Assessment and Treatment of People With a Disorder of Consciousness: An Account of Some Recent Studies

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This paper describes a number of studies looking at the assessment and treatment of people with disorders of consciousness (DOCs). These include patients in a coma, patients in a vegetative state (VS), and patients in a minimally conscious state (MCS). It is important to distinguish between these latter 2 states, as referral decisions may well be different for the 2 groups. A new version of a measure to assess patients with DOCs is described. The effects of posture on awareness are then considered. Provided that medical conditions do not contraindicate this, assessments should be carried out when the patient is in an upright position, as the majority of patients with DOCs show more behaviors when assessed on a tilt table or a standing frame. The third study looks at the effect of modafinil, an agent that maintains wakefulness and promotes attention and concentration, on levels of consciousness. Those with a traumatic brain injury (TBI) would appear to respond more to this than those whose DOC is caused by hypoxia. The fourth study compares the delayed recovery of 2 groups of patients, those with a TBI and those with hypoxic brain damage. Not surprisingly, those who showed delayed recovery of consciousness were far more likely to have sustained a TBI than hypoxic brain damage. The paper concludes with a case study of a man who had a DOC for 19 months before regaining consciousness and proceeding to regain skills so that he is now leading a reasonably normal life.

Keywords: disorder of consciousness, vegetative state, minimally conscious state, brain injury, rehabilitation

To be conscious, one must be both awake and aware. Obviously, when we are asleep with our eyes closed, we are not aware. Awareness requires, in the first instance, open-eyed wakefulness. To be aware, however, involves more than just wakefulness. Awareness “is the ability to have, and the having of, experience of any kind” (Royal College of Physicians [RCP], 2013, p. 2). People with a disorder of consciousness (DOC) are those in a coma, a vegetative state (VS), or a minimally conscious state (MCS): They have problems with both wakefulness and awareness. The RCP (2013) reports that coma is a “state of unrouseable unresponsiveness, lasting more than 6 hours in which a person cannot be awakened; fails to respond normally to painful stimuli, light or sound; lacks a normal sleep–wake cycle; and does not initiate voluntary actions” (p. 3). Thus, those in a coma are neither awake nor aware.

The VS is defined by evidence of sleep–wake cycles but, critically, no evidence of purposeful response to sensory or cognitive stimuli. The RCP (2013) defines this as a “complete absence of behavioral evidence for self—or environ-
mental awareness” (p. 3). Thus, people in a VS are awake because they open their eyes, but they are not, apparently, aware. The same guidelines (RCP, 2013) recognize that families do not like the term vegetative and frequently misinterpret it as vegetable (see Wilson, Dhamapurkar, & Rose, 2016). The European Task Force on Disorders of Consciousness recommended that the term unresponsive wakefulness syndrome (UWS) replace the term vegetative state. To date, this has not happened.

The MCS is defined by preserved eye opening and sleep-wake cycles and critically reproducible but inconsistent evidence of purposeful response to sensory and/or cognitive stimuli (Giacino et al., 2002). In the words of the RCP (2013) guidelines, the MCS is “characterized by inconsistent, but reproducible, responses above the level of spontaneous or reflexive behavior, which indicate some degree of interaction with their surroundings” (p. 3). There is behavioral evidence of self- or environmental awareness. Therefore, people in an MCS are awake and have minimal awareness.

The main causes of DOCs are severe traumatic brain injury (TBI), oxygen deprivation as a result of a stroke or TBI, cardiac and pulmonary arrests, hanging, drowning, suicide attempts from carbon monoxide poisoning, embolisms, drug overdoses, encephalitis, severe hypoglycemia, and advanced dementia or other neurodegenerative diseases (RCP, 2013; Wilson, 1996).

Most people recover from a coma within 4–6 weeks of their injury or illness, either regaining full consciousness or progressing to a VS or MCS. During the acute phase, they are likely to be assessed using the Glasgow Coma Scale (GCS; Teasdale & Jennett, 1976). This scale is the most widely used measure of wakefulness and awareness in the acute hospital environment. It consists of three subscales: Best Eye Response, Best Motor Response, and Best Verbal Response. Although Teasdale and Jennett (1976) suggest that the three subscales should be individually reported, it is common for the scores on each subscale to be summed, providing a maximum score of 15. Although a score of 7 or less on the GCS is consistent with the behavioral definition of the comatose state, someone in a VS could theoretically also score 7 or less on the scale, and someone in an MCS could score as little as 8. It is this lack of clear distinction—and thus sensitivity to the critical behavioral repertoire of these conditions of impaired consciousness—that has prompted the creation of alternative behavioral scales designed to detect specifically the key behaviors distinguishing these patients.

