

# Neurophysiology: An Extension of Clinical Assessment to Categorize Movement Disorders

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# Disclosures

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# Outline

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- Introduction – Why Neurophysiological Studies in Movement Disorders
- Extension of clinical assessment (history/examination)
- Diagnostic utility
  - Functional tremor
  - Bereitschaftspotential
  - In jerky movement disorders
  - In movement disorders practice

# Why perform neurophysiological studies in movement disorders patients?

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- Provide or confirm a clinical diagnosis
- Differentiate organic from functional movement disorders – provide objective evidence
- Assess the effect of medical or surgical treatment
- Better understand underlying pathophysiology for research purpose

# Disorders that could be investigated

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- Tremor
  - Functional (Psychogenic)
  - Orthostatic
  - PD / ET / enhanced physiological tremor
  - Dystonic
- Jerks
  - Myoclonus
  - Tics
  - Functional (Psychogenic)
- Dystonia and spasms

# Equipment needed

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## Basic

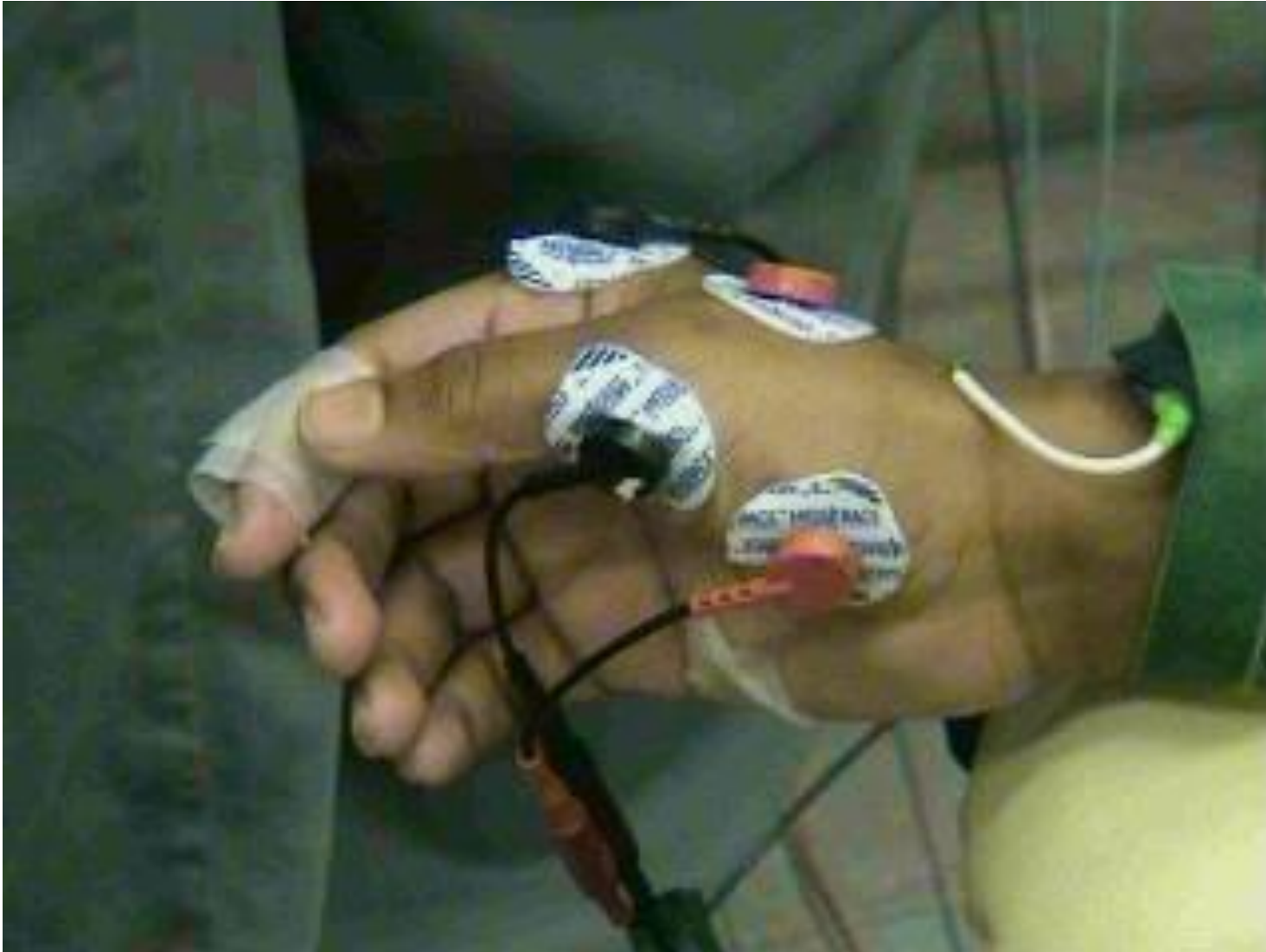
- EMG (surface), EEG
- Accelerometer (or goniometer, gyroscopes, inertial measurement device IMU)
- Simple software to store and analyze data (place cursors for latencies, amplitudes, averaging)

