Vegetative state, minimally conscious state, akinetic mutism and Parkinsonism as a continuum of recovery from disorders of consciousness: an exploratory and preliminary study

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Summary

The aim of this study was to review the usefulness of clinical and instrumental evaluation in individuals with disorders of consciousness (DOC). Thirteen subjects with severe acquired brain injury (ABI) and a diagnosis of DOC were evaluated using the Coma Recovery Scale in its revised version (CRS-R) and a new global disability index, the Post-Coma Scale (PCS). These instruments were administered both by a neutral examiner (professional) and by a professional in the presence of a caregiver. All patients were also scored using the International Classification of Functioning, Disability and Health (ICF).

A statistically significant correlation between CRS-R and PCS was demonstrated. However, there also emerged significant differences in responsiveness between professional versus caregiver-professional assessment using the two scales. The emotional stimulation provided by significant others (caregivers) during administration of DOC evaluation scales may improve the assessment of responsiveness.

KEY WORDS: disorders of consciousness, evaluation scales, minimally conscious state, vegetative state

Introduction

Clinical diagnosis of disorders of consciousness

Owen et al. (1) first described the case of “hidden islands of consciousness” in a young woman who sustained a severe head injury in a traffic accident and was afterwards diagnosed with vegetative state (VS). Although the patient was still unresponsive and unable to communicate five months after the accident, functional MRI showed that she retained some ability to process language and to produce some mental imagery on request, similarly to healthy subjects. In this case report, electroencephalography (EEG) showed some slow alpha frequencies, as can occur in locked-in syndrome (LIS) (1,2). Locked-in syndrome is a neurological condition, due to a ventral pontine lesion, in which patients are quadriplegic, mute, conscious and aware. Unlike patients with VS, they are able to communicate by means of vertical gaze, and/or upper eyelid movements (3), and their EEG activity is generally normal. However, some LIS patients are unable to communicate by eyelid movements because of palsy that extends to ocular motility (4-6). In such cases, which could possibly include the patient described by Owen and co-workers (1,2,5), clinical differentiation between VS and LIS may be very difficult.

It has been reported that some VS patients pass through a locked-in-like syndrome during recovery of consciousness. This process does not necessarily occur in the presence of structural brainstem lesions, given that diffuse axonal injury (DAI), too, can cause a functional disconnection syndrome (5,7). This possible evolution of the clinical picture may not be recognized, especially if the first attempts at functional communication by means of blinking occur several months after the onset of coma, evolving into VS.

Thus, it can be concluded that awareness (content of consciousness) cannot always be excluded on the basis of lack of interaction with the environment, even in chronic VS. The minimally conscious state (MCS) has been described as a condition in which the subject obeys simple commands inconstantly, or recovers eye tracking or functional use of objects while remaining unable to communicate (8,9). The MCS may follow coma or the VS as a continuum of consciousness recovery. Akinetic mutism (AM), first described as secondary to diencephalic lesions, consists of severe quadriplegia, mutism, akinnesia, visual fixation and pursuit (10). It is a rare condition that has been described as a subcategory of the MCS (11). Complete or nearly complete loss of spontaneity and initiative, resulting in uniform reduction of action, ideation, speech and emotion, are peculiar to AM (12). AM patients sporadically follow commands and

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perform after a long latency. The differential diagnosis between AM and MCS may be difficult, since the two conditions generally share the features of diffuse and severe motor deficit, fluctuations in the obeying of simple orders and the presence of eye tracking.

As recently reported, central thalamic deep brain stimulation (DBS) [a specific therapeutic approach to treat Parkinson’s disease (PD)] was found to improve the arousal regulation of functionally connected, but inconsistently active, cerebral networks, which may be present in some MCS patients but not in VS patients (13). In the VS patient described by Schiff and co-workers (13), who remained in the MCS for six years following a traumatic brain injury (TBI), bilateral thalamic DBS produced improvements in intelligible verbalization and functional limb control. Indeed, the clinical description of this case report seems to recall AM. The observed improvements in arousal level, motor control, and response initiative might reflect direct activation of frontal cortical and basal ganglia systems. These blurred boundaries between different disorders of consciousness (DOC) may explain the high frequency of misdiagnosis between VS and MCS (14,15).

