Movement Disorders
CLINICAL PRACTICE

RESEARCH ARTICLE

Utility of Neurophysiological Evaluation in Movement Disorders Clinical Practice

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Abstract: Background: Quantitative and objective neurophysiological assessment can help to define the predominant phenomenology and provide diagnoses that have prognostic and therapeutic implications for movement disorders.

Objectives: Evaluate the agreement between initial indications and final diagnoses after neurophysiological evaluations in a specialized movement disorders center.

Methods: Electrophysiological studies conducted for movement disorders from 2003 to 2021 were reviewed. The indications were classified according to predominant phenomenology and the diagnoses categorized in subgroups of phenomenology.

Results: A total of 509 studies were analyzed. 51% (259) of patients were female, with a mean age of 51 years (ranges 5 to 89 years). The most common reasons for referral were evaluation of functional movement disorders (FMD), followed by jerky movements, tremor and postural instability. Regarding FMD referrals, there was a diagnostic change in 13% of the patients after electrophysiological assessment. The patients with jerky movements as indication had a diagnosis other than myoclonus in 27% of them, and tremor was not confirmed in 20% of the cases. In patients with an electrophysiological diagnosis of FMD, it was not suspected in 30% of the referrals. Similarly, tremor was not mentioned in the referral of 17% of the patients with this electrophysiological diagnosis and myoclonus was not suspected in 13% of the cases.

Conclusions: Electrophysiological assessment has utility in the evaluation of movement disorders, even in patients evaluated by movement disorders neurologists. More studies are needed to standardize the protocols between centers and to promote the availability and use of these techniques among movement disorders clinics.

The use of neurophysiological techniques to evaluate movement disorders is well established for myoclonus, stiff-person syndrome and orthostatic tremor (OT).[1–3] There is growing interest in the field, and methods are being developed to evaluate tremor,[4] especially for the diagnosis of functional tremor,[5] and other movements such as dystonia and tics.[6–8] Quantitative and objective neurophysiological assessment can help to define the predominant phenomenology and provide diagnoses that have prognostic and therapeutic implications.[3,9,10] There are few neurophysiological units dedicated to movement disorders studies around the world, which limits the routine use of these assessments. To provide an overview of how a dedicated movement disorder neurophysiology unit impacted clinical practice, we reported data from our unit collected over 18 years. We explored how the neurophysiological testing contributed to the diagnosis of movement disorders in a clinical setting by retrospectively examining the indications and findings.

Methods

A total of 562 electrophysiological studies from January 2003 to December 2021 were reviewed. The service only accepts

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referrals from movement disorders specialists. We excluded 12 patients with indication other than movement disorder, 16 patients who were part of a research study and 25 patients who had missing information. The reasons for electrophysiological studies were extracted from the referral and classified first into two main groups: suspected functional or organic movement disorders. The studies were further classified based on the main phenomenology/reason for the referral such as tremor, postural instability, jerky movements, dystonia, periodic limb/facial movements, rigidity and spasms and other categories grouped as involuntary movement evaluation (Fig. 1). The techniques used in each examination were classified as surface and needle electromyography (EMG) and electroencephalography (EEG). Specific protocols using EMG, EEG, or a combination of these techniques are summarized in Table S1.

The diagnosis from electrophysiological testing were classified into specific types of movement disorders. The electrophysiological diagnostic criteria evolved during the 18 years as specific tests were proposed and are summarized in Table 1. Functional movement disorders (FMD) was classified by the most prominent movement disorder presentation at the time of recording. Patients with myoclonus were classified as cortical or subcortical, when it was not possible to distinguish between cortical and subcortical generators, myoclonus was classified as from undefined generator. Tremor was classified into the tremor syndromes such as intention, dystonic, essential, parkinsonian, enhanced physiological and Holmes tremor.[4] Some patients with cerebellar tremor were assessed with electrophysiological tools to determine the potential benefit of surgical intervention. Abnormal orthostatic movements were classified as tremor or myoclonus. Other less common classifications within the organic movement disorders group comprises dystonia, stiff-person syndrome, myorhythmia, painful legs/arms moving toes/fingers syndrome and myokymia. In some studies, no abnormalities were found, and they were considered as normal. Studies were considered inconclusive if the findings were insufficient to draw conclusions.

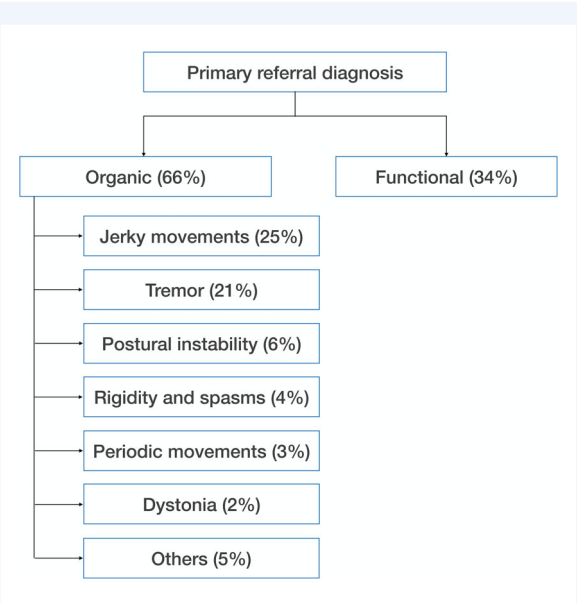

Figure 1. Classification and distribution of the different phenomenologies indicated in the referrals for electrophysiological studies.

