Clinical Neurophysiology of Myoclonus Subtypes
(and Startle Syndrome)

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Objectives

1. ***Overview of clinical characteristics of Myoclonus subtypes and Startle Syndromes.***

2. ***Overview of physiologic characteristics of Myoclonus subtypes and Startle Syndromes.***

3. ***Simplistic/Pragmatic diagnostic approach for physiologic evaluation of jerk-like movements and characterizing myoclonus subtypes.***

Myoclonus

- Myoclonus literally means “a quick movement of muscle.”
- Sudden, brief, shock-like jerks caused by active muscle contractions -Positive myoclonus.
- Brief lapses of muscle contraction in active postural muscles- Negative myoclonus or asterixis

Differential Diagnosis for ‘jerk-like movements/myoclonus

- Tics (simple tics)
- Components of Chorea/Ballism/Dystonia
- Tremors
- Peripheral disorders
  - Fasciculations
  - Myokymia
  - Hemifacial spams

Classification of Myoclonus

Axis Ia- Clinical Features

Axis Ib- Clinical Neurophysiology Features


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IAPRD new consensus classification of myoclonus

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Axis II- Etiology

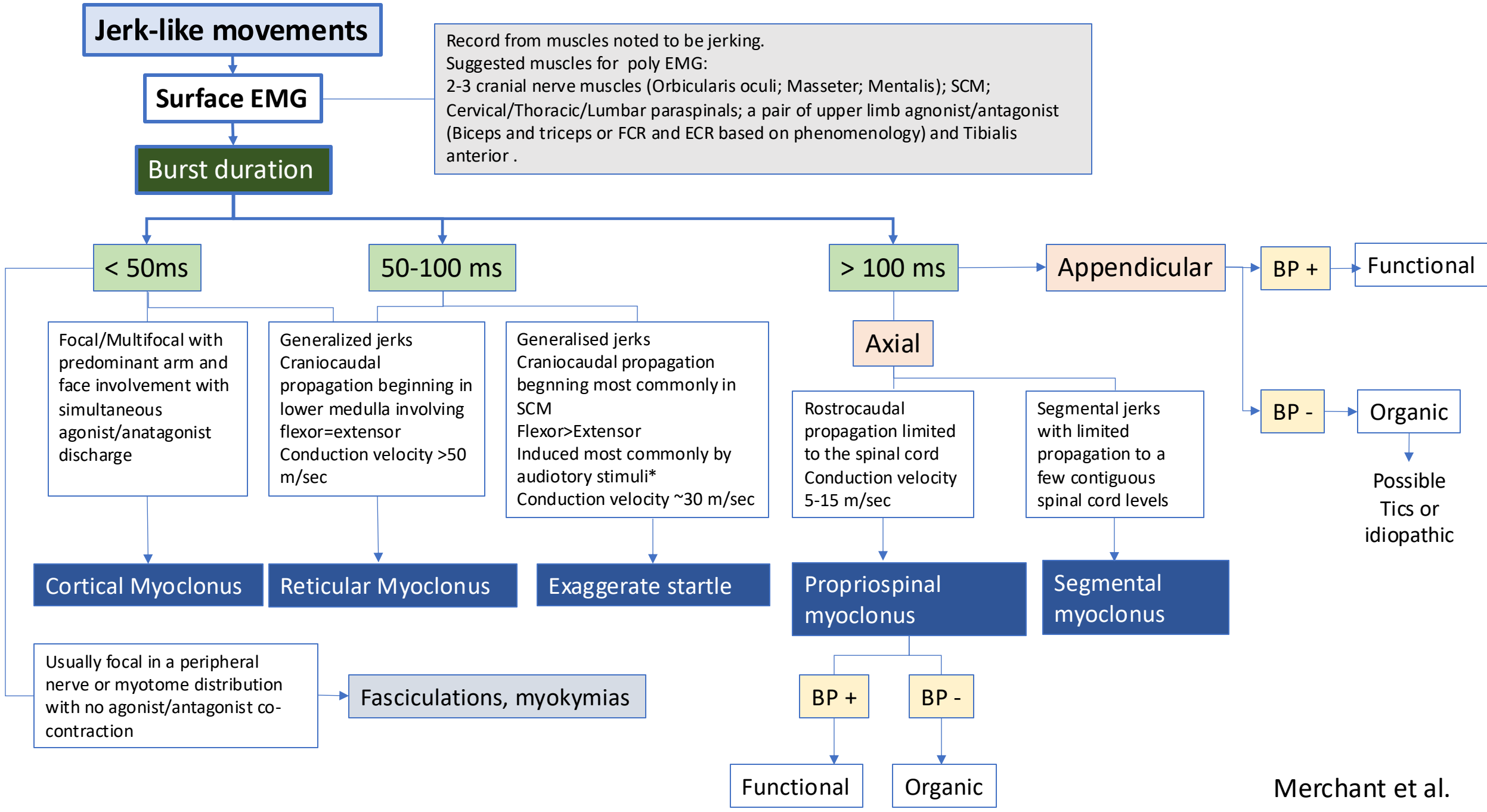
- Genetic
- Acquired (Toxic/metabolic/degenerative/infectious/paraneoplastic)
- Functional Neurological Disorder
- Idiopathic (Familial/Sporadic)
- Physiological (Hypnic/Fragmentary/Hiccup/anxiety, exercise-induced)

