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Movement disorders

To jerk or not to jerk: A clinical pathophysiology of myoclonus

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ABSTRACT

Myoclonus is a sudden brief (20–250 ms) contraction (positive myoclonus), or a brief and sudden cessation of tonic muscle (negative myoclonus) inducing a simple jerky movement of body part. Myoclonus could have different origins in almost every part of the nervous system, from the cortex to the peripheral nerve, sharing a large panel of etiologies. It is regarded as the paradigmatic movement disorder causing jerks, although not the sole. This paper aims to depict the clinical and neurophysiological characteristics of myoclonus. It shows how neurophysiological investigations including surface polymyography and methods exploring cortical excitability, namely conventional EEG, EEG – jerk-locked back-averaging, somatosensory evoked potentials and C-reflex studies are required to define the generator of myoclonus in the central nervous system and clearly classify myoclonus as cortical, corticothalamic, subcortical – resulting from lesions or dysfunctions of basal ganglia/reticular system – or spinal. This paper also enlightens other movement disorders that may mimic myoclonus appearances, including psychogenic jerks, simple motor tics, spasms and startle syndromes. Finally, it raises few unresolved questions regarding the propriospinal myoclonus or peripheral myoclonus entities, the role of the cerebellum in myoclonic diseases and the relationship between cortical and epileptic myoclonus.

As well as being one of the most common involuntary movements, myoclonus should also be regarded as being both the simplest and yet complex movement disorder. Myoclonus is simple, as it may be summarized as a brief and single movement with no specific pattern of motor activation. It causes a jerky, shock-like movement, and disappears as soon as it happens. Thus, it follows that myoclonus represents a movement disorder that can be difficult to observe and

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characterize due to its brevity. When isolated, only the passive part of the movement, bringing the involved limb back to its previous position, is witnessed. On the other hand, complexity is also present, as myoclonus may originate in almost any part of the nervous system, from the cortex to the peripheral nerves, and has a large panel of etiologies. In addition, it can easily be confused with many other movement disorders, as it cannot be assumed that everything that jerks is myoclonus. Thus, the present paper aims to describe the clinical and neurophysiological characteristics of myoclonus, and to clarify the other movement disorders that may mimic its appearances.

1. Jerks and pseudo-jerks: clinical definitions and boundaries

1.1. *Phenomenology of jerks*

As a quick, sharp and sudden movement, jerks appear to those looking at the patient as a brief change in posture or interruption of an ongoing action, making it imprecise and/or causing carried objects to be inadvertently dropped. Typically, jerks present as two components: the first part of the movement, which is only assumed to be pathological, corresponds to either a brisk muscle contraction (positive) or sharp decrease in ongoing muscle tone (negative); the second part is passive, causing the involved segment to return to its previous position or to rest. As jerks are movements, they are clearly and definitively distinguished from fasciculation, or myokymia, muscle contractions that are not associated with any visible movement except those caused by small muscle groups in, for example, the eyelids and digits. Although it is a simple, one-shock movement, myoclonus may be difficult to diagnose, as evidenced by a study involving movement disorder specialists [1]. This is clearly due to its brevity, as already mentioned. Clinically, every abnormal movement lasting 100 ms may appear to be myoclonic, although other characteristics may help in deciphering them. Simple motor tics, spasms, choreic movements, startle reactions or even shivering could be misdiagnosed as myoclonus. Moreover, some movements are defined as myoclonic, although their duration may be far longer than expected when recordings are available: psychogenic myoclonus is a good example of a sustained and prolonged muscle contraction, causing jerks (Fig. 1).

Nevertheless, should every muscle contraction that causes jerks be called 'myoclonus' until neurophysiological investigation determines its distinctive features? Clearly not: clinicians need to be aware of this discrepancy and be careful to use the term 'myoclonus' only when sudden, brief, involuntary jerks of a segment are evidenced and, if necessary, with appropriate neurophysiological tools. Otherwise, use of the term 'jerk' is convenient; accordingly, the term 'psychogenic myoclonus' should be abandoned in favor of 'psychogenic jerks'.

1.2. *Myoclonus*

As already pointed out, myoclonus is characterized by a sudden, brief and sometimes repetitive muscle contraction of body parts (positive myoclonus), or a sudden brief cessation of tonic muscle activation, followed by rapid recovery of tonus (negative myoclonus). While it is regarded as the paradigmatic

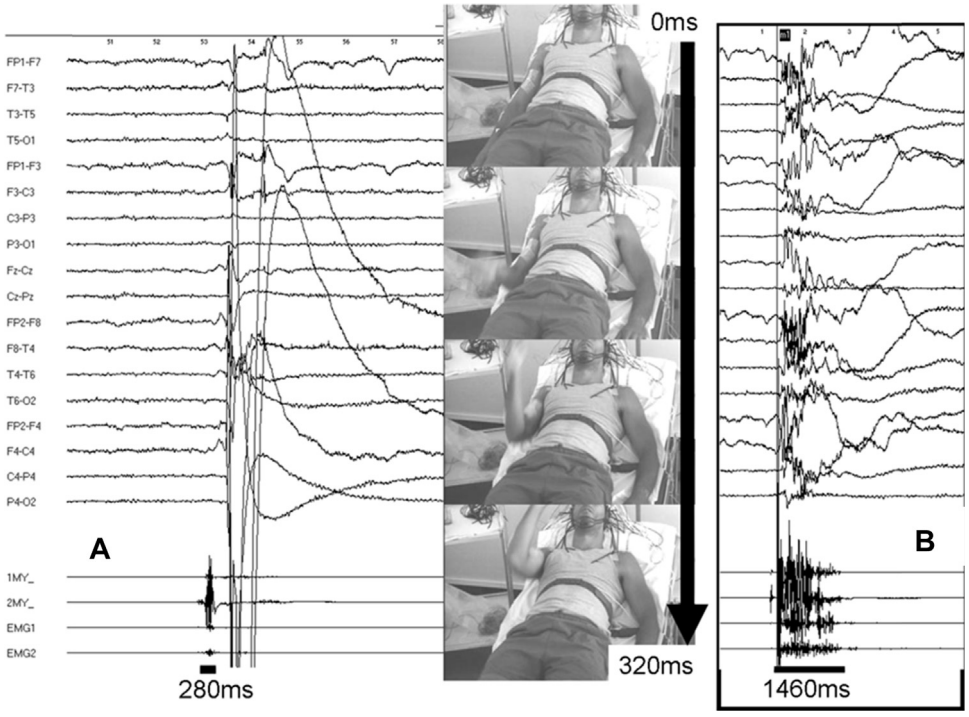

Fig. 1 – Psychogenic jerks recorded simultaneously by EEG and EMG. A. Jerk involving the right arm is a somewhat complex movement (shown in successive snapshots), while EMG discharges are brisk (lower four traces), despite lasting 280 ms, and EEG changes are movement artifacts that follow muscle activation. B. Another event, this time lasting > 1 s (see the last four traces).