We evaluated the agreement between the referral and the electrophysiological diagnoses (Table 2). When both diagnoses were FMD, they are regarded as concordant. If the study demonstrated organic movement disorders, agreement is established if the primary phenomenology was identical. For example, postural instability agreed with orthostatic tremor or orthostatic myoclonus, jerky movement agreed with myoclonus and tremor agreed with all subtypes of tremor. The main phenomenology could be associated with other phenomenologies in the electrodiagnostic diagnosis and is still considered as agreement.

Results

A total of 509 studies were analyzed. 51% (259) of the patients were female, with mean age of 51 years and ranges from 5 to 89 years old.

Reasons for Referral for Electrophysiological Studies

The most common reason for referral were evaluation of FMD, jerky movements, tremor, and postural instability (Table 2). When the movement disorder was not well defined in the referral, we referred to them as involuntary movement evaluation.

Functional Movement Disorders

Suspected FMD represented 34% (n = 172) of the referrals (Table 2). The movement phenomenology included functional jerky movements (n = 78), functional tremor (n = 71), functional dystonia (n = 10), functional postural instability (n = 3), functional rigidity and spasms (n = 1) and functional tics (n = 1).

Among the 172 cases, 13% (n = 23) had final diagnosis of an organic disorder without functional component. The diagnoses were tremor (n = 14), myoclonus (n = 6), dystonia (n = 2) and tic disorder (n = 1). In 16% (n = 27) of the cases, the diagnoses included both organic and functional movement, and 68% (n = 108) had a final diagnosis of only FMD. Fourteen studies (8%) were inconclusive (Table 2).

Polymyography was used in all the studies. Moreover, EEG back-averaging was performed in 25% (n = 43) of the studies to record the bereitschaftspotential, mostly in patients with jerky movements.[11] A smaller number of studies used somatosensory evoked potentials (SEP) (n = 7, 4%) and the long-latency reflexes (n = 4, 2%).

Jerky Movements

Jerky movements include possible myoclonus, tics, chorea, ballism, and tremors associated with "jerk-like" phenomenology.[12] Characterization of jerky movements was the reason for referral in 25% (n = 129) of the cases (Table 2).

Based on electrophysiological testing, the most common final diagnosis was myoclonus 60% (n = 73), divided into cortical

TABLE 1 *Summary of the electrophysiological diagnostic criteria used*

Functional Movement Disorder
• Signs of distraction, entrainment, variability of presentation (frequency/pattern of muscle contraction) or suggestibility
• Stimulus sensitive jerks with reaction times in the voluntary range (>100 ms) • Presence of bereitschaftspotential[34] • Schwingenshuch criteria for functional tremor were incorporated since 2016[5]
Myoclonus
Stimulus sensitivity (can be cortical or subcortical), triggered with a latency less than 100 ms, by: • Sound (loud clapping) • Electrical stimulation (three times the sensory threshold) • Tapping with the reflex hammer
Cortical[3] • EMG burst duration was 50 ms or lower • Cranio-caudal and predominately distal distribution; • Presence of EEG spike preceding the movements on backaveraging • Giant somatosensory evoked potential (SEP)
Subcortical • EMG burst duration >50 ms • Absence of EEG spike before movement • Absence of giant SEP
Tremor
Dystonic tremor[8,35,36] • Dystonic features such as co-contraction of the agonist and antagonist muscles during voluntary movements such as elbow/wrist flexion extension • Overflow of contraction to muscles not involved in the intended movement • Occurrence of tremor exclusively or with significantly increased amplitude with a specific posture
Enhanced physiological tremor[3,37] • Postural tremor with a frequency higher than 6 Hz • Loading test leading to a tremor frequency reduction of more than 1 Hz (peripheral generator)
Essential tremor[38,39] • Postural tremor with frequency higher than 4 Hz without functional features • Absence of latency for occurrence from rest to postural position of the arms, and frequency shift of less than 1 Hz with the loading test
Parkinsonian tremor[3,8,38] • Alternating EMG pattern of activation • Average frequency varying from 4 to 7 Hz • Occurrence at rest and possible association with a re-emergent postural component (with latency for recurrence after assuming outstretched posture)
Tremor assessment for surgical intervention[40,41] • Tremor secondary to multiple sclerosis or a cerebellar impairment with another etiology • Well-defined tremor peak from power spectral analysis higher than 3 Hz
Intention tremor[4] • Tremor recorded during voluntary movement (kinetic tremor) in which an increase in tremor amplitude occurs as the affected body part approaches its visual target
Holmes tremor[4,35,42] • Combination of rest, postural, and intention tremor with a frequency lower than 5 Hz • EMG burst duration usually longer than 150 ms

(Continues)

