

The Vegetative State

Guidance on diagnosis and management

Report of a working party of the Royal College of Physicians



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1 Introduction

Background

- 1.1 This guidance has been compiled to replace the recommendations published by the Royal College of Physicians in 1996,¹ in response to requests for clarification from the Official Solicitor. The guidance applies primarily to adult patients and older children in whom it is possible to apply the criteria for diagnosis discussed in the body of the document.

Wakefulness without awareness

- 1.2 Consciousness is an ambiguous term, encompassing both wakefulness and awareness. This distinction is crucial to the concept of the vegetative state, in which wakefulness recovers after brain injury without recovery of awareness.²⁻⁵

Definitions

The vegetative state

- 1.3 A patient in the vegetative state (VS) appears at times to be wakeful, with cycles of eye closure and eye opening resembling those of sleep and waking. However, close observation reveals no sign of awareness or of a 'functioning mind': specifically, there is no evidence that the patient can perceive the environment or his own body, communicate with others, or form intentions. As a rule, the patient can breathe spontaneously and has a stable circulation. The state may be a transient stage in the recovery from coma or it may persist until death. The vegetative state can follow a variety of severe insults to the brain, most commonly traumatic or hypoxic-ischaemic brain injuries.
- 1.4 The terms 'wakefulness' and 'awareness' require further clarification.

Wakefulness

- 1.5 Wakefulness refers to a state in which the eyes are open and there is a degree of motor arousal; it contrasts with sleep, a state of eye closure and motor quiescence. There are degrees of wakefulness. Wakefulness is normally associated with conscious awareness, but the VS indicates that wakefulness and awareness can be dissociated. This can occur because the brain systems controlling wakefulness, in the upper brainstem and thalamus, are largely distinct from those which mediate awareness.⁶

Awareness

- 1.6 Awareness refers to the ability to have, and the having of, experience of any kind. We are typically aware of our surroundings and of bodily sensations, but the contents of awareness

2 Criteria for diagnosis of the vegetative state

Preconditions

- 2.1 The following preconditions must apply before diagnosis of the VS can be considered.
- 1 The cause of the condition should be established as far as is possible. It may be due to acute cerebral injury, degenerative conditions, metabolic disorders, infections or developmental malformations.
 - 2 The possibility that the persisting effects of sedative, anaesthetic or neuromuscular blocking drugs are responsible in whole or part should be considered. Drugs may have been the original cause of an acute cerebral injury, usually hypoxic, but their continuing direct effects must be excluded either by the passage of time or by appropriate laboratory tests.
 - 3 The possibility that continuing metabolic disturbance is responsible for the clinical features must be considered and excluded. Metabolic disturbances may of course occur during the course of a VS.
 - 4 The possibility that there is a treatable structural cause should be excluded by brain imaging.

Clinical criteria

- 2.2 The following criteria are usually met.
- 1 The key requirement for diagnosis is that there must be no evidence of awareness of self or environment at any time; no response to visual, auditory, tactile or noxious stimuli of a kind suggesting volition or conscious purpose; no evidence of language comprehension or meaningful expression. These are all necessary conditions for the diagnosis.
 - 2 There are typically cycles of eye closure and eye opening giving the appearance of a sleep-wake cycle.
 - 3 Hypothalamic and brainstem function are usually sufficiently preserved to ensure the maintenance of respiration and circulation.
- 2.3 Criteria 2 and 3 are usually satisfied by patients in the vegetative state but, unlike the first criterion, they are not obligatory (thus, for example, a patient with cerebral injuries sufficient to cause the vegetative state might, incidentally, have third nerve palsies preventing eye opening, or injuries to the chest or medulla affecting respiratory function).

