

An Overview: Myoclonic Epilepsy Occurrence and Syndromes

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Perspective

Myoclonic epilepsy refers to a family of epilepsy that occurs in myoclonus. If myoclonic seizures are occasionally associated with abnormal EEG activity, it can be classified as a myoclonic seizure. Diagnosis of myoclonic epilepsy may be considered if abnormal EEG activity persists and is due to persistent seizures. Myoclonus can be described as a short cramp in the body. It can affect any part of the body, but most often it is found in the muscles of the limbs and face. Seizures are mostly involuntary and can lead to falls. EEG is used to read EEG activity. Spike activity produced by the brain usually correlates with the short spasms found in EMG or excess muscle artifacts. They usually occur without apparent loss of consciousness and can be generalized, topical, or focused in EEG recording. Myoclonic seizures can be epileptic or non-epileptic. Epileptic myoclonus is a basic electro clinical manifestation of epilepsy with descending neurons, and its spatial (dilated) or temporal (independent repetitive) amplification can cause overt epileptic activity. There are two syndromes and several related disorders:

Juvenile

Juvenile myoclonic epilepsy is accountable for 7% of cases of epilepsy; Seizures usually begin around puberty and usually have a hereditary.

Progressive

Progressive myoclonus epilepsy is a disorder associated with myoclonus, seizures, and other gait or conversation problems. These symptoms often worsen over time and can be fatal. MERRF syndrome is also known as

irregular fibrous myoclonic epilepsy. This rare hereditary disease affects muscle cells. In addition to myoclonus and epileptic seizures, MERRF features include ataxia, peripheral neuropathy, and dementia. Lafora disease is also known as Lafora progressive myoclonus epilepsy. It is an autosomal recessive disorder that causes recurrent seizures and diminished mental capacity. Lafora disease usually occurs in late childhood and usually dies about 10 years after the first signs of the disease. MERRF syndrome (or myoclonus epilepsy with irregular red fibers) is a mitochondrial disease. It is very rare and has different expressivity due to heterogeneous traits. MERRF syndrome affects different parts of the body, especially the muscles and nervous system. Signs and symptoms of this disorder appear at an early age, usually in childhood or adolescence. Although it is difficult to determine the cause of MERRF syndrome, it is a mitochondrial disease and can be caused by mutations in nuclear or mitochondrial DNA. [3] The classification of this disease varies from patient to patient, as many do not fall into a particular disease category. The main signs seen in people with MERRF include myoclonus, seizures, cerebellar ataxia, myopathy, and shabby red fibers (RRF) on muscle biopsy. Secondary symptoms are dementia, optic nerve atrophy, bilateral deafness, peripheral neuropathy, spasticity, or multiple lipomas. Mitochondrial diseases, including MERRFS, can occur at any age.

Unverricht-Lundborg disease is an autosomal recessive inherited disorder can be seen in kids as young as six years. It is usually correlated with possible loss of consciousness, rigidity, ataxia, dysarthria, declination of mental functioning, and involuntary shaking. Neuronal ceroid lipofuscinosis is a group of diseases that cause blindness, loss of mental abilities, and loss of movement. Majority diseases belonging in this group are Lysosomal-storage disorders leading to death roughly ten years after onset of the disease.

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