

*Lesson of the week***Benign sleep myoclonus in infancy mistaken for epilepsy**

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Benign sleep myoclonus in infancy is a distinctive but underdiagnosed disorder of quiet sleep, which according to our findings occurs from the first day of life up to age 3 years. Its main features are rhythmic myoclonic jerks when drowsy or asleep, which stop if the child is woken, and normal encephalograms during or after the episodes.^{1 2} When all these features are present the diagnosis should be clear cut. The diagnosis may be difficult if the association with sleep is not noted and if no attempt is made to stop the “seizures” by waking the child. We report on 15 patients in whom benign sleep myoclonus was initially mistaken for epilepsy.

Patients

All the patients were referred during the five year period 1996-2001 for investigation and treatment of prolonged “seizures”; some had been given anti-convulsants, without effect. The table summarises the clinical details of all the patients.

Illustrative case

A 14 day old boy (case 1; see table) was admitted after emergency helicopter transfer. In the previous week he had had several episodes of what was assumed to be status epilepticus but these had failed to respond to rectal and intravenous diazepam. Phenobarbitone and then phenytoin had been added but the seizures had continued. When he reached hospital he had been having generalised myoclonic movements for one hour. He was receiving three anticonvulsants in high doses. After the “seizure” ceased he appeared well but was drowsy. He fed well. Spontaneous movements and newborn and positional reflexes were normal.

Encephalograms during and after the episodes showed excess beta activity with generalised slowing, which was ascribed to his medication. There was no epileptic activity. The results of ultrasound and magnetic resonance imaging of his brain were normal, as were concentrations of plasma calcium, magnesium, glucose, ammonia, and urinary amino acids and organic acids.

Despite triple anticonvulsant therapy the “seizures” recurred and he was given repeated doses of diazepam, resulting in shallow and irregular respiration with periods of apnoea and oxygen desaturation needing intensive care. There the nurses observed that “seizures” occurred only when he was drowsy or asleep, never when he was awake. A severe form of benign sleep myoclonus was then suspected and the anticonvulsants were discontinued. He became less drowsy and both respiration and the frequency of “seizures” improved. Waking the child abolished the myoclonic episodes. He continued to have similar episodes until 3 months of age but developed normally and had no further “seizures” during four years of follow up.

Data on other cases

One child's mother had had benign sleep myoclonus during early infancy. In three other cases there was a

history of unusually strong sleep onset myoclonus affecting in one case the mother and in the two other cases a sibling. In all cases pregnancy and delivery had been normal.

All 15 patients had generalised rhythmic myoclonic “seizures” but eight had also had focal clonic episodes affecting various sites. Brief periods of oxygen desaturation were noted in four of the nine patients who were monitored.

“Seizures” lasted longer than 30 minutes in four children, 10 minutes in two, and 2-10 minutes in nine.

Seven children were receiving anticonvulsants when first seen; all were being given phenobarbitone and two children were receiving one and one child two additional drugs (see table). After diagnosis all anticonvulsants could be discontinued.

All patients developed normally during six months and four years of follow up. None has developed epilepsy.

Discussion

The prevalence of benign sleep myoclonus is unknown but our experience in two centres suggests the condition is being under-recognised. Since writing this report one of us (JE) has seen four more infants with the condition during one year at the Kinderspital Meran, which has 1300 births annually. During the same period neonatal epilepsy was diagnosed in three other infants.

Benign sleep myoclonus usually presents within a few days of birth. Rhythmic myoclonic movements appear while the infant is drowsy or asleep but they stop if the child is woken, and this characteristic feature confirms the diagnosis. We have not encountered any cases clinically diagnosed as having sleep myoclonus—stopping on waking—who later turned out to have epilepsy. Sleep myoclonus usually disappears after a period of weeks and has resolved in most cases by 3 months of age. This coincides with the period of rapid maturation in sleep patterns seen during the first 3 months of life, at the end of which the longest nocturnal sleep period occurs and the diurnal-nocturnal pattern is established. During the first 12 weeks of life the initial period of sleep gradually changes from REM to non-REM sleep, and the total REM sleep periods continue decreasing markedly during the first six months of life.^{3 4}

Although most patients with sleep myoclonus seem free of “seizures” by the age of 3 months, some may be having prolonged episodes of nocturnal myoclonus beyond that period unobserved because after that age prolonged sleep occurs mainly at night time when parents are asleep. Indeed the condition may persist for months and years, as the table shows—in patient 6 it lasted until age 3 years, and in patients 5, 9, and 11 it resolved at 7, 6, and 5 months respectively.

