

Misdiagnosis of the vegetative state: retrospective study in a rehabilitation unit

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See also
by Gaitard

Abstract

Objective—To identify the number of patients who were misdiagnosed as being in the vegetative state and their characteristics.

Design—Retrospective study of the clinical records of the medical, occupational therapy, and clinical psychology departments.

Setting—20 bed unit specialising in the rehabilitation of patients with profound brain damage, including the vegetative state.

Subjects—40 patients admitted between 1992 and 1995 with a referral diagnosis of vegetative state.

Outcome measures—Patients who showed an ability to communicate consistently using eye pointing or a touch sensitive single switch buzzer.

Results—Of the 40 patients referred as being in the vegetative state, 17 (43%) were considered as having been misdiagnosed; seven of these had been presumed to be vegetative for longer than one year, including three for over four years. Most of the misdiagnosed patients were blind or severely visually impaired. All patients remained severely physically disabled, but nearly all were able to communicate their preference in quality of life issues—some to a high level.

Conclusions—The vegetative state needs considerable skill to diagnose, requiring assessment over a period of time; diagnosis cannot be made, even by the most experienced clinician, from a bedside assessment. Accurate diagnosis is possible but requires the skills of a multidisciplinary team experienced in the management of people with complex disabilities. Recognition of awareness is essential if an optimal quality of life is to be achieved and to avoid inappropriate approaches to the courts for a declaration for withdrawal of tube feeding.

Introduction

The vegetative state is a rare disorder which is diagnosed by clinical examination. The clinical features were originally described by Jennett and Plum¹ and recently further clarified by the Multi-Society Task Force on PVS² and the Royal College of Physicians.³ The main characteristics are that the patient has a sleep-awake pattern, responds to stimulation only in a reflex way, and shows no evidence of meaningful response to the environment—that is, is awake but not aware.

It has been pointed out that neurodiagnostic tests can neither confirm the diagnosis of a vegetative state nor predict the potential for recovery.^{2,4,5} Giacino and Zasler have also pointed out the limitations of clinical assessment in the identification of "internal awareness" in a patient who otherwise lacks the motor function to show their awareness.⁶

The Royal College of Physicians' report on the permanent vegetative state³ supports the view expressed by others that the diagnosis requires regular assessment

and taking into account the observations by carers and family.⁵ However, even these conditions can result in misdiagnosis. Childs *et al* reported that 37% of patients admitted more than one month after injury with a diagnosis of coma or persistent vegetative state had some level of awareness.⁷ In a group of longer term patients in a nursing home, Tresch *et al* found that 18% of those diagnosed as being in the persistent vegetative state were aware of themselves or their environment.⁸

The diagnosis of the vegetative state can have a major influence on decision making about the level of care or services provided and may lead to an application being made to the courts for a directive on withdrawal of tube feeding. Clinicians should therefore be aware of the risk of misdiagnosis and the factors associated with it.

Method

The medical, occupational therapy, and clinical psychology records of all patients admitted to the rehabilitation unit for profound brain damage between 1992 and 1995 were examined retrospectively. Patients with a referral diagnosis of the vegetative state due to acute onset brain damage were included. The unit accepts patients from throughout the United Kingdom who are over the age of 16 years, have a diagnosis of acute onset brain damage causing profound physical and mental impairment, and are medically and surgically stable. Patients being ventilated are not accepted, though patients with a tracheostomy or enteral feeding are. No limit is placed on the time since brain damage, though early admission is encouraged.

The main data collected were specialty of the diagnosing doctor; cause of the brain injury; date of first inconsistent responses indicating awareness; date when consistent meaningful responses were achieved; highest cognitive level achieved; and rating on the Rancho Los Amigos cognitive function scale.⁹ Inconsistent responses were defined as following commands in a non-reflex way fewer than nine times out of 10 within any one session; consistent responses were defined as following commands at least nine out of 10 times within a session.

Patients received two half hour occupational therapy sessions a day for six weeks to assess responses to sensory stimulation and to identify the most reliable responses to command.

