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