Living for 11 years in a Permanent Vegetative State – case report

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Abstract

The Vegetative State, term introduced more than 40 years ago, is described as a disorder of consciousness. It represents the by-product of modern intensive care, an unintended consequence of the increased ability to manage neurological damage. The life span of patients in such a state is substantially reduced. For most, life expectancy ranges from 2 to 5 years and survival beyond 10 years reported as being unusual. This article presents a case report, therapeutic management and in-home patient care of a 55 year-old male, who lived in a permanent vegetative state for 11 and a half years.

Keywords: *vegetative state, life expectancy, in-home care*

Case report

We present a case report of a 55 year-old male who, on the 13th of February 2002, is found unconscious at home. The patient is obese, BMI between 30-35, heavy smoker and with no history of neurological disorders and trauma, known with previous history of type 2 Diabetes and Arterial Hypertension, diagnosed 2 years earlier.

Subsequently, he is brought by the ambulance into the Emergency Room, University Hospital Regensburg and the data extracted from the medical records indicate that a Cranial CT had been performed (Figure 1), revealing a massive cerebellar haemorrhage, invading the IIIrd and IVth ventricle, with internal hydrocephalus.

The subsequent operative care implied emergency suboccipital craniotomy and evacuation of the haematoma and placement of an external ventricular drainage.

The post-operative evolution however, was unfavourable with the patient being described as unresponsive. Numerous complications developed in the following weeks.

On the 24th of February the patient developed a cerebrospinal fluid (CSF) fistula. Three days later, a deep vein thrombosis (DVT) caused a pulmonary embolism complicated with cardiogenic shock, requiring thrombolysis.

The medical records show that on the 6th of March the patient was diagnosed with ventriculitis, on the 14th with multiple stress ulcers, on the 24th with left fronto-temporal subdural hygroma, subsequently drained and on the 3rd of April a CT scan showed a subdural haematoma on the site of the drained hygroma and dilation of the ventricular system.

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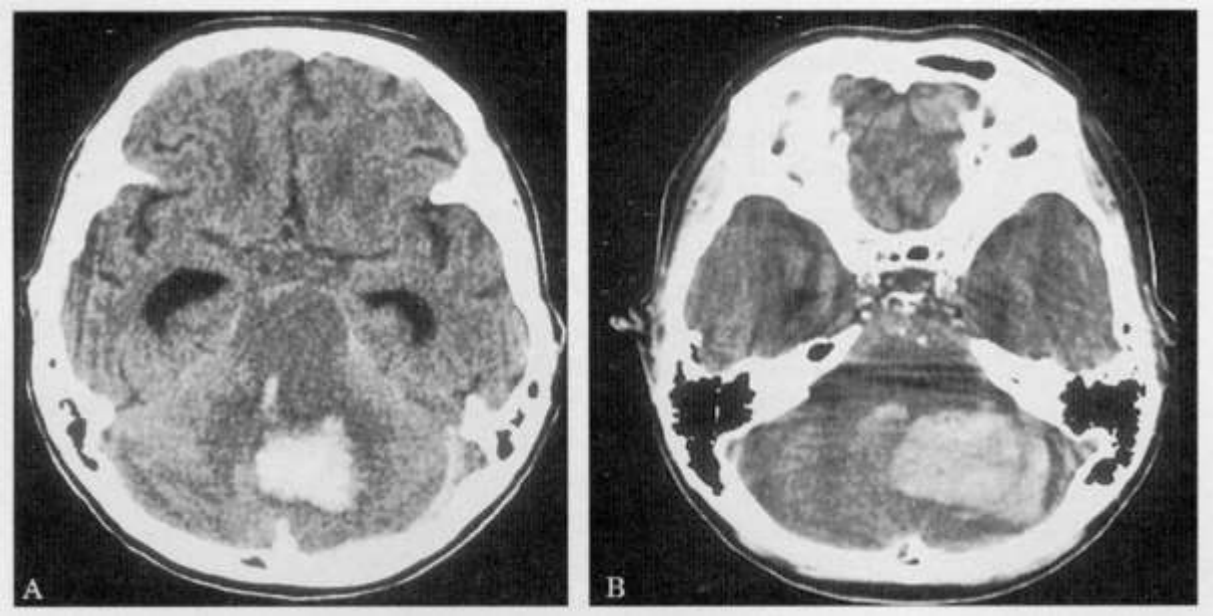

Fig. 1. CT scan images A and B, revealing a left hemispheric cerebellar haematoma, extending to the vermis, with haemorrhage invading into the IVth ventricle

In the period 4.04-26.04, the patient was admitted to the Neurological rehabilitation clinic in Regensburg, Germany with the diagnosis of diminished wakefulness, at the time of a remission phase of an apallic syndrome. A tracheostomy tube, percutaneous endoscopic gastrostomy (PEG) tube and suprapubic catheter were placed as part of the medical treatment. On the 10th of April, seizures originating in the right hemisphere and subsequently generalized were documented.

