Hospital Groningen, The Netherlands. The proband, a 20 year old woman, had an onset of attacks of 'swinging legs', dizziness, involuntary jerky limb movements, dysarthria, and gait ataxia beginning at 6 years of age. Attacks up to four times daily and lasting 10 seconds to 5 min occurred at rest, during exercise, or when startled, standing up, or running. Myokymia of the hands and semirhythmical movements of fingers were noted on examination, and EMG showed myokymic discharges. An attack provoked by knee bends consisted of rhythmic involuntary shaking of limbs, and tremor and dysmetria on finger-to-nose test. Acetazolamide prevented attacks, but treatment was limited by paraesthesiae and development of tolerance. A 22 year old brother was also affected from 6 years of age, and the myokymia and ataxia were complicated by paroxysmal kinesigenic dystonia at 15 years, after a mild head injury. Another family member also had attacks of paroxysmal choreoathetosis. Carbamazepine controlled the attacks of dystonia and choreoathetosis but not the ataxia. Data on 6 affected family members are tabulated. (Lubbers WJ, Brunt ERP, et al. Hereditary myokymia and paroxysmal ataxia linked to chromosome 12 is responsive to acetazolamide. J Neurol Neurosurg Psychiatry October 1995;59:400-405). (Respond: Dr ERP Brunt, Department of Neurology, University Hospital Groningen, PO Box 30.001, 9700 RB Groningen, The Netherlands).

COMMENT. Provisional diagnoses of basilar migraine and epilepsy had been made initially in two of three children with familial paroxysmal ataxia reported in the UK (Hawkes CH, 1992). The EEG was normal and the MRI showed atrophy of the superior cerebellar vermis. All responded to acetazolamide. (see Progress in Pediatric Neurology II, PNB Publ, 1994, p149-150). MRI findings were not reported in the above series. An overlap and relation between paroxysmal types of ataxia, myokymia, choreoathetosis and dystonia is strengthened by this report, although different responses to acetazolamide and carbamazepine may suggest separate etiologies.

SEIZURE DISORDERS

HEREDITARY ROLANDIC EPILEPSY AND SPEECH DYSPRAXIA

A syndrome of nocturnal oro-facio-brachial partial seizures, secondarily generalized partial seizures, centro-temporal epileptiform discharges, associated with oral and speech dyspraxia and cognitive impairment, is described in a family of 9 affected members in three generations reported from Austin Hospital, Heidelberg (Melbourne), the University of Melbourne, and the Royal Children's Hospital, Melbourne, Australia. All affected individuals had nocturnal rolandic seizures limited to midchildhood. Inheritance of epilepsy and speech dyspraxia was autosomal dominant, with 100% penetrance for speech dyspraxia. Clinical anticipation was noted across the three generations of affected individuals, with increasingly severe speech dyspraxia, epilepsy as well as cognitive impairment. The syndrome has features resembling benign rolandic epilepsy (BRE) and may help in identifying the gene for BRE which is unassociated with clinical anticipation. It differs from the syndromes of Landau-Kleffner and epilepsy with continuous spike and wave during slow-wave sleep. (Scheffer IE et al. Autosomal dominant rolandic epilepsy and speech dyspraxia: a new syndrome with anticipation. Ann Neurol October 1995;38:633-642).

(Respond: Dr Scheffer, Department of Neurology, Austin Hospital, Heidelberg (Melbourne), Victoria, Australia).

COMMENT. The authors suggest that this new syndrome, with its known genetic basis, may help to clarify the relationship between benign rolandic epilepsy, a benign syndrome, and Landau-Kleffner and CSWSS, more severe syndromes.

Symptoms and findings in the Landau-Kleffner (LKS) and continuous spike-and-wave during slow sleep (CSWSS) syndromes are compared in a report from the Department of Child Neurology, University of Helsinki, Children's Castle Hospital, Helsinki, Finland. (Granstrom M-L et al. Epilepsia 1995;36(suppl 4):123). Bilateral epileptiform activity was found in sleep EEGs in 5 of 6 LKS children and in all 11 children with CSWSS. All LKS children had auditory agnosia and deterioration of expressive language, 4 had attention deficit disorders, and 2 became clumsy or ataxic. Six children with CSWSS had deterioration of expressive language, motor skills and general intelligence, and 4 had hyperkinesia and delayed development. Epileptic seizures occurred in all LKS and in 8 CSWSS children. Mean age at diagnosis was 5 years for LKS and 6 and 1/2 years for CSWSS. MRI/CT was abnormal in 1 LKS and 5 CSWSS patients. LKS usually affects previously normal children whereas CSWSS occurs in children with pre- or perinatal pathology and previously abnormal development. An overnight EEG is recommended in children with developmental arrest, loss of speech, and/or major behavioral problems. Four additional papers on Landau-Kleffner syndrome will be presented at the Annual Meeting of the American Epilepsy Society, Baltimore, Dec 1-6, 1995.

VALPROATE, BRAIN ATROPHY AND REVERSIBLE DEMENTIA

Two children who developed severe cognitive and behavioral deterioration while being treated with sodium valproate for idiopathic epilepsy are reported from the Miami Children's Hospital, Miami, FL. Patient 1 presented at age 5 years with left focal seizures and right central sharp waves on the EEG, consistent with benign rolandic epilepsy (BRE). After his fourth seizure at age 8 years he received sodium valproate (Depakote), with blood levels of 91-106 mg/dl, and methylphenidate (MPH) 20 mg twice daily for an associated ADDH. MPH was replaced by thioridazine (Mellaril) and benztropine (Cogentin) at age 9 years. Impaired motor ability was noted at age 10 years. Activity level, speech, and IQ progressively deteriorated, with ataxia and marked obesity developing by 10 years 8 months. MRI showed enlarged ventricles and cortical sulci. Felbamate was substituted for valproate and his motor activity, speech, gait, weight, and IQ returned to normal within a few weeks. The MRI improved after 3 months and was normal at 1 year. Pemoline (Cylert) was introduced for ADDH without relapse of behavior or seizure recurrence. Patient 2 had an onset of a similar degenerative syndrome within 3 weeks of prescribing valproate for migraine and BRE. The condition resolved 4 to 6 months after substituting phenobarbital. No metabolic changes were uncovered. (Papazian O et al. Reversible dementia and apparent brain atrophy during valproate therapy. Ann Neurol October 1995;38:687-691). (Respond: Dr Papazian, 3200 SW 60th Ct, #302, Miami, FL 33155).

COMMENT. Patients treated with valproate need to be monitored for mental and motor deterioration in addition to liver dysfunction.