FATTY LIVER AND MCT DIET IN INTRACTABLE EPILEPSY

Fatty infiltration of the liver in 4 children treated with the medium chain triglyceride (MCT) diet is reported from the Depts of Paediatrics and Radiology, Leeds General Infirmary, Belmont Grove, Leeds, England. This was not associated with hepatic dysfunction and resolved after discontinuing the diet. The patient's ages ranged from 4 to 12 years. The seizures were astatic myoclonic in pattern and resistant to medications, including sodium valproate, clobazam, or carbamazepine. The duration of the diet was 2 to 3 years at time of diagnosis by ultrasound scan and, in one case, by liver biopsy. Triglyceride and cholesterol levels were normal in 2 and liver function tests normal in 3 patients tested. Seizure control was improved during treatment with the MCT diet. (Beverley D, Arthur R. Fatty liver and medium chain triglyceride (MCT) diet. Arch Dis Child July 1988;63:840-842).

> **COMMENT**. The mechanism of the fatty liver infiltration was undetermined. The standard ketogenic diet is not always associated with a significant increase in serum lipids but it is accompanied by a fall in blood pH, standard bicarbonate, and blood sugar. (Millichap JG et al. Amer J Dis Child 1964;107:593) The accumulation of excess hydrogen ion within the liver cell is cited as one possible mechanism of fatty change in the liver. The concomitant therapy with anticonvulsant drugs, particularly valproate, might also be contributory.

DEGENERATIVE DISORDERS

RETT SYNDROME

The case-histories of 7 girls and 1 woman, 2 to 25 years of age, are reported from the Clinical Genetics and Child Developmental Center, Department of Maternal and Child Health, Dartmouth Medical School, Hanover, NH. Diagnosis was made after 5 years of age in 4 of the patients when the characteristic hand wringing, hand washing movements were first noted. In addition to these stereotypic hand behaviors, other clinical features included loss of hand function, hyperventilation, bruxism, irritability or self-injury, sleep disturbance, strabismus, seizures, scoliosis, ataxia, and hypotonia in infancy. Head growth deceleration, short stature and retarded rate of growth occurred in almost all cases. Three were small for gestational age at birth and experienced neonatal feeding problems. Previous diagnoses included Angelman syndrome, encephalopathy, and encephalitis, and in 1 case the syndrome appeared to develop as a reaction to pertussis vaccine. Treatment was symptomatic and supportive. Music and motion such as rocking or riding in a car had a calming effect, particularly when screaming attacks and sleep disturbances were troublesome. The absence of a "biological marker" for Rett syndrome makes diagnosis difficult and may lead to confusion with other defined neurodegenerative disorders, such as leukodystrophies, spinocerebellar heredoataxias, neuronal ceroid lipofuscinosis, and ornithine transcarbamylase deficiency. (Moeschler JB et al. Rett Syndrome: natural history and management. Pediatrics July 1988;82:1-10).