- *People with life-long severe disabilities* – Some people with severe intellectual disabilities, commonly accompanied by severe physical disabilities, have limited capacity to respond to the outside world; but those close to them are clear that they do communicate and are aware, and may indeed have a rich internal life. Such people should not be classed as vegetative.
- *Locked-in syndrome* – Locked-in syndrome results from brainstem pathology which disrupts the voluntary control of movement without abolishing either wakefulness or awareness. Patients who are ‘locked in’ are substantially paralysed but conscious, and can communicate using movements of the eyes or eyelids.
- *Coma* – Coma is a state of unconsciousness in which the eyes are closed and sleep-wake cycles absent. Coma is usually transient, lasting for hours or days: the VS is one possible outcome.
- *Death confirmed by brainstem death testing* – This implies the irreversible loss of all brain stem functions: it is followed by cardiac arrest, usually within hours or days, despite intensive care.

2.6 The above distinctions are made primarily on clinical grounds. Brain imaging with computed tomography (CT) or magnetic resonance imaging (MRI) often helps to clarify the cause of these clinical syndromes, but the findings on imaging are not specific. Cerebral atrophy is commonly seen in patients in the VS.

2.7 Sophisticated techniques used to assess cortical function – positron emission tomography (PET), electroencephalography (EEG), magnetoencephalography (MEG) and evoked potential (EP) studies – can be used to shed light on the physiology of the VS, but are not yet routine diagnostic tools. Their use is not required for diagnosis of the VS, which remains essentially clinical.

The time course

2.8 The prognosis of patients in the persistent VS is influenced by age, the underlying cause of the VS and its current duration. People in a VS one month after trauma stand a slightly better than even chance of regaining awareness; in cases of the VS due to non-traumatic causes, fewer than one-fifth of people in a VS at one month recover awareness. The chances of regaining awareness fall as time passes. Beyond one year following trauma, and beyond six months in non-traumatic cases, the chances of regaining consciousness are extremely low. In the very small number of well documented cases, recovery has usually been to a state of exceptionally severe disability.³ Patients in the persistent VS should therefore be observed for 12 months after head injury (traumatic brain injury) and six months after other causes before the VS is judged to be ‘permanent’.

3 Management of the vegetative state

Medical care

- 3.1 Patients in the VS require a high quality of nursing care to avoid the preventable complications of their highly dependent state. Standard measures include adequate nutrition, often via a percutaneous endoscopic gastrostomy (PEG) tube, good skin care, passive joint exercises to minimise contractures, suction where necessary to help avoid aspiration, careful management of the doubly incontinent bladder and bowel, and attention to oral and dental hygiene. Until there is firm scientific evidence that treatment, in terms of specific medical, physiotherapeutic or rehabilitative activities, improves the outcome of patients in a VS, the use of these measures is a matter of clinical judgement. The medical and nursing staff must keep the relatives and carers well informed throughout the course of the VS.

Assessment

- 3.2 Both the initial diagnosis of the VS and the subsequent diagnosis of the permanent VS should be made with great care. There is evidence that the VS has been diagnosed in error.^{11,12} The explanations for misdiagnosis include confusion about the meaning of the term, inadequate observation in suboptimal circumstances, failure to consult those who see most of the patient (especially family members), and the inherent difficulty of detecting signs of awareness in patients with major perceptual and motor impairments.
- 3.3 Thus, when the diagnosis of the permanent VS is being considered, it is essential that the patient should be examined by at least two doctors both of whom are experienced in assessing disorders of consciousness. They should take into account the views of the medical staff, other clinical staff (including clinical neuropsychologists, occupational therapists and physiotherapists with expertise in assessing disorders of consciousness), carers and relatives about the patient's reactions and responses. They should undertake their clinical assessments separately and write the details of their assessments and their conclusions in the notes. They should consider the results of the investigations which have been performed to clarify the cause of the condition. As the patient's physical position can affect responsiveness, it may be valuable to assess the patient in more than one position. It may be helpful for nursing staff and relatives to be present during the examination.