## More advanced

- EEG backaveraging (epoching and averaging)
- Frequency (e.g FFT), coherence, phase analysis
- Motion tracking device (infra-red or magnetic field based)
- Transcranial magnetic stimulation

# Polygraphic EMG and accelerometry

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# Neurophysiology: Extension of Clinical Assessment

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- Traditional Clinical Neurophysiology methods such as electroencephalography (EEG), electromyography (EMG), evoked potentials are all considered extension of clinical assessment
- Movement Disorder Neurophysiology, combined with clinical history/examination, is more recently recognized as playing a role in establishing the diagnosis

# Introduction of “Laboratory supported” Definite Functional Movement Disorder

| Traditional   | Proposed revision   |
|---|---|
| Classification of degrees of certainty in diagnosis <sup>a</sup><br>1. Documented <sup>b</sup><br>Remittance with suggestion, physiotherapy, psychotherapy, placebos, ‘while unobserved’<br>2. Clinically established <sup>b</sup><br>Inconsistent over time/incongruent with clinical condition + other manifestations: other ‘false’ signs, multiple somatizations, obvious psychiatric disturbance<br>3. Probable<br>Inconsistent/incongruent – no other features<br>Consistent/congruent + ‘false’ neurological signs <sup>c</sup><br>Consistent/congruent + multiple somatizations <sup>c</sup><br>4. Possible <sup>d</sup><br>Consistent/congruent + obvious emotional disturbance <sup>c</sup> | Classification of degrees of certainty in diagnosis<br>1. Documented (as in original)<br>2a. Clinically established plus other features (as in original)<br>2b. Clinically established minus other features<br>Unequivocal clinical features incompatible with organic disease with no features suggesting another underlying neurological or psychiatric problem<br><br>1 + 2a + 2b = Clinically Definite<br><br><b>3. Laboratory-supported definite</b><br>Electrophysiological evidence proving a psychogenic movement disorder (primarily in cases of psychogenic tremor and psychogenic myoclonus) |

<sup>a</sup> Adapted from [12].

<sup>b</sup> Subsequently, Fahn and his coauthors [10] proposed combining categories 1 + 2 under ‘Clinically Definite’.

<sup>c</sup> We proposed to reclassify these patients under ‘Possible’.

<sup>d</sup> We also questioned the utility of retaining the ‘Possible’ category as this generally represents patients with organic movement disorders with additional psychiatric problems rather than a true ‘Possible psychogenic movement disorder’ [11].

# IAPRD new consensus classification of myoclonus

Anna Latorre<sup>a,1</sup>, S. van der Veen<sup>b,1</sup>, Ashley Pena<sup>c,1</sup>, Daniel Truong<sup>d,1</sup>, Roberto Erro<sup>e,1</sup>,  
Steven Frucht<sup>f,1</sup>, Christos Ganos<sup>g,1</sup>, Mark Hallett<sup>h,1</sup>, Belen Perez-Duenas<sup>i,1</sup>, Malco Rossi<sup>j,1</sup>,  
Emmanuel Roze<sup>k,1</sup>, Marie Vidailhet<sup>l,m,1</sup>, Marina AJ. de Koning-Tijssen<sup>n,1</sup>, John N. Caviness<sup>o,1,\*</sup>