To summarize, differential diagnosis among VS, MCS, AM and LIS is not as simple as reported in the international literature (9,16). Indeed, even though the recovery of visual fixation and pursuit is not universally considered equivalent to recovery of the ability to obey simple commands, this visual behavior is, nevertheless, an interactive behavior with the environment.

Similarly, psychomotor agitation and aggressiveness are frequent behaviors in individuals with severe brain injury who may still be unable to follow simple orders. Agitated patients often show stereotyped aggressive behaviors towards themselves or others. In these cases, the patients, because of extreme attentive lability or oppositional behaviors, are rarely able to obey simple commands or to interact with the environment by functional communication, even though the above behavioral features seem to be a favorable predictive factor for consciousness recovery (17). According to the definition of VS, i.e., “lack of any understandable behavior in response to external stimulus or inner need” (16,18), aggressiveness may also be interpreted as the result of physical, emotional or mental discomfort that the patient is unable to express otherwise. This is the reason why agitated non-communicative subjects should be diagnosed as MCS rather than VS, even if they are not able to follow commands.

In conclusion, VS can be defined as a name in search of a universally accepted definition rather than a syndrome “in search of a name”, as suggested by Jennett and Plum (16).

Pain perception in individuals with DOC

Two main studies on pain perception in VS patients, evaluating activation of specific areas by means of positron emission tomography (PET), show contradictory results. Laureys (19) demonstrated that the painful electrical stimulation of the medial nerve activated only primary CNS areas involved in pain perception, such as the brainstem, thalamus and primary somatosensory cortex. Using the same procedure, Kassubek (20) showed the activation of somatosensory cortical areas, either primary or secondary, including the anterior cingulate which is responsible for the affective and cognitive components of pain perception. The discrepancy between the above studies may depend upon the different amount of time that elapsed between injury and recording in the two populations of VS patients, being one month in the first and up to 18 months in the second. Furthermore, the disconnection between the secondary and primary cortical areas involved in pain perception, demonstrated at one month post-injury, may reflect the persistence of cerebral edema and possible diascysis and disconnection syndrome in the early phase after injury.

Central chronic pain after severe TBI, the most frequent cause of VS and MCS, has been widely described (21,22). As mentioned above, hyperpathy and neurogenic pain may well be responsible for psychomotor agitation during recovery from DOC (17) and contribute to the difficult evaluation of responsiveness in these agitated subjects.

Parkinsonism during and after DOC

Parkinsonian symptoms during recovery from DOC, previously defined as prolonged post-traumatic coma (17,23,24) or from VS, MCS and AM after severe TBI are very common, although rarely reported. The most frequent features are hypokinesia and rigidity, associated with amimia, non-extinguishable naso-glabellar reflex, seborrhea of the face, hypersalivation (sialorrhea) and extrapyramidal dysarthria (25,26). These parkinsonian features may improve after L-Dopa treatment along with different degrees of consciousness recovery, especially if the drug is used during the early recovery phases (27). Amantadine and dopamine agonists also show some efficacy in improving parkinsonian features and consciousness recovery (28-31). There are reports of motor slowness in subjects with severe TBI and clinical features resembling post-traumatic parkinsonism, associated with changes of specific anticipatory evoked potentials (“Bereitschaft potentials”) (32,33). In most of these patients there is evidence of DAI not always involving the rostral brainstem (34). During the early phase of coma recovery, survivors of severe TBI with cerebral MRI features of DAI may show extrapyramidal signs (35) similar to those observed in multi-infarct parkinsonism (36). Indeed, similarly to TBI, a potential role of excitotoxicity has also recently been reported in PD (37). Interestingly, not all patients with post-traumatic parkinsonism respond to L-Dopa treatment, as in other secondary parkinsonisms (36,38). In particular, akinesia and rigidity, possibly secondary to multiple lesions either in the substantia nigra or the caudate-putamen area (39,40), are common features after severe TBI (41). Interestingly, motor disabilities in these subjects are the consequence of traumatic lesions in the extrapyramidal pathway rather than an effect of systemic dopaminergic degeneration (42).