Axis Ib- Clinical Neurophysiology Features

- ***Cortical***
- *Cortical-Subcortical*
- *Subcortical*
- ***Brainstem (focal/generalized)***
- ***Spinal (focal) myoclonus***
- ***Propriospinal myoclonus***
- ***Peripheral myoclonus***
- ***Functional jerks***

Evaluation of Clinical Characteristics: Axis Ia

- History (Age of onset, temporal course)
- Primary etiology/Comorbid Neurological disorder
- Other localizing Neurological signs
- Phenomenology of movement disorder
- Pattern of activation
- Distribution of muscles involved (proximal vs distal; axial vs appendicular)
- Stimulus sensitivity/ Type of stimuli
- Habituation of stimulus sensitive response


Jerk-like movements
Surface EMG
Burst duration
Record from muscles noted to be jerking.
Suggested muscles for poly EMG:
2-3 cranial nerve muscles (Orbicularis oculi; Masseter; Mentalis); SCM; Cervical/Thoracic/Lumbar paraspinals; a pair of upper limb agnonist/antagonist (Biceps and triceps or FCR and ECR based on phenomenology) and Tibialis anterior .
< 50ms
50-100 ms
> 100 ms
Appendicular
BP +
Functional
BP -
Organic
Possible Tics or idiopathic
Axial
Focal/Multifocal with predominant arm and face involvement with simultaneous agonist/anatagonist discharge
Generalized jerks
Craniocaudal propagation beginning in lower medulla involving flexor=extensor
Conduction velocity >50 m/sec
Generalised jerks
Craniocaudal propagation begnning most commonly in SCM
Flexor>Extensor
Induced most commonly by audiotory stimuli*
Conduction velocity ~30 m/sec
Rostrocaudal propagation limited to the spinal cord
Conduction velocity 5-15 m/sec
Segmental jerks with limited propagation to a few contiguous spinal cord levels
Cortical Myoclonus
Reticular Myoclonus
Exaggerate startle
Propriospinal myoclonus
Segmental myoclonus
Usually focal in a peripheral nerve or myotome distribution with no agonist/antagonist co-contraction
Fasciculations, myokymias
BP +
BP -
Functional
Organic
Merchant et al.

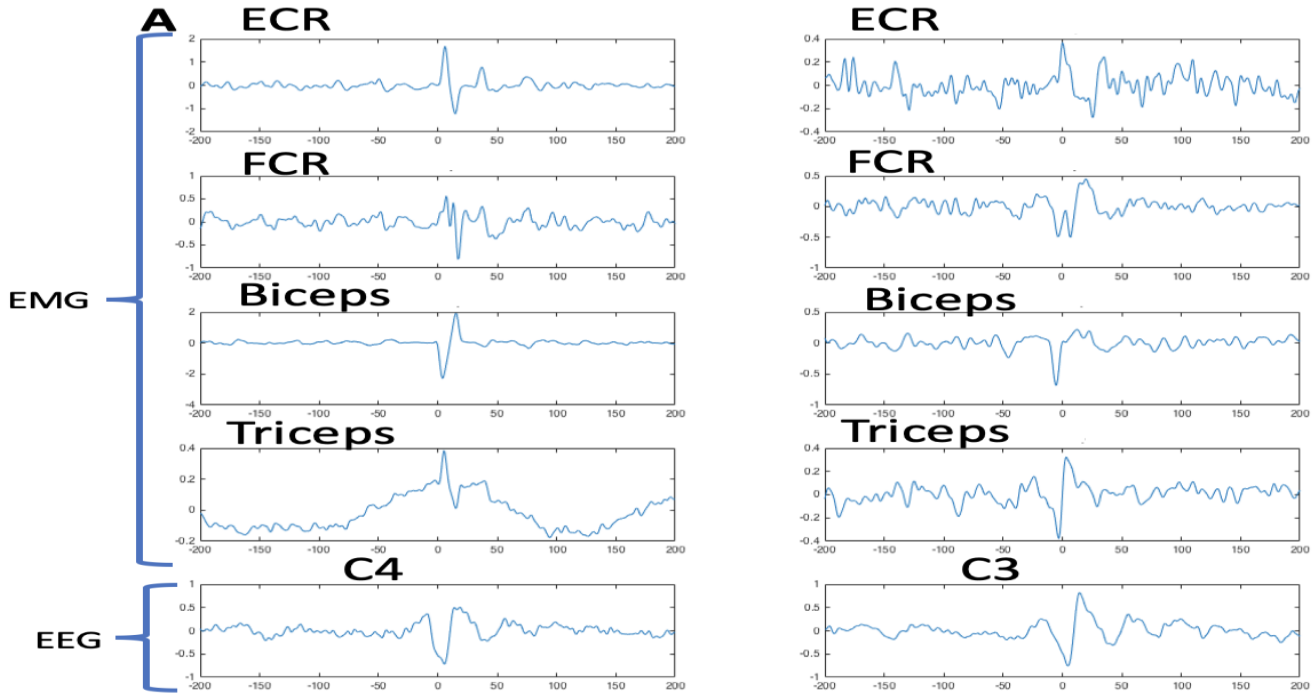
	Cortical Myoclonus	Brainstem/Reticular Myoclonus	Propriospinal Myoclonus	Startle Reflex
Phenomenology	Spontaneous jerks more commonly involving the face and distal upper extremities	Spontaneous jerks involving the entire body which can also be evoked by somatosensory stimuli to distal extremities	Arrhythmic, usually flexor, brief jerks involving the trunk, hips, knees in a fairly uniform pattern	Bilaterally synchronous flexor response to a startling stimulus
Reflex Physiology	Cortical onset short lasting EMG bursts (simultaneous agonist-antagonist discharge) with a cortical EEG correlate on back averaging.	Craniocaudal EMG discharge beginning at the lower medulla	Craniocaudal EMG discharge usually beginning at the level of the abdominal muscles; restricted to spinal cord propriospinal pathways	Early eyelid blink followed most consistently by SCM with subsequent cranio-caudal propagation
Afferent Stimulus	Spontaneous; but can be triggered by tactile or other somatosensory stimuli more commonly to distal upper extremities.	Spontaneous; can be induced by somatosensory stimuli, touch or muscle stretch of distal extremities	Spontaneous; but can be induced by tactile or auditory stimuli	Auditory; can also be elicited by visual, somatic and vestibular stimulation
Pattern of Muscle activation; Flexors vs Extensors	Mainly involving distal upper extremities and face (which have a large cortical representation)	Both flexors and extensors	Mainly flexors	Mainly flexors (but extensors also noted to be activated)
Site of origin	Cerebral cortex	Medullary reticular formation (likely nucleus reticularis gigantocellularis)	Limited to spinal cord (note that most cases are functional)	Caudal pontine reticular formation (nucleus reticularis pontis caudalis)
Velocity of bulbo-spinal efferent volley	~100 m/sec	> 50 m/sec	5-15 m/sec	~30 m/sec
EMG burst duration	< 50 msec	<50 msec	~150-450 msec (can be longer)	>70 msec