The main methods of showing awareness were the ability to follow a simple command to press a buzzer switch or look at a named object. Any available movement (finger, arm thrust, shoulder shrug, head movement) which was sufficient to press a simple touch sensitive switch to control a buzzer was used. The switch is particularly suitable for those patients who are able to generate only a very small amount of movement, which might otherwise go unnoticed. Correctly positioned to provide optimal potential for the movement of particular muscle groups, the patient was then taught to press the switch once for "yes" and twice for "no." When this was consistent without prompting

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Table 1—Characteristics of patients* misdiagnosed as being in the vegetative state

| Patient | Age (sex) | Months between injury and admission | Days from admission to first follow command (consistent response) | Blind or severely visually impaired | Final rating on Rancho scale | Showed ability in: | | | | | | |
|---------|-----------|-------------------------------------|---|-------------------------------------|------------------------------|--------------------|---------------------|-----------------------|-------------------|--|--|----------------|
| | | | | | | Making choices | Spelling to command | Free word generation† | Mental arithmetic | Orientation in time, place, and person | Recalling name after 15 minutes' delay | Writing letter |
| A | 25 (M) | 20 | 7 (50) | No | 6 | + | - | - | + | - | + | - |
| B | 23 (M) | 16 | 175 (175) | Yes | 6 | + | - | - | + | + | + | - |
| C | 64 (M) | 82 | 9 (9) | Yes | 8 | + | + | + | + | + | + | + |
| D | 20 (M) | 19 | 6 (6) | Yes | 8 | + | + | + | + | + | + | + |
| E | 55 (F) | 73 | 15 (26) | Yes | 7 | + | + | + | + | + | + | + |
| F | 21 (M) | 15 | 12 (21) | Yes | 8 | + | + | + | + | + | + | + |
| G | 39 (M) | 59 | 8 (42) | No | 5 | + | - | - | - | - | - | - |
| H | 21 (F) | 11 | 8 (36) | No | 6 | + | + | + | + | - | - | - |
| I | 43 (M) | 7 | 16 (18) | Yes | 6 | - | + | + | + | + | + | + |
| J | 29 (M) | 10 | 2 (2) | Yes | 8 | + | + | + | + | + | + | + |
| K | 32 (M) | 8 | 6 (45) | Yes | 8 | + | + | + | + | + | + | - |
| L | 19 (M) | 6 | 8 (46) | No | 5 | + | + | + | - | Person | - | - |
| M | 43 (M) | 6 | 4 (14) | No | 7 | + | + | + | - | + | + | ? |
| N | 18 (M) | 10 | 6 (8) | No | 5 | - | - | - | - | - | - | - |
| O | 38 (F) | 6 | 11 (54) | Yes | 8 | + | + | + | + | + | + | + |
| P | 56 (M) | 10 | 40 (46) | Yes | 7 | + | + | + | - | + | + | + |

*Only 16 of the 17 patients are included because the relatives of one patient did not wish any details to be included in the study.

†Generating own words using letter by letter spelling with buzzer (see text).

the patient was given simple biographical questions, with answers provided by the family, to test whether responses were appropriate.

More recently a "listener scanning" technique has been introduced. The therapist speaks the letters of the alphabet and the patient operates the buzzer when the desired letter is reached. Patients start by selecting letters of simple words such as "bed" to command before moving on to generating their own words and messages.

For visually impaired patients a variety of scanning devices, including computer based programs which speak the letters of the alphabet, were used.

Evidence of cognitive functioning had to be confirmed by at least two members of the team. In nearly all cases most members of the team and the family became proficient in the use of the communication technique.

To avoid confusing spontaneous recovery with misdiagnosis we did not include those patients admitted within six months of their brain damage, since spontaneous recovery is not uncommon during this period. Patients were considered to have been misdiagnosed if they could follow commands consistently within our initial assessment period of six weeks. Although we have previously shown in another group of our patients that recovery can occur after six months post injury,¹⁰ none of that group showed responses within the first six weeks of admission.