Nevertheless, post-traumatic parkinsonism seems to share specific neurophysiological patterns with PD, such as blink reflex habituation changes (35). Matsuda et al. (43) described three patients with persistent VS after TBI and parkinsonian features such as rigidity and hypokinesia, all of whom improved both in parkinsonism and consciousness following L-Dopa treatment. According to Jellinger
The clinical features of post-traumatic parkinsonism often resemble those of post-encephalitis parkinsonism, both showing akinesia, rigidity, hypomimia, rare tremor, and optomotor and autonomic disorders. Indeed, both the pattern of lesions and the therapeutic efficacy of long-term L-Dopa treatment in these conditions suggest dysfunction within the striato-nigral dopaminergic system.

Since severe brain injury and post-traumatic/post-hypoxic parkinsonisms may share a common midbrain (substantia nigra)-striatal-thalamic-frontal network dysfunction, VS, MCS, AM and parkinsonism might represent a recovery continuum from prolonged DOC. Responsiveness to L-Dopa in some patients and to DBS (13) in others might depend respectively on the integrity or impairment of the dopaminergic post-synaptic receptors (5, 44).

Responsiveness evaluation

Although recent evidence has indicated the relevance of advanced neurophysiological and neuroimaging techniques in the evaluation of DOC (45-47), clinical evaluation and qualitative monitoring of VS and MCS patients remain the strongest diagnostic tools (8, 9). Among others, the Coma Recovery Scale, in its original (CRS) and revised (CRS-R) versions (48-50), is the most commonly used evaluation scale in VS and MCS patients. Although the CRS has sufficient sensitivity to monitor and detect minimal changes during consciousness recovery, it pays little attention to the subjects’ emotional responsiveness, which is, in fact, considered a supplementary item, rather than a main one. Furthermore, this instrument does not attach enough importance to responses following stimuli given by a close relative or caregiver. Differences between physician and caregiver evaluations have also been reported in Alzheimer’s disease (51). The clinical evaluation of VS and/or MCS patients may, in fact, differ when the assessment is administered by a neutral examiner or by caregivers. Such differences are not captured by the CRS, unless it is administered with the help of a close relative or caregiver. These observations suggest that there is a need for new clinical tools to evaluate behavioral emotional responsiveness in DOC patients. A Post-Coma Scale (PCS) (Appendix) (52) has been proposed as a targeted and tailored tool, additional to the CRS scoring system, for verifying the emotional responsiveness to caregiver-presented stimuli.

Materials and methods

We recruited a sample of severe acquired brain injury subjects hospitalized at the Post-Coma Unit at the Fondazione Santa Lucia Rehabilitation Hospital. The sample of 14 patients included 7 males and 6 females, with a mean age of 38 years (range 21-52 yrs), who presented with a Glasgow Coma Scale (GCS) score ≤8 (53) in the emergency room at the time of injury. The diagnosis of VS, MCS, AM or LIS was made following a period of coma due to stroke (hemorrhagic stroke=5; ischemic stroke=2), cerebral anoxia (AH=1) or traumatic brain injury (TBI=5). At the time of evaluation, the mean interval that had elapsed since injury was eight months. All subjects underwent at least one brain CT or MRI scan revealing diffuse brain damage. The evaluation performed for the purpose of this study did not interfere with the usual medical practice, or with the everyday rehabilitation therapies and was part of a routine screening of severe brain-injured subjects during their rehabilitation program. Two assessments were obtained for each patient, one with the evaluation scales administered by a neutral examiner (professional), the second performed in the presence of a caregiver. Patients were diagnosed as VS (n=9), MCS (n=2), AM (n=1) and LIS (n=1) and scored with both the CRS-R and the PCS. Demographic data are shown in Table I.

Table I - Demographic data and CRS-R and PCS scores obtained by standard professional evaluation

