EARLY MYOCLONIC ENCEPHALOPATHY

Early myoclonic encephalopathy almost always starts in the neonatal 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 of them definitely have a very rare condition called non-ketotic hypoglycaemia. 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 (sudden bending or extension) movements; partial motor seizures or rarely tonic spasms when stiffness occurs. The seizures tend to be very frequent throughout the day.

The EEG (electroencephalogram) test which records the ongoing electrical activity in the brain shows evidence of abnormal discharges or spikes and waves along 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.

TREATMENT

The seizures are often resistant to medication. If the syndrome is suspected it is probably unwise to use Valporate (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 itself can be treated. The actual choice of medication is difficult and it may be safer to use some of the older antiepileptic drugs such as Phenobarbitone before others are tried.

PROGNOSIS

Infants with early myoclonic encephalopathy make very little development 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 disabled.