

Neurological Bulletin

Volume 3 | Issue 1

Article 5

January 2012

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Recommended Citation

Dolgovin M, David S, Harth CE. Undulating Toe Flexion Sign in Brain Death. *Neurological Bulletin* 2011;3:33-35, http://dx.doi.org/10.7191/neurol_bull.2012.1031

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NEUROLOGICAL BULLETIN

FEATURING ARTICLES BY TRAINEES IN NEUROLOGY & NEUROSCIENCE

Undulating Toe Flexion Sign in Brain Death

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Case Report

A 30-year-old man with a history of opioid and benzodiazepine abuse was found unresponsive in asystole and then resuscitated for 15 minutes. He was subsequently brought to the emergency department, intubated and placed on mechanical ventilation. The patient was admitted to the medical intensive care unit where hypothermia protocol was initiated. Head CT showed cerebral edema with complete loss of gray-white matter differentiation, complete effacement of the sulci within the bilateral cerebral and cerebellar hemispheres, and effacement of all of the cisterns. Brain death protocol was initiated and completed 5 days after arrest once the criteria for normal electrolytes and adequate blood pressure were met.

Vital signs at the time of evaluation for brain death were blood pressure 128/64, pulse 80, respiratory rate set at 12 on the ventilator, and temperature 97.2 °F. The exam revealed no meaningful motor response to noxious stimuli, fixed and dilated pupils, absent oculocephalic reflex (Doll's eyes), absent corneal reflex, no spontaneous respiration (confirmed by apnea test), no spontaneous movement, absent gag reflex, and absent

flexor plantar response.¹ Vestibulo-ocular reflex was not assessed with cold caloric testing due to ruptured tympanic membranes and hemotympanum. Of note, the patient was found to have a reflexive undulating toe flexion sign bilaterally upon plantar tactile stimulation. The patient remained in the medical intensive care unit awaiting organ procurement for transplant. The accompanying video was made 6 days after the initial arrest.

Because the vestibulo-ocular reflex could not be assessed, 16 channel portable EEG was used as a confirmatory test and was performed according to standards set by the American Electroencephalographic Society, American Academy of Neurology, and American Neurological Association. Three traces were reserved for EKG, respiration and extracerebral monitoring. Standard electrode placement was used, recording from electrode pairs at least 10 cm apart. Inter-electrode impedances were greater than 100 and less than 10,000 ohms. High sensitivity (2 μ v/mm or better) was used for at least 30 minutes. The low frequency pass filter was at 1 Hz or lower, and the high frequency filter at 30 Hz or above. EEG revealed no discernible activity of cerebral origin, either spontaneous or in response to somatosensory

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Keywords: brain death, coma, undulating toe sign, hypoxic ischemic encephalopathy, movements in brain death

or auditory stimulation, consistent with electrocerebral silence and supporting the diagnosis of brain death.

Discussion

Brain dead patients, who by definition have lost all cortical and brain stem function, exhibit a wide array of spontaneous movements and reflexes, including triple flexion response, “Lazarus” sign, upper limb pronation/extension reflex, finger jerks, flexor plantar response, unilateral facial myokymia and many more.² Most abnormal movements are observed in the first 24 hours after declaration of brain death, which is defined as the time at which all brain death criteria have been met. Undulating toe flexion movements are more likely to be seen in the first 12 hours after the diagnosis of brain death. In our patient the abnormal movement was present at the time the patient was pronounced brain dead, 5 days after the initial cardiac arrest, and persisted for over 24 hours after declaration of brain death. Dösemeci and colleagues reviewed 134 brain dead patients. Of these patients, 18 had abnormal spinal reflexes, the majority (44%) of which were finger and toe jerks (8 of 18).³ In contrast, Saposnik and colleagues found undulating toe flexion movements to be the most common. They examined 107 brain dead patients, in which 47 had abnormal spinal reflexes, and the majority (23%) was the undulating toe flexion sign (25 of 47).⁴

The undulating toe flexion sign was first described in 1992.⁵ Upon tactile or noxious stimulation to the plantar area, the great toe flexes, followed by brief and sequential flexion of the second, third, fourth and fifth toes (refer to video 1).⁵ The undulating toe flexion sign was found to be more common in patients with intracerebral hemorrhage as opposed to tumor, subdural hematoma, or encephalopathy.⁴ Since in brain death there is no evidence of cortical or brainstem activi-

ty, it is believed that the undulating toe flexion sign represents a spinal reflex likely originating between the L5 to S1 segments.⁴ A review of the 1995 and 2010 guidelines from the American Academy of Neurology warns that complex, non-brain mediated reflexes can mimic retained brain function.^{1,6} There is an established guideline for the diagnosis of brain death where specific spinal movements do not exclude a diagnosis of brain death. These movements include: (1) spontaneous movements of limbs other than pathological flexion or extension response, (2) respiratory like movements (shoulder elevation and adduction), back arching, intercostal expansion without significant tidal volume, (3) deep tendon reflexes, superficial abdominal reflexes, triple flexion response, (4) Babinski reflex.² These signs should not preclude the diagnosis of brain death, and it is critical for clinicians to understand and recognize the clinical appearance of these neurological phenomena.

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Disclosure: the authors report no conflicts of interest.

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