TABLE 1 *Continued*

Orthostatic tremor/myoclonus
Orthostatic tremor[2] • Classical orthostatic tremor: 13 to 18 Hz tremor in lower limbs while standing with high coherence between homologous muscles • Slow OT: frequency lower than 13 Hz and lower coherence • Reduced amplitude during leaning forward with the weight supported by the arms
Orthostatic myoclonus[2,31] • Irregular bursts in the lower limbs with a mean EMG burst durations of 20–100 ms that appeared while standing with frequency of 3–7 Hz • Reduced amplitude during leaning forward with the weight supported by the arms
Stiff- person syndrome
• Continuous motor unit activity at rest assessed by surface EMG from agonist–antagonist muscle pairs such as paraspinal and rectus abdominis[3] • Failure to relax the antagonist muscle while performing a movement (e.g. trunk flexion, knee flexion/extension)[3] • Exaggerated exteroceptive reflex[33,43]
Painful legs/arms moving toes/fingers
• Semi rhythmic contractions at rest, with EMG burst durations varying from 80 to 2000 ms[44] • Average frequency ranging from 0.5 to 2 Hz[9,45] • Can be partially suppressed volitionally[45]
Myorhythmia
• Rhythmic discharges of motor units with normal morphology (approximately 200 ms duration) occurring at a cranial or limb muscles at rest or during action[4,46] • Average frequency from 1 to 4 Hz[4,46]
Myokymia
• Spontaneous discharges of a single motor unit potentials firing at 5–150 Hz, appearing as doublets, triplets, or multiplets[47] • Short periods of silence between discharges, followed by the repetition of the same pattern at regular or irregular intervals[47]

Abbreviation: EMG, Electromyography; SEP, Somatosensory evoked potential; OT, Orthostatic tremor.

myoclonus (n = 40), non-localized myoclonus (n = 23) and subcortical myoclonus (n = 10). In 10% of the patients (n = 14), the final diagnosis was tremor which included dystonic tremor (n = 6), non-specific tremor (n = 5), intention tremor (n = 2) and task-specific tremor (n = 1). FMD was diagnosed in 14% of patients (n = 18). Dystonia was the diagnosis of two cases, and tic disorder was the diagnosis of one patient. Some cases (n = 19, 15%) had more than one diagnosis. In 13% of the cases, the assessment was inconclusive (n = 10) or recorded no abnormal movement (n = 7).

The techniques used included polymyography in all cases, and in some cases EEG recording with SEP (n = 25, 19%), back-averaging (n = 17, 13%), or peripheral stimulation with assessment of long-latency reflex[13–15] (n = 25, 19%) were used to localize the origin of myoclonus.[12,16]

Tremor

In 21% (n = 106) of the cases, tremor evaluation was the reason for referral (Table 2). These include clarification of the primarily phenomenology and pre-surgical evaluations. The electrophysiological diagnosis in cases of presurgical evaluation was only descriptive, such postural or intentional tremor. Only 20% of the patients (n = 21) had a final diagnosis with a phenomenology different from tremor, which included FMD (n = 18), dystonia (n = 1), clonus (n = 1), and myoclonus (n = 1). Inconclusive results were encountered in 7% (n = 7).

The remaining 73% (n = 78) patients were diagnosed with tremor syndromes. Most of them had essential tremor, followed by intention tremor, non-specific tremor and dystonic tremor (Table 2). Polymyography was used to characterize the tremor in all cases. Two cases also required needle EMG for differential diagnosis with dystonia. Needle EMG was used to accurately access deep/small muscles and allow coherence analysis between agonist and antagonist muscles EMG, which can contribute to the diagnosis of dystonia. Functional tremor overlapping with organic tremor was found in 4% (n = 4) of the patients.

Postural Instability

Postural instability was the reason for referral in 6% (n = 30) of cases. The characteristics of the patients are provided in Table 2. The majority had the diagnosis of orthostatic myoclonus (OM) 40% (n = 12), while OT was the diagnosis in 23% (n = 7). One patient had OM associated with functional tremor. The other patients had diagnosis of other types of tremor (10%, n = 3), functional tremor (7%, n = 2), and other types of myoclonus (7%, n = 2). No abnormalities found in 10% (n = 3).

TABLE 2 *Distribution of primary referral diagnoses and agreement between the referral and electrophysiological diagnoses*

Primary referral diagnosis	Mean age (range) Sex	Agreement between referral diagnosis and electrophysiological diagnosis
Functional Movement Disorder (n = 172)	50.5 (12–83) years F 53% M 47%	**Positive (79%)** FMD, n = 108 FMD associated with an organic component, n = 27 **Negative (13%)** Organic etiology, n = 23 **Non applicable (8%)** Normal study/Inconclusive, n = 14
Jerky movements (n = 129)	48 (5–87) years F 43% M 57%	**Positive (60%)** Myoclonus, n = 55 Myoclonus associated with tremor, n = 12 Myoclonus associated with dystonia, n = 4 Orthostatic myoclonus, n = 3 Myoclonus associated with tremor and dystonia, n = 1 Myoclonus associated with tics, n = 1 Myoclonus associated with FMD, n = 1 **Negative (27%)** FMD, n = 18 Tremor, n = 14 Dystonia, n = 2 Tics, n = 1 **Non applicable (13%)** Normal study / Inconclusive, n = 17
Tremor (n = 106)	49.7 (13–80) years F 48% M 52%	**Positive (73%)** Essential tremor, n = 18 Intention tremor, n = 16 Undefined tremor – rest, postural, kinetic, n = 18 Dystonic tremor, n = 8 Parkinsonian tremor, n = 6 Task specific tremor, n = 3 Enhanced physiological tremor, n = 3 Tremor associated with FMD, n = 4 Tremor associated with mycolonus, n = 1 Orthostatic tremor, n = 1 **Negative (20%)** FMD, n = 18 Myoclonus, n = 1 Clonus, n = 1 Dystonia, n = 1 **Non applicable (7%)** Inconclusive, n = 7
Postural instability (n = 30)	70.7 (48–89) years F 70% M 30%	**Positive (67%)** Orthostatic myoclonus, n = 12 Orthostatic tremor, n = 7 Orthostatic myoclonus associated with FMD, n = 1 **Negative (23%)** Myoclonus, n = 2 Dystonic tremor, n = 1 Postural tremor, n = 1 Essential tremor, n = 1 Functional, n = 2 **Non applicable (10%)** Normal study, n = 3