One patient (patient B) fell outside this criterion but was included since it was felt that he had been misdiagnosed by even the experienced team on the unit and therefore warranted inclusion.

Consent to publication was given by the six patients who were mentally alert enough to give consent; relatives of 10 of the remaining 11 patients gave permission to the publication of details of individual cases.

Results

Of 97 patients with profound brain damage admitted to the unit between 1992 and 1995, 40 (41%) had been diagnosed by the referring clinician as being in a vegetative state.

OUTCOME OF VEGETATIVE GROUP

Of the 40 patients diagnosed as being in the vegetative state, 10 (25%) remained vegetative, 13 (33%) slowly emerged from the vegetative state during the rehabilitation programme, and 17 (43%) were con-

sidered to have been misdiagnosed as vegetative. The identification of misdiagnosis was more common in the later part of the study period: two were recognised in 1992, one in 1993, four in 1994, and 10 in 1995.

Table 1 shows that seven patients had been considered to be vegetative for longer than one year, with three of these being treated as being vegetative for between four and seven years. The 10 other patients had been considered to be vegetative for between six and 12 months.

CAUSE OF BRAIN DAMAGE

Ten (59%) of the misdiagnosed group had sustained brain damage from trauma, four (23.5%) from anoxia, two (12%) from vascular causes, and one (6%) from encephalitis.

PERCEPTUAL DISORDERS

One striking finding was that 11 (65%) of the "misdiagnosed" patients were either blind or very severely visually impaired, with visual field defects or visual perceptual disorders, or both. Since all patients followed verbal commands it is assumed that none were deaf or had severe hearing impairment.

PHYSICAL DISABILITY

All 17 misdiagnosed patients were at the "severe" level of the Glasgow outcome scale,¹¹ being totally physically dependent for all care needs. For 15 (88%) patients, pressing a buzzer was the only functional movement, though one patient later developed an ability to point with a finger and another patient became able to write words; the other two patients communicated by eye pointing.

COGNITIVE OUTCOME

Fifteen of the 17 misdiagnosed patients showed the ability to respond to a command (such as "Press the buzzer" or "Look at the [object]") in a non-reflex way within 16 days of admission. The exception was patient B; we did not identify his responses until 25 weeks after his admission, though it was obvious from subsequent conversations with him that he had not been vegetative for some time. This patient was admitted with very severe joint contractures which required surgical release and a prolonged physical management programme

before he could be seated appropriately in a special seating system. Only when he was satisfactorily seated was it identified that he had a slight shoulder shrug which could be used for communication purposes.

In all patients, inconsistent responses were followed by consistent responses at a time between the same day and 43 days later (table 1). All patients were able to correctly answer simple biographical questions that required a yes/no response such as "Do you have two sons?" or "Do you come from [place]?" The numbers of questions requiring positive and negative responses were equal.

Table 1 shows the number of patients achieving consistent (at least 9 out of 10 correct responses) reproducible responses to various cognitive tasks. Fifteen patients were able to make choices, such as selecting their previously preferred music tapes from a choice of three. Twelve (71%) patients were able to spell out their own short messages using the listener scanner technique.

Eleven patients (65%) were able to carry out simple one stage and two stage mental arithmetic tasks such as "What is 10 divided by 5?" and "What is 8 minus 4 minus 1?" Eleven patients (65%) were oriented in time, place, and person, and one patient was only oriented in person. Thirteen patients (76%) were able to recall a name from a choice of three options; and eight (47%) patients were able to use listener scanner techniques to write a letter to relatives.

Two patients used eye pointing, rather than the buzzer, to discriminate between a choice of two objects, pictures, colours, numbers, letters, and words and to make choices about their daily care. One patient, although reliable and consistent using yes/no buzzer responses, did not progress beyond this level; he withdrew responses during treatment sessions.