<table>
<thead>
<tr>
<th>Patient</th>
<th>Gender/Age</th>
<th>Etiology</th>
<th>CRS-R scores</th>
<th>PCS score</th>
<th>Diagnosis</th>
<th>Time elapsed since injury (months)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>M/30</td>
<td>TBI</td>
<td>4-5-5-2-1-2</td>
<td>81</td>
<td>MCS</td>
<td>13.5</td>
</tr>
<tr>
<td>2</td>
<td>F/21</td>
<td>TBI</td>
<td>1-1-2-0-0-1</td>
<td>51</td>
<td>VS</td>
<td>12.7</td>
</tr>
<tr>
<td>3</td>
<td>M/50</td>
<td>IS</td>
<td>1-0-0-0-0-1</td>
<td>34</td>
<td>VS</td>
<td>3.3</td>
</tr>
<tr>
<td>4</td>
<td>M/43</td>
<td>HS</td>
<td>1-2-2-2-0-2</td>
<td>63</td>
<td>AM</td>
<td>10.5</td>
</tr>
<tr>
<td>5</td>
<td>F/45</td>
<td>HS</td>
<td>1-1-2-0-0-2</td>
<td>52</td>
<td>VS</td>
<td>15.4</td>
</tr>
<tr>
<td>6</td>
<td>M/25</td>
<td>HS</td>
<td>1-1-2-1-0-2</td>
<td>39</td>
<td>VS</td>
<td>5.2</td>
</tr>
<tr>
<td>7</td>
<td>F/50</td>
<td>CA</td>
<td>1-1-2-2-0-2</td>
<td>48</td>
<td>VS</td>
<td>5</td>
</tr>
<tr>
<td>8</td>
<td>F/50</td>
<td>HS</td>
<td>4-5-1-2-2-3</td>
<td>72</td>
<td>LIS</td>
<td>10.1</td>
</tr>
<tr>
<td>9</td>
<td>F/20</td>
<td>TBI</td>
<td>2-1-2-0-0-2</td>
<td>55</td>
<td>VS</td>
<td>4.2</td>
</tr>
<tr>
<td>10</td>
<td>M/52</td>
<td>HS</td>
<td>1-0-2-1-0-1</td>
<td>43</td>
<td>VS</td>
<td>3.8</td>
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<tr>
<td>11</td>
<td>F/63</td>
<td>IS</td>
<td>2-3-2-0-0-1</td>
<td>62</td>
<td>MCS</td>
<td>10.6</td>
</tr>
<tr>
<td>12</td>
<td>M/22</td>
<td>TBI</td>
<td>0-0-1-1-0-1</td>
<td>33</td>
<td>VS</td>
<td>3.1</td>
</tr>
<tr>
<td>13</td>
<td>M/23</td>
<td>TBI</td>
<td>1-0-2-0-0-2</td>
<td>38</td>
<td>VS</td>
<td>3</td>
</tr>
</tbody>
</table>

Abbreviations: M=male; F=female; CRS-R=Coma Recovery Scale-Revised; PCS=Post-Coma Scale; TBI=traumatic brain injury; IS=ischemic stroke; HS=hemorrhagic stroke; CA=cerebral anoxia; MCS=minimally conscious state; VS=vegetative state; AM=akineti- c mutism; LIS=locked-in syndrome.
All the patients were also scored using the International Classification of Functioning, Disability and Health (ICF) (54) criteria in order to evaluate all factors related to disability, namely the patients’ own resources and features of their environment during the rehabilitation period, considered either as facilitators or barriers.

In five patients, DOC was re-evaluated after two months, again using both the CRS-R and PCS administered both by a professional and by a professional in the presence of a caregiver, to detect minimal changes in responsiveness. Pearson’s correlation was used to compare CRS-R and PCS values.

**Results**

Demographic data and CRS-R and PCS standard evaluation scores are shown in Table I.

A statistically significant correlation between CRS-R and PCS scores was demonstrated ($p_{xy}=0.9$; Fig. 1). However, there also emerged significant differences in responsiveness between professional versus professional+caregiver assessment using the two scales (Table II).

Indeed, in four of the five patients in whom CRS-R and PCS scales were repeated after an interval of two months, the evaluation by a neutral examiner (professional) differed from the professional+caregiver assessment, higher CRS-R scores being recorded when the evaluation was performed in the presence of the caregiver (Table II). Similarly, in this group of patients in whom DOC was re-evaluated after two months, four of the five subjects recorded an improved overall PCS score (change of diagnosis from VS to MCS), whereas in the other case it remained unchanged (Table III). With respect to the CRS-R, the sub-scales most sensitive to the presence of the caregiver turned out to be the “auditory function scale”, the “oromotor/verbal function scale” and the “arousal scale”. In the PCS, in which some items also include responsiveness changes following caregiver stimulation, the most sensitive sub-scales were: “vigilance”, “reaction to auditory stimulation”, “mobility of the eyes”, “emotional responsivity”, “spontaneous motility and posture”, “verbal communication”, “behavior” and “interaction with the environment”.