Re-assessment

- 3.4 There is no urgency in making the diagnosis of the permanent VS. If there is any uncertainty in the mind of the assessor, the diagnosis should not be made and the patient should be reassessed after an interval. Structured observation may help to reveal signs of awareness in doubtful cases.¹³⁻¹⁵ The key consideration in making the diagnosis is whether the patient might be aware to some degree: it is always important to seek the views of nursing staff, relatives and carers on this issue.

Appendix 1

Checklist for the diagnosis of the permanent vegetative state

The diagnosis of the permanent vegetative state requires prolonged observation, experience in the assessment of disorders of consciousness, and discussion with relatives and with medical and paramedical staff. It cannot be made by following a simple protocol. However, we hope that this checklist will be of some practical help by highlighting the key steps on the way to the diagnosis.

- 1 Has at least one year elapsed since the onset in cases due to head injury?
or
- 2 Have at least six months elapsed since the onset in cases due to other causes?
- 3 Has the cause been established? (It should be established 'as far as possible'.)
- 4 Have effects of drugs been excluded?
- 5 Have effects of metabolic disturbance been excluded?
- 6 Has the possibility of a treatable structural cause been excluded by brain imaging?
- 7 Have two doctors who are experienced in the assessment of disorders of consciousness, independently confirmed that there is no evidence of:
 - awareness of self or environment
 - purposeful movement
 - any attempt to communicate?
- 8 Do medical staff, nursing staff and other therapists agree?
- 9 Do family and friends agree?*
- 10 In case of doubt, has an expert clinical neuropsychological assessment been carried out?

Where the answer is 'yes' to all these questions, the diagnosis of the permanent vegetative state is confirmed.

First assessing doctor:	Second assessing doctor:
Name _____	Name _____
Qualifications _____	Qualifications _____
Signature _____	Signature _____
Date _____	Date _____

* Sometimes, even when all other members of the family and friends of the patient are in agreement, one individual may be unable to agree with the general conclusion that the patient lacks awareness. Any evidence of awareness should be examined very seriously, but in these circumstances the continuing disagreement of one individual with the conclusion of health professionals and others close to the patient is not a bar to the diagnosis of the permanent vegetative state.

in spastic muscle tone in her when they move her arms to wash her, making their task a little easier.

Comment: The combination of discriminating responses to environmental and bodily stimuli with a changing level of responsiveness over the past two months suggests the possibility of a minimally conscious state, and therefore a further two-month period of observation is required.

3. A man aged 38 who was found unconscious after taking an overdose of antidepressant drugs

A year ago, two months after admission to hospital, he had shown early signs of emergence from coma (following people with his eyes, reliably squeezing his right hand to command and occasionally mouthing single words when in apparent discomfort). However, at this point he developed a serious urinary tract infection during which he had a prolonged episode of status epilepticus, requiring anticonvulsant drug treatment. Subsequently only reflex patterns of responses have been seen. Two months ago his anticonvulsant drugs were withdrawn, since when there has been no change in his level of responsiveness.

Comment: The history suggests a further episode of hypoxic brain damage during the episode of status epilepticus. Two months is ample time for any sedative effects of the anticonvulsant drugs to have disappeared and the observed pattern of responsiveness is now consistent with permanent VS.

4. A woman aged 25 injured in a high-speed road accident, resulting in coma and the need for assisted ventilation lasting three weeks

Three months after spontaneous respiration had returned, she started to show movements of the right arm and leg and grinding of the teeth, principally in response to noxious stimuli (eg having a blood sample taken, being stretched by the physiotherapist, or having a foot-drop splint applied in order to prevent calf muscle contractures). At times she would consistently follow certain members of staff and family members with her eyes for periods of up to five minutes but at other times her eyes would remain closed. Ward staff have noticed that each time she has been formally assessed by the visiting neurologist, her eyes have closed within 1–2 minutes of his arrival and she then makes no signs or responses indicating any conscious awareness and her face remains expressionless until after he has left. When the physiotherapist first enters her room she usually grinds her teeth and the limbs stiffen. When in bed or sat in a chair she has not been observed to make any limb movements for which a volitional purpose could be deduced, but in the gymnasium, moving her to an upright position or rolling her over a ball reliably induces an increased state of alertness with more visual tracking and apparent 'protective' volitional (not reflex) movements of the right arm; in this aroused state she might smile when teased by the therapists.

