Clinical Neurophysiological to Differentiate Jerky Movements: Basic Physiology

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Krembil
Relentless.

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UHN
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Toronto Western
Princess Margaret
Toronto Rehab

Disclosures

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Myoclonus

- Sudden, brief, jerky, shock-like movements involving extremities, face and trunk
- Unassociated with loss of consciousness
- Most myoclonus is due to muscle contraction (positive myoclonus)
- Negative myoclonus - sudden cessation of muscle contraction

Cortical myoclonus

- Spontaneous cortical myoclonus
- Cortical reflex myoclonus - elicited by stimuli (touch, muscle stretch, electrical stimulation)
- Cortical tremor/minipolymyoclonus
- Epilepsia partialis continua

Causes

Progressive myoclonic epilepsy, Alzheimer's disease, Creutzfeldt-Jakob disease, metabolic encephalopathy, corticobasal degeneration, Familial cortical myoclonic tremor with epilepsy (FCMTE), post-anoxic

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Cortical myoclonus

- 26 year old women
- Progressive history of tremoulousness of the arms and face, ataxia, seizure, cognitive decline and speech disturbances

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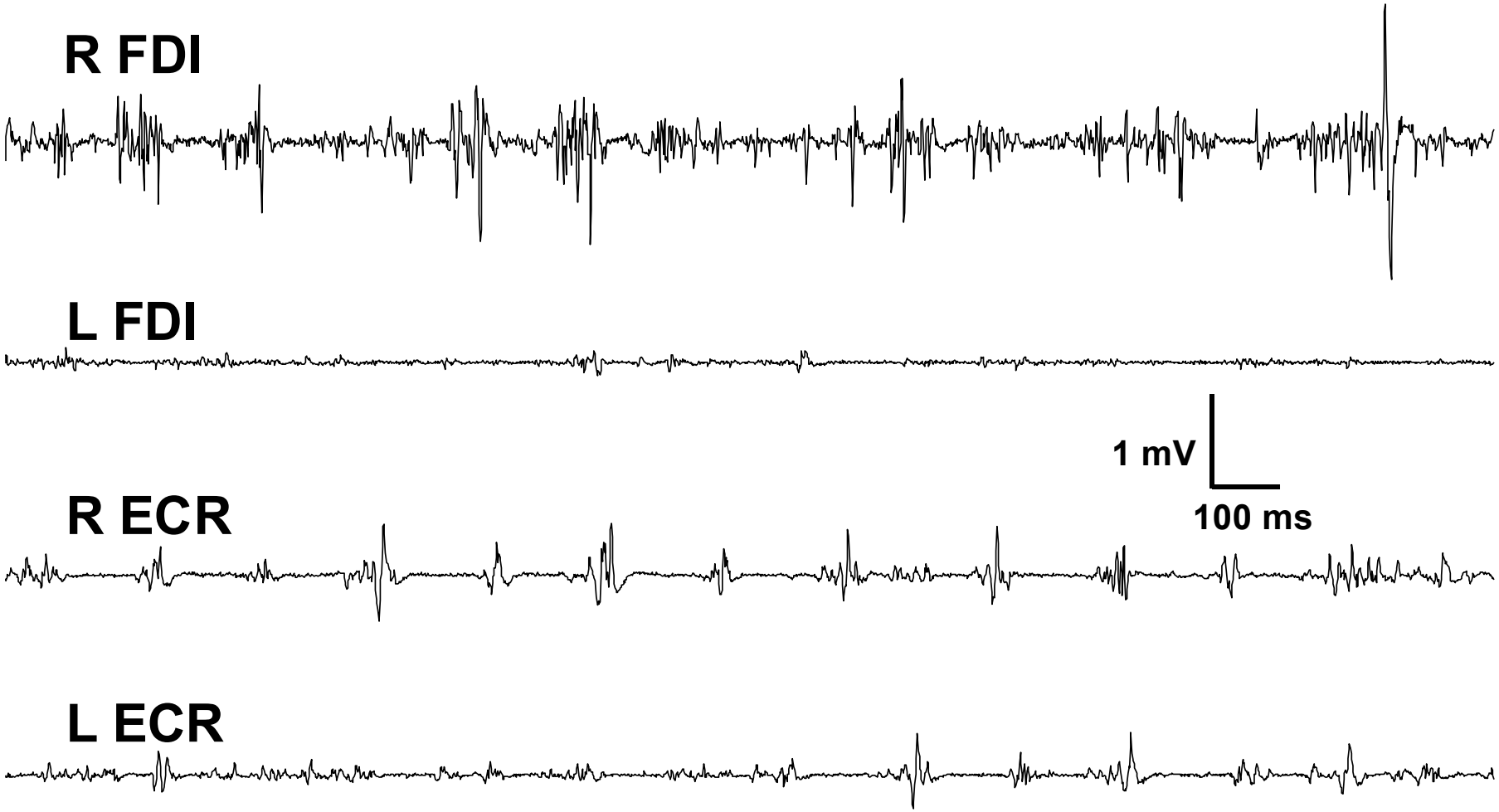
Cortical Myoclonus

EMG – cortical myoclonus



Cortical myoclonus
Polymyography - EMG findings

- Duration: short duration EMG bursts (20-70 ms),
- Recruitment pattern: Cranial-caudal (like TMS) rapid conduction – near synchronous EMG discharges in nearby muscles
- Distribution: Distal - most prominent in hands (feet), face

EMG burst in cortical myoclonus

Substantiating the Short Burst Duration in Cortical Myoclonus

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- 8 patients with cortical myoclonus
- 19 patient with non-cortical myoclonus (myoclonus-dystonia) phenotype