The patients’ ICF scores are reported in Table IV. As expected, the values reflect the severity of the functional impairment in all the subjects. In accordance with the ICF criteria, the close relatives provided information regarding their caregiving role. Twelve patients were found to receive concrete support from their families and therefore the family is to be considered a facilitator.

**Table II - Individual scores of five patients in whom the results of the assessment differed according to whether the scales were administered by the professional alone or by the professional together with the caregiver.**

<table>
<thead>
<tr>
<th>Patient</th>
<th>CRS-R scores (P)</th>
<th>CRS-R scores (P/C)</th>
<th>PCS score (P)</th>
<th>PCS score (P/C)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>3-5-5-2-1-2</td>
<td>4-5-5-2-1-2</td>
<td>79</td>
<td>81</td>
</tr>
<tr>
<td>7</td>
<td>1-1-2-0-0-1</td>
<td>1-1-2-0-0-2</td>
<td>46</td>
<td>48</td>
</tr>
<tr>
<td>9</td>
<td>3-3-3-0-1-2</td>
<td>2-3-3-0-1-2</td>
<td>57</td>
<td>60</td>
</tr>
<tr>
<td>12</td>
<td>1-3-2-0-0-1</td>
<td>1-3-2-0-0-2</td>
<td>39</td>
<td>44</td>
</tr>
<tr>
<td>13</td>
<td>1-0-2-0-0-1</td>
<td>1-0-2-0-0-2</td>
<td>38</td>
<td>38</td>
</tr>
</tbody>
</table>

Abbreviations: CRS-R=Coma Recovery Scale-Revised; PCS=Post-Coma Scale; P=professional; P/C=professional and caregiver.

**Table III - Longitudinal DOC evaluation after two-month interval**

<table>
<thead>
<tr>
<th>Patient</th>
<th>I Diagnosis</th>
<th>CRS-R scores (VS)</th>
<th>PCS score (VS)</th>
<th>II Diagnosis *</th>
<th>CRS-R scores (MCS)</th>
<th>PCS score (MCS)</th>
</tr>
</thead>
<tbody>
<tr>
<td>2</td>
<td>VS</td>
<td>1-1-2-2-0-1</td>
<td>51</td>
<td>MCS</td>
<td>4-5-5-2-1-2</td>
<td>75</td>
</tr>
<tr>
<td>4</td>
<td>AM</td>
<td>1-2-2-2-0-2</td>
<td>63</td>
<td>AM</td>
<td>3-3-5-3-1-2</td>
<td>78</td>
</tr>
<tr>
<td>7</td>
<td>VS</td>
<td>1-1-2-2-0-2</td>
<td>48</td>
<td>MCS</td>
<td>3-1-4-3-1-3</td>
<td>69</td>
</tr>
<tr>
<td>9</td>
<td>VS</td>
<td>2-1-2-0-0-2</td>
<td>55</td>
<td>MCS</td>
<td>3-3-3-0-1-2</td>
<td>60</td>
</tr>
<tr>
<td>12</td>
<td>VS</td>
<td>0-0-1-1-0-1</td>
<td>33</td>
<td>MCS</td>
<td>1-3-2-2-0-2</td>
<td>44</td>
</tr>
</tbody>
</table>

Abbreviations: VS=vegetative state; AM=akinetic mutism; MCS=minimally conscious state; CRS-R=Coma Recovery Scale-Revised; PCS=Post-Coma Scale; * mean interval=two months.
**Discussion**

The complexity of the neurological signs commonly associated with DOC demands a global disability evaluation scale that, in addition to responsiveness, also evaluates neuromotor and neuropsychological deficits. Moreover, some prognostic information such as interval from coma onset and DOC duration may also be useful. It is common experience that individuals with DOC recover their first interaction with the environment through significant emotional stimulation, mostly provided by close relatives or caregivers. However, few evaluation scales include or require the presence of meaningful persons during the examination of responsiveness in persons with DOC. In this respect, caregivers should be considered possible facilitators of expression of responsiveness, on the basis of the ICF criteria (54). Responsiveness evaluation of persons with DOC should take into account emotional stimulation provided by close relatives or caregivers, in order to avoid misdiagnosis between VS and MCS. In fact, the borders between these two conditions are subtle, and therefore diagnosis must be based on accurate evaluations. Including ICF values may be helpful to obtain better classification in DOC evaluation, especially in those individuals receiving substantial caregiving from significant others. In addition, follow-up evaluations of individuals with DOC might also seek to verify the possible efficacy of specific scales in predicting long-term outcome in severe brain-injured patients.

