



Practice Guideline Update Recommendations Summary: Disorders of Consciousness

Report of the Guideline Development, Dissemination, and Implementation Subcommittee of the American Academy of Neurology; the American Congress of Rehabilitation Medicine; and the National Institute on Disability, Independent Living, and Rehabilitation Research

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Abstract

Objective: To update the 1995 American Academy of Neurology (AAN) practice parameter on persistent vegetative state and the 2002 case definition on minimally conscious state (MCS) and provide care recommendations for patients with prolonged disorders of consciousness (DoC).

Methods: Recommendations were based on systematic review evidence, related evidence, care principles, and inferences using a modified Delphi consensus process according to the AAN 2011 process manual, as amended.

Recommendations: Clinicians should identify and treat confounding conditions, optimize arousal, and perform serial standardized assessments to improve diagnostic accuracy in adults and children with prolonged DoC (Level B). Clinicians should counsel families that for adults, MCS (vs vegetative state [VS]/ unresponsive wakefulness syndrome [UWS]) and traumatic (vs nontraumatic) etiology are associated with more favorable outcomes (Level B). When prognosis is poor, long-term care must be discussed (Level A), acknowledging that prognosis is not universally poor (Level B). Structural MRI, SPECT, and the Coma Recovery Scale—Revised can assist prognostication in adults (Level B); no tests are shown to improve prognostic accuracy in children. Pain always should be assessed and treated (Level B) and evidence supporting treatment approaches discussed (Level B). Clinicians should prescribe amantadine (100–200 mg bid) for adults with traumatic VS/UWS or MCS (4–16 weeks post injury) to hasten functional recovery and reduce disability early in recovery (Level B). Family counseling concerning children should acknowledge that natural history of recovery, prognosis, and treatment are not established (Level B). Recent evidence indicates that the term chronic VS/UWS should replace permanent VS, with duration specified (Level B). Additional recommendations are included.

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This article presents practice guideline recommendations developed by the American Academy of Neurology (AAN), the American Congress of Rehabilitation Medicine (ACRM), and the National Institute on Disability, Independent Living, and Rehabilitation Research (see complete guideline at <http://www.archives-pmr.org/>). A companion article summarizes systematic review conclusions.¹

Recommendations

Unless noted, recommendations apply to individuals with prolonged disorders of consciousness (DoC) (i.e., ≥ 28 days). Recommendation rationales are presented; tables summarize recommendations for adults (tables 1–3) and children (table 4).

Recommendation 1 rationale

Our systematic review highlights the complexities of caring for patients with prolonged DoC (i.e., ≥ 28 days) at every stage. Such patients may be misdiagnosed due to confounding neurologic deficits² or inexperience in examining patients for subtle signs of consciousness.³ Accurate diagnosis is important to educate families about patients' level of consciousness and function, inform prognostic counseling, and guide treatment decisions. Knowledge gaps often lead to overestimation or underestimation of prognosis by nonspecialists.⁴ In addition, patients with prolonged DoC frequently experience significant medical complications that can slow recovery and interfere with treatment interventions.⁵ In view of this risk, patients are likely to have a better chance for recovery if care is

provided in a specialized setting managed by clinicians who are knowledgeable about the risks associated with DoC and are capable of initiating timely treatment. This is supported by findings from a large retrospective trauma registry, which found that cumulative mortality at 3 years postdischarge is significantly lower for patients discharged to home or inpatient rehabilitation facilities than those discharged to skilled nursing facilities, even after adjusting for covariates.⁶ Care for patients with prolonged DoC may benefit from a team of multidisciplinary rehabilitation specialists, including neurologists, psychologists, neuropsychologists, physiatrists, physical therapists, occupational therapists, speech pathologists, nurses, nutritionists, internists, and social workers (supplemental data, available online only at <http://www.archives-pmr.org/>).

Recommendation 2 rationale

The range of physical and cognitive impairments experienced by individuals with severe DoC complicates diagnostic accuracy and makes it difficult to distinguish behaviors that are indicative of conscious awareness from those that are random and non-purposeful. Interpretation of inconsistent behaviors or simple motor responses is particularly challenging. Fluctuations in arousal and response to command further confound the reliability of clinical assessment.^{7,8} Underlying central and peripheral impairments, such as aphasia, neuromuscular abnormalities, and sensory deficits, may also mask conscious awareness.^{9–11} Clinician reliance on non-standardized procedures, even when the examination is performed by experienced clinicians,^{2,12,13} contributes to diagnostic error, which consistently hovers around 40%. Diagnostic error includes misdiagnosing the locked-in syndrome for vegetative state/unresponsive wakefulness syndrome (VS/UWS) and minimally conscious state (MCS).^{14,15} Accurate diagnosis of the level of consciousness has implications for prognosis and management.

Recommendation 2a rationale

In view of the range of clinical challenges to accurate and reliable diagnosis of DoC, standardizing the assessment of patients with DoC can assist in recognizing key diagnostic features that may be missed on ad hoc examinations.^{12,16} The validity and reliability of standardized neurobehavioral assessment scales for diagnosis of DoC subtype have been previously reviewed.¹⁷ Other techniques

List of abbreviations:

AAN	American Academy of Neurology
ACRM	American Congress of Rehabilitation Medicine
CI	confidence interval
CRS-R	Coma Recovery Scale—Revised
DoC	disorders of consciousness
DRS	Disability Rating Scale
FDG	fluorodeoxyglucose
MCS	minimally conscious state
TBI	traumatic brain injury
UWS	unresponsive wakefulness syndrome
VS	vegetative state

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