indicator of mitochondrial disease in patients with encephalopathic disorders.

The treatment of congenital lactic acidosis is reviewed from the Center for Inherited Disorders of Energy Metabolism, Case Western Reserve University, Cleveland, OH (Kerr DS. Int Pediatr 1995;10:75-81). Treatments have included diet, vitamins, use of enzyme activators, and enzyme replacement. None has been very successful.

See Progress in Pediatric Neurology I and II (Millichap JG, Ed. PNB Publ, 1991 and 1994) for further articles on mitochondrial cytopathies. The diagnosis of mitochondrial disorder should be considered with the following: 1) an unexplained association of symptoms; 2) an early onset and rapidly progressive course; and 3) involvement of unrelated organs sharing no common embryologic origin and no common biological functions.

PRENATAL DIAGNOSIS OF LESCH-NYHAN SYNDROME

The results of carrier and prenatal diagnosis for Lesch-Nyhan syndrome by carrier testing of 83 women and prenatal analysis of 26 pregnancies are reported from the Department of Molecular and Human Genetics, Baylor College of Medicine, Houston, TX. Mutation detection and linkage analysis were used for probands and their families and biochemical measurement of HPRT enzyme activity for at-risk pregnancies. Mutations in the HPRT gene of affected males were detected in 100% cases. Forty five (56%) at-risk women were found not to carry their family's HPRT gene mutation. (Alford RL et al. Lesch-Nyhan syndrome: Carrier and prenatal diagnosis. Prenat Diagn April 1995;15:329-338). (Respond: Dr RL Alford, Department of Molecular and Human Genetics T-528, Baylor College of Medicine, One Baylor Plaza, Houston, TX 77030).

COMMENT. Lesch-Nyhan syndrome is an X-linked recessive disorder characterized by hyperuricemia, choreoathetosis, mental retardation, self-mutilatory behavior, and a deficiency of the enzyme hypoxanthine guanine phosphoribosyltransferase (HPRT). Molecular diagnostic studies of affected males and carrier testing prior to pregnancy have been shown to demonstrate genetic risks and unnecessary prenatal tests may be avoided

5-HYDROXYTRYPTOPHAN IN FRIEDREICH'S ATAXIA

The effect of the levorotatory form of 5-hydroxytryptophan (approx 1 gm/day/orally) on cerebellar symptoms in 26 patients with Friedreich's ataxia was evaluated in a double-blind drug-placebo study by the Ataxia Research Center, Hopital Neurologique, Lyon and 11 other research hospitals in France. Of 19 completing the study, 11 were treated with 5-hydoxytryptophan and 8 with placebo. A significant decrease of the kinetic score and improvement in coordination was observed in the active treatment group after 6 months but not at 4 months, indicating a progressive drug effect. Five subtests demonstrating improvement included finger-nose test, heel-knee, and Archimedes' spiral. A trend toward acceleration of the speed of speech was also observed. Gastrointestinal symptoms were the main side-effects of treatment. (Trouillas P et al. Levorotatory form of 5-hydroxytryptophan in Friedreich's ataxia. Arch Neurol May 1995;52:456-460). (Reprints: Dr Trouillas, Ataxia Research Center and Cerebrovascular Unit, Faculte Alexis Carrel, Universite Claude Bernard, Hopital Neurologique, 59 Boulevard Pinnel, 69003 Lyons, France).

COMMENT. The effect of 5-hydroxytryptophan was only partial, improving kinetic ataxic symptoms but not the static scores involving posture. In another double-blind crossover study at the Medical University of Lubeck, and other centers in Germany, Wessel K et al reported no significant effect of hydroxytrytophan on cerebellar symptoms in 19 patients with Friedreich's ataxia (Arch Neurol May 1995;52:451-455). Currier RD, in an editorial, concludes that "the levorotatory form of 5-hydoxytryptophan may have an effect that is minimal, selective, and difficult to detect. The question of clinical usefulness is not settled."

INFECTIOUS DISORDERS

BACTERIAL MENINGITIS OUTCOME

The neurologic, psychological, and educational outcomes of bacterial meningitis in 130 children evaluated at a mean age of 8 years, and 6 years after their meningitis, are reported from the Department of Paediatrics and Clinical Epidemiology and Biostatistics Unit, University of Melbourne, and the Royal Children's Hospital, Victoria, Australia. Compared to controls, children with meningitis as a group were at greater risk (26.9%) for abnormal neurologic and audiologic sequelae, had lower IQs and neuropsychologic performance, and behavior and adaptive difficulties at school. Eleven (8.5%) had major deficits (IQ <70, seizures, hydrocephalus, spasticity, blindness, or severe to profound hearing loss); and 24 (18.5%) patients compared to 14 (10.8%) controls had minor deficits (IQ 70-80, inability to read, some hearing loss, speech problems, and behavior disorders). Those who suffered acute neurologic symptoms with the meningitis had a poorer outcome than those with uncomplicated meningitis or controls (39% vs 18% vs 11%). (Grimwood K, et al. Adverse outcomes of bacterial meningitis in school-age survivors. Pediatrics May 1995;95:646-656). (Reprints: Dr Keith Grimwood, Royal Children's Hospital, Parkville, Victoria 3052, Australia).

COMMENT. Even with optimal treatment, one in four children who recover from meningitis may have severe or functionally significant disabilities which affect academic performance. The poor outcome is not restricted to those having acute neurologic complications. All children recovering from meningitis should be followed carefully until school age to exclude learning, hearing, and neurologic disorders that may require treatment.

PERINATAL HIV ENCEPHALOPATHY

The characteristics and survival of 178 children with perinatally acquired human immunodeficiency virus (HIV) infection and encephalopathy are reported from the Centers for Disease Control and Prevention, Public Health Service, US Department of Health and Human Services, Atlanta, GA. Ten percent of HIV-infected children and 23% of children with AIDS had HIV encephalopathy that was diagnosed at a median age of 19 months. The estimated risk of HIV encephalopathy by age 1 year was 4%, and by age 4 years it was 14%. HIV encephalopathy correlated with an increased risk of cardiomyopathy, more hospitalizations, and with severe immunodeficiency. Estimated median survival after diagnosis was 22 months. (Lobato MN et al.