(Continues)

TABLE 2 *Continued*

Primary referral diagnosis	Mean age (range) Sex	Agreement between referral diagnosis and electrophysiological diagnosis
Rigidity and spasms (n = 20)	48.2 (19–77) years F 65% M 35%	**Positive (40%)** Stiff person syndrome, n = 7 Stiff leg syndrome, n = 1 **Negative (10%)** Spasticity, n = 1 FMD, n = 1 **Non applicable (50%)** Normal study/Inconclusive, n = 10
Periodic limb/facial movements (n = 14)	53.9 (37–67) years F 35% M 65%	**Positive (43%)** Painful legs moving toes, n = 2 Painful arms moving fingers, n = 1 Myorhythmia, n = 1 Myokymia, n = 1 Facial tremor (Essential tremor), n = 1 **Negative (33%)** Exagerated spinal reflex, n = 1 Dystonia, n = 1 FMD, n = 4 **Non applicable (14%)** Normal study/Inconclusive, n = 2
Dystonia (n = 12)	59.6 (42–85) years F 58% M 42%	**Positive (16%)** Dystonia, n = 1 Dystonic tremor, n = 1 **Negative (34%)** Spasticity, n = 2 Myokymia, n = 1 FMD, n = 1 **Non applicable (50%)** Normal study / Inconclusive, n = 6

Abbreviation: F, Female; FMD, Functional movement disorder; M, Male.

The muscles evaluated usually included the quadriceps, hamstrings, gastrocnemius, and tibialis anterior. In most cases, upper limb muscles such as the triceps were also evaluated to test the effects of leaning with weight supported by the arms. For these evaluations, surface EMG was the only technique needed.

Rigidity and Spasms

Investigation of rigidity and spasms was the reason for referral in 20 patients (4%). Half of the studies were inconclusive. Stiff-person syndrome was diagnosed in seven patients and stiff-leg syndrome in one patient, comprising 40% of studies in this category. One patient had diagnosis of spasticity and one had FMD. The techniques used were polymyography and exteroceptive reflex evaluation.

Periodic Limb Movements

We have seven cases (1.4%) referred for assessment of period limb movements, which included a variety of potential causes. Two cases had a diagnosis of painful leg moving toes and one with painful arm moving fingers. One patient was considered to have repetitive movement secondary to exaggerated spinal reflex. The movements were characterized by dorsiflexion of the right big toe followed by ankle dorsiflexion and knee flexion. One patient had FMD, one patient had spasticity, and two did not show abnormalities. The techniques used include polymyography and exteroceptive reflex studies.

Facial Movements

Evaluation of periodic facial movements was the reason for referral in seven cases (1.4%). Two cases were diagnosed with FMD, one case was diagnosed with myorhythmia, one with myokymia, one with dystonia, one with facial tremor secondary to essential tremor and one study was inconclusive. In addition to the polymyography, the protocol used included needle EMG to record from small muscles and EEG recording with back-averaging technique.

Dystonia

Dystonia was the reason for referral in 12 cases (2%). The study was inconclusive in six cases (50%). One patient was diagnosed