It is important to differentiate between these conditions because management, referral, and rehabilitation decisions might be made on the basis of the cerebral function and capacity inferred from such measures. It is often difficult, however, to differentiate between VSSs and MCSs, and a high rate of misdiagnoses has previously been highlighted (Gill-Thwaites, 2006; Schnakers et al., 2009). Neuropsychologists can help distinguish between the two. McMillan (1997) describes a young woman who was thought to be in a VS but when carefully observed showed evidence of cognition. McMillan assessed this patient because the health authority was considering going to court to request permission to remove feeding and hydration. A further assessment of the same woman a few days later (Shiel & Wilson, 1998) confirmed McMillan’s findings. This evidence of cognition was in contrast to some medical opinion at the time. One of the main reasons for the difference of opinion was that the psychological observations had taken place over 2 days rather than a half hour or so, and systematic trials were conducted. The case was immediately dropped. McMillan and Herbert (2004) presented a 10-year follow-up of this patient. She was living in the community with 24-hr support, was speaking, and was initiating conversation; had a spontaneous sense of humor; expressed clear and consistent preferences; used an electric wheelchair; ate solid food; and could drink through a straw.

This early work contributed to the now-accepted assessment procedure for such patients. Patients with impaired consciousness following brain injury are now assessed over a prolonged period of observation (>6 weeks) by at least two independent doctors supported by the observations of all professionals and family members in contact with the patient (Bates, 2005). It is accepted that observations should take place at different times of the day when the patient is in different positions (elevated or rolled vs. supine) and that the observations of everybody in contact with the patient should be recorded and taken into account.
Assessment of People With DOCs

There are a number of assessment tools designed for use with adults who have very severe brain injury. These include the Glasgow Outcome Scale (Jennett & Bond, 1975), the Rancho Los Amigos Scale (Malkmus, Booth, & Kodimer, 1980), the Neurobehavioural Rating Scale (Levin et al., 1987), the JFK Coma Recovery Scale—Revised (JFK CRS–R; Giacino, Kalmar, & Whyte, 2004; Kalmar & Giacino, 2005), the Sensory Modality Assessment and Rehabilitation Technique (SMART; Gill-Thwaites & Munday, 2004), and the Wessex Head Injury Matrix (WHIM; Shiel, Wilson, McLellan, Horn, & Watson, 2000). The RCP (2013) recommends the JFK CRS–R, the SMART, and the WHIM. These are probably the most widely used assessment tools for people with DOCs in the United Kingdom.

The JFK CRS–R has 25 hierarchically arranged items with six subscales (Auditory, Visual, Motor, Oromotor, Communication, and Arousal). Scoring is based on the presence or absence of specific behavioral responses to stimuli presented in a standardized manner, from reflexive responses to cognitively facilitated responses. The JFK CRS–R helps differentiate between the diagnosis of VS and that of MCS.

The SMART identifies the consistency and quality of behavioral response to all sensory modalities. Using these results, the tool guides treatment plans to optimize the patient’s potential. The WHIM is a behavioral observation tool. It consists of 62 items in a roughly hierarchical order from least to most difficult, which provides a sequential framework of behavior covering a patient’s level of responsiveness and interaction with his or her environment. In clinical practice, these two measures provide complementary information.

This paper describes some recent studies addressing the assessment and management of patients with a DOC. We look at (a) the WHIM, which is currently undergoing a revision; (b) the effects of posture on awareness; (c) whether a drug that maintains wakefulness and promotes attention and concentration can improve awareness; (d) differences in delayed recovery between those with a DOC due to a TBI and those with a DOC due to hypoxic brain damage; and (e) a case study to illustrate an unexpectedly good outcome in a man who had a DOC for 19 months.