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- Axis 1a: Clinical features
- Axis 1b: Clinical Neurophysiology Features
- Axis II. Etiology

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# Functional Neurological Disorders

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- Functional Neurological Disorder – Neurological symptoms that cannot be explained by other neurological or medical conditions
- Functional Movement Disorder - Previously termed psychogenic movement disorders
- Movement considered involuntary by the patient, but utilize voluntary motor pathways

# Functional tremor - Physiology

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- Distraction
- Entrainment
- Distraction with ballistic movements
- Co-activation sign
- Increase in tremor amplitude (or frequency) with loading of limb
- Variability in tremor frequency – spontaneous or with task performance
- Side-to-side coherence
- Suggestibility

# 10 Item Test Battery

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Tapping performance at 1 Hz

Tapping performance at 3 Hz

Tapping performance at 5 Hz

Tapping response at 1 Hz

Tapping response at 3 Hz

Tapping response at 5 Hz

Ballistic movement response

Tonic coactivation

Coherence test

Loading test

## Validation of “Laboratory-Supported” Criteria for Functional (Psychogenic) Tremor

Petra Schwingenschuh, MD,<sup>1,2\*</sup> Tabish A. Saifee, MRCP,<sup>3</sup> Petra Katschnig-Winter, MD, MSc,<sup>1</sup> Antonella Macerollo, MD,<sup>3</sup> Mariella Koegl-Wallner, MD,<sup>1</sup> Valeriu Culea, MD,<sup>1</sup> Christine Ghadery, MD, PhD,<sup>1</sup> Edith Hofer, PhD,<sup>1,4</sup> Tamara Pendl, MD,<sup>1</sup> Stephan Seiler, MD,<sup>1</sup> Ulrike Werner, PhD,<sup>1</sup> Sebastian Franthal, MD,<sup>1</sup> Natasha M. Maurits, PhD,<sup>5</sup> Marina A. Tijssen, MD, PhD,<sup>5</sup> Reinhold Schmidt, MD,<sup>1</sup> John C. Rothwell, PhD,<sup>3</sup> Kailash P. Bhatia, MD, FRCP,<sup>3</sup> and Mark J. Edwards, FRCP, PhD<sup>3,6</sup>

- 38 functional and 38 organic tremor patients
- Good interrater and test-retest reliability
- Composite score of 3 (out of 10) points has sensitivity of 89.5% and specificity of 95.9% for diagnosis of Functional Tremor

