Pan American Movement Disorders Clinical Neurophysiology Course
May 1-3, 2025 | BMO Education & Conference Centre

Clinical cases of “other” hyperkinetic disorders

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Faculty/Presenter Disclosure

- **Relationships with commercial interests:**
  - **None**

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Disclosure of Commercial Support

- This program has received financial support from Abbvie, Ipsen, Natus, Cadwell and Medtronic in the form of sponsorship to support logistic costs.

- Potential for conflict(s) of interest:
  - None

Case 1

- 71 year-old woman

Case 1

- 71 year-old woman

Case 1

- “Restless” for 20 years noted by friends but she was unaware of the movements
- Worsening in the recent years, movements started to bother her
  - Impairs her daily activities
- No urge preceding the movement and no relief sensation following it
- Able to volitionally suppress the movements, but has to actively contract the muscle

- **Phenomenology?**

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Case 1

- Chronic headache
- **Long history of Depression** (since her 20's)
  - Venlafaxine, Nortriptyline, Gabapentin

- Mother had similar movements

- Genetics (HD, C9orf72) - Negative

Chorea vs. Tardive dyskinesia

- **Chorea**
  - Continuous and random flow of muscle contractions, characterized by floating, chaotic movements.
  - Unpredictable, Suppressible?

- **Tardive dyskinesia**
  - Involuntary movements of the mouth, jaw, tongue, face, trunk, or extremities, often in combination
    - Oral–buccal–lingual, “piano-playing”movements of fingers, toes, possible involvement of trunk

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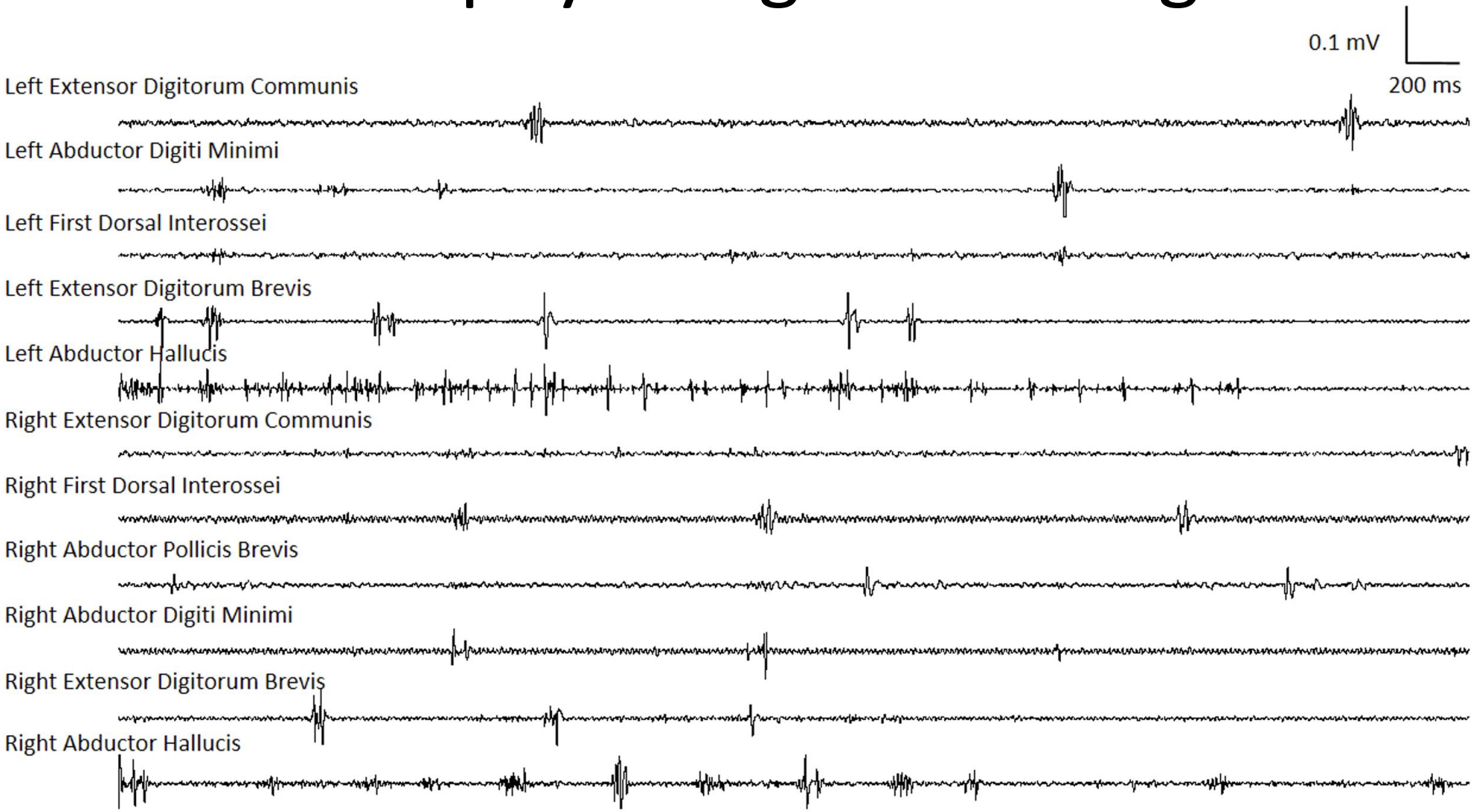
Hauser RA et al. CNS Spectr. 2022