The WHIM: Origins and a New Version

The WHIM was developed following observations of 88 patients recovering from severe TBI and was a joint project between psychologists, a physiotherapist, an occupational therapist, and a neurologist or medical rehabilitation specialist. Three types of behaviors were observed: (a) naturally occurring behavior such as a patient scratching his or her nose, (b) a response to something in the environment such as looking at a nurse entering the room, and (c) an elicited response such as tracking a flashlight. In total, 162 different items of behavior were observed from the 88 patients. As some behaviors only occurred once and others seemed to overlap with each other, 62 items of behavior were selected for the final scale (Shiel et al., 2000). There are several advantages to this scale. Improvements in functioning following severe brain injury are often very gradual and unless one is looking for them, they can be missed. Failure to notice can lead to poor motivation among staff and poor patient care, whereas good measures to identify progress can influence clinical practice. The WHIM requires no formal training, and it is flexible; the stimuli employed are meaningful, and it is possible to use stimuli personalized to the individual patient. It can be used in any environment, and it has the potential to discriminate between a VS and an MCS.

Wilson, Coleman, and Pickard (2008) compared the scores of 20 patients with a DOC, 10 of whom were in a VS and 10 who were in an MCS. All were assessed on the GCS and the WHIM. Each patient scored 10 on the GCS and therefore looked very similar to all the other patients in the study. On the WHIM, however, patients’ scores were distinctly differentiated from each another, suggesting that the WHIM is more sensitive than the GCS. This finding was in agreement with those of Majerus, Van Der Linden, and Shiel (2000) and Shiel et al. (2000), who argue that the WHIM is more sensitive than the GCS at measuring recovery and monitoring subtle changes.

Despite its use in both clinical and research settings, it has become clear that the WHIM...
needs a few modifications. It was developed with patients in the acute stage yet is often used with those in the chronic stage. Clinicians feel that the order of the 62 items needs some adjustment. The original WHIM included only TBI patients, yet clinically it is used for those with a DOC from other causes; in addition, because it was believed that patients with a TBI and hypoxic patients may show a different order of recovery, the order of items may need to be changed depending on the cause of the DOC. Furthermore, there are no guidelines regarding how long to wait for a response (and some people in a VS or an MCS may take a long time to respond), there are no guidelines as to how to present stimuli, and there is an overreliance on visual behaviors. Work on the new version is well under way. The research consists of two parts: a retrospective study (completed) and a prospective study (Shiel, Leahy, Morrissey, Leonard, & Wilson, 2014). In the retrospective study WHIM score sheets from several different centers were scrutinized in order to determine a) which was the most likely order for each behavior to occur and b) whether those with a TBI showed a different order of recovery than those with a non-TBI. More than 700 sheets were scored, with the mean number of assessments for each patient being more than 11. The new order is slightly different for those with or without a TBI. The prospective study (still ongoing) involves the observation of over 100 patients with the reordered WHIM to see if the predictions from the retrospective study are borne out. The new WHIM is expected to be published in 2017.

In addition to a proper assessment, appropriate and sensitive management of people with a Prolonged Disorder of Consciousness (PDOC) is essential. Therefore, the following sections address this issue.