A CT scan on the 24th of April revealed the progression of the ventricular dilatation, prompting external ventricular drainage. As a consequence, between 26.04-12.06.2002, the patient was again admitted to the Neurosurgery Ward with the evolution again marked by complications such as multiple revisions of the ventricular drainage that resulted in the placement of a ventriculo-peritoneal (VP) shunt, deep vein thrombosis and methicillin-resistant *Staphylococcus aureus* (MRSA) colonization.

This was followed by another two months (12.06-14.08.2002) spent in Neurological Rehabilitation Clinic, where we have records of a more thorough examination. The patient was described with diminished wakefulness and present sleep-wake cycles. Upon examination, there was noted an inability to talk, express willpower by blinking or head movements, inconsistent visual fixation, spontaneous and on verbal requests, presence of grunting and moaning, inconsistent withdrawal reflex on noxious stimuli, and no reaction on verbal or acoustic stimuli. Cranial nerves exam revealed bilateral absence of ciliary reflex and right facial nerve palsy. We also observed the absence of spontaneous movement and exaggerated left biceps reflex, otherwise normal. Noxious stimulation determined an increase in respiratory rate, but the sensibility could not be tested. Autonomic nervous system examination revealed bladder and bowel incontinence. Serial electroencephalograms (EEGs) were interpreted as having general alterations of moderate severity: the presence of generalized delta and theta waves, mainly in the right hemisphere, typical epileptiform waves, indicative of seizure activity, originating in the right fronto-temporal lobe, all in all, an increased cerebral excitability. Somatosensory evoked potentials reached cortical components, but were severely delayed.

Auditory evoked potentials were unable to obtain, suggesting severe bilateral pre- or cochlear lesion.

The endoscopic evaluation of swallowing that followed revealed the absence of deglutition reflex, with permanent saliva aspiration, therefore impossible for the tracheostomy tube to be removed. No signs of improvement were noted throughout this period and the criteria were met for the diagnosis of persistent vegetative state. The patient was discharged with further recommendations of physical therapy and ergotherapy.

Afterwards, between 14.08.2002 and 02.04.2003, the patient was sent to an elderly care facility in Parsberg, Germany. From 02.04.2003, on his wife's request he was transferred to another elderly care facility in Regensburg, until 01.07.2007.

From that day onwards the patient was brought by his family in Arad, Romania, for in-home care (Figure 2). This required round o'clock care by the wife and with the help of a retired nurse (24/7).