Van der Veen et al, Mov Disord 2024

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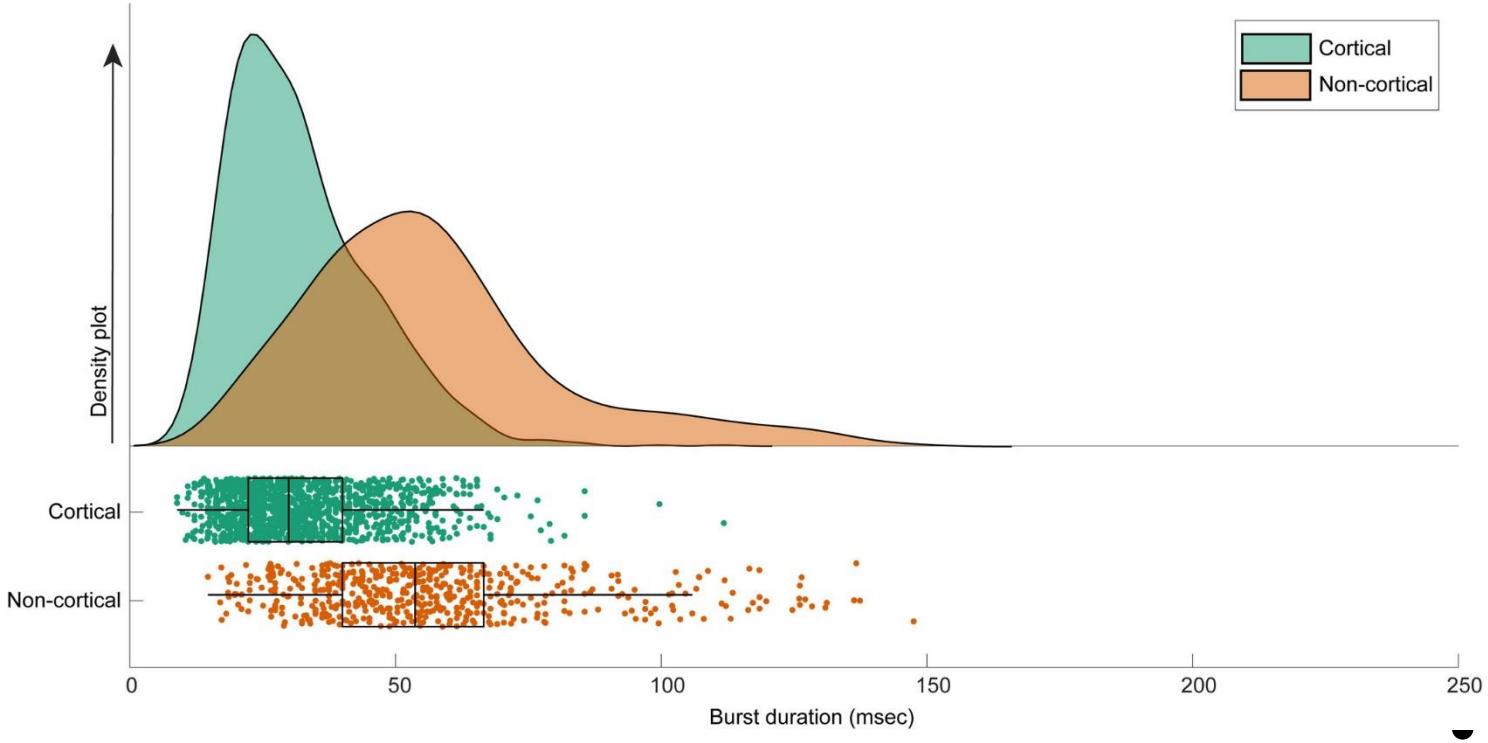
EMG burst in cortical myoclonus



- Cortical myoclonus 31.1 ms
- Non-cortical myoclonus 56.7 ms
- Threshold of 45 ms – sensitivity 100%, specificity 89.5%
- Confirm short duration EMG burst in cortical myoclonus

