Parkinsonism and Related Disorders 132 (2025) 107216

Contents lists available at ScienceDirect

Parkinsonism and Related Disorders

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

IAPRD new consensus classification of myoclonus

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A B S T R A C T

Introduction: Recent new advances in myoclonus characterization and etiology justify an update of the 40-year-old respected classification of myoclonus proposed by Marsden, Hallett, and Fahn. New advances include genetic studies and clinical neurophysiology characterization.
Methods: The IAPRD appointed an expert panel to develop a new myoclonus classification. The Delphi Method of consensus determination was employed using a panel of fifteen international experts in myoclonus. In an in-person meeting, an Axis approach, previously used for dystonia and tremor was ratified by the panel: Axis I included clinical and neurophysiology features, Axis II included etiology categories. As a unique part of our Axis approach, Clinical Neurophysiology was included as Axis Ib. The first Delphi survey round queried agreement on major headings in Axes Ia and Ib, myoclonus clinical syndromes, and Axis II. In the second round, the full expert panel was surveyed on constituents and specific characteristics of each feature that had consensus in the first round.
Results: In the first round, the percentage of agreement for the fifty-three out of the 56 items was greater than 60.0 %, indicating strong consensus among expert panel members. In the second round, for Axis Ia, Axis Ib, and Axis II, strong agreement was also achieved. For both rounds, Physiological Myoclonus had the lowest agreement. Comments from the whole panel were incorporated into the consensus results.
Conclusion: This Myoclonus Classification, which reached consensus using the Delphi Method, will facilitate a collaborative effort among myoclonus investigators to find better diagnostics and treatment for myoclonus patients.

1. Introduction

Myoclonus is defined as a sudden, brief, lightning-like muscle contraction ("positive myoclonus"), or contraction inhibition ("negative myoclonus"). Although this definition might seem straightforward, myoclonus has often been described as an enigmatic entity owing to the wide breadth of phenotypes and causes [1]. Since first recognition in the late 1800s, there has been ongoing debate as to what constitutes myoclonus and how different forms of myoclonus should be classified.

The first descriptions of myoclonus date back to 1881, when Professor Nikolaus Friedreich described a case of stimulus-induced multifocal muscle jerks distinct from the previously described chorea or

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https://doi.org/10.1016/j.parkreldis.2024.107216
Received 8 October 2024; Received in revised form 17 November 2024; Accepted 20 November 2024
Available online 4 December 2024

epileptic spasms [2]. He termed the movement disorder "paramyoklonus multiplex," which subsequently became abbreviated to "myoclonus." In the following decade, the term "myoclonus" was used to describe a variety of jerk or twitch like movements, some of which today would be known as tics, fibrillations, or otherwise. At the turn of the 20th century, the term "myoclonus" became further refined by Unverricht and Rabot describing cases of familial progressive myoclonus and epilepsy. They proposed that myoclonus could be classified according to three categories: Symptomatic, Essential, and Familial Myoclonic Epilepsy [3]. Throughout the first half of the 20th century, there were several additional descriptions of myoclonic epilepsies, including reports of myoclonus associated with cerebellar atrophy. In 1960, Aigner and Mulder (Myoclonus: Clinical Significance and an Approach to Classification) proposed a classification scheme, including four groups, i.e., myoclonus with: I) seizures and neurologic deficit; II) seizures without neurologic deficits; III) no seizures or neurologic deficits; IV) neurologic deficit but no seizures. It was posited that the association of myoclonus with other nervous system disorders portended a worse prognosis, and therefore the value of such a classification system would be its ability to assist with clinical prognostication [4].

In 1982, Marsden, Hallett, and Fahn (The nosology and pathophysiology of myoclonus) proposed a classification system that has been widely used for over 40 years, and it is still the most used myoclonus classification for all etiologies (Table 1) [5]. The first category was designated as *Physiologic* myoclonus, or myoclonus that occurs in the context of normal physiological functions. The second category of *Essential* myoclonus consists of myoclonus without other neurological deficits and without a secondary cause. The third category includes *Epileptic* myoclonus, where seizures predominate the clinical picture rather than other deficits such as encephalopathy, and myoclonus is seen as a major clinical part or accompaniment. The fourth and broadest category, like the 1903 classification of Lundborg, is termed *Symptomatic* and refers to myoclonus caused by another disease state where static or progressive encephalopathy is the predominant clinical feature.

