The permanent vegetative state

REVIEW BY A WORKING GROUP CONVENED BY THE ROYAL COLLEGE OF PHYSICIANS AND ENDORSED BY THE CONFERENCE OF MEDICAL ROYAL COLLEGES AND THEIR FACULTIES OF THE UNITED KINGDOM

The working group was convened following a recommendation of the House of Lords Select Committee on Medical Ethics that PVS should be defined and a code of practice developed relating to its management [1].

Defining the vegetative state

The Working Group recognises that the commonly used acronym 'PVS" can denote either the 'persistent vegetative state' [2] or the 'permanent vegetative state' and could thus lead to confusion. It is therefore recommended that the following terms and definitions be used:

The vegetative state

A clinical condition of unawareness of self and environment in which the patient breathes spontaneously, has a stable circulation and shows cycles of eye closure and eye opening which may simulate sleep and waking. This may be a transient stage in the recovery from coma or it may persist until death.

The continuing vegetative state (CVS)

When the vegetative state continues for more than four weeks it becomes increasingly unlikely that the condition is part of a recovery phase from coma and the diagnosis of a continuing vegetative state can be made.

The permanent vegetative state (PVS)

A patient in a continuing vegetative state will enter a permanent vegetative state when the diagnosis of irreversibility can be established with a high degree of clinical certainty. It is a diagnosis which is not absolute but based on probabilities. Nevertheless, it may reasonably be made when a patient has been in a continuing vegetative state following head injury for more than 12 months or following other causes of brain damage for more than six months [3,4]. The diagnosis can be made at birth only in infants with anencephaly or hydranencephaly. For children with other severe malformations or acquired brain damage, observation for at least six months is recommended until lack of awareness can be established.

Criteria for diagnosis of permanent vegetative state

Preconditions

- There shall be an established cause for the condition. It may be due to acute cerebral injury, degenerative conditions, metabolic disorders or developmental malformations.
- The persisting effects of sedative, anaesthetic or neuromuscular blocking drugs shall be excluded. It is recognised that drugs may have been the original cause of an acute cerebral injury, usually hypoxic, but their continuing direct effect must be excluded either by passage of time or by appropriate analysis of body fluids.
- Reversible metabolic causes shall be corrected or excluded as the cause. Metabolic disturbance may occur during the course of a vegetative state and are an inevitable consequence of the terminal stage but should have been ruled out as causative.

Clinical criteria

- 1 There shall be no evidence of awareness of self or environment at any time. There shall be no volitional response to visual, auditory, tactile or noxious stimuli. There shall be no evidence of language comprehension or expression.
- There shall be the presence of cycles of eye closure and eye opening which may simulate sleep and waking.
- 3 There shall be sufficiently preserved hypothalamic and brain stem function to ensure the maintenance of respiration and circulation.

These **three** clinical requirements shall **all** be fulfilled for the diagnosis to be considered.

Other clinical features are:

- There will be incontinence of bladder and bowel, spontaneous blinking and usually retained pupillary responses and corneal responses. The response to ice water caloric testing will be a tonic eye movement which can be conjugate or dysconjugate.
- There will not be nystagmus in response to ice water caloric testing, the patient will not have visual fixation, be able to track moving objects with the eyes or show a 'menace' response.
- There may be occasional movements of the head and eyes towards a peripheral sound or movement

and there may be movement of the trunk and limbs in a purposeless way; some patients may appear to smile and the eyes may water, there may be a 'grimace' to painful stimuli. There may be startle myoclonus. These motor activities shall be inconsistent, non-purposeful and explicable as a reflex response to external stimuli. Deep tendon reflexes may be present and reduced, normal or brisk; plantar responses may be flexor or extensor; there may be clonus and other signs of spasticity. There may be roving eye movements.

Differential diagnosis (Table 1)

It is essential to distinguish the vegetative state from brain stem death, coma and the locked-in syndrome. The differentiation of these conditions is on clinical grounds; there is no evidence at present that electroencephalography, evoked potentials, computed tomography (CT) of the cranium or magnetic resonance imaging (MRI) of the cerebrum improve upon the clinical diagnosis. Patients who are in a permanent vegetative state may show changes of cortical atrophy and hydrocephalus on CT head scan, and positron emission tomography (PET) will show a reduction in cerebral metabolism; but neither finding is diagnostic of the permanent vegetative state.

The time course

There is evidence that the factors which influence the prognosis of patients in a continuing vegetative state are the cause of the condition and the length of time for which it has continued. In patients who are in a continuing vegetative state following causes other than head injury there is very little hope of recovery of sentience after three months and none after six months. In patients who are in a continuing vegetative state after head injury the chances of recovery after six months are extremely low and, after 12 months non-existent [3, 4]. It is suggested that, whenever head injury is present, even when there is additional severe trauma, the longer of these time intervals be taken before the continuing vegetative state is termed 'permanent'.