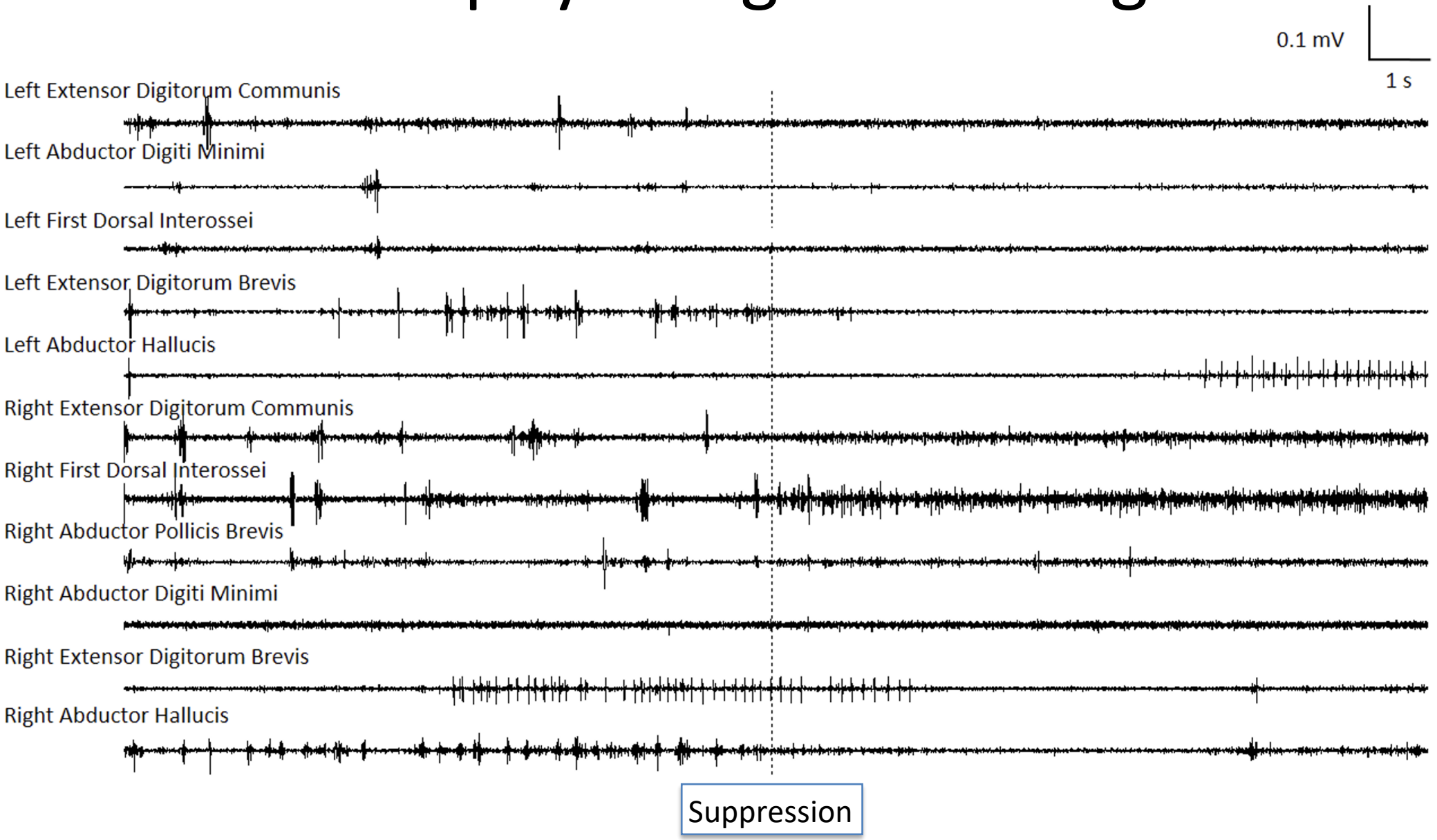
Cardoso F. Mov Disord Clin Pract. 2014

Neurophysiological Findings



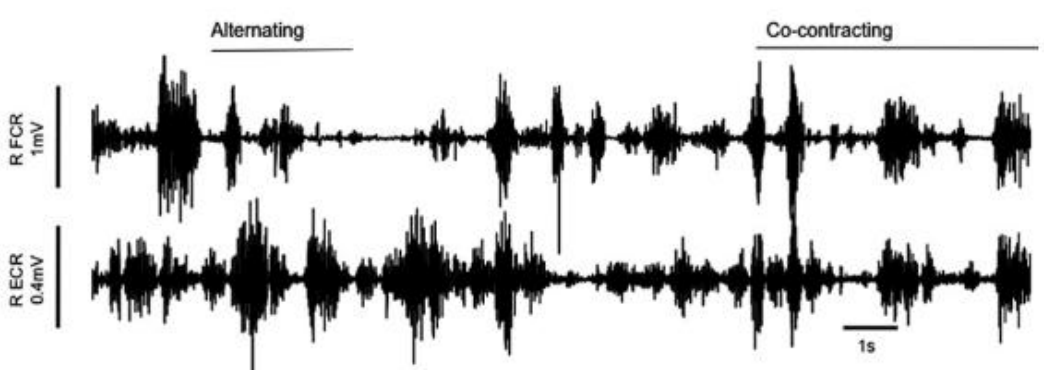
Duration: 60 - 160 ms

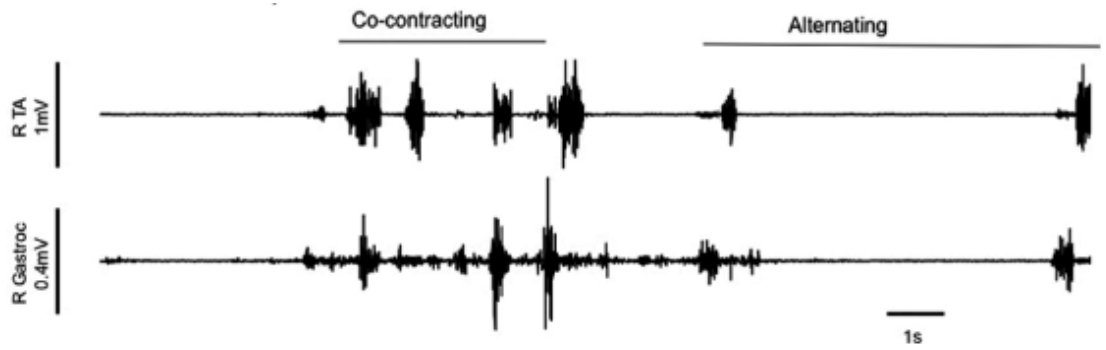
Neurophysiological Findings



Neurophysiology of Chorea

- Surface EMG:
  - Continuously changing pattern of activation in different muscles, with constant varying duration of the EMG bursts, unpredictable;
  - Duration approximately 100 ms, up to 1500 ms in ballismus (only case series):





Hallett M et al. J Neurol Neurosurg Psychiatry. 1981

Tai YC, et al. Mov Disord Clin Pract. 2016

Partial volitional supressibility - Chorea

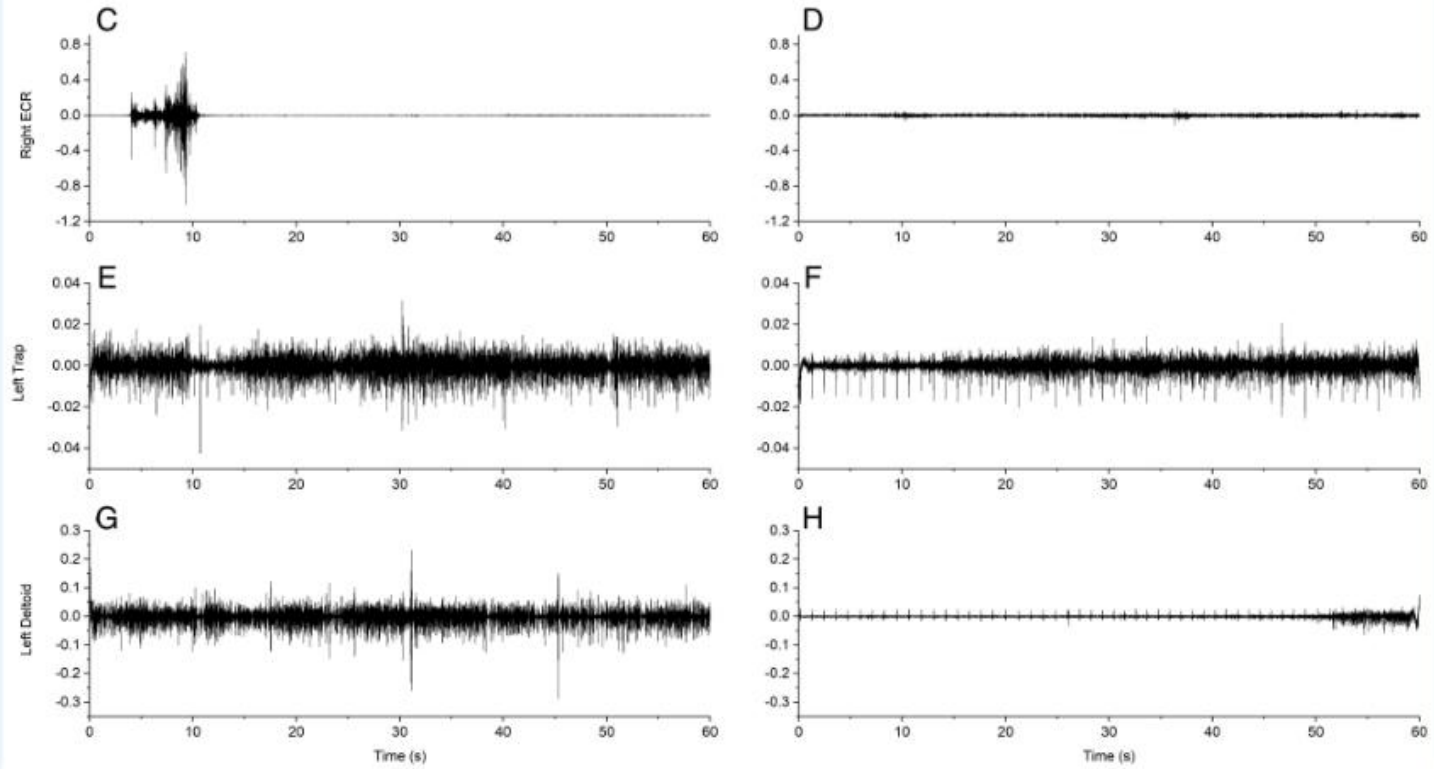


FIG. 1. EMG recording of case 3 during free to move condition on the left column (A,C,E,G), and involuntary movements' suppression condition on the right column (B,D,F,H). ECR, extensor carpi radialis; SCM, sternocleidomastoid; Trap, trapezius.

Hallett M et al. J Neurol Neurosurg Psychiatry. 1981

Tai YC, et al. Mov Disord Clin Pract. 2016

Case 2

- 21 year-old man

Case 2

- 21 year-old man

Case 2

- Paroxysmal febrile weakness episodes since he was 2.5 years old
- Involuntary jerky movements of neck and trunk started after a weakness episode, when he was 12 years old
  - Feels relief of a build-up sensation after performing the movement, but he has no sense of agency and cannot suppress it
  - Movements worsen when he is stressed or anxious
  - Pathogenic ATP1A3 variant (p.R756L)
- **Phenomenology?**