As our understanding of myoclonus has evolved in terms of defining syndromes, etiologies, and pathophysiological underpinnings, additions to the classic classification scheme have been proposed. The addition of myoclonus entities and subcategories has resulted in a lack of consensus as to which added items should be widely used and how these entities best integrate. In addition, advances in electrophysiological testing have given rise to neurophysiological classifications of myoclonus [6–8]. Along with these growing bodies of knowledge, placement of new entities of myoclonus under the old categories has produced debate and confusion. For example, terms such as "essential" or "primary" have been used but are nonspecific and may no longer apply in some situations as the genetics of myoclonus have become better understood. As transformational amounts of genetic information have been discovered due to the advent of whole genome sequencing, this has given rise to a genetic classification of myoclonus, and a new nomenclature for these syndromes [9]. Lastly, the relationship of seizures to myoclonus needs ongoing clarification, and the previous category of "Epileptic myoclonus" no longer suffices. Under the new International League Against Epilepsy (ILAE) Classification system, the seizure type term "Myoclonic" exists under both focal and generalized onset with multiple variations [10].

Further, the growing realm of functional neurological disorders (FNDs) is not well accounted for in myoclonus classifications despite comprising many referrals in clinical practice [11]. Although the terms "functional myoclonus" and "functional jerks" have been proposed to refer to jerky movements resembling myoclonus that result from a functional neurological disorder, there has not been consensus as to how these movements are best termed and how these movements should be included within the phenomenology of myoclonus.

Our understanding of the genetic and pathophysiological underpinnings of myoclonus must be synthesized with important clinical information to be applicable to the practice of medicine. As such, there is

Table 1
Commonly Used Previous Classification of Myoclonus by Etiology [5]. From:C. D. Marsden, M. Hallett, S. Fahn, The nosology and pathophysiology of myoclonus, in: C.D. Marsden, S. Fahn (Eds.), Movement Disorders, Butterworth Scientific, London, 1982, pp. 196–248.

Physiologic myoclonus		Hypnic jerks
		Anxiety-induced
		Exercise-induced
		Hiccough (singultus)
		Benign infantile myoclonus with feeding
Essential myoclonus		Hereditary
		Sporadic
Epileptic myoclonus	Fragments of epilepsy	Isolated epileptic myoclonus
		Epilepsia partialis continua
		Idiopathic stimulus sensitive myoclonus
		Photosensitive myoclonus
		Myoclonic absenses
	Childhood myoclonic epilepsy	
	Progressive myoclonic epilepsy	Infantile spasms
		Severe myoclonic epilepsy of infancy (Dravet syndrome)
		Myoclonic astatic epilepsy (Doose syndrome)
		Cryptogenic myoclonus epilepsy (Aicardi syndrome)
		Benign myoclonic epilepsy of infancy
		Juvenile myoclonic epilepsy
	Familial cortical myoclonic tremor with epilepsy	
Symptomatic myoclonus	Storage disease	Lafora body disease
		GM2 gangliosidosis
		Tay-Sachs disease
		Gaucher disease
		Krabbe leukodystrophy
		Ceroid-lipofuscinosis
		Sialidosis types 1 and 2
	Spinocerebellar degeneration	Progressive myoclonus ataxia
		Friedreich ataxia
		Ataxia telangectasia
		Other spinocerebellar degeneration
	Basal ganglia degeneration	Wilson disease
		Torsion dystonia
		Neurodegeneration with brain iron accumulation (NBIA)
		Progressive supranuclear palsy
		Huntington's disease
		Parkinson's disease
		Multiple system atrophy
		Corticobasal degeneration
		Dentatorubropallidoluysian atrophy (DRPLA)
	Dementias	Creutzfeld-Jakob disease
		Alzheimer's disease
		Dementia with Lewy bodies
		Frontotemporal dementia
		Rett syndrome
	Infectious/ Postinfectious	Subacute sclerosing panencephalitis
		Viral encephalitis (Herpes simplex, arbovirus, etc)
		Human immunodeficiency virus
		Malaria
		Cryptococcus
		Lyme disease
		Syphilis
		Whipple's disease
		Progressive multifocal leukoencephalopathy
		Other postinfectious encephalitis
	Inflammatory/ Paraneoplastic	Opsoclonus-myoclonus syndrome
		Connective tissue disease
		Steroid-responsive autoimmune encephalopathy with autoimmune thyroiditis
		Rasmussen encephalitis

(*continued on next page*)

Table 1 (*continued*)

	Anti-neuronal nuclear antibody
	Voltage-gated potassium channel antibody
	N-methyl-D-aspartate receptor antibody
	Glutamic acid decarboxylase antibody
	Glycine receptor antibody
	Celiac disease
Metabolic	Renal failure
	Dialysis syndrome
	Hepatic failure
	Hypoglycemia
	Hyperglycemia
	Hyponatremia
	Hyperthyroidism
	Multiple carboxylase deficiency
	Hypoxia
	Metabolic alkalosis
	Vitamin E deficiency
	Biotin deficiency
Toxic	Opioids
	Anti-epileptic medications (carbamazepine, phenytoin, pregabalin, lamotrigine)
	SSRIs
	Lithium
	Heavy metals
	Anesthetics
	Levodopa
	Antiarrhythmic medications
	Medication withdrawal
Physical encephalopathies	Posthypoxic injury
	Post-traumatic
	Heat stroke
	Electric shock
	Decompression injury
Focal nervous system damage	Post-stroke
	Post-thalamotomy
	Neoplasm
	Trauma
	Inflammatory (e.g. multiple sclerosis)
	Developmental (e.g. dysplasia)
Exaggerated startle syndrome	Hereditary
	Sporadic
Multiple system degeneration	Mitochondrial disorders
	Allgrove syndrome
	DiGeorge syndrome

need to outline a classification system that synthesizes and organizes the vast knowledge we have accrued. A new consensus myoclonus classification scheme will serve as an important guide in determining and outlining diagnostic approach and treatment for clinicians. In addition, it will outline opportunities to adjust and expand the classification as advances in myoclonus occur.

