

## NEUROCRITICAL CARE THROUGH HISTORY



# A Comatose State in Search of a Name: A History of Its Terminology and Semiology

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Notwithstanding the pertinent legal and societal implications of applying neuroimaging and new research to “responsiveness” in prolonged coma [1, 2], neurologists and neurosurgeons for decades have recognized states from which no one (or almost no one) substantially recovered or even awakened. Clearly, a large number of the patients who did recover were initially misdiagnosed and received an unduly bleak prognosis. However, rehabilitation physicians in particular [3] recognized that referred patients who had been “unresponsive” became more awake, albeit minimally so. The history of the semiology of prolonged unconsciousness can be best summarized as initial multiple attempts to describe the indescribable and indefinable, a presumed resolution after Jennet and Plum’s article in 1972 [3], and a Multi-Society Task Force recommendation in 1994 [4, 5]. Why was it so difficult to reach the skills to make this diagnosis and where did we fall short? A survey of the past may provide insights.

### Phrasing of Prolonged Coma

The terminology has changed considerably, and we are still struggling to name this elusive neurologic state. For many neurologists, the destruction of the brain was not debatable, but when comparing equally severe injuries, why did they find some patients in a prolonged state and others not?

Clinicians would not make this diagnosis in the immediate aftermath of the acute brain injury but after the patient had been in the state for months or even years. In Germany, the psychiatrist Ernst Kretschmer (1888–1964), better known for his concept of constitutional

body types, introduced the term *Das apallische Syndrom* in 1940 (Fig. 1 [6]). Kretschmer coined the term similar to apraxia and agnosia [6]. Apallia referred to a lesion (literally absence) of the pallium, the mantle of gray matter forming the cortex; in fact, his patients had suffered a catastrophic injury to the cortex from anoxic-ischemic injury.

In France, the terms included *la stupeur hypertonique post-comateuse* or *vie vegetative*. Dejerine published a large work on neurologic semiology [7]. He labeled the more severe types of comas as *coma carus*, which he applied to patients who were barely arousable, who were nearly immobile, and who could only opening their eyes to a strong stimulus. A strange case is Dejerine’s *coma vigil*, with eyes spontaneously open; from the description, it seems like a severe depression or abulia: “*Le coma vigil est un assemblage paradoxal de dépression et d’excitation psychiques, d’accablement et de délire, de sommeil et de veille. Le malade a les yeux fermés, mais les ouvre au moindre appel; il lors mais il s’agit et parle.*” (“The waking coma is a paradoxical assembly of mental depression and excitement, dejection and delirium, and sleep and wakefulness. The patient has his eyes closed but opens them at the slightest call; but he moves and talks”).

A new term, “unresponsive wakefulness syndrome,” is another misnomer. The patient is responsive to noxious stimuli (albeit reflexive), and eye opening is different from wakefulness or arousal. The term was introduced because of the perceived connection of the word *vegetative* with the term *vegetable* and it blurs distinctions with a minimally conscious state, locked-in syndrome, or other levels of severe disability. A similar homophone occurs in Spanish, in which the term is *vegetativo*, which sounds like “*vegetal*”. Several countries refer to patients in this state as “plants” that must be watered from time to time. All these popular-language associations are seriously

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# Das apallische Syndrom.

Von

Prof. Ernst Kretschmer, Marburg.

(Eingegangen am 14. März 1940.)

Die Beschreibung der psychischen Begleiterscheinungen cerebralneurologischer Vorgänge bei ausgebreiteten Gehirnerkrankungen, die in erster Linie die Großhirnrinde betreffen, ist an gewissen Stellen noch lückenhaft; statt einer klaren Syndromheraushebung muß man sich dann entweder mit der Aufreihung verzetzelter und für sich allein nichtsagender Einzelsymptome begnügen oder man ist gezwungen, benachbarte Syndrombezeichnungen hilfsweise heranzuziehen, die dann den Tatbestand schief und ungenau decken.