The Effects of Posture on Awareness

Elliott et al. (2005) published a paper titled “Effect of Posture on Levels of Arousal and Awareness in Vegetative and Minimally Conscious Patients: A Preliminary Investigation” (p. 298). Twelve patients—five in a VS and seven in an MCS—were assessed with the WHIM when supine and when upright on a tilt table or standing frame. Wilson, Dhamapurkar, Tunnard, Watson, and Florschutz (2013) replicated and extended these findings by including a third position: sitting. Sixteen patients (eight in a VS and eight in an MCS) with mixed aetiologies were assessed, and their scores in three different positions—supine, sitting, and standing—were compared. Several assessments were conducted in each position, and the best score in each position was used for comparison. Most patients (75%) showed more behaviors when in the upright position compared to lying down ($p < .003$). Twelve patients showed more behaviors when standing compared to being in a supine position (75%, $p < .05$). Ten showed more behaviors when sitting compared to being in a supine position (63%, $p = .32$), and 11 patients showed more behaviors when standing compared to sitting (69%, $p = .13$). For people who had different sitting and supine scores, their higher score was more likely to be when sitting ($p = .05$). For people who had different standing and supine scores, their higher score was more likely to be when standing ($p = .002$). When data for the VS and the MCS patients were looked at separately, only 4 of the 8 VS patients showed a higher standing score compared to their sitting score ($p = 1.00$), whereas 7 of the 8 MCS patients showed more behaviors when standing than when sitting ($p = .07$). These findings are similar to those seen in the study reported by Elliott et al. (2005). Both studies found that 75% of patients showed more behaviors when assessed in a standing position. The amount of difference found, however, was not as large in the Wilson et al. (2013) study as that seen in the Elliott et al. (2005) study. This may be because the patients in the Wilson et al. study were in the chronic stage, with most sustaining their brain injury many months (in one case, 36 years) earlier. Another possibility is that all people in the Elliott et al. (2005) study had sustained a TBI, whereas patients in the 2013 study had several different reasons for their low-awareness states. Nevertheless, both studies found that patients with DOCs are likely to be more responsive when assessed in an upright position.

The 2013 study also looked at WHIM scores when patients were assessed while seated in a wheelchair. With regard to sitting, 62.5% of patients were more responsive when assessed sitting in a wheelchair ($p < .05$) than in a supine position, and almost 69% were more responsive if assessed in an upright position compared to
sitting. This was particularly true for patients in an MCS, where 87.5% did better if assessed on a tilt table or standing frame compared to sitting, suggesting that positional changes can have an effect on the level of arousal and awareness among patients in a VS or MCS. The following year, this study was extended even further (Wilson, Rose, Dhamapurkar, & Florschutz, 2014). In a paper titled “Improving the Behavioral Responses of Vegetative and Minimally Conscious Patients Through Changes in Position,” four more patients in a VS and a further seven in an MCS were assessed in the same situations. Similar results were obtained. There was a statistically significant difference between the three positions. Most patients showed more behaviors when in the upright position and the fewest behaviors when lying down (p < .001). This was particularly true for those in an MCS, where standing scores were higher than lying-down scores (p = .004), but there was no difference between lying down and sitting (p = .13) or sitting and standing (p = .71). However, using the sign test, 10 of the 11 people (91%) in the MCS group who had different sitting and lying-down scores had a higher sitting score, and those with different sitting and lying-down scores were more likely to have a higher score for sitting than for lying down (Sidak adjusted p = .035). Nine of the 12 people who had different standing and sitting scores (75%) in the MCS group had a higher score for standing. Eleven of the 12 (92%) people who had different standing and lying-down scores in the MCS group had a higher score for standing.

In conclusion, these postural studies confirm earlier findings that positional changes can have an effect on the level of arousal and awareness among patients in states of low awareness. Provided that patients are medically stable, an upright position will help increase their level of alertness. These findings can impact therapy sessions. As suggested by Wilson et al. (2013), feeding could commence with a semireclined position; the tilt table could be used during grooming sessions, and patients could gradually be taught to tolerate being upright. This would also give patients the opportunity to benefit from auditory and visual input through use of a mirror and spoken communication. It is very difficult to receive this input when in bed being cared for in a supine position. Use of verbal prompts and hand-over-hand guidance would add additional benefits in terms of tactile and proprioceptive feedback. In physiotherapy, patients could initially begin standing on a tilt table at 45° of inclination and then gradually progress up to 90° depending on the level of tolerance. Adopting this approach would potentially enhance weight bearing and could also help prevent the early onset of osteoporosis.

### The Effect of Modafinil on Level of Consciousness: A Pilot Study

Cognitive enhancers are, arguably, the main focus for improving cognitive skills. Hopefully, these drugs not only alter but also significantly improve mental ability. One such enhancer is modafinil, a medication used for the treatment of narcolepsy, which maintains wakefulness and promotes attention and concentration. Of course, there are other drugs used to improve wakefulness for patients with a DOC, such as zolpidem (Thonnard et al., 2013; Tucker & Sandhu, 2015). However, the patients we were asked to assess had been prescribed modafinil, so we focused on the efficacy of this medication.