Among the limitations of the proposed Post-Coma Scale there is the high number of items to be administered and the need to train those who will administer the scale. Moreover, its inter-assessor reliability has still not been assessed, nor has the consistency of the results been investigated. Therefore, further studies on larger patient series are needed to validate the scale.

**Acknowledgements**

This work is supported by the European ICT Programme Project FP7-247919 (DECODER). This paper only reflects the authors’ views and funding agencies are not liable for any use that may be made of the information contained herein.

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**Table IV - ICF scores obtained in the population studied**

<table>
<thead>
<tr>
<th>Patient</th>
<th>ICF - consciousness</th>
<th>ICF - voluntary movements</th>
<th>ICF - brain injury</th>
<th>ICF - family care</th>
<th>ICF - eye tracking</th>
<th>ICF - non-verbal communication</th>
<th>ICF - ability to listen</th>
</tr>
</thead>
<tbody>
<tr>
<td>MCS</td>
<td>B110.4</td>
<td>B760.3</td>
<td>S110.370</td>
<td>E310+2</td>
<td>D110.02</td>
<td>D335.33</td>
<td>D115.04</td>
</tr>
<tr>
<td>VS</td>
<td>B110.4</td>
<td>B760.3</td>
<td>S110.470</td>
<td>E310+4</td>
<td>D110.43</td>
<td>D335.33</td>
<td>D115.34</td>
</tr>
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<td>B760.4</td>
<td>S110.473</td>
<td>E310+4</td>
<td>D110.44</td>
<td>D335.44</td>
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</tr>
<tr>
<td>AM</td>
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<td>B760.2</td>
<td>S110.470</td>
<td>E310+2</td>
<td>D110.03</td>
<td>D335.44</td>
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<td>D110.44</td>
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<td>B760.4</td>
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<td>D110.04</td>
<td>D335.44</td>
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<td>B760.3</td>
<td>S110.470</td>
<td>E310+2</td>
<td>D110.03</td>
<td>D335.88</td>
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<td>VS</td>
<td>B110.4</td>
<td>B760.2</td>
<td>S110.370</td>
<td>E310+4</td>
<td>D110.04</td>
<td>D335.33</td>
<td>D115.33</td>
</tr>
<tr>
<td>VS</td>
<td>B110.4</td>
<td>B760.2</td>
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<td>E310+3</td>
<td>D110.04</td>
<td>D335.44</td>
<td>D115.33</td>
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<td>MCS</td>
<td>B110.4</td>
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<td>VS</td>
<td>B110.4</td>
<td>B760.4</td>
<td>S110.450</td>
<td>E310+4</td>
<td>D110.04</td>
<td>D335.88</td>
<td>D115.44</td>
</tr>
</tbody>
</table>

Abbreviations: MCS=minimally conscious state; VS=vegetative state; AM=akinetic mutism; LIS=locked-in syndrome; B=body functions; S=body structures; E=environmental factors; D=activity and participation.
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APPENDIX

Post-Coma Scale (0-100)
(R. Formisano et al., 1996)

a) **Vigilance**

4 = normal
3 = fatigability
2 = soporous
1 = long periods of vigilance
0 = short periods of vigilance

b) **Reaction to auditory stimulation**

4 = obeys simple commands
3 = looks towards the stimulus
2 = blinks in response to meaningful or familiar stimulation
1 = fluctuating blinking in response to the stimulation
0 = no reaction

c) **Reaction to painful stimulation**

4 = intentional motor response
3 = localizes, but does not ward off, painful stimulation
2 = mild motor reaction
1 = decerebrated/decorticated reaction
0 = no reaction

d) **Position of eyelids and ocular bulbi**

4 = eyes open spontaneously
3 = eyes open in response to verbal or tactile stimulation
2 = ptosis - strabismus
1 = eyes open in response to painful stimulation
0 = fixed divergence of the bulbi

e) **Motility of the eyes**

4 = visual pursuit
3 = visual fixation
2 = blinks in response to threat
1 = presence of oculocephalic reflex
0 = pendular movements of the eyes/absence of oculocephalic reflex
f) Emotional responsiveness

