SEIZURE DISORDERS

EPILEPTIC SYNDROMES AND PHOTOSENSITIVE SEIZURES

The clinical features of different types of photic-induced seizures and epileptic syndromes characterized by visual sensitivity are reviewed from the University of Pisa, Italy, and Centre St Paul, Marseille, France. Seizure types associated with clinical photosensitivity include eyelid myoclonus, generalized myoclonic jerks, tonic-versive seizures, absence, generalized tonic clonic, and focal seizures. Epileptic syndromes with photic-induced seizures include benign myoclonic epilepsy in infancy, absence epilepsy, juvenile myoclonic epilepsy, epilepsy with myoclonic-astatic seizures, primary reading epilepsy, severe myoclonic epilepsy of infancy, photosensitive occipital lobe epilepsy, and progressive myoclonus epilepsies (PME). PME with photic sensitivity are symptoms of neuronal ceroid lipofuscinosis, Lafora's disease, Unverricht-Lundborg disease, and myoclonus epilepsy and ragged red fibers (MERRF). Visually induced seizures can be generalized or focal, idiopathic or symptomatic, or represent a pure reflex photosensitive epilepsy. (Guerrini R, Genton P. Epileptic syndromes and visually induced seizures. **Epilepsia** January 2004;45 (Suppl 1):14-18). (Reprints: Dr R Guerrini, Division of Child Neurology and Psychiatry, University of Pisa & IRCCS Fondazione Stella Maris, via dei Giacinti 2, 56018 Calambrone, Pisa, Italy).

COMMENT. The treatment of photosensitive epilepsies involves preventive measures and antiepileptic medications (AED). (Covanis A et al. **Epilepsia** Jan 2004;45(Suppl 1):40-45; Bureau M et al. **Epilepsia** Jan 2004;45(Suppl 1):24-26). Preventive measures include the following: avoid stimuli (eg TV, videogames); use small TV, 100-Hz screen, remote control, sit >2 m away from screen, wear spectacles, avoid stress and fatigue. Usually a combination of avoidance of stimuli and an AED is necessary. Valproate (VPA) is the AED of first choice, and lamotrigine is second choice. Other drugs recommended are clobazam, levetiracetam, ethosuximide, and topiramate.

CEREBRAL CYSTICERCOSIS AND SEIZURES

A 15-year-old Peruvian girl with neurocysticercosis is reported from Cornell University, New York, and Institute Neurologicas, Lima, Peru. She had a 3-month history of headache, vomiting, and visual obscuration and a one month history of incoherent speech, confusion, and visual and auditory hallucinations. Examination revealed papilledema, neck stiffness, and psychomotor retardation. MRI of brain showed numerous diffuse cystic areas with a dramatic "Swiss cheese" appearance. The cysts in the cerebral cortex were bright on T2-weighted images, and were also apparent in the tongue and ventricles on T1 scans. FLAIR demonstrated a bright image of a scolex in the cysts. Stools showed ova of the pork tapeworm, *Taenia solium.* Western blot analysis was positive for cysticercosis. Treatment with albendazole (15 mg/kg/day) and prednisone (60 mg/day) for one month was effective. At 8 month follow-up, recovery was complete and MRI showed resolution of the cysts. (Sander HW, Castro C. Neurocysticercosis. **N Engl J Med** January 15, 2004;350:266).

(Respond: Dr Howard W Sander, Weill College of Medicine, Cornell University, New York, NY).

COMMENT. The tapeworm is responsible for 10 percent of seizure admissions to emergency departments of large urban hospitals in New Mexico and California. Cysticercosis is the most common parasitic disease of the CNS worldwide, and the leading cause of late-onset epilepsy in many developing countries (Maguire JH. Tapeworms and seizures – treatment and prevention. **N Engl J Med** Jan 15, 2004;350:215-217). A double-blind, placebo-controlled trial of albendazole (800 mg/day) and dexamethasone (6 mg/day), for 10 days in 60 adult patients with cysts, reduced the number of seizures with generalization during the 2nd to 30th month after treatment (Garcia HH, Pretell EJ, Gilman RH et al. A trial of antiparasitic treatment to reduce the rate of seizures due to cerebral cysticercosis. **N Engl J Med** Jan 15, 2004;350:249-257). A 41% reduction in the number of partial seizures was not significant, but a 67% reduction of seizures with generalization was significant.

HEREDITARY ATAXIAS

FEBRILE EPISODIC ATAXIA WITH NOVEL MUTATION

An episodic ataxia type 2 (EA2) kindred with ataxic spells induced by fever or high environmental temperature and a novel *CACNA1A* mutation were identified and reported from the Universities of Mississippi and Minnesota. The proband was a 75-year-old woman with episodes beginning in childhood of ataxia, vertigo, weakness, and migraine lasting several hours, and provoked by fever, heat, stress, or sudden movements. The proband's father and sister were similarly affected. In 11 patients with episodic ataxia, age of onset varied from infancy to the twenties. Episodes ranged from daily to 2 annually, and lasted minutes to days. They were sometimes accompanied by headaches, diplopia, nausea, and vertigo. Those with the mutation had interictal cerebellar deficits. Early cerebellar dysfunction in EA2 results from the mutations in the neuronal calcium-channel gene and not a degenerative process. (Subramony SH, Schott K, Raike RS et al. Novel CACNA1A mutation causes febrile episodic ataxia with interictal cerebellar defects. **Ann Neurol** December 2003;54:725-731). (Respond: Dr Christopher M Gomez, Box 295, Departments of Neuroscience and Neurology, 420 Delaware St SE, Minneapolis, MN 55455).

COMMENT. Episodic ataxia type 2 (EA2) is a dominantly inherited disorder, characterized by spells of ataxia, dysarthria, vertigo, and migraine, associated with mutations in the neuronal calcium-channel gene CACNA1A. Attacks are precipitated by stress, exercise, or alcohol. Some patients have nystagmus between spells and some develop a progressive ataxia in adulthood. Twenty one CACNA1A mutations have been described in EA2. The above kindred study adds a further mutation and clinical syndrome in which ataxic spells are precipitated by fever or overheating, and patients develop signs of cerebellar dysfunction between attacks. Other neurologic disorders caused by mutations in the CACNA1A gene are the dominantly inherited progressive spinocerebellar ataxia (SCA6), and familial hemiplegic migraine.