H et al. Pediatr Neurol April 2001;24:303-305). The authors suggest a link between the chronic steroid treatment in an immunosuppressed patient and the development of ADEM.

VASCULAR DISORDERS

PROGNOSTIC FACTORS IN ISCHEMIC ARTERIAL STROKE

The predictive value of presenting symptoms, MRI and CT findings, and etiology in the outcome of ischemic arterial childhood stroke was determined in a consecutive series of 31 patients followed at the University Hospital, Rotterdam, The Netherlands. Hemiparesis was the most common presenting symptom (74%), seizures occurred in 19%, altered level of consciousness in 16%, and ataxia in 7%. Location of infarction on neuroimaging was in the territory of the middle cerebral artery (MCA) in 27 cases, basilar artery (BA) in 4, and in the cerebellum involving the posterior inferior cerebellar artery (PICA) in 2. Three MCA and 2 MCA and ACA strokes (19%) were complete. Etiology was identified in 24 (77%), including cardiac surgery complications in 6, varicella zoster-related in 5, mitochondrial disease in 2, migraine-related in 2, and Moya-Moya, Kawasaki disease, factor V Leiden, sickle-cell disease, and hyperthyroid crisis in 1 each. Risk factors at presentation that correlated with a poor prognosis were an altered level of consciousness, seizures, and a completed stroke of the MCA. Etiology, age at presentation, or gender showed no significant correlation with outcome. (Delsing BJP, Catsman-Berrevoets CE, Appel IM. Early prognostic indicators of outcome in ischemic childhood stroke. Pediatr Neurol April 2001;24:283-289). (Respond: Dr Catsman-Berrevoets, Child Neurologist, Dept of Child Neurology, Dr Molewaterplein 60, 3015 GJ Rotterdam, The Netherlands).

COMMENT. Almost one-half the patients in this study died or had severe residual morbidity. The early risk factors for this poor outcome were an altered level of consciousness at presentation, seizures, and MRI evidence of complete or end-zone MCA infarction.

NEUROMUSCULAR DISORDERS

EXPANDED MOBIUS SYNDROME

An infant born with Mobius syndrome died at 22 days and was found at autopsy to have more widespread involvement of brainstem and cranial nerve nuclei than usual, resulting in an "expanded Mobius syndrome," as reported from the University of Iowa Hospital, Iowa City, IA. At emergency cesarean section, performed at 33 weeks gestation because of fetal distress and arm tremor, a 1672 gm male infant required continuing ventilatory support. At neurologic examination, the diagnosis was expanded Mobius syndrome with diffuse cranial nerve and brainstem involvement. CT and MRI revealed diffuse cerebral atrophy. Postmortem examination showed bilateral pneumonia secondary to aspiration. The brain was of normal weight and its surface appeared normal. Cranial nerve rootlets VI-XII were absent. Microscopic examination showed bilateral brain, basal ganglia, and brainstem gliosis and mineralization. Neurons in the nuclei of cranial nerves III-XI were absent. There was lesser involvement of the spinal cord, cerebral white matter, and cerebellum. No inflammatory cells or evidence of infection were evident. (Peleg D, Nelson GM, Williamson RA, Widness JA. Expanded Mobius syndrome. Pediatr Neurol April 2001;24:306-309). (Respond: Dr Widness, Department of Pediatrics, University of Iowa Hospital and Clinics, 200 Hawkins Drive, Iowa

City, IA 52242).

COMMENT. Mobius syndrome is a congenital disorder usually limited to a bilateral paralysis of cranial nerves VI and VII, resulting in facial diplegia and impaired abduction of the eyes in lateral gaze. Other cranial nerves, V, IX, and XII are occasionally involved also. In some cases the involvement of the brainstem nuclei, nerves and muscles is more diffuse, leading to an expanded form of the syndrome. Limb and other craniofacial malformations may occur. In the case reported above, the pathology was particularly extensive and the infant died soon after birth. The syndrome has multiple presumed causes. As in the present report, a possible prenatal vascular insuffiency or hypoxic/ischemic event involving embryonic subclavian artery branches may cause gliosis and mineralization of selected cranial nerve nuclei or a more extensive pathology. A dysgenesis of cranial nerve nuclei and hypoplasia of nerves and muscles (oromandibular limb hypogenesis) or a primary myopathy involving facial and external eye muscles are alternative theories,. The timing of the suggested embryonic insult is in the early first trimester, Abuse of benzodiazepines, chorionic villus sampling, and placental bleeding have been invoked as causes. The term Mobius sequence is sometimes preferred to syndrome, better describing a cascade of events secondary to a proposed embryonic insult. Mobius sequence is preferred by the authors of the following report.

MOBIUS SYNDROME AND AUTISTIC SPECTRUM DISORDER

The relation between autistic spectrum disorder and Mobius syndrome was studied in 25 patients (18 males and 7 females) included in a multidisciplinary study of Mobius sequence at the Department of Child and Adolescent Psychiatry, Goteborg University, Sweden. Autistic spectrum disorder and learning disability were present in more than one third of patients. The findings point to a possible brainstem insult in early pregnancy as a cause in some cases of autism. (Johansson M, Wentz E, Fernell E et al. Autistic spectrum disorders in Mobius sequence: a comprehensive study of 25 individuals. Dev Med Child Neurol May 2001;43:338-345). (Respond: Maria Johansson MD, Department of Child and Adolescent Psychiatry, Gotegorg University, Kungsgatan 12, SE-411 19 Goteborg, Sweden).

COMMENT. Brainstem dysfunction has been proposed as a possible mechanism of autism (Gillberg et al, 1983). The present study offers some support for this hypothesis.

JUVENILE DERMATOMYOSITIS WITH GENERALIZED EDEMA

A 7-year-old girl presenting with an 8-week history of fatigue, myalgia, dyspnea, and generalized, nonpitting edema of the extremities, face, chest, and abdomen, is reported from the Walter Reed Army Medical Center, Washington, DC. A heliotrope and malar rash had developed, and muscles were tender to palpation. A severe proximal muscle weakness, elevated serum muscle enzymes, and heliotrope rash were characteristic of juvenile dermatomyositis (JDM). Other diseases characteristic of generalized anasarca, such as nephrosis, liver disease, hypothyroidism, and malignancy, were excluded. Seventeen additional cases of JDM and anasarca were cited in the literature. Response to oral steroids was poor, and high dose long duration IV therapy was needed. (Mitchell JP, Dennis GJ, Rider LG. Juvenile dermatomyositis presenting with anasarca: a possible indicator of severe disease activity. J Pediatr June 2001;138:942-945). (Reprints: Jeanne P Mitchell MD, Department of Rheumatology and Clinical Immunology, Walter Reed Army Medical Center, 6900 Georgia Ave NW, Ward 77, Washington, DC 20307).