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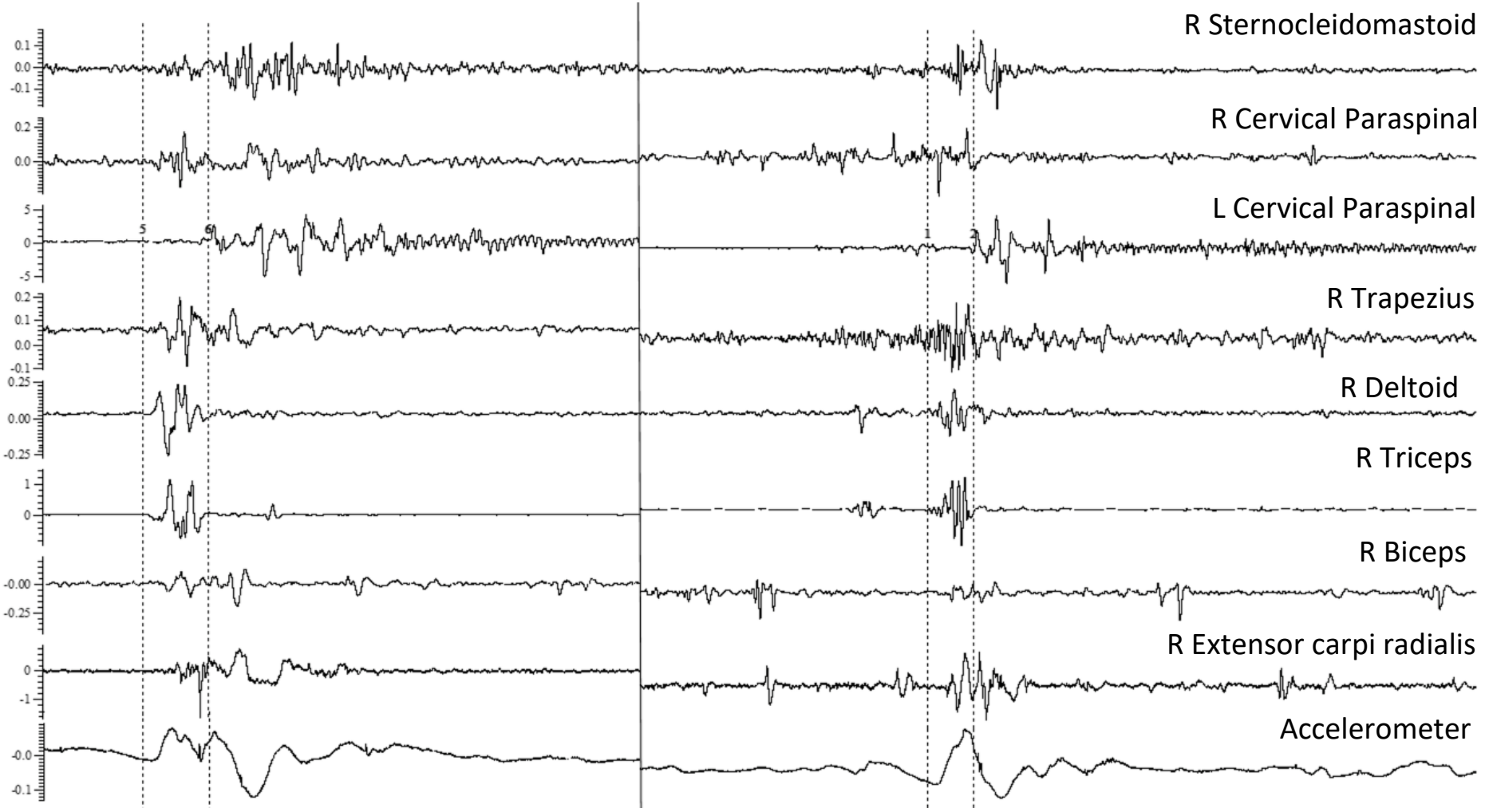
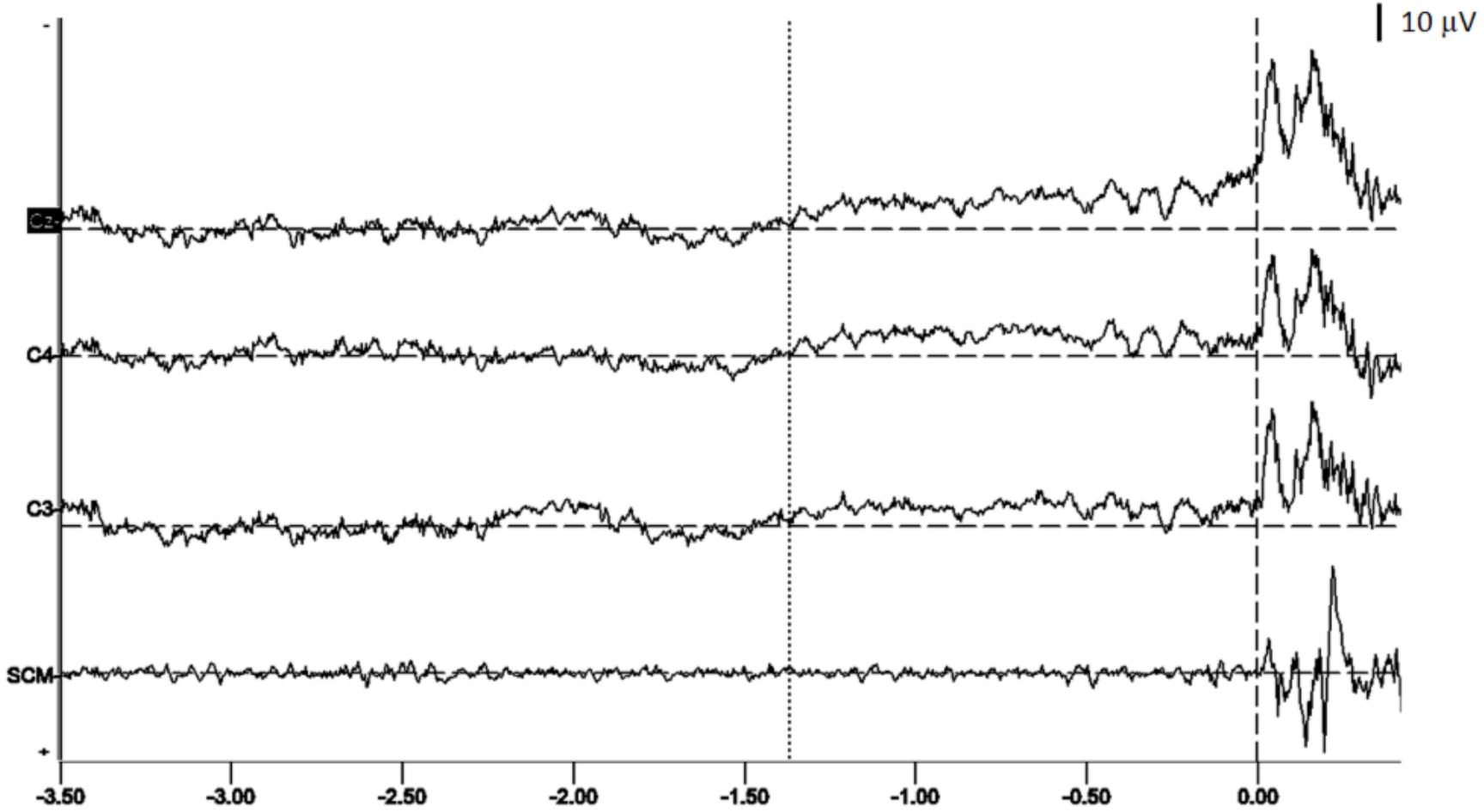
Neurophysiological Findings



150 ms burst duration, similar muscle spread pattern in every occasion

Neurophysiological Findings



Premotor potential short duration (1.4s)

Case 3 and 4

Courtesy of Dr. Saleh

Case 3

- 47 years old
- Stereotyped movements of blinking, head-turning, shoulder abduction, and elbow extension, mainly on the left side but also on the right side started gradually after car accident (4 years ago)
- No clear premonitory urge, presence of relief sensation after movement performance
- Volitional suppression of the movements
- **Phenomenology?**

Case 4

- 42 years old
- Involuntary jerky movements of the shoulder for 25 years
- No sense of agency
- No clear premonitory urge or relief sensation after movement performance
- Wife report movement during sleep
- **Phenomenology?**

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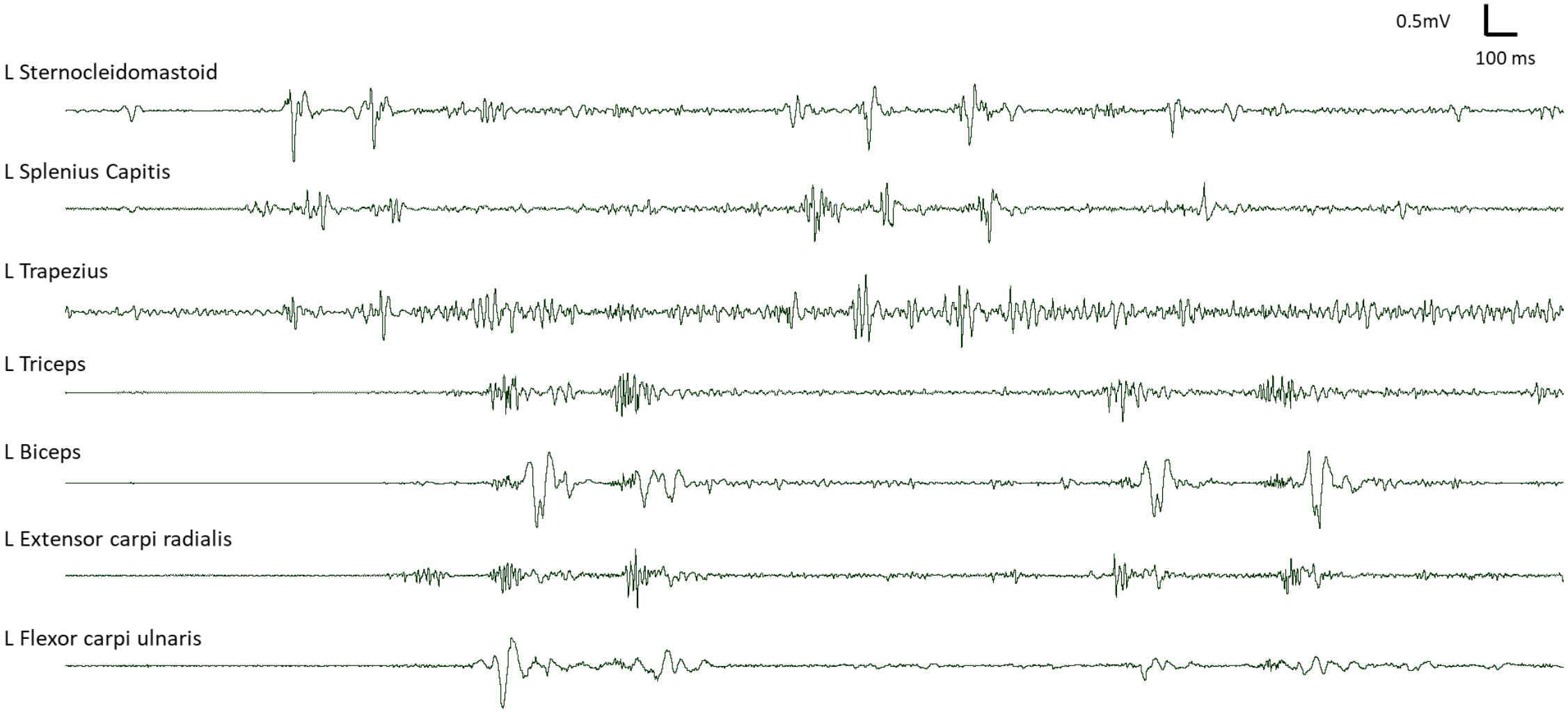
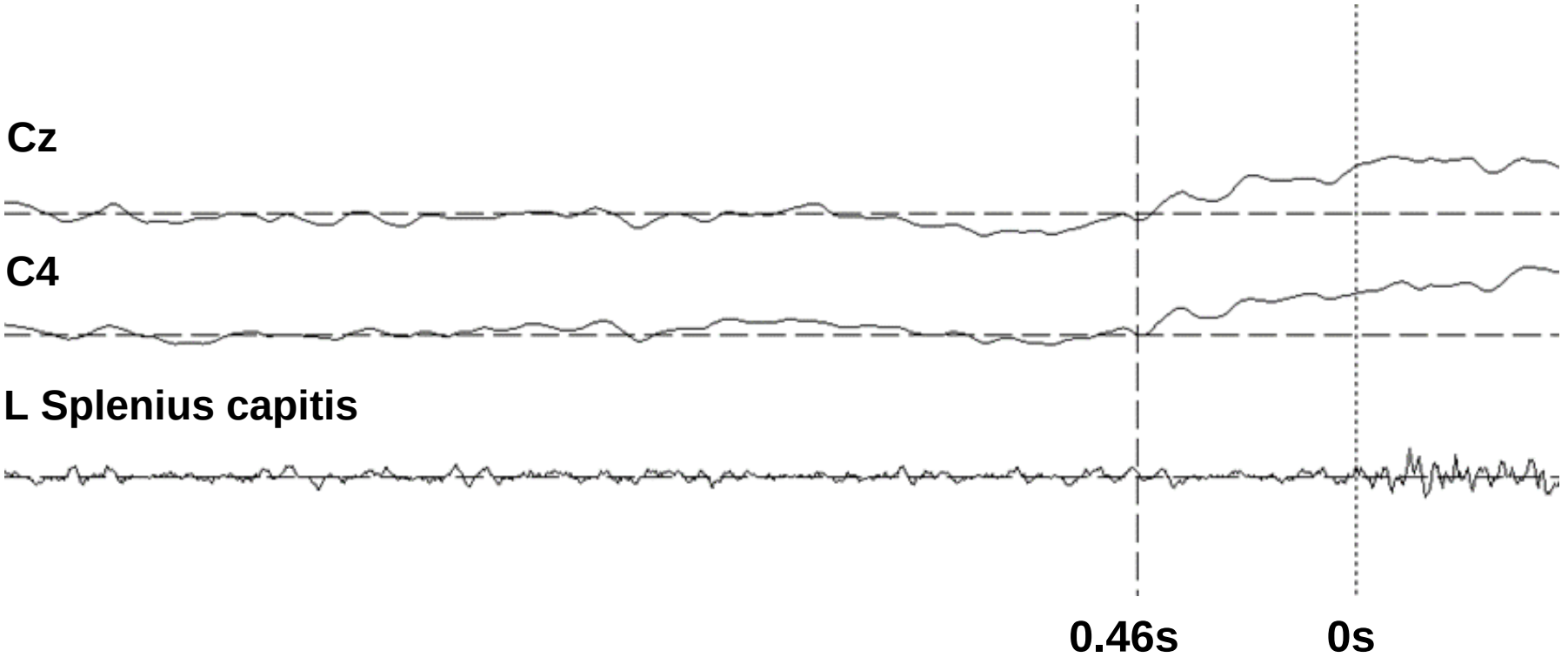
Neurophysiological Findings - Case 3