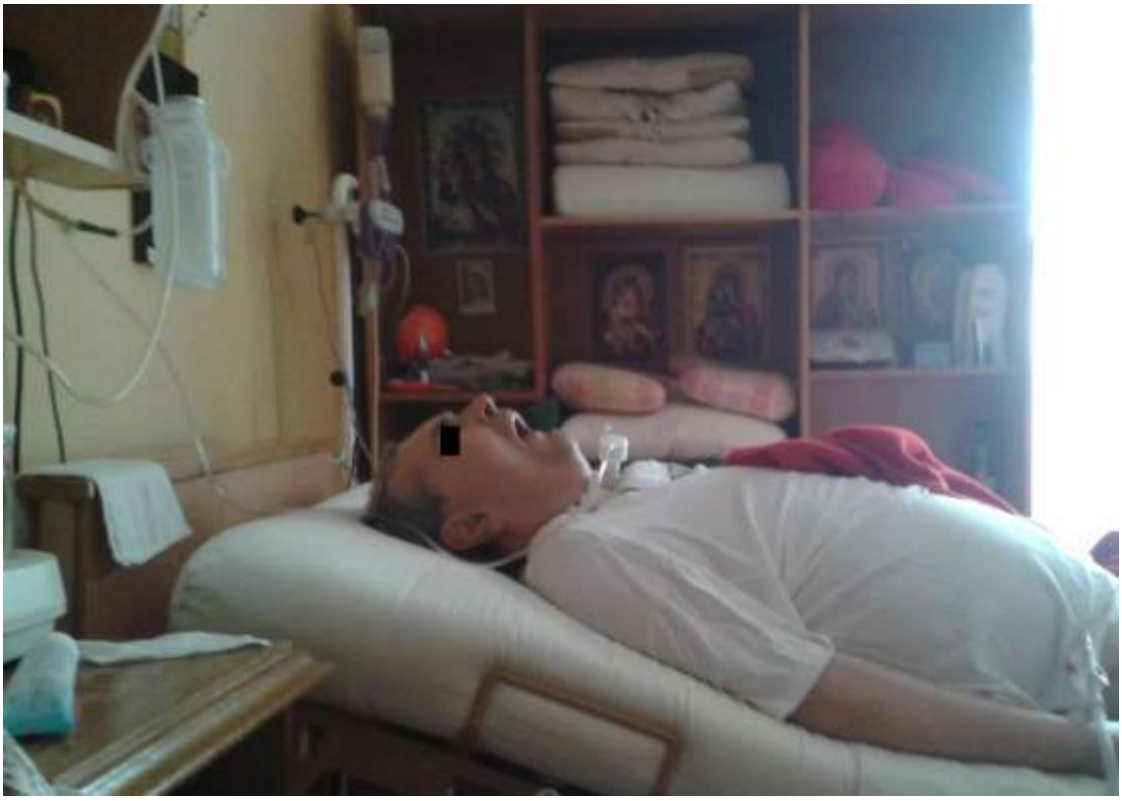

Fig. 2. Patient in 2013, at home, cared for by the wife and nurse. Note the tracheostomy tube and dressing around the neck, with a catheter connected to an oxygen apparatus, because of the anaemia. On the far left there is a small container of the suction apparatus, used for saliva aspiration. On the upper left side is the bottle of Glucerna with the apparatus used for PEG feeding. On the far right, the suprapubic catheter.

The recommendations made upon discharge from the German facilities are of level III of care. This is characterized by a need for assistance for at least 300 minutes daily, with basic care needs of at least 240 minutes per day. The main daily activities comprised of: whole body washing, oral, skin, hair, nails hygiene, intertrigo prophylaxis, wound care, aseptic PEG dressing change, composing a nutrition plan, administration of fluids and feeding over PEG tube, blood sugar and fluid balance measurements (Table 1).

Table 1. Scheduled activities on a daily basis, during the 6 and a half years of in-home care

3 am	Changing patient's position, 500 ml Glucerna (special food) in the course of 5 hours
6 am	Patient awakes Changing patient's position
8 am	250 ml water + 250 ml Glucerna in the course of 4 hours
9 am	Changing patient's position Oral, skin, hair, nails hygiene, shaving approx. 2 hours
12 am	Changing patient's position 250 ml of water + 250 ml Glucerna in the course of 3 hours
1 pm	Whole body wash, whole body massage and mobilization of the joints for approx. 4 hours
4 pm	500 ml Glucerna in the course of 5 hours
6 pm	Changing patient's position
9 pm	Changing patient's position Patient falls asleep Feeding pause until 3 am
12 pm	Changing patient's position

The patient required round o'clock care, frequent saliva aspiration and was never left unattended. Medication was administered over the PEG tube. The tracheostomy tube was replaced every one month and the suprapubic catheter every 6 weeks.

Preventable complications regarding basic care needs were successfully avoided with the high-quality nursing. However, due to permanent tracheostomy tube, pneumonias developed approximately twice a year. Prolonged catheterization resulted in frequent urinary tract infections. The patient often had epileptic seizures as a result of the neurological damage.

On the 10.05.2012, the patient developed a severe aregenerative anaemia (Hb=4.7 g/l), of an undetermined cause, most likely anaemia of chronic disease. From then on, blood transfusions were required approximately every 6 weeks (when Hb<8g/l). After approximately one and a half years, the life-prolonging treatment in the form of blood transfusion was stopped at wife's request. On the 12.09.2013, 11 and half years after the brain injury, patient died of respiratory failure.

Discussion

The first mention of this syndrome was in 1940, when Ernst Kretschmer called it "apallic syndrome" [1]. However, it wasn't until 1972, that the term "Vegetative State" was first introduced. Jennet and Plum used it to accurately describe a progressed coma to a state of wakefulness without detectable awareness [2].

