NUTRITIONAL DISORDERS

WERNICKE’S ENCEPHALOPATHY

Two cases of Wernicke’s encephalopathy (WE) diagnosed at autopsy are reported from the Depts of Pediatrics and Pathology, British Columbia’s Children’s Hospital, and the University of British Columbia, Vancouver, Canada. A 5 1/2 year old child was persistently febrile due to repeated pulmonary infections and died of respiratory failure after 9 weeks in coma following cold-water submersion. Her average daily nutritional intake (1,200 calories, 2.2 mg thiamine) was deficient in calories and her thiamine intake, adequate for a healthy child, was apparently insufficient for a severely ill child with high carbohydrate intake. In a 9 month old infant with Zellweger syndrome who died of hemorrhage from esophageal varices due to cirrhosis of the liver, feeding was made difficult because of frequent seizures and no vitamin supplements had been given. The mamillary bodies and periventricular areas in the brainstem showed spongy change, persistence of neurons, and astrocytosis characteristic of WE. The diagnosis was not suspected during life. (Sear MD, Norman MG. Two cases of Wernicke’s encephalopathy in children: An underdiagnosed complication of poor nutrition. Ann Neurol July 1988;24;85-87).

COMMENT. WE in alcoholic adults classically presents with ataxia, confusion, ophthalmoplegia, and coma and death if untreated. The onset and course may be acute, subacute, or chronic. In 6 cases cited in infants, deterioration was rapid, with lethargy, apneic spells, hypertonia, and hypothermia. The authors emphasize that WE often occurs without alcoholism, it is preventable and treatable with thiamine supplements, and the diagnosis should be suspected in malnourished infants, especially those with persistent vomiting.