Dhamapurkar, Wilson, Rose, and Florschutz (2015a) wanted to determine whether modafinil improved the level of consciousness in brain-injured patients.

Previous studies (e.g., Ishizuka, Murotani, & Yamatodani, 2010; Joo, Tae, Jung, & Hong, 2008; Kumar, 2008) have indicated that modafinil has the potential for cognitive enhancement and has been studied for its primary effect in sleep regulation among healthy individuals. However, the potential of modafinil as a cognitive enhancer in brain-injured patients has yet to be fully explored.

Twenty-four patients were included in the study, 13 of whom had sustained a TBI and 11 of whom had suffered hypoxic brain damage. Their levels of consciousness were measured by the WHIM and by behavioral observations from staff. The measures were taken before and after the administration of modafinil, with the exception of two patients who were already on modafinil when admitted. Dosage was between 100 and 400 mg/day. In addition to looking at changes in scores following the administration of modafinil, scores for TBI and hypoxic patients were compared. All patients also received...
treatment as usual. The effects of other medications, infections, nutrition, and other confounding factors were considered.

Of the patients diagnosed with TBI, nine showed a significant improvement after the introduction of modafinil. Cognitive improvements were noticed within the domains of wakefulness, awareness, concentration, following commands, and tracking. It was also noted that four of the TBI and three of the hypoxic patients progressed from a VS to an MCS when the modafinil dose was increased. Two TBI patients showed no changes before and after being administered modafinil. The two cases who were already on modafinil when admitted continued to improve, so their improvement could be due to natural recovery. The results from the pilot study suggested that modafinil appears to be beneficial for enhancing cognition in patients with DOCs. TBI patients seemed to benefit more in certain cognitive domains when compared to hypoxic patients. These findings, however, need to be replicated with a larger group of patients. As for those with hypoxic damage, it is possible that these patients might respond to a larger dose of modafinil. This could be the focus of a future study.

**Differences in Delayed Recovery Between Those With a DOC Due to a TBI and Those With a DOC Due to Hypoxic Brain Damage**

The recent guidelines from the RCP (2013) suggest that those who are in a VS for more than 12 months can be considered to be in a permanent VS. Yet we know that a small percentage do show considerable improvement. How many patients who remain in a state of low awareness for several months go on to regain consciousness? There are conflicting answers to this question, ranging from 9% after 6 months to 0% after 12 months (Giacino & Kalmar, 1997) to less than 14% (The Multi-Society Task Force on PVS, 1994) to 20% (Giacino & Whyte, 2005) and to a high of 33% (Luauté et al., 2010). Most studies seem to agree that the outcome for patients in a VS is worse than for those in an MCS and that those who are in a state of low awareness following TBI do better than those with anoxic or cerebrovascular damage (Giacino & Whyte, 2005).

Dhamapurkar, Wilson, Rose, and Florschutz (2015b) looked at patients with a DOC lasting 12 months or more to see if recovery was more likely for those who had sustained a TBI compared to those whose DOC resulted from other causes (mainly hypoxia). All patients (28 total) with a DOC admitted to a rehabilitation center over a 3-year period were assessed with the WHIM and the Disability Rating Scale (DRS; Hall, Hamilton, Gordon, & Zasler, 1993) to determine if they had emerged from a DOC. Four of the patients with hypoxic brain injury died. Eighteen patients remained with a DOC for another 12 months or more (12 in a VS and six in an MCS). Five patients (18%) emerged from a DOC—that is, they showed delayed recovery. Of these, four had sustained a TBI, and one had hypoxic damage. These results suggest that survivors of a TBI are more likely to show delayed recovery than those with hypoxic brain damage. As 18% of patients do not remain in a persistent state of low awareness, however, this study implies that we should not discontinue rehabilitation of such patients too early.

**A Case Study to Illustrate an Unexpectedly Good Outcome in a Man Who Had a DOC for 19 Months**

In October 2011, Gary, then 28 years old, was beaten with metal poles and pieces of wood while protecting his father John from a gang of about 30 teenagers. He sustained several fractures, including left orbital and left temporoparietal skull fractures, and developed an acute subdural hematoma. A craniectomy was performed. He lost the sight in his left eye, and blood had accumulated behind his right eye. He emerged from a coma after 1 week and was responding to his family but then had a fall and deteriorated. A Computerized Tomography (CT) scan showed hydrocephalus, and a shunt was inserted. Gary later developed seizures and cellulitis over the valve.

