Wahringergurtel 18-20 A-1090 Vienna,Austria).

COMMENT. Plasma pipecolic acid determination may aid in diagnosis, and a persistent elevation may avoid premature pyridoxine withdrawal in infants with pyridoxine-dependent epilepsy (normal range, 0.7-2.6 mcmol/L). The authors report an additional case of this syndrome in a 7 year-old child.

Pipecolic acid elevation in pyridoxine-dependent epilepsy may be caused by a deficiency of pyridoxal-dependent a-aminoadipic acid transaminase, a step in the degradation of lysine to acetoacetyl-CoA in the brain. Pipecolic acid also accumulates in peroxisomal disorders, such as Zellweger syndrome.

AUTOSOMAL DOMINANT PARTIAL EPILEPSIES

The clinical, electrophysiologic, and genetic characteristics of autosomal dominant partial epilepsy were studied in 71 patients and 33 non-epileptic at-risk family members in 19 European families followed at the Hopital Universitaire de Geneve, Switzerland, and centers in Strasbourg, Paris, Grenoble, Nice, Marseille, Rome, and Pisa. Families were subdivided into those with autosomal dominant nocturnal frontal lobe epilepsy (ADNFLE) (n = 8), familial temporal lobe epilepsy (7), and autosomal dominant partial epilepsy (4).

Familial partial epilepsies show great intrafamilial variability, and up to 30% may be resistant to antiepileptic medication. Some affected family members may have only EEG abnormalities, without clinical seizures, reflecting incomplete penetrance. Genetic studies found no mutation in the a4 and B2 nicotinic acethylcholine receptor subunits, but positive lod scores were obtained in 4 families with markers from the candidate region on chromosome 10q. (Picard F, Baulac S, Kahane P et al. Dominant partial epilepsies. A clinical, electrophysiological and genetic study of 19 European families. Brain June 2000;123:1247-1262). (Respond: Dr Fabienne Picard, EEG, Department of Neurology, Hopital Universitaire de Geneve, 24, rue Micheli-du-Crest, 1211 Geneva 14, Switzerland).

COMMENT. Familial partial epilepsies may originate in frontal, temporal, and other variable locations. Clinical and surface EEG findings may provide conflicting localizing evidence, and an overlap of partial epilepsy syndromes within families may occur, with genetic heterogeneity.

Frontal lobe origin of absence seizures is discussed by Pavone A and Niedermeyer E (Clin Electroencephalogr July 2000;31:153-156). The distinction between partial and absence seizures of frontal lobe origin is important from a therapeutic standpoint. Carbamazepine is the agent of choice for partial, and valproate for absence seizures.

HHV-6, INFANTILE SPASMS, AND CEREBELLAR ASTOCYTOMA

A case of human herpesvirus-6 encephalitis, carditis, infantile spasms, and a subsequent cerebellar astrocytoma containing the HHV-6 genome, is reported from the University of Oulu, Finland. A 14-month-old girl presented with fever, hypotonia, and a diffuse urticarial exanthem. After admission, she developed encephalitis and status epilepticus, followed by myocarditis. MRI of the brain showed thin subdural effusions but no tumor. After 11 weeks from onset, convulsions changed to infantile spasms, resistant to vigabatrin and ACTH. A repeat MRI at 11 months after the primary illness showed a cystic astrocytoma located near the vermis. Five months after surgical removal of the tumor, the patient has hypotonia, poor social communication, and daily infantile spasms. HHV-6 DNA was detected by PCR in the tumor tissue. (Rantala H, Mannonen L,