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# A syndrome in search of a name

When Jennett and Plum in 1972 coined the term persistent vegetative state, in a *Lancet* paper subtitled 'A syndrome in search of a name' (1), they were neither the first to describe this condition nor the first to propose a name. In 1899, Rosenblath had reported a 15-year-old tightrope walker who after two weeks in coma following a fall from his wire recovered 'to become strangely awake'; he died after 8 months being tube fed in this state (2). In 1940, a German psychiatrist Kretschmer proposed the term **the apallic syndrome** to describe patients who were awake but unresponsive (3). As examples he described a case with a gunshot wound of both cerebral hemispheres and one of panencephalitis subacuta, thereby indicating that this state could result from either acute or chronic progressive brain damage. Although several authors in continental Europe have used this term (4) it has never caught on in English-speaking countries.

In 1952 an American neurosurgeon commented that when brain damage deprived patients of the intuitive and protective functions necessary for survival they rarely lived more than 2–3 weeks (5). However, he went on to describe five patients who had survived for months with periods of wakefulness without ever being aware, but he did not suggest a name for this state. In 1956, Strich reported the pathological findings in five cases from the Oxford Neurosurgical Unit who had what she called **severe traumatic dementia** (6). She commented on the similarity between the severe white matter degeneration that she found and that previously reported by Rosenblath in his case. Since then others have used the term **post-traumatic dementia** or **encephalopathy**, but these terms have never acquired the strict definition now associated with the vegetative state. In fact, in her expanded series of 20 such patients examined pathologically, Strich noted that several had spoken a few words and some had even obeyed commands during the stage of partial recovery before they died (7). Similarly, those using the apallic label frequently referred to partial or

incomplete forms of the syndrome (8). By contrast, Jennett and Plum recommended an absolute distinction between patients who did not make any consistently understandable response to those around them, whether by word or by gesture, and those who never did. The former should be regarded as very severely disabled and not as in a lesser degree of the vegetative state. Recently the terms **minimally responsive state** (9) or **minimally conscious state** (10) have emerged to describe those patients who have regained very limited conscious responses (p. 23).

The terms **permanent**, **irreversible** or **prolonged coma** or **unconsciousness** have been used at various times to describe vegetative patients, and still sometimes appear in articles (or headlines) by journalists. However, physicians now generally accept that **coma** should be confined to describing patients whose eyes are continuously closed and who cannot be aroused to a wakeful state. Of course many patients in a vegetative state following an acute insult will have been in coma for some time before regaining wakefulness, although some nontraumatic cases may become vegetative after only a day or so in coma. This is because there has been no element of widespread temporary depression of the reticular activating system that is a consistent feature of severe head injury. **Unconsciousness** is taken to imply lack of awareness of the self or the environment. The President's Commission in 1983 accepted several types of patient as having permanent loss of consciousness (11). These included those in a vegetative state (duration undefined), those in coma from acute brain damage till death, in coma from untreatable mass lesions or in the end stage of degenerative conditions such as Alzheimer's disease, and infants with anencephaly. Both patients in coma and those in a vegetative state are unconscious but this term fails to distinguish between the two because it does not acknowledge that arousal and awareness can be independently affected. Some have suggested that vegetative patients be described as having prolonged **post-traumatic unawareness** (12) or **postcomatose unawareness** (13).

These patients have sometimes been described as in a **decerebrate** or **decorticate state**. These terms are most often used to describe types of motor dysfunction rather than the mental state implied by the term vegetative. Moreover, these terms tend to imply structural lesions that do not correspond to the pathological findings in all vegetative patients. Physiologists commonly use decerebration to describe the state of animals after upper brain stem transection and this would be anatomically misleading when applied to patients in the vegetative state.

**Coma vigile** was sometimes used in the older French literature to describe some patients with severe typhus and typhoid fever, and although it is neatly descriptive of one aspect of the vegetative state it does not adequately encompass the syndrome as a whole (14).