Van der Veen et al, Mov Disord 2024

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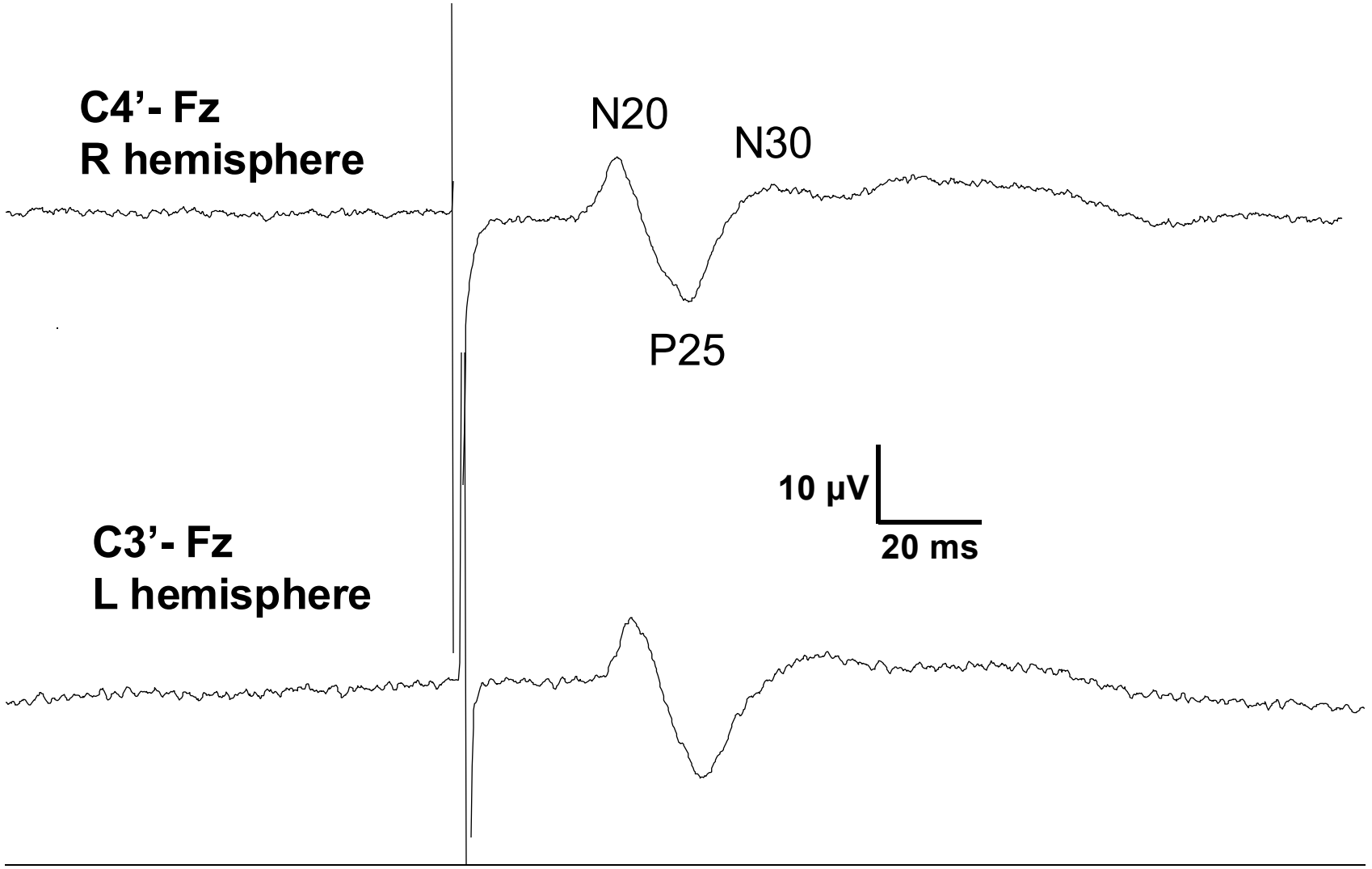
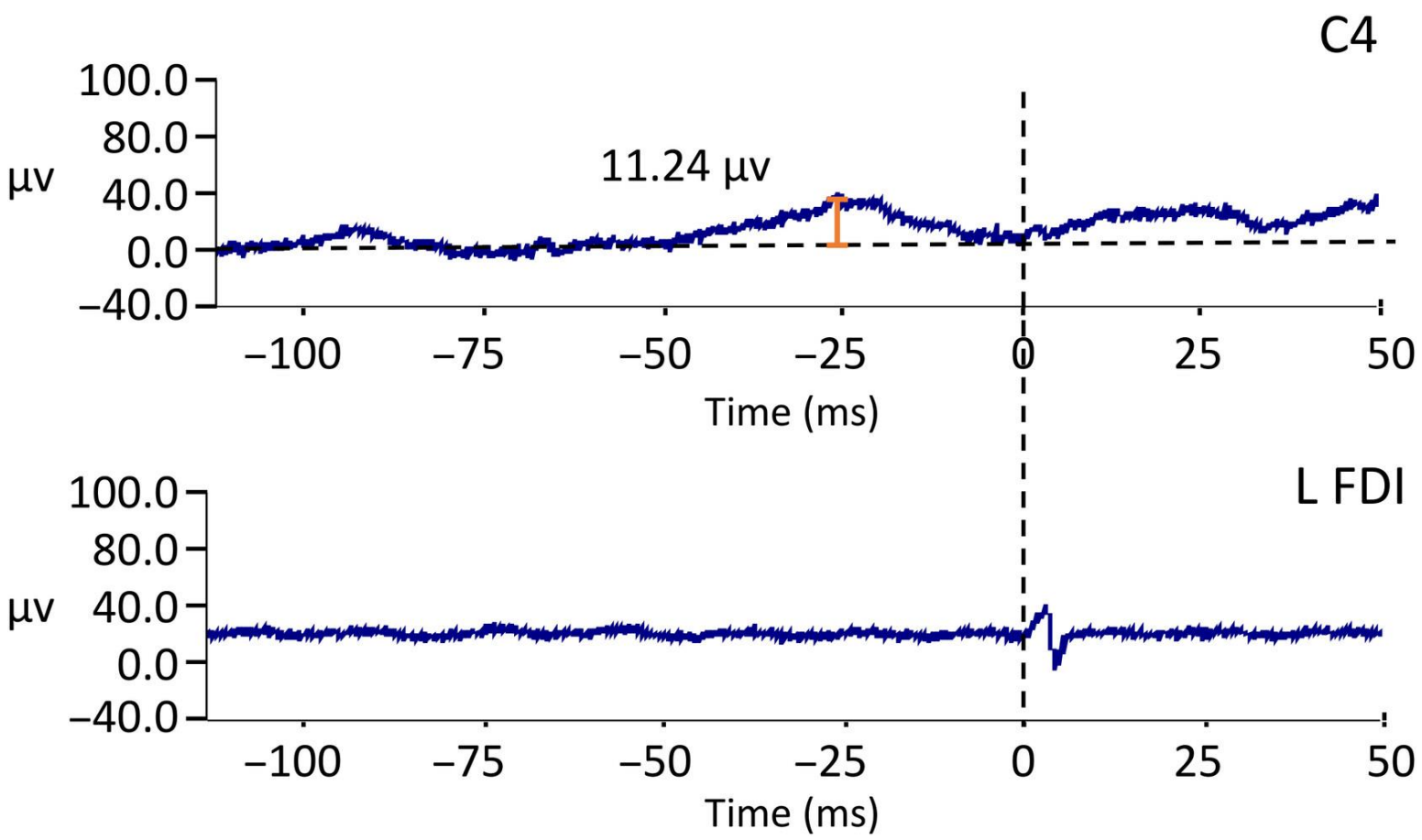
Cortical myoclonus
EEG – EMG (polymyography) finding

- Spike ± wave in EEG
- “Giant” SEP
- EEG backaveraging – time-locked short latency (~ 30 ms for hand) cortical correlate preceding movement

Giant median nerve SEP



Jerk lock backaveraging



- 54 year old woman
- Jerky movements and cortical myoclonus
- Spontaneous contractions in the left FDI muscle
- Average of 18 trials

