Journal of Clinical Cases and Reports

Clinical Review | Vol 3 Iss 3

Myoclonus Considerations on Their Classification and Treatment

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Abstract

Myoclonus is defined as involuntary, sharp and short-term movements of one or more muscular groups, stemmed from any structure of the central nervous system. Even though the study the myoclonus to motivated great quantity of scientific articles is not known with precision the physiopathological mechanisms that produce them. There are numerous classifications of the myoclonus that consider semiologic aspects, etiological, nosological, or physiological. Within them it takes on great importance the cortical myoclonus which occurs after the cortical origin of discharge transmitted through the pyramidal way.

Keywords: ***Myoclonus; Classification; Cortical myoclonus; Treatment***

Received Date: September 12, 2019; **Accepted Date:** September 20, 2019; **Published Date:** September 27, 2019

Introduction

Myoclonus consists of an involuntary, sharp and brief contraction of a muscle or group of muscles that is expressed by a shake from 20 ms to 120 ms of duration and very variable amplitude [1]. The myoclonic crisis occurs through a cortical or subcortical generator that acts on the muscles by polisinaptics mechanisms, most frequently that by the cortical-spinal monosinaptic way, as has been demonstrated upon studying the latencies between the beginning of the complex and the potential electromyography deltoid muscle [2].

Myoclonus can be located, isolated or repeated and rhythmic or arrhythmic mass that can appear without triggering cause (spontaneous myoclonus) or induced by sensory or somestesics incentives (reflex myoclonus) or by muscular activity (action myoclonus).

Its clinical meaning is variable and observed in normal subjects in different situations or circumstances, or physiological myoclonus, or as expression of a dysfunction of the central nervous system in which the myoclonus is a symptom more within a pathological context more or less complex, or the predominant symptoms.

Citation: Julio López Argüelles, Myoclonus Considerations on Their Classification and Treatment. J Clin Cases Rep 3(3): 75-81.

Given importance and frequency of this movement disorder, we intend to review the principal forms to classify the myoclonus and their treatment.

Discussion

According to the semiology the myoclonus can be classified in accordance with its topographical distribution in generalized, segmentary, focal, and multifocal, according to its temporary pattern in rhythmic and arrhythmic, with regard to its form of induction in spontaneous, action and reflex (Table 1).

In accordance with the topography
Generals
Segmentary
Focal
Multifocal
In accordance with its temporary pattern
Rhythmic
Arrhythmic
De acuerdo a su forma de induccion
Spontaneous
Action

Table 1: Semiologic classification of myoclonus.

According to its physiopathology can be divided into positive and negative, being divided: the first in accordance with its origin at different levels of the nervous system in: cortical, subcorticals, cortical-subcortical, spinal and more recently has been included within these the stemmed from the peripheral nerve [3,4] (Table 2).

A. Positive
1. Cortical
Ø reflex
O Photic stimuli
O Somatosensorials stimuli
Ø spontaneous
Ø action
Ø mixed
2. Subcorticals
Ø Basal ganglia
Ø brainstem
Ø reticulate
3. Espinales
Ø spinals
Ø segmentary
4. Cortico-subcorticals
B. Negatives
1. Cortical
2. Subcortical

Table 2: Physiopathological classification.

Within this group it takes on great importance the cortical myoclonus which occurs after the cortical origin of discharge transmitted through the pyramidal way. The first association description between myoclonus and cortical activity is due to Gibas et al. that in 1935 describe activity end wave in the EEG associated with myoclonic shakes [5].

The cortical myoclonus can be characterized from the electrophysiological standpoint by the following criteria:

1. Brief duration of the muscular discharge (10 ms-50 ms)
2. Focal, multifocal, or generalized distribution.
3. If is generalized continues to be a diffuse pattern cephalocaudal, with initial discharge of the most cranial muscles, closer to the cortex [6,7].
4. Reflex myoclonus to the cutaneous stimulation, proprioceptive o electric of a nerve in muscles of the promoted member. Can also be triggered multiple discharges to a single stimulus higher than the 20 Hz [6,8-10].
5. Cortical components of the somatosensory evoked potentials for great amplitude [11,12].

