March 2001;138:332-337). (Reprints: Eugenio Mercuri MD, Department of Paediatrics, Hammersmith Hospital, Du Cane Rd, London W12 0HS, UK).

COMMENT. A standardized neurologic examination at approximately 1 year of age in an infant suffering hypoxic-ischemic encephalopathy at birth is a useful measure of functional motor outcome in later infancy and early childhood. The examination at 1 year correlates with the MRI findings at or soon after birth.

Prognostic value of EEG and MRI combined in full-term infants with acute encephalopathy. The correlation between an early EEG (within 72 hours after birth) and an MRI (end of 1 week) in assessing outcome of acute neonatal encephalopathy was studied in 25 full-term infants treated at Hammersmith Hospital, London, UK. (Biagioni E, Mercuri E, Rutherford M et al. Pediatrics March 2001;107:461-468). Both EEG and MRI were predictive of the outcome at 2 year follow-up. Seven infants with normal EEG background activity and 14 with discontinuous abnormality showed normal and abnormal outcomes, respectively. Eight with normal MRI or minimal abnormalities had a normal outcome, whereas 17 with moderate to severe MRI lesions were moderately to severely impaired neurologically and 5 died.

Hypothermic therapy for hypoxic-ischemic encephalopathy. The neurodevelopmental outcome at 18 month follow-up of 40 term infants with HIE treated with head cooling and systemic hypothermia or standard normothermic care was determined at the National Women's Hospital, Auckland, New Zealand. (Battin MR et al. Pediatrics March 2001;107:480-484). The mildly cooled groups showed no worsening of outcome and a trend toward better outcome (26% adverse outcome cf 44% among controls). The authors conclude that the short-term safety of cooling is demonstrated but the long-term efficacy is not proven. Multicenter trials are in progress.

METABOLIC DISORDERS

NEUROLOGIC OUTCOME OF INFANTILE HYPOGLYCEMIA

The neurologic development of 90 patients with persistent hyperinsulinemic hypoglycemia was studied retrospectively at Hospitalier Universitaire Necker-Enfants Malades, Paris, France. Treatment was surgical in 63 and medical in 27. Of 54 neonates, 8 were treated medically and 46 surgically. Of 36 with infancy-onset hyperinsulinism, 19 were treated medically and 17 had pancreatectomy. Development was normal in 74%. Severe mental retardation occurred in 8% of patients, and intermediate psychomotor disability in 18%. Epilepsy occurred in 18% of the total (in 24% of 54 neonates and 8% of 36 infants). The incidence of epilepsy was 7% among those with normal mental development and 57% in the severely retarded, at a mean follow-up age of 8 years. Microcephaly occurred in 10% of normal and 57% of severely retarded children. Neonatal onset was the main risk factor for severe retardation or epilepsy. Patients treated medically were less severely affected than those treated surgically. The outcome was not different in patients with diffuse and focal adenomatous hyperplasia with hyperinsulinism. (Menni F, de Lonlay P, Sevin C et al. Neurologic outcomes of 90 neonates and infants with persistent hyperinsulinemic hypoglycemia. Pediatrics March 2001;107:476-479). (Reprints: Jean-Jacques Robert MD, PhD, Diabete de l'Enfant et de l'Adolescent, Hopital Necker-Enfants Malades, 149 rue de Sevres, 75743 Paris Cedex 15, France).

COMMENT. Hyperinsulinism in infancy causes recurrent episodes of profound hypoglycemia, often <1 mmol/L, with diverse neurologic sequelae, including psychomotor retardation, learning disability, seizures, and microcephaly. Neonatal onset is associated with more complications and a greater need for surgical treatment. Early diagnosis and onset in infancy result in a slightly better prognosis.

In contrast to the frequent risk of mental impairment in neonates and infants with hyperinsulinemic hypoglycemia, young adults with insulin dependent diabetes and recurrent episodes of hypoglycemia have either a mild or negligible risk of cognitive impairment. (Deary IJ, Frier BM. BMJ 1996;313:767-8).

Offspring of diabetic Japanese mothers are at increased risk of lowered IQ scores at 3 years of age (Yamashita Y et al. Acta Paediatr 1996;85:1192-6). The risk of hypoglycemic brain damage is related inversely to the age of the patient. (See Progress in Pediatric Neurology III, 1997;p311).

MITOCHONDRIAL ENCEPHALOMYOPATHIES: INCIDENCE & DNA

The incidence, mortality, clinical features and DNA abnormalities of mitochondrial encephalomyopathies (ME) were evaluated in a population-based study of children from western Sweden conducted at The Queen Silvia Children's Hospital, Goteborg, Sweden. Thirty two patients under 16 years of age were diagnosed from 1984-1998. The incidence of ME in preschool children was 1 out of 11000, and the point prevalence in children <16 years of age was 1 out of 21000. Leigh's syndrome occurred in 1/32000 preschoolers, and Alper's syndrome and cytochrome C oxidase deficiency in 1/51000. Infantile onset of ME was frequent, the course severe, and mortality high. Patients with infantile onset ME had a median survival of 12 years. Complex I and IV deficiencies were the most common biochemical defects. The spectrum of disorders in children was different from that reported in adult hospital-based patients. Encephalopathies were more frequent in children, and mtDNA mutations were identified less frequently. Mitochondrial DNA point mutations, DNA deletions, and nuclear mutations in the SURF1 gene were identified in 4, 2, and 2 cases, respectively. (Darin N, Oldfors A, Moslemi A-R, Holme E, Tulinius M. The incidence of mitochondrial encephalomyopathies in childhood: Clinical features and morphological, biochemical, and DNA abnormalities. Ann Neurol March 2001;49:377-383). (Respond: Dr Niklas Darin, The Queen Silvia Children's Hospital, Department of Pediatrics, Sahlgrenska University Hospital-East, Goteborg University, S-416 85 Goteborg, Swedern).

COMMENT. The authors conclude that mitochondrial encephalomyopathies are relatively common neurometabolic disorders in childhood.

Among 51 patients with mitochondrial respiratory chain disease analyzed at the University of Newcastle upon Tyne, UK (Jackson MJ, Bindoff LA et al. Brain 1995;118:339-357), presenting symptoms in order of frequency included ptosis and ophthalmoplegia (20), lactic acidosis (10), seizures (6), myopathy (6), failure to thrive (6), and ataxia (5). The most useful confirmatory diagnostic test was histochemical analysis of muscle, and elevated CSF lactate was a good indicator of mitochondrial encephalopathy. (see Progress in Pediatric Neurology III, 1997;pp542-3).