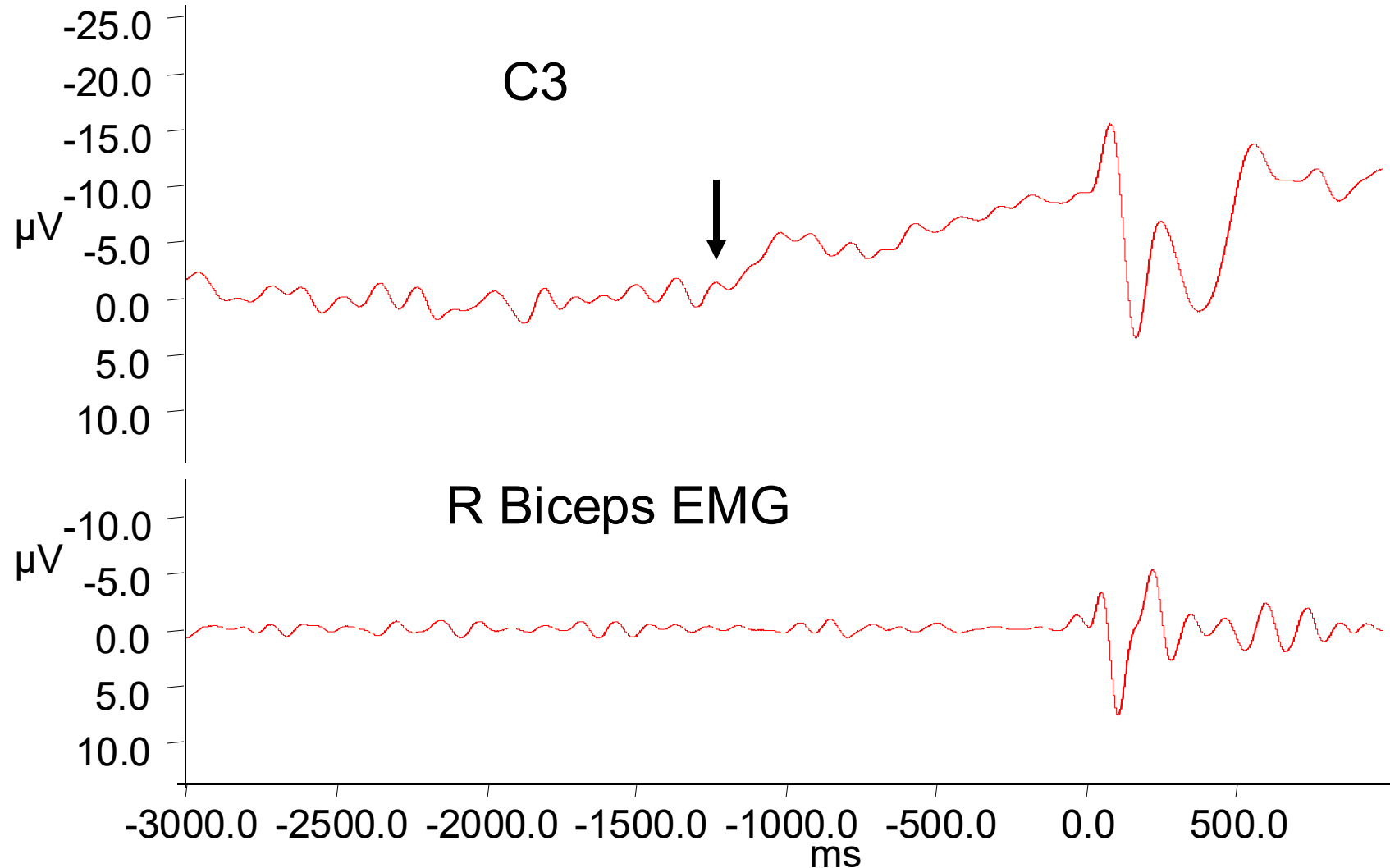


# Functional tremor - Physiology

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- Very useful in objectively documenting functional (psychogenic) features
- Individual findings not always present
- A battery of tests should be performed
- Beware of co-existence of functional and organic tremor

# Movement muscle jerks – cortical premovement potentials (BP)



# The Bereitschaftspotential in jerky movement disorders

Sandra M A van der Salm,<sup>1</sup> Marina A J Tijssen,<sup>1,2</sup> Johannes H T M Koelman,<sup>1</sup>  
Anne-Fleur van Rootselaar<sup>1</sup>

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## Presence of BP

- Psychogenic (functional) 86% (25/29)
- Tourette 43% (6/14)(with shorter duration)
- Myclonus 0% (0/5)
- Controls 100% (25/25)(with voluntary movement)

van der Salm et al, J Neurol Neurosurg Psychiatry 2012

# Electrophysiologic testing aids diagnosis and subtyping of myoclonus

Rodi Zutt, MD, Jan W. Elting, MD, PhD, Jonathan C. van Zijl, MD, J. Han van der Hoeven, MD, PhD, Christiaan M. Roosendaal, MD, Jeannette M. Gelauff, MD, Kathryn J. Peall, MD, PhD, and Marina A.J. Tijssen, MD, PhD

## Correspondence

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- 72 patients with myoclonus
- 66 underwent electrophysiological testing (6 too subtle to test)
- Polymyography, EEG-EMG backaveraging, coherence, SEP
- Change in initial diagnosis of myoclonus or subtype: 53%

Zutt et al. *Neurology* 2018





# Diagnostic Utility of Clinical Neurophysiology in Jerky Movement Disorders: A Review from the MDS Clinical Neurophysiology Study Group