From the etiological standpoint the myoclonus can be: physiological, as the hiccup, those which appear at the beginning the sleep or those which are associated states of anxiety or fatigue. Exist also myoclonus of epileptic mechanism. The essential myoclonus are rare; they tend to be arrhythmic and erratic, distributed by the whole body; can be spontaneous or family. Finally, the symptomatic mioclonías are part of broader neurological syndromes, of highly diverse types [13-15] (Table 3).

Table 3. Etiological Classification
I. Physiological myoclonus
Ø Entry in sleep
Ø States of anxiety-weariness
Ø Hiccup
II. Essential myoclonus
Ø Essential family members
Ø Sporadic
Ø Nocturnal
III. Symptomatic myoclonus
1. Deposit diseases
Ø With bodies of Lafora
Ø Tay-Sachs and Krabbe diseases
Ø Lipofucsinosis ceroide
Ø Gaucher´s disease
2. Spinals and cerebellar degenerations and related diseases
Ø Mitochondrial Encephalomyopathy (MERFF)
Ø Kufs´s disease
Ø Celiac disease
Ø Friedreich´s disease
Ø Telangiectasia ataxia
Ø Unverricht-Lundborg disease
Ø Dentato-rubro-pallido-luysian atrophy
Ø Olivopontocerebelose atrophy
3. Degenerative diseases
Ø Progressive supraduclear paralysis
Ø Huntington's disease
Ø Alzheimer's disease
4. Infectious diseases
Ø Creutzfeld-Jakob´s disease

Ø Sclerosing subacute panencephalitis
Ø Encephalitis by virus
Ø Polimyoclonc child with opsoclonus (Kinsbourne)
Ø Whipple´s disease
5. Encephalopathies and metabolic diseases
Ø Wilson´s disease
Ø Liver, renal, respiratory, dialysis, hyponatremia,
Ø Hyperglycemia, etc.
6. Toxic encephalopathies
Ø Bismuth, methyl bromide, etc.
Ø Drugs : levodopa, lithium, inhibitors of the MAO
Ø Tricyclic antidepressants, etc.
7. Physical encephalopathies
Ø Postanóxicas, postraumatic, electrocution
8. Focal injuries
Ø Stroke
Ø Tumors
Ø Injuries
Ø Medullary diseases
IV. Myoclonic Epilepsies.

Table 3: Etiological classification.

Myoclonic Epilepsies

In 1986, Fejerman [3], establishes a classification of the myoclonus, in relation to the association or not to encephalopathy and/or epilepsy, divided into five groups: mioclonías without encephalopathy nor epilepsy; encephalopathies with non-epileptic myoclonus; progressive encephalopathies with epileptic myoclonus; encephalopathies with myoclonic crises, and myoclonic epilepsies.

The myoclonus and epilepsies are closely related on numerous occasions; the most practical example of this is the heterogeneous group of so-called myoclonic epilepsies. Myoclonic epilepsies constitute a very controversial group within the epileptology, since the myoclonic crises can occur in many epileptic patients with different syndromes and can appear in all ages, from the newborn up to the elderly person [16].

For the discussion of myoclonic epilepsies, will follow a recent classification based on the one accepted by the International League against Epilepsy [18] (Table 4).