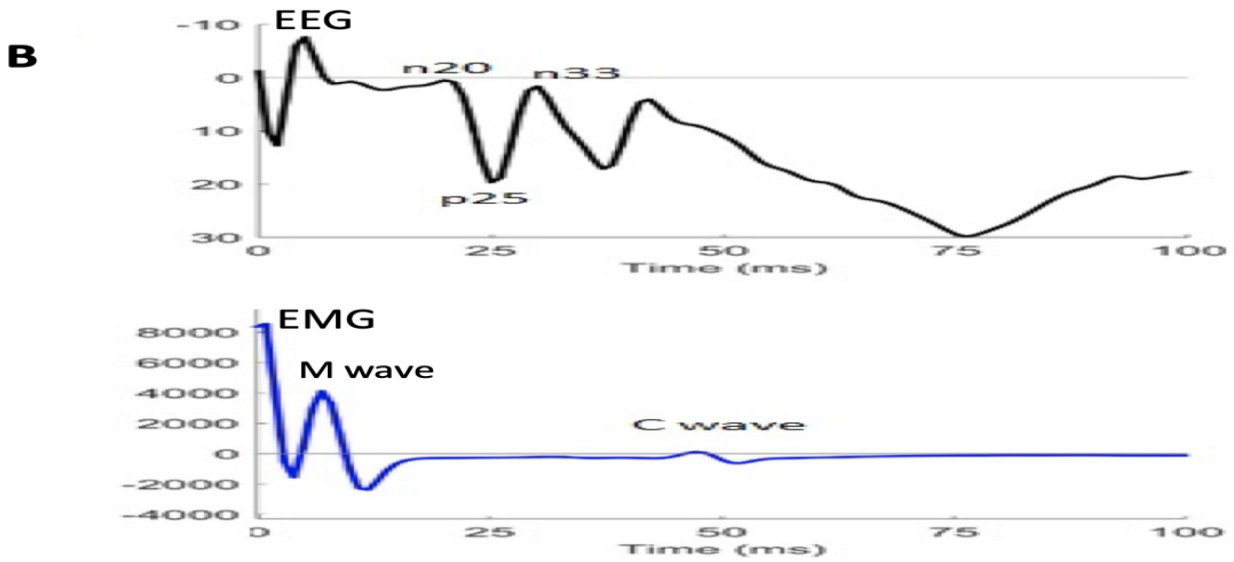
Original case of Post-anoxic myoclonus (Lance & Adams)