**Akinetic mutism** was coined by Cairns et al. in 1941 (15) to describe the intermittent depression of consciousness observed in an adolescent with a brain tumour (craniopharyngioma). Her condition was one of silent immobility, with the eyes open and apparently attentive, and 'giving the promise of speech' – indeed she sometimes did whisper in monosyllables. Her state was three times reversed by aspirating fluid from the tumour. A subsequent review by Skultety (16) found this term to be used rather loosely to describe reversible disorders of responsiveness in which akinesis and mutism did not always go together – some patients spoke or used sign language. Nor were the limbs in the spastic posture associated with the vegetative state. He ascribed it to functional depression of critical amounts of the afferent or efferent systems, or of the activating reticular formation. Again, the term is descriptive of only part of the behaviour of vegetative patients and it is not an acceptable synonym.

**Neocortical necrosis** is a pathological term that applies only to the subset of vegetative patients who have suffered anoxic or hypoglycaemic damage resulting in loss of cortical neurones. The term **cognitive death** has some attraction in that it invites comparison with, but a distinction from, brain death, but the term death implies irreversibility.

The phrase **pie vegetative** was used by Arnaud et al. in 1963 to describe some survivors of head injury (17), and **vegetative survival** was one outcome category for severe head injuries reported by the Finnish neurosurgeons Vapalahti and Troupp in 1971 (18), but their condition was not clearly defined. The term persistent vegetative state (PVS) came the following year, with arguments that it was preferable to all previous names (1). Its acceptance into medical terminology in many countries probably owes much to its being one of the four categories of survival in the Glasgow Outcome Scale proposed by Jennett and Bond in 1975 (19). This scale has been widely adopted by neurosurgeons and neurologists for reporting the outcome in survivors of either traumatic or nontraumatic coma.

Persistent vegetative state (PVS) was recommended as the term of choice in the 1993 report of the American Neurological Association (20) and in the 1994 statement of the Multi-Society Task Force (21), and it has been widely adopted also by philosophers, lawyers and others outside medicine. As Jennett and Plum (1) stated, the word vegetative itself is not obscure. **To**

**vegetate** is defined in the Oxford English dictionary as 'to live a merely physical life, devoid of intellectual activity or social intercourse (1740)', and **vegetative** is used to describe 'an organic body capable of growth and development but devoid of sensation and thought (1764)'. It suggests even to the layman a limited and primitive responsiveness to external stimuli, whilst it reminds the doctor that there is relative preservation of autonomic regulation of the internal milieu of the body.

In seeking a name for this syndrome we wished to have one that did not presume a particular anatomical abnormality or pathological lesion because these vary considerably from case to case and can seldom be known with certainty at the bedside. A term that described behaviour seemed appropriate as the essence of the definition is observed behaviour, and is independent of special investigations that may not always be available, and that in any event do not show consistent abnormalities in vegetative patients (pp. 25–8). As our intention was to provide a term that would facilitate communication about this state between doctors and the patient's relatives, moralists and lawyers it seemed advantageous to have one that avoided medical jargon. Moreover such a broad descriptive term, indicating only absence of observed cognitive function, invited further clinical and pathological investigations rather than giving the impression of a problem already fully understood.

In recent years there has, however, been increasing concern about the ambiguity of the 'persistent' component of this term because it may seem to suggest irreversibility, although Jennett and Plum had made it clear that this should not be implied. Recovery of varying degrees after weeks and sometimes months in a vegetative state is now widely recognized, but confusion is evident in occasional statements that the diagnosis of PVS cannot be made until a year after an acute brain insult. This is to confuse diagnosis with prognosis. There is no doubt that the label PVS in the first few weeks after a brain insult can result in suboptimal rehabilitation efforts at a stage when active treatment is important, because recovery is still possible. For this reason expert groups in the US and Europe have suggested using only the terms 'vegetative state' and 'permanent vegetative state' (22,23,24,25). Indeed, the authoritative code of practice published by the Royal College of Physicians of London in 1996 was titled 'The permanent vegetative state' (25). This recommended using 'the vegetative state' for the condition soon after the insult, the 'continuing vegetative state' when it had lasted for more than four weeks and 'permanent vegetative state' when

it was considered to have become irreversible (by agreed criteria). In the rest of this book PVS will be used only to refer to the permanent vegetative state, but it must be accepted that this abbreviation is still widely used to mean persistent rather than permanent.