Idiopathic focal epilepsies ➢ Epilepsy with rolandic paroxysms (negative myoclonus) ➢ Epilepsy of the reading (myoclonic variant) **Symptomatic focal epilepsies** ➢ Continuous partial epilepsy ➢ Negative myoclonus in focal epilepsies
Idiopathic generalized epilepsies ➢ Benign myoclonic epilepsy of the infant ➢ Other child myoclonic syndromes ➢ Palpebral Myoclonus with absences ➢ Juvenile myoclonic epilepsy and related syndromes ➢ Photogenic epilepsies

Symptomatic generalized epilepsies ➢ Epilepsy with myoclonic absences ➢ Epilepsy myoclonic astatic (Doose´s syndrome) ➢ Myoclonic variant of the LennoxGastaut´s syndrome ➢ Child myoclonic encephalopathies ➢ Progressive myoclonic epilepsies ➢ Family myoclonic epilepsy of the adult ➢ Myoclonic epilepsies of the elderly person Linked with Alzheimer's disease Related to Down syndrome
Indeterminate epilepsies ➢ Severe myoclonic epilepsy of the infant
Others ➢ Myoclonic epilepsies related to static encephalopathy Angelman´s syndrome

Table 4: Epileptic syndromes that attend with myoclonic crises.

Treatment of Myoclonus

The treatment of the myoclonus will depend obviously on the knowledge of its cause. Thus the myoclonus epileptic and many cases of myoclonus essential should be treated with antiepileptic drugs and some symptomatic myoclonus could be treated specifically. But cannot sometimes be carried out more than a purely symptomatic treatment and to this end is advisable to know the neurochemistry of the myoclonus. Thus the therapeutic approximation to the myoclonus should be individualized.

In addition, should be taken into account that on some occasions the treatment should be with multiple drugs, basically in cases of symptomatic myoclonus [17].

First line
Clonazepam
Valproic acid
Piracetam
Second line
Lisuride
Trihexyphenidyl
Opioids Agonists
Tetrabenazine

Table 5: Drugs utilized in the treatment of the myoclonus.

Clonacepán is a benzodiazepine with gabergic and serotoninergic action. The effective dose is very variable, habitually between 6 mg/day and 20 mg/day, being always recommended titling it with minimum and gradual increases (to begin with 0 mg/d, 5 mg/d-1.5 mg/d, and to increase 0.5 mg-1.5 mg every week). It is probably the antimioclonus drug of choice. It has the drawback of which acute tolerance (tachyphylaxis) can occur after some weeks of treatment even though the initial response is good. In these cases it is recommended adding another drug to the treatment or suspending it temporarily in order to reintroduce it subsequently [15].

The valproic acid is a drug with action on different neurotransmitters (it increases the serotonin, GABA, aspartic and glycine). The dose to utilize is in addition variable (habitually 1500 mg/d -2000 mg/d) being able to be made the same recommendations that obtained the clonacepam. Tachyphylaxis is not developed [20].

Piracetam is a drug of mechanism of little known but effective action in some cases. The dose to utilize is habitually of 12 gr/d -16 gr/d. It is a drug that agrees always to have "in mind" by its effectiveness and because it practically is free from side effects [22].

Lisuride is a dopaminergic agonist with serotoninergic action. Perhaps, thus is the dopaminergic agonist most useful in the treatment of the myoclonus. Is particularly useful in reflex epilepsy and can be it in posthypoxic myoclonus [19].

Trihexyphenidyl is an anticholinergic drug that is especially effective in the treatment of velopalatines myoclonus (rhythmic myoclonus that affect the velopatatine musculature due to an injury in the Mollaret triangle trained by the dentated nucleus, bulbar olives and red nucleus), considered by some as a way of tremor. Its effectiveness has recently been described in cases of essential myoclonus.

Have been described cases of spinal myoclonus sensitive to tetrabenazine, a presinaptic dopaminergic antagonist.

Conclusion

Myoclonus is kept as one of the most various involuntary movements, with great clinical and topographical variability. For its correct treatment it is indispensable initially to carry out an adequate diagnosis and classification, in order to subsequently begin with the prescribed drug.

Acknowledgement

None.

Conflict of Interest

There was no conflict of interest in the research governance and conduct of the research, nor in the reporting arrangements.

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