months in 4 patients and 6 years in one, without apparent development of drug tolerance. (Fink JK et al. Dystonia with marked diurnal variation associated with biopterin deficiency. Neurology May 1988;38:707-711).

COMMENT: The familial cases described here are similar to those with hereditary progressive dystonia reported by Segawa et al and characterized by diurnal variation and extreme sensitivity to levodopa. (Adv Neurol 1976;14:215). Response to levodopa is generally seen in only 10% of patients with dystonia.

THERAPY FOR TOURETTE SYNDROME

A 12 year-old male with Tourette Syndrome whose symptoms improved with gum chewing is reported from the Division of Pediatric Neurology, Hahnemann University, Philadelphia, PA. With onset at 3 years of age, symptoms had included blinking, head nodding, neck twisting, shoulder shrugs, snorting, sniffing and blowing, and coprolalia. Haloperidol controlled tics but was associated with excessive drowsiness. Clonidine was of some benefit. After chewing gum, using 2-3 sticks to make a large wad, the urge to move and vocalize was decreased and jaw movements made noise-making more difficult. The author recommends gum chewing as an adjunct therapy in Tourette Syndrome to decrease stress, facial movements, and particularly vocalizations. (Brill CB. Gum chewing as therapy for Tourette syndrome. Pediatr Neurol April 1988;4:128).

COMMENT: Provided that teachers and parents are tolerant of gum chewing, this form of therapy should be recommended. The prevalence of Tourette syndrome was discussed in a previous issue (Ped Neur Briefs March 1988;2:24). Tics were controlled by haloperidol in 13 of 18 children requiring treatment; the disorder was mild and treatment was unnecessary in 23 (56%) patients (Neurology 1988;88:472).

LESCH-NYHAN SYNDROME

Five boys with Lesch-Nyhan syndrome and varying degrees of dystonia, chorea, spasticity, ataxia, dysarthria, and mental retardation were studied at the Depts of Neurology and Medicine, Baylor College of Medicine, and Depts of Neurology and Pediatrics, University of Texas Health Science Center, Houston, TX. Four showed reduction of homovanillic acid (HVA) in the CSF and all had low CSF phenethylene glycol, indicating reducee dopamine and norepinephrine turnover. Three children had high CSF 5-hydroxyindoleacetic acid (HIAA), suggesting increased serotonin turnover. The patient with the most severe chorea had the lowest CSF HVA value, whereas the patient with the least amount of self-mutilation had the highest CSF 5HIAA. Only one patient improved with carbidopa-levodopa, whereas all 5 showed some lessening of self-mutilatory or hyperkinetic behavior in response to tetrabenazine, a monoamine-depleting agent. The study was thought to support the theory of abnormal central monoamine metabolism in Lesch-Nyhan syndrome. (Jankovic J et al. Lesch-Nyhan syndrome: A study of motor behavior and cerebrospinal fluid neurotransmitters. Ann Neurol May 1988;23:466-469).

COMMENT: Lesch-Nyhan syndrome is an X-linked recessive disorder