- Video

Courtesy Dr. Hallett

Cortical Myoclonus Physiology





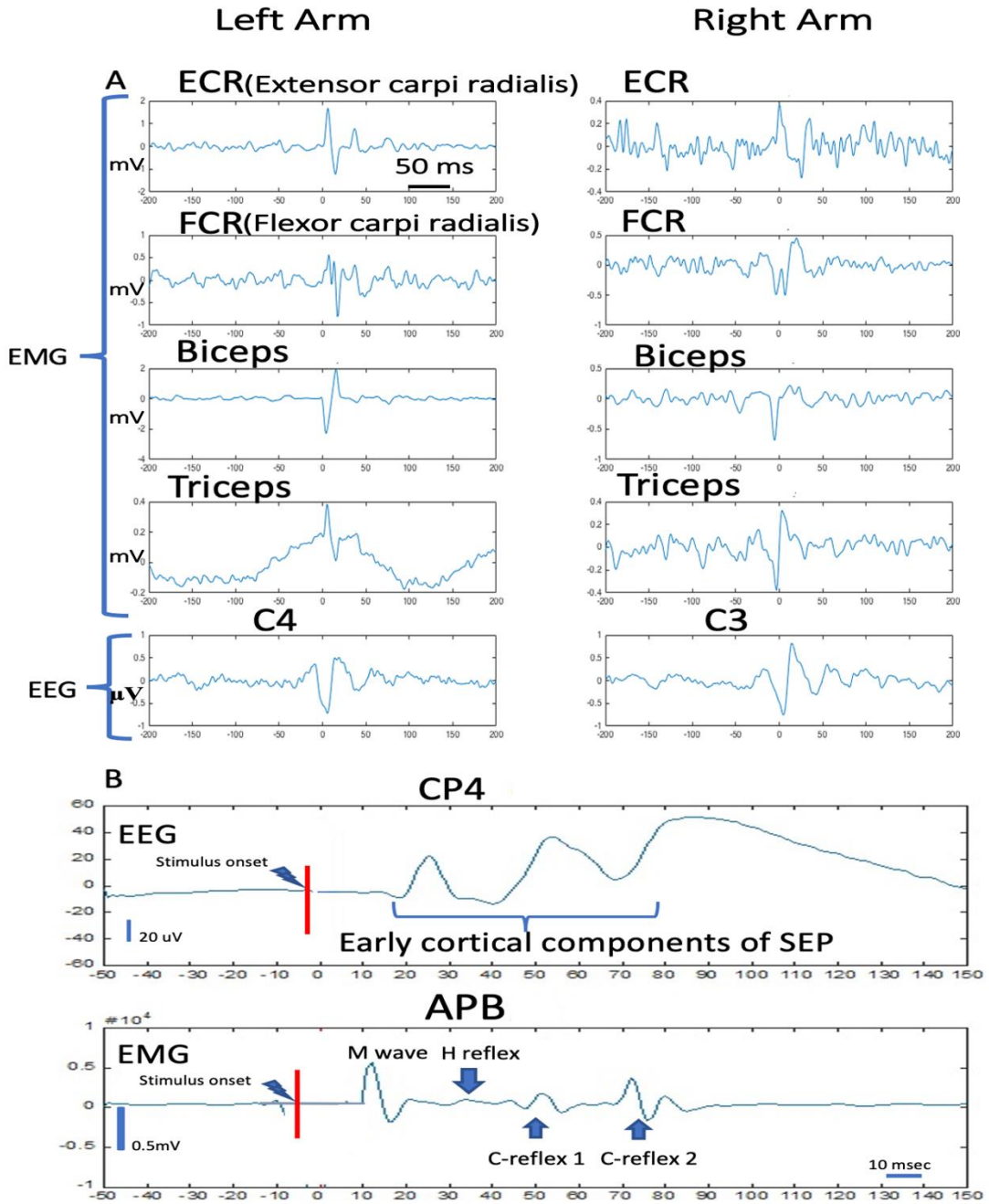
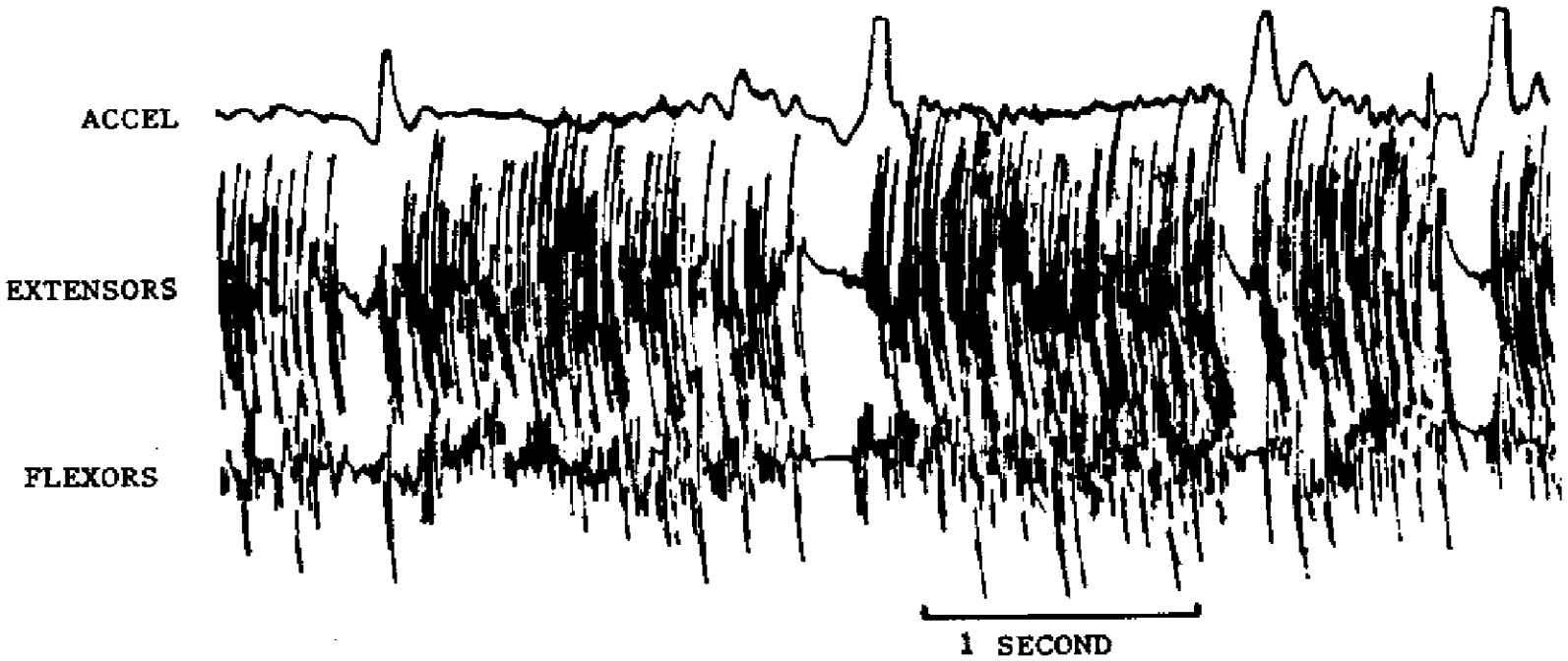


