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

Medical Center, Bronx, New York. Epilepsy occurred in 14% of autistic children and 8% of dysphasic children. Girls were affected more frequently than boys (24% cf 11%). The seizure types included generalized tonic-clonic, atypical absence, atonic, myoclonic, infantile spasms and partial. Infantile spasms occurred in 12% of autistic patients with epilepsy. The major risk factors for epilepsy were severe mental deficiency and the combination of mental deficiency with a motor deficit. When cognitive and motor disabilities were excluded, the risk of epilepsy in autistic children was only 6% and similar to that found in dysphasic non-autistic children (Tuchman RF, Rapin I, Shinnar S. Autistic and dysphasic children. II: epilepsy. Pediatrics Dec 1991; 88:1219-1225.) (Reprints: Dr. Rapin, Rose F. Kennedy Center for Research in Mental Retardation, Room 807, 1410 Pelham Parkway South, Bronx, NY 10461.)

COMMENT. The rates of epilepsy reported in autistic children have ranged from 11% to 42%. Associated cognitive and motor deficits, a higher ratio of girls, and the complication of verbal auditory agnosia in some series might explain the higher rates of epilepsy. As a group, autistic children are more likely than dysphasic children to have language subtypes affecting central processing and a history of regression of language and behavior. Girls with autism are more likely than boys to have severe mental deficiency and a motor deficit.

Epilepsy occurred in 8 (17%) of 46 children with autism and deafness (Jure R et al. Hearing impaired autistic children. Dev Med Child Neurol Dec 1991; 33:1062-1072).

SURGERY OF EPILEPSY

The surgical outcome in 34 patients between 2 and 15 years of age who were operated on for medically intractable seizures is reported from the Neurosurgical Clinic, University Clinical Center, Visegradska 26, Belgrade, Yugoslavia. Temporal epileptic foci were present in 9 patients, extratemporal foci in 6, infantile hemiplegia in 16 and epilepsia partialis continua in 3. Detectable brain lesions were present in 30 (88%). At 1-14 years post-operative follow-up (mean, 4 years), 21 (62%) are seizure free, 8 (23%) have improved, and 5 (15%) have shown no improvement. Patients with temporal foci had a partial temporal lobectomy or restricted temporofrontal resection; those with extratemporal foci had cortical resection; and patients with infantile hemiplegia had a partial hemispherectomy, extensive cortical resection or functional hemispherectomy of Rasmussen type. (Ribaric II et al. Surgical treatment of epilepsy: our experiences with 34 children. Child's Nerv Syst Nov 1991; 7:402-404.) (Reprints: Dr. Ribaric, University Clinical Center, Visegradska 26, YU-11000 Belgrade, Yugoslavia.)

COMMENT. Of 16 patients with infantile hemiplegia who were subjected to hemispherectomy, 12 (75%) were seizure free. Lindsay J et al. from the Park Hospital for Children, Oxford, found a similar incidence of seizure control following hemispherectomy in 17 children with hemiplegic epilepsy (Dev Med Child Neurol 1987; 29:592. See