Neurology Asia 2022; 27(3) : 791 – 794

Practical utility of electrophysiologic tests in diagnosing functional myoclonus: An illustrative case

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Abstract

We describe the case of a 22-year-old female who was diagnosed with a functional movement disorder. The diagnostic processes she underwent elegantly illustrates the practical utility and limitations of multi-channel electromyography/polymyography and EEG back-averaging in correlating with and demonstrating the clinical features of functional movement disorders, providing a vital avenue through which a clinician may find useful when navigating the imposing diagnostic minefield of functional movement disorders.

Keywords: functional movement disorder; functional myoclonus; movement disorder; EMG; neurodiagnostic test

CASE REPORT

A 22-year-old university student presented with involuntary jerks of the left upper limb, and referred to our facility with the diagnosis of partial seizures. These intermittent movements began abruptly 2 months before, were more prominent when stressed, but were unaltered by visual, cutaneous, and auditory stimuli, and were absent when asleep (video 1). She had no medical and psychiatric history, but was described by her parents to easily anxious. She displayed arrhythmic (5-10 jerks per minute), sudden, brief jerks over the left shoulder and elbow, resulting in the forward flexion and adduction of the left shoulder, with concomitant extension of the left elbow. These movements were entrainable (video 2). There were no significant abnormalities of the cranial nerves. Examination of the limbs demonstrated normal tone, reflexes, sensorimotor, and cerebellar functions. Slit-lamp examination was negative for Kayser-Fleischer rings. Ancillary tests inclusive of serum caeruloplasmin and urinary copper levels were negative for thyroid, metabolic, and electrolyte derangements. Magnetic resonance imaging (MRI) of her brain and cervical spine were unremarkable. A 30-minute electroencephalogram (EEG) returned normal, despite frequent occurrence of the movements during the study.

In view of the atypical characteristics of the involuntary movements, she underwent a multi-channel electromyography (EMG)/ polymyography, during which recordings were obtained from the deltoid, extensor carpi radialis (ECR), and tibialis anterior (TA) muscles on the left, and the first dorsal interosseous (FDI) muscle on the right. When at rest, multiple bursts of EMG activity were recorded over the left deltoid and ECR muscles at a frequency of 0.5Hz (Figure 1A). They were synchronous, but had variable amplitudes and durations (250 – 600ms). While performing contralateral ballistic movements (hitting the table with her right hand), EMG activity was attenuated for 1.2s (Figure 1B). The amplitudes and frequencies of the EMG bursts were reduced whilst performing cognitive tasks (serial-7 subtractions), and entrainment of the EMG bursts was demonstrated whilst tapping the contralateral hand at 1, 2 and 3 Hz (Figures 1C-E). Suggestibility was observed with a vibrating 128Hz tuning fork pressed gently against the patient's left ulnar styloid, with the patient being told prior that such an act may either exacerbate or attenuate the involuntary movements (Figure 1F).

In search for the Bereitschaftpotential (BP), EEG back-averaging (scalp electrodes at C3, C4, Cz, A1 and A2) was performed over the left deltoid muscle. In our centre, at least 50 jerks or epochs are required for EEG back-averaging analysis, during which EEG potentials occurring between 20-40ms before the onset of EMG

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Date of Submission: 28 December 2021; Date of Acceptance: 11 April 2022

https://doi.org/10.54029/2022iyp

signals are deemed representative of cortical myoclonus. This technique can be limited by the frequency of the jerks: jerks that are infrequent may not generate enough trials for analysis, while jerks which are excessively frequent can result in an unstable baseline which renders back-averaging analysis difficult. The latter limitation was evident in our patient, during the excessive jerks (>200 episodes) observed over 30 minutes resulted in an unstable EEG baseline, hindering the detection of pre-movement BP (Figure 1F). For the same reason, recording of event-related desynchronization (ERD) could not be performed. Despite these limitations, multi-channel EMG/polymyography clearly demonstrated variability, distractibility, entrainment, and suggestibility of her movements, consistent with the diagnosis of functional myoclonus (FM). The diagnosis and investigative findings were shared with the patient, after which the patient reported spontaneous improvement of her symptoms. Outpatient psychiatric consultations were subsequently planned on discharge. Her involuntary movements have improved significantly when reviewed a month later, with infrequent subjective sensation of 'tightening' over the left shoulder, but without discernible movements.