Four months postinjury, Gary was admitted to a rehabilitation center. At this point, his eyes were open, but he was not responding to stimuli. Thus, he was in a VS. He was referred for a neuropsychological assessment, and the WHIM was used as the assessment tool. Gary was seen on nine occasions from 4 to 7 months postinjury. He was observed in several different locations at different times of the day and with
different people present. The locations included Gary’s room, the physiotherapy room, and the art therapy room. He was seen with his mother, other members of his family, the art therapist, two physiotherapists, his occupational therapist, and others. Observation sessions lasted between 20 and 30 min. Gary was usually awake when assessed and remained quiet, although sometimes he made a sound (a hiccup or an oh sound), and sometimes his left arm would shake. The report concluded that he was in a VS. He was reassessed at 11 months postinjury when the report said no real change. He remained in a VS for nearly 14 months.

Throughout this time, Gary had daily physiotherapy and occupational therapy. He was regularly put onto a tilt table so he was in an upright posture and often received sensory stimulation. In addition, he received music and art therapy twice a week. In these sessions, the therapists used hand-over-hand guidance. Eventually, Gary emerged from a VS into an MCS and remained in this state for an additional 5 months. In addition to the therapies just described, ongoing treatment included gradual reduction of antiepileptic drugs, cranioplasty, and maintenance of hydration and nutrients. Observations, monitoring, and assessments continued.

After 19 months, Gary emerged from the MCS and continued to improve. He received inpatient rehabilitation for an additional 17 months (3 years in total). Toward the end of this 3-year period, Gary moved into a supported living flat within the grounds of the rehabilitation center as part of his discharge plan. He started spending longer periods at his mother’s house, during which time he attended the rehabilitation center daily for outpatient rehabilitation. He had several operations during this time, including a cranioplasty, an operation to restore the sight in his right eye, and an operation on his foot to enable him to learn to walk again. He applied to college to study electrical engineering; he is walking, talking, and making jokes; his speech is good; and his memory is not too bad. On some neuropsychological tests, he is in the average or even the above-average range (e.g., block design). Many people like Gary are sent to a nursing home. If this had happened, he would probably have been left in a low-awareness state for the rest of his life. Fortunately, he was referred to a rehabilitation center where he received excellent care and continuous rehabilitation. The main message, once again, is do not stop rehabilitation for people in a state of DOC too soon. Gary’s story is told in detail in Wilson et al. (2016).

Conclusions

A number of studies looking at the assessment and treatment of people with DOCs are portrayed. Following a description of a coma, a VS, and an MCS, assessment of people with DOCs is considered. This paper highlights a number of ethical considerations that those working in the field of DOC cannot ignore—namely, accuracy of diagnosis, provision of suitable care, and access to neurorehabilitation for an appropriate length of time. It is essential to assess people with a PDOC properly because improvements in functioning following severe brain injury are often very gradual and can be missed if they are not looked for. This may lead not only to low motivation among staff and poor patient care, but measures to identify progress can also influence and improve clinical practice. It appears that patients who have sustained a TBI may show a different order of recovery compared to those who have sustained hypoxic brain damage, so one would not expect to see the same behaviors occurring in the same order for the two groups of patients. In addition, it is important to distinguish between those in a VS and those in an MCS, as referral decisions may well be different for the two groups. Provided that medical conditions do not contraindicate this, assessments should be carried out when the patient is in an upright position. Those in an MCS may show more behaviors when assessed on a tilt table or a standing frame, and they may benefit from a cognitive enhancer such as modafinil. We know that a small percentage of those in an MCS for 12 months or longer are more likely to regain consciousness compared to those in a VS and that this is more likely to be true for TBI patients than for hypoxic patients. A case study of a man who sustained severe brain damage following an assault and who was in a state of disordered consciousness for 19 months is used to illustrate one example of a good recovery. We should ensure that all those with a DOC receive the best possible care and that rehabilitation does not stop too soon.
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Received November 5, 2015
Revision received April 7, 2016
Accepted April 25, 2016