4 = smiling or crying in response to significant stimulation
3 = smiling or crying in absence of significant stimulation
2 = presence of responses only to familiar stimulation
1 = presence of sporadic responses with fluctuations
0 = absent

g) Oral movements

4 = follows commands with the lips or tongue
3 = bite tendency and/or oral reflex activity of the mouth area
2 = trismus, jaw contracture
1 = primitive oral automatisms (yawning, sucking, bruxism, etc.)
0 = absent

h) Spontaneous motility and posture

4 = functional use of objects
3 = intentional motor activity
2 = stereotyped movements/myoclonic jerks/grasping reflex
1 = decortication/decerebration
0 = flaccidity

i) Verbal communication

4 = comprehensible
3 = aphasia
2 = aphonia/dysphonia or dysarthria
1 = confused and stereotyped words/confabulations/incomprehensible sounds
0 = absent

j) Behavior

4 = collaborative
3 = psychomotor agitation/aggressiveness
2 = oppositional behavior/reduction of psychomotor initiative
1 = spastic crying and/or laughing
0 = no psychomotor initiative/complete inertia

k) Interaction with the environment

4 = verbal
3 = by gesture or writing
2 = by eyelid closure or upwards/downwards gaze
1 = by mimic reactions
0 = absent

l) Duration of consciousness disorder

4 = less than 1 month
3 = less than 3 months
2 = less than 6 months
1 = less than 1 year
0 = longer than 1 year
m) Breathing

4 = normal
3 = tachypnea or stertorous
2 = with pauses/periodic or superficial breathing
1 = intubation/tracheostomy/tracheal stenosis
0 = assisted ventilation

n) Autonomic nervous system functions/immune system

4 = normal
3 = orthostatic hypotension
2 = vegetative crises (increased sweating, arterial hypertension, tachycardia)
1 = recurrent infections
0 = subcontinuous hyperthermia

o) Feeding

4 = normal
3 = dysphagia
2 = food refusal (inconstant feeding by mouth)
1 = stomach tube/gastrostomy (PEG)
0 = parenteral nutrition

p) Epilepsy

4 = no epilepsy
3 = early epilepsy
2 = late epilepsy (1-2 attacks)
1 = late epilepsy (more than 2 attacks)
0 = previous or current epileptic status

q) Neurosensory deficits

4 = none
3 = hyposmia and/or hypogeusia
2 = sensory disturbances (hypoesthesia, dysesthesia, paresthesia)
1 = deafness
0 = not assessable

r) Visual disorders

4 = none
3 = visual field deficits
2 = diplopia
1 = blindness
0 = not assessable

s) Peripheral nervous system disorders

4 = none
3 = facial nerve palsy
2 = peripheral palsies upper limbs
1 = peripheral palsies lower limbs
0 = peripheral palsies upper and lower limbs (CIP = critical illness polyneuropathy)
t) Motor coordination disorders

4 = normal
3 = slight intentional tremor/slight ataxia
2 = moderate intentional tremor/moderate ataxia
1 = severe intentional tremor/severe ataxia
0 = not assessable

u) Extrapyramidal symptoms

4 = none
3 = hypomimia, facial seborrhea, hypersalivation
2 = parkinsonian posture, resting or postural tremor
1 = rigidity and bradykinesia
0 = not assessable

v) Cognitive disorders

4 = spatial and temporal disorientation
3 = mild/moderate cognitive disorders (attention/memory)
2 = hemineglect
1 = severe cognitive disorders
0 = not assessable

w) Sphincter control

4 = normal
3 = sporadic incontinence and/or retention
2 = urine condom/napkin
1 = vesicostomy/intermittent catheterization
0 = no control (urine catheter and fecal incontinence)

x) Cutaneous trophism

4 = normal
3 = small non-infected bedsore
2 = large non-infected bedsore
1 = infected bedsore or multiple bedsores
0 = multiple large and/or infected bedsores

y) Muscle contractures/ankylosis

4 = normal
3 = mild or moderate spasticity
2 = severe spasticity
1 = muscle contractures/subankylosis
0 = ankylosis and/or para-articular ossifications (PAO)

z) Interval from brain injury to evaluation

4 = less than 1 month
3 = less than 3 months
2 = less than 6 months
1 = less than 1 year
0 = longer than 1 year