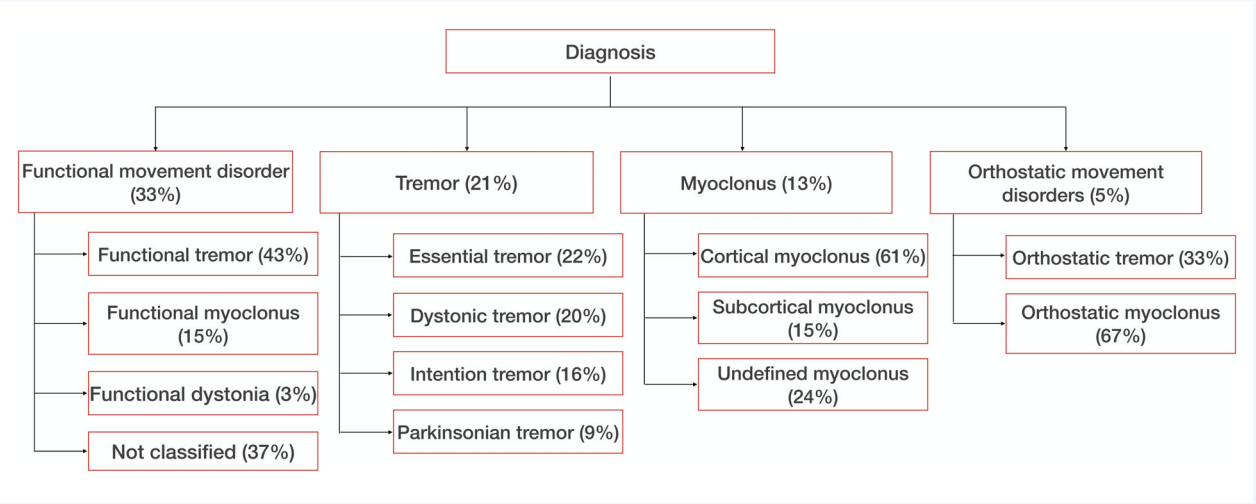

Figure 2. Main categories and distribution of diagnoses after electrophysiological studies.

with dystonia and one patient was diagnosed with dystonic tremor. Two patients were diagnosed with spasticity, one with myokymia, and one with functional tremor. The assessments included polymyography and in one case needle EMG was used to demonstrate myokymia.

Other Indications

This category includes less well-defined categories grouped as involuntary movement disorders evaluations (n = 23). It also includes two patients for evaluation of hiccups, and one patient for hypothenar spasm. The two cases of hiccups were confirmed to be of diaphragmatic origin using needle EMG, and the hypothenar spasm was due to idiopathic palmaris brevis syndrome.

Diagnosis

The most common electrophysiological diagnoses in our study were FMD, tremor and myoclonus (Fig. 2).

TABLE 3 *Description of the different categories of electrophysiological diagnoses and the reasons for disagreement with the referral diagnosis*

Electrophysiological diagnosis	Mean age (range)/Sex	Disagreement with primary referral and electrophysiological diagnosis
Functional (n = 167) 43% Tremor 37% Not classified 15% Myoclonus 3% Dystonia 2% Others*	50.8 (12–87) years F 50% M 50%	Functional features not noted (n = 50), 30%
FMD associated with an organic component (n = 24)	58 (28–81) years F 67% M 33%	Functional features not noted (n = 6), 25%
Tremor (n = 108)	51.6 (20–89) years F 43% M 57%	Functional (n = 14), 13% Jerk movement (n = 1), 1%
Myoclonus (n = 66)	48.5 (48–89) years F 44% M 66%	Functional (n = 5), 8% Tremor (n = 1), 2%
Orthostatic myoclonus (n = 14)	72.3 (45–88) years F 57% M 43%	None
Orthostatic tremor (n = 8)	67.8 (60–75) years F 63% M 37%	None

*One orthostatic tremor, one stiff-person syndrome, and one combination tremor and dystonia.
Abbreviation: F, Female; FMD, Functional movement disorder; M, Male.

Functional Movement Disorder

A total of 167 patients were diagnosed with only FMD. Among these, 71 (43%) had tremor phenomenology, 62 (37%) were not classified, 25 (15%) had myoclonus, five (3%) dystonia, one orthostatic tremor, one with hyperlordosis, paraspinal and leg stiffness associated with leg spasms suggestive of a stiff-person syndrome presentation, one combination of tremor and myoclonus, and one tremor and dystonia (Table 3). Regarding the initial diagnosis of these patients, in 30% (n = 50) FMD was not mentioned in the referral. Another 24 patients had both organic and FMD as the diagnosis as shown in the case described in Figure 3. Six (25%) of these patients did not have a suspicion of FMD in the referral.

Tremor

Tremor was the only diagnosis in 108 patients (Table 2). The majority were diagnosed with essential tremor (22%, n = 24), followed by dystonic tremor (20%, n = 22), intention tremor (16%, n = 17) and Parkinsonian tremor (9%, n = 10). The less common tremor diagnoses included task-specific tremor (4%, n = 4), enhanced physiological tremor (3%, n = 3) and Holmes tremor (1%, n = 1). Some patients did not reach an etiological diagnosis and were classified based on the phenomenology, with occurrence of tremor at rest, during maintenance of posture or kinetic tremor (25%, n = 27). Tremor was present in association with another diagnosis in 32 (30%) patients, which included dystonia, myoclonus, and FMD. In 14% of the cases, the initial

