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Myoclonus

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Continuing Education Activity

Myoclonus is defined as rapid, brief, jerky, or shock-like movements involving muscle or group of muscles. Among all hyperkinetic movement disorders, Myoclonus is considered to be the most rapid and brief. When caused by sudden muscle contraction, it is known as "positive myoclonus," while a brief loss of muscular tone results in "negative myoclonus" as in asterixis. Myoclonus is classified in different ways according to its physiology, anatomical site of origin, and etiology. As with most movement disorders, myoclonus can be focal, multifocal, segmental, or generalized, and can be one of the signs in a wide variety of nervous system disorders.

Objectives:

- Describe the pathophysiology of myoclonus, depending on the underlying etiology.
- Outline the components of proper evaluation and assessment of a patient presenting with myoclonus, including any indicated laboratory and/or imaging studies.
- Discuss available treatment and management options for myoclonus, based on underlying etiologies.
- Review the importance of improving care coordination among interprofessional team members to improve outcomes for patients affected by myoclonus.

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Introduction

Myoclonus is defined as rapid, brief, jerky, or shock-like movements involving muscle or group of muscles. Among all hyperkinetic movement disorders, myoclonus is considered to be the most rapid and brief. When caused by sudden muscle contractions, it is known as 'positive myoclonus', while a brief loss of muscular tone results in 'negative myoclonus' such as in asterixis.[1][2]

Myoclonus is classified in different ways according to its physiology, anatomical site of origin, and etiology. As with most movement disorders, myoclonus can be focal, multifocal, segmental, or generalized.[3]

Myoclonus is one of the signs in a wide variety of nervous system disorders such as dystonia, multiple sclerosis, Parkinson's disease, Creutzfeldt-Jakob disease (CJD), serotonin toxicity, Huntington disease, subacute sclerosing panencephalitis, Alzheimer's disease, and Gaucher disease.

Etiology

Etiologically, myoclonus can be primary or secondary. Primary myoclonus can further be subdivided into physiological myoclonus (hypnic jerks which occur in normal individuals), essential myoclonus (idiopathic or

hereditary), and epileptic myoclonus, while secondary myoclonus is also called 'symptomatic myoclonus, where the myoclonus is secondary to an underlying disorder.[1]

Moreover, myoclonus can be subdivided by its anatomical origin into the following types:[4]

1. Cortical (includes post-hypoxic action myoclonus, primary generalized myoclonic seizures, primary generalized epileptic myoclonus, juvenile myoclonic epilepsy)
2. Subcortical (includes essential myoclonus, reticular reflex myoclonus, opsoclonus-myoclonus syndrome, hyperekplexia)
3. Spinal (includes propriospinal myoclonus and segmental spinal myoclonus)
4. Peripheral (includes hemifacial spasm)

Since functional myoclonus is among the most common functional movement disorders, it is crucial to differentiate it from organic myoclonus. This can be done through the localization of the movements. Features in favor of an organic cause over a functional cause of myoclonus include consistent phenomenology, insidious onset, or response to benzodiazepines or antiepileptic medication.[5] The presence of spontaneous periods of remission, pronounced reduction of the myoclonus with distraction, acute onset, and sudden resolution favors the functional diagnosis of myoclonus.[4]

Epidemiology

In the United States, myoclonus is not common, with a lifetime prevalence of 8.6 per 100,000 population. After 50 years of age, the prevalence increases. In about a quarter of cases (27%), myoclonus is transient and usually drug-induced. About 8% of the myoclonic cases are functional.[6] The average annual incidence of myoclonus (from 1976 to 1990) was 1.3 cases per 100 000 person-years.[4]

The most common type of myoclonus is symptomatic myoclonus representing 72% of all patients with myoclonus,[7] followed by epileptic myoclonus with around 17%, while essential myoclonus represents only 11% of all patients with myoclonus.[6]

Pathophysiology

Myoclonus can arise from several levels in the nervous system, ranging from the cerebral cortex to the peripheral nerves. Cortical myoclonus predominantly affects body regions with the largest cortical representations, such as the hands and face. As the motor areas of the cerebral cortex are mostly involved during voluntary actions, the jerks mostly manifest on the action (i.e., action myoclonus).[4]

Brainstem motor systems are mainly involved in axial and bilateral movements and are near subcortical reflex centers. Thus, brainstem myoclonus is characterized by being generalized and stimulus sensitive (especially auditory stimuli). Its hallmark is auditory reflex jerks as in startle syndrome and brainstem reflex myoclonus.[4][8][4]

History and Physical

History-taking in a patient with myoclonus should focus on the age of onset, course, and duration of disease, family history, precipitating factors such as drugs, triggers (auditory stimulus, movement, tactile, visual, or emotional stimuli), the body parts involved, and the rhythmicity of the myoclonus. The presence of associated neurological signs/symptoms or disorder help to identify the underlying etiology and/or pathology.[6]

The main clinical characteristics demarcating myoclonus from other movement disorders are its rapid onset, brief duration, and the single-event type of muscle activation.[9] Myoclonus can occur spontaneously (at rest) or during movement (i.e., action myoclonus) and can be provoked by external tactile or acoustic stimuli (i.e. reflex myoclonus).