Anna Latorre, MD, PhD,<sup>1,\*</sup> Christos Ganos, MD,<sup>2</sup> Masashi Hamada, MD, PhD,<sup>3</sup> Nicolas Phielipp, MD,<sup>4</sup> Lorenzo Rocchi, MD, PhD,<sup>5</sup> Shabbir Merchant, MD,<sup>6</sup> Marina A. Tijssen, MD, PhD,<sup>7,8</sup> Sterre van der Veen, MD, PhD,<sup>7,8</sup> and Robert Chen, MBBChir, MSc, FRCP<sup>9,10</sup>

**TABLE 1** Clinical neurophysiological studies in cortical myoclonus

| Technique                               | Subjects studied over a total sample of 227 (%) | Subjects studied showing positive findings (n) and percentage (%) |
|---|---|---|
| Jerk locked back-averaging              | 138 (60%)                                       | 70 (50%)  |
| SEP                                     | 144 (63%)                                       | 67 (46%)  |
| C-reflex                                | 68 (29%)  | 49 (72%)  |
| Cortico-muscular coherence <sup>a</sup> | 49 (21%)  | 19 (38%)  |

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- Neurophysiology can be used to identify myoclonus and its subtypes
- Single test not sensitive enough – a battery of tests needed
- Standardized protocols needed

# Clinical Utility of Neurophysiologic Classification (and Declassification) of Myoclonus

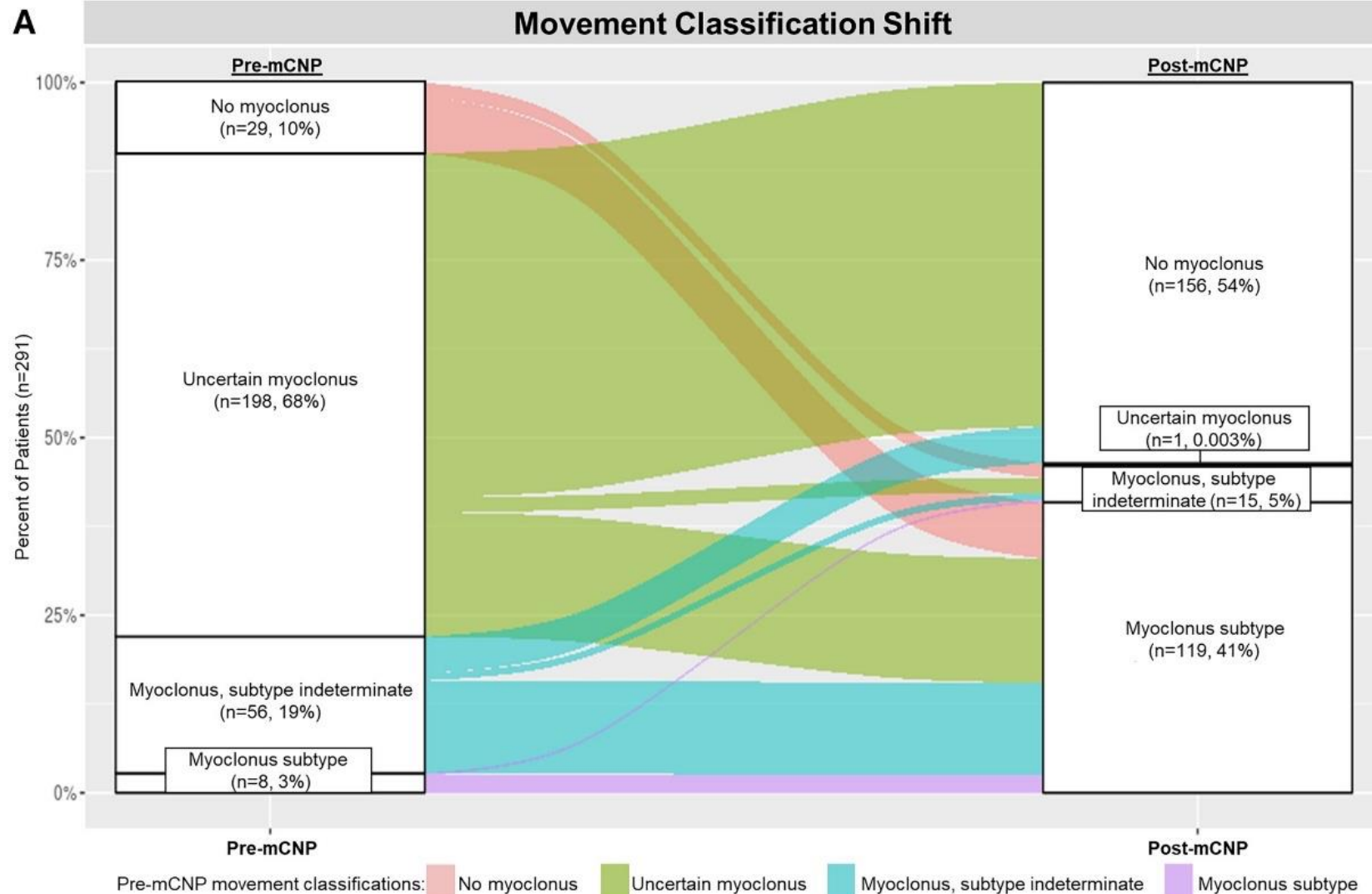
Marcus N. Callister, MD,<sup>1,2\*</sup> Molly C. Klanderman, PhD, MS,<sup>3</sup> Alyssa Stockard, BS,<sup>4</sup> Charles Van Der Walt, BS,<sup>3</sup>  
Ashley B. Pena, MD,<sup>1,5</sup> and John N. Caviness, MD<sup>1</sup>