2. Methods

This consensus effort for myoclonus classification was initiated and sponsored by the International Association of Parkinsonism and Related Disorders (IAPRD).

Method and Panel Selection: In 2020, the IAPRD appointed a panel of 15 myoclonus experts, including a chair and co-chair (JNC&MAJT), with a mandate to develop a new classification for myoclonus. The Delphi Method was chosen to achieve consensus on a new classification of myoclonus [12,13]. Precision Consulting, LLC, with Delphi Method expertise, provided statistical and survey programming support.

Delphi Rounds: Two rounds were conducted. Prior to the first round, a five-member subgroup conducted a literature review of salient issues and drafted a proposal for the full expert panel to review and discuss at an in-person full panel meeting. This included the recent classifications for dystonia and tremor which used a two-axis approach, with Axis I for clinical features and Axis II for etiology [14,15]. The clinical features in Axis I were used to derive clinical syndromes. The same general scheme was proposed for this myoclonus classification. An additional Axis Ib was proposed for neurophysiology of myoclonus and its subtypes, because of its importance for myoclonus characterization. Thus, Axis I designates clinical and neurophysiology features. This axis approach and some major components of each axis were ratified in an in-person full expert panel meeting at the World Congress on Parkinson's Disease and Related Disorders on May 2, 2022.

The full expert panel in-person discussion was incorporated into the first-round survey. The Axes Ia and Ib, myoclonus clinical syndromes, and Axis II were proposed for agreement. In addition, major components of each axis and a list of myoclonus clinical syndromes were included in the survey. The full expert panel was invited to make edits, additions, and comments on all items in the first survey round. A priori agreement of 60 % was defined as consensus for both survey rounds. The survey was administered electronically to the email of the 15 expert panel members, and responses were collected electronically by Precision Consulting, LLC, who conducted all statistical analyses.

In the second round of the Delphi Method, the full expert panel was surveyed on constituents and specific characteristics of each feature that had consensus in the first round. In addition, comments from the first round were used to modify items for the second round. A report summarizing the outcomes of the initial survey round was provided to the panel with the second survey round. Again, the full expert panel was invited to make edits, additions, and comments on all items. Alongside the quantitative analysis of the full expert panel's responses, qualitative feedback provided through comments was also carefully considered in determining the final classification. The second survey round was administered electronically in the same manner as the first.

A draft of this manuscript was sent to the full expert panel, and the comments were incorporated into the final draft of this manuscript.

3. Results

First round. Panel response was 100 % with all fifteen expert members responding. The percentage of agreement for the fifty-three out of the 56 items was greater than 60.0 %, indicating strong consensus among expert panel members. Agreement was not obtained for the following three items: "Epileptic myoclonus" as an alternative to Cortical-Subcortical myoclonus as an Axis Ib neurophysiological subtype; "Normal myoclonus" as an alternative to Physiologic myoclonus in the Fahn-Marsden classification; and "Unknown" as an item under Physiological myoclonus.

The overall average percentages of agreement across Axis Ia Clinical Features were as follows: Historical Features (87.5 %); Myoclonus (exam) Characteristics (93.8 %); and Diagnostic Evaluation (78.1 %). For the listed suggested Myoclonus Clinical Syndromes there was 84.4 % agreement overall, all Clinical Myoclonus Syndromes met agreement, and no new Clinical Myoclonus Syndromes were contributed by the full expert panel. For Axis Ib (Clinical Neurophysiology), there was 75.6 % agreement overall. Under Axis II (Etiology): Genetic and its items had 80 % agreement overall, with Acquired (85.6 %), Idiopathic (89.6 %) and "Physiological Myoclonus" (62.5 %) overall. Moreover, the overall percentage of agreement across all items in the first round was 81.0 %, showing strong consensus.

Second round. Eleven of 15 (73 %) expert panel members responded to the second-round survey, which was acceptable to Delphi Method standards (>70 %). In addition to using the consensus items from the first round, comments from the whole panel were incorporated where possible. The purpose of the second survey round was to: 1) assess agreement on more detailed items under items that were agreed to in the first round; 2) assess agreement on characteristics of items that were agreed to in the first round. The percentage of agreement for all 129 items was 91.5 % overall, indicating strong levels of consensus among the panel experts for all the items included in the second-round survey.

