



Early Myoclonic encephalopathy (sometimes called neonatal myoclonic encephalopathy)

Epilepsy Action is indebted to Dr Richard Appleton, a consultant who specialises in children's epilepsy, and his associates, at Alder Hey Hospital, Liverpool, who have kindly written this fact sheet.

For further information about epilepsy or anything mentioned in this factsheet, 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.

Early myoclonic encephalopathy

Early myoclonic encephalopathy almost always starts in the newborn period or in very early infancy. It is a syndrome with several causes but it is thought that most of the infants have an underlying biochemical disorder even if this cannot be identified. Some infants definitely have a very rare condition called non-ketotic hyperglycinaemia. Affected infants have extremely poor development.

Symptoms

The seizures can be myoclonic jerks which affect small areas of the body from time to time, massive myoclonic movements (sudden flexion or extension), partial motor seizures or, rarely, tonic spasms (when stiffness occurs). The seizures are very frequent throughout the day. After several months seizures usually change to*infantile spasms (West syndrome).

The electroencephalogram (EEG), which records the electrical activity ongoing in the brain, shows evidence of abnormal discharge or spikes and waves interspersed with periods of flatness. The type of record is referred to as suppression-burst and is similar to that seen in *Ohtahara's syndrome.

The infants are all neurologically very abnormal. Often they are extremely floppy and excessively sleepy.

Treatment

The seizures are often resistant to medication. If this syndrome is suspected it is probably unwise to use valproate (Epilim), since in this group of babies an underlying biochemical disorder is quite likely. It is important to test the babies thoroughly for a possible chemical disorder, in case this in itself can be treated. The actual choice of medication is difficult and it may be safer to use some of the older anti-epileptic drugs such as phenobarbital before others are tried. All babies should receive a

trial of pyridoxine (vitamin B6) in case they have
*pyridoxine dependent seizures.

Prognosis (outlook)

Infants with early myoclonic encephalopathy make very little developmental progress. They remain totally dependent and often feed poorly. More than half of the infants who have been reported in the literature to have this condition die within the first year of life. Those who survive remain severely developmentally delayed.

* Factsheet on infantile spasms, Ohtahar's syndrome and pyridoxine dependent seizures are available from Epilepsy Action.

Epilepsy Action makes every effort to ensure the accuracy of information in its publications but cannot be held liable for any actions taken based on this information.

If you are reading this publication more than 12 months after the date shown below, please contact Epilepsy Action to check whether it is still current.

October 2006

S017

Epilepsy Action is a working name of British Epilepsy Association
New Anstey House, Gate Way Drive, Yeadon, Leeds LS19 7XY
tel: 0113 210 8800 **fax:** 0113 391 0300 **epilepsy helpline freephone:** 0808 800 5050
www.epilepsy.org.uk **email:** helpline@epilepsy.org.uk

Registered Charity in England (No. 234343) © Copyright Epilepsy Action 2006