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

seizure. Clinical characteristics can be used to distinguish epileptic and nonepileptic phenomena. Epileptic seizures are not stimulus sensitive, not suppressible by restraint and are likely to be accompanied by autonomic phenomena. If nonepileptic the behavior is 1) stimulus sensitive, 2) may be suppressed by gentle passive restraint, and 3) is not accompanied by autonomic changes such as tachycardia and an increase in blood pressure. Epileptic seizures should be treated because of the threat of brain injury from the seizures per se. Nonepileptic events, e.g. generalized tonic posturing or subtle clinical phenomena without autonomic accompaniments and EEG seizure activity, should not be treated with anticonvulsant drugs. Nonepileptic events rarely interfere with respiration or circulation, they are not harmful to the brain, and unusually high blood levels of phenobarbital are needed to suppress this clinical activity. More data are needed to resolve the issue of a danger of electrical seizure activity to the brain of the newborn in the absence of clinical activity suppressed by anticonvulsant drugs. The author does not attempt to eliminate all electrographic seizure activity because the high doses of medication needed may impair ventilation and/or cardiac function. He discontinues all drugs except phenobarbital as soon as the acute illness is completed (usually when intravenous lines are discontinued) and if the neurologic examination is normal. The etiology of the seizures is important: the risk of recurrence of seizures is 100% with cerebral cortical dysgeneses and 30% with hypoxic ischemic injury and nil with transient metabolic disturbances. He believes that infants should be maintained with phenobarbital for as brief a period as possible. (Volpe, JJ. Neonatal seizures: Current concepts and revised classification. Pediatrics September 1989; 84: 422-428).

COMMENT. Brain damage as a result of seizures per se in the neonate is controversial. The present author believes that there is strong indirect evidence that repeated neonatal convulsions may result in brain injury whereas Lombrozzo and Freeman hold that there is no proof that neonatal seizures can induce brain damage. The degree of emergency in the treatment of neonatal convulsions and the best therapeutic techniques remain in dispute. It seems reasonable to treat all recurring seizures, maintaining the phenobarbital at nontoxic levels. (For a review of this subject see Aicardi J. Epilepsy in children. Raven Press New York. 1986). Although anticonvulsant treatment is often the only therapeutic option available, care must be taken to exclude causes amenable to specific therapy, e.g. hypocalcemia, hypomagnesemia, pyridoxine dependency, biotin deficiency or some cases of hypoglycemia. The use of nasopharyngeal electrodes in electroencephalographic studies of neonatal seizures is potentially hazardous and should be discouraged. The clinical manifestations are usually sufficient to distinguish epileptic and nonepileptic phenomenon. In the absence of routine electrographic confirmation electrodes placed on the facial zygoma may be sufficient to record from the temporal lobe of an infant. In doubtful and persistent cases a trial of anticonvulsant medication would be justified.