autosomal dominant inheritance. Ann Neurol Sept 1992; 32:404-407.) (Correspond: Dr. Boylan, Mayo Clinic, 4500 San Pablo Road, Jacksonville, FL 32224.)

COMMENT. Werdnig-Hoffmann disease is generally regarded as an autosomal recessive disorder with linkage to chromosome 5q11.2-13.3. Other studies have suggested genetic heterogeneity.

The prognosis of patients with Werdnig-Hoffmann disease and a clinical scoring system are evaluated by Russman BS et al. (Ann Neurol Sept 1992; 32:439 (abstr)). The 7 criteria for a poor prognosis were poor movement in utero, presence of tongue fasciculation, a poor cry, inability to hold the head at 3 months, inability to roll over by 6 months, loss of function, and diaphragmatic breathing. Five patients with scores of 4-7 died prior to 15 months of age, while 1 patient with a score of 1 died at 31 years. Patients whose scores during the first 6 months of life were 0 or 1 are living and range in age from 3-27 years. Death prior to age 2-4 years is not invariable.

A unique presentation of neonatal adrenoleukodystrophy as a progressive spinal muscular atrophy is reported in 2 siblings who later developed the symptoms and signs of encephalopathy (Miles DK et al. Ann Neurol Sept 1992; 32:466 (abstr)). Peroxisomal diseases must be considered in the differential diagnosis of Werdnig-Hoffmann disease.

HYPEREKPLEXIA REVISITED

The August 1992 issue of Ped Neur Briefs included 3 articles on hyperekplexia. An additional report from the Hammersmith Hospital, London illustrates the difficulties of clinical diagnosis of startle disease (hyperekplexia). At 1 hour after birth the infant had generalized jerks diagnosed as convulsions and treated with phenobarbitone. She had pronounced truncal hypotonia, but on day 3 she developed hypertonia in the limbs, exaggerated tendon reflexes, and jitteriness. The hypertonia diminished during sleep but increased when she was touched. Jerks and stiffening were precipitated by handling, tapping, and especially by turning to the prone position. Pyridoxine and phenobarbitone reduced the frequency and severity of the attacks but did not relieve the touch-induced myoclonus. Clonazepam introduced on day 28 controlled the symptoms. Forcible flexion of the infant also stopped the jerks and stiffening. The measurement of CSF GABA concentration may be helpful in the diagnosis. (Dubowitz LMS et al. Low cerebrospinal fluid concentration of free gamma-aminobutyric acid in startle disease. Lancet July 11, 1992; 340:80-81.) (Correspondence: Dr. L.M.S. Dubowitz, Department of Paediatrics and Neonatal Medicine, Hammersmith Hospital, Du Cane Road, London W12 OHS, UK.)

COMMENT. I am startled by the unexplained rash of recent articles on hyperekplexia! Ryan SG et al. have studied 4 large clonazepam-responsive families with a hereditary hyperekplexia, using a panel of chromosome 5q polymorphic DNA markers. "Multilocus linkage analysis in each family confirms the previous chromosomal assignment

of hyperekplexia to chromosome 5q. The hypothesis that a mutation in a GABA-receptor subunit causes hyperekplexia is particularly attractive in view of the long term benzodiazepine responsiveness of the disorder."

EYE MOVEMENT DISORDERS

PERIODIC ALTERNATING GAZE AND CEREBELLAR DYSGENESIS

Two children with periodic alternating gaze deviation, presenting at birth in 1 patient and at 2 months in the other, are reported from the New England Medical Center, Tufts University, Boston, MA. The eyes conjugately drifted from one side to the other. With the head stabilized, a regular nystagmus of low frequency and large amplitude was seen when the patient attempted to direct the eyes away from the gaze bias. Compensatory head turning was noted in both patients. MRI showed multiple congenital abnormalities of the posterior cranial fossa. In 1 patient a left renal aplasia and a right supra-adrenal neuroblastoma were diagnosed by ultrasonography and by resection at 4 weeks of age. (Legge RH et al. Periodic alternating gaze deviation in infancy. Neurology Sept 1992; 42:1740-1743.) (Reprints: Dr. Thomas R. Hedges III, 750 Washington Street, Box 381, Boston, MA 02111.)

COMMENT. The periodic alternating gaze phenomenon is rare and points to cerebellar disease, with malformation of the inferior cerebellar vermis as the specific underlying lesion. The association with neuroblastoma in 1 patient is a coincidence since symptoms of opsoclonus were not described.

OCULOMOTOR PARESIS AND BASILAR ANEURYSM

The clinical and pathologic findings in a 10 month old girl with congenital heart diseasewho died after rupture of a congenital distal basilar artery aneurysm are reported from the University of Connecticut Health Center, Farmington, CT and the Children's Hospital of Philadelphia, PA. She was hospitalized at 10 months of age after having had 4 syncopal episodes within a 6 week interval. Twenty-four hours after cardiac catherization the infant developed an adduction deficit of the left eye which resolved over the next 36 hours. Five days after surgical correction of the cardiac defect she had a massive subarachnoid hemorrhage. (DiMario FJ Jr., Rorke LB. Transient oculomotor nerve paresis in congenital distal basilar artery aneurysm. Pediatr Neurol July/Aug 1992; 8:303-306.) (Communications: Dr. DiMario, Division of Neurology, University of Connecticut Health Center, 263 Farmington Avenue, Farmington, CT 06030.)

COMMENT. Transient third nerve paresis in an infant with a history of recurrent syncope and congenital heart disease should suggest a possible intracranial arterial aneurysm. The authors found 6 previous reports of basilar aneurysm preceded by a transient third nerve dysfunction. A migraine variant is another explanation. (Shevell MI et al. Ann Neurol Sept 1992; 32:484 (abstr).)