ATAXIA SYNDROMES

ACUTE CEREBELLAR ATAXIA: COURSE AND OUTCOME

A study of 73 consecutive children with acute cerebellar ataxia seen over a 23 year-period is reported from the Departments of Pediatrics and Neurology, Washington University School of Medicine and St Louis Children's Hospital. Mean age at onset was 5 years (range, 1 to 21 years); 60% were 2 to 4 years at onset. Prodromal illnesses identified in 57 children included chicken pox in 26%, other presumed viral illness in 52% and immunization related in 3%. No prodrome was recognized in 19%. Epstein-Barr virus was identified in 2 children. Gait ataxia was most severe in patients with varicella, EBV, and vaccination. Other neurologic abnormalities included dysmetria, nystagmus, cranial nerve palsies, and corticospinal tract signs. WBC counts were elevated in half the postviral ataxia cases and normal in the remainder. CSF protein averaged 24 mcg/dl and the mean WBC count was 10 (range, 0-107/mm^3). Pleocytosis >5 was present in one half. Brain scans were normal with one temporary exception. Recovery was complete in 91% of 60 followed for 4 months or longer; 100% in post-varicella cases and 89% in children with non-varicella-related ataxia. Transient behavioral or intellectual difficulties occurred in 20%, and learning problems persisted in 5 (8%). Four children had recurrences of acute ataxia, usually after another presumed viral illness. (Connolly AM, Dodson WE, Prensky AL, Rust RS. Course and outcome of acute cerebellar ataxia. Ann Neurol June 1994;35:673-679). (Respond: Dr Rust, University of Wisconsin School of Medicine, Department of Neurology-H6/546, 600 Highland Avenue, Madison, WI 53792).

> COMMENT. The prognosis for non-varicella cases in this study was superior to that reported by Weiss and Carter (Neurology 1959;9:711) who found 33% of 18 cases with persistent gait disturbance at follow-up. Findings in the above St Louis study previously unreported include the following: 1) boys are affected more frequently than girls (57%/43%), have more severe ataxia, and more frequent cranial nerve palsies and nystagmus; 2) varicella related cases have worse ataxia but more rapid and complete recovery than non-varicella cases; 3) recurrences are not rare and may affect 5% of patients.

TOXIC DISORDERS

LONG-TERM EFFECTS OF METHYLMERCURY POISONING

The clinical, neuropsychological, and radiological features of a family, and the toxicological and neuropathological findings of one family member, who were acutely and severely intoxicated with methylmercury are reported after a 22-year follow-up from the Albuquerque Veterans Affairs Medical Center, the University of New Mexico School of Medicine, and the Environmental Health Sciences Center, the University of Rochester School of Medicine, NY. In 1969 a family in New Mexico had consumed pork containing methylmercury. Three children and a neonate developed severe neurological signs. At 22-year follow-up, the 2 oldest patients, ages 42 and 35 years, had cortical blindness, impaired stereognosis and graphesthesia, poor hand coordination, ataxia, choreoathetosis, dysarthria, and attentional deficits. MRIs showed loss of tissue in calcarine cortices, parietal lobes, and cerebellar folia. The 2 youngest were quadriplegic, blind, and mentally retarded and they died

at ages 29 and 21 years. The brain of the patient poisoned at 8 years and dying at 29 showed cortical atrophy, neuronal loss and gliosis. Total mercury level in the occipital cortex was 1,974 ng/gm, 50 times that of a control; the Hg was mainly inorganic. Hair and systemic organs had Hg levels comparable to controls. (Davis LE et al. Methylmercury poisoning: Long-term clinical, radiological, toxicological, and pathological studies of an affected family. Ann Neurol June 1994;35:680-688). (Respond: Dr Davis, Chief, Neurology Service (127), Albuquerque VA Hospital, 2100 Ridgecrest Drive SE, Albuquerque, NM 87108).

COMMENT. Methylmercury crosses the blood-brain barrier easily while inorganic mercury does not. Biotransformation to inorganic Hg over time may account for the high level of inorganic Hg and absence of methyl Hg in the patient's brain at autopsy. The possible role of inorganic Hg in the brain damage is debatable; it is usually considered to be inert and nontoxic. See Environmental Poisons in Food , Chicago, PNB Publishers, 1993, for an account of the sources, metabolism, epidemiology, clinical manifestations, treatment, and prevention of mercury poisoning. Accidental exposure to mercury vapor is a persisting hazard in nurseries with broken thermometers and in school science labs. The symptoms of mild exposure, *micromercurialism*, are subtle and difficult to diagnose without a high index of suspicion. Acrodynia, or Pink disease, is a relatively rare occurrence, but a diagnosis which should be familiar to the pediatric neurologist and pediatrician.

ATTENTION DEFICIT AND LEARNING DISORDERS

ADDH AND METHYLPHENIDATE RESPONSE

The dose-response, clinical effectiveness, and response prediction in 76 children with ADDH treated with methylphenidate (MPH) were evaluated by a double-blind, placebo-controlled, crossover study at the Department of Psychology, University of Hawaii, Honolulu. Four dose levels (5, 10, 15, and 20 mg) were employed. Effects on classroom functioning, (on-task attention, correct completion of assignments, and teacher ratings), were linear and dose related. Accuracy was enhanced at all dose levels, and task completion was significantly greater at doses above 5 mg. Academic improvement was associated with behavioral gains on teacher ratings. In children failing to respond to low dose MPH, attention changes were responsive to dose increments whereas academic and behavioral improvements failed to occur. A significant subset failed to gain academically from treatment with MPH. (Rapport MD et al. Attention deficit disorder and methylphenidate: Normalization rates, clinical effectiveness, and response prediction in 76 children. J Am Acad Child Adolesc Psychiatry July/Aug 1994;33:882-893). (Respond: Dr Rapport, Dept of Psychology, University of Hawaii, 2430 Campus Rd, Gartley Hall, Honolulu, HI 96822).

COMMENT. Methylphenidate is again proven effective in the treatment of children with ADDH, and improvements in behavior, attention, and academic functioning may be expected in a large percentage. Response to MPH is related to the dose, especially in tasks requiring attention. In one subset, however, academic performance is unrelated to attentional and behavioral response to MPH. In another subset, MPH is ineffective