



Eyelid myoclonia with absences (EMA)

Epilepsy Action is indebted to Dr Richard Appleton, Consultant Paediatric Neurologist who specialises in children's epilepsy at Alder Hey Hospital, Liverpool, who has kindly written this factsheet.

For further information about epilepsy or anything mentioned in this fact sheet, please contact the Epilepsy Helpline freephone 0808 800 5050 or helpline@epilepsy.org.uk.

What is a syndrome?

A syndrome is a group of signs and symptoms that, added together, suggest a particular medical condition. In epilepsy, examples of these signs and symptoms would be things like the age at which seizures begin, the type of seizures, whether the child is male or female and whether they experience difficulties with learning.

Eyelid myoclonia with absences (EMA)

This epilepsy syndrome is rare and may be difficult to diagnose. It is sometimes mistaken for a much more common type of epilepsy called childhood absence epilepsy*. EMA is sometimes called Jeavons syndrome, after the doctor who first described a very small number of patients with this type of epilepsy.

In EMA, most children start having seizures around two or three years of age. However, because the absences last only a few seconds and may not be noticed by the family, it may not be diagnosed until much later.

Symptoms

As the name suggests, the main seizure type is a very brief absence, which usually lasts less than one or two seconds. At the same time as the absence, the eyelids or eyeballs may quickly roll or, more usually, jerk backwards and upwards so that the white part of the eyes is seen. This is the 'eyelid myoclonia' part of the seizure. The quick upward jerk of the eyelids in this epilepsy syndrome is different to the eyelid flickering or fluttering that happens in many children with childhood absence epilepsy. Also, the absences and eyelid flickering in childhood absence epilepsy usually last for between 10 and 15 seconds.

Absences with EMA may happen only infrequently (now and then) in the first few weeks after the epilepsy starts. However, very quickly, the absences increase in frequency to the point where they may happen 10, 20 or even 30 (or more) times every day.

Children with EMA are also photosensitive*. This means that the seizures are triggered by flickering light. The seizures may even be triggered by simply going out of a dark room into sunlight.

Rarely, children with this type of epilepsy may also have another type of seizure, called a tonic-clonic

seizure*. This usually only begins in later childhood or adulthood.

No cause has been found to explain this epilepsy. It is possible that future research might show it to be a genetic type of epilepsy (inherited, in the genes). However, research may also show it not to be an epilepsy syndrome but just part of another type of epilepsy.

A child's development and learning is usually unaffected by EMA, even though the seizures happen many times a day.

Diagnosis

The diagnosis is made by taking a very careful account of what happens during the seizures, and especially at what age the seizures start. Sometimes it can be very helpful to take a video recording of the seizures and show this to the doctor. However, this may be difficult because the seizures only last a few seconds. An electroencephalogram (EEG) may also be helpful, particularly if the child has one of their seizures while the EEG is being done. The EEG should also show the photosensitivity that happens in this type of epilepsy.

Treatment

The anti-epileptic drugs sodium valproate (Epilim), lamotrigine (Lamictal) and ethosuximide (Zarontin) may help to control most of the seizures. Sometimes a combination of two of these drugs may be more effective than a single drug. Other medications such as levetiracetam (Keppra) and clobazam (Frisium) may also be helpful. It is also important to know that some other anti-epileptic medications will make EMA worse and should **not** be used. The medications that should not be used

are carbamazepine (Tegretol), phenytoin (Epanutin) and vigabatrin (Sabril).

Prognosis (Outlook)

EMA is unlikely to be completely controlled with anti-epileptic treatment. It is also likely that the epilepsy will continue throughout childhood and into adult life. Fortunately, most children with this type of epilepsy will have no learning difficulties or behaviour problems.

* Factsheets are available from Epilepsy Action.

For information on seizure types, anti-epileptic drugs and general aspects of epilepsy please contact the Epilepsy Helpline freephone 0808 800 5050, helpline@epilepsy.org.uk

Support organisation(s)

Contact a Family, 209-211 City Road, London, EC1V 1JN, telephone 0808 808 3555, www.cafamily.org.uk

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