COMMENT. Congenital central hypoventilation syndrome, characterized by absent ventilatory and arousal sensitivity to hypercapnia and variable sensitivity to hypoxemia, has been reported with Hirschsprung disease, neural-crest tumors, and mild cerebral atrophy on CT or MRI. Many require ventilatory support particularly while asleep. Seizures and neurodevelopmental delays may bring these patients to the attention of the pediatric neurologist.

The effects of phenobarbital on sleep behavior in children with febrile seizures were evaluated by Hirtz DG, Farwell JR et al. at Bethesda, MD and Seattle, WA (Neurology April 1992; 42(Suppl 3):367). Night awakenings were not more common in children on phenobarbital except for those who were poor sleepers initially. Lengthy night awakenings were not more frequent and total sleep time was not altered. At least one behavioral disturbance previously assigned to the use of phenobarbital in the prophylaxis of febrile seizures has been negated by this study.

Horne J from the Sleep Research Laboratory, Loughborough University, Leicestershire, U.K. discusses epilepsy in sleep in a paper on sleep and its disorders in children (J Child Psychol Psychiat March 1992; 33:473-487). In addition to febrile convulsions and benign rolandic childhood epilepsy, other paroxysmal disorders that occur during sleep include the complex partial seizures of frontal origin, usually misdiagnosed as nightmares (Stores G, 1991) and sometimes referred to as "nocturnal paroxysmal dystonia" (Lugaresi et al. 1986).

PAROXYSMAL DISORDERS

PHENYTOIN AND NEUROPSYCHOLOGICAL PERFORMANCE

A comprehensive neuropsychological assessment of young adults (mean age 32) before and after treatment with phenytoin (PHT) for a 5 year study period is reported from the Regional Epilepsy Center, University of Washington School of Medicine, Seattle, Washington. In 3 groups of 11 patients each, the following medications were not changed and the median number of seizures per subject was 2: (a) PHT monotherapy; (b) PHT and other drugs; (c) drug regimens excluding PHT. Of 20 neuropsychological variables, none showed losses and 3 showed statistically significant improvements (Trail Making Part B, WAIS Performance IQ, WAIS Full-Scale IQ). The mental abilities of the groups were similar to each other at both the beginning and the end of the study. With normal therapeutic serum levels and well controlled seizures there was no evidence for any cognitive losses in patients treated with phenytoin over extended periods. (Dodrill CB, Wilensky AJ.Neuropsychological abilities before and after 5 years of stable antiepileptic drug therapy. Epilepsia March/April 1992; 33:327-334.) (Reprints: Dr. C.B. Dodrill, Regional Epilepsy Center, Harborview Medical Center, 325 Ninth Avenue, Seattle, WA 98104.)

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.