Für bestimmte Formen weitgehender Ausschaltung gesamtpsychischer Leistungen, bei gleichzeitigem Erhaltensein der notwendigsten vegetativen Steuerungen des Hirnstamms, haben wir den Begriff der „hypnoiden Syndrome“, den wir in seinen schweren Ausprägungen bekanntlich mit dem Ausdruck „Koma“, in seinen leichteren Graden als „Somnolenz“, „Benommenheit“ usw. quantitativ mit einer Reihe von Bezeichnungen abstufen können. Man könnte sie auch als „Bewußtseinsstörungen“ im engeren Wortsinn zusammenfassen, wenn der Ausdruck „Bewußtsein“ nicht allzu vieldeutig wäre. Ursächlich betrachtet sind der Prototyp dieser Gruppe die pathologisch-physiologischen Störungen, die bei Überschwemmung des Gehirns mit Toxinen eintreten, denen normalphysiologisch die Schlafreaktion auf Ermüdungstoxine entspricht. Wesentlich für alle in die hypnoide Gruppe gehörigen Phänomene ist die in ihnen enthaltene Störung der *Wach-Schlafsteuerung* im Sinne einer herabgesetzten „Helligkeit“ des Bewußtseins bis zum Grade tiefer pathologischer Schlafzustände. Wesentlich für den subjektiven Tatbestand ist immer das einfache diffuse Dunkel- und Unscharfwerden des Erlebens mit nachfolgender Erinnerungstrübung bzw. Amnesie. Objektiv ist, wenn dies nicht gezeigt werden kann, mindestens ein zeitweiliges oder angedeutetes Auftreten schläfriger Symptome in der Motorik, hinsichtlich Gesichtsinervation, Lidsenkung, Gähnen, Atmungstypus, Kopffasomotorium, sei es mit passivem, ungezieltem, verträumtem Bewegungsspiel (Delir), sei es nur mit Wegsinken der Aufmerksamkeit (Amentia), oder zeitweiliger Desorientierung zu fordern, sofern der Ausdruck „Bewußtseinstrübung“ etwas Präzises besagen soll.

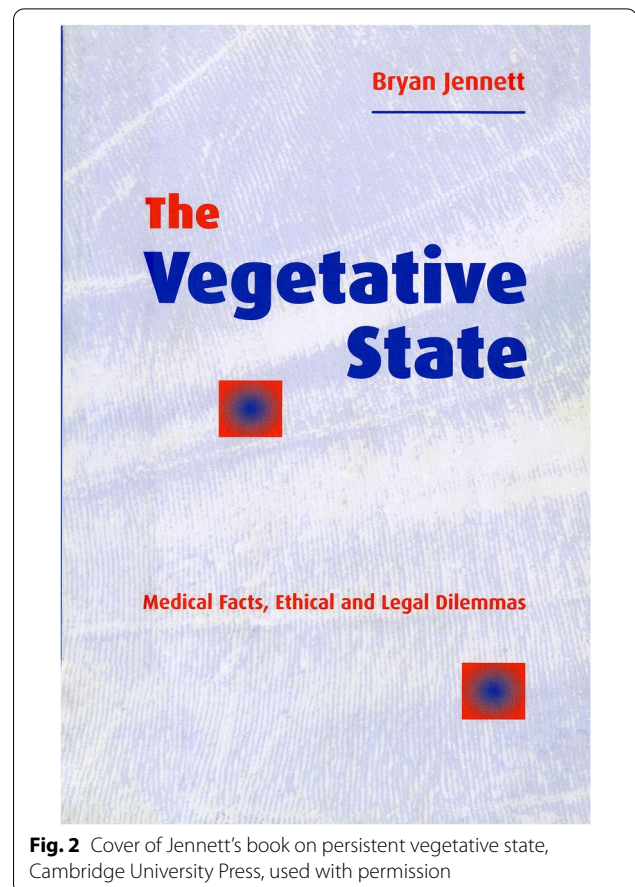
problematic and derogatory, but the reason the term “vegetative” was chosen is not commonly known.

In the UK, the first attempts to define this syndrome clinicopathologically (“neocortical death”) came from the Institute of Neurological Sciences in Glasgow, but this term was applied only to patients after cardiac arrest. Jennett and Plum then proposed the term “persistent vegetative state” (PVS) and described clinical features that distinguished it from other, less severely affected disabling neurological states. In their 1972 *Lancet* communication, they wrote “the word ‘vegetative’ itself is not obscure: vegetate is defined in the *Oxford English Dictionary* as “to live a merely physical life, devoid of intellectual activity or social intercourse; and vegetative is used to describe ‘an organic body capable of growth and development but devoid of sensation and thought’” [3].