Figure. Cortical Myoclonus Physiology; A) EMG traces showing short duration (<30 msec) simultaneous agonist-antagonist bursts with cortical EEG spike correlate. B) Median nerve SEP showing giant early components of SEP on the EEG. Corresponding EMG recordings showing an M wave, followed by H-reflex response proceeded by exaggerated long-loop reflexes (C-reflex), reflective of disinhibited sensorimotor cortex.

Asterixis or Negative Myoclonus



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Reticular Myoclonus Physiology

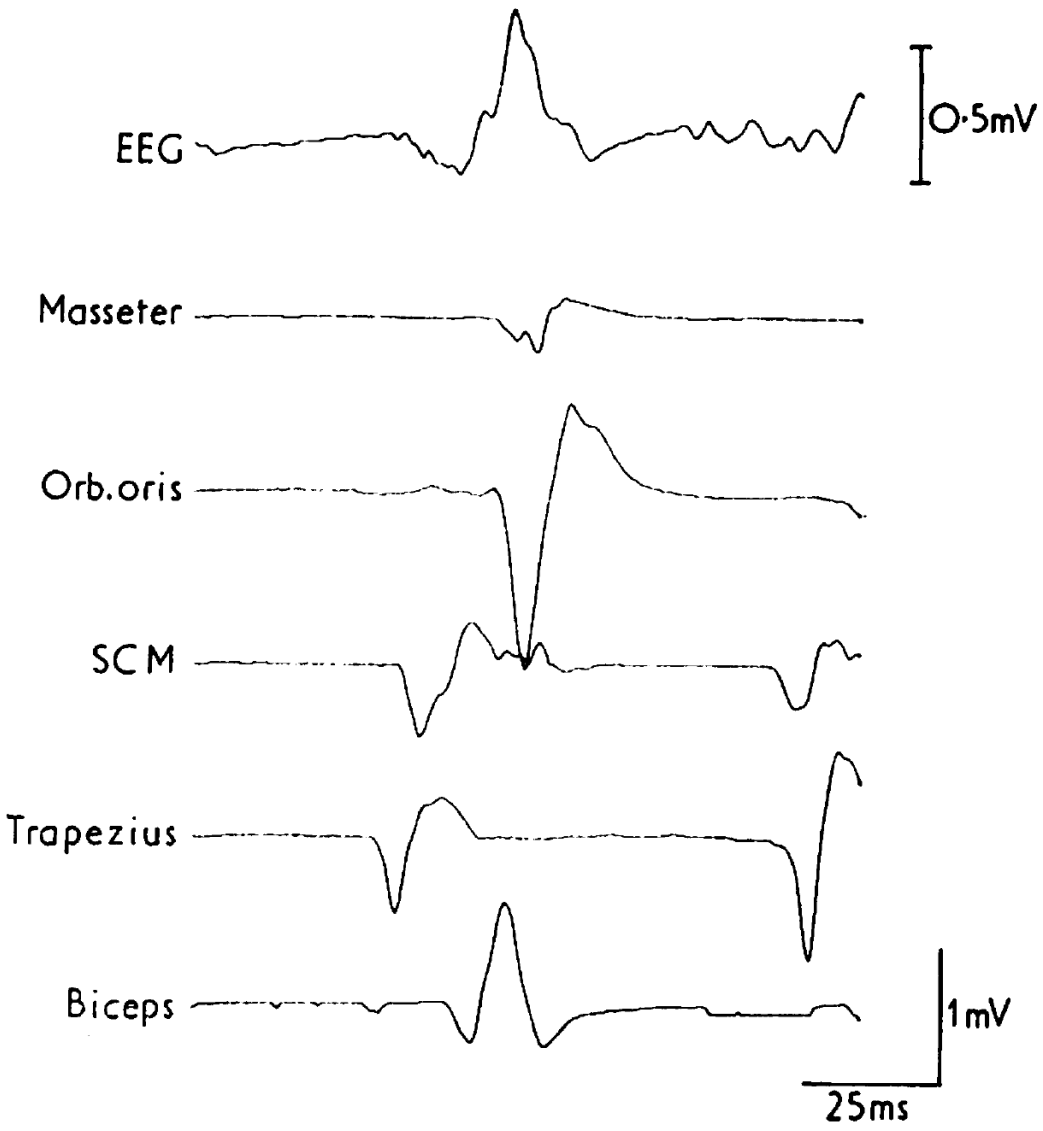

Fig. 2 *Electrical record of spontaneous myoclonic jerk including activity in cranial nerve muscles. The electroencephalographic recording (EEG) is from a point 1 cm to the left and 2 cm behind the vertex referred to a mid-frontal electrode (a positive deflection is downward). Other records are from the right masseter, left orbicular oris (Orb. oris), left sternocleidomastoid (SCM), right trapezius, and right biceps. Note the