The comatose state, minimally conscious state and vegetative state are all traditionally described as disorders of consciousness. However, the term consciousness nowadays is ambiguous and, for a scientific purpose, has been divided in two distinct components: awareness and wakefulness. Central to the definition of the Vegetative State (VS) is a paradoxically dissociation between the two components [3]. Together, these are

fundamental for a healthy, living person but Vegetative State implies one existing without the other. By wakefulness, we mean a state of arousal, in which one's eyes are open and usually associated with a degree of motor activity [4], whereas awareness is understood to be one's ability to experience the environment [5]. Therefore, the patients have been described as awoken yet unaware [2].

The diagnostic criteria for the Vegetative State were drawn up in 1994, by the US Multi-Society Task Force on PVS (Table 2) [5]. It relies solely on physical examination, the criteria representing clinical findings, as there is no specific test, clinical sign or laboratory investigation for diagnosing awareness [4].

Table 2. Diagnostic criteria for the Vegetative State as formulated by the US Multi-Society Task Force [5]

No evidence of awareness of self or environment and an inability to interact with others
No evidence of sustained, reproducible, purposeful, or voluntary behavioral responses to visual, auditory, tactile, or noxious stimuli
No evidence of language comprehension or expression
Presence of sleep-wake cycles
Sufficiently preserved hypothalamic and brainstem autonomic functions to permit survival with medical and nursing care
Bowel and bladder incontinence
Variably preserved cranial-nerve and spinal reflexes

There are a number of additional possible features such as: grunting, moaning, crying, smiling and lack of visual fixation. While none of those require conscious awareness, such signs may have a distressing impact on family members or clinical staff, but they do not alone exclude the presence of Vegetative State [6].

The Multi-Society Task Force on PVS states that neurological testing can neither confirm the diagnosis, nor predict the potential for recovery. Neuroimaging often reveals diffuse or multifocal cerebral disease involving the grey and white matter. EEG shows diffuse generalized polymorphic delta or theta activity.

Typical epileptiform activity is unusual in patients in a Persistent Vegetative State, as is seizure activity [5]. However, recently it has increasingly clear proven that neuroimaging can be used to rule out a Vegetative State and may even yield information about prognosis (in the form of quantitative measurement of brain activity) [7-9].

Statistics about Vegetative State are scarce. One study in Austria gives us a glimpse of how many patients there actually are. One hundred and fourteen out of 889 long-term care facilities and nursing homes cared for 269 patients in a VS indicating a prevalence of 3.36 patients per 100,000 [10].

Persistent Vegetative State is defined after 4 weeks spent in VS. The VS is deemed to be permanent after 12 months in the case of a traumatic brain injury and 6 months for other causes [4].

Although the term 'permanent' implies that awareness will never be recovered, and certainly has a prognostic significance, the prediction cannot be made with an absolute certainty [4]. Literature has described a few outstanding cases that recovered after a very long time (19 years in VS) [11].

However, this may be due to misdiagnosis in the first place and, as studies have demonstrated, chances of recovery are

considerably diminished as the time spent in VS increases [12]. Hence, there is a distinction to be made in the abbreviation of PVS, which has since been discouraged as the Royal College of Physicians has given recommendations on this matter [4], later reinforced by Jennett (2002) [13].

The causes for the VS are several and have been divided into 3 distinct categories: a. acute brain injuries, traumatic or non-traumatic, b. severe congenital abnormalities of the central nervous system and c. degenerative and metabolic disorders [5].

As for the presented patient, he had a stroke, which represents a major cause of non-traumatic brain injury, while intraparenchymal haemorrhage accounts for approximately 8-13% of all strokes [14]. Additionally, cerebellar haemorrhage comprises for 5-13% of all parenchymal intracranial haemorrhages, with hypertension being the underlying cause (60-80%).

The clinical course and prognosis of a Vegetative State depends on its cause (Figure 3). Progress in medicine has resulted in an increased number of patients who are able to survive severe acute brain damage [15].

