11.9-21.4 mcM). Treatment with valproic acid was not responsible for the low TC. TC was unrelated to a low blood glucose or time to achieve ketosis. A decrease in TC in the first months of diet treatment was followed by stabilization and a slight increase with long term treatment. None developed clinical signs of carnitine deficiency, and only 19% of patients received supplements of carnitine. An increased acyl/free carnitine ratio is dependent on the level of ketosis during KD treatment, and is not a useful measure for carnitine insufficiency. (Berry-Kravis E, Booth G, Sanchez AC, Woodbury-Kolb J. Carnitine levels and the ketogenic diet. Epilepsia Nov 2001;42:1445-1451). (Reprints: Dr E Berry-Kravis, Rush-Prebyterian-St Luke's Medical Center, 1725 West Harrison St, Suite 718, Chicago, IL 60612).

COMMENT. The majority of patients on the ketogenic diet do not develop low carnitine plasma levels, and those with low levels are asymptomatic. TC should be checked at 1 to 6 months of treatment, especially for patients with low-normal TC at KD initiation.

PAROXYSMAL TONIC UPGAZE: AGE OF ONSET AND PROGNOSIS

Six children who developed paroxysmal tonic upgaze (PTU) at 2.6 to 7.4 years of age were examined at least once per year for a 10 year period of follow-up at the Departments of Paediatrics and Ophthalmology, University of Chieti, Italy. Bouts of sudden sustained upward deviation of the eyes lasted for 10 to 20 seconds each, they appeared in small clusters, and were not associated with loss of consciousness. PTU was typically exacerbated by fatigue and relieved by sleep. Three patients were male and 3 female. Two had a febrile illness preceding the onset, 2 had a personal history of febrile seizures, 1 a father with a history of febrile seizures, and 1 a mother with migraine. One had an epileptiform interictal EEG, with temporo-occipital spikes, mainly in the left hemisphere. MRI, neurologic, and psychological examinations, including WISC-R or Stanford-Binet Intelligence Scales were normal. Episodes of PTU resolved spontaneously after 1 to 4 years, without treatment. (Verrotti A, Trotta D, Blasetti A et al. Paroxysmal tonic upgaze of childhood: effect of age-of-onset on prognosis. Acta Paediatr Nov 2001;90:1343-1355). (Respond: Dr Albert Verrotti, Department of Pediatrics-Policlinico Colle Dell'Ara, University G D'Annunzio, Via dei Vestini 5, IT-66013 Chieti, Italy).

COMMENT. The first description of the syndrome as "benign paroxysmal tonic upgaze of childhood" was reported in 4 patients by Ouvrier RA and Billson F (J Child Neurol 1988;3:177-180). Some subsequent reports have included associated neurologic abnormalities, including ataxia (Deonna T et al. 1990), developmental delay, and abnormal brain MRI. Improvement following levodopa therapy in one child suggested a possible dystonia (see Progress in Pediatric Neurology II, PNB Publ, 1994;p146). Most patients have an early spontaneous resolution of the disorder. The authors suggest that the outcome may be age-dependent, their older patients having a more benign form of the syndrome than that described in younger infants. An immature cortico-mesencephalic control of vertical gaze is postulated as the pathophysiology.

MOVEMENT DISORDERS

RIGHT FRONTAL WHITE MATTER AND TOURETTE SYNDROME

An MRI volumetric analysis of frontal and nonfrontal gray and white matter was performed in 11 boys with Tourette syndrome (TS) only, 14 with TS + ADHD, 12 with ADHD only, and 26 healthy boys, at the Kennedy Krieger Institute,