activation of the cranial nerves up the brain stem, and the onset of activity in trapezius before that in the EEG. The upper voltage calibration refers to the EEG record and the lower voltage calibration to all of the EMG records.*

Hallett et al. 1977

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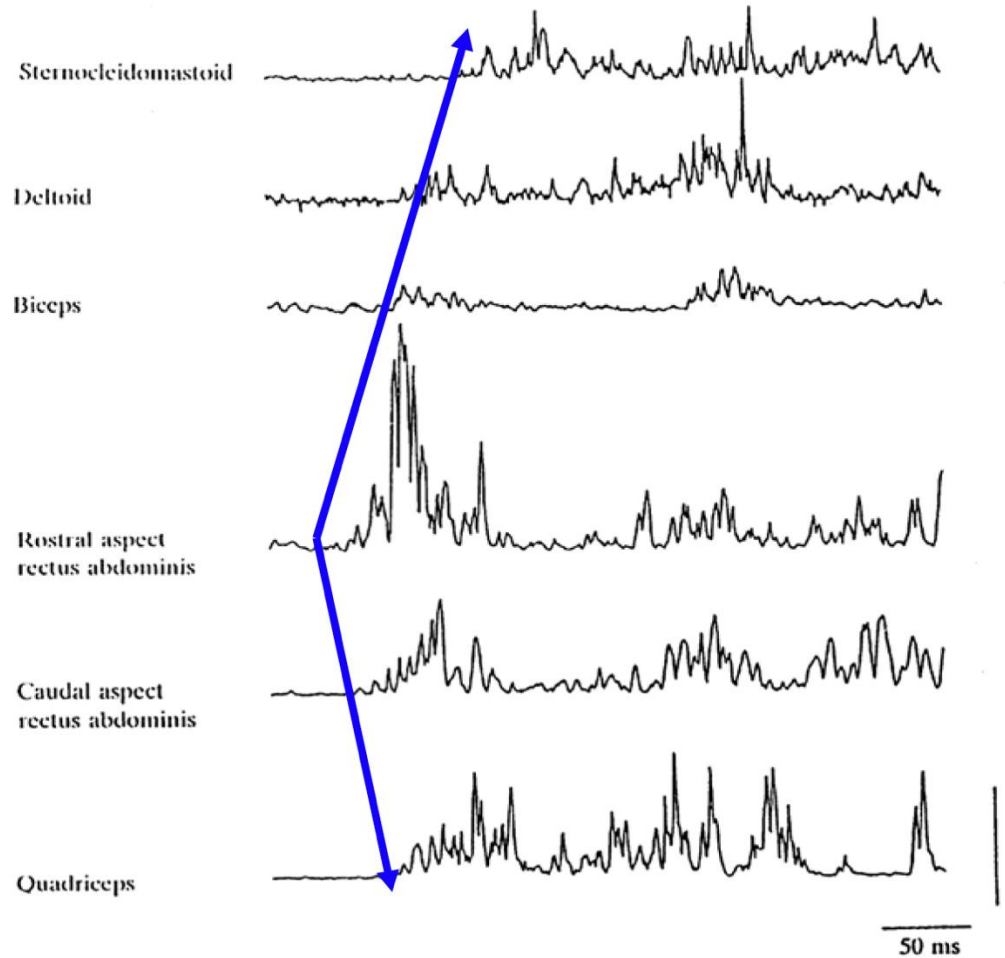
Propriospinal Myoclonus



Brown et al. 1991

Video PMD

Physiologic Features of Functional jerk-like movements (Psychogenic myoclonus)

- ***Longer burst duration >150 ms; similar to voluntary movements***
- ***Inconsistent spatial-temporal pattern***
- ***Variable pattern of spread (different origin and spread features)***
- ***Greater latencies than those seen with cortical or reflex myoclonus of cortical and brainstem origin***
- ***Variable latencies***
- ***Latencies can be longer than the fastest voluntary reaction time (Thompson et al. 1992)***
- ***Can find a normal looking BP (Bereitschaftspotential) prior to the movements (Terada et al. 1995)***