For Axis Ia (Clinical Features): The average percentage of agreement

across all item characteristics for Historical Features was 87.4 %; with Myoclonus Characteristics (92.2 %); and Diagnostic Evaluation (99.2 %). Average agreements for Axis Ib (Neurophysiology of myoclonus) characteristics were as follows: Cortical myoclonus (95.4 %); Cortical-Subcortical myoclonus (85.5 %); Subcortical myoclonus (95.5 %); Brainstem myoclonus (focal/generalized) (93.5 %); Spinal myoclonus (focal) (85.5 %); Propriospinal myoclonus (90.9 %); Peripheral myoclonus (89.1 %); and Functional Jerk Features (97.4 %). As in the first round, no Clinical Myoclonus Syndromes were rejected or added. For Axis II (Etiology), the average percentage of agreements across all items were as follows: Genetic (72.7 %); Acquired was (95.4 %); Nervous System Lesions (93.7 %); Idiopathic (95.4 %); and Physiological Myoclonus (81.8 %).

The new consensus Classification of Myoclonus is given in Table 2.

4. Discussion

We have proposed a new consensus-based classification for myoclonus that integrates previous classifications and considers clinical features, anatomical distribution, neurophysiological markers, and etiology. This consensus was achieved using the Delphi Method, a systematic process for developing agreement among a panel of experts that is especially valuable in areas where statistical model-based evidence is lacking, knowledge is uncertain and incomplete, and consensus expert judgment is more dependable than individual opinion [16]. Alongside the quantitative analysis of the full myoclonus expert panel's responses, qualitative feedback provided through comments was also carefully considered in determining the final classification. By synthesizing both quantitative and qualitative inputs, the Delphi Method facilitated a comprehensive evaluation process, ensuring that the resulting new myoclonus classification accurately reflected the consensus and expertise across all myoclonus expert panel members, who were selected based on their diverse backgrounds to achieve a broader perspective and generalization of consensus.

The Delphi consensus supported an axis approach reflected in recent published classifications for dystonia and tremor [14,15]. Accordingly, the classification is based on two axes: Axis I, which includes a detailed clinical characterization of the patient with myoclonus, and Axis II, which encompasses its etiology. The rationale behind this scheme is to encourage clinicians to recognize the syndrome or phenotype (a combination of signs and symptoms that occur together) and define a particular condition - Axis I (a&b), leading to the identification of one or more underlying etiologies (Axis II). However, our consensus has notable differences tailored to myoclonus. The most notable of these differences is the inclusion of Axis Ib for Clinical Neurophysiology features which also contributes to defining clinical myoclonus syndromes along with Axis Ia Clinical features.

4.1. Axis Ia. Clinical features

The Clinical features in Axis Ia contains historical features, myoclonus (exam) characteristics, and diagnostic evaluation. All attained high agreement among the expert panel. Multiple articles have emphasized important clinical features in identifying different subtypes of myoclonus and myoclonus syndromes [6,17]. Obtained from the clinical history of the myoclonus received from the patient, the historical features drive a targeted investigation for neurological pathophysiology and ultimately, etiology. For myoclonus, age of onset is crucial for differential diagnosis, as early-onset myoclonic syndromes rarely present in adulthood, and vice versa. In addition, temporal course and mode of onset have implications for the pathophysiology of the underlying etiology. Basic neurological principles apply. Thus, a relatively acute onset may suggest an inflammatory, metabolic, drug-induced myoclonus, functional jerks, but a chronic and progressive course may suggest a neurodegenerative disease. The existence of co-morbid medical or neurological conditions is particularly important since secondary etiologies of myoclonus are quite common. Lastly, medication-induced myoclonus is prominent, and its possibility should always be considered.

Myoclonus characteristics derived from physical examination are key to inform clinical description as well as to guide diagnostic considerations. The anatomical distribution, activation state, and temporal profile observed provide a current status documentation of the myoclonus [18]. For instance, focal or multifocal myoclonus of the face or distal limbs (particularly upper extremities which have a larger cortical representation) is typical of cortical myoclonus, which is often stimulus-sensitive. In contrast, highly stimulus-sensitive generalized or axial and proximal limb myoclonus is likely reticular in origin. When myoclonus is focal and follows a nerve or plexus distribution, peripheral myoclonus should be considered. The unified myoclonus rating scale (UMRS) is the currently used scale to measure myoclonus clinical severity [19].

The diagnostic evaluation should be guided by the historical features and myoclonus (exam) characteristics; however, laboratory and imaging are almost always essential. If the diagnostic etiology is not obvious, genetic testing and electrodiagnostic testing should be liberally employed as indicated. Guidelines on a staged evaluation of a patient's myoclonus are available [18,20].