Thus, the diagnosis of the permanent vegetative state should not be made before six months following non-head injury brain damage or 12 months following head injury.

The management of the patient in a vegetative state

Medical care Prior to the diagnosis of a permanent vegetative state it is imperative that patients have a high quality of care with appropriate nursing or home care and that oxygenation, circulation and nutrition are maintained and complicating factors such as hypoglycaemia and infection corrected. Until there is firm scientific evidence that treatment, in terms of specific medical, physiotherapeutic or rehabilitative activities improves the outcome of patients in a continuing

vegetative state it is a matter of clinical judgement as to the most appropriate measures, their application and the length of time they should be pursued. The medical staff must advise the relatives and carers of the situation during the continuing vegetative state.

Examination When the diagnosis of a permanent vegetative state is being considered it is obligatory that the patient should be examined by two medical practitioners experienced in assessing disturbances of consciousness. They should undertake their own assessment separately and should write clearly the details of that assessment and their conclusion in the notes. They must ask medical and other clinical staff and relatives or carers about the reactions and responses of the patient and it is important that the assessors shall take into account the descriptions and comments given by relatives, carers and nursing staff who spend most time with the patient. The medical practitioners shall separately perform a formal neurological examination and consider the results of those investigations which have been undertaken to identify the cause of the condition. It is helpful for nursing staff and relatives to be present during the examination; their role as responsible witnesses who spend a much longer time with the patient than can the medical practitioners must be recognised.

Re-assessment It is to be emphasised that there is no urgency in making the diagnosis of the permanent vegetative state. If there is any uncertainty in the mind of the assessor then the diagnosis shall not be made and a reassessment undertaken after further time has elapsed. The most important role of the medical practitioner in making the diagnosis is to ensure that the patient is not sentient and, in this respect, the views of nursing staff, relatives and carers are of considerable importance and help.

Final definitive diagnosis and decisions concerning life support

When the diagnosis of a permanent vegetative state has been established by (a) identification of the cause for the syndrome; (b) the clinical state of the patient; and (c) the lapse of time, recovery cannot be achieved and further therapy is futile. It merely prolongs an insentient life for the patient and a hopeless vigil for relatives and carers. The clinical team of doctors and nurses, augmented when necessary by colleagues, should formally review the clinical evidence. The decision, when made on full evidence that the situation is, in lay terms, 'hopeless' should be communicated sensitively to the relatives who are then given time to consider the implications, including the possibility of withdrawing artificial means of administering food and fluid [5,6,7]. At present the courts require, as a matter of practice, that the decision to withdraw nutrition and hydration, resulting in the inevitable death of the patient, should be referred to the court before any action is taken [8]. A decision to withdraw other life

Condition	Vegetative state		Coma		Brain stem death		Locked-in syndrome
Self-awareness	Absent		Absent		Absent		Present
Cyclical eye opening	Present		Absent		Absent		Present
*Glasgow coma scale	A4,B1-4,C1		A1-2,B1-4,C1-2		A1,B1-2,C1		A4,B1,C1
Motor function No purposeful movement		No purposeful movement		None or only reflex spinal movement		Eye movement preserved in the vertical plane and able to blink volitionally	
Experience of pain	No		No		No		Yes
Respiratory function	Normal		Depressed or varied		Absent		Normal
EEG activity	Polymorphic delta or theta —sometimes slow alpha		Polymorphic delta or theta		Electrocerebral silence or theta		Normal or minimally abnormal
Cerebral metabolism	Reduced by 50% or more		Reduced by 50% or more		Absent or greatly reduced		Minimally or moderately reduced
Prognosis	Depends on cause and length		Recovery, vegetative state, or death within 2–4 weeks		No recovery		Depends on cause though recovery unlikely
[‡] Glasgow Coma Scale:	A	Eye opening	В	Motor fun		C	Verbal
	1 No response 2 To pain		1 No respon 2 Extension 3 Flexion				None Grunts
							Inappropriate words
	3 4	To voice Spontaneously	4	Flexion		4	Confused

sustaining medication such as insulin for diabetes may also need to be referred to the courts because the legal position on this is uncertain. By contrast, decisions not to intervene with cardio-pulmonary resuscitation or to prescribe antibiotics, dialysis and insulin are clinical decisions. Further, those responsible for the patient's care must take account of, and respect, the patient's own views when known, whether these are formally recorded in a written document (or advance directive) or not [9]. When the medical team is agreed on the course to be taken the relatives should be counselled and their views sought, but (subject to court involvement) the decision is for those professionals who have the responsibility for the care of the patient.

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