- 206 patients referred for possible myoclonus
- 105 (40%) had myoclonus, 156 (59%) had no myoclonus
- Commonest alternative classification: functional jerks and tremor, and 1 was uncertain
- 29 studies identified myoclonus without clinical suspicion
- 119/113 (89%) specific neurophysiologic subtype identified, most common cortical (64, 54%)
- Medication change in 151 (52%), improvement 35/51 (67%)

Callister et al, Mov Disord, 2024

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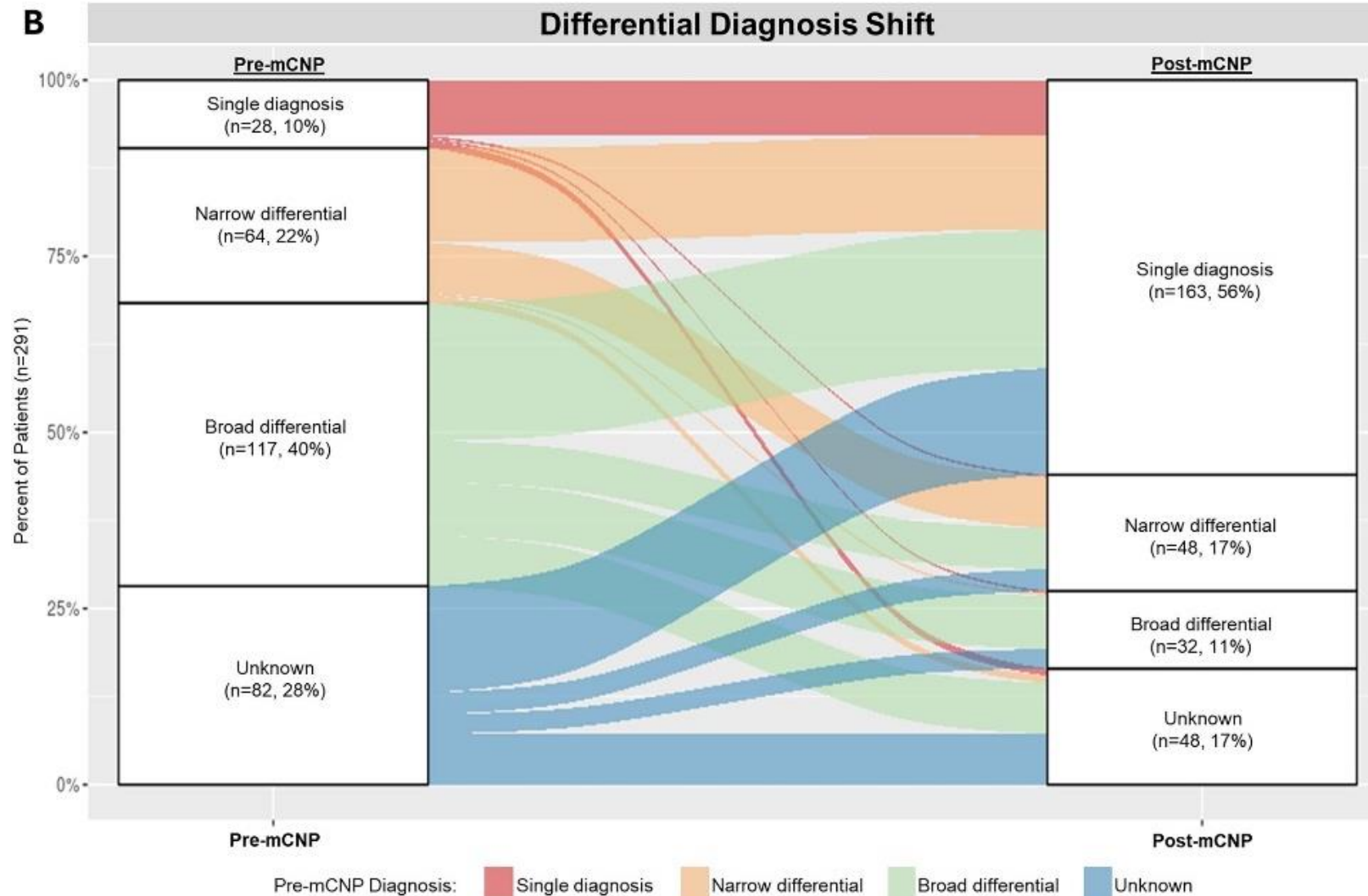