Benign sleep myoclonus may be mistaken for neonatal epilepsy due to a serious underlying disorder, or

Benign sleep myoclonus may be mistaken for epilepsy; prompt diagnosis prevents unnecessary investigations and treatments

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Clinical details of 15 infants with sleep myoclonus

Case	Sleep myoclonus		Rhythmic myoclonus		Electroencephalography		Neuroimaging		Treatment	Outcome
	Onset (day of life)	Age resolved	General	Focal	Between episodes	During episodes	Ultrasound	CCT/MRI		
1	7	3 months	Present	Present	Normal	Normal	Normal	MRI normal	Phenobarbitone, phenytoin, diazepam	Normal
2	2	6 weeks	Present	Absent	Normal	Normal	Normal	ND	Phenobarbitone	Normal
3	4	3 months	Present	Absent	Normal	Normal	Normal	ND	Phenobarbitone	Normal
4	8	2 months	Present	Present	Normal	ND	Normal	ND	None	Normal
5	4	7 months	Present	Present	Normal	ND	Normal	ND	Phenobarbitone, phenytoin	Normal
6	7	3 years	Present	Absent	Normal	Normal	Normal	ND	Phenobarbitone, carbamazepine	Normal
7	7	4 weeks	Present	Absent	Normal	Normal	Normal	ND	None	Normal
8	5	3 months	Present	Present	Normal	ND	Normal	ND	None	Normal
9	7	6 months	Present	Present	Normal	ND	Normal	ND	None	Normal
10	7	4 months	Present	Present	Normal	ND	ND	ND	None	Normal
11	28	5 months	Present	Present	Normal	Normal	ND	ND	None	Normal
12	5	9 weeks	Present	Absent	Normal	ND	Normal	CCT normal	Phenobarbitone	Normal
13	8	4 months	Present	Absent	Normal	ND	Normal	CCT normal	Phenobarbitone	Normal
14	7	3 months	Present	Absent	Normal	ND	Normal	ND	None	Normal
15	2	3 months	Present	Present	Normal	ND	Normal	ND	None	Normal

ND = investigation not done; MRI = magnetic resonance imaging; CCT = cranial computed tomography.

for benign neonatal or familial neonatal seizures. When the myoclonic jerks are unilateral, a more serious condition is often suspected and the diagnosis of benign sleep myoclonus may not be considered. All our 15 patients had generalised jerking and eight had unilateral jerks as well. Investigations other than an encephalogram are not helpful. Ultrasound scans of the brains of all of our patients were normal, and so was cranial computed tomography in two and magnetic resonance imaging in one. Ultrasound examination of the brain is justified, especially if there are doubts about the diagnosis and to allay parents' anxiety, but neither the irradiation from tomography nor the risks of anaesthesia for magnetic resonance imaging can be justified in this self limiting condition.

Seven of our patients received anticonvulsants for periods ranging from three months to seven years, without benefit. Anticonvulsants are ineffective in sleep myoclonus, and indeed may be harmful: by causing drowsiness, they may increase the frequency of fits: there is no indication for giving them. The episodes

can be effectively managed by waking the child, and parents have become expert in this by simple measures such as changing nappies or gently squeezing extremities. It is important to tell parents not to waken their child by shaking.

Mistaking this benign self limiting condition for epilepsy may result in unnecessary investigations, unnecessary treatment, and unnecessary parental anxiety.

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Fig 1 Chemical burns at three days after exposure to giant hogweed sap



Fig 2 Giant hogweed (*Heracleum mantegazzianum*)

Know your weed

A 45 year old man spent two days clearing an overgrown area in the garden of a French villa. He worked in bright sunlight wearing only shorts, vest, gloves, and boots. Two days later he presented to the accident and emergency department with chemical burns (fig 1).

The plant he was handling was *Heracleum mantegazzianum*, better known as the giant hogweed (fig 2), whose sap contains furanocoumarin, which renders the skin photosensitive. In this case, exposure resulted in partial-thickness burns, which were successfully treated conservatively with topical ointments leaving minimal residual scarring. Although increasingly rare in Britain, giant hogweed is commonly found on the continent.

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