Clinically, cortical myoclonus is more prominent in the hands and face and more commonly provoked by tactile stimuli. Moreover, cortical myoclonus is generally action-induced. There is a frequent occurrence of negative myoclonus as well.

Brainstem reflex myoclonus, a subtype of subcortical myoclonus, typically presents with jerks that are more prominent in the axial region and proximal extremities.[10] Startle syndrome involves movements that occur bilaterally and are characterized by raising flexed arms over the head and closing eyes. [11]

Spinal myoclonus tends to present as unilateral arrhythmic jerking in the arm and/or trunk, whereas repetitive jerking of only the trunk and abdomen, in the supine position and bilaterally, may be due to propriospinal myoclonus.[12]

Since a variety of classifications exist, there is considerable overlap between the subtypes of different classifications. [3]

The presences of cognitive impairment, epilepsy, ataxia, other movement disorders, or neurological signs usually point towards symptomatic myoclonus. Meanwhile, acute or subacute onset should orientate towards a toxic, metabolic, or infectious etiology. A progressive course should orientate toward a degenerative process, static encephalopathy, or progressive metabolic disease.[7]

Evaluation

The determination of the generator site in myoclonus is an important clue for diagnostic orientation. All neurophysiologic tools (electromyography [EMG], electroencephalography [EEG], evoked potentials) may be helpful. [13] EEG is useful for the identification of both ictal and interictal patterns in epileptic myoclonus (for example, generalized spikes, and 3 to 6 Hz spike and wave patterns are typical for primary generalized myoclonic epilepsy).[4]

In cortical myoclonus, the technique of EEG-EMG back-averaging (correlation of electrical activity in the brain with muscle jerking) shows cortical discharge preceding the activity on electromyography. Other findings may be enlarged somatosensory evoked potentials.[14] The EEG in cases of subcortical myoclonus is devoid of back-averaged cortical activity or giant somatosensory evoked potentials.[3]

Imaging of the brain and spinal cord may be done to determine structural or focal lesions.

Advanced testing for specific and rare diagnoses should be considered, including, but not limited to, a cerebrospinal fluid exam, tissue biopsy of skin, and genetic testing for inherited disorders.[4]

Treatment / Management

Clinicians mainly manage myoclonus by treating symptoms. The previously mentioned classification of myoclonus into cortical and subcortical helps in the guidance of pharmacotherapy since spinal myoclonus fails to respond to medications effective for cortical myoclonus and vice versa.[12]

In cortical myoclonus, first-line medications include valproate, clonazepam, and levetiracetam with topiramate and zonisamide used as second-class agents.[15][16][17][18] Meanwhile, antiepileptic drugs as phenytoin, carbamazepine, and lamotrigine may cause a worsening of cortical myoclonus symptoms.[19] On the other hand, subcortical/spinal myoclonus responds to both clonazepam and levetiracetam as first-line and second-line agents, respectively.[12]

Botulinum toxin can be offered as a treatment option in focal peripheral myoclonus.[18][20]

In regards to deep brain stimulation (DBS) in myoclonus, the most commonly reported indication is myoclonus–dystonia syndrome, and the target is usually both internal globus pallidus (GPi) and thalamic nuclei.[3][21]

Differential Diagnosis

Differential diagnosis of myoclonus includes similar movement disorders presenting as rapid and jerky movements. They vary in amplitude from mild, not causing movements across the joint as fasciculations and myokymia to stronger movements as tremor, tics, and chorea, all of which need to be distinguished from myoclonus.[12]

The differential diagnosis can include:

- Dystonia
- Multiple sclerosis
- Parkinson
- Creutzfeldt–Jakob disease
- Serotonin toxicity
- Huntington disease
- Subacute sclerosing
- Panencephalitis
- Alzheimer disease
- Some forms of Gaucher disease

Prognosis

The degree to which myoclonus improves with treatment depends on the underlying etiology. Certain types of myoclonus may be secondary to potentially treatable etiologies as an inflammatory disorder or a metabolic disturbance. For example, in drug-induced myoclonus, stopping the offending drug can be sufficient for complete resolution of myoclonic jerks.[16]

Complications

Myoclonus can be the cause of significant disability with impairment in activities of daily living and maybe depressive symptoms. This results from jerky movements occurring during rest, with muscle activation or by external stimuli as sound resulting in interference with performing or starting the desired correct movement for a specific task. [18]

Enhancing Healthcare Team Outcomes

When a primary caregiver or nurse practitioner comes across a patient with a movement disorder, referral to a neurologist is recommended as the differential diagnosis is broad and the treatments do vary for each disorder. The interprofessional team may consist of a neurologist, neurosurgeon, neuroscience nurse, and a pharmacist. The nurse can monitor patient response, provide education, and keep the team apprised of changes in patient status. The pharmacist should educate the patient about medication use, compliance, and side effects. The best outcomes will be achieved with a team approach. [Level 5]

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