the same family. An autoimmune etiology is suggested. (Dauvilliers Y, Mayer G, Lecendreux M et al. Kleine-Levin syndrome: an autoimmune hypothesis based on clinical and genetic analyses. Neurology December (1 of 2) 2002;59:1739-1745). (Reprints: Dr Mehdi Tafti, HUG, Belle-Idee, Biochemistry and Genetics, Chemin du Petit-Bel-Air, 2, CH-1225 Chene-Bourg, Switzerland).

COMMENT. The etiology of Kleine-Levin syndrome is unknown. Periodic hypothalamic dysfunction, focal encephalitis, and an autoimmune process are possibilities. The disorder is rarely familial, only in 2 of 30 patients in the above study, and in 2 siblings in the following report. Levels of CSF orexin (hypocretin), decreased in some narcoleptic patients, were normal in the following familial cases.

Familial Kleine-Levin syndrome is reported in 2 siblings with unusually long hypersomnic spells, from Tufts University, Boston, MA (Katz JD, Ropper AH. Arch Neurol Dec 2002;59:1959-1961). Both patients shared the HLA-DR2, DQ1, and DR5 haplotypes. Treatment with lithium, methylphenidate, modafinil, clonazepam, and other drugs was ineffective in preventing or reducing the duration of hypersomnic spells. Patient 1, a 15-year-old boy, developed flu-like symptoms without fever, he slept excessively for 9 days, and was irritable when awake. Eight similar attacks occurred during the following 10 months, and 16 hypersomnic spells in 5 years. Compulsive symptoms followed the episodes. Patient 2, the sister of patient 1, began having hypersomnic spells at age 13 years, the first preceded by a flu-like illness. Between attacks, MRI, EEG, Ebstein-Barr virus titer, ESR, and CSF were normal.

VASCULAR DISORDERS

TIME LAG TO DIAGNOSIS OF STROKE

Time to diagnosis was determined in 41 children, 0 to 18 years, with ischemic or hemorrhagic stroke documented prospectively or by retrospective chart review of the last 2 years database, at State University of New York, Stony Brook, NY. Twelve neonates diagnosed in the neonatal period were excluded from analysis. Of the remaining 29 (mean age at presentation 8.67 years), 24 had accurate time records, and 28 events were recorded. Ischemic stroke occurred in 21 events and hemorrhagic stroke in 7. The cause was idiopathic in 18 (46%) and embolic in 4 (surgery and atrial myxoma); vascular malformation was present in 3, arterial dissection in 2, inherited coagulopathy in 3, and moyamoya or other syndrome in 4. An underlying known disorder was found in 38%, and a predisposing disorder in an additional 15%. Initial symptoms were headache in 32%, motor in 60%, sensory in 7%, aphasia in 14%, seizures in 10%, and mental status change in 21%. Time from clinical onset to first medical exam was an average of 28.5 hours, and time to diagnosis of stroke averaged 35.7 hours. Unless hemorrhagic, stroke in children is rarely diagnosed in <3 to 6 hours from onset, a major criterion for inclusion in therapeutic trials of thrombolytic or neuroprotective agents. (Gabis LV, Yangala R, Lenn NJ. Time lag to diagnosis of stroke in children. Pediatrics November 2002;110:924-928). (Reprints: Lidia V Gabis MD, Division of Developmental Disabilities, South Campus, Putnam Hall, State University of New York, Stony Brook, NY 11794).

COMMENT. Stroke with massive hemorrhage demands immediate medical attention. Ischemic stroke is less dramatic and medical attention is delayed. Compared to the 35 hour delay in children, stroke in adults is treated as an

emergency and the total delay time, symptom onset until CT completion, is only 4 hours (range 2.3-8.3 hours) (Morris DL et al. 2000). Reasons for excessive diagnostic delay in children include their inability to describe symptoms such as headache, sensory or cerebellar symptoms, and difficulty in recognition of some neurologic signs, eg aphasia. Increased awareness of predisposing causes (eg cardiac, sickle cell disease) should lead to earlier diagnosis and treatment.

Risks of posterior circulation stroke are evaluated in 22 cases (17 boys) identified in a retrospective study at University College and Great Ormond Street Hospitals, London, UK. (Ganesan V, Chong WK, Cox TC et al. Neurology Nov (2 of 2) 2002;59:1552-1556). Vertebrobasilar arterial abnormalities (vertebral artery dissection in 10) were present in 20, multifocal in 12. Cardiac abnormalities with embolism were present in only 4, hypertension in 9, and factor V Leiden and other gene mutations in 6. Two had subluxation of the upper cervical spine. During follow-up for 6 months to 11 years, 5 (20%) had recurrence and 7 had TIA; 12 (50%) had no residual deficits. Investigations include MRI, cerebral angiography, echocardiography, and cervical radiography. In contrast to anterior circulation stroke, where 50% have a preexisting disorder, the majority with posterior circulation stroke are previously healthy.

OUTCOME OF HEMISPHERECTOMY IN STURGE-WEBER SYNDROME

A questionnaire was mailed to the parents of 70 patients identified by the Sturge-Weber Foundation as recipients of hemispherectomy between 1979 and 2001, and responses obtained from 32 (46%) were analysed at the Johns Hopkins Hospital, Baltimore, MD. The mean age at onset of seizures was 4 months, and the median age at surgery was 1.2 years. Eighty one percent were seizure free, and anticonvulsants were discontinued in 53%. Seizure control was not related to the age at seizure onset, but it did correlate with an older age at operation. Six with continued seizures were operated on at 1.3 years, and in 26 currently seizure-free operation was delayed until age 3.1 years (p=0.05). Hemiparesis was not worsened following surgery. Cognitively, 2 were normal, 10 had mild learning impairment, 14 were moderately learning disabled, and 6 were severely disabled. Cognitive outcome was not related to age at operation or seizure control. (Kossoff EH, Buck C, Freeman JM. Outcomes of 32 hemispherectomies for Sturge-Weber syndrome worldwide. Neurology Dec (1 of 2) 2002;59:1735-1738). (Reprints: Dr Eric H Kossoff, Department of Pediatric Neurology, The Johns Hopkins Hospital, 600 N Wolfe St, Jeffedrson 128, Baltimore, MD 21287).

COMMENT. The main benefit of hemispherectomy in this population is improved seizure control, and the likelihood of seizure freedom is higher when operation is delayed until the child is older.

CHOROID PLEXUS A-V MALFORMATION PRESENTING WITH IVH

A term infant presenting on the second day with apnea and decerebrate posturing had an intraventricular hemorrhage (IVH), the result of an arteriovenous malformation of the choroid plexus demonstrated angiographically on the 3rd day and reported from Johns Hopkins Hospital. The AVM was not demonstrated by ultrasonogram and MR angiogram. Hydrocephalus was managed by acetazolamide and ventricular taps, and a ventriculoperitoneal shunt was placed at 6 weeks. Definitive therapy is delayed until after 1 year. (Heck DV, Gailloud P, Cohen HL et al. Choroid plexus arteriovenous malformation presenting with intraventricular hemorrhage. J Pediatr Nov 2002;141:710-711). (Reprints: Donald V Heck MD, Johns Hopkins Med Ctr, 600 N Wolfe St, Baltimore, MD 21287).