Callister et al, Mov Disord, 2024



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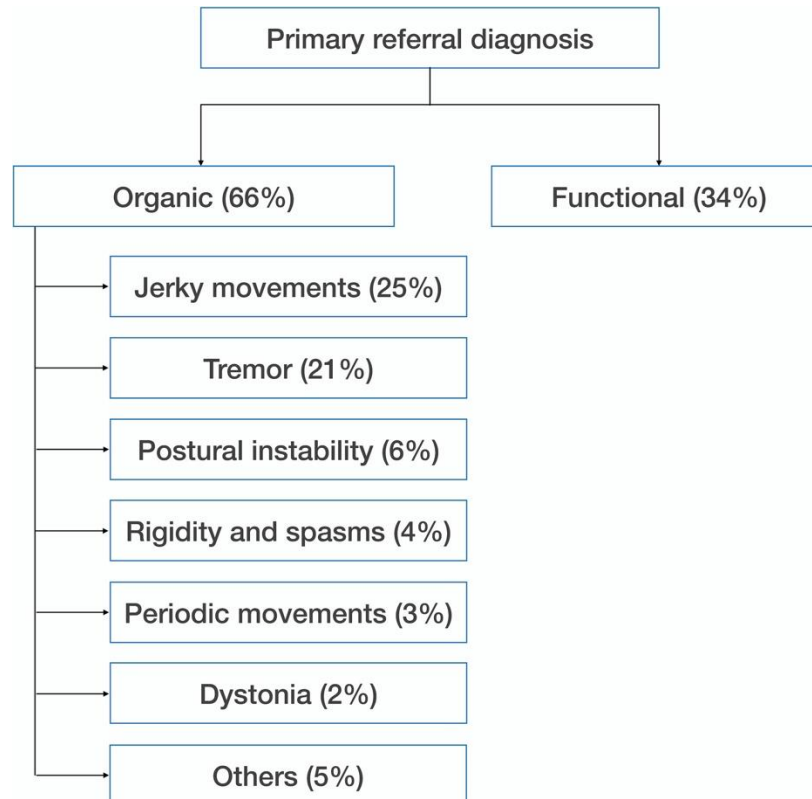


