Disorders of Consciousness and Disability Law

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Abstract

In 2018, the American Academy of Neurology, the American Congress of Rehabilitation Medicine, and the National Institute on Disability, Independent Living, and Rehabilitation Research published a systematic evidence-based review and an associated practice guideline for improved assessment, treatment, and rehabilitation of patients with disorders of consciousness. Patients with disorders of consciousness include individuals in the vegetative and minimally conscious states, as well as others with covert consciousness and cognitive motor dissociation. These landmark publications (concurrently published in *Neurology* and *Archives of Physical Medicine and Rehabilitation*) supplant the 1994 *New England Journal of Medicine* Multi-Society Task Force report on the vegetative state and the 2002 criteria establishing minimally conscious states. The guideline re-designates the permanent vegetative state as chronic. In our article, we consider the legal and ethical implications of the practice guideline for clinical practice and explain the vulnerability of these patients who suffer from high rates of misdiagnosis, inadequate medical surveillance, undertreatment of pain, inadequate rehabilitation, and segregation in chronic care. We argue that these deficiencies in medical care are inconsistent with our growing appreciation of the dynamic nature of these brain states and an emerging standard of care as articulated by the national guideline. These deficiencies also violate domestic and international disability law. To substantiate this latter claim, we apply disability law to this population, focusing on key Americans with Disabilities Act mandates, the relevance of the 1999 Supreme Court, *Olmstead v. L.C.*, and the utility of *Olmstead* enforcement actions to integrate the care of these individuals into the medical mainstream.

Terry Wallis resided in a nursing home, receiving what is euphemistically called “custodial care,” following a car accident in 1984 which left him in the permanent vegetative state.¹ For decades, his family had asked for him to be seen by a neurologist, but they were told there was no hope for recovery.

In 1993, Terry’s mother, Angilee Wallis, received a call from the nursing home.¹ An aide told her that Terry was “not right.” Overnight, Terry’s elderly roommate with dementia became tangled in his sheets and asphyxiated himself. The aide, unschooled in the nature of the vegetative state, sensed Terry’s distress. She could not know that his diagnosis precluded awareness of self, others, or the environment. At the risk of sanction, she called Angilee and told her to rush over.

Angilee recalls Terry “… laying there with his eyes open wide, he would not go to sleep.”¹ She was convinced he had been awake and distressed by the overnight events, although such awareness would have been inconsistent with the vegetative state.

It would take nearly a decade for a consensus panel to correctly characterize Terry’s brain state that night. In 2002, an expert panel codified the Aspen Criteria for the minimally conscious state (MCS), a state of liminal consciousness in which patients have awareness of self, others, and their environment — a condition distinct from the wakeful unresponsiveness that characterizes a vegetative state.² It can be challenging to distinguish between the MCS and the vegetative state because MCS patients’ behavioral manifestations — a proxy for
consciousness — are episodic and intermittent. Moreover, patients may transition from a vegetative state to MCS unpredictably — and often after an interval of many years. Studies have found that MCS patients are misdiagnosed as vegetative at rates upwards of 40%. Many of these errors are due to a lack of will, not methodology, as the Coma Recovery Scale—Revised has been repeatedly validated as a reliable behavioral assessment tool and found to reliably distinguish MCS from the vegetative state when properly used.

So it was with Terry Wallis. In 2003, a year after the promulgation of the Aspen Criteria, Wallis spoke for the first time in 19 years. Although his case generated international media attention and was heralded as a miracle awakening, closer scrutiny of his medical records supplied by the Wallis family suggested that he had been in MCS within months of his accident. His story is a parable about nosologic humility. Despite the evolution of his brain state, medicine did not yet have a diagnostic category to characterize his recovery of consciousness.

The years since the publication of the Aspen Criteria have been marked by progress in the classification and treatment of disorders of consciousness (DoC). Neuroimaging can now identify covert consciousness in some patients who behaviorally appear vegetative but are able to follow volitional commands. Therapeutics are emerging: in a randomized clinical trial, amantadine was shown to accelerate the recovery of consciousness and zolpidem has been reported effective in case reports. Experimental neuromodulation using deep brain stimulation, non-invasive ultrasound, transcranial magnetic stimulation, and vagal nerve stimulation have shown proof of principle. With proper specialized neurorehabilitation, the provision of assistive devices, and emerging pharmacologic treatments, upwards of 21% of patients with disorders of consciousness admitted to acute inpatient rehabilitation were able to regain functional independence in an observational study involving the National Institute on Disability and Rehabilitation Research Traumatic Brain Injury (TBI) Model Systems Programs. Although rehabilitation was not proven as causal in this observation study, and the care provided by exemplary programs in the TBI Model Systems Programs is not the usual standard of care, the data is promising when resources and commitment to care are brought together.

