

PEDIATRIC NEUROLOGY BRIEFS

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SEIZURE DISORDERS

JUVENILE MYOCLONIC EPILEPSY

A case-report from the Dept of Medicine, University of Peradeniya, Sri Lanka, concerns a young adult with myoclonic epilepsy since 15 yrs of age whose attacks were precipitated while playing with the Rubik's cube, a three-dimensional colored puzzle. The EEG at rest and with photic stimulation was normal; playing the cube produced generalized 3Hz S/W accompanied by myoclonic jerks and mental blocking. Arithmetic, draughts (checkers), card games, and reading precipitated EEG epileptiform discharges but not jerks. Diazepam prevented the seizures, but made him drowsy. The patient preferred to disengage his mind from the puzzle when he felt a jerk. (Senanayake N. Epileptic seizures evoked by the Rubik's cube. J. Neurol Neurosurg Psychiatry 1987; 50:1553-1559).

COMMENT. Having played the Rubik's Cube and Rubik's Magic unsuccessfully myself and given up in despair, I can imagine that frustration could be a factor responsible for the patient's seizures as much as "spatial and decision making processes". The same author has described seizures evoked by card games, draughts, and a local game with sea shells called "punchi" (Epilepsia 1987; 28:356). Ch'en, Chi'in, and Ch'u were the first to describe Chess and Card epilepsy! (Chinese Med J 1965;84:470).

NEONATAL SEIZURES

Investigators in the sections of Neurophysiology and Pediatric Neurology, Baylor College of Medicine, Houston, TX, have characterized and classified 415 clinical seizures recorded in 71 neonates and 11 with EEG seizure activity without clinical accompaniments. A crib-side EEG/polygraphic/video monitoring system was employed. Seizures with a close association to EEG seizure discharges were focal clonic (58 in 14 patients), myoclonic (38 in 4 patients), focal tonic (8 in 2 patients), and apneic (4 in 1 patient). Seizures with an inconsistent or no relationship to EEG seizure discharges were motor automatisms ("subtle seizures") involving mouth, eyes, limb pedaling and swimming (140 in 22

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patients), generalized tonic (90 in 13 patients), and myoclonic (66 in 13 patients). Only 2 neonates had infantile spasms. Clonic seizures with focal EEG seizure activity correlated with focal brain lesions such as infarction or intracerebral hemorrhage and a favorable short-term outcome. Seizures with no or inconsistent relationship to the EEG were correlated with diffuse hypoxic-ischemic encephalopathy and a poor prognosis (>50% with abnormal neurologic exams at discharge, and 20% died). Those with myoclonic seizures had high morbidity (35%) and mortality (29%) compared to those with clonic seizures (71% normal at discharge). The authors question the use of potentially neurotoxic anticonvulsant drugs in neonates with nonepileptic seizures or "behaviorisms" not accompanied by EEG seizure activity. (Mizrahi EM, Kellaway P. Characterization and classification of neonatal seizures. Neurology 1987; 37:1837-1844).

COMMENT. I contacted Dr. Gerald Fenichel at Vanderbilt Univ Sch of Med, an authority on neonatal seizures, for his opinion regarding the use of anticonvulsants in the treatment of "subtle" seizures. Unless the motor automatisms or tonic posturings can be correlated with a simultaneous recording of EEG seizure activity, he considers these subtle seizures as non-epileptic and advises against treatment with anticonvulsants. Patients with epileptiform seizures associated with acute hypoxic-ischemic encephalopathy are given IV phenobarbital in a loading dose of 20-30 mg/kg to provide a serum level of 40 ug/ml. If the patient is free from seizures on recovery from the acute illness and at the time of discharge from hospital, anticonvulsants are discontinued. (personal communication). Despite the enthusiastic promotion of newer antiepileptics, there is a growing and greater awareness of the potential toxicity of anticonvulsants in general. Any practical, less hazardous alternative or safe means of withholding medication must be considered seriously, especially in the neonate and young child. See Brent et al (Pediatrics 1987; 80 (Dec):909) re phenobarbital-induced depression and suicidal behavior in epileptic children, another area for concern in the long-term use of anticonvulsant drugs.

PHENACEMIDE IN COMPLEX PARTIAL SEIZURES

The anticonvulsant phenacemide (phenylacetylurea), discarded for 30 years because of serious toxicity, has been resurrected and used in the treatment of 13 children with refractory complex seizures at Loyola University, Maywood, and Christ Hospital, Oak Lawn, Illinois. Twelve responded, nine were seizure free for 2-12 months, and one developed nausea and vomiting necessitating drug withdrawal. Other side-effects included aggressive behavior in 1, drowsiness (2) ataxia (2), headache (1), and elevated SGPT and GGT in one child aged 3 yrs with tuberous sclerosis. A liquid chromatography assay developed to determine plasma phenacemide concentrations showed a linear relationship between drug peak height and plasma concentration over a range of 0-150 ug/ml. After a single oral dose the peak concentration in a 16 year old patient was at 1 to 2 hours and in a 40 year old volunteer, at 5 hours.