Benign myoclonic epilepsy in infancy

Epilepsy Action is indebted to Dr Richard Appleton, a Consultant Paediatric Neurologist who specialises in children's epilepsy, and to Dr Rachel Kneen, Consultant Paediatric Neurologist and Dr Stewart Macleod, Specialist Registrar in paediatric neurology, at Alder Hey at Alder Hey Children’s Hospital, Liverpool, who have kindly prepared this fact sheet.

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.

Benign myoclonic epilepsy in infancy
This is a very rare form of epilepsy which is more common in boys than girls. Seizures begin from four months to three years of age.

Symptoms
The seizures themselves called myoclonic seizures are brief and may not initially be noticed. They involve head nodding and usually occur several times a day. They may cause loss of balance but only rarely will the child fall. As the child’s head drops forward onto their body, their arms tend to move upwards and outwards, while their legs may flex. They sometimes roll their eyeballs.

Diagnosis
A full account of the seizures and of what happens during the seizures is essential for the doctor making the diagnosis.

The electroencephalogram (EEG) test which records the electrical activity in the brain shows evidence of abnormal discharges of spikes and waves or polyspikes and waves, particularly as the child is falling asleep. The abnormalities can be made worse by exposure to flashing lights.

Treatment
The seizures virtually always respond to sodium valproate (Epilim). Clonazepam (Rivotril) may also be effective.

Prognosis (outlook)
Approximately half the children who have benign myoclonic epilepsy in infancy have normal behaviour and intellectual development later. However, there is some risk of delayed learning. In particular there may be delayed language development. It is not possible to say at an early stage which children will have later difficulties in learning. Some children also have behavioural problems.