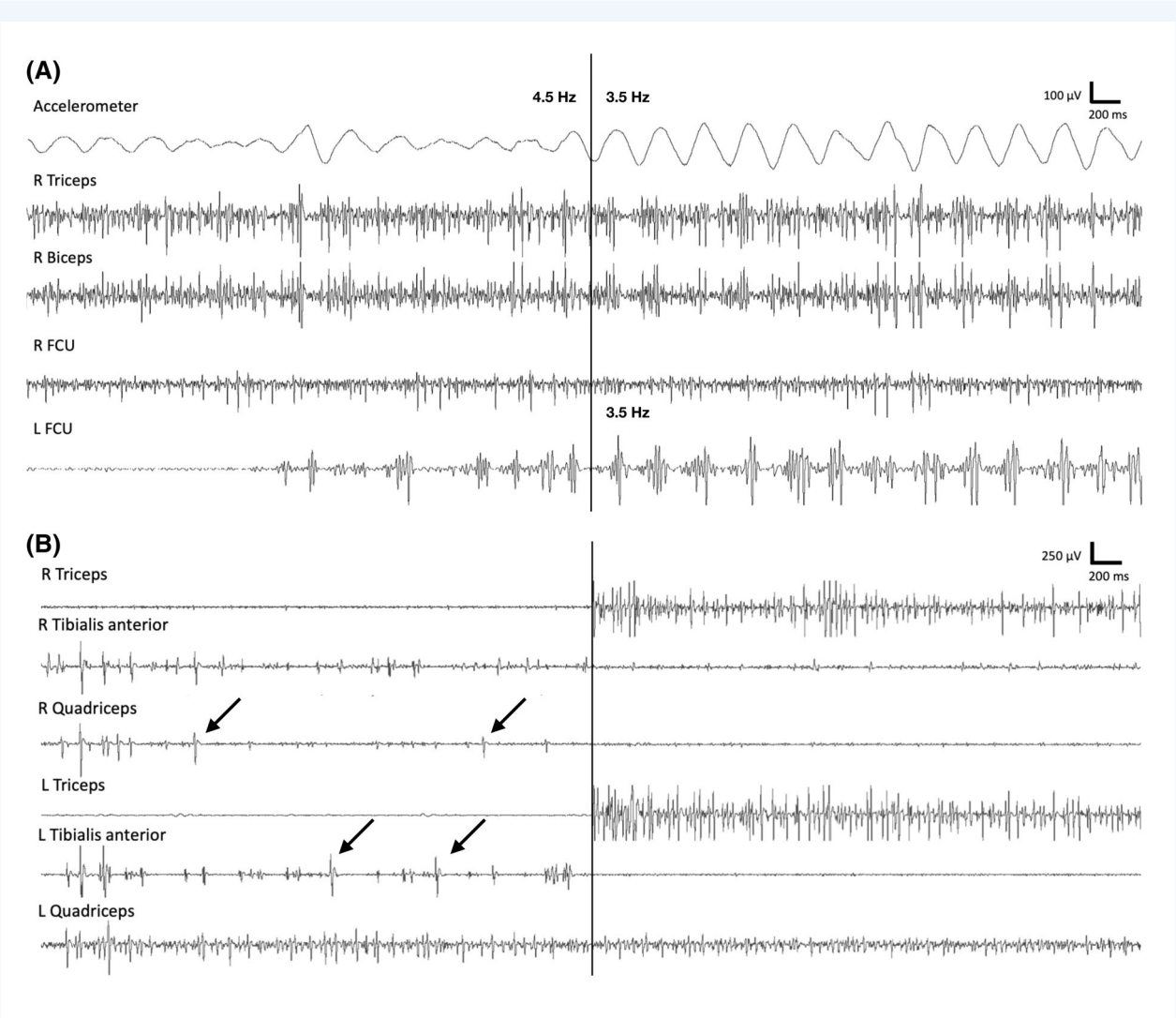

Figure 3. A 49-year-old woman with referral diagnosis of functional tremor and electrophysiological diagnosis of orthostatic myoclonus and functional tremor. Multichannel surface electromyography (EMG) and accelerometer (placed at the dorsum of the right hand) recordings. (**A**) Left: Postural tremor recorded from the right arm with arms outstretched. There were rhythmic, synchronous EMG bursts with mean frequency of 4.5 Hz involving the biceps and triceps muscles. Right: Entrainment of the tremor with left hand tapping at 3.5 Hz (**B**) Left: spontaneous myoclonus with EMG burst duration of ~50 ms recorded from both lower limbs while standing (arrows). Right: Reduction of myoclonic activities in the lower limbs while the patient was leaning forward with weight supported by the arms. FCU, flexor carpi ulnaris; L, left; R, right.

diagnosis was different from tremor, as illustrated in Figure 4 in a case referred as possible myoclonus.

Myoclonus

Sixty-six patients had the diagnosis of myoclonus, including 61% with cortical myoclonus (n = 40) (an example is shown in Fig. 5), 15% with subcortical myoclonus, and 24% were not classified between cortical or subcortical myoclonus (n = 16). Moreover, myoclonus was present in association with other movement disorders, including tremor, dystonia, tic disorder and functional component in 26 patients. In 10% of the cases, the initial diagnosis was different from myoclonus (Table 3).

Orthostatic Movement Disorders

These movements occurred in 24 patients, with orthostatic myoclonus in 67% of the cases (n = 16), orthostatic tremor was present in 33% of the patients (n = 8). The initial diagnosis described as postural instability was always in accordance with the final diagnosis in this group (Table 3).

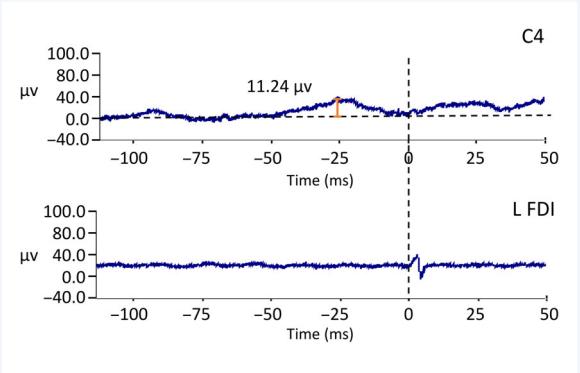

Figure 5. EEG backaveraging from a 24-year-old woman with referral diagnosis of jerky movement and electrophysiological diagnosis of cortical myoclonus. The traces show backaveraging from the onset of spontaneous contractions in the left first dorsal interosseous (FDI) muscle recorded with surface EMG (defined as time 0) from 18 trials. The top recording shows EEG from C4 electrode with the linked ear as reference. A negative discharge occurred at 25 ms before onset of EMG activities in the FDI muscle. The bottom recording shows the average of spontaneous activations of the left FDI muscle from the same 18 trials.