In August 2018, the American Academy of Neurology (AAN), the American Congress of Rehabilitation Medicine (ACRM), and the National Institute on Disability, Independent Living, and Rehabilitation Research (NIDILRR) published a systematic evidence-based review and a derivative practice guideline for patients with DoC. These were published concurrently in Neurology and Archives of Physical Medicine and Rehabilitation.

The guideline supplants earlier statements on the vegetative and minimally conscious states. It calls for the establishment of standards of care for this historically vulnerable population. Recognizing the dynamic nature of brain states — and that 20% of vegetative patients might have late recoveries to MCS — the guideline recommends that the permanent vegetative state be re-designated as the chronic vegetative state. This represents a major shift in nosology. Because the right to die took root in cases involving the vegetative state, it also has cultural import beyond medical practice. These cases include the 1976 New Jersey Supreme Court’s opinion on Quinlan and the highly divisive 2005 proceedings concerning Terri Schiavo. Given the clinical, ethical, and legal salience of these cases, the re-designation of the permanent vegetative state is remarkable. It was the perceived permanence of the vegetative state that became the ethical and legal justification to remove life-sustaining therapy. But even as we appreciate that 20% of patients once thought permanently vegetative may recover additional functionality, 80% will not. Moreover, a commentary accompanying the AAN/ACRM/NIDILRR guideline notes that...
many of those thought to recover from the vegetative state had in fact been covertly conscious.\textsuperscript{25,26} Thus, they were not recoveries from the vegetative at all but rather patients who were not initially identified as being in an MCS. A small number may have had late structural changes undergirding the return of consciousness.\textsuperscript{27}

Based on the evidence review, the guideline calls for improved diagnostic accuracy of brain states and the identification and amelioration of confounding conditions that might obscure the diagnosis of covert consciousness or adversely affect morbidity and mortality if unaddressed. Diagnosis should be informed by the use of serial examinations with the vetted Coma Recovery Scale—Revised\textsuperscript{28} and emerging imaging technologies such as functional magnetic resonance imaging as well as single-photon emission computed tomography, although these remain supplementary means of assessment. For DoC patients, within 4 to 16 weeks of TBI, the guideline calls for the use of amantadine hydrochloride.\textsuperscript{9,17,18} It recommends family counseling, and the discussion of long-term care when the prognosis is grim, but notes that “prognosis is not universally poor” if comorbid conditions are addressed and proper rehabilitation is provided.\textsuperscript{17,18,29}

This is an exceedingly important caveat and one that should not be dismissed. Many thoughtful commentators look at patients with disorders of consciousness who are unable to communicate as a fate worse than death. Some have referred to living in that state as “torture.”\textsuperscript{30} Imagining this plight creates a sense of dread as families, clinicians, and societies contemplate the isolation of these patients trapped in their own heads. Although we appreciate the motivations for such arguments, we caution against the implicit power of a potentially unrecognized “disability paradox.”\textsuperscript{31}

Clinical care in these cases should be guided by the patient’s prior wishes and the views of the surrogates as well as an accurate assessment of the risks and benefits of continued treatment and rehabilitation, as in all cases of surrogate decision-making. The goal is to preserve choice and affirm the possibility of care\textsuperscript{32} through a paradigm of informed consent. This paradigm provides families with accurate information about what might be achieved with proper rehabilitation.

We may differ with the medical mainstream on two salient points. First, we believe that the status quo is undertreatment, which is equal to therapeutic nihilism. There is a long-term association of futility with the right-to-die dating to the prominent role that vegetative state played in landmark cases such as Quinlan, Cruzan, and Schiavo.\textsuperscript{1} We believe that the ineffectuality of treatment found in cases where the patient is indisputably and chronically vegetative has been generalized to other similar but distinct brain states where there may be benefit in ongoing treatment and rehabilitation. Our goal is to offer a corrective approach drawing on new knowledge of brain resilience.

Second, we believe in the importance of the patient’s wishes. We urge a doctrine of informed consent and ask clinicians to think about how to help conscious individuals who are trapped in their head by adhering to the recent practice guidelines, which offer some promise in a difficult clinical landscape.

