deficiency in skeletal muscle, heart and liver but not in kidney and brain. There was no abnormality in mitochondrial DNA. The disease was thought to result from a nuclear oxidative phosphorylation gene mutation. (Zheng X et al. Evidence in a lethal infantile mitochondrial disease for a nuclear mutation affecting respiratory complexes I and IV. Neurology Sept 1989; 39:1203-1209).

COMMENT. Mitochondrial encephalomyopathies attributed to mutations in the mitochondrial DNA include MERRF and Kearns-Sayre syndrome with onset in childhood through adulthood. In the neonatal period some mitochondrial myopathies have a benign course and some are lethal and a variety of oxidative phosphorylation deficiencies have been associated with these disorders.

PROGRESSIVE SPASTIC CEREBRAL ATAXIA

A syndrome of diabetes insipidus followed by progressive spastic cerebellar ataxia is reported in four boys from the Departments of Neurology, Pediatrics and Psychiatry, UCLA School of Medicine, Los Angeles, CA. In two patients central nervous system histiocytosis was detected. CT scan showed bilateral calcification of the cerebellar dentate nuclei and multiple hypodense areas in the skull; a biopsy confirmed the diagnosis of histiocytosis. A trial of Prednisone was beneficial. (Birnbaum DC et al. Idiopathic central diabetes insipidus followed by progressive spastic cerebral ataxia. Arch Neurol September 1989; 46:1001-1003).

COMMENT. Each of these patients developed idiopathic central diabetes insipidus between the ages of two and six years and all responded to intranasal Desmopressin. Spastic cerebellar ataxia developed eight to ten years later. Histiocytosis accounts for 8-16% of cases of diabetes insipidus in children. Patients with this syndrome may benefit from treatment with corticosteroids.

SEIZURE DISORDERS

NEONATAL SEIZURES

The current concepts and revised classification of neonatal seizures are the subject of a special article from the Division of Pediatric Neurology, Washington University School of Medicine, St. Louis, MO. The clinical classification includes the following: 1) subtle, 2) clonic (focal, multifocal), 3) tonic (focal, generalized), and 4) myoclonic (focal, multifocal, generalized). The focal and multifocal clonic, focal tonic, and generalized myoclonic seizures are commonly associated with simultaneous electrographic seizures. Some varieties of subtle seizures have simultaneous EEG seizure discharges. Neonatal seizures not usually accompanied by EEG seizure activity include: certain subtle seizures, most generalized tonic seizures, and the focal and multifocal myoclonic seizures. These may represent "brain stem release phenomena" and may occur in infants with hydranencephaly and anencephaly. The absence of EEG seizure activity does not rule out an epileptic origin for a clinical