The 17 misdiagnosed patients ranged (table 1) in cognitive ability from level 5 (confused, inappropriate, non-agitated) to level 8 (purposeful-appropriate) on the Rancho scale⁹; in other words, from aware but severely cognitively impaired to nearly normal. All but one of the patients had been referred by a hospital consultant, and there were records in most cases of the diagnosis being made by a neurologist, neurosurgeon, or rehabilitation specialist—all of whom could have been expected to have experience of vegetative state.

Discussion

These findings show how difficult it can be, even for experienced clinicians, to diagnose cognitive ability in the presence of profound physical disabilities. The Royal College of Physicians' recommendations emphasise the importance of seeking information from carers and family about possible responses and reactions.³ As far as we could identify, none of the professional carers had recorded any evidence of meaningful responses.

The vegetative state is extremely uncommon and therefore few clinicians gain the necessary experience for appropriate assessment and clinical management. Even those clinicians who see a number of such patients are rarely responsible for, or trained in, the longer term management of brain damaged people.

EMERGENCE FROM THE VEGETATIVE STATE

It could be argued that the clinical team had introduced a successful treatment programme to bring about the emergence from the vegetative state. Although a significant change in clinical management often took place (such as improvement of nutritional state, better postural management, provision of specialist seating support systems, control of infection, and the introduction of sensory regulation programmes), it is our opinion that these patients were not vegetative at the time of admission. For some of these it was obvious shortly after admission to the unit but for others it gen-

Key messages

- Assessing awareness in brain damaged patients who have profound physical disabilities requires skill, time, and repeated observations
- Many patients who are misdiagnosed as being in the vegetative state are blind or have severe visual handicap; thus lack of eye blink to threat or absence of visual tracking are not reliable signs for diagnosing the vegetative state
- Any motor activity, no matter how slight, that can be used for communication by the profoundly disabled patient should be identified at an early stage and repeated at regular intervals
- Identification of awareness in the presence of profound and complex neurological disabilities requires the skills of a multidisciplinary team experienced in long term management of disability due to brain damage

erally took several weeks to confirm that they were aware, presumably due to the need for them to accommodate to communicating again, especially through technological aids and after a long period of non-communication.

All of the misdiagnosed patients were severely physically disabled, often with contractures, and were anarthric. Since demonstration of awareness needs a motor response, such profound physical disability complicates assessment of awareness. The very high prevalence of severe visual impairment, to the best of our knowledge not previously reported, is an additional complicating factor since clinicians making the diagnosis of the vegetative state place great emphasis on the inability of the patient to visually track or blink to threat.

IDENTIFYING AWARENESS

It is of note that more "misdiagnoses" were detected by the team in 1995 than the previous years, although there has been no obvious change in referral characteristics over the period. This increase in detection rate is probably due to the team's increased sensitivity owing to the accumulation of experience over several years, coupled with the development of more effective assessment methods.

On this unit the level of the patient's awareness is nearly always identified first by the occupational therapists and then by the clinical psychologist, and only later is communication achieved by the other members of the team. This has important implications since it is usually on the basis of bedside observations by a physician or surgeon that decisions are made to refer a patient for specialist treatment programmes, or decisions are made to apply to the courts to withhold or withdraw medical treatment or artificial nutrition and hydration.

It is disturbing to think that some patients who were aware had for several years been considered to be, and treated as being, vegetative. It must be extremely distressing to be aware but unable to make contact with family or clinical carers. It is possible that we have been referred an unrepresentative sample of patients. This is possible since the unit is the only one in Britain specialising in the management of this group of people. However, similar figures for misdiagnosis have been described for patients at an earlier stage after brain damage,⁷ and figures of about half our level have been reported for patients in long term care.⁸ These findings are not a criticism of the referring clinician but emphasise both the complex nature of profound brain damage and the difficulties of caring for patients experienced by staff who see very few patients in this condition.

We also emphasise that a quarter of those diagnosed as vegetative by the referring team remained vegetative and were almost certainly, from our experience, likely to remain so. These findings are therefore not an argument against the withdrawal of artificial nutrition and hydration but do emphasise the importance of accurate diagnosis of the vegetative state being made after expert assessment and provision of a rehabilitation programme by a very experienced team.