Some commentators, including the Pro-Life Committee of Catholic Bishops in the US (26), have expressed concern that the word vegetative can suggest that the patient is a vegetable and therefore subhuman, and they have urged the medical profession to seek a less discriminatory and demeaning alternative. Several physicians share this concern and some US and UK experts have suggested as an alternative 'the wakeful unconscious state' (13). They did so without much confidence that the term vegetative state was likely to be replaced because it is now so widely used by many different disciplines.

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# Diagnosis

The definition of the vegetative state and the description of its features have evolved over the years as individuals have reported surveys of patients and as various medical organizations have produced consensus statements. Criteria used for two widely quoted Japanese epidemiological surveys in the late 1970s now seem very imprecise (Tables 2.1 and 2.2), but they may have lost something in translation. However, it is clear that the definition of Sato et al. (2) allowed inclusion of patients who could obey some commands and who would therefore have been excluded by later definitions. The responses of 250 child neurologists (3) who were asked in 1991 to comment on the relative importance of ten features that had been proposed as an operational definition of vegetative state by Nelson and Bernat (4) showed a marked lack of consensus (Table 2.3). An estimate of the prevalence of the vegetative state in children in California in 1991 was based on a survey of State residents registered as developmentally disabled (5). To identify residents who might be considered to be in a vegetative state 15 items were selected from the adaptive behavioural section of the Client Development Evaluation Report form (Table 2.4). However, this form had been devised for other purposes and there must be some doubt as to how accurately this group of items corresponds with more formal definitions of the vegetative state that have emerged since then.

A committee of the American Neurological Association (ANA) (6) published a set of diagnostic criteria in 1993 (Table 2.5). Then came the 1994 report from The Multi-Society Task Force on PVS (7), which included representatives from five American professional bodies – the Academy of Neurology, the Neurological Association, the Association of Neurological Surgeons, the Academy of Pediatrics and the Child Neurology Society. The report included a list of criteria (Table 2.6), which represented codified elaborations of the more descriptive accounts given by Jennett and Plum in 1972 (8), by the American Academy of Neurology

**Table 2.1.** Epidemiological survey in Japan 1977

| Arbitrary criteria for study |
|---|
| 1. Defect of verbal and behavioural communication |
| 2. Loss of expression of intention |
| 3. Absence, or at least reduction, of emotional expression |
| 4. Urinary and faecal incontinence |
| 5. Complete loss of self-supportability |

Higashi et al. (1).

**Table 2.2.** Epidemiological survey in Japan 1978

| |
|---|
| 1. Unable to move by himself |
| 2. Able to vocalize but unable to make any meaningful speech |
| 3. Can barely respond to such a simple order as 'open your eyes', 'squeeze my hand', etc. but no further communication is possible |
| 4. Eyes can follow an object but cannot recognize it |
| 5. Unable to take a meal by himself |
| 6. Be in a state of rectal and urinary incontinence |

Based on promulgation of Japanese Neurosurgical Society, Sato et al. (2).

(AAN) in 1989 (9) and by the American Medical Association (AMA) in 1990 (10). The Quality Standards Subcommittee of the American Academy of Neurology (11) subsequently endorsed the Task Force criteria, and they have become the benchmark for American practice. In 1996, the Royal College of Physicians of London produced a statement (12) in response to a request from a parliamentary committee set up after the *Bland* case (p. 154), and this included diagnostic criteria (Table 2.7). It is to be hoped that these authoritative declarations will limit the variation in definition that was evident in some early reports.