Progress in the field is tempered by data from the evidence-based review which state that “the number of patients with prolonged DoC admitted to inpatient rehabilitation settings has progressively declined over the past 15 years.”\textsuperscript{15,16} Paradoxically, although the science has advanced, access to rehabilitative care is less available than it once was.\textsuperscript{25,26}

Patients with DoC are as vulnerable to misdiagnosis and neglect as Terry Wallis was. A family member of a patient with DoC who was interviewed as part of an ongoing narrative study worried that, “human error and incompetence are going to interfere with my wife’s recovery and not the nature and the extent of her brain injury and that is something I cannot accept.”\textsuperscript{33} If patients are mistakenly thought to be vegetative when they have covert consciousness, care providers might presume them to be insensate; therefore, the patient might be deprived of neuropalliative care and pain.
and symptom management. Neuroimaging data have shown that patients who are in the vegetative state do not have the integrative and widely distributed neural networks necessary to perceive pain. This in contrast with patients with MCS who have integrative cortical function. The ability of MCS patients to perceive pain coupled with their inability to reliably cry out when they are in distress makes this diagnostic error of omission more chilling. This creates a clinical and ethical imperative to differentiate vegetative patients from those who retain consciousness, as patients who are conscious are sensate even when they may not be able to ask for relief.

The fact that patients with covert consciousness, who often live in isolation, feel pain but are unable to ask for relief should strike us as ethically unacceptable. Their profound vulnerability should raise the question of how this population might be protected from continued neglect.

Clinical prescriptions are becoming clear in light of a consensus statement from leading clinical societies and the National Institutes of Health. Clinical care should seek to avoid complications to allow patients the opportunity to recover. Protected patients should have access to rehabilitation and evolving therapeutics that might accelerate the recovery of consciousness or unmask covert consciousness when it is present. Given the dynamic nature of these conditions, we must appreciate that it takes time for the injured brain to heal and provide longitudinal rehabilitation. As recent research has shown, patients with DoC undergo axonal sprouting and pruning reminiscent of normal development for years after injury. These changes suggest the possibility of an ongoing regenerative process. In one study, structural changes were correlated with functional improvement in a patient with MCS who was later able to communicate and interact with the environment.

This course of recovery makes it important for patients to be assessed longitudinally from acute care to rehabilitation to chronic care in order to track the evolution of their brain states over time. Some part of diagnostic error rates attendant to this population can be explained by outdated diagnoses which were accurate before dynamic changes occurred within the injured brain. To surveil patients over time, access should be provided to specialized centers where patients can undergo neuroimaging and neuropsychiatric testing that is not now routinely available in the skilled nursing homes where many of these patients reside.

While the recently published guideline makes clear the importance of an accurate diagnosis and outlines good clinical practice for this population, we still must endeavor to ensure that health systems provide patients with DoC proper medical care and rehabilitation. After all, it has been nearly 2 decades since MCS was described; however, misdiagnosis of MCS patients remains routine. We know that patients with access to proper rehabilitation are more likely to recover consciousness, yet many patients do not have access.

Although one might reasonably view our call for change as a matter of ensuring access to high-quality medical care through health insurance reform, we believe that it is critical to view the needs of this population through the lens of disability rights.

Reframing care in terms of disability rights not only shows respect for members of this marginalized group, it also implicates laws prohibiting discrimination against and mandating equal treatment for persons with disabilities, including those within health care settings. Some may think it is obvious that disability law applies to this population, but recent research has shown that practicing physicians do not understand their legal obligations to their patients with disabilities, including how to accommodate them.

There is not only a professional responsibility to engage in good clinical practice or an ethical obligation to recognize the worth of individuals with a DoC; following guideline recommendations can also be viewed as a legal requirement under federal disability law. With legal duties come legal consequences or remedies for violating these requirements.

The most important of the disability rights laws is the Americans with Disabilities
Act (ADA). Enacted in 1990, the ADA prohibits discrimination against people with disabilities. The statute defines discrimination to include the failure to make “reasonable modifications” to policies or practices where necessary to accommodate a disability. Accommodations must be made unless they present an “undue burden.” The ADA applies to both public and private hospitals and care settings, and in contrast to other civil rights laws (such as those prohibiting discrimination on the basis of race), the ADA requires physicians and health care organizations to be proactive. Individuals with DoC clearly count as having a “disability” under that statute because they have a physical or mental “impairment” that “substantially limits” consciousness, a “major bodily function.” Critically, in light of ADA provisions, provision of the care recommended by the practice guideline would not be so costly as to constitute an “undue burden” and would require the delivery of routine medical care and rehabilitation.