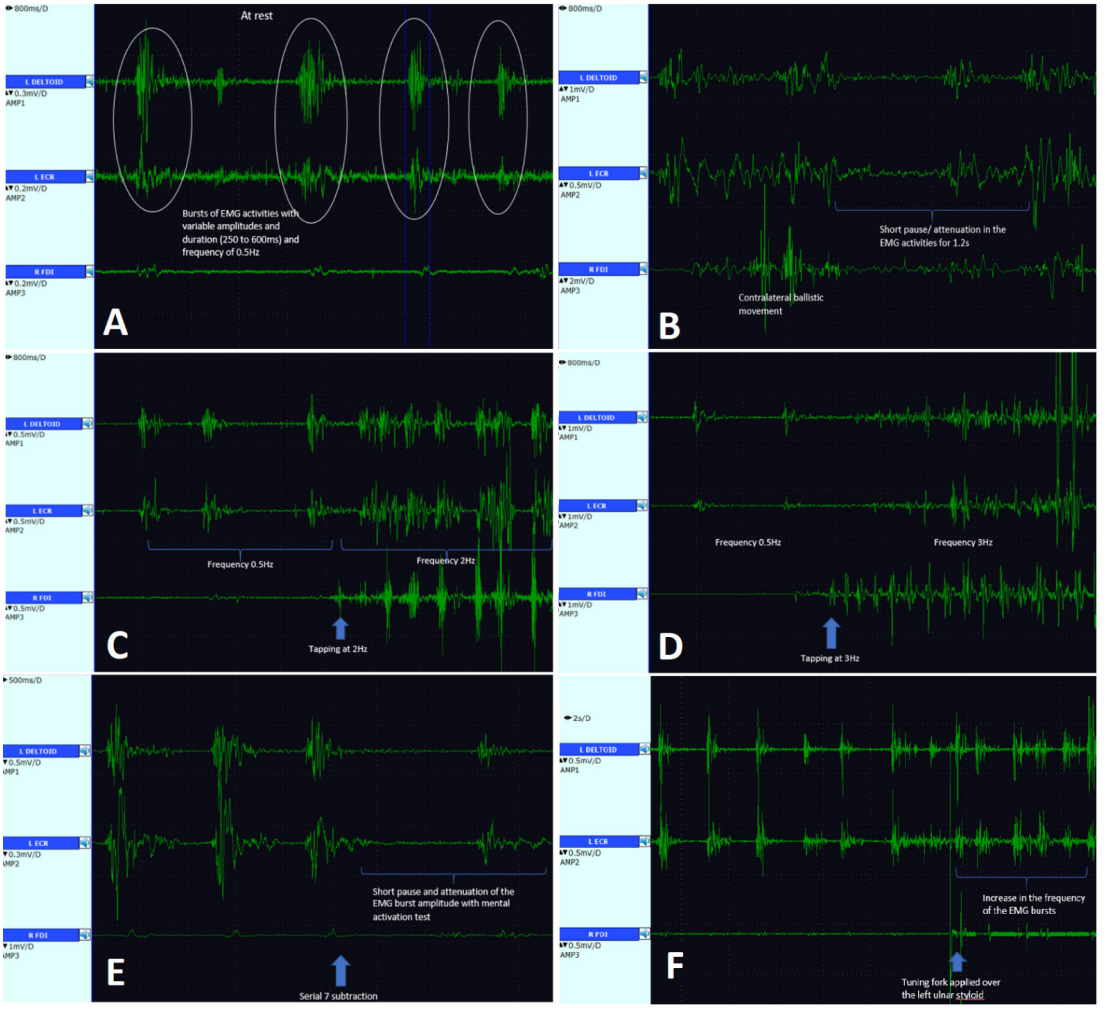

Figure 1. Multi-channel polymyography. (A) At rest, multiple EMG bursts of varying amplitudes and durations (250-600ms) were synchronously recorded over the left deltoid and left extensor carpi radialis (ECR), at a frequency of approximately 5 EMG bursts per 10-second period. (B) When performing ballistic movements with the contralateral arm, attenuation of EMG bursts was observed for 1.2s. (C, D) Tapping of the contralateral hand at 2 and 3Hz resulted in the entrainment of EMG activities over the left deltoid and ECR. (E) Mental activation whilst performing cognitive tasks reduced the amplitudes and frequency of the EMG bursts. (F) Application of a vibrating 128Hz tuning fork on the ipsilateral ulnar styloid increased the frequency of EMG bursts (suggestibility).

DISCUSSION

FM has been reported in 30-50% of patients presenting with myoclonus, and can be difficult to discern from organic myoclonus.[1] Earlier authors have described the characteristic profile of a young female of at least average intelligence, similar to our patient's.[2] Typical clinical traits hinting at the movements' non-organic origins include the abrupt onset of symptoms, the varying amplitudes and frequencies, and them being entrainable, suggestible, and distractible.[3] Although entrainment is a commonly-described feature of functional tremors, referring to the shifts in tremor frequency so as to match concomitant yet separate competing rhythmic movements of another body part, a similar phenomenon may be observed electromyographically and clinically in patients with functional myoclonus, as in the case of our patient, during which the frequency of the myoclonic movements over her left upper limb shifted to match the tapping movements of the right-sided fingers, and the rhythmic clenching of the right first (Figure 1 and Video 2), providing another potential avenue for differentiating functional and organic myoclonus.

