

months) was favorable in 30% of the study population. The remaining 70% (28 of 40) with an unfavorable outcome included mortality in 33% and severe morbidity in 38%. Epilepsy occurred in 56% of survivors (15 of 27), development delay - 67%, and cerebral palsy - 63%. Factors influencing the outcome were the etiology of the seizures, the age at the onset of seizures, birth weight and neurologic examination results. Asphyxia, meningitis and cerebral dysgenesis carried a poor prognosis. Onset of seizures within the first day of life and a severely abnormal neurologic exam significantly predicted an unfavorable outcome. An Apgar score less than 5 at 5 minutes was not significantly related to a poor prognosis. The EEG background activity was the only EEG parameter of value in prognosis: an abnormal background carried an unfavorable outcome in 81% and a normal background was predictive of a favorable prognosis in 67%. (Legido A et al. Neurologic outcome after electroencephalographically proven neonatal seizures. Pediatrics September 1991; 88:583-596).

COMMENT. In this retrospective study of neonates with EEG proven seizures, the mortality rate was 33% and the morbidity rate was 56%.

The long-term outcome of an infant with pyridoxine-dependent neonatal seizures is reported from the British Columbia's Children's Hospital, Vancouver, BC, Canada (Connolly MB et al. Intravenous immunoglobulin and pyridoxine-dependent seizures. Neurology September 1991; 41:1524). Generalized clonic seizures began at 6 days of age followed by partial motor seizures becoming secondarily generalized at 1 month, and episodes of status epilepticus lasting 1 hour at 2-1/2 months. Seizures were controlled and the EEG normalized by 100 mg pyridoxine daily. The patient remained seizure free until age 20 months and then developed episodes of status epilepticus twice a week, always associated with infections. Monthly IV gamma globulin therapy 400 mg/kg infused over 6 hours resulted in prevention of infections and control of convulsions. IVIg therapy may be helpful in the treatment of pyridoxine-dependent seizures beginning in the neonatal period and may improve the long-term outcome.

SURGERY IN WEST SYNDROME

The results of surgical removal of a porencephalic cyst in an 18-month old child with infantile spasms and hypsarrhythmia are reported from the Departments of Neurology and Neurosurgery, VA Medical Center and University of Florida; the State University of Stony Brook, NY; and University of Michigan Medical School, Ann Arbor, MI. Infantile spasms were evaluated first at 4 months of age. The seizures were generalized but the neurologic examination showed a left hemiparesis and a left visual field deficit. ACTH provided a transient control of spasms for 3 months. At 18 months the seizures had become refractory to medical treatment. Postoperatively, seizures have been controlled for 12 months and the use of the paretic extremities has improved. Preoperative anti-epileptic medications

were continued. (Uthman BM et al. Outcome for West Syndrome following surgical treatment. Epilepsia Sept/Oct 1991; 32:668-671).

COMMENT. This report indicates that infantile spasms and hypsarrhythmia of a diffuse pattern may be associated with a focal cerebral lesion amenable to surgery. Infantile spasms associated with COFS syndrome (cerebro-oculo-facial-skeletal syndrome) in a 3-month-old child is reported from the Department of Neurology, New York Medical College, New York, NY (Harden CL et al. Pediatr Neurol July/Aug 1991; 7:302-304). COFS syndrome is a rare autosomal recessive condition characterized by microcephaly, microphthalmia and/or cataracts, neurogenic arthrogryposis and multiple congenital anomalies. The infantile spasms and hypsarrhythmia resolved during ACTH therapy.

DIAGNOSIS OF JUVENILE MYOCLONIC EPILEPSY

Factors contributing to the misdiagnosis of juvenile myoclonic epilepsy (JME) in an epilepsy clinic have been examined in 70 patients at the Division of Neurology, King Khalid University Hospital, Riyadh, Saudi Arabia. More than 90% were undiagnosed on referral and 33% were not recognized initially in the epilepsy clinic. The delay in diagnosis was 8 years from onset and 17 months from the first evaluation in the clinic. Factors responsible for the delayed diagnosis include the following: myoclonic jerks rarely reported by patients; generalized tonic-clonic seizures may be nocturnal without circadian relation to awakening from sleep; unilateral jerks may suggest simple partial seizures; absence seizures may antedate jerks and GTCS seizures by 4.5 years and are frequently unrecognized. The EEG was significant in confirming the diagnosis in 63% of patients. Valproate is considered the treatment of choice and clonazepam is used as an adjunctive treatment. (Panayiotopoulos CP et al. Juvenile myoclonic epilepsy: factors of error involved in the diagnosis and treatment. Epilepsia Sept/Oct 1991; 32:672-676).

COMMENT. Failure to recognize JME may result in improper choice of anticonvulsant therapy, resultant status epilepticus, and failure to provide appropriate genetic counseling. This study reemphasizes the atypical history in some cases and the frequency of occurrence of absence seizures as the initial manifestation.

MEMORY AND LEARNING DISABILITIES

ANATOMY OF MEMORY

Studies of the anatomy and function of the brain system for memory in humans and animal models are reviewed from the Veterans Affairs Medical Center, San Diego and the Department of Psychiatry, University of California, San Diego, La Jolla, CA. Patients who underwent temporal lobe surgery developed memory impairment only when the removal extended far enough posteriorly to include the hippocampus and the parahippocampal gyrus. Horel