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MYOCLONIC EPILEPSIES OF INFANCY AND CHILDHOOD

Myoclonus and myoclonic seizures

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Myoclonus refers to sudden, brief, shock-like involuntary movements, caused by muscular contractions (positive myoclonus) or inhibitions (negative myoclonus) arising from the central nervous system. This definition excludes the muscular twitches or fasciculation due to lesions of the lower motor neurons.[1] Myoclonus has been observed in patients with various neurological conditions including epilepsy, and the clinical manifestations, etiology or other associated symptoms are not always homogeneous. Various attempts to classify myoclonus have been made on the basis of clinical appearance, predominant distribution of the myoclonus, temporal pattern, etiology and, most recently, the anatomical origin of the jerks based on the underlying neurophysiological mechanism.[2] Although there have been a few proposals of anatomical classification, they have largely subdivided the origin into the cortex, subcortical structures including reticular formation, and spinal cord.

Although myoclonus is a major and distinct seizure type in epileptology, the neurophysiological background of myoclonic seizures has long remained uninvestigated. Recent progress in the neurophysiological understanding of myoclonus has thus prompted the ILAE to reconsider the classification of myoclonic seizures from an epileptological standpoint, and to propose the classification of myoclonus into cortical myoclonus, thalamo-cortical myoclonus, reticular reflex myoclonus and negative myoclonus.[3]

Cortical myoclonus has a proven cortical origin for which a computer-assisted averaging technique can only identify the preceding small cortical spike discharges, thus, the underlying mechanism is similar to focal seizures.[2,3] Some epilepsia partialis continua can be classified into cortical myoclonus. Thalamo-cortical myoclonus corresponds to MS, the most common form of myoclonus in epilepsy.[3] The ictal EEGs of such patients consist of generalized bilaterally synchronous spike and wave complexes, which are presumably generated by thalamo-cortical hyper-excitability. Reticular reflex myoclonus has a brain stem origin, in which the myoclonus impulse spreads along the body away from the source in a time-related sequential fashion.[2,3] There seems to be no counterpart in epileptic seizure classification, although generalized tonic seizures and epileptic spasms may be candidates. Negative myoclonus is a special type of myoclonus producing negative motor phenomena, including epileptic negative myoclonus, and may be a brief form of atonic seizures, as a counterpart in epileptic seizure classification. It is difficult to make a definite diagnosis of cortical and reticular reflex myoclonus because it requires a sophisticated computer-assisted technique.

Among the four types of myoclonus, thalamo-cortical myoclonus and negative myoclonus have long received attention in epileptology, because they are the main seizure types in various important epileptic syndromes developed during infancy and early childhood. However, there has been some confusion in determining the seizure type diagnosis solely based on history-taking or visual inspection, because several other seizure types including epileptic spasms, brief tonic seizures and non-epileptic myoclonus produce apparently similar clinical pictures indistinguishable from myoclonic seizures, epileptic negative myoclonus and atonic seizures, respectively. Recent advances in video-EEG systems together with polygraphic techniques have enabled us to distinguish these seizure types precisely, and offer important bases for constructing syndromic classification. Myoclonic seizure has been found to be a main seizure type in juvenile myoclonic epilepsy, severe and benign myoclonic epilepsies, and cryptogenic myoclonic-astatic epilepsy, etc., although there are some clinical and EEG differences in myoclonic seizures among these syndromes. Epileptic negative myoclonus and atonic seizures are the predominant seizure type in children with cryptogenic focal and generalized epilepsies including myoclonic-astatic epilepsy, respectively.

In juvenile myoclonic epilepsy, myoclonic seizure occurs singly or successively, during wakefulness, corresponding to a burst of generalized bilaterally synchronous spike and wave complexes at 4-5 Hz lasting up to 4 seconds.[4] Myoclonic events are momentary, and involve either the distal or proximal muscle predominated. In benign myoclonic epilepsy, myoclonic seizures occurs singly and infrequently during wakefulness and/or sleep and corresponds to generalized bilaterally synchronous spike and wave complexes at 1.5-3 Hz. MS involves the proximal muscle predominantly. Myoclonic EMG potentials last approximately 200 to 300 msec, longer than those of juvenile myoclonic epilepsy.

Severe myoclonic epilepsy patients have at least four types of myoclonic attacks detectable by means of video-EEG or polygraphic recordings.[5] The first seizure type is characterized by a more or less symmetrical momentary jerking or twitching of the proximal muscles and eyelids, corresponding to irregular generalized bilaterally synchronous spike and wave complexes at 3 to 3.5 H. They tend to occur successively during wakefulness. The second type of myoclonic attack consists of successive myoclonic twitching involving mostly the head, and eyelids, and results in rhythmic retropulsion of the head. The third seizure type is massive myoclonia, which is infrequent and sometimes appears only before the onset of generalized tonic clonic convulsion, i.e., generalized tonic clonic convulsion seizure. These seizures are frequently observed during sleep, and are considered to be a fragment of generalized tonic clonic convulsion seizures. The final type is non-epileptic segmental and multifocal myoclonus, often seen during the period of frequent generalized tonic clonic convulsion seizures.

Myoclonic seizures in cryptogenic myoclonic-astatic epilepsy occurs singly and infrequently during wakefulness and/or sleep and corresponds to generalized bilaterally synchronous spike and wave complexes at 1.5-2 Hz. At times, Myoclonic seizures involves the axial muscles, giving rise to massive flexion of the trunk at the waist, leading to patients being thrown forward on the floor (drop attacks due to flexor spasms).[6]

Atonic seizures, another form of astatic seizures observed in myoclonic-astatic epilepsy, are characterized by sudden collapse of the body forward, when patients are sitting.[6] When they are standing, they collapse straight downward onto the buttocks with an immediate recovery. Polygraphic study has revealed that generalized bilaterally synchronous spike and wave complexes at 1.5 to 2 Hz corresponds to a sudden interruption of ongoing EMG activity, lasting from 300 to 500 msec.

Epileptic negative myoclonus is characterized by a brief lapse in the postural tone of a body segment, which is time-locked to an epileptic spike or wave over a contralateral central region without evidence of preceding positive myoclonia on agonist and antagonist muscles, as defined by Tassinari.[7] In epileptic negative myoclonus observed in patients with benign focal epilepsy, there was the frequent dropping of one arm, which corresponded to sharp slow wave complexes on contralateral centroparietal regions. The silent-locked averaging of polygraphy demonstrated that the latency between the peak of the spike and wave complexes and the onset of epileptic negative myoclonus reached approximately 50 msec.[8]

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