

A Sixteen-Year-Old in PVS Who Develops Cushing's Disease: Medical and Ethical Issues

Introduction

We present the medical and ethical concerns we encountered when a young adolescent in permanent vegetative state (PVS) developed Cushing's disease.

History and Examination

A previously healthy boy of 16 years presented with two tonic clonic seizures due to an arteriovenous malformation (AVM). He underwent embolisation, which was complicated by catastrophic postoperative bleeding resulting in severe brain injury. On transfer to the neurological rehabilitation unit three months later he appeared to have no awareness of himself or his environment. His tracheostomy had been removed just prior to his transfer and he was maintaining normal respiration and circulation. He was fed through a percutaneous endoscopic gastrostomy tube. He exhibited minimal spontaneous movements in his limbs. Tone was generally increased with flexion contractures in upper limbs, extensor posturing in lower limbs and his feet were fixed in equinovarus. He had sleep-wake cycles with non-purposeful roving eye movements, chewing movements and reflex response to noxious stimuli. His Wessex Head Injury Matrix (WHIM) score was 1/7.

Rehabilitation goals

The main aims of his rehabilitation were, firstly, to establish whether there was any evidence of recovery that might result in him improving from a vegetative state to a higher level of awareness, with the attendant possibility of communication; secondly, to establish a clear nursing and postural management plan, including managing issues related to growth. Thirdly, it was important to provide his family with appropriate emotional and psychological support.

Diagnosis of PVS

In order to confirm the diagnosis of PVS, it was necessary to review his treatment and cognitive state. His clinical condition did not improve over the following nine months in our unit. For the diagnosis of vegetative state, he underwent formal and systematic assessment of the three major sensory systems (auditory, visual and somatic) and the motor system.^{2,8} The awareness assessment consisted of ten sessions over a 14-day period, at varying times of the day and lasting approximately 40 minutes. This process was repeated two months later after a multimodal sensory stimulation programme. He showed a generalised startle response to loud noise, occasional slow but inconsistent localisation of sound by turning his head and no response to verbal commands. He had normal pupillary responses, spontaneous eye movements, spontaneous blinking but no consistent tracking and no response to visual threat. There was reflex flexion to pain in the legs but no response to pain in the arms or face. Some grunting noises were made, but not with any consistency or

meaning. His level of awareness was therefore rated at level two on the Rancho Los Amigos Scale, which is a cognitive functioning scale⁷ (Table 1).

In view of the significance of a diagnosis of PVS, a second opinion⁹ was sought from an experienced clinician from another institution. He independently confirmed our clinical findings.

His medication was reviewed to consider the potential for sedative side-effects.⁵ He was on baclofen and carbamazepine. The baclofen was stopped to judge any change in his level of arousal, with no positive effect. The dosage of carbamazepine was extremely low and felt unlikely to be contributory.

A brain CT scan at six months confirmed the presence of widespread ischaemia in both hemispheres (see figure).

In summary he was found to have intact primary sensory pathways and primary motor output but no evidence of sustained, reproducible, purposeful or voluntary behavioural responses to normal or noxious visual, auditory or tactile stimuli, no language comprehension or expression. He only had reflex motor responsiveness with no evidence of co-ordinated motor activities. As his neurological status remained unchanged for one year after catastrophic brain injury, the diagnosis of PVS was made.^{1,2,5}

Presentation and diagnosis of Cushing's Syndrome

At the time the patient was being assessed for PVS one year after his haemorrhage he was found to have hypertension with no obvious cause, unexplained weight gain and rapidly progressive purple striae on his arms, trunk and thighs. He was also noted to be severely osteopenic on an ankle x-ray.

Four 24-hour urine collections showed elevated uri-



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Table 1: Rancho Los Amigos Scale Level 2: Generalised Response: Total Assistance

1	Demonstrates generalised reflex response to painful stimuli.
2	Responds to repeated auditory stimuli with increased or decreased activity.
3	Responds to repeat external stimuli with generalised physiological changes, gross body movement with or without purposeful vocalisation.
4	Responses noted above may be same regardless of type and location of stimulation.
5	Responses may be significantly delayed.

Original Scale co-authored by Chris Hagen, PhD, Danese Malkmus, MA, Patricia Durham, MA Communication Disorders Service, Rancho Los Amigos Hospital, 1972. Revised 11/15/1974 by Danese Malkmus, MA and Kathryn Stenderup, OTR.

