HEADACHE

ENVIRONMENTAL FACTORS IN HEADACHE

The environmental conditions related to headache in 38 children are reviewed from the Illinois Institute of Technology and the Franciscan Children's Hospital and Rehabilitation Center, Boston, MA. Parents filled out the Children's Headache Assessment Scale (CHAS) at an evaluation for behavioral medicine treatment and the parent rating questionnaire was completed again after therapy. The frequency of specific CHAS categories (stress antecedents, physical antecedents, attention consequences, escape consequences, coping responses, and medication use) varied widely and were changed by behavioral medicine treatment. There was less interference of headache with school attendance and improved coping responses after treatment. (Budd KS, Kedesdy JH. Investigation of environmental factors in pediatric headache. Headache October 1989; 29:569-573).

> COMMENT. The Children's Headache Assessment Scale focuses on situations and events surrounding the headaches rather than questioning parents about the symptoms. This study shows that the parent rating questionnaire is of value in behavioral assessment of pediatric headache and in following responses to behavioral treatment.

NARCOLEPSY

SYMPTOMATIC NARCOLEPSY WITH DIENCEPHALIC LESIONS

Three patients with symptomatic narcolepsy are reported from the Departments of Neurology and Psychiatry, University of Michigan Medical Center, Ann Arbor, MI. One was a girl who developed polyphagia, weight gain, decreased growth, headaches with visual blurring, and excessive daytime sleepiness with frequent, irresistible brief naps at age 7½ years. At age nine, she developed hyperprolactinemia and galactorrhea. At age 11 she had diabetic ketoacidosis, temperature dysregulation, hypothalamic hypothyroidism, partial diabetes insipidus, and hepatitis. The CT was normal. MRI showed mild diffuse brain substance loss. Following treatment with methylphenidate, sleepiness and irresistible sleep attacks improved. Tissue typing was positive for HLA-DR2 and HLA-DQw1. The authors refer to ten additional cases of symptomatic narcolepsy with documented brain lesions reported in the literature. (Aldrich, MS, Naylor, MW. Narcolepsy associated with lesions of the diencephalon. Neurology November 1989; 39:1505-1508).

> COMMENT. In these cases the REM sleep abnormalities and excessive daytime sleepiness were documented by polysomnography and Multiple Sleep Latency Tests. Two of the three patients were HLA-DR2 positive while the third was negative. In an addendum, the authors note an additional report of symptomatic narcolepsy in a nine year old HLA-DR2-positive boy following removal of a craniopharyngioma (Kowatch RA, et al. Sleep Res 1989; 18:250). Other brain lesions associated with narcolepsy have included

midbrain glioblastoma, cerebral sarcoidosis, pontine infarcts, 3rd ventricle glioma, pituitary adenoma, 3rd ventricle colloid cyst, multiple sclerosis, encephalitis, ischemia, head trauma, as well as craniopharyngioma.

MUSCLE DISORDERS

SLEEP BREATHING PATTERNS AND MUSCULAR DYSTROPHY

The breathing patterns and $HbSaO_2$ changes during nocturnal sleep were monitored in 11 chair-bound Duchenne muscular dystrophy patients at the Institutes of Neurology and Respiratory Diseases, University of Pavia, Italy. Nocturnal sleep had no significant adverse effects on the nighttime polygraphic sleep recordings and respiration. Infrequent central apneas occurring in six patients were associated with falls in $HbSaO_2$ greater than normal and correlated with functional residual capacity values. The blood oxygen balance was relatively preserved but unstable during nocturnal non-REM and REM sleep in patients with Duchenne muscular dystrophy, mean age 15 years (range 10-21) even in advanced stages of the illness. (Manni R et al. Breathing patterns and $HbSaO_2$ changes during nocturnal sleep in patients with Duchenne muscular dystrophy. J Neurol October 1989; 236:391-394).

COMMENT. Acute respiratory failure is an important factor contributing to death in most patients with Duchenne muscular dystrophy. During the later stages of the illness, a restrictive lung disease related to the progressive inspiratory muscle weakness and rib cage deformities develops. In this study, nocturnal sleep did not seem to have a significant adverse effect on respiration and no pathological breathing patterns were observed. Unimpaired diaphragmatic function might account for the relatively preserved arterial oxyhemoglobin desaturation during REM sleep in the population studied.

BECKER'S DYSTROPHY

Two brothers affected with Becker's muscular dystrophy in whom the disease followed completely different courses are reported from the Departments of Neurology/Neurosurgery and Genetics, Washington University Medical School, St. Louis, MO. The oldest sibling died at 37 following many years of severe disability whereas the younger sibling, now 26, has normal muscle strength. Symptoms began between 10 and 12 years of age in both patients. Analysis of the DNA from each revealed a similar deletion at the 5' end of the dystrophin gene. The younger brother had epilepsy from age 13 and had been treated with phenytoin continuously for 13 years. (Medori R, Brooke MH, Waterston RH. Two dissimilar brothers with Becker's dystrophy have an identical genetic defect. Neurology November 1989; 39:1493-1496).

COMMENT. The long term treatment with phenytoin from the onset of the muscle symptoms may have influenced the clinical course of the younger brother. The authors suggest that the action of a membrane stabilizer such as phenytoin may prevent the degeneration of the muscle fibers lacking dystrophin.