Discussion

We retrospectively investigated how clinical electrophysiological testing can help in the diagnosis in a specialized movement disorders center. The most common indication was FMD evaluation, followed by jerky movement assessment, tremor classification and postural instability investigation. We evaluated the diagnostic process in 509 patients to understand what the main indications were, and if the electrophysiological evaluation confirmed, changed or was not able to help in the diagnosis. Overall, the electrophysiological assessment had a high agreement with the referral diagnosis. However, electrophysiological evaluations changed the diagnosis in 13% of the patients with FMD indication, in 27% of the jerk movements referrals, 20% of the patients with tremor, and 23% of the patients with postural instability. These results highlight the importance of these tests in clarifying the clinical diagnosis.

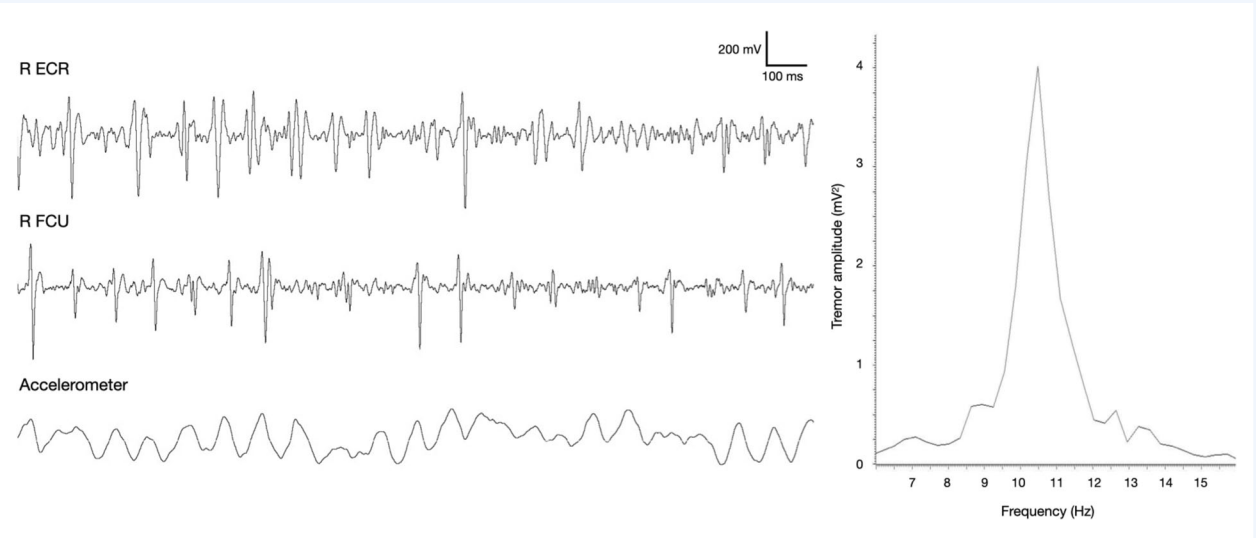

Figure 4. Recordings from a 44-year-old woman with referral diagnosis of jerky movement and electrophysiological diagnosis of high frequency tremor. Left: Multichannel surface electromyography (EMG) and accelerometer (placed at the dorsum of the right hand) recordings showing an alternating tremor with mean frequency of 10.5 Hz while the arms were in a semi-supinated position. Right: Power spectral analysis of the accelerometer recording. The high frequency tremor made the movement appeared jerky. ECR, extensor carpi radialis; FCU, flexor carpi ulnaris; R, right.

To understand how the electrophysiological assessment can add information to the initial clinical suspicion, we evaluated the disagreement between the primary referral diagnosis and the electrophysiological diagnosis. In FMD, functional features were not noted in 30% of cases, tremor was referred as another phenomenology in 14% of the cases, and myoclonus was referred as FMD, or tremor in 10% of the times. Orthostatic tremor or myoclonus had no disagreement with the initial diagnosis as postural instability.

The use of electrophysiological assessment to diagnose FMD was encouraged in the recent consensus and reviews on this topic.[17–20] As highlighted in the example shown in Fig. 3, electrophysiological evaluation can reveal an organic component in suspected FMD that could change the choice of treatment. Moreover, the electrophysiological evaluation provides objective evidence of functional origin of abnormal movements that increases diagnostic certainty and potentially facilitate patients in accepting their diagnosis. These results facilitate the discussion of the diagnosis with patients, may contribute to increased adherence to proposed treatment.

