Decision making for primary prevention in disorders other than coronary artery disease and dilated cardiomyopathy is generally driven by observational data and judgment based on clinical observations. Controlled clinical trials providing evidence-based indicators for ICDs are lacking for these smaller population subgroups. In general, for the rare disorders listed above, indicators of arrhythmic risk such as syncope, documented ventricular tachyarrhythmias, aborted cardiac arrest, or a family history of premature SCD in some conditions, and a number of other clinical or ECG markers, may be used as indicators for ICDs.

SECTION 3 NEUROLOGIC CRITICAL CARE

328 Coma

Allan H. Ropper

Coma is among the most common and striking problems in general medicine. It accounts for a substantial portion of admissions to emergency wards and occurs on all hospital services. It demands immediate attention and requires an organized approach.

There is a continuum of states of reduced alertness, the most severe form being coma, defined as a deep sleeplike state from which the patient cannot be aroused. Stupor refers to a higher degree of arousability in which the patient can be transiently awakened by vigorous stimuli, accompanied by motor behavior that leads to avoidance of uncomfortable or aggravating stimuli. Drowsiness, which is familiar to all persons, simulates light sleep and is characterized by easy arousal and the persistence of alertness for brief periods. Drowsiness and stupor are usually accompanied by some degree of confusion (Chap. 34). A precise narrative description of the level of arousal and of the type of responses evoked by various stimuli as observed at the bedside is preferable to ambiguous terms such as lethargy, semicoma, or obtundation.

Several conditions that render patients unresponsive and simulate coma are considered separately because of their special significance. The vegetative state signifies an awake-appearing but nonresponsive state in a patient who has emerged from coma. In the vegetative state, the eyelids may open, giving the appearance of wakefulness. Respiratory and autonomic functions are retained. Yawning, coughing, swallowing, and limb and head movements persist, and the patient may follow visually presented objects, but there are few, if any, meaningful responses to the external and internal environment—in essence, an "awake coma." The term *vegetative* is unfortunate because it is subject to misinterpretation. There are always accompanying signs that indicate extensive damage in both cerebral hemispheres, e.g., decerebrate or decorticate limb posturing and absent responses to visual stimuli (see below). In the closely related but less severe minimally conscious state, the patient displays rudimentary vocal or motor behaviors, often spontaneous, but some in response to touch, visual stimuli, or command. Cardiac arrest with cerebral hypoperfusion and head injuries are the most common causes of the vegetative and minimally conscious states (Chaps. 327 and 330). The prognosis for regaining mental faculties once the vegetative state has supervened for several months is very poor, and after a year, almost nil; hence the term *persistent vegetative state*. Most reports of dramatic recovery, when investigated carefully, are found to yield to the usual rules for prognosis, but there have been rare instances in which recovery has occurred to a severely disabled condition and, in rare childhood cases, to an even better state. The possibility of incorrectly attributing meaningful behavior to patients in the vegetative and minimally conscious states creates inordinate problems and anguish. On the other hand, the question of whether these patients lack any capability for cognition has been reopened by functional imaging studies that have demonstrated, in a small proportion of posttraumatic cases, meaningful cerebral activation in response to verbal and other stimuli.

Apart from the above conditions, several syndromes that affect alertness are prone to be misinterpreted as stupor or coma. Akinetic mutism refers to a partially or fully awake state in which the patient is able to form impressions and think, as demonstrated by later recounting of events, but remains virtually immobile and mute. The condition results from damage in the regions of the medial thalamic nuclei or the frontal lobes (particularly lesions situated deeply or on the orbitofrontal surfaces) or from extreme hydrocephalus. The term abulia describes a milder form of akinetic mutism characterized by mental and physical slowness and diminished ability to initiate activity. It is also usually the result of damage to the frontal lobes and its connections (Chap. 36).

Catatonia is a curious hypomobile and mute syndrome that occurs as part of a major psychosis, usually schizophrenia or major depression. Catatonic patients make few voluntary or responsive movements, although they blink, swallow, and may not appear distressed. There are nonetheless signs that the patient is responsive, although it may take ingenuity on the part of the examiner to demonstrate them. For example, eyelid elevation is actively resisted, blinking occurs in response to a visual threat, and the eyes move concomitantly with head rotation, all of which are inconsistent with the presence of a brain lesion causing unresponsiveness. It is characteristic but not invariable in catatonia for the limbs to retain the postures in which they have been placed by the examiner ("waxy flexibility," or catalepsy). With recovery, patients often have some memory of events that occurred during their catatonic stupor. Catatonia is superficially similar to akinetic mutism, but clinical evidence of cerebral damage such as Babinski signs and hypertonicity of the limbs is lacking. The special problem of coma in brain death is discussed below.

The locked-in state describes yet another type of pseudocoma in which an awake patient has no means of producing speech or volitional movement but retains voluntary vertical eye movements and lid elevation, thus allowing the patient to signal with a clear mind. The pupils are normally reactive. Such individuals have written entire treatises using Morse code. The usual cause is an infarction or hemorrhage of the ventral pons that transects all descending motor (corticospinal and corticobulbar) pathways. A similar awake but de-efferented state occurs as a result of total paralysis of the musculature in severe cases of Guillain-Barré syndrome (Chap. 460), critical illness neuropathy (Chap. 330), and pharmacologic neuromuscular blockade.

THE ANATOMY AND PHYSIOLOGY OF COMA

Almost all instances of diminished alertness can be traced to widespread abnormalities of the cerebral hemispheres or to reduced activity of a special thalamocortical alerting system termed the reticular activating system (RAS). The proper functioning of this system, its ascending projections to the cortex, and the cortex itself are required to maintain alertness and coherence of thought. It follows that the principal causes of coma are (1) lesions that damage the RAS in the upper midbrain or its projections; (2) destruction of large portions of both cerebral hemispheres; or (3) suppression of reticulocerebral function by drugs, toxins, or metabolic derangements such as hypoglycemia, anoxia, uremia, and hepatic failure.

The proximity of the RAS to midbrain structures that control pupillary function and eye movements permits clinical localization of the cause of coma in many cases. Pupillary enlargement with loss of light