autosomal dominant epilepsy with FS plus, generalized epilepsy with FS plus, and with severe myoclonic epilepsy in infancy that often begins with a prolonged FS. Various Na^+ channels and $GABA_A$ receptors in the brain are probably involved in the pathogenesis of frequent FS and also, in simple FS. Genetic linkage analyses have mapped FS to four loci, *FEB1,2,3 and 4,* in chromosomes 8q13, 19p, 2q23-q24, and 5q14-q15, respectively. (Hirose S, Mohney RP, Okada M et al. The genetics of febrile seizures and related epilepsy syndromes. Brain Dev August 2003;25:304-312). (Respond: Dr Shinichi Hirose, Department of Pediatrics, School of Medicine, Fukuoka University, 45-1, 7-chome Nanakuma, Jonanku, Fukuoka 814-0180, Japan).

COMMENT. The determination of the molecular genetic mechanism of FS may lead to more specific therapies. FS occur with increased frequency among family members of patients with FS. Tsuboi (1977) reported 17% of parents and 22% of siblings of FS probands affected; 30% of siblings are affected if one parent has a history of FS (Hauser WA. In: Febrile Seizures. Ed by Nelson and Ellenberg, Raven Press, 1981). An analysis of 2,109 patients with FS reported between 1948 and 1963 in 12 different publications showed a mean familial incidence of 17% (range 2 to 58%) (Millichap, 1968).

GENETICS OF SEVERE MYOCLONIC EPILEPSY OF INFANCY

The role of *SCN1A* gene mutations in the etiology of severe myoclonic epilepsy of infancy (SMEI) was investigated in 93 patients followed at the Hopital Saint Vincent de Paul, Paris, and other centers in France and Italy. *SCN1A* mutations occurred in 33 patients (35%). Parents of three patients (10%) who carried the inherited mutations were asymptomatic or had a milder form of epilepsy. Patients with the mutations had a greater frequency of unilateral motor seizures than those without and a more frequent family history of epilepsy. (Nabbout R, Gennaro E, Bernadina BD et al. Spectrum of *SCN1A* mutations in severe myoclonic epilepsy of infancy. Neurology June (2 of 2) 2003;60:1961-1967). (Reprints: Dr Rima Nabbout, Hopital Saint Vincent de Paul, Paris, France).

COMMENT. SMEI caused by *SCN1A* mutations is characterized by unilateral motor seizures, and 10% of cases are inherited from an asymptomatic or mildly affected parent. SMEI has a mean age of onset of 5 months, more than half the seizures are febrile, seizures are of all types, absence seizures with a myoclonic component are often triggered by photic stimulation, status epilepticus occurs in 75% and is often precipitated by fever, ataxia develops in >80%, psychomotor delay is common to all, and seizures are refractory to medication.

EARLY ONSET ABSENCE EPILEPSY

Neuropsychological evaluations, behavior patterns, and outcomes were studied in a retrospective analysis of 10 patients (7 girls, 3 boys) with onset of absence epilepsy before age 3 years, referred between 1986 and 2002 at Hopital Henri Gastaut, Marseille, and Hopital des Enfants, Toulouse, France. At >2 years of follow-up (range, 2 years 8 months to 9 years 4 months), only two had typical absences with good seizure control and good cognitive outcome. In the remaining 8 cases, 8 had cognitive delays and behavioral

difficulties, and 5 had persisting absence seizures, some with asymmetric interictal EEGs. None had abnormal neurologic signs, and neuroimaging was unremarkable. (Chaix Y, Daquin G, Monteiro F et al. Absence epilepsy with onset before age three years: a heterogeneous and often severe condition. Epilepsia July 2003;44:944-949). (Respond: Dr Yves Chaix, Centre Saint-Paul, Hopital Henri Gastaut, Marseille, France).

COMMENT. Early onset absence epilepsy is a rare heterogeneous condition with a generally poor outcome.

TEMPORAL LOBECTOMY FOR EPILEPSY WITH CONGENITAL PORENCEPHALY AND HIPPOCAMPAL SCLEROSIS

The clinical characteristics and surgical outcome of 6 patients with intractable epilepsy and coexisting extratemporal porencephaly and hippocampal sclerosis are presented from the University of Alabama, Birmingham, AL. Of 24 patients with congenital porencephaly and epilepsy, 6 had a temporal lobe epileptogenic focus, and temporal lobe resection was performed in 5. The mean age at surgery was 31 years (range, 15-42 years), and the time from onset of epilepsy was 27 years (range, 14-41 years). Mean age of seizure onset was 4.3 years (range, 6 months to 10 years). Porencephalic cyst volume was 11% of total intracranial volume (range, 1% to 32%). Freedom from seizures occurred in all five patients, at mean follow-up of 47 months (range, 22-67 months). Antiepileptic drug therapy was continued but at lower doses. Hippocampal sclerosis was confirmed histopathologically. Children with congenital porencephaly and intractable epilepsy should be evaluated early, and temporal lobectomy should be considered if clinical, MRI, and EEG findings indicate a temporal lobe origin for seizures. (Burneo JG, Faught E, Knowlton RC et al. Temporal lobectomy in congenital porencephaly associated with hippocampal sclerosis. Arch Neurol June 2003;60:830-834). (Reprints: Ruben Kuzniecky MD, University of Alabama at Birmingham Epilepsy Center, Civitan International Research Center (CIRC) 312, 1719 Sixth Ave S, Birmingham, AL 35294).

COMMENT. A common ischemic cause for the congenital porencephalic cyst and hippocampal sclerosis is postulated, involving perinatal occlusion of the posterior cerebral artery. The most frequent origin of seizures associated with the dual pathology is the temporal lobe. The selective resection of the temporal epileptogenic focus might be considered as an alternative to hemispherectomy, which carries a higher morbidity in these patients.

HEADACHE DISORDERS

QUALITY OF LIFE IN CHILDHOOD MIGRAINE

The Pediatric Quality of Life Inventory (PQLI), Version 4.0 and a standardized headache assessment were completed by children and parents, in a survey of 572 consecutive patients (mean age, 11.4 +/- 3.6 years) who presented with headaches at the Cincinnati Children's Hospital Medical Center, Ohio. Most (99%) had a clinical diagnosis