

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.

Sleep problems associated with epilepsy in AS may be related to the severity of seizures and the use of anticonvulsant medication. In 290 individuals with AS, decreased nightly hours of sleep, and a difficulty initiating sleep were significantly correlated with epilepsy. (Conant KD, Thibert RL, Thiele EA. *Epilepsy and the sleep-wake patterns found in Angelman syndrome. **Epilepsia** Nov 2009;50(11):2497-2500.*

Genetic testing in AS demonstrates a molecular mechanism causing lack of expression of the UBE3A gene with abnormalities of chromosome 15, inherited from the mother. (Dan B. Angelman syndrome: Current understanding and research prospects. ***Epilepsia** Nov 2009;50(11):2331-2339.* Three characteristic EEG patterns are described: A. high amplitude delta with spikes anteriorly without clinical correlation; B. diffuse theta not associated with drowsiness; and C. high amplitude delta mixed with spikes in posterior regions on eye closure. Boyd SG and associates described the EEG features in early diagnosis of AS. (***Eur J Pediatr** 1988;147:508-513.*

LANGUAGE DISORDERS

LANGUAGE AND READING DISORDERS IN EPILEPSY

The severity and range of linguistic impairments in youths with epilepsy were studied at UCLA, Los Angeles, State Fullerton University, and UC at Irvine, California. Tests of language, intelligence, achievement, and psychiatric interviews were administered to 182 youths with epilepsy, ages 6.3-8.1, 9.1-11.7, and 12.0-15.2 years, and to 102 normal children. Parents provided demographic, seizure-related and behavioral information. Language scores 1 SD below average were significantly more frequent in epilepsy subjects than in controls. Intermediate and adolescent epilepsy groups had significantly lower mean language scores compared to controls. The older group had more language impairment. Longer duration of epilepsy, absence epilepsy, psychiatric diagnosis, and socioeconomic status were associated with linguistic deficits in the young group. Prolonged seizures, lower Performance IQ and minority status predicted low language scores in the intermediate age epilepsy group. Poor seizure control, decreased Performance IQ, and lower socioeconomic status correlated with language impairment in the adolescent group. Linguistic and reading deficits were significantly related in each epilepsy group. (Caplan R, Siddarth P, Vona P, et al. *Language in pediatric epilepsy. **Epilepsia** Nov 2009;50(11):2397-2407.* (Respond: Rochelle Caplan MD, Semel Institute for Neuroscience and Human Behavior, 760 Westwood Plaza, Los Angeles, CA 90024. E-mail: rcaplan@ucla.edu).

COMMENT. Linguistic and reading impairment in pediatric and adolescent epilepsy increases with age, and predictors of impairment vary with each age group. Language assessment and intervention are important in children with epilepsy.