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Myoclonus subtypes in tertiary referral center. *Cortical myoclonus and functional jerks are common*

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HIGHLIGHTS

- Specific clinical features are helpful in distinguishing myoclonic subtypes.
- More than 40% of myoclonus patients are diagnosed with a functional movement disorder.
- Electrophysiological testing is important to verify the clinical diagnosis.

ABSTRACT

Objective: To evaluate the accuracy of clinical phenotyping of myoclonus patients and to determine differentiating clinical characteristics between cortical (CM), subcortical (SCM), spinal (SM), peripheral (PM) myoclonus, and functional jerks (FJ).
Methods: Clinical notes for all patients with myoclonus over an 8-year period (2006–2014) were reviewed retrospectively. We used the conclusion of electrophysiological testing as definite diagnosis of myoclonus or FJ.
Results: 85 patients were identified suffering from CM (34%), SCM (11%), SM (6%), PM (2%), and 47% FJ. The clinical diagnosis of myoclonus was confirmed by electrophysiological testing in 74% and its subtype in 78% of cases. CM was characterized by an early age of onset, facial myoclonus, and provocation by action. Differentiating features of FJ were an abrupt onset, preceding contributing events and provocation by a supine position.
Conclusion: The majority of clinical myoclonic jerk cases were functional in our heterogeneous tertiary clinic cohort. CM was the main anatomical myoclonic subtype. Clinical diagnosis was accurate in the majority of cases, although electrophysiological testing was important to verify the clinical classification.
Significance: In patients with jerky movements a functional diagnosis should be considered. Determination of the myoclonic subtypes is important to initiate tailored treatment.

1. Introduction

Myoclonus is a hyperkinetic movement disorder caused by an abrupt muscle contraction (positive myoclonus) (Friedreich, 1881) or interruption of muscle activity (negative myoclonus) (Lance and Adams, 1963).

Myoclonic jerks can be classified according to origin, i.e. generated in the cortex, subcortical areas (including basal ganglia and brainstem), spinal cord or peripheral nerves. In addition, myoclonus can also be the result of a functional movement disorder; i.e. FJ. CM is considered most frequent (Caviness and Brown, 2004) but little is known about the epidemiology. Even less information is available on the sensitivity and specificity of clinical features in patients with myoclonus. Differentiating between subtypes of myoclonus is important, as each subtype can be linked to an

Abbreviations: BM, brainstem myoclonus; CM, cortical myoclonus; FJ, functional jerks; SCM, subcortical myoclonus; SM, spinal myoclonus; PM, peripheral myoclonus; PSM, propriospinal myoclonus; UMCG, University Medical Center Groningen.

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etiological differential diagnosis and guides treatment selection (Zutt et al., 2014, 2015).

Accurate clinical diagnosis of myoclonus remains challenging (van der Salm et al., 2013) and electrophysiological tests are often required to distinguish myoclonus from other hyperkinetic movement disorders and subsequently, to define its anatomical subtype. Video-polymyography is the electrophysiological test in clinical practice to make the diagnosis of a jerky movement based on burst duration and muscle recruitment. (Shibasaki et al., 1978) Additional, more sophisticated testing can be performed such as EEG–EMG back-averaging (Shibasaki et al., 1978) or coherence analysis, (Grosse et al., 2002; Brown et al., 1999) to detect a cortical origin in CM or a bereitschaftspotential in FJ (Shibasaki and Hallett, 2006). Furthermore, somatosensory evoked potential (SEP) can be useful to detect a giant potential pointing towards cortical hyperexcitability (Shibasaki et al., 1990).

The aim of this study is to evaluate the accuracy of clinical phenotyping in a heterogeneous cohort of myoclonus patients and to determine differentiating clinical characteristics.