Startle Video

Startle reflex

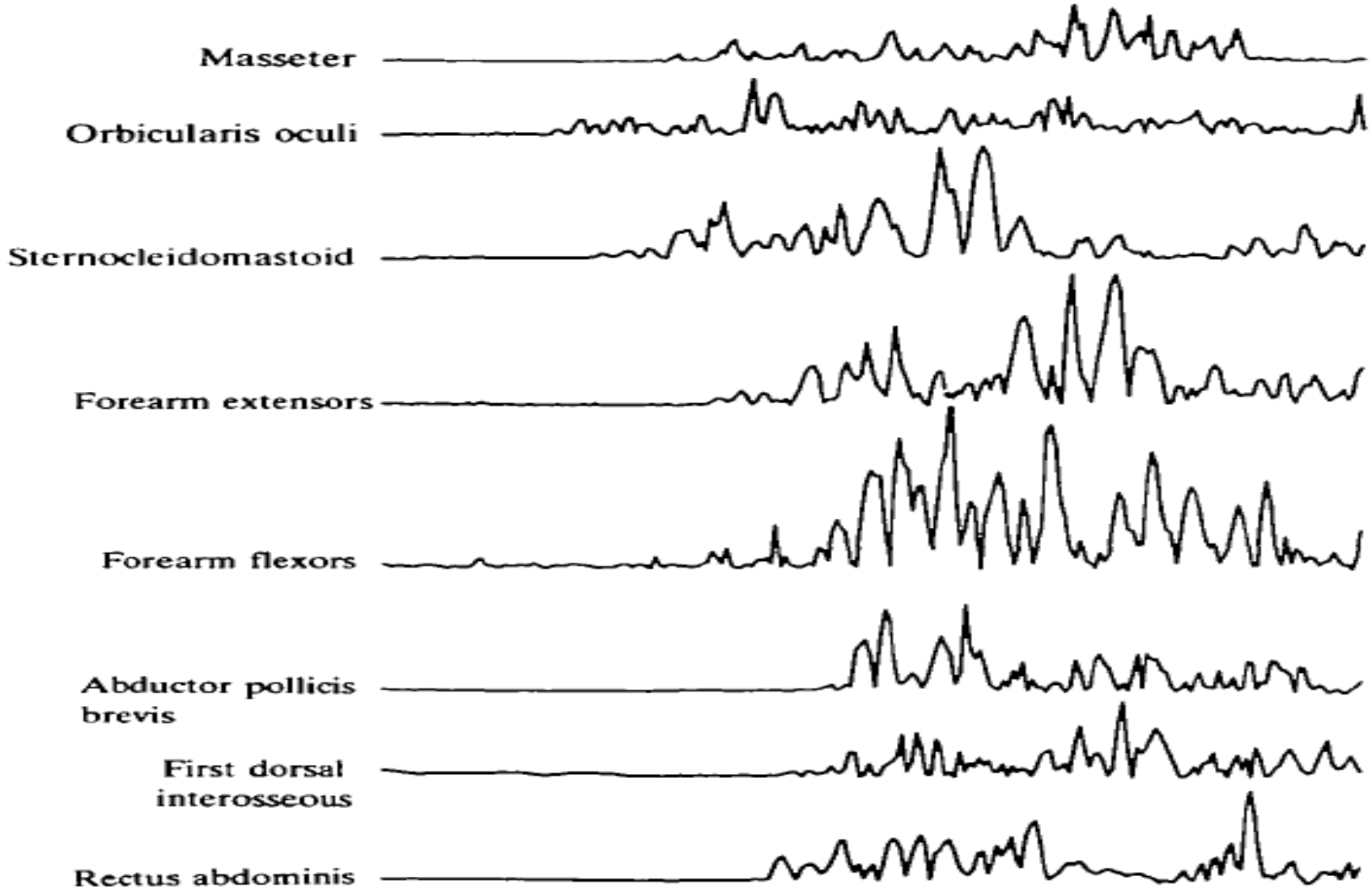
- **Clinical Characteristics of normal startle**
  - Eye closure, facial grimacing
  - Neck/trunk flexion
  - Arm abduction
  - Elbow flexion
  - Forearm pronation

Exaggerated Startle

- Clinical characteristics
  - Excessive and more widespread muscle activation
  - Lower thresholds for response
  - Impaired habituation

- Hereditary Hyperekplexia
  - Three cardinal features
    - Generalized stiffness at birth
    - Excessive startling
    - Temporary generalized stiffness after being startled

- Physiologic Characteristics
  - Excessive EMG bursts
  - Higher amplitudes and burst duration
  - Impaired habituation



Brown et al 1991

Mixed disorders (Myoclonus>Tremor)

