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DEVELOPMENTAL DISORDERS

PILEPTOGENICITY OF CORTICAL DYSPLASIAS AND TUMORS

The epileptogenic characteristic of focal cortical dysplasias and dysembryoplastic neuroepithelial tumors explored by depth electrodes and stereoelectroencephalography is quantified using an epileptogenicity index, in a study of 36 patients with focal drug-resistant epilepsy at Universite de la Mediterranee and other centers in Marseille and Rennes, France. The epileptogenic zone is organized as a simple focal lesional site or as a complex 'epileptogenic network' extending beyond the lesion. Epileptogenicity index (EI) values range from 0 (none) to 1 (maximal epileptogenicity). The mean EI in lesional regions was 0.87, and 0.29 in non-lesional structures. A single focal lesion was found in 31% of patients, and more than one epileptogenic region in 25 patients (64%) (a network organization in 61% and bilateral epileptogenic zone organization in 8%). Distant structures are often involved, and in mesio-temporal epilepsy, the number of epileptogenic structures increases with epilepsy duration. None patient with bilateral organization became seizure-free, while 87% with focal organization and 57% with network organization were seizure-free. The EI is of value in the delineation of the epileptogenic zone with brain lesions and in the definition of the extent of surgical resection. (Aubert S, Wendling F, Regis J, et al. Local and remote epileptogenicity in focal cortical dysplasias and neurodevelopmental tumours. **Brain** Nov 2009;132:3072-3086). (Respond: Pr Fabrice Bartolomei, MD, PhD, Service de la Neurophysiologie Clinique, CHU Timone-264 Rue st Pierre, 13005-Marseille, France. E-mail: fabrice.bartolomei@ap-hm.fr).

COMMENT. The epileptogenicity index (EI) is a method of quantifying and defining the epileptogenicity in and around focal cortical dysplasias and neurodevelopmental tumors during stereo-EEG with depth electrodes. Defining the extent of the epileptogenic zone is

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important in prognosis and in surgical resection. The November 2009 issue of **Ped Neur Briefs** reviews a report of a novel immunocytochemical test for epileptogenic brain tissue, independent of the histological findings (Sarnat HB, Flores-Sarnat L. **Can J Neurol Sci** 2009;36:566-574). With this added quantification of the epileptogenicity zone by stereo-EEG and the EI, surgical resection for refractory epilepsies should become more accurate and effective.

EPILEPSY IN ANGELMAN'S SYNDROME

The natural history and response to treatment of epilepsy in a large population of Angelman syndrome (AS) patients were studied by detailed electronic survey conducted through the AS Foundation by pediatric neurologists at Massachusetts General Hospital, Boston; Texas Southwestern Medical Center; and Rady Children's Hospital, San Diego, CA. Approximately 1000 families of individuals with AS were asked to complete a questionnaire online. The survey was available for 3 months, Feb–May 2007, and questions included the description of seizures, and response to various medications and their side effects. Responses were obtained from family members of 461 individuals with AS, a 40-50% response rate. The average age of patients was 11.8 years (1.3-45 years) at time of survey, and an average age of 5.3 years (<1-35 years) at diagnosis; 56% were male. Multiple seizure types were reported, most commonly atonic seizures (41%), generalized tonic-clonic (40%), atypical absence seizures (37%), and complex partial (32%). Myoclonic seizures occurred in 12% and infantile spasms in 2%. Control of seizures was reported in 34% for a median period of 3.2 years, usually beginning at 8.8 years of age. Of 396 with current epilepsy, only 46% of those age <3 years had seizures, whereas 53-64% ages 3-18 years had seizures; 35% had regression in development, and 12% had experienced convulsive status epilepticus. In 64% of subjects with epilepsy, emergency lorazepam or diazepam was used for prolonged seizures or clusters of seizures.

Rates of epilepsy differed among genetic subtypes; those with maternal deletions (89%) and unknown subtypes (90%) had the highest rates of epilepsy, whereas those with imprinting defects (55%) were least affected. Of all subjects, 65% had a maternal deletion, and 18% had an unknown subtype. Most commonly prescribed anticonvulsant medications (AED) were valproic acid (63%) and clonazepam (34%), but lamotrigine (24%) and levetiracetam (20%) had similar efficacy and tolerability. Only 15% responded to the initial AED, and an additional 8% responded to the second agent; 77% had refractory seizures. Ketogenic diet was effective in 11 of 31 subjects, and vagus nerve stimulation in 8 of 16. (Thibert RL, Conant KD, Braun EK, et al. Epilepsy in Angelman syndrome: A questionnaire-based assessment of the natural history and current treatment options. **Epilepsia** Nov 2009;50(11):2369-2376). (Respond: Dr Elizabeth Thiele MD PhD, Pediatric Epilepsy Program, Department of Neurology, Massachusetts General Hospital, 175 Cambridge Street No 340, Boston, MA 02114. E-mail: ethiele@partners.org).

COMMENT. Epilepsy is a common problem in AS and is refractory to treatment. Although often considered a generalized form of epilepsy, partial seizures are fairly frequent. Newer AEDs, lamotrigine and levetiracetam, are as effective as conventional medications and have fewer serious side effects. Genetic analyses are correlated with response to therapy.