Letter to the editor

'Epileptic blindness in tuberous sclerosis complex'

SIR–We read with interest the case report by Shahar and Hwang[1] describing the existence of a status epilepticus amauroticus. We would like to report our experience of cerebral visual impairment in a female infant with tuberous sclerosis complex (TSC). TSC was diagnosed antenatally based on cardiac rhabdomyoma demonstrated by ultrasonography. Two days postpartum, seizures were noticed. Cerebral MRI revealed multiple tubers and subependymal noduli; a large tuber was found in the right parieto-occipital region. Antiepileptic treatment was started but was not successful. At follow-up, the child did not show any visual behaviour: no fixation nor following eye movements could be obtained. Seizures consisting of deviation of eyes and head to the right combined with nystagmoid eye movements occurred daily. A mild left-sided hemiparesis was found.

Ophthalmological examination did not show any retinal abnormalities and electroretinogram was normal. Visually evoked responses demonstrated normal latencies with low amplitudes. EEG at the age of 5 months showed a long series of epileptiform activity in the right temporo-occipital region, 15–20 times a day, with clinical signs of nystagmus and clonus of the left arm and leg. As the epilepsy proved to be drug resistant and psychomotor development stopped, at the age of 7 months an occipital lobectomy was performed on the child. Postoperatively she was seizure free. Three weeks after the operation she started to show visual behaviour, improving quickly in the following months. Seven years postoperatively, she suffers minor non-disabling seizures.

We agree with Shahar and Hwang[1] that epileptic cerebral visual impairment can occur in the presence of a unilateral epileptic focus. Even in tuberous sclerosis, in which results of epilepsy surgery are variable, resection should be considered in order to improve neurological functioning.

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Reference

1. Shahar E, Hwang PA. (2001) Prolonged epileptic blindness in an infant associated with cortical dysplasia. *Developmental Medicine & Child Neurology* **43:** 127–9.

Abstract

Analysis by gestational age of cerebral palsy in singleton births in north-east England, 1970–1994
PM Drummond and AF Colver
Paediatric and Perinatal Epidemiology (2002) **16:** 172–80.

This study reports from a well-established cerebral palsy (CP) register the changes in CP rates by gestational age for singleton births over a 25-year period in north-east England. Cases associated with a known post-neonatal insult were excluded. The denominator for the calculation of CP rate is the number of neonatal survivors. The gestational ages of numerators and denominators are of a high accuracy dating back to 1970. This is because academic units in paediatrics and obstetrics were studying the assessment of gestational age in individual infants and the distribution of gestational age across all births in the north-east from the 1960s. The total population is approximately 770 000. The rate of CP rose from 1.6 per 1000 singleton neonatal survivors between 1970 and 1975 to 2.3/1000 from1990 to 1994, a rise of 0.7/1000 (95% CI 0.2–1.3). There was little change in the rate of CP in term infants whereas in preterm infants (<37 weeks) it rose from 5.5 to 16.8, a rise of 11.3/1000 (95% CI 5.9–16.8). Rises occurred in the three preterm gestational age bands <28, 28–31, and 32–36 weeks with the most marked rise in those <28 weeks from 0 to 112.7. The proportion of all cases of CP arising in the preterm group rose from 19 to 45%; and the proportion of the severest cases arising in the preterm group rose from 8 to 55%. In those born after 32 weeks, there is a preponderance of small-for-gestation infants, with 10% of infants more than 2SDs below the mean. All types of CP are more common in infants below average weight for gestation and this is most marked for the non-spastic types which are almost only seen in term, small-for-gestation infants. Gestational age is the crucial determinant of rate of CP and the increase in prevalence seen over the past 25 years is due to increased rates in preterm infants, not term infants. Both conclusions, suspected from birthweight analyses, are now demonstrated conclusively, with the contribution coming from infants of 32–36 weeks' gestation as well as very preterm infants.

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