Grippe & Chen, Mov Disord Clin Pract. 2023

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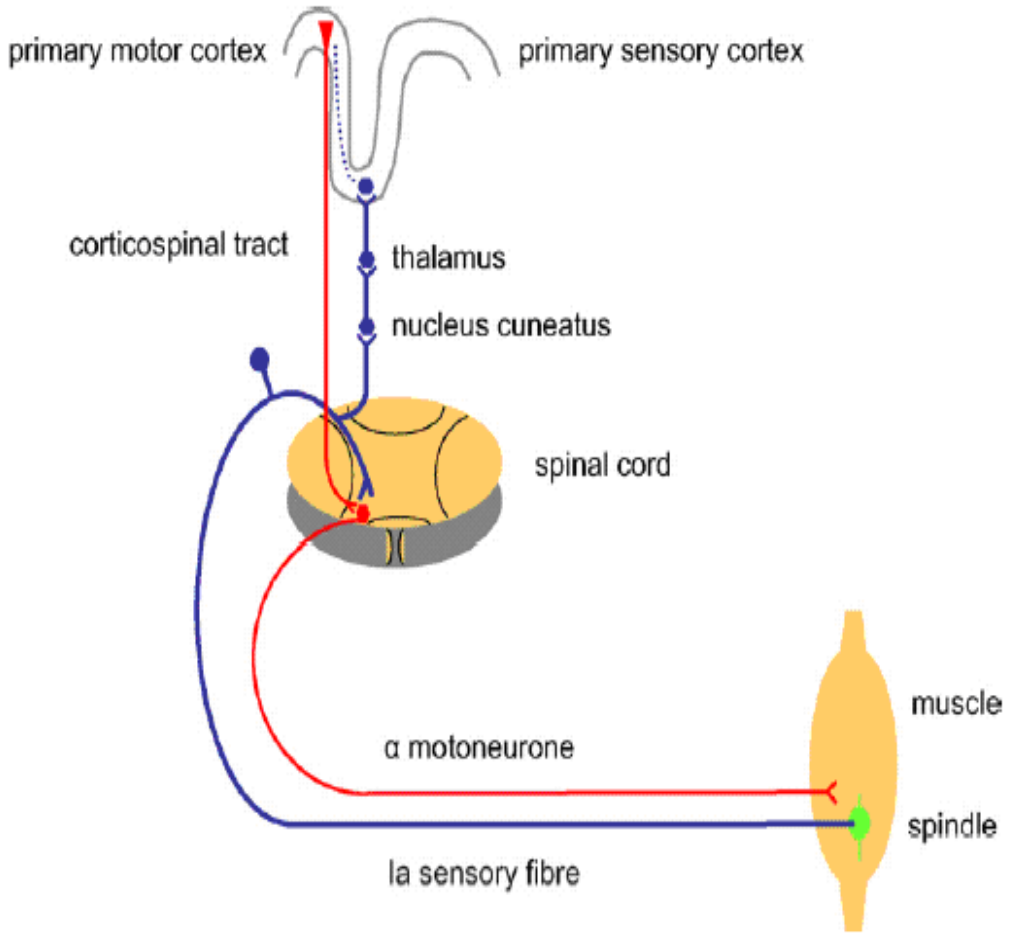
Stimulus- sensitive myoclonus

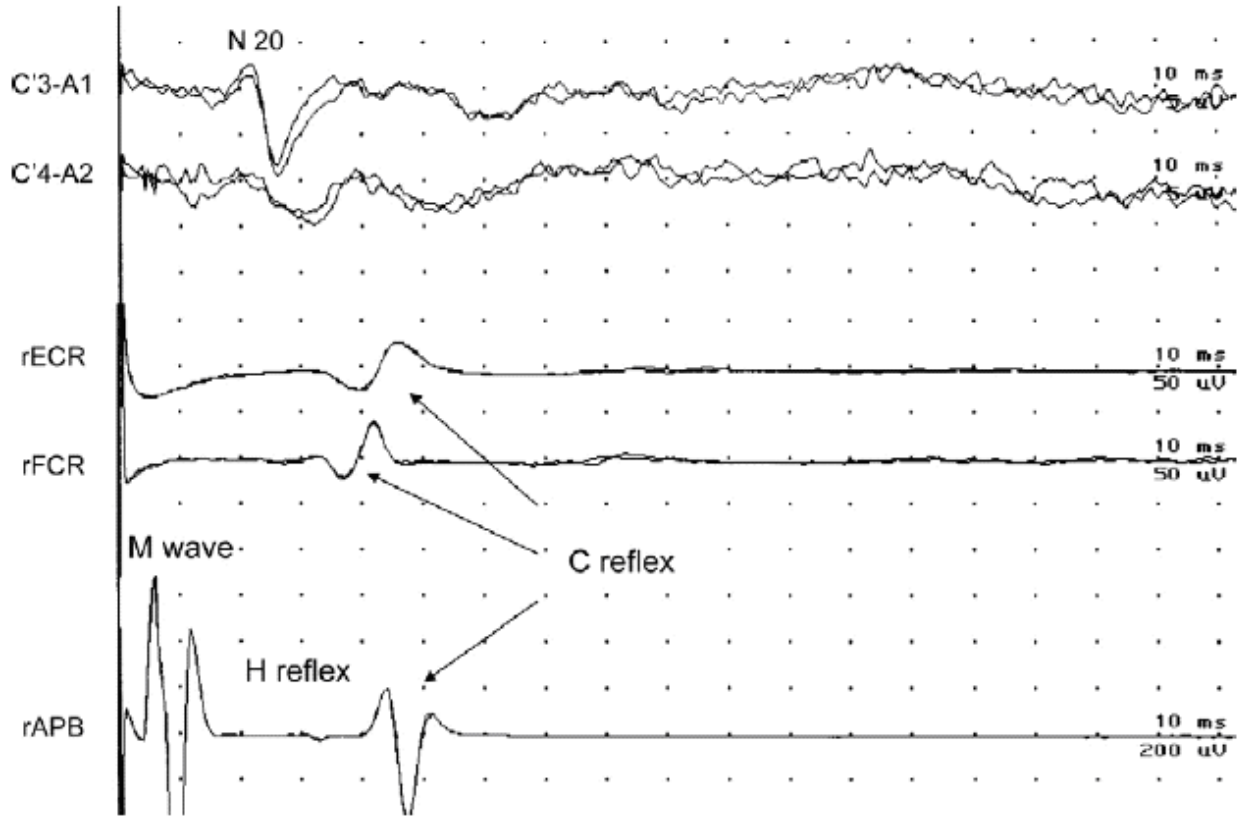
- 65-year-old women with stimulus sensitive myoclonus