4.2. Axis Ib. Clinical neurophysiology features

The inclusion of detailed clinical neurophysiology features has not been present in previously published classifications of movement disorders. Historically, myoclonus has been closely tied to its neurophysiology characteristics and presumed sources of pathophysiology. Ideally, neurophysiology studies should be performed by those with formal neurophysiological training in EEG and/or EMG and with experience in recording data directly from myoclonus patients. Multiple authors have published neurophysiological characteristics of different myoclonus types and their classification [6,7,21]. The sensitivity and specificity of most of the electrophysiological features is lacking [8], most clinicians rely on the case series published and their own experience. In this study, characteristics of various clinical neurophysiology features reached agreement among the expert panel.

In the first survey round, an anatomic-physiological classification of myoclonus was proposed, and consensus was reached on the following myoclonus subtypes: cortical, cortical-subcortical, subcortical, brainstem, spinal, propriospinal, peripheral, and functional jerks. Although the last category implies an etiological diagnosis, i.e., FND, it was included due to its clinical relevance and the presence of neurophysiological markers that assist in distinguishing it from myoclonus.

Cortical myoclonus is by far the most common neurophysiological type of myoclonus, and its features reached the highest panel agreement among the Clinical neurophysiology categories. Cortical myoclonus confirmation is the most well accepted, with a jerk-locked back-averaged pre-myoclonus EEG transient providing the gold standard of its confirmation. The myoclonus EMG duration, muscle recruitment pattern, presence of long latency reflex type I, and enlarged somatosensory evoked potential (SEP) are deemed supportive by many authors, but they have not been extensively studied for validation [21]. The relationship between cortical myoclonus and motor seizures, both focal and secondarily generalized, has been described [22]. These authors put forth that all these entities, when taken together, exhibit a "spectrum" of varying combinations of stimulus-sensitive myoclonus, spontaneous myoclonus, muscle activation myoclonus, epilepsia partialis continua, focal motor seizures, and secondarily generalized convulsions [22–24]. Further, these authors posited that patients along this spectrum represented subtle differences in the site of abnormality in sensorimotor cortical neuronal mechanisms. Hallett has divided myoclonus into epileptic and non-epileptic [25], suggesting that cortical reflex myoclonus is a fragment of partial epilepsy, reticular reflex myoclonus is a fragment of generalized epilepsy, and primary generalized epileptic myoclonus is a fragment of primary generalized epilepsy.

Table 2
CLASSIFICATION OF MYOCLONUS. The new consensus classification for myoclonus via the Delphi Method.

Axis Ia. Clinical features	
Historical Features	
Age of onset	1. Infancy (birth-2 years) 2. Childhood (3–12 years) 3. Adolescence (13–20 years) 4. Early adulthood (21–50 years) 5. Late adulthood (>50 years)
Temporal course	1. Stable course 2. Progressive course 3. Improving course
Temporal onset	1. Gradual onset 2. Sudden onset
Co-morbid medical or neurological conditions	
Medication	
Family history	
Myoclonus Characteristics	
Anatomical distribution	1. Focal 2. Multifocal 3. Generalized 4. Hemi 5. Distal 6. Proximal limb 7. Axial
Activation state	1. Rest/spontaneous 2. Action/intention 3. Stimulus induced (Tactile) 4. Stimulus induced (Visual) 5. Stimulus induced (Auditory) 6. Stimulus induced (Startle) 7. State-specific (Sleep) 8. State-specific (Sleep transitions)
Temporal profile	1. Constant 2. Paroxysmal 3. Rhythmic
Other neurological features	1. Isolated 2. Combined
Diagnostic Evaluation	
Laboratory studies (non-genetic)	1. Electrolytes 2. Organ function 3. Toxins/drugs 4. Infections 5. Antibodies
Genetic testing	
Neuroimaging	
Electrodiagnostic testing	1. Surface Electromyography (sEMG) 2. Electroencephalography (EEG) 3. Evoked potentials 4. Corticomuscular coherence 5. Reflex testing 6. Back-averaging for fast or slow potential (jerk-locked or EMG discharge)

Axis Ib. Clinical Neurophysiology Features	
Cortical myoclonus	
Duration	1. sEMG discharges <100 ms (ms) (usually <50 ms) 2. Negative myoclonus corresponding to a silent period of 100–400 ms within a tonic sEMG discharge
Muscle recruitment pattern	1. Often focal or multifocal 2. Co-contracting sEMG discharges among agonist-antagonist muscle groups; cranial-caudal progression may be seen
EEG	1. Time-locked cortical sharp wave on EEG corresponding to a sEMG discharge in a corresponding anatomical distribution 2. Cortical sharp wave precedes the onset of the sEMG myoclonus discharge (with 15–22 ms latency in upper extremity, ~40 ms in lower extremities)
Other	1. Presence of long latency reflex type I 2. Presence of enlarged somatosensory evoked potential (SEP)
Cortical-Subcortical myoclonus	
Duration	sEMG discharges of 50–100 ms
Anatomical distribution	Generalized bilateral synchronous
EEG patterns observed	1. EEG correlate is polyspike, polyspike and wave (4–6 Hz spike and wave), with variable EEG-EMG latency 2. sEMG discharges time-locked with generalized spike and wave discharges on EEG (e.g. absence seizures with myoclonus) 3. EEG correlate is bifrontal/fronto-central/bifrontal negativity to myoclonus EMG discharge.
Subcortical myoclonus	
Duration	Normally >100 ms, but could be 50–100 ms
Muscle recruitment	Variable
EEG	Absence of time-locked cortical potential
Other	Absence of giant SEP/Absence of long latency reflex type I
Brainstem (focal/generalized) myoclonus	
Reticular	1. Contiguous sequential activation of brainstem innervated structure

