infants recover when dietary chloride supplements are provided. Previous follow-up reports have drawn attention to developmental delay (Chutorian et al. Pediatr Neurol 1985; 1:334-341) and the present study demonstrates specific cognitive, language and attentional deficits in later childhood.

MAPLE SYRUP URINE DISEASE AND CEREBRAL EDEMA

Cerebral edema causing death in four children with maple syrup urine disease (MSUD) is reported from St. Christopher's Hospital for Children, Temple University School of Medicine, Philadelphia, PA. An intercurrent infection that caused severe dehydration and acidosis precipitated the cerebral edema when the children were three to five years of age. All four had been adequately managed with few problems before the intercurrent illness. The diagnosis of cerebral edema was established by autopsy in one patient and demonstrated by CT in two, one also having a subarachnoid hemorrhage. The authors recommend early hospitalization and cautious rehydration in all children with MSUD in whom intercurrent infections develop in association with decreased nutrient intake or vomiting, or both. Early treatment of dehydration and acidosis may prevent the catastrophic consequences observed (Riviello JJ Jr et al. Cerebral edema causing death in children with maple syrup urine disease. J Pediatr July 1991; 119:42-45).

COMMENT. Acute metabolic decompensation in MSUD during otherwise minor illnesses has generally been presumed to result from massive release of leucine from protein catabolism. The dynamics of protein metabolism and implications for management are reported from the Murdoch Institute and the Department of Dietetics, Royal Children's Hospital, Melbourne, Australia, and the Nutrition Research Group, Clinical Research Centre, Harrow, UK (Thompson GN et al. J Pediatr July 1991; 119:35-41). Fasting appeared to be a more important cause of increased leucine levels than the catabolic effect of infection. Branched-chain amino acid restriction should be commenced at the start of minor illness in children with MSUD and the intake of other nutrients should be increased. Dietary supplementation reduces the risk of metabolic decompensation during acute illnesses and early and meticulous treatment of MSUD results in intellectually normal children according to a study from the Division of Biochemical Development and Molecular Diseases, Children's Hospital of Philadelphia, University School of Medicine, Philadelphia (Kaplan P et al. J Pediatr July 1991; 119:46-50). Affected children treated presymptompatically had higher IQ scores than their siblings treated when their disease was symptomatic.

NON-MENKES TYPE COPPER DEFICIENCY

A two year old girl with copper deficiency progressive neuronal disorder and granulocytopenia is reported from the Department of Pediatrics, Kyoto University, Japan. A familial granulocytopenia was noted at one month

of age and she developed truncal ataxia and progressive neurologic regression at age two years, shortly after a ten day fever. She was admitted with a diagnosis of acute cerebellar ataxia and later developed dysarthria, mild dysphagia and was then unable to speak two word sentences. She was referred to the Utano National Hospital, Kyoto, three months after the onset. She was well nourished, her hair was normal, and she had no hepatosplenomegaly. Truncal and limb ataxia, horizontal nystagmus, increased reflexes and extensor plantar reflexes were noted. CT showed cerebellar atrophy and symmetric low-density areas in the bilateral periventricular white matter. Sural nerve biopsy revealed axonal degeneration. Blood lactate levels were high (25.8/mg/dl) and serum levels of copper and ceruloplasmin and urinary excretion of copper were low. Cultured skin fibroblasts showed normal copper uptake. Copper given orally 0.1 mg/kg daily for three days resulted in increased serum copper and ceruloplasmin levels, normal granulocyte count, hemoglobin level, blood lactate and pyruvate levels, but neither clinical improvement nor normalization of copper, lactate and pyruvate levels in the CSF were noted. The patient deteriorated slowly, the MRI showed extension of high signal T_2-weighted images and newly appearing lesions in the basal ganglia. At age three years (11 months after onset) she had spastic tetraplegia with respiratory difficulty and was placed on a respirator. (Fujii T et al. Non-menkes-type copper deficiency with regression, lactic acidosis, and granulocytopenia. Neurology August 1991; 41:1263-1266).

COMMENT. The clinical and MRI features of this patient ruled out the diagnosis of Wilson's disease, and Menkes' disease was unlikely in the absence of pili torti and with a normal copper uptake in cultured fibroblasts. The authors speculated that copper transport across the intestinal wall and across the blood brain barrier was impaired in their patient, and that long-term copper deficiency in the brain might have caused irreversible damage to cytochrome *c* oxidase production. (Dr. Fujii is presently at The Burke Rehab Ctr., White Plains, NY).

FOLIC ACID AND NEURAL TUBE DEFECTS

The results of a randomized double-blind prevention trial conducted at 33 centers in seven countries to determine the effects of folic acid supplements around the time of conception in the prevention of neural tube defects is reported by the MRC Vitamin Study Research Group, Department of Environmental and Preventive Medicine, Medical College of St. Bartholomew's Hospital, London EC1, UK. A total of 1,817 women at high risk of having a pregnancy with a neural tube defect were allocated at random to one of four groups - folic acid, other vitamins, both, or neither. Of 27 mothers giving birth to a child with a neural tube defect, six were in the folic acid groups and 21 in the two other groups, a 72% protective effect. Other vitamins showed no significant protective effect. Capsules for those in the folic acid group contained 4 mg of folic acid, one a day until 12 weeks of pregnancy (Wald N et al. Prevention of neural tube defects: results of the Medical Research Council vitamin study. Lancet July 20, 1991; 338:131-137).