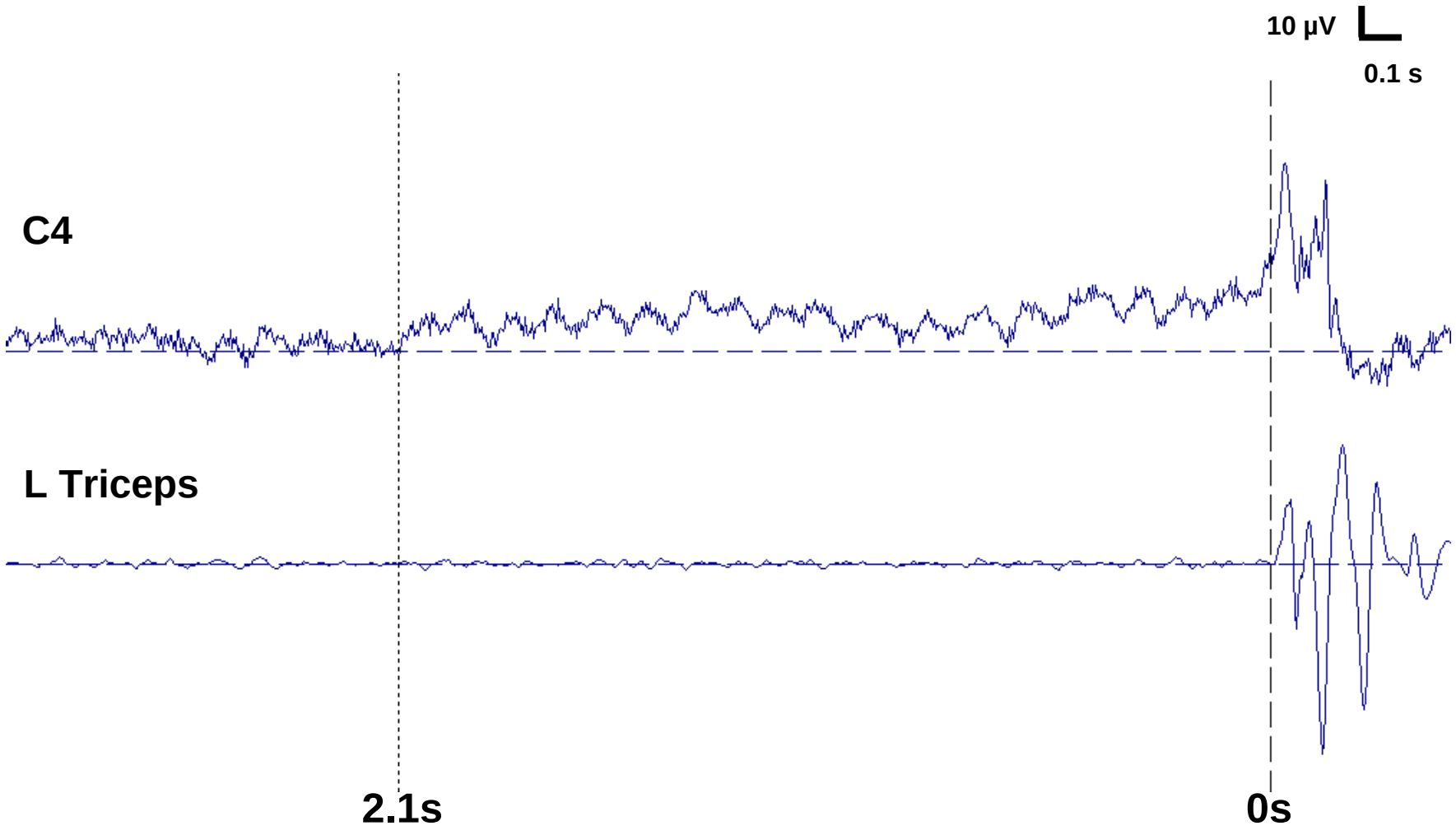
Stereotyped movement with repeated pattern and EMG burst duration of 130 - 250 ms

Neurophysiological Findings - Case 3



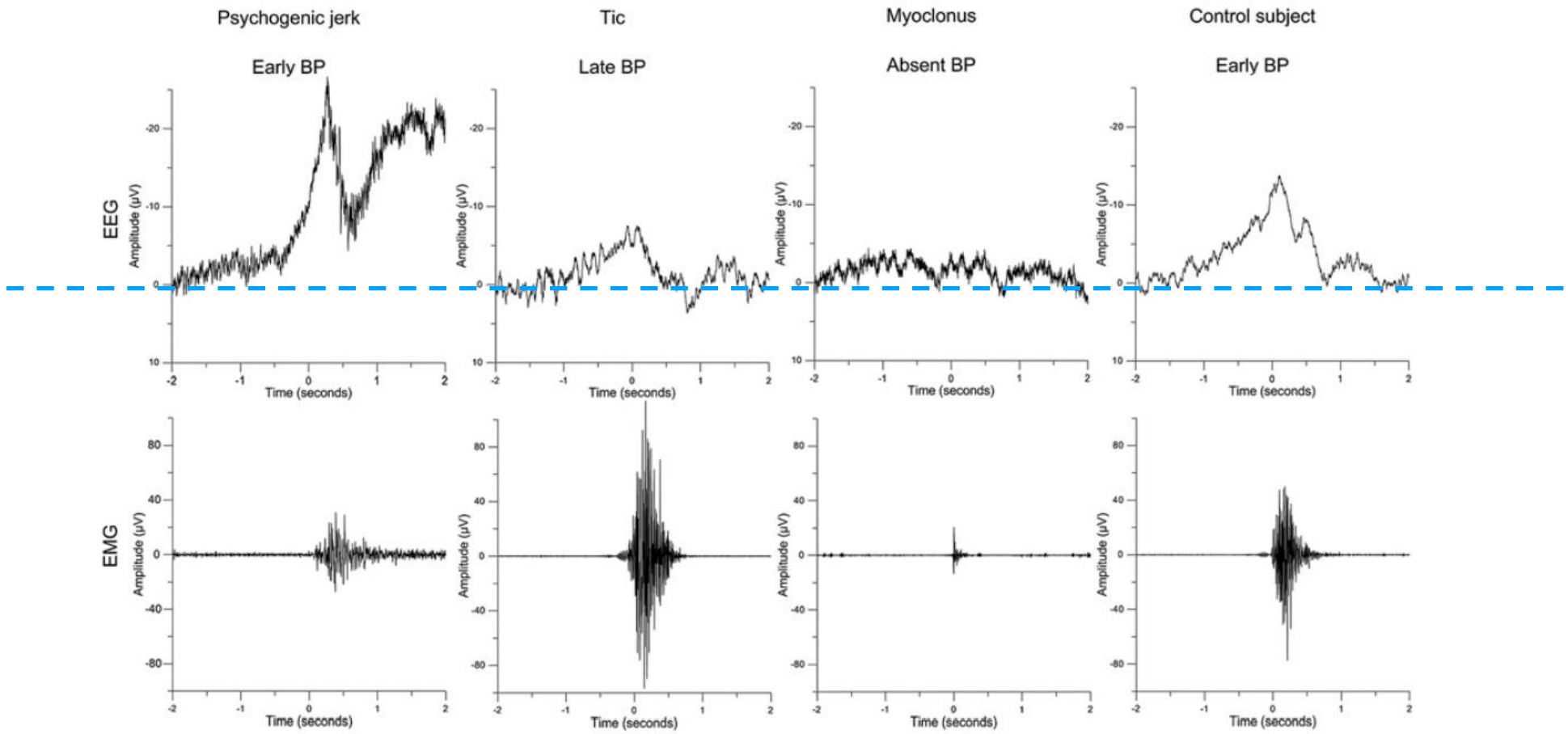
Premotor potential - Late component

Neurophysiological Findings - Case 4



Premotor potential

Neurophysiology of Tics



van der Salm SM et al. J Neurol Neurosurg Psychiatry. 2012

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Neurophysiology of Tics

Movement disorders

RESEARCH PAPER

The bereitschaftspotential in jerky movement disorders

Sandra M A van der Salm,[1] Marina A J Tijssen,[1,2] Johannes H T M Koelman,[1] Anne-Fleur van Rootselaar[1]