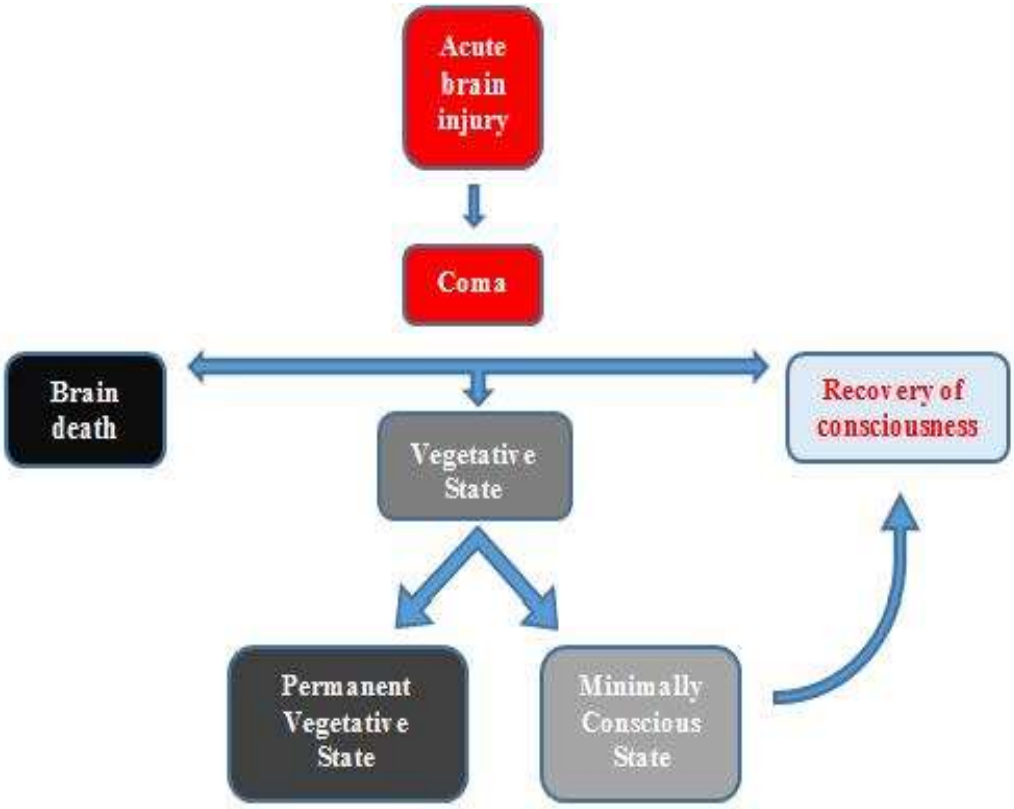

Fig. 3. Different clinical entities that may follow acute brain injury. Patients recovering wakefulness but not awareness are deemed in a Vegetative State. The term "Permanent" is added after 12 months from a traumatic brain injury or 6 months for other causes [4]. However, there is the possibility of transition to Minimally Conscious State (MCS) from the Vegetative State. Afterwards, return of functional communication or object use is translated as emergence from the MCS. Adapted from Laureys (2007) [16].

After a traumatic or non-traumatic brain injury, while recovering from a coma, wakefulness may recover while awareness may not. A sketchy explanation of the dissociation is related to the fact that brain systems controlling wakefulness are located in the upper brain and thalamus, whereas largely distinct from the areas that are responsible for awareness. The injury involves cortical or white matter and thalamus, rather than brain stem structures [4]. The capacity for survival in a Permanent Vegetative State resides in the

preservation of hypothalamic and brain-stem autonomic functions. Most patients who survive for a long time maintain normal body temperature, the ability to breathe spontaneously, and a functioning cardio-vascular system [5].

Age, type of injury and time spent in VS are the main factors influencing the prognosis. Time spent in vegetative state is inversely correlated with chances of recovering consciousness and independence and positively correlated with the probability of remaining in VS [17]. A report shows that recovery rate of independence, at one year for patients over 40 years old, is virtually 0%.

Related to the type of brain injury, reports show that in terms of recovery of consciousness, traumatic brain injuries are associated with better outcomes that non-traumatic brain injuries (52 vs. 13%, at one year) [12]. Overall, the available data (based on 251 patients in four large series) indicate that the mortality rate for adults in a persistent vegetative state after an acute brain injury is 82% at three years and 95% at five years [18-21]. Life expectancy ranges from 2 to 5 years; survival beyond 10 years is unusual [22].

Conclusion

In conclusion, this case represents evidence that prolonged survival in a persistent vegetative state in the setting of in-home care is a real possibility. Moreover, it suggests that autonomic function is sufficient to maintain long-term internal regulation as long as the patient's needs receive constant attention. However, the ethical aspect of life-prolonging treatment remains debatable and family's wishes are of utmost importance in the matter.

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