

# LOCKED-IN SYNDROME AS A NEURODIVERGENCE

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## The Traditional Definition of Locked-In Syndrome (LiS)

Locked-in Syndrome (LiS) was initially defined as a combination of quadriplegia, lower-cranial nerve paralysis, and mutism with preservation of consciousness. Even in 1966, it was clear by way of this definition that persons experiencing LiS were not experiencing a lack of awareness. By 1986, the mutism component was replaced with anarthria, a complete inability to speak marked by a loss of motor control. This is because 'mutism' in reference to a conscious patient could simply be the result of an unwillingness to communicate (Smith and Delargy, 2005), further emphasizing the retention of awareness in LiS.

Importantly, although we now standardly distinguish LiS as presenting in one of three subtypes (classical, incomplete/partial, and complete/total), the guiding feature for these subtype distinctions is the degree to which paralysis is present (Das, 2023; Halan et al., 2021), not an LiS patient's degree of conscious awareness. Inversely, the more a person with LiS retains an ability for voluntary muscle control (in the classical presentation, typically limited to blinking and vertical movement of the eyes), the more profound a person's LiS subtype.

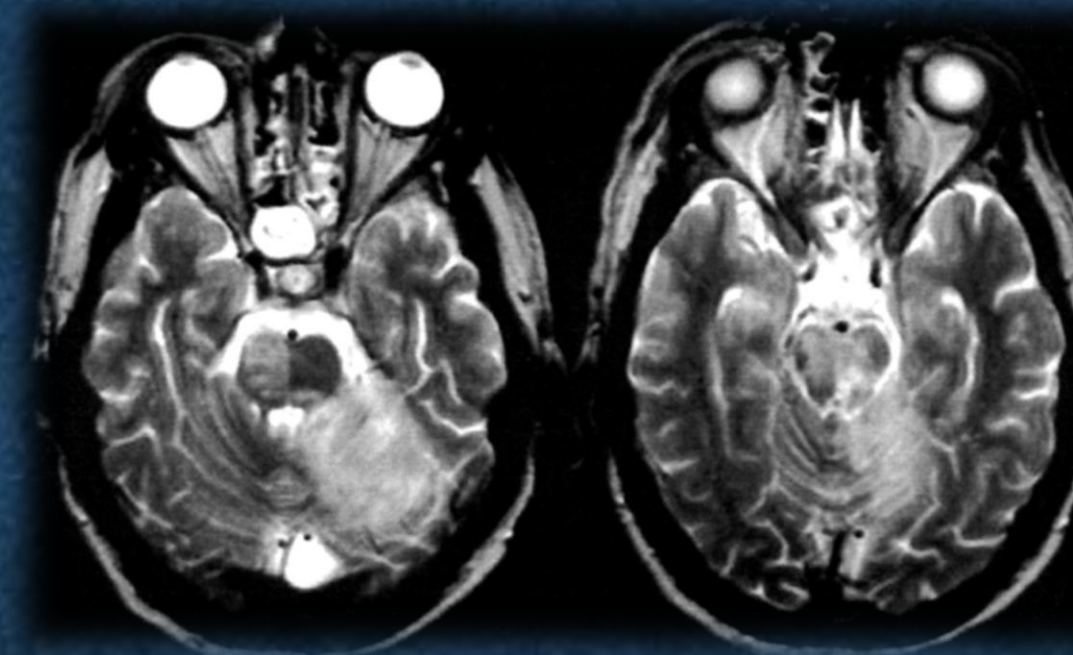
Despite LiS being grouped with coma and Unresponsive Wakefulness Syndrome (UWS) as a **disorder of consciousness**, LiS does not destroy nor disrupt conscious awareness.

This misnomer is puzzling given that intact awareness is a historically-defining clinical feature of the syndrome, i.e., the concept is predicated on behavioral and electroencephalographic evidence that LiS patients retain an intact awareness coupled with the essential diagnostic element from a neurostructural standpoint of bilateral lesions to the ventral pons due to e.g., vascular lesions, masses, infections, traumas, and demyelinating disorders affecting the brainstem (Das, et al., 2023), a region of the brain not involved in generating representational states (i.e., the perceptual or conceptual content of consciousness).

## Locked-In Syndrome, Consciousness, and Personhood

We don't need to be fully-invested higher-brain theorists (e.g., in support of higher-brain neurologic criteria for the determination of death) to support the intuitive and neuropsychologically-backed proposition that personhood is grounded in consciousness. With the irreversible loss of the conscious mind, comes the loss of the person. This is perhaps not so plainly given, nevertheless I believe it is a premise most cognitive scientists and philosophers would tacitly accept.

Given those experiencing LiS retain their consciousness, and the above conceptual relation, LiS patients also retain their personhood. Thus, unlike in cases of coma or UWS wherein agency is lost along with a genuinely disordered consciousness, ethically, in cases of LiS the principle of respect for persons still applies.



Seemingly, however, the manner by which the autonomy of such persons is treated in the neurointensive context often mirrors approaches taken with those genuinely suffering from an often-irreversible loss or fragmentation of consciousness and is thus lacking ethical justification (e.g., excluding LiS patients from conversations regarding goals of care, including code status, assuming their decisional incapacity, inadequate attention to palliation, assumptions regarding subjective quality of life (e.g., the disability paradox), and other basic violations of their rational autonomy).

## Locked-In Syndrome as a Neurodivergence

Whereas LiS cannot be either evidenced or logically supported as a disorder of consciousness, it does carry the potential to affect various cognitive capacities, e.g., attention, executive function, intellectual ability, perception, as well as memory.

The concept of **neurodivergence** is a socially-normative, theoretical posit designed to reframe the many different ways in which minds can function in accordance with differences in their neurologic structure (McDermott, 2022). **Causal factors for neurodivergence are also accepted as varied, e.g., differences are not only marked by neurologic development but by disease and trauma.** Its normativity lies in its efforts to reedify social understanding and thus stigmatized perceptions of neurologic conditions as resulting only in deficits, in order to see them instead as mere variances within an expanded realm of **normal human function and experience.**

## Conclusion

I conclude that whereas there stands no logical (conceptual) nor empirically (behaviorally) supportable argument for LiS to be considered a disorder of consciousness, its effects do include cognitive changes that can be both behaviorally evidenced as well as neurologically supported to be demonstrably consistent with the socially-normative category of disease- or trauma-induced neurodivergence. This reconception not only makes our understanding of LiS a rationally and empirically defensible one, but makes clear the ethical imperative to appropriately attend to those experiencing LiS as one that prioritizes the principle of respect for persons, with the added force and significance of this principle when it operates in regard to vulnerable populations.

## References

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