years of age. The above report identifies a kindred with PM-Like D, clinically resembling PMD, but with intact myelin and with mutations at a new locus on the X chromosome.

SPINAL MUSCULAR ATROPHY AND ARTHROGRYPOSIS

Four infants with neurogenic arthrogryposis who died of respiratory failure before 1 month of age had DNA testing of autopsy specimens for *SMN*^{T} gene deletion in a study at the Children's Hospital of Philadelphia, PA, and the Children's Hospital at Dartmouth, Lebanon, NH. All infants had clinical, pathologic, or EMG evidence of motor neuron disease. In addition to anterior horn cell loss, autopsies showed a more extensive neurodegeneration involving central sensory neurons in Clarke's column and the thalamus. *SMN*^{T} deletion was identified in two of the cases. (Bingham PM, Shen N, Rennert H et al. Arthrogryposis due to infantile neuronal degeneration associated with deletion of the *SMN*^{T} gene. Neurology Sept 1997;49:848-851). (Reprints: Dr PM Bingham, Division of Child Neurology, Children's Hospital of Philadelphia, Philadelphia, PA 19104).

COMMENT. Arthrogryposis in association with infantile spinal muscular atrophy (Werdnig-Hoffmann disease) was first reported by Byers and Banker (1961). DNA analysis for *SMN*^{T} deletion in cases of neurogenic arthrogryposis may uncover a diagnosis of spinal muscular atrophy or SMA variant and facilitate genetic counselling. Some cases of infantile SMA may have degenerative changes in sensory neurons in addition to the classical anterior horn cell loss.

Congenital axonal neuropathy with *SMN* deletion is reported in three newborn siblings presenting with generalized weakness, asphyxia, facial diplegia, and external ophthalmoplegia, and studied at Pediatric University Hospital, Mathildenstr, Freiburg, Germany (Korinthenberg R, Sauer M, Ketelsen U-P et al. Ann NeurolSept 1997;42:364-368). EMG, NCV, and nerve biopsies confirmed an axonal neuropathy. The electrophysiological and biopsy findings, together with the *SMN* gene deletion, were diagnostic of a severe spinal muscular atrophy, complicated by involvement of brainstem nuclei and sensory nerves. Contrary to accepted criteria, weakness of extraocular muscles and facial weakness do not exclude the diagnosis of SMA.

VASCULAR DISORDERS

FACTOR V LEIDEN MUTATION AND NEONATAL STROKE

Three infants with familial factor V Leiden mutation and neonatal cerebrovascular disorders are reported from the Children's Hospital of Philadelphia, PA. One had placental thrombosis. Activated protein C resistance caused by factor V Leiden mutation is an important cause of in utero ischemic infarction and hemorrhagic stroke and may present with neonatal hemiplegic cerebral palsy. (Thorarensen O, Ryan S, Hunter J, Younkin DP. Factor V Leiden mutation: an unrecognized cause of hemiplegic cerebral palsy, neonatal stroke, and placental thrombosis. Ann Neurol Sept 1997;42:372-375). (Dr Younkin, Division of Neurology, 6th Floor, Wood Bldg, Children's Hospital of Philadelphia, 34th St and Civic Center Blvd, Philadelphia, PA 19104).

COMMENT. Infants with hemiplegic cerebral palsy caused by a vascular accident should be tested for factor V Leiden mutation, especially if a parent

has suffered a stroke, venous thrombosis, or heart attack. Factor V Leiden may coexist with hereditary homocystinuria, another prothrombotic disorder.

The role of hyperhomocysteinemia in stroke is emphasized in a study of 125 consecutive adults at the University of Munster, Germany. (Evers S, Koch H-G, Grotemeyer K-H et al. Features, symptoms, and neurophysiological findings in stroke associated with hyperhomocysteinemia. Arch Neurol Oct 1997;54:1276-1282). The prevalence was 20% in all patients with stroke, and impaired cognition was more pronounced in those with hyperhomocysteinemia.

INFECTIOUS DISORDERS

CAT-SCRATCH ENCEPHALOPATHY

A 9-year-old girl with cat-scratch disease complicated by encephalopathy and seizures is reported from the Kaiser Foundation Hospital, Los Angeles, CA. The patient was admitted to hospital after a 2-week history of cervical adenitis, a 2-day history of low-grade fever, diarrhea, and headache, and an 11-day course of oral antibiotics with no response. A generalized tonic-clonic seizure occurred within hours of admission and initiation of i.v. antibiotics. Following the 2 minute seizure she became combative, delirious, and comatose. CSF showed protein of 72 mg/dl and normal cells. Recovery began after 24 hours and was complete in 5 days. Serum serology and polymerase chain reaction (PCR) analysis of lymph node tissue were positive for *Bartonella henselae*. The child had 4 kittens but no observed scratches or bites. (Wheeler SW, Wolf SM, Steinberg EA. Cat-scratch encephalopathy. Neurology Sept 1997;49:876-878). (Reprints: Dr Sheldon M Wolf, 1505 N Edgemont Street, 5th Floor, Los Angeles, CA 90027).

COMMENT. Neurologic complications of cat-scratch disease are uncommon, although there are several reports of encephalopathy, generally with complete recovery, and isolated reports of myelopathy, cranial nerve palsies, optic neuritis, chorea, and cerebellar ataxia. A cat scratch is not always identified, but the cat is the principal reservoir for the infecting organism, *Bartonella henselae*. A cat flea may account for the transmission in some cases. See Progress in Pediatric Neurology II (Millichap JG, ed. Chicago, PNB Publishers, 1994;pp421-423) for further reports of neurologic complications of cat-scratch disease, including one series of 76 patients. The differential diagnosis includes Lyme encephalitis.

FACIAL PALSY AND LYME BORRELIOSIS

The value of CSF examinations for intrathecal antibody production to *Borrelia burgdorferi* in the diagnosis of neuroborreliosis in children with peripheral facial palsy (PFP) was examined at the University Children's Hospital of Zurich, Switzerland. Twenty (95%) of the children with PFP had immunoglobulin (Ig)M or IgG in the acute-phase serum, but serologic assays showed discrepancies in one third. Intrathecal antibody to *B. burgdorferi* was present in 5 of the 20 seropositive children. Seroconversion in convalescent sera was found in all 5 with intrathecal antibody, and in 8 of 10 without intrathecal specific-antibody production. Patients showing intrathecal antibodies or seroconversion had lymphocytic pleocytosis in the acute phase of PFP. (Albisetti M, Schaer G, Good M, Boltshauser E, Nadal D. Diagnostic value of cerebrospinal fluid examination in children with peripheral facial palsy