Physiological studies

- Median nerve SEPs normal
- EEG backaveraging not possible because there was no spontaneous jerks
- Action myoclonus: short duration (20-70 ms), synchronous EMG discharges
- Long latency, or C (cortical) reflex: Stimulate median nerve at wrist (mixed nerve) or cutaneous nerve at middle or index finger

Median nerve stimulation: Long latency reflex





Cutaneous reflexes

- Cutaneous (long latency, or C (cortical)) reflex: Stimulate middle finger at 3x sensory threshold, average 50-100 trials

- Long latency (transcortical reflex) latencies

  - hand 40-50 ms
  - foot ~90 ms

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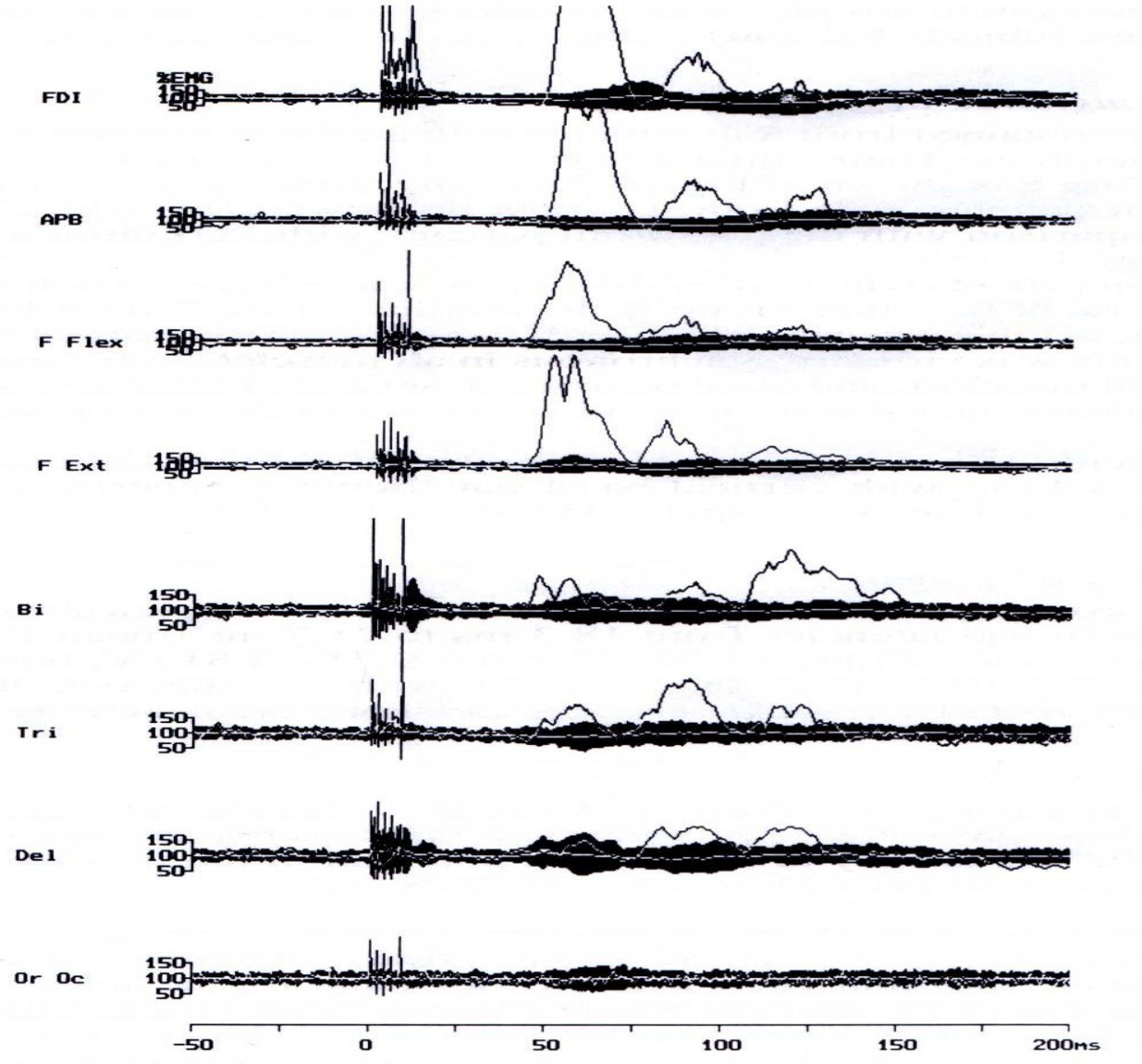
Myoclonus in corticobasal syndrome -Exaggerated transcortical reflex



