

MAGNETIC RESONANCE SPECTROSCOPY IN CANAVAN'S DISEASE

The neuroradiological evaluation of Canavan's disease in a 38 month-old girl is reported from the Alfred I. duPont Institute, Wilmington, DE and Children's Hospital, Philadelphia, PA. A female child of Ashkenazic Jewish heritage presented at age 5 months with psychomotor retardation and macrocephaly. At age 7 months cerebral brain biopsy confirmed a suspected diagnosis of Canavan's disease. Over the last 30 months she developed severe spastic quadriplegia, optic atrophy and dysphagia which was treated by gastrostomy at age 27 months. CT showed diffuse symmetrical low attenuation values of the subcortical and deep cerebral white matter. MRI demonstrated symmetrical diffuse low signal density on T1-weighted images and high signal intensity on T2-weighted images. Magnetic resonance spectroscopy showed elevated levels of N-acetylaspartic acid in the occipital lobe. The in vivo measurement of N-acetylaspartic acid in the brain by magnetic resonance spectroscopy offered an additional non-invasive diagnostic test for Canavan's disease (Marks H G et al. Use of computer tomography, magnetic resonance imaging, and localized 1H magnetic resonance spectroscopy in Canavan's disease: a case report. Ann Neurol July 1991; 30:106-110).

COMMENT. Canavan's disease or spongy degeneration of the brain in infancy is a rare autosomal recessive leukodystrophy most prevalent among Ashkenazi Jews and Saudi Arabians. Matalon and co-workers have demonstrated an excessive amount of acetylaspartic acid in urine, blood, CSF and a deficiency of aspartoacylase in cultured skin fibroblasts and brain tissue in children with Canavan's disease (Matalon R et al. Am J Med Genet 1988; 29:463). The diagnosis may now be confirmed by magnetic resonance imaging followed by localized 1H magnetic resonance spectroscopy.

REYE SYNDROME AND ANTI-EMETICS

A drug-induced encephalopathy mainly by anti-emetics in two children with a diagnosis of Reye syndrome is reported from the Department of Paediatrics, University Hospital, Gasthuisberg, Leuven, Belgium. A 6 year-old girl presented with fixed stare, stupor alternating with agitation, confusion and wild delirium and signs of liver disease. The illness was preceded by headache, muscular aching, abdominal pain and anorexia. Vomiting had been treated with domperidone and metoclopramide. Dystonic reactions and oculogyric crises occurred one day before referral. When medication was omitted the neurological examination became normal 24 hours later. The preceding viral syndrome was due to influenza-A. The patient recovered completely after 10 days. Case 2 consisted of an 11 month-old baby who later proved to have cytomegalovirus infection. Anti-emetic domperidone had been prescribed for severe vomiting and 60 hours later bizarre arm movements and eye movements developed. A diagnosis of Reye syndrome prompted admission to hospital. The infant had impaired consciousness, stupor and