Hands-on Course 8

MDS-ES/EAN: Neurophysiological study of tremor - Level 1

Orthostatic tremor, cortical tremor, dystonic tremor and psychogenic tremor: Electro-clinical semiology

Emmanuelle Apartis
Paris, France

Email: emmanuelle.apartis@sat.aphp.fr
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The author has no conflict of interest in relation to this manuscript.

Key learning objectives

The key learning objectives of the session are to identify the clinical and electrophysiological core signs of uncommon tremors: orthostatic tremor, cortical tremor, dystonic tremor and psychogenic tremor, in order to assess an appropriate positive diagnosis and to differentiate them from other movement disorders, thus leading to adequate care.

Essential knowledge

1. Orthostatic tremor

Primary orthostatic tremor (OT) is a rare disorder that remains under-diagnosed. OT arises from a central generator located in the posterior fossa with pathological ponto-cerebello-thalamo-cortical activations. OT predominantly affects women over the age of 50. The cardinal symptoms, which are highly suggestive of the diagnosis, are feelings of instability, leg tremor in the standing position, improvement with walking and disappearance while sitting. OT remains mis-diagnosed as vestibular or cerebellar disorder or psychogenic balance disorder. Yet, the diagnosis of OT can be very easily assessed by the recording of the typical tremor activity in the 13-18 Hz range in the legs while standing. This char-
acteristic electro-clinical picture differs from those of two looking alike conditions - tremor in orthostatism and orthostatic myoclonus - frequently related to neurodegenerative disorders and parkinsonian syndromes.

**Signs of OT**²,³:
- The symptoms appear a few seconds to a few minutes after standing. They disappear when walking and when sitting at rest. They are improved when supporting against a wall or a table.
- The major symptom of OT is the feeling of instability, even more than tremor in the legs. No falls but fear of falling.
- At clinical examination, the tremor is hardly visible, and may be palpable. The walk and the neurological examination are otherwise normal.
- **Neurophysiology:** Very regular tremor of high frequency (13-14 Hz) evidenced in both legs when standing, disappearing at rest and composed of very short synchronous EMG bursts (20-40 ms). The higher the tremor amplitude on polymyography, the higher the disability, both increasing with the duration of standing. Extension of OT to the trunk and upper limb muscles when standing continues. High inter-limb and inter-muscular coherence at tremor frequency.

**Signs of tremor in orthostatism:**
- Tremor is the major complaint, instability can be mild or absent
- Tremor involves one or two legs when standing
- Tremor is visible at clinical examination
- Tremor can persist when walking or at rest when sitting
- Neurological examination may be abnormal - rigidity, akinesia, cerebellar signs -

- Associated conditions: idiopathic Parkinson’s disease, hydrocephalus, spino-cerebellar ataxia, brainstem or cerebellar focal lesions, advanced essential tremor

- **Neurophysiology:** Regular tremor at slower frequency (3-12 Hz) than OT, with longer burst duration (70-130 ms) when standing. Associated proximal or distal rest tremor in the lower limb at the same frequency. Possibly, re-emergent parkinsonian tremor of the lower limbs, with posture or when standing.

**Signs of orthostatic myoclonus 4:**

- Similar symptoms than OT, with frequent feeling of instability, sometimes feeling of “shaky-legs”, more than tremor.

- Persistence at walk, fear of falling and falls

- Neurological examination is frequently abnormal - rigidity, akinesia, cerebellar signs, cognitive signs

- Associated conditions: mainly idiopathic Parkinson’s disease, also Lewy body dementia, Alzheimer’s disease, multiple system atrophy, progressive supra-nuclear palsy

- **Neurophysiology:** Irregular myoclonic bursts, in the lower limbs, asynchronous, bursts duration (40 to 100 ms), frequency range (6-11 per seconds), occurring predominantly when standing, sometimes persisting at walk or with outstretched legs. No inter-limb coherence at a given frequency. The generator may be either cortical or sub-cortical, depending on the underlying disease.
2. Cortical tremor

Familial Cortical Myoclonic Tremor (FCMT) is a rare condition with an action tremor to be distinguished from essential tremor, considering the different therapeutic issues for both diseases. FCMT is an autosomal dominant disease characterized by a slow or non-progressive adult-onset, involuntary repetitive myoclonic jerks of the distal upper limbs, looking like an irregular tremor enhanced by outstretched posture and action. Progressive myoclonic epilepsy is excluded by the absence of ataxia and mental retardation and by the benign course of the disease. In this background, the association of an upper limb postural tremor with generalized epilepsy or a familial history of epilepsy, and its resistance to beta-blockers should question this diagnosis, considering the good response of FCMT to anti-epileptic drugs. Focal cortical tremor may also occur with congenital or acquired lesions of the cortical fronto-central area. Electrophysiological findings in FCMT and focal cortical tremor fulfil criteria for cortical myoclonus.