Callister et al, Mov Disord, 2024

# Utility of Neurophysiological Evaluation in Movement Disorders Clinical Practice

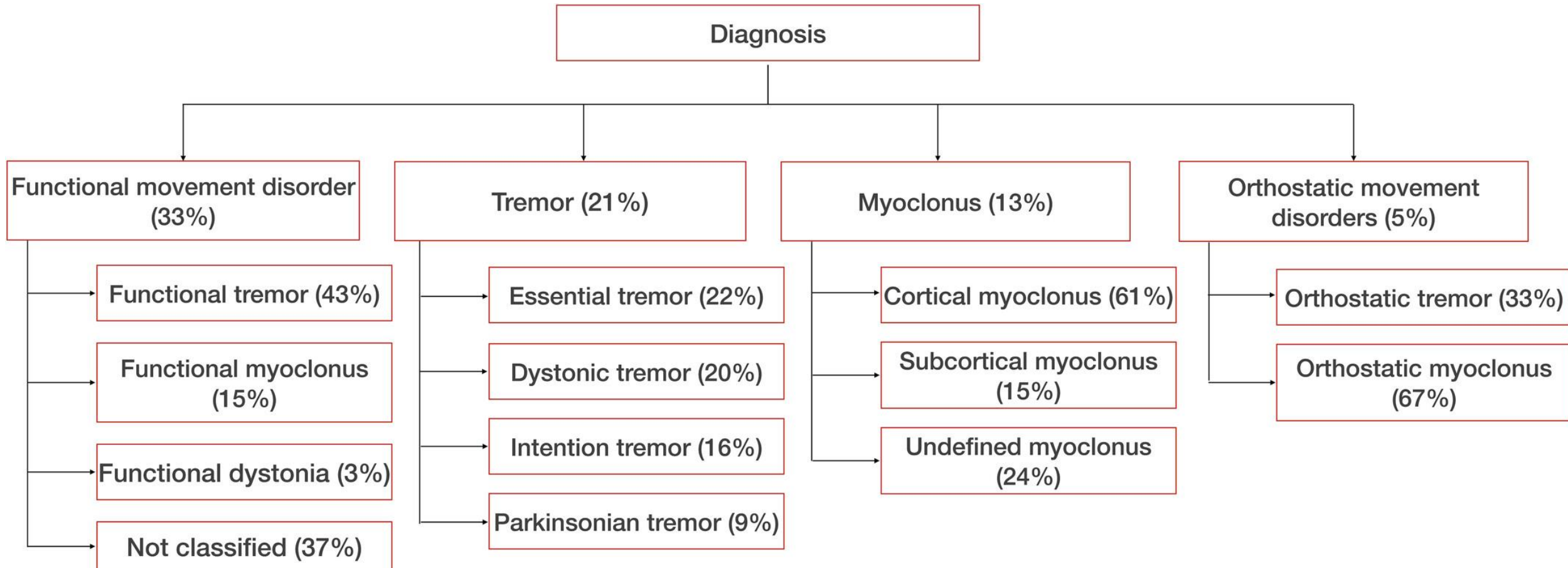
2023

Talyta Grippe, MD, MSc<sup>1,2,3</sup> and Robert Chen, MBBChir, MSc, FRCPC<sup>1,3,4,5,\*</sup>



- 509 studies over 18 years

# Utility of Neurophysiological Evaluation



# Utility of Neurophysiological Evaluation

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Change in diagnoses based on referral diagnoses

- Functional movement disorders: 13%
- Jerky movements: Other than myoclonus 27%
- Tremor: Not confirmed in 20%

Neurophysiological diagnoses not initially suspected

- Functional movement disorders: 30%
- Tremor: 17%
- Myoclonus: 13%

Has utility in patients evaluated by movement disorders specialist

Grippe & Chen, Mov Disord Clin Pract. 2023

# Summary

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- Clinical electrophysiology: Extension of Clinical Assessments
- Clinical utility in assessment of
  - Functional movement disorder
  - Tremor
  - Muscle jerk/myoclonus
  - Orthostatic movement disorders
- Establishment of standardized protocols will be useful