While there is no gold-standard confirmatory test, assessments such as surface EMG, EEG, and EEG back-averaging, hold significant value in the objective evaluation of patients with FM. Typical surface EMG features include variable and inconsistent order of muscle recruitment, variable burst duration of more than 100ms, distractibility when performing ballistic movements with the contralateral limb, and the presence of suggestibility and entrainment.[4] Demonstration of BP on EEG back-averaging, and ERD on time-frequency decomposition have recently been reported in recent literature to be important biomarkers supportive of functional myoclonus.[4] BP is a negative, slow-rising potential that precedes voluntary movement by 1.5 to 2.5s, and is purported to reflect movement preparation at the supplementary motor area, and is thus most prominent at the Cz electrode. The presence of BPs strongly supports the diagnosis of FM, but its absence does not exclude the diagnosis of functional movement disorders.[5] ERDs describe the suppression of beta or mu rhythms over the cortical motor areas during preparation of volitional movements, and are best observed 1.5 to 2s before EMG onset at the C3 and C4 electrodes.[4] Their presence also support the diagnosis of functional movement disorders, conferring an additional diagnostic gain of 71% when combined with the qualitative and quantitative demonstration of BPs.[6] In our patient's case, her frequent movements resulted in an unsteady EEG baseline, hindering the demonstration of BP and ERDs, reflecting their practical limitations in the assessment of functional movement disorders.

In conclusion, our report elegantly illustrates the practical utility of electrodiagnostic tests when assessing a patient with clinical features suggestive of functional movement disorders. Characteristics unique for underlying non-organic processes are demonstrated electrophysiologically using multichannel EMG/polymyography: multiple EMG bursts of varying amplitudes and durations, prolonged bursts of EMG (>70ms), distractibility, entrainment of EMG activities, and suggestibility.[7] Despite our inability to detect BPs and ERDs due to an unstable EEG baseline from the patient's frequent and vigorous movements, her polymyographic findings were sufficient in differentiating functional from organic myoclonus through the demonstration of the aforementioned findings, although the diagnostic sensitivity and specificity based only on the performance of multichannel EMG/polymyography are currently undescribed in literature and await future elucidation. Regardless, clinical and electrophysiology findings remain strongly consistent with functional myoclonus, permitting the initiation of apposite treatment measures, whilst avoiding unnecessary investigations, and inappropriate therapies.

DISCLOSURE

Financial support: None

Conflict of interest: None

Video 1: The patient displayed sudden, brief jerks of the left shoulder and elbow with varying frequencies and amplitudes, resulting in the arrhythmic forward flexion and adduction of the left shoulder, with concomitant extension of the left elbow. https://neurology-asia.org/content/27/3/neuroasia-2022-27(3)-791-v1.mp4

Video 2: The involuntary movement was entrainable. When asked to rhythmically grasp with the contralateral (right) hand, the involuntary movement was entrained to the frequency similar to the voluntary hand movement on the right. https://neurology-asia.org/content/27/3/neuroasia-2022-27(3)-791-v2.mp4

REFERENCES

1. Zutt R, Elting JW, van der Hoeven JH, Lange F, Tijssen MAJ. Myoclonus subtypes in tertiary referral center. Cortical myoclonus and functional jerks are common. *Clin Neurophysiol* 2017;128(1):253-9. doi: 10.1016/j.clinph.2016.10.093.
2. Ford B, Williams DT, Fahn S. Treatment of psychogenic movement disorders. In: Kurlan R, Ed: Treatment of movement disorders. J.B. Lippincott Company, 1995: 475-85.
3. Miyasaki JM, Sa DS, Galvez-Jimenez N, Lang AE. Psychogenic movement disorders. *Can J Neurol Sci* 2003;30 (Suppl 1):S94-100. doi: 10.1017/s0317167100003292.
4. Chen KS, Chen R. Principles of Electrophysiological Assessments for Movement Disorders. *J Mov Disord* 2020;13(1):27-38. doi: 10.14802/jmd.19064.
5. van der Salm SM, Tijssen MA, Koelman JH, van Rootselaar AF. The bereitschaftspotential in jerky movement disorders. *J Neurol Neurosurg Psychiatry* 2012;83(12):1162-7. doi: 10.1136/jnnp-2012-303081.
6. Beudel M, Zutt R, Meppelink AM, *et al*. Improving neurophysiological biomarkers for functional myoclonic movements. *Parkinsonism Relat Disord* 2018;51:3-8. doi: 10.1016/j.parkreldis.2018.03.029.
7. Eberhardt O, Topka H. Myoclonic disorders. *Brain Sci* 2017;7(8):103. doi: 10.3390/brainsci7080103.