J Neurol Neurosurg Psychiatry 2012;**83**:1162–1167. doi:10.1136/jnnp-2012-303081

Clinical Neurophysiology Practice 1 (2016) 33–37

Contents lists available at ScienceDirect

Clinical Neurophysiology Practice

journal homepage: www.elsevier.com/locate/cnp

Case report

Clinical neurophysiological evaluation for simple motor tics

Pattamon Panyakaew[a,b], Hyun Joo Cho[a], Mark Hallett[a,*]

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[b] *Department of Medicine, Faculty of Medicine, Chulalongkorn Center of Excellence on Parkinson Disease and Related Disorders, Chulalongkorn University and King Chulalongkorn Memorial Hospital, Thai Red Cross Society, Bangkok 10330, Thailand*

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Physiological and introspective antecedents of tics and movements in adults with tic disorders

Antonio I. Triggiani[a,1], Kaya Scheman[a,1], Sarah Pirio Richardson[a,b], Masao Matsuhashi[a,c], Elizabeth Peckham[a,d], Fatta Nahab[a,e], Zoltan Mari[a,f], Shashi Ravindran[a], Mark Hallett[a,*]

OPEN ACCESS Freely available online

Tremor and Other Hyperkinetic Movements

New Observations Letter

Purposely Induced Tics: Electrophysiology

Patrick McGurrin[1], Sanaz Attaripour[1,2], Felipe Vial[1,3] & Mark Hallett[1*]

[1]Human Motor Control Section, NINDS, National Institutes of Health, Bethesda, MD, USA, [2]Department of Neurology, University of California, Irvine, Irvine, CA, USA, [3]Facultad de Medicina Clínica Alemana, Universidad del Desarrollo, Vitacura, Región Metropolitana, CL

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Case 5 and 6

Case 5

- 73 years old
- Intermittent twitching movements of bilateral toes asscoiated with internal “crawling sensation” and pain
  - Movements briefly suppressible
  - Wife reports possible movements during sleep
  - Normal nerve conduction studies
- **Phenomenology?**

Case 6

- 27 years old
- Involuntary movements of the toes started after ankle injury, associated with increased sweating, pain and discomfort in the feet
  - Unable to suppress the movement
  - Right> Left
  - Normal nerve conduction studies
- **Phenomenology?**

Functional etiology of PLMT

Table 1. Features for and against functional movement disorder in PLMTS

In Favor of OMD	In Favor of FMD
Movements not voluntarily reproducible	Previous reports of movements diminishing with voluntary activity of same limb
Lack of comorbid functional neurological symptoms	Overlap in some patients with complex regional pain syndrome (in which movement disorder is functional in nature8). Painful dystonia is a red flag for an FMD.
Atypical older age range	Later onset FMD in the absence of traditional risk factors increasingly recognized
Duration of bursts reported in the literature in some patients (as short as 50 ms) are outside the range of voluntary movement.3	Most reported durations (e.g., 0.5-2.0 seconds) and frequencies (2 Hz) are within the range of voluntary movement.2, 4

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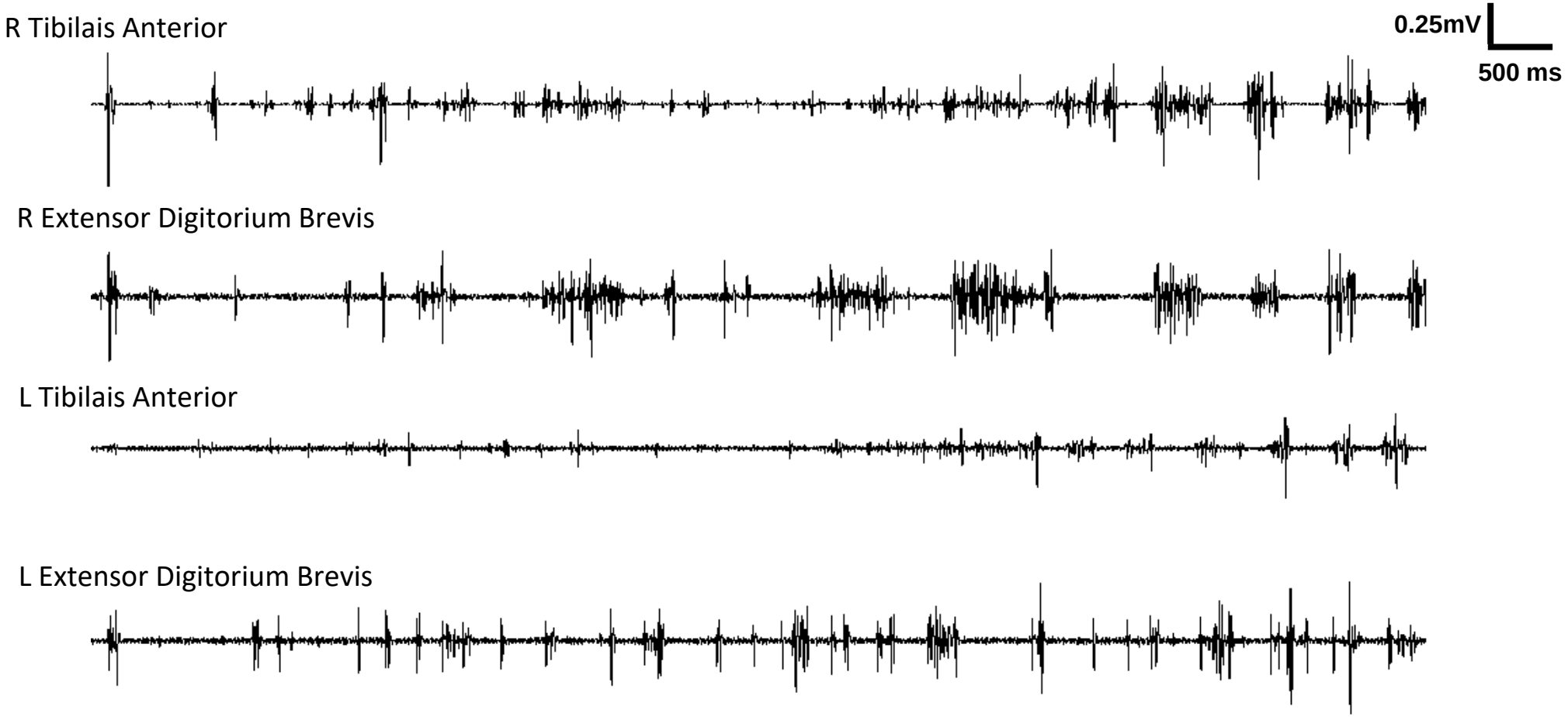
Vanegas-Arroyave N et al. Tremor Other Hyperkinet Mov (N Y). 2016

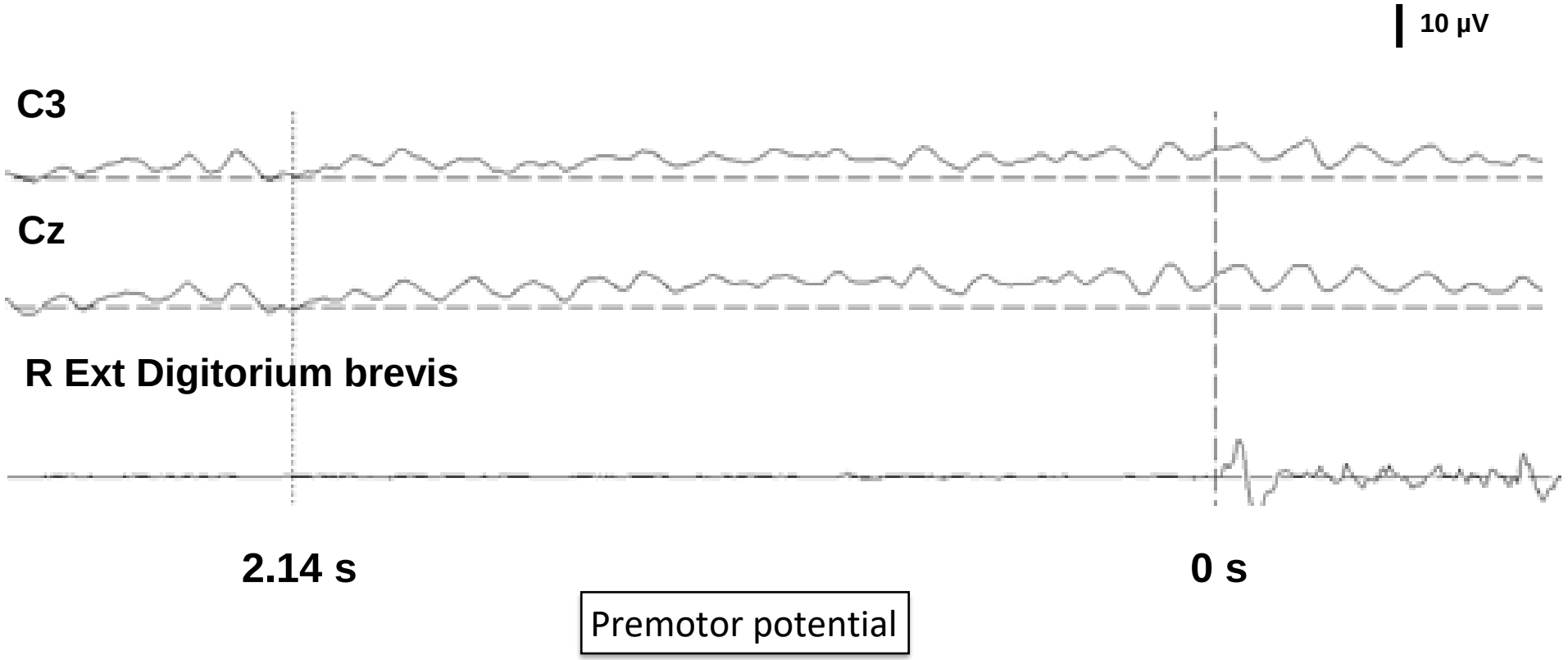
Stone J et al.Mov Disord. 2014.

