

Preface

Who could claim they've never experienced trembling at least some point in their lifetime? Indeed, everybody has perceived some tremor some time in life. For instance, the postural tremor observed when using the pointer during one's first lecture... In fact, a slight, rapid, and postural physiological tremor is permanently present, as shown when placing a piece of paper on the hand when the upper limb is extended. Tremor is thus a very common phenomenon when one looks around carefully.

Medical doctors are aware that the observation of an unexpected tremor in a given subject can result either from diseases of non-neurological origin (hyperthyroidism, drug treatments, etc.) or from an affliction of the nervous system. The most known of the latter are Parkinson's disease (PD), although the classical rest tremor is not always present, and Essential Tremor (ET), characterized by a postural/kinetic tremor and whose prevalence is six times higher than the prevalence of PD. By contrast with what is usually believed, the diagnosis of tremor is far from being easy. When its intensity is minimal, it is often difficult to distinguish ET from physiological tremor (for instance in subjects pertaining to ET families). When tremor is severe, its large amplitude may wrongly orient towards other disorders such as repetitive movements observed in PD treated by levodopa. Even more difficult, a tremor can mimic rhythmic myoclonus as seen in dystonic patients or depressed patients overtreated with various medications (lithium, etc.). In all these difficult cases, a polygraphic recording of tremor by an experienced clinical physiologist can be very helpful. Although tremor is a remarkable sign to perform an accurate diagnosis during daily clinical practice, it is often overlooked. There are many biases and drawbacks, for example, the amalgam between tremor and senility, or the wrong idea that tremor is often associated with alcoholism (I recall the case of a waiter who was considered alcoholic, although he had in fact a severe ET that he tried to improve by drinking several glasses of wine before serving the guests).

When possible, the treatment of tremor depends primarily on the treatment of the condition causing the tremor (hyperthyroidism for example). However, this is exceptionally the case in most neurological disorders. In PD patients, tremor is rarely disabling, except in few forms of the diseases, and it is usually largely attenuated by the administration of levodopa provided the doses of the administrated

amino acid are high enough, which is not always possible. In most severe cases of parkinsonian tremor, the neurosurgical approach (high-frequency stimulation of the thalamus, thalamotomy) can be extremely helpful. Bilateral stimulation of the subthalamic nucleus has not only the advantage of abolishing the contralateral rest tremor, but also to markedly improve the most disabling akineto-rigid syndrome. In patients with ET, one has to clearly distinguish two clinical situations. In benign cases, when the symptom starts to bother the patient in his daily life, the administration of drugs such as beta-blockers or primidone is required provided there is no contraindication. In patients with severe ET, i.e., when the amplitude of the tremor is interfering with the most elementary gestures of daily life, the medical treatment becomes ineffective, and the best option, when acceptable, is neurosurgery. High-frequency stimulation of the Vim of the thalamus is the treatment of choice, but the destructive approach (thalamotomy) can be considered in fragile, aged, or non-cooperative patients.

These comments are obviously oversimplified and will be extensively developed in the book. Whether benign, needing a simple follow-up, or severe, implying a sophisticated treatment, the clinical aspects of the various types of tremor need to be perfectly identified as it is the only way to ensure an optimal management of patients. To become a good semiologist in the field of tremor is necessary, but it is not sufficient! One needs also to be an excellent physiologist. Nowadays, as the mechanisms of the different categories of tremor start to be understood, this is now possible. In this field, the practitioner needs to keep in mind three main ideas (1) tremor can result from the dysfunction of all parts of the nervous system: the cerebral cortex (rhythmic myoclonus), the basal ganglia (PD rest tremor), the brain stem (Holmes tremor), the cerebellum (ET), the spinal cord (in fact segmental myoclonus), and peripheral nerves (Charcot–Marie–Tooth diseases); (2) several groups of neurons are tremorogenic, giving rise to various rhythmic oscillations in the brain (12–14 Hz in the olive; 3–6 Hz in the basal ganglia); (3) there is no unique “center of tremor” explaining the rhythm, the speed, and the amplitude of tremor, which also depends on the tension of the implicated muscles. In most cases, even if the lesions are selectively confined in the brain, tremor results from the dysfunction of various neuronal circuits, thereby giving rise to different symptomatic aspects of tremor.

Why this new book then? The reason is that it provides an extensive state of the art of the available clinical and scientific knowledge related to tremor. The numerous chapters, provided by the best experts in the field, will allow the clinicians to base their diagnosis, prognosis, and treatment on an updated clinical and pathophysiological basis, with bridges between fundamental aspects and clinical approaches.

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