## Features of the vegetative state

What characterizes the vegetative state is the combination of periods of wakeful eye opening without any evidence of a working mind either receiving or projecting information, a dissociation between arousal and

**Table 2.3.** Responses of 250 child neurologists to commonly used features of PVS (% of respondents)

| | Apply | Supportive | Necessary |
|---|---|---|---|
| Wakefulness without awareness | 95 | 16 | 84 |
| Eyes-open unconsciousness | 94 | 33 | 67 |
| No 'voluntary' action or behaviour | 91 | 23 | 77 |
| No 'cognitive' response | 90 | 22 | 78 |
| No 'voluntary' language | 84 | 29 | 71 |
| No commands followed | 83 | 22 | 78 |
| No sustained eye tracking | 83 | 47 | 53 |
| Intact brain stem reflexes, sleep/wake cycles | 75 | 53 | 47 |
| Breathing intact, chewing and swallowing impaired | 75 | 65 | 35 |
| Bowel and bladder incontinence | 53 | 53 | 47 |

From Ashwal et al. (3).

awareness. Following acute insults the eyes open spontaneously after a period that varies according to the mechanism of the brain insult. Head injury involves a concussive effect on the brain-stem reticular formation that takes time to recover, and it is usually 2–3 weeks, sometimes as long as 12 weeks, before the eyes open and coma ends. After nontraumatic coma, when there is no concussion, the eyes open much sooner, in more than half the patients in the first week, in some within 24 hours of the insult (13). However, in Higashi's series of 110 cases who had all been vegetative for at least 3 months, half of them for more than a year, 14% still had their eyes closed at this time and this was still so for 4% at follow-up 3 years later (1). It is exceptionally rare for true coma to be so prolonged; it never lasted more than a month in a large series of survivors of nontraumatic coma (13). It may, however, occur with a lesion in the posterior hypothalamus (14). Another possible explanation for failure of the eyes to open for a long period after head injury is that focal damage has produced bilateral third nerve lesions resulting in ptosis (paralysis of the muscle that opens the eyelids). Once the eyes do open, patients in a vegetative state have prolonged periods of being awake, alternating with sleep (from which they can be roused by vigorous stimulation). Whether this periodicity reflects normal diurnal rhythm is difficult to assess, because such patients are usually

**Table 2.4.** Items from Client Development Evaluation Report used to define vegetative patients in survey of disabled in California

1. Rolling and sitting: does not lift head when on stomach; no rolling or sitting
2. Hand use: no functional hand use
3. Arm use: no functional arm use
4. Eating: does not feed self, must be fed completely
5. Level of bladder control: no control
6. Level of bowel control: no control
7. One-to-one interaction with peers: does not enter into interaction
8. Auditory perception: does not react to sounds
9. Visual perception: does not explore visually; includes continuous staring
10. Associating time with events and actions: does not associate events and actions with time
11. Word usage: no use of words
12. Expressive nonverbal communication: no expressive nonverbal communication
13. Receptive nonverbal communication: does not demonstrate understanding of gestures (tactile or visual) or facial expressions
14. Receptive language: does not understand speech
15. Expressive language: makes no sound

From Ashwal et al. (5).

in continual light and frequently stimulated, as part of their active nursing care.

Whilst eye opening is a positive and uncontroversial feature of the vegetative state, the crux of the rest of the definition is essentially negative – the lack of any evidence of awareness, by meaningful responses or activity. However, the wide range of reflex responsiveness in vegetative patients, and the tendency for this to become more marked as time passes in most patients, can give rise to some such activity being interpreted as evidence of returning consciousness. Because some vegetative patients do recover consciousness it is important to recognize when that boundary is reached and the patient can be declared no longer vegetative but in a minimally conscious state (p. 24) or even better than that. What is clear is that some patients can regain a wide repertoire of reflex responsiveness without going on to recover any evidence of awareness when followed for months or years, whilst some 'recovered' patients may not progress beyond a state of minimal consciousness. The only detailed report of the frequency of