(*continued on next page*)

Table 2 (*continued*)

Axis Ib. Clinical Neurophysiology Features	
	2. Stimulus sensitivity
	3. If EEG potential exists, it begins after beginning sEMG discharge
Startle	1. Contiguous sequential activation of brainstem innervated structures
	2. Stimulus sensitivity
	3. Early auditory blink response
Focal	Localized activation brainstem innervated structures at 1–3 contiguous levels
Spinal (focal distribution) myoclonus	
Duration	>100 ms sEMG discharges
Muscle recruitment	1. sEMG discharges localized to 1–3 contiguous levels of the spinal cord
	2. Co-contracting
EEG	Absence of time-locked cortical potential
Other	Absence of giant SEP/Absence of long latency reflex type I
Propriospinal myoclonus	
Duration	>100 ms
Muscle recruitment	sEMG discharges arising from trunk muscles followed by rostral and caudal activation through propriospinal pathways (slow conduction
consistent	velocity)
EEG	Absence of time-locked cortical potential
Other	Absence of giant SEP/Absence of long latency reflex type I
Peripheral myoclonus	
EMG Duration	Variable, 50–100 ms
Muscle recruitment	1. sEMG discharges localized to muscles innervated by single peripheral nerve or nerve root
	2. sEMG co-contracting discharges across a specifical peripheral nerve distribution
EEG	Absence of time-locked cortical potential
Other	Absence of giant SEP/Absence of long latency reflex type I
Functional jerk features	
Duration	Most of the times >200 ms sEMG burst duration
Muscle recruitment commonly shows variable pattern	
EEG	1. Bereitschaftspotential present on EEG-EMG back averaging
	2. Event-related desynchronization in broad beta band with a reduction of beta and low gamma oscillations prior to cued and self-paced movement
Other	1. Distractible
	2. Entrainable
	3. Non-physiologic stimulus latency

Clinical Myoclonus Syndromes (derived from Axis Ia and Ib)	
1.	Myoclonus-Dystonia
2.	Progressive myoclonus epilepsy
3.	Progressive myoclonus ataxia
4.	Cortical myoclonus tremor
5.	Juvenile myoclonic epilepsy
6.	Opsoclonus-myoclonus-ataxia
7.	Functional jerks

Axis II. Etiology[a]
Genetic
Prominent myoclonus
Combined myoclonus with other movement disorders
Disorders that usually present with other phenotypes but can present with prominent myoclonus
Acquired
Metabolic (non-genetic)
Toxic- and drug-induced
Neurodegenerative disease
Nervous system lesions
Static
Progressive
Infections/post-infectious
Inflammatory/paraneoplastic (autoimmune)
Other medical (systemic) disorders
Functional neurological disorder
Idiopathic
Familial (genetic basis undetermined)
Sporadic
Physiological
Hypnic jerks
Fragmentary (sleep)
Exercise-induced
Hiccup
Anxiety-induced

[a] Rapid change of etiology examples prevents a reasonable current listing.

There was agreement for a Cortical-Subcortical myoclonus neurophysiology category during the Delphi rounds. This physiology reflects the abnormal bidirectional excessive neuronal activity widespread between cortical and subcortical circuits, producing the diffuse excitation. As such, this physiology is dissimilar from localized Cortical myoclonus mentioned above, and thus in a different neurophysiology category. Evidence from functional imaging and animal models exist [27,28]. Because this excitation over the sensorimotor cortex is simultaneously widespread, the myoclonus is commonly generalized. Thus, this physiology correlates with diffuse excitation of cortex, such as with

generalized spike, polyspike, spike and wave EEG discharges. Myoclonus associated with primary generalized epilepsy (e.g., Juvenile Myoclonic Epilepsy) is the most common example. Moreover, Juvenile Myoclonic Epilepsy is the most common etiology of myoclonus from an epidemiology standpoint [26].

Subcortical myoclonus reached agreement among the expert panel, but its exact definition remains elusive. Myoclonus-Dystonia is the classic and best-known example [29]. Its features reflect an absence of cortical myoclonus features, without a specific neurophysiological trait. In addition, surface EMG recruitment pattern is not indicative of another physiology or anatomical source. The features approved by the expert pattern are common, but they are nonspecific. More work is needed to define specific subcortical myoclonus physiology.

