

# Disorders of Consciousness

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## KEYWORDS

- Disorder of consciousness • Rehabilitation • Coma • Vegetative state
- Minimally conscious state • Traumatic brain injury

## KEY POINTS

- Disorders of consciousness (DOC) are altered states of pathologic consciousness, which can be subdivided into coma, vegetative state, and minimally conscious state (MCS) based on neurobehavioral function.
- The Coma Recovery Scale-Revised assessment scale is recommended in DOC for clinical practice and research.
- Emergence from MCS is defined as reliable and consistent functional object use and functional communication.
- In a randomized, double-blinded, placebo controlled study, Amantadine improved functional recovery in patients with DOC.

## INTRODUCTION

Annually, approximately 2.5 million people sustain a traumatic brain injury (TBI) in the United States, and more than 5.3 million people live with a TBI-related disability. TBI not only impacts the life of an individual and their family but also has a large societal and economic toll. The estimated economic cost of TBI in 2010, including direct and indirect medical costs, was approximately \$76.5 billion. In addition, the cost of fatal TBIs and TBIs requiring hospitalization accounts for approximately 90% of the total TBI medical costs.<sup>1</sup> Approximately 0.3% severe TBIs can result in Disorders of Consciousness (DOC).<sup>2</sup> DOC is a state of prolonged altered consciousness, which can be

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categorized into coma, vegetative state (VS), and minimally conscious state (MCS). DOC can prove difficult to diagnose and treat and can result in increased burden of care for families and facilities. In this article, the authors review the definition, diagnosis, imaging, and treatment interventions for this difficult patient population.

## CONSCIOUSNESS

Historically, the concept of human consciousness has been difficult to describe on both a philosophic and a scientific level. Previous models often describe this phenomenon as a subjective experience, which consequently poses a diagnostic challenge in patients with a DOC. However, recent advances in modern medicine have allowed for improved survivability of acute brain injury and have secondarily imparted insight into the neural correlates of consciousness. Clinically, the 2 components that separate consciousness from unconsciousness are arousal and awareness.<sup>3</sup> Wakefulness is a state of arousal, which can be assessed by the presence of eye-opening and brainstem responses. The depth of wakefulness can be evaluated objectively using measures such as the Glasgow Coma Scale.<sup>4</sup> On a neuroanatomic level, arousal is mediated by the ascending reticular activating system of the upper brainstem. Activation of the cerebral cortex occurs with passage of sensory information from the upper brainstem via reticulothalamocortical and extrathalamic pathways. From a neurobiologic perspective, the conscious awake state is associated with a high energy demand and electrical activity within the corticothalamic system. This is further supported by electroencephalogram recordings (EEG), which show that increasing levels of arousal are associated with increased frequency of electrical activity in the cerebral cortex.<sup>4</sup> Conversely, a decline in arousal is associated with reduction in excitatory neuromodulatory influences. The global deafferentation and disruption of the corticothalamic networks could explain the dysfunction in arousal seen in severe brain injuries.<sup>5</sup> Awareness refers to the ability of an individual to respond to both external and internal stimuli in an integrated manner. It is inferred by command following and neurobehavioral assessment.<sup>6</sup> On a neuroanatomic level, the connectivity of frontoparietal regions and the thalamus appears to play a role in the maintenance of consciousness.<sup>7</sup> This is supported by functional MRI (fMRI) studies, which suggest dysfunctional cerebral connectivity in widespread areas of the frontoparietal networks in patients with DOC.<sup>8</sup> In a healthy individual, an increase in arousal is associated with an increase in awareness in a linear fashion along the continuum of conscious states.<sup>9</sup> A dissociation of these 2 components of consciousness is seen in pathologic states, such as in the VS and MCS.

## CLINICAL ENTITIES

### *Brain Death*

In 1995, the American Academy of Neurology (AAN) provided practice guidelines for the determination of brain death. They emphasized 3 clinical findings that indicate cessation of brain function: (1) coma (of known cause), (2) absence of brainstem reflexes, and (3) apnea. Before this determination, other causes for brainstem dysfunction should be excluded, including shock/hypotension, hypothermia, central nervous system depressants, spinal cord injury, and electrolyte and/or endocrine abnormalities. There is no consensus regarding the timing of follow-up testing, but clinicians must use judgment and perform serial evaluations to exclude the possibility for recovery. A diagnosis of brain death is ominous, and there have been no reports of neurologic recovery once determined by the 1995 AAN practice parameters.<sup>10,11</sup>

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