Chen et al., Brain, 1992

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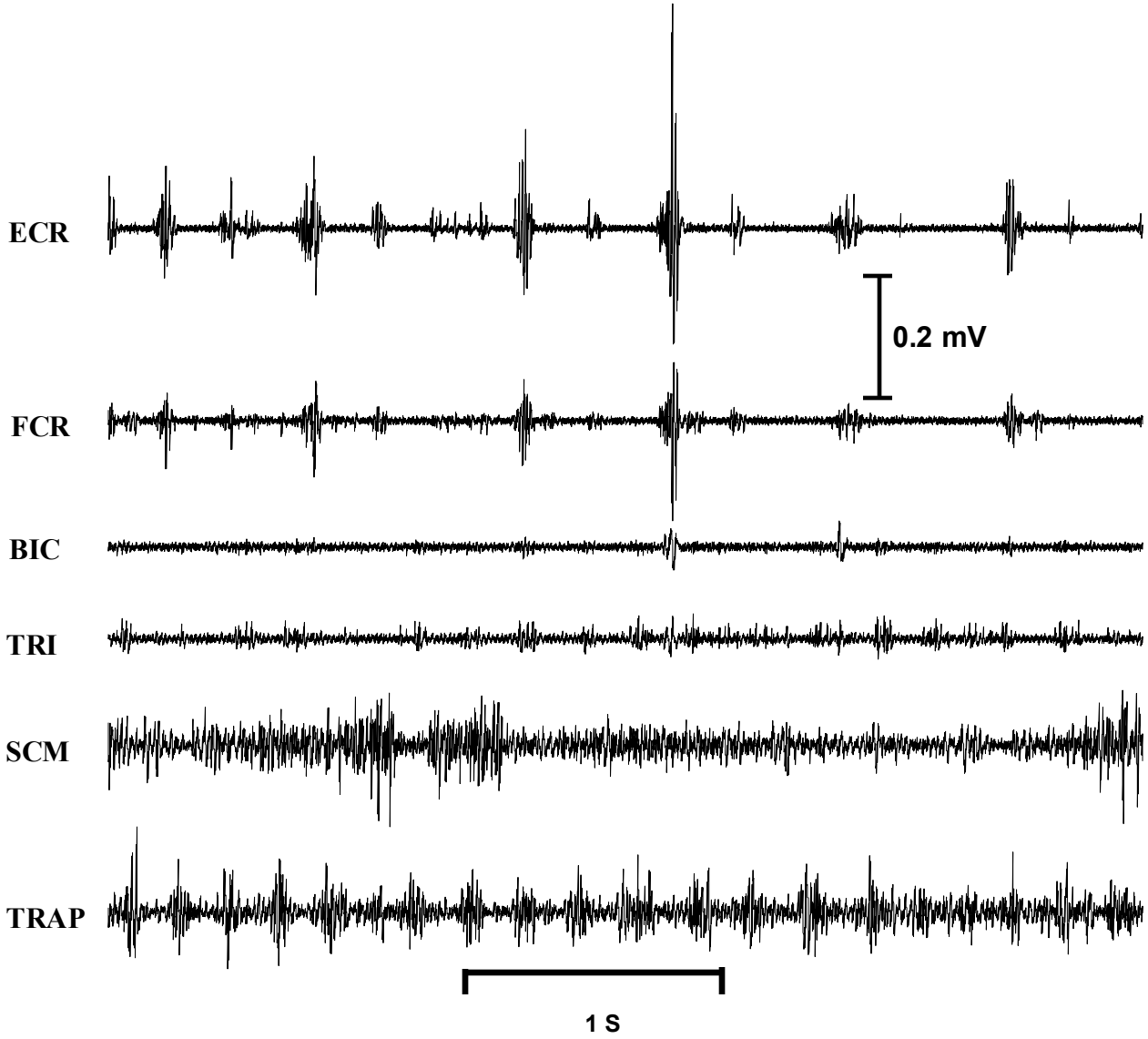
Myoclonus-Dystonia (essential myoclonus)

- EMG burst duration ranged from 30 to 700 ms
- No giant SEP, jerk-locked cortical correlates, exaggerated long-latency reflexes
- Transcranial magnetic stimulation studies showed normal cortical inhibition
- Subcortical myoclonus

Myoclonus-Dystonia (essential myoclonus)



Li et al, Mov Disord 2008

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Jerks- R arm movement

- 61 year old man
- 7 year history of abrupt movements of the right side

Jerks R arm (video)

Question:
This patient has

A Cortical myoclonus

B Tics

C Dystonia

D Functional movement disorder

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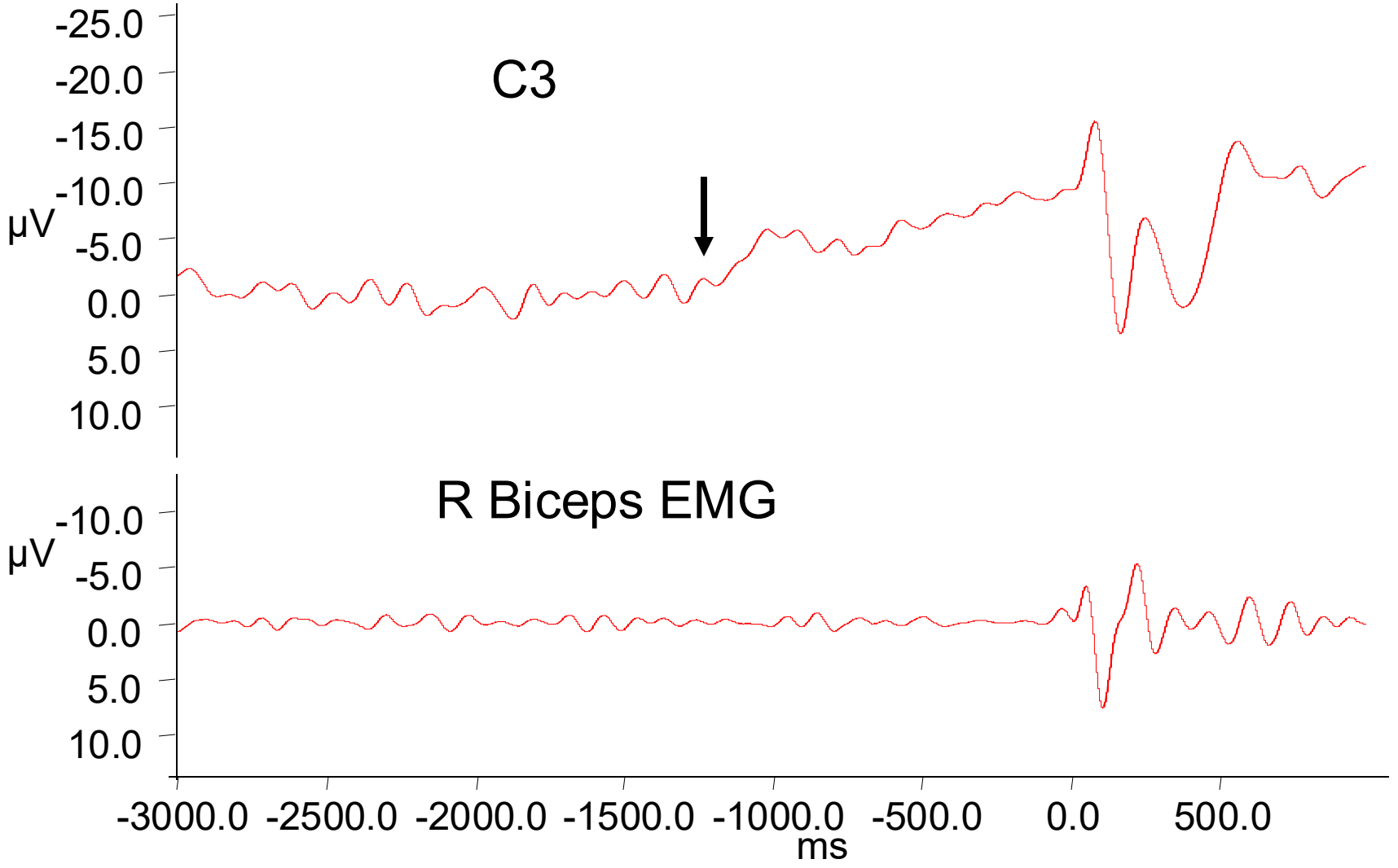
Jerks- R arm movement