**Neurophysiology of FCMT:**

- Irregular postural tremor recorded on the accelerometer (5-10 per second)
- Brief polymyographic bursts lasting less than 50 ms in the distal muscles
- Predominant at postural maintenance, absent at rest, not enhanced with intention
- Neurophysiological signs of cortical hyperexcitability and evidence for a cortical generator of the myoclonus:
3. Dystonic tremor

Dystonic tremor is classically defined as tremor in a body part that is affected by dystonia. Tremor is a common feature in patients with adult-onset focal dystonia and may involve several different body parts - i.e. cervical dystonic tremor, upper limb dystonic tremor, truncal tremor -. Dystonic features may be severe but also mild. Dystonic tremor may also be isolated, without overt clinical signs of dystonia. Dystonic tremor should be distinguished from tremor that is associated with dystonia, i.e., that occurs in a body part not affected by dystonia. In addition to its diagnosis value, neurophysiological recording can be helpful in assessing the more active muscles and guiding the botulinum toxin injections when dystonic tremor is highly focal;

**Classical signs of dystonic tremor:**

- Typically, postural or action tremor, rarely manifesting at rest
- Suppression by sensory tricks ("geste antagoniste")
- Clinical dystonic features in the same body part
  o Abnormal posture
  o Motor overflow defined as unintentional muscle contraction, affecting the electivity of a voluntary movement
- Tremor elicited by a specific task or a selective position of the body part
- Neurophysiology:
  o Irregular tremor, with variable duration (50-300 ms) and amplitude of consecutive bursts
  o Variable frequencies (3-12 Hz)
  o Sustained contractions, spasms or myoclonus associated to tremor
  o Co-contraction of agonistic and antagonistic muscles (i.e. wrist flexors and wrist extensors)

Special features of cervical dystonic tremor:
- In addition to the criteria mentioned above,
- Recording of the neck muscles allows comparing the activities of the coupled muscles that rotate the head to the left - left splenius and right sterno-cleido-mastoideus - to those that rotate the head to the right - right splenius and left sterno-cleido-mastoideus -.
- An asymmetrical location of the tremor in rotator muscles or elective involvement of one couple argues for dystonic cervical tremor and against essential cervical tremor.
Signs of pure writing tremor:
- Regular or irregular tremor occurring specifically with writing
- Disappearance of tremor during postural maintenance, during non-specific actions and other tasks than writing
- Mirror writing tremor elicited at rest by writing with the contralateral hand
- Lack of overt dystonic features in the affected hand

3. Psychogenic tremor
Psychogenic tremor frequently involves the upper limbs, but may affect every part in the body including neck, legs and trunk. It may occur under different conditions, rest, posture, action, intention, and even with orthostatism. After evidence of the clinical clues suggestive of psychogenic tremor, neurophysiological tools are necessary to assess the diagnosis by revealing objective positive signs in comparison to the classical characteristics of tremors of organic origin. Psychogenic tremor should be distinguished chiefly from pyramidal clonus and organic tremors such as enhanced physiologic tremor, parkinsonian tremor, dystonic tremor, and odd tremors. In some cases, these physiological data combined with the patient insight during the tests, are beneficial in persuading the patients of the accuracy of the diagnosis and of the potential reversibility of their symptoms.

Clinical clues for psychogenic tremor:
- Tremor with a sudden onset
- Tremor with a variable course, periods of complete normality
- Distractibility
- Fluctuations of intensity, or sudden shifts of the localization or the axis of tremor, particularly if they are provoked by external manipulations (i.e., passive immobilization of the hand inducing the shifting of tremor to the shoulder or to another body part)

- Inconsistencies

**Neurophysiological signs of psychogenic tremor**\(^{10,11}\).

Among criteria listed by R Helmhich (part 1 of the HoC: Neurophysiological study of tremor: how to do it in clinical practice), the strongest are related to the tremor frequency analysis and manipulation by the examiner:

- The spontaneous variability of tremor frequency

- The frequency driving induced by contralateral rhythmic tasks,

By contrast, organic tremors have a stable frequency that cannot be driven to another one. Tips and tricks adapted to the body part affected by the psychogenic tremor, and pitfalls in tremor variability assessment and interpretation will be highlighted, with a special attention paid to the differential diagnosis of dystonic tremor.
References and suggested reading:


