GELASTIC EPILEPSY AND HYPOTHALAMIC HAMARTOMA

Three patients with hypothalamic hamartoma and gelastic seizures were studied using ictal single-photon emission computed tomography and EEG recordings with depth electrodes at the University of Alabama at Birmingham Epilepsy Center. Precocious puberty developed by age 7, and the patients were large for their age, above the 95th percentile. Cognitive and behavioral abnormalities were revealed on neurologic examination, with impulsivity, irritability, and aggressive tendencies. Laughing seizures began in the newborn period in 2 patients and before 6 years in a third. The frequency of attacks ranged from 1 to 5 minor episodes daily to 10 complex partial seizures each month. Interictal EEGs showed rare sharp waves from various areas. Ictal EEGs revealed diffuse theta. MRIs showed hypothalamic hamartomas, with displacement of the floor of the third ventricle. Ictal SPECT showed focal hyperperfusion of the hamartoma and thalamus. Depth electrode EEG within the hamartoma recorded focal spikes. Stereotactic radiofrequency surgical treatment of the hamartoma of one patient resulted in seizure remission. (Kuzniecky R, Guthrie B, Mountz J et al. Ann Neurol July 1997;42:60-67). (Respond: Dr Kuzniecky, Department of Neurology, University Station, Birmingham, AL 35294).

COMMENT. Gelastic (*'gelos'* Gr for mirth) seizures in children with precocious puberty and hypothalamic hamartoma originate from the hamartoma and involve adjacent diencephalic areas. Seizures occur more commonly when the tumor displaces structures adjacent to the hypothalamus. See Progress in Pediatric Neurology II, PNB Publ, 1994;p41, for further articles on gelastic epilepsy. The anterior cingulate region may also be involved in some cases.

NONCONVULSIVE STATUS AND RING CHROMOSOME 20

Six cases of epilepsy and ring chromosome 20 are reported and 20 additional cases in the literature are reviewed from the National Epilepsy Center, Shizuoka Higashi Hospital, Japan. Seizures consisted of a prolonged confusional state, and the ictal EEG showed bilateral high-voltage slow waves with occasional spikes. Neurological exam and MRI were usually normal. Mental retardation or bordeline IQ was present in 5 of 6 cases. Seizures were refractory to AEDs. (Inoue Y, Fujiwara T, Matsuda K et al. Ring chromosome 20 and nonconvulsive status epilepticus. A new epileptic syndrome. Brain June 1997;120:939-953). (Respond: Dr Yushi Inoue, National Epilepsy Center, Shizuoka Higashi Hospital, Urushiyama 886, Shizuoka 420, Japan).

COMMNENT. Chromosome analysis may be indicated in children with drug refractory epilepsy and learning disabilities, despite the absence of dysmorphic features. Ring chromosome 20 in a child with gelastic and partial complex seizures was included in a section on mental retardation syndromes in Progress in Pediatric Neurology III, 1997;p384.

GENETICS OF EPILEPSIES WITH FOCAL SHARP WAVES

The clinical manifestations and genetic aspects in 147 children with benign focal sharp waves were evaluated at the Epilepsy Center, Raisdorf, Germany. Seizures in 134 patients included febrile convulsions (26%), rolandic

seizures (24%), neonatal (6%), generalized tonic-clonic (49%), and rare complex partial. Transmission of FC showed a maternal preponderance. The phenotypic expression of the genetic basis for focal sharp waves was remarkably variable. The clinical and EEG findings suggested a multifactorial pathogenesis for epilepsies characterized by benign focal sharp waves. (Doose H, Brigger-Heuer B, Neubauer B. Children with focal sharp waves: Clinical and genetic aspects. Epilepsia July 1997;38:788-796). (Reprints: Dr H Doose, Epilepsie-Zentrum, 24223 Raisdorf, Germany).

COMMENT. Focal sharp waves may be characteristic of a variety of different seizure patterns, including febrile convulsions, and not only that of rolandic epilepsy. In addition to epileptic syndromes, focal sharp waves may be found in children with learning disabilities such as dyslexia and dysphasia. The genetics of these syndromes appears to be multifactorial.

Genetics of febrile convulsions. Doose H and Maurer A have also reported on genetic factors in febrile convulsions. (Seizure risk in offspring of individuals with a history of febrile convulsions. Eur J Pediatr June 1997;156:476-481). The risk was 10%; only FC in 64% of affected offspring. Offspring of females with affected parents were at increased risk.

VAGUS NERVE STIMULATION FOR EPILEPSY

The Therapeutics and Technology Assessment Subcommittee of the American Academy of Neurology reports on vagus nerve stimulation (VNS) for epilepsy. A total of 104 articles and responses in 130 patients were reviewed. Data on VNS in children was sparse; 5 of 12 children with intractable partial, generalized, and mixed seizures, followed for 2 to 14 months in an open-label study, had a 90% reduction in seizure frequency. In a randomized prospective trial in 114 adult patients, high levels of stimulation showed a significant response compared to low stimulation. The degree of improvement was modest, with a mean partial seizure reduction of 31%, and 39% of patients having a >50% reduction in seizure frequency. Young patients with auras and idiopathic epilepsy had more than a 50% responder rate, while secondarily generalized seizures were not controlled. The mechanism of VNS was undetermined. Left-sided implantations were safer, since right VNS caused bradycardia. Electrodes around the vagus were connected to an infraclavicular generator pack. Serious complications were rare, but hoarseness, throat pain, and cough were common complaints during stimulation. The device is costly, and improvement in quality of life for the patient needs to be determined. (Fisher RS, Krauss GL, Ramsay E, Laxer K, Gates J. Assessment of vagus nerve stimulation for epilepsy: Report of the Therapeutics and Technology Assessment Subcommittee of the American Academy of Neurology. Neurology June 1997;49:293-297). (Reprints: Dr Robert S Fisher, Barrow Neurological Institute, 350 West Thomas Road, Phoenix, AZ 85013).

COMMENT. The panel considers vagal nerve stimulation a promising treatment for intractable partial seizures, but further controlled studies are recommended. Some predictors of a favorable response include age less than 34 years, idiopathic epilepsies, and early signs of effectiveness.

KETOGENIC DIET REVISITED IN THE LABORATORY

The effect of a ketogenic diet (KD) on kindled seizures and behavior in adult rats was studied at the Children's Hospital, Harvard Medical School, Boston, MA. In a kindling model, KD provided transient protection against