Physiological findings

- EMG burst durations >100 ms

Movement muscle jerks – cortical premovement potentials (BP)



Functional myoclonus/jerks

Physiological studies

- “Organic” findings (e.g. giant SEP, short latency cortical discharge before EMG onset) cortical absent
- Variable pattern – variable onset and intermuscle latency
- EMG duration prolonged (> 100 ms)
- Distractibility
- May be preceded by a premovement (Bereitshaftspotential) or desynchronization of EEG
- Suggestibility

Cases

Abnormal abdominal movements

Case 1

- 60-year-old construction worker
- 6 months: confusional episode, unsteadiness – seizure
- 5 months: memory alterations, stopped working due to poor balance and weakness
- 3 months: irritable, sleep for 2-3 hours, episodic urinary incontinence
- 1 month: episodes of hiccups and bizarre behavior
- Presented with worsening behavior and cognitive function, aggressive behavior

Examination:

- Able to speak but disoriented
- Slight dysmetria in arms and legs, stooped posture, wide based gait

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ELECTROPHYSIOLOGICAL RECORDING

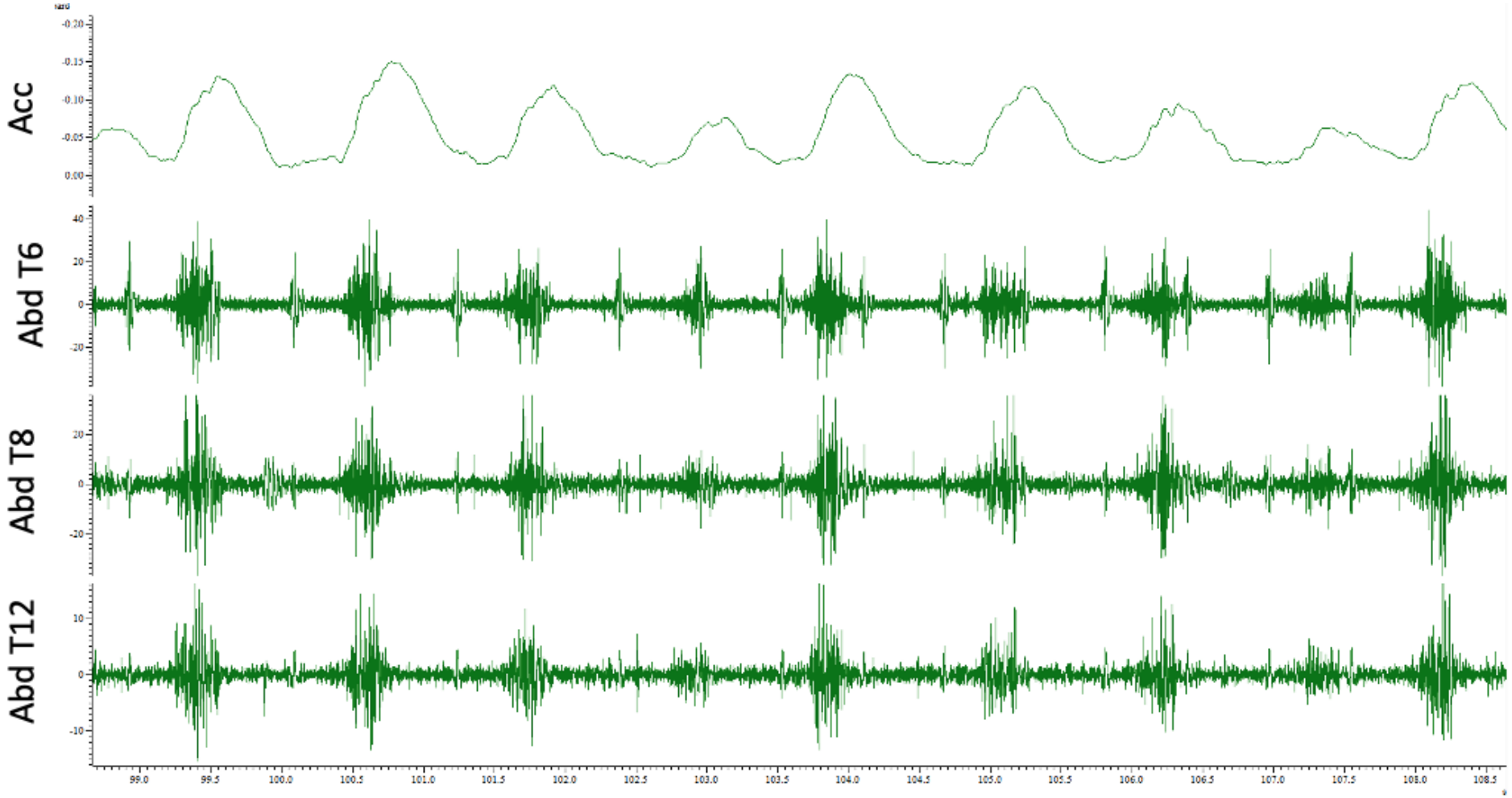

Figure 1: Surface EMG (3 bottom channels) and accelerometer (top channel) show contraction of the rectus abdominis muscles with a mean burst duration of 350ms and frequency of 1Hz.