Neurophysiological findings - Case 5



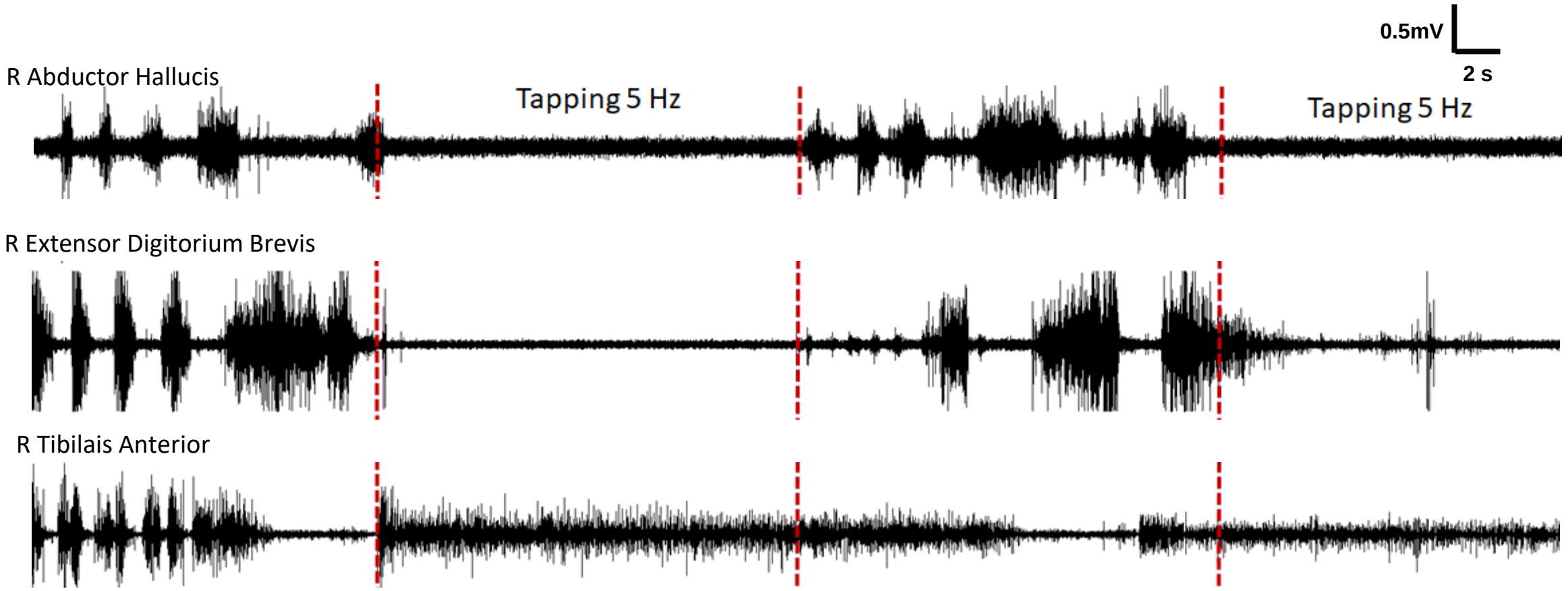
Semi-rhythmic contractions (~2Hz) of the both EDB and TA with EMG burst durations of 100 - 600ms

Neurophysiological findings - Case 5



Courtesy of Dr. Justus Chen

Neurophysiological findings - Case 6



Semi-rhythmic contractions (~2-4Hz) of the both EDB and TA with EMG burst durations of 150 - 500ms
Suppression while tapping with the left foot / Absence of Premovement potential

Courtesy of Dr. Justus Chen

Neurophysiology of PLMT

- Random, irregular EMG bursts lasting 50 to 1.000 ms in the distal limb muscles.
- The movements would at times group into semirhythmic or pseudorhythmic brief of approximately 0.5 to 1 Hz frequency (data from case series, no standardized values)

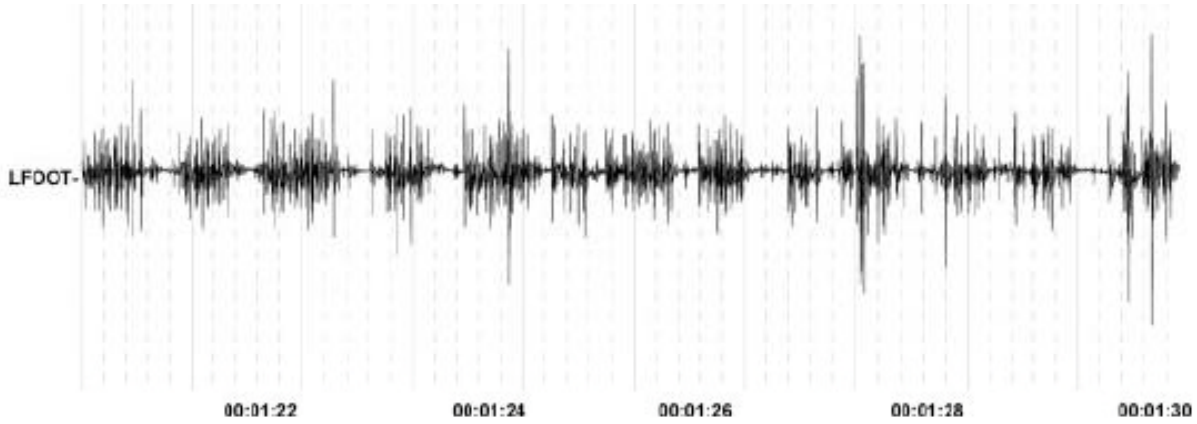


FIG. 1. Surface electromyography (EMG) in a 74-year-old man with painful legs and moving toes. The time between solid vertical lines is 1 second. The surface EMG shows semirhythmic EMG bursts lasting 500–1,200 milliseconds in the left extensor digitorum brevis.

Alvarez MV, et al. Mov Disord. 2008

Case 7

- 57 years old

Alvarez MV, et al. Mov Disord. 2008

Case 7

Alvarez MV, et al. Mov Disord. 2008

Case 7

- 18-month history of imbalance, dizziness, shock-like pain in the left part of her face, stiffness in both legs, and increased urinary frequency.
- Followed by the initiation of jerky movements of the upper and lower limbs that occur at rest but are exacerbated during action
- SCA 8 (allele 2 - >200 repetitions)
- **Phenomenology?**

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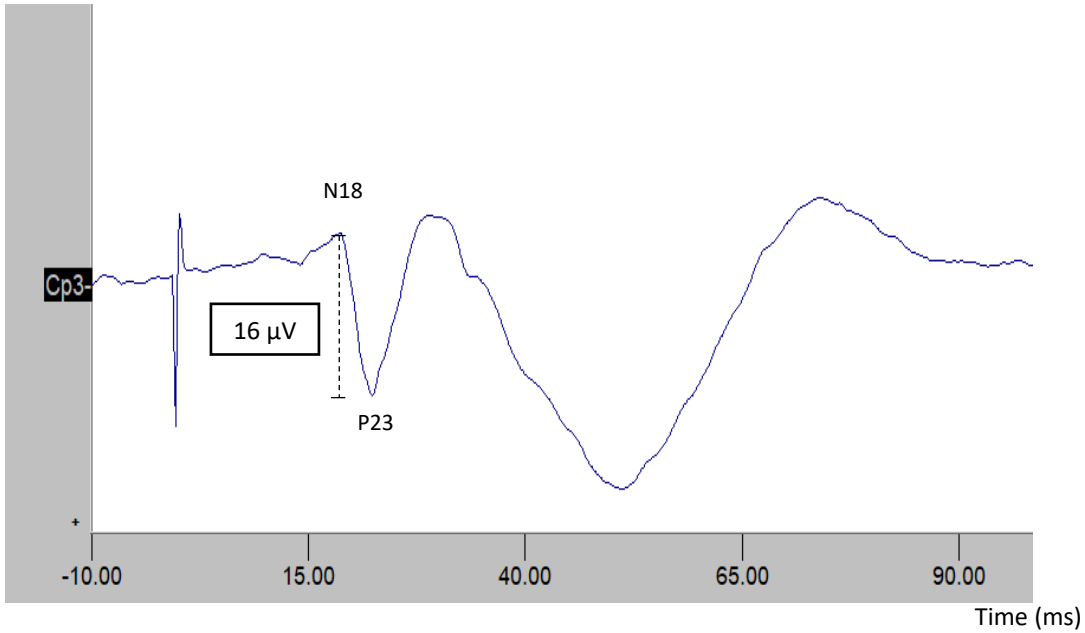
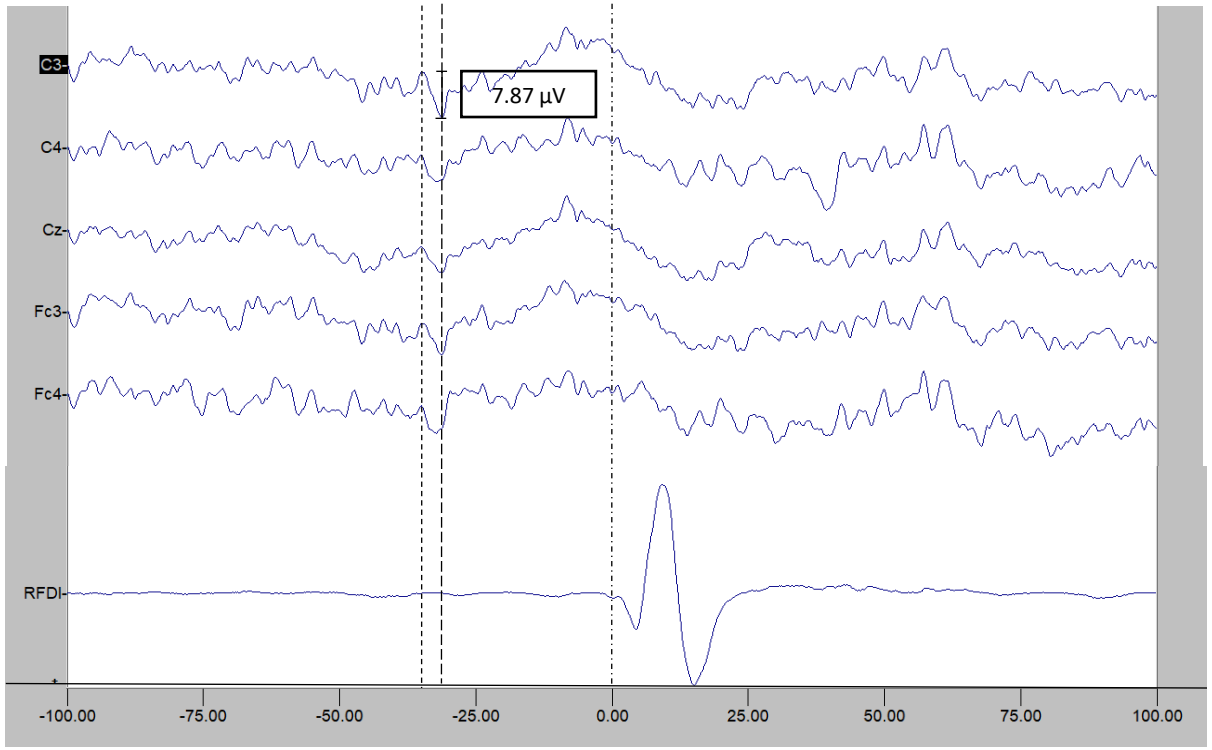
Neurophysiology - Case 7

