macrocephaly, a white retina (lipid retinalis), and hepatosplenomegaly, may be amenable to treatment with a low fat diet that corrects the hyperchylomicronemia.

MEGALENCEPHALIC CYSTIC LEUKOENCEPHALOPATHY

A 41 year-old Japanese male diagnosed with megalencephalic leukoencephalopathy (MLC) with subcortical cysts (van der Knaap disease), presenting at 1 year of age with macrocephaly, slowly progressing after 5 years, seizures beginning at 11 years, completely bed ridden at 18 years, and with long survival, is reported from Tokyo Metropolitan Higashiyamato Medical Center for the Severely Disabled, Japan. His motor function had severely deteriorated and his cognitive function was at a 2 year level. MRI revealed marked cerebral atrophy, ventricular enlargement, and large cysts in frontoparietal and anterior temporal areas. A homozygous missense mutation was detected in the *MLC1* gene. (Saijo H, Nakayama H, Ezoe T et al. A case of megalencephalic leukoencephalopathy with subcortical cysts (van der Knaap disease): molecular genetic study. Brain Dev August 2003;25:362-366). (Respond: Dr Harumi Saijo, Tokyo Metroplitan Medical Center, 3-44-10 Sakuragaoka, Higashiyamato, Tokyo 207-0022, Japan).

COMMENT. Van der Knaap and colleagues described an infantile onset leukoencephalopathy with swelling and a discrepantly mild clinical course in 8 children in 1995. Mutations of MLC1 gene were identified as a cause of the leukoencephalopathy with subcortical cysts in 2001 (Leegwater et al). The changes in size of basal ganglia and diffuse white matter abnormalities that characterize Canavan and Alexander diseases are not evident in MLC.

ACUTE NECROTIZING ENCEPHALOPATHY

A 14-month-old girl who presented with sudden onset of unreponsiveness following a fever for 2 days developed decerebrate posturing within 9 hours after admission to Children's Memorial Hospital, Chicago, IL. She had no response to deep pain, her reflexes were exaggerated, and plantar responses were extensor. EEG showed slowing but no epileptiform discharges. Blood counts and chemistries were normal, except for elevated transaminases, with an aspartate aminotransferase level of 190 IU/L (normal, 22-59 IU/L). CSF analysis was normal. Initial head CT scan was normal, but 10 hours later, a second CT showed new bilateral thalamic hypodensities. MRI with MR angiography and venography, FLAIR imaging, and apparent diffusion coefficient (ADC) mapping confirmed thalamic and periventricular white matter involvement. Infectious, vascular, metabolic, and other etiologies were excluded. When discharged two weeks later to a rehabilitation facility with a diagnosis of acute necrotizing encephalopathy of childhood (ANEC), the child was blind, she responded only to noxious stimuli, and had diffuse spasticity. (Bassuk AG, Burrowes DM, McRae W. Acute necrotizing encephalopathy of childhood with radiographic progression over 10 hours. Neurology 13 May 2003;60:1552-1553). (Reprints: Dr Alexander G Bassuk, Children's Memorial Hospital, Division of Neurology, Box 51, 2300 Children's Plaza, Chicago, IL 60614).