prognostic value in assessment of neurological outcome. (See **Progress in Pediatric Neurology**, Millichap JG ed, 1991, page 333)

Cerebro spinal fluid examination in symptom-free infants with risk factors for infection was evaluated in 284 newborns at the Department of Pediatrics, Rush Presbyterian-St. Luke's Medical Center, Chicago, IL (Fielkow S et al. J Pediatr Dec 1991; 119:971-973). Positive cultures without pleocytosis in 5 infants (1.8%) were contaminants and none of the symptom-free infants had meningitis. The authors conclude that CSF examination is not indicated in the diagnostic evaluation of symptom-free infants born to mothers with chorioamnionitis or other risk factors for neonatal infection.

NEUROCUTANEOUS SYNDROMES

VISUAL LOSS IN TUBEROUS SCLEROSIS

Visual loss in 4 patients with tuberous sclerosis complicated by subependymal giant-cell astrocytoma is reported from the Departments of Ophthalmology, Neurology, and Radiology, University of Michigan Medical Center, Ann Arbor, MI. The patients presented at 12-20 years of age with obstructive hydrocephalus. Surgery relieved elevated pressure in all cases but 2 patients became blind and 1 has severe visual field loss from the effects of chronic papilledema on the optic nerves. Early surgical decompression prevented visual loss in the fourth patient whose tumor was removed at 12 years of age and who required further resection of a giant-cell astrocytoma at 20 years. (Dotan, SA et al. Visual loss in tuberous sclerosis. Neurology Dec 1991; 41:1915-1917.) (Reprints: Dr. J.D. Trobe, W.K. Kellogg Eye Center, 1000 Wall St., Ann Arbor, MI 48105.)

COMMENT. Periodic opthamologic examination and brain imaging are advisable in tuberous sclerosis patients with subependymal nodules. The timely relief of increased intracranial pressure may arrest or prevent loss of vision.

FAMILIAL SPINAL NEUROFIBROMATOSIS

The clinical features and genetic linkage analysis of two pedigrees with familial spinal neurofibromatosis (NF) are described from the Divisions of Neurology and Medical Genetics, Cedars-Sinai Medical Center, UCLA School of Medicine, Los Angeles, CA; the Neurofibromatosis Institute, Pasadena, CA; and Department of Medical Informatics, University of Utah, Salt Lake City, UT. On clinical grounds, it was difficult to assign the two families to NF1 or NF2 based on the criteria established by the National Institutes of Health. Cutaneous tumors, Lisch nodules or acoustic tumors were absent. Cafe-au-lait spots were present in 1 family and absent in the other. The inheritance pattern was autosomal dominant in both pedigrees. Genetic linkage analysis was performed with markers linked to the NF1 gene on chromosome 17 and markers linked to the NF2 gene on chromosome 22. The

location for the mutation in the first family was in the NF1 gene, whereas that in the second family was excluded from the NF1 locus, although the phenotype was similar to that of family 1. (Pulst SM, Riccardi VM et al. Familial spinal neurofibromatosis: clinical and DNA linkage analysis. Neurology Dec 1991; 41:1923-1927.) (Reprints: Dr. Pulst, Div Neurol, Cedars-Sinai Medical Center, 8700 Beverly Blvd., Los Angeles, CA 90048.)

COMMENT. Studies of families in which all affected individuals express the same subset of the NF phenotype allow a correlation between the mutation and its clinical characteristics. Symptomatic spinal neurofibromas occur in less than 5% of patients with NF1, but are commonly found in NF2. This may be the first report of familial spinal neurofibromatosis in families with NF1. One individual in each pedigree developed a neurofibrosarcoma and died of its complications. This is a rare complication in NF1, explained by the authors as a familial predisposition related to the mutations.

CUTANEOUS GRANULOMAS AND ATAXIA-TELANGIECTASIA

Development of cutaneous granulomas in 8 patients with ataxia-telangiectasia is reported from the Departments of Pediatrics and Dermatology, Northwestern University Medical School, Chicago, IL; the Hospital for Sick Children, Toronto, Canada; Henry Ford Hospital, Detroit, MI; and University of North Carolina, Chapel Hill. The granulomas were atrophic lesions that frequently became encrusted and ulcerative. Their appearance differed significantly from the well circumscribed annular plaques or skin colored subcutaneous nodules of typical granuloma annulare. The lesions were persistent rather than self-limited and no infectious organisms were demonstrated. Treatment with intravenous immune globulin, topical antibiotic therapy and topical corticosteroid therapy were unsuccessful. The authors postulate that the granulomas were an attempt to localize antigen in patients with a dysfunctional immune system. (Paller AS et al. Cutaneous granulomatous lesions in patients with ataxia-telangiectasia. J Pediatr Dec 1991; 119:917-922.) (Reprints: Dr. Paller, Division of Dermatology, Children's Memorial Hospital, 2300 Children's Plaza, Chicago, IL 60614.)

COMMENT. Patients with ataxia-telangiectasia have dysfunction of humoral and cell-mediated immunity as demonstrated by an immature thymus; absent or small tonsils; low levels of immunoglobulins A, E, G2 and G4; and a poor response to antigenic stimulation.

SEIZURE DISORDERS

AUTISM AND EPILEPSY

The prevalence and clinical manifestations of epilepsy in 302 autistic and 237 dysphasic, non-autistic children were studied at the Departments of Neurology and Pediatrics, Albert Einstein College of Medicine and Montefiore