of age with incomplete clinical signs, 2) in NF-1 families for prenatal testing, and 3) when complete clinical examination is impracticle. (Hofman KJ, Boehm CD. J Pediatr March 1992; 120:394-398.)

METABOLIC AND TOXIC DISORDERS

METHYLGLUTACONIC ACIDURIA

Two siblings with 3-methylglutaconic aciduria with normal enzyme activity and neurological abnormalities are reported from the Department of Pediatrics, Beilinson Medical Center, Petah Tiqva, Sackler School of Medicine, Tel Aviv University, Israel. Patient 1, an 11 month old infant was referred for evaluation of deterioration of motor development beginning at 5 months of age. There was an arrest of weight gain and head circumference growth. He was restless and had choreoathetoid movements of hands, head and mouth, deep tendon reflexes were hyperactive, plantar responses were extensor and optic discs atrophied. At 2 years of age he sat with support, crawled and walked with help. MRI and CT showed prominent frontal lobe atrophy. Urinary organic acid analysis showed a prominent peak of 3-methylglutaconic and 3-methylglucrylic acid. Patient 2, the 14 year old sister of patient 1, developed ataxia of gait at age 2 years, and optic atrophy at 8 years of age. Examination showed variable muscle tone with brisk tendon reflexes and extensor plantar responses. Choreoathetoid movements were prominent in the upper limbs, head and mouth and funduscopic examine showed prominent optic atrophy. Her IQ on the WISC was 87. CT and MRI showed generalized brain atrophy. Urinary organic acid excretion showed a prominent peak of methylglutaconic and methylglutaric acids. The patients represent a new clinical variant of the methylglutaconic aciduria syndrome with a relatively favorable prognosis. (Zeharia A, Weitz R et al. 3-methylglutaconic aciduria: a new variant. Pediatrics June 1992; 89:1080-1082.) (Reprints: Avraham Zeharia, M.D., Department of Pediatrics B, Beilinson Medical Center, Petah Tiqva 49100, Israel.)

COMMENT. 3-Methylglutaconic aciduria is the hydrolysis product of 3-methylglutaconyl coenzyme and an intermediate in the degradation pathway of leucine. Two syndromes are described - one with deficient methylglutaconyl coenzyme A and the other with normal enzyme activity, but prominent neurological deterioration. The present case reports were unique in the relatively normal cognitive and intellectual development and the relatively mild neurological manifestations. The boy had demonstrated developmental improvement in his second year of life and his sister developed well with normal school performance.

COCAINE ADDICTION AND EEG IN INFANTS

Thirty-five consecutive infants of cocaine-addicted mothers hospitalized for a comprehensive health assessment and 51 healthy, age-matched infants were studied with electroencephalography at the Children's Hospital of Philadelphia, PA. No definite EEG seizures were recorded in any of the patients. In infants of cocaine-addicted mothers there was a tendency for

electroclinical sleep discordance, and the frequency of early acquisition of a mature pattern of quiet sleep (continuous, slow-wave sleep) was significantly higher than in the comparison group below conceptional age 45 weeks. The significance of precocious maturity of quiet sleep was unclear, but may indicate accelerated biological development. (Legido A et al. Electroencephalographic and behavioral-state studies in infants of cocaine-addicted mothers. AJDC June 1992; 146:748-752.) (Reprints: Dr. Legido, St. Christopher's Hospital for Children, Section of Neurology, Erie Avenue at Front Street, Philadelphia, PA 19134-1095.)

COMMENT. The authors suggest that the longitudinal assessment of sleep disturbance and its relation to later development might permit tracking of the long-term effects of prenatal exposure to cocaine. Other complications of cocaine exposure in utero are small head circumference, abnormal behavior, cerebral infarction or hemorrhage, seizures and SIDS.

CEREBELLAR SYMPTOMS IN CRIGLER-NAJJAR TYPE I DISEASE

Three children with Crigler-Najjar (CN) type I disease who had cerebellar symptoms as the initial manifestation of kernicterus are reported from the Hopital Antoine Beclere, Clamart Cedex, France. Patient 1, a 3600 gram boy, was admitted with jaundice and a serum total bilirubin of 507 umol/L unconjugated on day 2. After two exchange transfusions and continuous phototherapy the jaundice persisted and a therapeutic trial with phenobarbital was of no benefit. The bilirubin UDPG-T hepatic activity assayed at 3 months was absent. At age 6 years during an episode of fever and headache with respiratory viral infection the jaundice increased and ataxia and dysmetria developed. Liver transplantation was performed with apparent success. In all 3 children the cerebellar symptoms began after an infectious episode or an interruption of phototherapy. The neurotoxicity was associated with an elevated saturation of albumin by bilirubin of 61% to 75%. In 8 patients without neurotoxicity this parameter remained below 58%. (Labrune PH et al. Cerebellar symptoms as the presenting manifestations of bilirubin encephalopathy in children with Crigler-Najjar type I disease. Pediatrics April 1992; 89:768-770.) (Reprints: P.H. Labrune, M.D., Service de Pediatrie, Hopital antoine Beclere, 157 rue de la Porte de trivaux, 92141 Clamart Cedex, France.)

COMMENT. Cerebellar symptoms are rare features of kernicterus despite the known occurrence of bilirubin staining of the dentate nuclei in pathological studies. The absence of reports of cerebellar involvement in the neonate may be explained by the difficulty in detection of cerebellar signs. Since neurological impairment may be permanent and is potentially preventable, patients with CN-1 disease are candidates for liver transplantation performed when phototherapy is ineffective and prior to the development of kernicterus. Therapy of jaudice in the newborn is reviewed by Newman TB and Maisels MJ in Pediatrics May 1992; 89:809-818.