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Commentary: The importance of patients' consent for publication

Richard Smith

This paper made us think hard about the issue of getting consent from patients for publication of identifiable information about them. Obvious identifying information has been removed, but these patients cannot be considered to be truly anonymised. After much discussion, the editorial team and the authors agreed that we should get witnessed consent for publication from those patients in the series who were capable of giving consent and agreement to publication from the relatives of those not able to give informed consent. The authors did this, and one set of relatives did not agree to publication. The details of the patient whose relatives did not agree have been removed from the paper.

Getting consent for the publication of a series of patients in medical journals has not been usual, and readers, future authors, and other editors may find it useful if we describe the thinking that went into getting consent for this paper. We start from the premise that information that emerges from the doctor-patient relationship is confidential information. The doctor-patient relationship is built on trust, including the trust of patients that information they disclose will remain confidential. This information should not be revealed to third parties in an identifiable form unless patients give consent or there is overwhelming reason to break the confidentiality without patients' consent. It is hard to imagine circumstances in which publication in a medical journal would be so important that confidentiality could be broken without the patient's consent, although minimising the danger of just one patient being misdiagnosed as being in a vegetative state might to some people justify breaking patient confidentiality. Another premise is that everybody has a right to privacy. This right applies beyond the doctor-patient relationship. That right may be overridden in some circumstances, but why should publication of details about you in a medical journal override your right to privacy?

A problem with publication of a series of cases as in this paper is that these are unusual cases, and they have all been managed at some stage in one hospital. Some people will be able to recognise these patients. The media have already gathered details. Clearly there is much discussion about the definition of identifiable—do we mean identifiable to the average reader, one or two readers, the patient, other family members, friends, or the nursing staff who cared for the patient? We have experienced cases where only the patient has recognised the report but has nevertheless been deeply disturbed.

There were arguments against getting consent. The first argument is that under English law, relatives cannot give consent for patients who cannot themselves give

consent. But would we want to publish details of a patient if the relatives did not want us to? We decided we would not. A second argument is that by requesting consent we complicate the process of publishing medical papers. This may result in some papers not being published. We could not convince ourselves that these steps would stop important papers being published. Most patients, we have discovered, will give consent. A third argument is the slippery slope argument. Where will we draw the line? If we have an epidemiological paper with data on 5000 individuals, will we require consent from all of these people? The answer will always be no when, as is usual, the data are presented in a combined form: no individual is identifiable.

The fourth argument came in various forms but was essentially the argument that getting consent was a lot of trouble: the patients are now scattered; it might take a long time to get consent from some of them; it would be necessary to be clear who could give consent and who couldn't; publication would be slowed; and the media were already pursuing the authors and editors. Expediency is not a good reason to override ethical issues.

So our conclusion was that we must get consent from patients and agreement to publication from relatives, and we are grateful to the authors for doing that.

Problems of confidentiality arise in many series of cases, and authors and editors need to sensitise themselves to the problems. There is a particular problem with family pedigrees, in which information may be disclosed on relatives of patients. Indeed, the possibility arises that family members may learn something important to themselves for the first time through publication of pedigrees in medical journals.

A current issue where problems may arise surrounds the publication of data on Creutzfeldt-Jakob disease. The British government announced at the end of June that it would publish data on definite and probable cases every three months. There will obviously be intense interest in these data, particularly in the cases of the new variant of the disease. And this is a topic where in the early days of what may prove an epidemic every case will matter greatly. Leaving out just one or two cases from the data may confuse the developing picture. There were three deaths among people with new variants of the disease in 1995 and six so far in 1996. "Clearly," said Sir Kenneth Calman, the chief medical officer, "with the present intense interest in the disease, there is a need to put statistics into the public domain more frequently. Personal details will not be included. There is a difficult balance to be struck between the legitimate need for public information and patient confidentiality."