Vegetative State (Unresponsive Wakefulness)

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DEFINITIONS 1
ETIOPATHOLOGY 1
CLINICAL FEATURES 2
DIAGNOSTIC STUDIES 3
TREATMENT 3
PROGNOSIS 3
STATE OF VIRGINIA 4

DEFINITIONS

VEGETATIVE STATE (VS) – state of ***arousal without inner or outer awareness***;
broader but less definite terms are *AKINETIC MUTISM* and *COMA VIGIL*.

PERSISTENT VEGETATIVE STATE (diagnostic category) - vegetative state *present for ≥ 1 month*.

PERMANENT VEGETATIVE STATE (prognostic category) - vegetative state *present for ≥ 3 months* (if brain injury was medical) or *> 12 months* (if brain injury was traumatic) - further improvement is unlikely!

ETIOPATHOLOGY

- patient enters VS *after period of coma* (usually traumatic or anoxic), when CNS re-establishes consciousness (with supportive care, brain stem may recover more rapidly than cerebral cortex).
- VS may not begin with coma but can develop as *end stage of dementing neurodegenerative diseases* or accompany *severe developmental abnormalities* (e.g. anencephaly).
- histopathology - *loss of cortex* with *preservation of brainstem RAS*.
  N.B. necessary condition for VS is relatively intact brain stem!
  A) **diffuse axonal injury** (e.g. in **trauma**).
  B) **laminar necrosis of cortical mantle** (i.e. death of selectively vulnerable cortical neurons), e.g. secondary to cerebral hypoperfusion from **cardiac arrest**.
  C) **cortical injury** following purulent meningitis / encephalitis, exposure to nervous system toxins (esp. CO), prolonged hypoglycemia.
  D) **bihemispheric infarctions**
  E) **thalamic necrosis** (e.g. as sequelae of cerebral hypoperfusion; such thalamic necrosis was major cause of VS of widely reported patient Karen Quinlan).

Pseudolaminar necrosis of cerebral cortex (typical of persistent vegetative state on life-support systems) - cortical ribbon is very thin because of cortex loss:

Source of picture: "WebPath - The Internet Pathology Laboratory for Medical Education" (by Edward C. Klatt, MD) >>

CLINICAL FEATURES

- ***do not respond to any stimuli*** (external or internal)* – absolute inability to communicate → total lack of cognitive function - "awake coma" ("coma vigil").
  *except primitive responses to pain (decorticate or decerebrate postures or fragments of these movements).
  N.B. *brain stem startle reflexes are preserved*, so eye opening occurs with loud sounds and blinking may occur with bright lights.
  Diagnosis is inaccurate and unreliable in patients less than 2 yr of age!
- **pupils** react normally.
- **eyes** open and close, move spontaneously, appear to track objects about room*.
  N.B. spontaneous eye opening is sign of arousal, not awareness!
  **spontaneous roving eye movements are particularly characteristic* (very slow movements of constant velocity, uninterrupted by saccadic jerks, and cannot be volitionally mimicked);
  – *particularly distressing to family members* as patients appear to be looking about room and at some point roving eyes are pointed at observer, who may perceive patient to be "looking at" or following him throughout room;
  – brain stem origin of eye movements is documented by their being *readily redirected by doll's-eyes reflex*.
- intact brainstem reflexes (e.g. may **chew & swallow** food placed in mouth).
- manifests **sleep-wake** cycling (intact RAS)! vs. comatose patients
- intact **cardiorespiratory**, **thermoregulatory**, **neuroendocrine** control (may be subject to periods of overactivity).
- **limbs** may move randomly, but not purposefully or only primitively purposefully (e.g. grasping object that contacts hand); limbs show signs of extensive damage to both cerebral hemispheres - Babinski signs, decerebrate or decorticate posturing.

- incontinence of **bladder & bowel**.

Criteria for Determination of Vegetative State

1.	No awareness of self or surroundings. Eye opening (reflex or spontaneous) may occur.
2.	No communication (auditory or written) between examiner and patient that is meaningful and consistent. Target stimuli not followed visually, but sometimes visual tracking present. No emotional response to verbal stimuli.
3.	No comprehensible speech or mouthing of words.
4.	Emotions (smiling, frowning, crying) inconsistently related to any apparent stimulus.
5.	Sleep-wake cycles present.
6.	Brain stem and spinal reflexes variable, e.g. preservation of sucking, rooting, chewing, swallowing, pupillary reactivity to light, oculocephalic responses, grasp or tendon reflexes.
7.	No voluntary movements or behavior, no matter how rudimentary; no motor activity suggesting learned behavior; no mimicry. Withdrawal or posturing can occur with noxious stimuli.
8.	Usually intact cardiorespiratory function. Incontinence of bladder and bowel.

In *AKINETIC MUTISM*, patient doesn't move and doesn't speak; awareness is present, but mentation is very slow.

DIAGNOSTIC STUDIES

- **imaging studies** depict sequelae of causative injury but are not diagnostic of VS.
- **MR spectroscopy** - decrease in neuronal marker of *N*-acetyl aspartate.
- **PET** - decreased glucose utilization and cerebral blood flow.
- **EEG** - diffuse polymorphic delta or theta activity with no response to stimuli (except painful); some patients demonstrate alpha coma pattern (it is more specific for VS).
- **evoked responses** are absent.

TREATMENT

- SUPPORTIVE:
    1) adequate **fluids & nutrition** (frequently by gastrostomy)
    2) management of **pain and discomfort** (rarely apparent)
    3) comfortable and nurturing **environment**.
    4) preventing pneumonia (incl. tracheostomy), pressure ulcers, contractures of extremities.
- physician should introduce concept of "*do not resuscitate*" and review prognosis and management with family on regular basis.
- technologic feasibility of indefinitely prolonging life without consciousness has generated considerable ethical debate.
- **DBS** for thalamic intralaminar nuclei and adjacent paralaminar regions improves EEG patterns (an increase in desynchronization and the power spectrum of electroencephalogram), patient awareness (but only slightly), decreases spasticity, opsoclonus, and myoclonus.
    - suitable only for patients with preserved anatomic connectivity.

PROGNOSIS

- varies with ***etiology*** (mechanism of injury), ***duration*** of VS and ***age*** of patient.
    - spontaneous improvement is observed within 2 years.
    - vegetative state persisting 3 months after *anoxic* injury is expected to persist indefinitely.

- vegetative state at time of hospital discharge after head *trauma* has 50% likelihood of some **awareness recovery** over next 12 months.
- prognosis for **functional recovery** (returning to work or to school) is much worse;
  ≈ 1/3 trauma patients < 30 years of age who awaken after prolonged vegetative state meet these goals (but only some patients > 30 years of age do so).
- mean survival 2-5 years (described cases of survival for 37 and 41 years!).

State of Virginia

PVS - state of unconsciousness that involves lack of response to external stimuli in any other than a reflex activity.

- there is no legal requirement as to how a PVS is diagnosed or that specific personnel make diagnosis.

BIBLIOGRAPHY see p. S30