The ADA also requires that services be provided to individuals with disabilities in “the most integrated setting appropriate to the needs of the individual.” The key case interpreting the ADA’s integration mandate is the 1999 US Supreme Court decision in Olmstead v L.C. In this case, two women with mental illness and developmental disabilities who had been hospitalized in state institutions sued the state of Georgia to obtain services in community-based settings. The court held that the state was required to serve the plaintiffs in the community “when the State’s treatment professionals have determined that community placement is appropriate, the transfer from institutional care to a less restrictive setting is not opposed by the affected individual, and the placement can be reasonably accommodated, taking into account the resources available to the State …”

Olmstead v L.C. importantly held that the rights of people with disabilities to community and reintegration were robust enough to require the state to provide community-based services even if it entailed additional costs. Of course, there are limits to what constitutes reasonable accommodation given scarce resources and competing goals, but we would argue that the minimal standard of care has evolved with the publication of the new AAN/ACRM/NIDILRR guidelines. The restoration of voice also allows patients to express a choice about their wishes regarding care and placement, so that they feel comfortable and safe. Olmstead v L.C. enforcement actions have since been used as a means to integrate people with disabilities back into their communities.

Segregation is a key issue for persons with DoC. Segregated in the chronic care sector, far from the advances that inform the new practice guideline, these patients often receive substandard care. This may mean that they are deprived of interventions that could enable a transfer to a less restrictive environment, such as group homes, or returning back to their families, which may violate the ADA’s integration mandate.

Substandard treatment can mean that patients are deprived of interventions that might restore their ability to functionally communicate. When this happens, broken ties with family — which might be repaired if they could communicate — remain severed, perpetuating separation from their community, and violating their human rights as detailed in international law. Lack of access to these interventions which could assist in communication, known as “auxiliary aids and services” in the statute, may also violate the ADA. Here we view these interventions as a means to restore the patients’ voice and allow for functional communication. With communication comes the prospect for community and community reintegration, which could mean returning home or better engagement with family and friends while still receiving institutional care.

Whereas Olmstead v L.C. enforcement actions have not always had the support necessary to ensure access to community-based services to which they are entitled, an Olmstead v L.C.—based approach could be instrumental for patients with DoC. With improved treatments, some patients may be able to move from institutional to
community settings, as researchers have shown is possible for approximately one-fifth of patients with DoC.14 Under a conventional reading of Olmstead v L.C., these patients have the right to treatment and the de-institutionalization that could follow, should they and their guardians (if they are under guardianship) concur. By restoring their voice and perhaps their ability to choose, improved treatments can help ensure that these patients can express their own preferences so that guardians are not the only ones speaking for them.

What about those patients who still require institutional care? In a number of recent cases, federal courts have held that Olmstead v L.C. is not just about where people with disabilities live but about how they live and in particular about whether they have the opportunity to engage with others and make decisions about their lives. In a 2016 decision, a federal court of appeals held that a state’s failure to provide services that enabled a disabled plaintiff to interact with nondisabled individuals to the greatest extent possible would violate the integration mandate. That was true even though the state was serving the plaintiff in his own home rather than an institution.52 In 2012, a federal district court held that the state’s failure to provide integrated employment opportunities for people with developmental disabilities would violate Olmstead v L.C., even if they did not live in institutions.53 The same principle applies in this context. Even for those patients with DoCs who require care in an institutional setting, treatments that enable greater interaction with nondisabled relatives and friends serve a moral good that should be pursued.

By affirming the legal rights of patients with DoC, society recognizes their social worth and right to equitable treatment. Indeed, to ensure a just allocation of scarce resources, we are morally compelled to consider the plight of conscious individuals who have been systematically deprived of care that might make a difference for their future and that of their families.

As the US Supreme Court noted “... new insights and societal understandings can reveal unjustified inequality within fundamental institutions that once passed as unnoticed and unchallenged...”54 With the advance of clinical science pertaining to disorders of consciousness,24,55 medicine and society can no longer plead ignorance when it comes to the needs of people whose consciousness is present but not always apparent. The status quo of misdiagnosis and neglect perpetuates segregation and sanctions therapeutic nihilism,56 which may violate both the law and evolving ethical norms.54 The AAN/ACRM/NIDILRR disorders of consciousness practice guideline17,18 provides an evidence base for practice reform, and federal law provides a road map for equitable inclusion in the least restrictive and most integrated care setting appropriate.37

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Abbreviations and Acronyms: AAN = American Academy of Neurology; ACRM = American College of Rehabilitation Medicine; ADA = Americans with Disabilities Act; DoC = disorders of consciousness; MCS = minimally conscious state; NIDILRR = National Institute on Disability, Independent Living and Rehabilitation Research; TBI = traumatic brain injury

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REFERENCES


43. 42 U.S.C § 12102.

44. 42 U.S.C § 12102.


46. 42 U.S.C § 12102(b)(1)(B); 28 C.F.R. § 35.130(e).


52. Steimel v. Wernert, 823 F3d 902 (7th Cir. 2016).


54. Obergefell (135 S. Ct. 2584 [2015]).