2. Methods

A retrospective analysis was performed of patients who visited our tertiary referral center between February 2006 and May 2014 and in whom video-polymyography was part of the diagnostic work-up. Patients were identified with the use of an electronic database from the department of Clinical Neurophysiology at the UMCG, the Netherlands. The database contains all electrophysiological test results since 2006. Registrations were analysed by two experienced clinical neurophysiologists (JWE and JvdH). The Ethical Board of the University Medical Center Groningen (UMCG) approved the study (Number M14.157933). We selected all cases with myoclonus as referring clinical diagnosis for video-polymyography. The definite diagnosis used in our study was the diagnosis based on electrophysiological testing.

Electrophysiological tests included continuous recordings of surface EMG (maximum of nine channels) and video in all cases. In a subset of patients EMG–EEG back-averaging, coherence analysis and/or SEP was applied.

EMG was recorded with Ag/AgCl pairs of surface electrodes placed at affected muscles. Myoclonus was measured during rest and action, action was defined by posture and specific tasks (finger to nose and knee to heel test).

The EEG was recorded with Ag/AgCl surface electrodes placed at the scalp according to the 10–20 International System and acquired by a computerized system (All data was recorded with BrainRT software (OSG BVBA, Rumst, Belgium)) using a sample frequency of 1000 Hz.

The electrophysiological characteristics of myoclonus and its subtypes were applied as described in literature and used in our laboratory to draw conclusions (Table 1). Besides the techniques of back-averaging and coherence analysis, all EEGs were analysed for epileptiform abnormalities.

Table 1
Electrophysiological criteria of myoclonus and its subtypes used in this study.

Myoclonus and its subtypes		Electrophysiological criteria based on polymyography	Importance of criteria
Myoclonus		Abrupt muscle contraction or interruption of tonic muscle activity	Required
		Synchronous contraction of agonists and antagonists muscles	Supportive
Cortical		Burst duration of positive myoclonus <100 ms	Required
		Multifocal/focal distribution	Supportive
		Presence of negative myoclonus	Supportive
		Positive cortical spike back-averaging	Diagnostic
		(more reliable if >100 jerks, not performed if <25 jerks)	
		Presence of a "time-locked" biphasic potential >2SD above baseline on the contralateral motor cortex preceding the jerks seen on the EMG according to the conduction time of corticospinal pathways (15–25 ms for jerks in the arms and by ±40 ms for jerks in the legs)	
		Positive cortico-muscular coherence	Diagnostic
		(frequencies >10 Hz–60 Hz)	
		Occurrence of significant cortico-muscular coherence in the alpha and beta band with a phase difference consistent with a cortical generator (i.e. cortex leads muscle) in coherence analysis.	
		Presence Giant SEP	Diagnostic
		The P27 and N35 peaks had large amplitudes above 5uV and had a suitable shape	
Subcortical	Brainstem	Burst duration >100 ms	Supportive
		Simultaneous rostral and caudal muscle activation at brainstem level	Supportive
	Myoclonus–Dystonia	Burst duration >100 ms	Supportive
		Do not meet criteria other categories	
Spinal	Segmental	Burst duration >100 ms	Supportive
		Distribution according to one or two contiguous spinal segments	Required
		Rhythmic (1–2/min–240/min)	Supportive
	Propriospinal	Burst duration >100 ms	Supportive
		Initiation in mid thoracic segments followed by rostral and caudal activation	Required
		Propagation with slow velocity (5–15 m/s) in cord	Required
Peripheral		Burst duration <50 ms	Required
		Large MUAPs	Required
		Minipolymyoclonus or fasciculations/myokymia	Required
		Accompanied by weakness/atrophy	Supportive
Functional jerks		Variable muscle recruitment	Supportive
		Variable burst duration	Supportive
		Burst duration >100 ms	Supportive
		Distractibility and or/ entrainment (rhythmical myoclonus)	Supportive
		Bereitschaftspotential	Diagnostic (ex. tics)
		(performed if >40 jerks, less than 1 every 5 s)	
		Presence of a clear slow negative electrical shift over the central cortical areas that increased over time with amplitudes of at least 5 μV 1–2 s before movement onset	