Brainstem myoclonus physiology is predominantly defined by a characterizable surface EMG recruitment pattern and absence of preceding EEG changes indicative of another physiology. The EMG recruitment pattern is typically characterized by earliest activation of the trapezius and sternocleidomastoid muscles, followed by simultaneous rostral to caudal as well as caudal to rostral progression, with latencies compatible with the distance of the muscles from brainstem origin [30]. In agreement with the literature, some critical differences between reticular myoclonus and startle myoclonus were recognized by the panel, aiding in their distinction [31]. Similarly to Brainstem myoclonus, Spinal (focal) myoclonus, and Propriospinal myoclonus are also defined by surface EMG recruitment pattern [32].

The term “focal myoclonus” was preferred by the expert panel rather than “segmental”. Segmental myoclonus has historically referred to both distribution and physiology, so its absence in the new classification may lead to some confusion. Peripheral myoclonus, although rare, did reach agreement among the expert panel. It is mostly dependent on surface EMG characteristics that are confined to a peripheral nerve distribution.

The neurophysiology features of functional jerks have undergone much study [33]. EEG features of Bereitshaftspotential and event-related desynchronization are useful for evidence of functional jerks. Other surface EMG features in the expert panel consensus are supportive of a functional jerk pattern. However, the absence of these features does not rule out a FND etiology.

4.3. Clinical myoclonus syndromes (derived from Axis Ia and Ib)

The syndromes that were proposed to the full panel were selected from classic myoclonus syndromes. A strong literature presence exists for all of them. All seven syndromes reached consensus agreement. Not all the Myoclonus Syndromes have the same degree of myoclonus presence. Moreover, each Myoclonus Syndrome will have variable clinical presentation among different cases, including prominence of myoclonus and other dyskinesias. The cortical myoclonus tremor syndrome has myoclonus that is relatively rhythmic. A full description of each Myoclonus Syndrome is available elsewhere [11,24,34–40]. This classification allows for these syndromes to have multiple etiologies.

4.4. Axis II. Etiology

Defining etiology in a patient with myoclonus is critical for both patient satisfaction and treatment options. Symptomatic treatment is best derived from the Clinical Neurophysiology classification category [6], while potential curative treatment is derived from etiology. Potential curative treatments are likely to increase in coming years. In addition, naming an etiology reassures the patient that the cause of their illness has been found. Specific examples of etiologies are not listed in the classification. This is because the list is long and ever changing. Myoclonus reviews have listed etiology examples under the previous classification [6,18]. All the etiology category items reached high consensus, except for Physiological myoclonus that had a slightly lower agreement compared to the other categories, and they are divided as follows: genetic, acquired, nervous system lesions, FND, idiopathic, physiological.

Genetic etiologies related to myoclonus have grown in recent years, and definition of such genetics etiologies will increase even further in future years. A robust genetic classification of myoclonus has been published [9], with the recommendation to allocate the genetic syndromes, according to their clinical presentation, into one of the following groups: *prominent myoclonus syndromes* (genetic disorders that present with prominent myoclonus in the majority of cases); *combined myoclonus syndromes* (genetic disorders that present with prominent myoclonus and another prominent movement disorder -eg, dystonia/ataxia-in the majority of cases); and *disorders that usually present with other phenotypes but can manifest as a prominent myoclonus syndrome* (genetic disorders that present with prominent myoclonus only in a minority of cases as part of the phenotypic spectrum of this disorder). Whole genome-wide sequencing is becoming an integral part of myoclonus etiology evaluation. Advances in genomics will allow multiple enhancements of this classification. As genetic-based treatments are increasingly developed, such treatment will directly benefit the corresponding myoclonus patients.

Acquired etiologies of myoclonus have historically been the most identified etiologies of myoclonus. An important reason to create a new classification of myoclonus was that many myoclonus etiologies that were previously thought to be “acquired” have genetic etiologies now defined instead. Moreover, the former “Essential myoclonus” category consists mostly of genetic etiology cases; many of these have Myoclonus-Dystonia syndrome, and thus “Essential myoclonus” has been less used in recent years. Nevertheless, metabolic, toxic, and drug-induced etiologies seem justly called acquired. However, neurodegenerative disorders, as currently understood have genetic influences. More elucidation of the neurodegenerative disorder subcategory etiologies is needed to classify these myoclonus cases more clearly.

Nervous system lesions comprise classic and as well as some rare etiologies of myoclonus. Hypoxic brain injury is one of the best described myoclonus etiologies, and it can be divided into acute and chronic (Lance-Adams) types [41]. Specific antibodies have increasingly been able to be defined in myoclonus cases, and they may provide a basis for treatment approach in autoimmune cases. Research has defined both genetic and acquired influences in antibody associated syndromes, so further consideration of category taxonomy may be justified. Systemic disorders of diverse types can cause myoclonus, and definition depends on the accurate diagnosis of the specific systemic disorder.