After use of lorazepam 1mg

ELECTROPHYSIOLOGICAL RECORDING

- No involvement of the paraspinal muscles and hamstrings (no visual movements observed)
- No consistent muscle activation order
- No movement triggered by sound or sensory stimulation
- Occurs during sleep

Myorhythmia/
Spinal segmental myoclonus

DIAGNOSIS: ANTI-CASPR2 ENCEPHALITIS

Abnormal abdominal movements Case 2

- 81-year-old women with cardiac pacemaker
- Abnormal abdominal movements for 3 months
- Intermittent, no known triggering factor
- Found the movement bothersome, but otherwise no other complain

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Abnormal abdominal movements

Case 2

This patient has

A Tics

B Spinal segmental myoclonus

C Diaphragmatic myoclonus (tremor)

D Functional (psychogenic) movement disorder

E Pacemaker lead displaced, pacing the diaphragm

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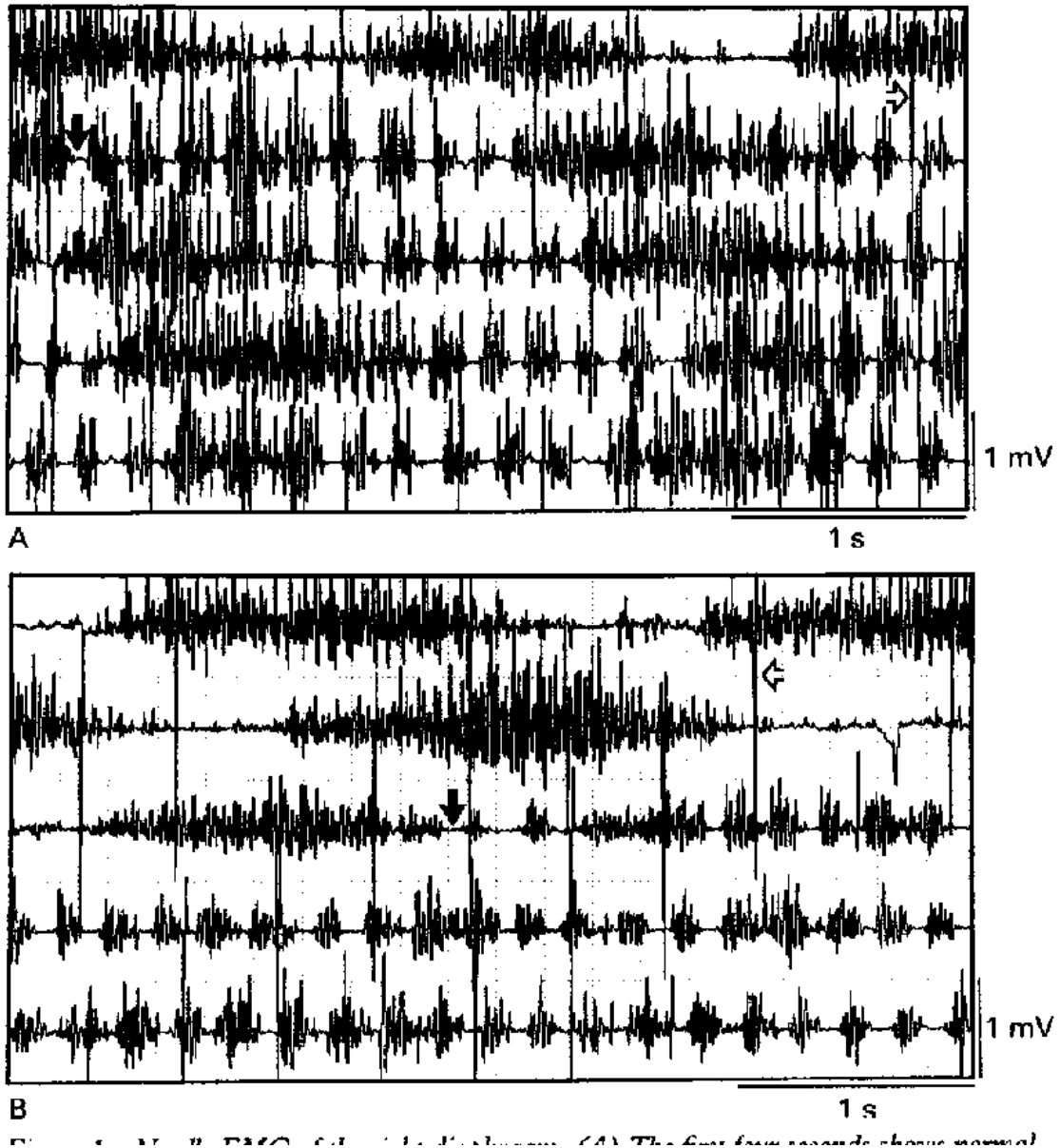
Abnormal abdominal movements

Diaphragmatic myolconus (tremor)



Chen et al, JNNP 1995

Isolated diaphragmatic tremor

Is there a spectrum in "respiratory myoclonus"?

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VIDEO

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ABSTRACT

Background: Respiratory myoclonus or diaphragmatic flutter is an unusual movement disorder with abnormal diaphragmatic activity, which may be associated with respiratory symptoms. The effects of distracting maneuvers on diaphragmatic activity have not been investigated.

Methods: Two patients with nondisabling abdominal movements of suspected diaphragmatic origin were studied with surface and needle electromyography (EMG).

Results: The abdominal movements resulted from isolated, rhythmic diaphragmatic contractions with variable EMG burst duration, suppressibility with breath-holding and distracting maneuvers, and other attributes of volitional control.

Conclusion: "Respiratory myoclonus" may be a heterogeneous disorder ranging from synchronous movements of the diaphragm and other respiratory muscles associated with respiratory compromise, to diaphragmatic movements under at least some volitional control with no respiratory or functional disability. The latter group could be designated phenomenologically as "isolated diaphragmatic tremor." *Neurology*® 2007;69:689–692

Summary

Useful clinical electrophysiological techniques

Muscle Jerks

1. EMG: Burst duration, muscle recruitment order
2. Jerk locked back averaging
3. Long latency reflex (C-reflex)
4. SEP studies

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Neurophysiological studies and jerks

Neurophysiological studies can determine

- Is it myoclonus?
- Site of origin
  1. Cortical
  2. Subcortical
  3. Spinal
     - Segmental myoclonus
     - Propriospinal myoclonus