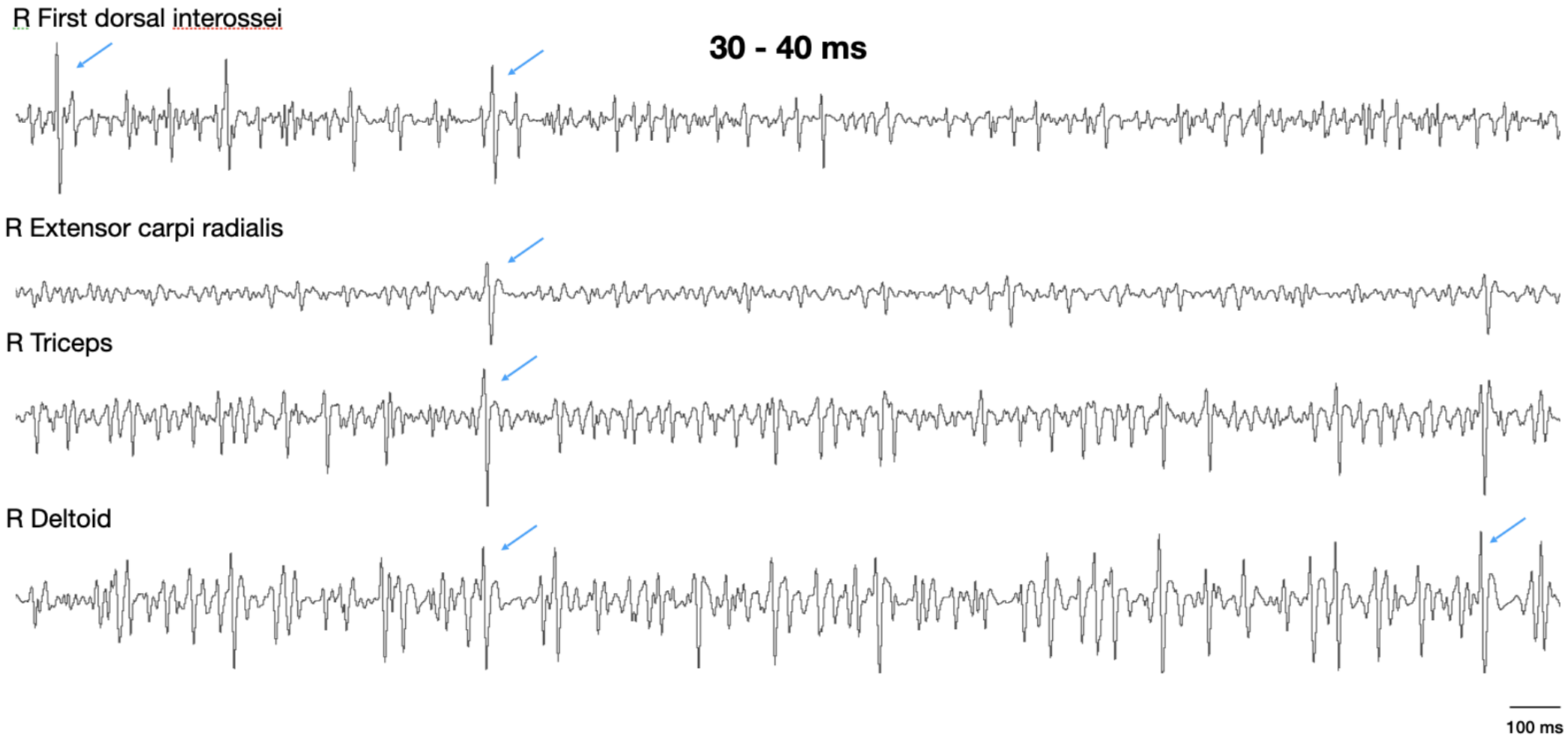


Giant SSEP

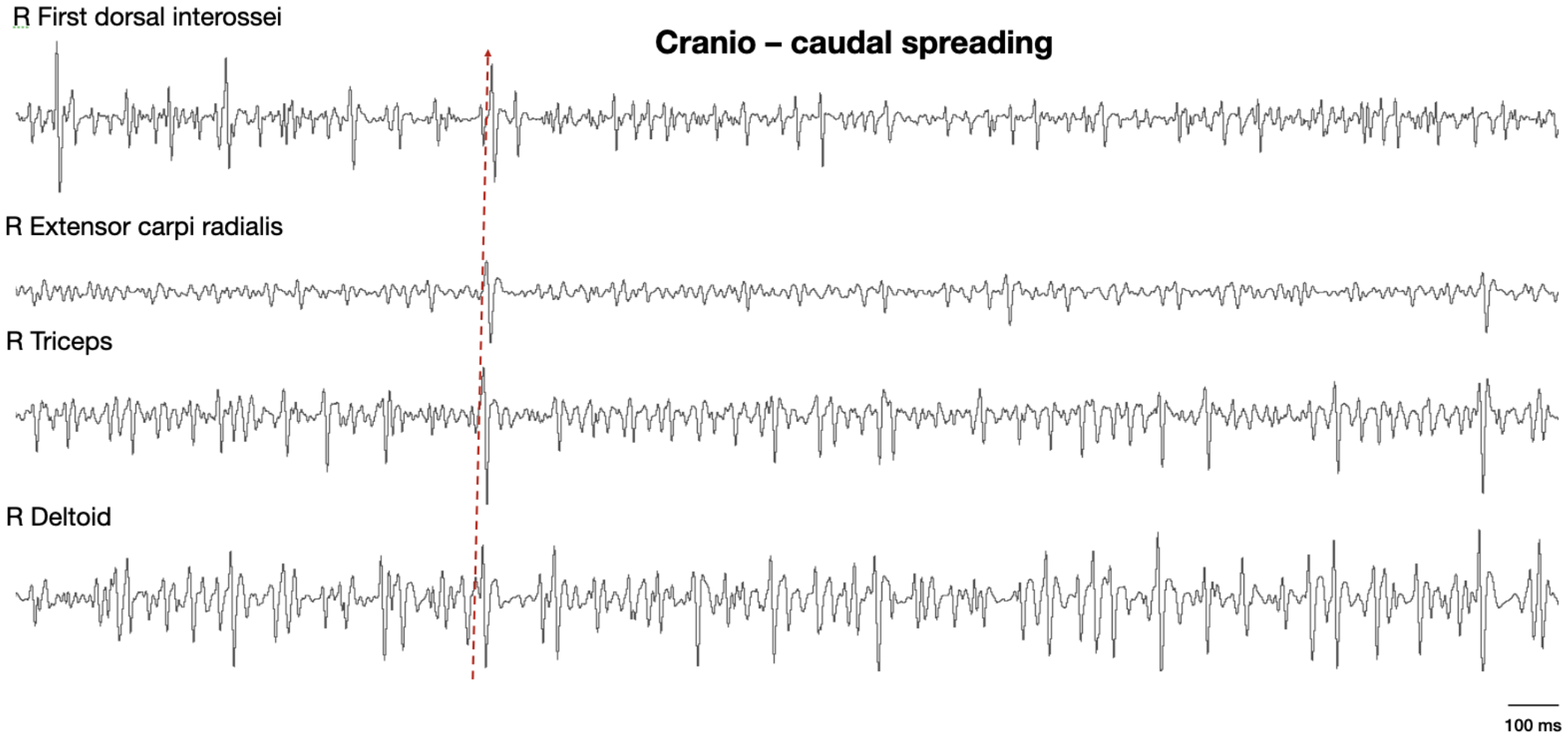


Focal EEG transient 31ms before the myoclonus

Neurophysiology - Case 7



Neurophysiology - Case 7

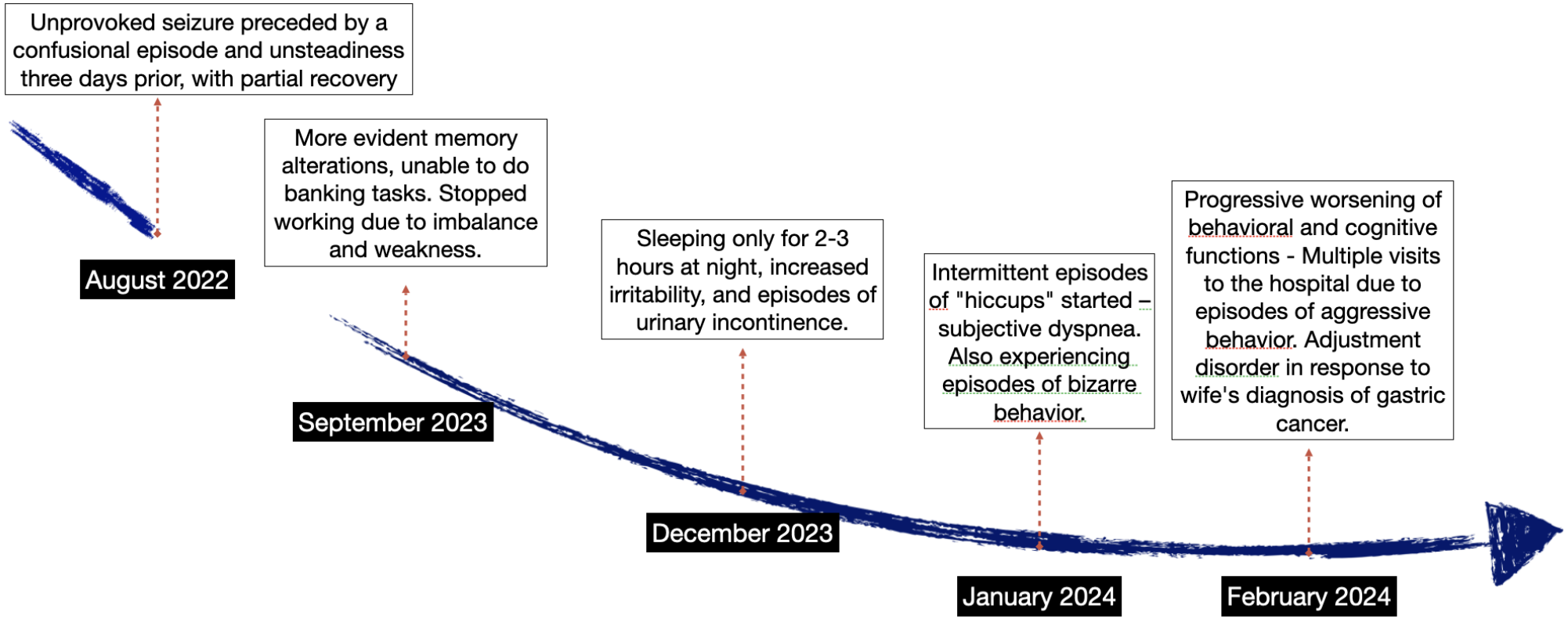


Case 8

- 60 years old

Alvarez MV, et al. Mov Disord. 2008

Case 8


Unprovoked seizure preceded by a confusional episode and unsteadiness three days prior, with partial recovery
August 2022
More evident memory alterations, unable to do banking tasks. Stopped working due to imbalance and weakness.
September 2023
Sleeping only for 2-3 hours at night, increased irritability, and episodes of urinary incontinence.
December 2023
Intermittent episodes of "hiccups" started – subjective dyspnea. Also experiencing episodes of bizarre behavior.
January 2024
Progressive worsening of behavioral and cognitive functions - Multiple visits to the hospital due to episodes of aggressive behavior. Adjustment disorder in response to wife's diagnosis of gastric cancer.
February 2024

Case 8

PAST MEDICAL HISTORY

- Type 2 diabetes
- Depression/Anxiety using sertraline since 2014 (history of previous aggressive behavior in the past/suicidal ideation)
- Hepatitis C (incomplete treatment)
- Alcohol abuse (reduced to 3-4 drinks in 2005 – stopped 2023)
- Smoker - 2 packs/day for the past 40 years
- Hypertension, Dyslipidemia

Case 8 - Movement features

- Semirhythmic, slow, continuous during sleep, involving abdominal muscles and hip adductor muscles;
