COMMENT. Chronic use of phenytoin did not result in cognitive deterioration in young adults with well controlled seizures with onset after 16 years of age. Previous reports suggesting PHT-related cognitive deterioration were complicated by poor seizure control and were limited by less sophisticated tests than those used in the above study. Mikati M et al. at the Department of Neurology, Children's Hospital, Harvard Medical School, have found significant differences in potency and plasma concentrations of brand name PHT and generic PHT monotherapy (Epilepsia March/April 1992; 33:359-365). Variability in capsule content may be an important factor to be considered in trials of medications such as PHT that manifest nonlinear pharmacokinetics.

FAMILIAL PAROXYSMAL ATAXIA: ACETAZOLAMIDE THERAPY

A dramatic response to acetazolamide in 3 patients with familial paroxysmal ataxia is reported from the Ipswich Hospital, Neurological Centre, Suffolk, England. Case 1 developed mild squint at 3 years and nystagmus at 6 years. Subsequently she experienced episodes of dysarthria, ataxia and vertigo often accompanied by nausea and vomiting and followed by headache and drowsiness. The attacks lasted from 2 to 24 hours and occurred every 2 weeks. When assessed at 26 years of age the attacks were occurring up to 5 times a week. The EEG was normal and an MRI showed atrophy of the superior cerebellar vermis. Acetazolamide 250 mg bid reduced attacks dramatically over an 8 month observation period. Cases 2 and 3, sons of Case 1, developed identical episodes at 1 year and 6 weeks of age, respectively. Both responded to acetazolamide. The father of the index case was ataxic and then chairbound for 13 years and died at age 67. This family was the first described in the U.K. but many may be mislabeled as epilepsy or migraine (Hawkes CH Familial paroxysmal ataxia: report of a family. J Neur Neurosurg and Psych March 1992; 55:212-213.) (Correspondence: Dr. CH Hawkes, Ipswich Hospital, Neurological Centre, Heath Road, Ipswich, Suffolk IP4 5PD, England.)

COMMENT. Provisional diagnoses of basilar migraine and epilepsy had been made initially in 2 of these cases. The accurate diagnosis in 1 individual will often reveal similarly affected family members. Inheritance is autosomal dominant. Acetazolamide is also effective in the treatment of paroxysmal dystonia (tonic seizures) as a presenting manifestation of multiple sclerosis (Sethi KD et al. Neurology April 1992; 42:919-921). Koller W et al. have used acetazolamide successfully in the treatment of essential tremor (Neurology April 1992; 42(Suppl 3):322).

LANDAU-KLEFFNER SYNDROME: LONG-TERM PROGNOSIS

The long-term follow up of 6 patients and a review of the recent literature on the Landau-Kleffner syndrome ("acquired aphasia with convulsive disorder") are reported from the Department of Neurology, University Hospital Dijkzigt-Rotterdam, the Netherlands. The age at onset was 3-5½ years, with epilepsy as the first sign in 3 and comprehension deficit in 3.

Seizures were absence, generalized tonic-clonic, partial and complex partial in patterns. Seizures had disappeared at follow-up in 3 and epileptic activity in the EEG was absent in 4. The course of the aphasia was linked to the appearance and disappearance of electrical status epilepticus during slow sleep (ESES) which was found in 3 patients. Lasting EEG abnormalities and persisting clinical seizures, combined with the presence and duration of ESES, may impede language recovery. The disappearance of ESES may be associated with an improvement of language functions. The course of the aphasia was variable: slow and continuous improvement in 3, rapid recovery in 1, fluctuating in 1, and deterioration followed by slow improvement in 1. The follow-up period varied from 3 to 19 years. The response of the seizures and aphasia to anticonvulsants was variable; some benefit was obtained with phenytoin and phenobarbital in 1 patient, ethosuximide and valproate were helpful in 2. Prednisone in 1 patient had a variable and inconclusive effect (Paquier PF, Van Dongen HR, Loonen MCB. The Landau-Kleffner Syndrome or "acquired aphasia with convulsive disorder" Arch Neurol April 1992; 49:354-359). (Reprints: Dr. Van Dongen, Department of Neurology, Room EE 2287, University Hospital Dijkzigt-Rotterdam, P.O. Box 1738, 3000 DR Rotterdam, the Netherlands.)

COMMENT. The correlation between ESES and acquired epileptic aphasia has been emphasized previously (Jayakar PB, Seshia SS. J Clin Neurophysiol 1991; 7:299-311; Deonna TW. J Clin Neurophysiol 1991; 7:288-298). See also Ped Neur Briefs Sept 1991; 5:71; Jan 1991; 5:4-5). The beneficial effects of ACTH and cortico steroids have been reported from Tel Aviv University (Lerman P et al. See Ped Neur Briefs April 1991; 5:28-29). Cortico steroids administered early were more effective than delayed treatments.

MOVEMENT DISORDERS

PROGNOSIS OF TOURETTE'S SYNDROME

A retrospective study of 58 adults with a diagnosis of Gilles de la Tourette's syndrome (TS) during childhood is reported from the Department of Neurological Sciences, Rush-Presbyterian-St. Luke's Medical Center, Chicago, IL. The 41 men and 17 women had a mean current age of 27.1 years and a mean age of tic onset of 6.9 years. Forty six percent had a first degree relative with tics. All patients had tics as adults and most had both motor and phonic tics. Coprolalia occurred in 4% of adults. No patient experienced extreme adult disability and tics caused minimal to mild intrusion into private and professional life. All had received specialized medical care for TS in childhood, but only 26% were currently seeing a physician for tic disorders. Maximal disability had occurred during adolescence in most subjects, but 98% graduated high school, and 90% were full time students or fully employed. Childhood tic severity and coprolalia during development had no predictive value for risk of adult moderate/severe tics. Mild tic severity in adulthood correlated with mild tics during adolescence. (Goetz CG et al. Adult tics in Gilles de la Tourette's syndrome: description and risk factors. Neurology