Cortical Tremor/ Myoclonus

- CT Video

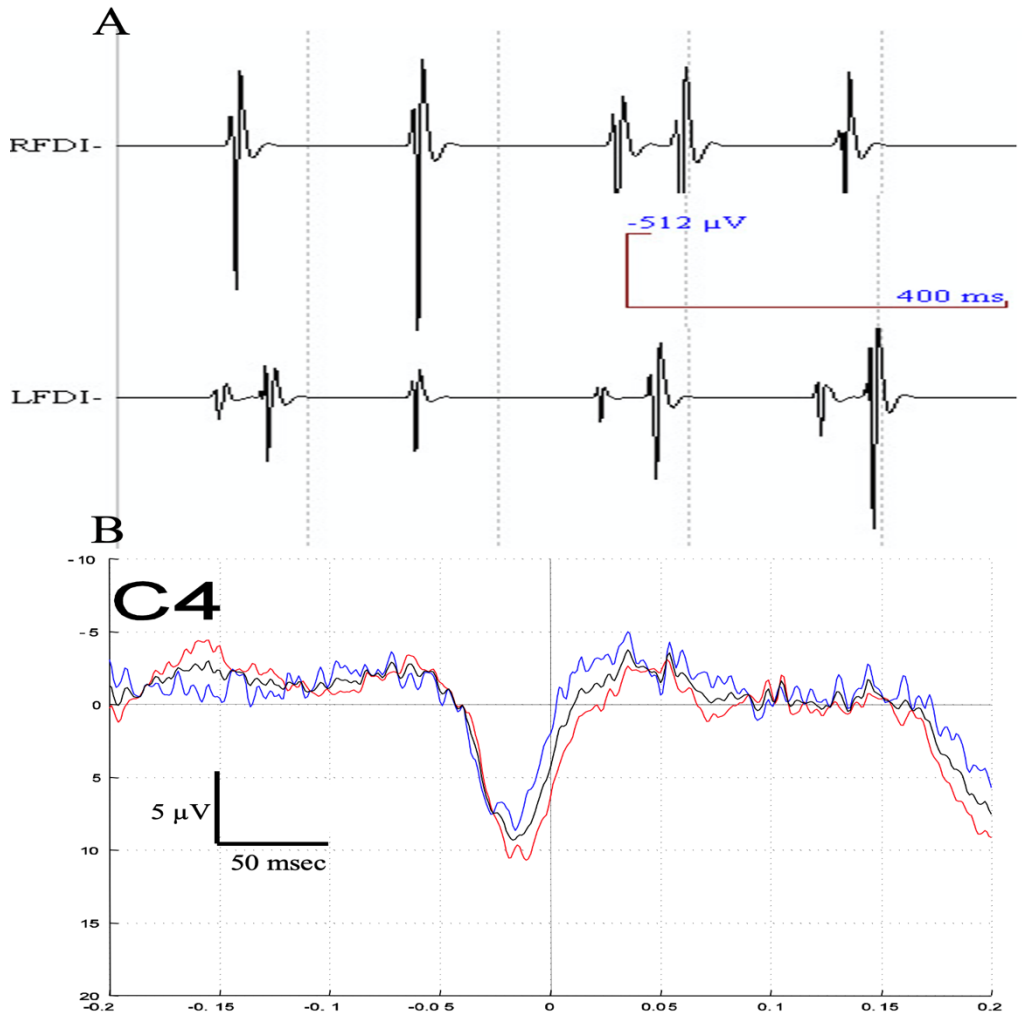


Figure . A. Short duration (<30 µsec bursts) arrhythmic EMG bursts recorded from bilateral FDI muscles at a frequency of ~10 Hz, B. Back-averaged cortical spike recorded from the contralateral sensorimotor cortex.

Brainstem (Palatal Myoclonus)

- Occurs fairly regularly (0.5-3 Hz) and typically presents with uvula and soft palate contractions.
- The tongue, face, larynx or neck can also be involved.
- Two forms
  - Essential palatal myoclonus
  - Symptomatic palatal myoclonus

- EPT video

Essential Palatal Myoclonus

Typically slower, limited to palatal movements (more prominent soft palate movements)

Tensor Veli Palatini

+Ear clicks (more common)

Disappear during sleep

Etiology: Idiopathic /Possibly functional

- SPT video

Symptomatic Palatal Myoclonus

Relatively faster, involves other muscles; pendular nystagmus, involvement of pharynx

Levator Veli Palatini

+Ear clicks (less common); more commonly present with other symptoms.

Persist during sleep

Etiology: Pathology in triangle of Guillain Mollaret; hypertrophic olives (Strokes, Neurodegenerative, MS, vasculitis)

Physiologic and Other Forms

- **Hiccups**
  - Myoclonus of the diaphragmatic muscles.
- **Sleep myoclonus**
  - Occurs in many normal people
  - Hypnagogic/hypnopompic
- **Toxin/drug-induced myoclonus**
  - Opiates, especially when combined with serotonin reuptake inhibitors.

Summary

- *Many different movement disorders have 'jerk-like' phenomenology and can superficially look the same.*
- *Different myoclonus subtypes have distinctive spatial and temporal physiologic characteristics which can be identified using surface EMG.*
- *Adding EEG and reflex/stimulus induced response can be useful and specific for certain types of myoclonus*
- *Neurophysiology is current 'gold standard' for classifying myoclonus subtypes.* ***Axis Ib***
- *CNP can provide useful pathophysiologic insights to guide workup for possible etiologies and guide treatment.*