- Not triggered by sound or sensory stimulation;
- Not altered with respiratory cycles;
- Persistent during sleep.

Case 8 - Movement features

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- Not triggered by sound or sensory stimulation;
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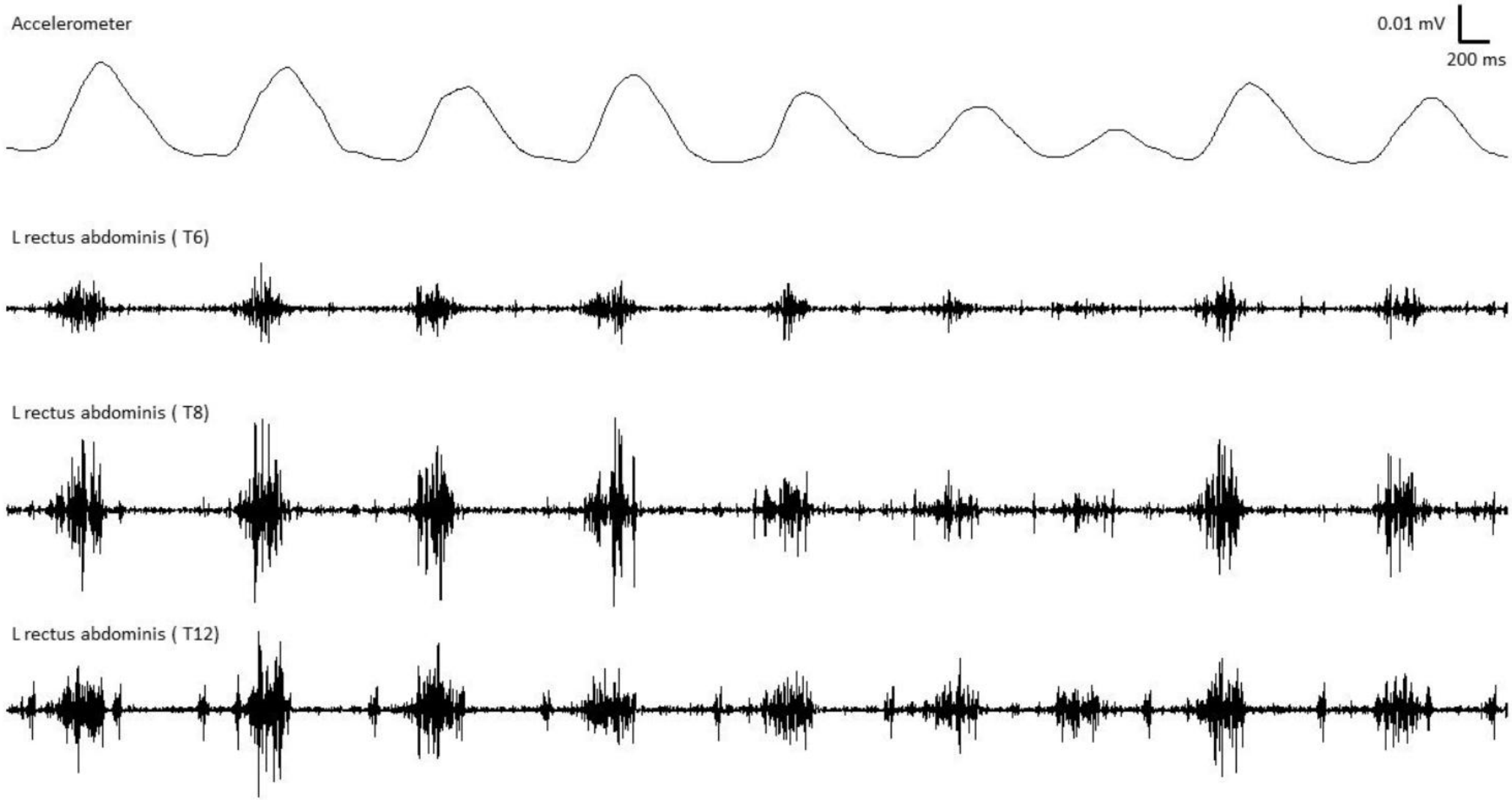

Synchronous contractions of the rectus abdominis muscles with a mean EMG burst duration of 350ms and mean frequency of 1Hz at rest while the patient was lying down.

MYORHYTMIA

After use of lorazepam 1mg

Case 8

CSF analysis: RBC 20,000/ WBC 12, 64% neutrophils, 27% lymphocytes / Protein **2.17** (ref 0.45)/ Glucose **6.8** (ref: 4.4)/ lactate 1.7

- Meningoencephalitis panel negative

EEG: Mild intermittent non-epileptiform disturbance of cerebral activity was recorded over the left frontotemporal region. **No definitive epileptiform abnormality was recorded.**

Spinal and Brain MRI: No significant alteration

Case 8 - Movement features

RAPIDILY PROGRESSIVE COGNITIVE SYMPTOMS + PSYCHIATRIC SYMPTOMS + SLEEP DISORDER + CEREBELLAR SYMPTOMS + ABDOMINAL SEGMENTAL SPINAL MYOCLONUS

DIAGNOSIS: ANTI-CASPR2 ENCEPHALITIS

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CASPR2 proposed phenotypes

Paroxysmal ataxia	Segmental myoclonus	Paroxysmal orthostatic myoclonus
Limbic encephalitis	Morvan syndrome	Isolated

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Thank you!