Jennett and Plum coined the term to emphasize the “vegetative or noncognitive components of the nervous system.” Plum mentioned that “the term persistent autonomic state could have been employed almost equally well,” but the term “would have been less understood by the patient’s family.” Jennett and Plum were also quick to point out the following:

*New methods of treatment may, by prolonging the lives of patients with conditions which were formerly fatal, result in situations never previously encountered. And new situations call for new names if they are to be accurately understood and discussed... There is a group of patients who never show evidence of a working mind. This concept may be criticized on the grounds that observation of behavior is insufficient evidence on which to base a judgment of mental activity; it is our view that there is no reliable alternative available to the doctor at the bedside, which is where decisions have to be made.*

Jennett in his classic work (Fig. 2 [8]) noted that patients demonstrated no evidence of awareness of themselves or their environment. They were incapable of interacting and absence of sustained, reproducible, purposeful, or voluntary behavioral responses to visual, auditory, tactile, or noxious stimuli. They had mostly preserved cranial nerve (pupillary, oculocephalic, corneal, vestibulo-ocular, gag, and cough) reflexes. They demonstrated the presence of sleep–wake cycles (and often had eyes open during the day). Careful examination of the eye movements carries a high priority. Eyes may open wide when the patient is touched, but visual pursuit—smoothly following an object—is absent or momentary and not reproducible. Visual fixation is absent, although it can appear later and mostly at random without other signs of improvement. A visual orienting reflex may occur with head turning when



**Fig. 2** Cover of Jennett’s book on persistent vegetative state, Cambridge University Press, used with permission

family members or nursing staff move in the room. Large objects or persons suddenly approaching the patient may cause the patient to turn the eyes briefly, suggestive of target focusing, but the response extinguishes quickly. The eyes are typically roving and nystagmoid. After the term PVS was coined in 1972, it became an established neurologic condition. It was still called *das komplette appallische* syndrome in Germany and Austria the same year [9] and was later called *wachcoma*.

PVS became an issue in legal proceedings, in which the “persistent” label of the condition was debated. In the 1990s, position articles by major US and UK medical organizations moved toward the definitions of medical treatment in these patients. Both nasogastric and intravenous fluid administration were considered forms of medical treatment. This then implied that family or a legal guardian could stop these interventions. Interruption of nutrition and hydration in children also was considered acceptable but more problematic.

To address the need for a comprehensive review, a Multi-Society Task Force formed in 1991 included representatives of the American Academy of Neurology, Child Neurology Society, American Neurologic

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Association, American Association of Neurological Surgeons, and American Academy of Pediatrics. Consultants to the task force included leading experts in traumatic head injury, anoxic brain injury, and bioethics. The review quickly gained notoriety for introducing the new term “permanent vegetative state.” “Permanent” indicated irreversibility, whereas “persistent” could mean “a condition from the past and continuing disability with an uncertain future.” The task force also finally concluded that recovery from a posttraumatic PVS was unlikely after 12 months. Recovery from a nontraumatic PVS was rare after 3 months. The Task Force on PVS noted a 70% mortality in 3 years and an 84% mortality in 5 years. All these numbers have been seriously questioned and not confirmed in modern intensive care units around the world. There was a high probability of mortality from untreated infections or overwhelming sepsis in the first 3 years. Prolonged survival can be achieved only with meticulous care and aggressive medical intervention with each complication [4, 5].

### And What Now?

In a later article, Jennett summarized the current state of the terminology:

*What attracts attention and curiosity is the dissociation between arousal and awareness—the combination of periods of wakeful eye opening with lack of any evidence of a working mind either receiving or projecting information. The advantage of the term “vegetative state” is that it simply describes observed behavior, without implying specific structural pathology [10].*

The reliability of neurological examination in PVS has withstood the test of time, although errors by nonneurologists are still numerous. There are the usual questions: Is our neurological examination reliable? Do we have better ways to assess “consciousness”? Can functional magnetic resonance imaging (MRI) scans predict recovery or provide evidence of awareness not detected clinically? This last question cannot yet be answered with certainty. Brain activation on functional

MRI (or electroencephalogram changes temporally related to verbal commands) does not necessarily prove consciousness, although some neuroscientists are convinced that functional MRI can uncover “willful brain behavior” in patients otherwise unable to show it. Functional MRI can show thalamic integrity suggestive of recovery potential. It remains to be seen if newer technology leads to more granular definitions and endotypes. Throughout the world there has been insufficient academic interest in patients in “PVS” who reside in nursing homes. But first, a workable lexicon is warranted.

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