Epileptic Encephalopathy: Regression May Be Selective

BY JEFF EVANS
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BALTIMORE — Childhood regression may be related to epileptic encephalopathy only among children with language regression alone, rather than among those with autistic regression, Dr. John F. Mantovani said at a meeting on developmental disabilities sponsored by Johns Hopkins University.

But study data do not rule out the possibility of a continuum of regressive disorders in which epileptic encephalopathies underlie the process in some children who do not fit the general pattern, said Dr. Mantovani, medical director of the St. John's Mercy Child Development Center, St. Louis.

The evidence for an association between epileptic encephalopathy and autistic regression seems plausible based on the known prevalences with which regression, seizures, and epileptiform EEG activity occur in both children with autistic spectrum disorders (ASD) and those with Landau-Kleffner syndrome (LKS) and other regressive conditions in childhood.

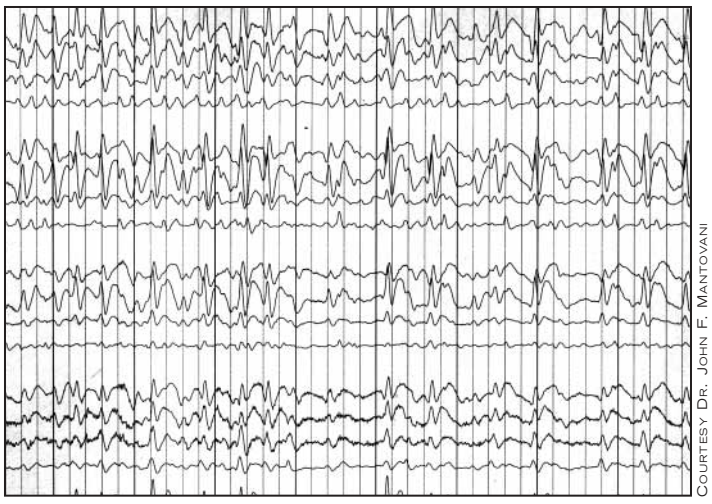
LKS usually occurs at 3-8 years of age in previously normal children; these children may have isolated language regression and variable behavioral abnormalities occurring with severely epileptiform EEGs and/or clinical seizures.

About 20%-40% of children with autism have experienced autistic regression. The prevalence of seizure history in children with ASD increases with age, so they have occurred in 20%-30% by 30 years of age.

Epileptiform activity on EEGs—which is highly associated with, but not specific for, epilepsy—also occurs in about 15% of children with ASD who do not have seizures during brief testing, and in up to 60% of these children during prolonged, overnight testing. It is also known that LKS is caused by epileptic encephalopathy even without the presence of clinical seizures, Dr. Mantovani said.

But reports have shown that children with autistic regression before 2-3 years of age are significantly less likely to have epileptic encephalopathy than are children who develop language regression alone at 2-3 years of age or older, which is similar to the pattern described for children with LKS. Clinical seizures and epileptiform EEGs occur more than twice as often in children with language regression alone, compared with those with the broader profile of autistic regression, said Dr. Mantovani, of the neurology and pediatrics departments at Washington University, St. Louis.

There is a lack of temporal correlation between seizures and ASD. People with ASD, especially the nonsyndromic or idiopathic type, tend to have few seizures at early ages but then start to have them at older ages. Yet autistic regression mostly occurs before 2 years of age. There also is a lack of EEG and clinical specificity to associate epileptic encephalography with autistic regression, because most children with autistic regression on overnight EEG testing do not have epileptiform activity and there is no clinical difference between children with autistic regression and those with nonregressive ASD. Dr. Mantovani said it is reasonable to get an overnight EEG for children who develop isolated language regression at ages older than 2 years, or global regression at ages older than 3 years. ■



COURTESY DR. JOHN F. MANTOVANI

An EEG obtained in an overnight sleep study shows the characteristic pattern of high voltage, frontally dominant constant spike and wave activity consistent with epileptic encephalopathy.