Comment: In this case, eye closure might sometimes represent a volitional withdrawal response to unwanted stimuli. The evidence of consistent discrimination in the range of responses made to different individuals varying with the state of arousal suggests the presence of awareness.

Are brain scans needed to make the diagnosis of vegetative state?

Techniques like electroencephalography (EEG) and positron emission tomography (PET) are not needed to make a diagnosis of vegetative state. However, simple brain imaging should be performed to ensure that there is no unexpected treatable cause (this will generally have been done anyway).

What movements do people in a vegetative state usually make?

In most people in a vegetative state, there are cycles of eye closing and eye opening giving the appearance of sleeping and waking. Most people have normal circulation and can breathe without aid.

Patients in a vegetative state may make a range of spontaneous movements including chewing, teeth grinding, swallowing, roving eye movements and purposeless limb movements. They may make facial movements such as smiles or grimaces, shed tears, or make grunting or groaning sounds for no obvious reason. They may react automatically by reflex responses to various stimuli: for example, they may gag when being fed, or their eyes may move when their head is turned from side to side. Things like noise can also cause a response, such as faster breathing, grimaces or movement of limbs. Their eyes may turn fleetingly to follow a moving object or person, or towards a loud sound, and their hands may appear to grasp objects placed in them. None of these responses require conscious awareness.

In addition to occasionally following moving objects or people with their eyes, patients in a vegetative state have also been known to utter a single inappropriate word. Behaviour of this kind should lead to a careful search for awareness, but responses like these may occur because small 'islands' in the brain have survived but they are no longer able to work together to generate awareness.

What can hospital staff do for patients in a vegetative state?

Staff will carefully monitor and record patients' responses on a daily basis.

They will give high-quality nursing care to avoid any preventable complications developing. This care will include things like adequate nutrition (often via a feeding tube), good skin care, joint exercises, careful management of bowel and bladder incontinence, and oral and dental hygiene.

Then all possible treatable causes of the state must have been ruled out. The patient then has to be examined by at least two doctors who are both experienced in assessing this kind of disorder. They must conduct their assessments independently of one another, and write their results in the notes. As the patient's physical position can affect responsiveness, the doctors may assess the patient in more than one position. They should take into account views about the patient's reactions and responses from the medical staff involved (eg nurses), from other clinical staff (eg physiotherapists with experience in making such assessments), and from those close to the patient.

If the two doctors think that the patient is in a permanent vegetative state, then they will discuss their opinion with other medical staff, nursing staff, therapists and those close to the patient. If there is broad agreement, then a diagnosis of permanent vegetative state will be made, meaning that the evidence indicates that the patient will never recover their awareness.

The diagnosis of permanent vegetative state will not be rushed. If there is any doubt about it, the patient will be re-assessed at a later date. In doubtful cases, the staff may use structured observation (watching closely for specific time periods at set intervals) to try to see if there are any signs of awareness in the patient.

How are decisions made about withdrawing nutritional support?

When the diagnosis of a permanent vegetative state has been made, it may be decided that recovery cannot reasonably be expected and that continued support and treatment is futile. Prolonging an insentient life would have no benefit for the patient, and would mean a hopeless vigil with major emotional costs for those close to the patient.

In these circumstances, the medical team, with the help of colleagues when required, will formally review the evidence. After confirming the diagnosis of a permanent vegetative state, they will discuss this with those close to the patient, and give them time to consider the implications, including the possibility of withdrawing the means of providing food and water.

At present, any decision to stop giving food and water by tube must be referred to the courts before any action can be taken.

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