AED treatment of PS and rolandic epilepsy (RE). A clinical practice survey in the UK among 590 pediatricians who treat epilepsy found that 40% of 132 respondents reported non-treatment of PS and RE because of low frequency of seizures and parent/child preferences. They estimated 233 new cases of PS and 751 new RE cases, annually. Carbamazepine is the preferred older, and levetiracetam the preferred newer AED in randomized controlled trials [4].

References.
1. Berg AT, et al. Epilepsia. 2010 Apr;51(4):676-85.
2. Moseley B, et al. Epilepsy Behav. 2013 Mar;26(3):375-85.
3. Millichap JG, Lombroso CT, Lennox WG. Pediatrics. 1955 Jun;15(6):705-14.
4. Mellish LC, et al. Arch Dis Child. 2014 Sep 8. [Epub ahead of print].

KCNB1 MUTATIONS IN EPILEPTIC ENCEPHALOPATHY

Researchers at Scripps Research Institute, San Diego, and other centers in California; Johns Hopkins, Baltimore; Vanderbilt University, TN; and Northwestern University Feinberg School of Medicine, Chicago, IL; searched for de novo mutations in a family with a sporadic case of epileptic encephalopathy. The cause was determined using whole exome sequencing (WES) and whole genome sequencing (WGS). A de novo missense mutation in KCNB1 was identified that encodes the K2.1 voltage-gated potassium channel. Subsequently, 2 additional patients were identified with epileptic encephalopathy and de novo KCNB1 missense mutations that cause a similar pattern of K2.1 dysfunction. Clinical WES may be useful for diagnosis of epileptic encephalopathies of unknown etiology. (Torkamani A, Bersell K, et al. De novo KCNB1 mutations in epileptic encephalopathy. **Ann Neurol** 2014 Oct;76(4):529-40).

COMMENTARY. Researchers at University of Arizona, Tucson, previously explored the utility of WES and identified causal de novo variants in genes of 7 of 10 children with sporadic epilepsy, refractory seizures, developmental delay, or epileptic encephalopathy. These probands all presented with seizures within the first 6 months of life, and 6 have intractable seizures. The genes affected included SCN1A, CDKL5, EEF1A2, and KCNH5 [1]. The present finding of de novo KCNB1 mutations as a cause of K2.1 dysfunction expands the locus heterogeneity associated with epileptic encephalopathies [2].

References.
1. Veeramah KR, et al. Epilepsia. 2013 Jul;54(7):1270-81.
2. Torkamani A, et al. Ann Neurol. 2014 Oct;76(4):529-40.

TRANSITION CARE TO ADULT EPILEPSY CENTERS

Epileptologists at University of Toronto and University of Saskatchewan, Canada, evaluated the complexity of epilepsy patients transitioned from child to adult care between tertiary centers compared to patients transferred from the community. Patients aged from 18 to 25 years were divided into 2 groups: Group 1 comprised 170 patients referred from the pediatric tertiary center; and Group 2 had 132 patients referred from the

community. Retrospective comparison of patients in the 2 groups found that patients in Group 1 had earlier seizure onset, longer epilepsy duration, and more patients with symptomatic etiologies, epileptic encephalopathy, and cognitive delay ($p<0.001$). Group 1 patients required more referrals to other specialties, and more frequent treatment with polytherapy, epilepsy surgery, ketogenic diet, and vagus nerve stimulator ($p<0.001$). Adult neurologists (n=86) and pediatric neurologists (n=29) surveyed indicated that adult neurologists have lower levels of confidence in diagnosing and treating severe forms of childhood-onset epilepsies ($p<0.001$) and epilepsies associated with cognitive delay ($p<0.001$). (Borlot F, Tellez-Zenteno JF, Allen A, Ali A, Snead OC, Andrade DM. Epilepsy transition: Challenges of caring for adults with childhood-onset seizures. **Epilepsia** 2014 Oct;55(10):1659-66).

COMMENTARY. Patients from tertiary centers present more complex needs and require more resources than age-matched patients from the community, and adult neurologists feel less confident in diagnosing and treating adult patients transferred with some childhood-onset epilepsies. The researchers propose that adult epileptologists should receive training in childhood-onset epilepsies. Alternatively, pediatric epileptologists should continue to follow more complex childhood-onset epilepsies in the adult or pediatric epilepsy clinic, provided the patient is seen regularly by an internist to attend to general systemic needs.

The most challenging childhood-onset epilepsies for adult neurologists are the epileptic encephalopathies with intractable seizures and cognitive delay. The well-known syndromes include West, Dravet, and Lennox Gastaut. Patients with Lennox-Gastaut syndrome followed for >10 years have severe cognitive delay in 75-99% cases and refractory seizures in 92%.

References.

1. Camfield PR, Camfield CS. Pediatr Neurol. 2014 Jul;51(1):17-23.
2. Camfield PR, et al. Epilepsia. 2014 Aug;55 Suppl 3:24-8.

DIFFERENTIAL DIAGNOSIS OF LENNOX-GASTAUT SYNDROME

Epileptologists from Children's Hospital, Boston, and UCLA, California, discuss approaches to the differential diagnosis of Lennox Gastaut syndrome (LGS) and identification of a possible underlying etiology. The classic diagnostic criteria for LGS consist of a triad of features: multiple seizure types, abnormal EEG, and cognitive impairment. Onset is commonly between 3 and 5 years of age, with slight male preponderance, and is sometimes preceded by West syndrome. Tonic seizures during sleep are the classic feature used for diagnosis but in fact LGS has multiple concurrent seizure types: tonic, atypical absence, atonic, and myoclonic jerks. Nonconvulsive status epilepticus, lasting days to weeks, occurs in 50% patients, and sudden tonic or atonic falls, or "drop attacks," occur with the same frequency. The abnormal EEG shows slow spike-wave complexes (known originally as petit mal variant) at 2-2.5 Hz during wakefulness, and paroxysmal fast rhythms (10-20 Hz) during REM sleep. The MRI is abnormal in two thirds or more of patients with LGS. The IQ deteriorates over time, with 99% having cognitive delay by adolescence. Nonconvulsive status epilepticus is the most