Functional jerks, although not conventionally considered myoclonus, are a critical consideration due to their variable phenomenology, uncertain pathophysiology, high incidence, and significant impact in clinical practice [11]. This classification incorporates FND etiology into the myoclonus classification. Functional jerks were consensually included by the expert panel, but definition as to the exact nature of the FND etiology is needed. Similarly to other FNDs, the diagnosis of functional jerks relies on the presence of positive clinical signs, for which neurological expertise is mandatory. However, functional jerks offer the advantage of being evaluated through specific neurophysiological tests that assess the planning and execution of voluntary movements at the cortical level, differentiating functional jerks from involuntary movements [33].

Idiopathic etiology, although necessary to include, is problematic. This is because these disorders may just represent “etiologies waiting to be discovered and defined.” The expert panel agreed to include Familial and Sporadic under this subcategory.

The Physiological myoclonus category brought the most controversy among the expert panel. However, the expert panel did find agreement on entities that can be thought to exist as normal myoclonus phenomena. The expert panel did comment that such normal manifestations of myoclonus can abnormally increase in frequency and/or amplitude. When this happens, it may be appropriate to designate this occurrence as an abnormal, rather than a normal occurrence.

5. Conclusion

This new Myoclonus Classification, as determined by the Delphi Method provides a beginning foundation for further refinement and advancements in the approach to myoclonus. It contains an axis approach, like what has been published for other movement disorders. This approach has numerous advantages. Notably, it facilitates clinicians in defining the appropriate myoclonus syndrome. By using Axis I (a & b) features, clinicians can identify the relevant symptoms and signs, framing the Clinical Myoclonus Syndrome as a starting point to determine the possible etiology. Moreover, the classification also incorporates Clinical Neurophysiology Features (Axis Ib) which are key to both the diagnosis of the myoclonus syndrome and/or etiology, as well as providing symptomatic treatment guidance. Axis II Etiology is condensed in terms of heading terms, in view of the long and constantly evolving known etiology list. However, it is more encompassing and modern compared with previous myoclonus classification. The new Genetic Etiology category takes advantage of genetic advances in myoclonus and will further grow. Finally, the incorporation of a "functional jerk" reference in both Axis Ib and Axis II reflects current comprehensive thinking of jerky movement disorders and underscores the urgent need to better define this common condition. Lastly, it is acknowledged that etiologies may have mixed influences (e.g. genetic and acquired), and we look to etiology research to guide more exact classification.

The potential of this new Myoclonus Classification will be realized by ongoing updates, but also important validation work on the listed features and their characteristics. This is particularly important for Axis Ib Clinical neurophysiology, where validation of sensitivity and specificity of features is generally lacking, despite their usefulness in clinical practice [42,43]. The addition of feature criteria (and validation) for items under both axes and Myoclonus Syndromes will be important to move myoclonus research forward. The relationship between myoclonus and seizures/epilepsy deserves attention [44]. The disparate thinking on this relationship, and with regard to ILAE classification, has produced confusion, so special consideration should be given to this in future iterations of this classification. Consideration should be given to a society or foundation sponsored database to contain current Axis II Etiology examples on an ongoing basis. We advocate for further research to include large cohorts of patients, recruited from diverse centers and with different myoclonus subtypes, to clarify the myoclonus criteria. It is hoped that this Myoclonus Classification, which reached consensus using the Delphi Method, produces a collaborative effort among myoclonus investigators to find better diagnostics and treatment for myoclonus patients.

CRediT authorship contribution statement

Anna Latorre: Writing – review & editing, Formal analysis. **S. van der Veen:** Writing – review & editing, Formal analysis. **Ashley Pena:** Writing – review & editing, Formal analysis. **Daniel Truong:** Writing – review & editing, Formal analysis. **Roberto Erro:** Writing – review & editing. **Steven Frucht:** Writing – review & editing. **Christos Ganos:** Writing – review & editing. **Mark Hallett:** Writing – review & editing. **Belen Perez-Duenas:** Writing – review & editing. **Malco Rossi:** Writing – review & editing. **Emmanuel Roze:** Writing – review & editing. **Marie Vidailhet:** Writing – review & editing. **Marina AJ. de Koning-Tijssen:** Writing – review & editing, Formal analysis, Conceptualization. **John N. Caviness:** Writing – original draft, Supervision, Project administration, Methodology, Investigation, Formal analysis, Data curation, Conceptualization.

Declaration of competing interest

The authors have no conflicts of interest to disclonse for this article.

Acknowledgements

This project was initiated and funded by the International Association of Parkinsonism and Related Disorders (IAPRD). The Christian Truong Foundation funded the in-person whole expert panel meeting at the World Congress on Parkinson's Disease and Related Disorders, in Memorial of Mr Norman Seiden for his contribution to research on Myoclonus.

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