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