Jerky movement is a challenging phenomenology that may encompass different movements alone or in combination.[11] In some cases, even after carefully clinical examination in different positions and using a variety of maneuvers, it was still unclear if the movement represents tremor, myoclonus, dystonia, chorea or if there was a functional component. Ancillary evaluation with electrophysiology can help to clarify the phenomenology in these cases as illustrated in Fig. 4, in which electrophysiological evaluation revealed a high frequency tremor (10.5 Hz) that was provisionally diagnosed as myoclonus. No patient had chorea as the electrophysiological diagnosis in the jerky movement referrals. This may be related to the absence of specific criteria to characterize chorea using surface EMG[21] and the low prevalence of this movement disorder. Moreover, the referring movement disorders neurologists may not feel that electrophysiological testing is sensitive for the diagnosis of chorea.

Another important utility of the electrophysiological assessment is to differentiate between the cortical and subcortical generators in myoclonus as illustrated in Fig. 5, which shows an EEG spike preceding jerky movements in a patient with cortical myoclonus. This finding helps to guide additional investigations and therapeutic options.[11,22–24] However, in 24% of the cases, the generator could not be identified. Some cases had features suggestive of cortical myoclonus such as short EMG burst durations and distal predominance, but jerk-locked backaveraging was negative and there were no exaggerated long-latency reflexes. The EEG backaveraging technique is often limited by low number of epochs available due to rare spontaneous jerks, artifacts such as EMG artifacts from facial movements, and it could not be performed in patients who had only stimulus sensitive jerks but no spontaneous jerks. Moreover, some patients have features suggestive of both cortical and subcortical combined origin of myoclonus.[8]

Patients with tremor showed the importance of electrophysiological assessment in selected cases. Some cases initially suspected of having tremor were found to have myoclonus, dystonia or FMD, demonstrating the role of electrophysiology in clarifying the phenomenology in more challenging presentations.[25] Another important utility is classification of specific syndromes, which can guide the therapeutical approach. The finding of the most common tremor diagnosis being essential tremor followed by dystonic and intentional tremors is similar to the findings at another movement disorders center, which had essential tremor as most prevalent followed by parkinsonian and intention tremor.[26]

Evaluation of postural instability is another important contribution of the electrophysiological studies. Previous reports highlighted essential tremor, action myoclonus, generalized polymyoclonus, myoclonic tremor, exaggerated clonus, parkinsonism and normal pressure hydrocephalus as differential diagnoses.[27–29] The diagnosis most commonly considered with the complaint of shaking when standing is OT. However, in our sample, orthostatic myoclonus (OM) was more prevalent than OT. Without electrophysiological assessment, it is not possible to differentiate between these entities,[2,27,29,30] highlighting the utility of this assessment. OM is a relatively recently described and has a broad spectrum of presentation with similar clinical presentation as OT, but sometimes without clear clinical finding of shaking. The differentiation between OT and OM is essential since they are managed differently. Our population of OT and OM has similar features to those previously described, with higher prevalence after the age of 60 and female predominance.[31]

Other less common but notable indications for electrophysiological studies include rigidity and spasms, periodic limb and facial movements. Electrophysiological assessment is important to confirm the diagnosis of stiff-person syndrome, which has the main features of continuous muscle motor unit firing,[32] co-contraction of agonist–antagonist muscles and exaggerated exteroceptive reflexes.[3,33] Electrophysiological evaluation plays an important role in distinguishing between stiff-person syndrome, spasticity and FMD, which were found in some patients with the provisional diagnosis of stiff-person syndrome in our study. Periodic movements can be difficult to classify based solely on clinical observation. Electrophysiological studies permit identification of more specific entities such as myorhythmia, myokymia, and painful legs moving toes syndrome.

Limitations

The limitations of this study include the retrospective nature of the study and referral bias in a tertiary referral center for movement disorders, which contributed to the high prevalence of challenging presentations and potentially the high rate of disagreement between the clinician and electrophysiological diagnosis. In addition, although we used generally accepted electrophysiological criteria for the diagnoses, most of these criteria are based on small studies and they have not been vigorously validated in large number of cases.

In conclusion, this retrospective study demonstrates the importance of electrophysiological studies in the diagnosis of a diverse set of movement disorders in clinical practice. Electrophysiological assessment has many utilities, such as helping to

confirm diagnosis of FMD and stiff-person syndrome, and to distinguish the phenomenologies of tremor, myoclonus, tic, and dystonia. Moreover, it can help to define tremor syndrome and the origin of myoclonus. It is also crucial for elucidating the type of periodic movement and separating OT and OM. More studies are needed to standardize the protocols between centers, and to promote the availability and use of these techniques in other movement disorders clinics.

Author Roles

(1) Research project: A. Conception, B. Organization, C. Execution; (2) Statistical Analysis: A. Design, B. Execution, C. Review and Critique; (3) Manuscript Preparation: A. Writing of the first draft, B. Review and Critique.

T.G.: 1A, 1B, 1C, 2A, 2B, 3A.

R.C.: 1A, 1B, 2C, 3B.

Disclosure

Ethical Compliance Statement: The study was approved by the University Health Network Research Ethics Board (reference number 22–5841). The Research Ethics Board determined that informed patient consent was not necessary for this work. We confirm that we have read the Journal's position on issues involved in ethical publication and affirm that this work is consistent with those guidelines.

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Supporting Information

Supporting information may be found in the online version of this article.

TABLE S1. Summary of the electrophysiological techniques used.