felt to be similar to healthy controls, although inadequate pain relief is associated with suicidal thoughts (9). Because the etiology is often restricted to the brainstem, LIS typically does not affect cognition (10-12).

**Diagnosis:** Diagnosis is dependent upon the physical examination, but unless the examiner is familiar with LIS, LIS may be mistaken for coma (eyes closed, does not follow commands) or a vegetative state (eyes may open and move, but not to command) (7). Therefore, physical exam is best performed by a neurologist. If LIS is suspected, clinicians should assess for abnormal brainstem respiratory patterns such as central neurogenic hyperventilation (rapid and deep breaths 20-40/min), apneustic breaths (prolonged inspiratory pause) or ataxic respirations (irregularly irregular). A complete coma exam including cranial nerves and volitional eye/eyelid movements should be performed. Cranial imaging is typically performed to elucidate the diagnosis with magnetic resonance imaging as the preferred modality.

**Care Decisions:** The common care decisions in LIS are related to the consequences of the severe impairment of muscles that control eating and breathing. Thus, decisions about the use of mechanical ventilation, artificial nutrition and hydration, and 24 hour nursing care will be paramount. Considering the communication challenges clinicians may encounter, consultation with neuropsychology to assess decision-making capacity should be done early in the patient's course, especially since delirium can be a confounding variable. Though prior wishes expressed in advance care planning documents may be useful, misunderstandings regarding LIS patient's cognition and care preferences are common.

**Communication Strategies:** As portrayed in the 1997 book *The Diving Bell and the Butterfly*, written by a locked-in patient, communication is possible but it requires patience (13) and for the patient, it may be limited to vertical/lateral eye movements or blinking of the upper eyelid (2). Hence, consultation with speech language pathology is advised. The following communication strategies are recommended:

- Establish a reliable and consistent communication method, such as a vertical eye movement up means "yes," and vertical eye movement down means "no."
- Phrase questions so that the answers must be "yes" or "no."
- Present a list of words (i.e. symptoms), and allow the patient time to respond.
- To improve reliability of the decisionality assessment, present questions in both an affirmative and negative manner: "Do you want a PEG tube?" and "Do you want to refuse PEG tube placement?" An orientation question might be, "Is the year 1999? 2019? 2015?" Comprehension can be tested with questions such as "Can a shark fly? Can a hammer pound a nail?"
- Apply the basic principles of determining decisionality as in any other patient (see *Fast Fact #55*)
- Utilize augmentative communication tools such as alphabet boards or eye-tracking devices when available.
- Permit extra time for fluctuating arousal and fatigue.
- Family members and/or primary caregivers may have insights into communication and subtle signals of distress on the part of the patient. These can be elicited and posted at the bedside.

### Cautions:

- LIS patients are at risk for being talked about at the bedside as if they are dead or in a coma by hospital staff (1). Always assume the patient can hear you.
- Assess decisionality and the patient's care preferences as soon as possible. If an advance directive is available, review for care preferences that may guide decision making if the patient is deemed non-decisional.
- Be aware that surrogates and clinicians may wrongly assume that quality of life is poor and not worth living and thereby advocate for the early withdrawal of life sustaining therapies (7, 14-16).
- Clinicians should be aware of their own values and personal responses to a LIS patient, and be careful not to assume they are shared by the patient. Efforts should be made to insure alignment of goals of care between the patient and surrogate throughout the disease trajectory.
- Clinicians should be transparent with patients/surrogates about what the future may look like and discuss a full range of care options such as life prolongation (PEG tube, tracheostomy, nursing home placement) vs a comfort plan of care.

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