Follow-up ranged from 12 months to 8 years. No patient died. Time to biochemical diagnosis (high urine sulfite and low plasma urate, high urine purine metabolites [xanthines] and 5-sulfocysteine) ranged from 4 days to 4 years (median 3 months). EEG abnormalities included burst suppression pattern and seizure activity. Brain MRI showed cerebral infarction in all except one with atypical onset. Distinctive features seen in an early brain MRI were acute infarction of the globus pallidi and subthalamic regions with older cerebral hemisphere infarction, chronic lesions suggestive of a prenatal insult, pontocerebellar hypoplasia and retrocerebellar cyst, and a band at the cortical/subcortical white matter. Sequential imaging showed progressive pontine atrophy and enlargement of retrocerebellar cyst. Early brain MRI (<1 week) abnormality may lead to early prenatal diagnosis, mutational confirmation and therapeutic intervention.

CO-ENZYME Q10 AND L-CARNITINE IN CYCLIC VOMITING

Researchers at Childrens Hospital Los Angeles, CA conducted a retrospective chart review of 42 patients treated with co-enzyme Q10 and L-carnitine, and the addition of amitryptiline (or cyproheptadine for <5 year olds) in refractory cases. Patients ate 3 meals and 3 snacks a day and avoided fasting. Treatment was monitored with blood levels: co-enzyme Q10 >3.0 mg/L; L-carnitine >40 micromolar; amitryptiline >150 ng/ml; cyproheptadine dose 0.5 mg/kg/day. In 30 cases with available outcome data, vomiting episodes resolved in 23, and improved by >50-75% in 4 cases. Treatment with mitochondrial-targeted cofactors (co-enzyme Q10 and L-carnitine) plus amitryptiline was highly effective in the prevention of vomiting episodes. (Boles RG. High degree of efficacy in the treatment of cyclic vomiting syndrome with combined co-enzyme Q10, L-carnitine and amitryptiline, a case series. BMC Neurology 2011;11:102-107). (Respond: E-mail: rboles@chla.usc.edu).

COMMENT. The author observes that these findings need confirmation by a prospective blinded trial in unselected cases. Cyclic vomiting is usually regarded as a form of migraine, and mitochondrial dysfunction may be a factor in etiology of both cyclic vomiting and migraine. Cyclic vomiting, sometimes associated with epilepsy and epileptiform EEG, may also respond to treatment with antiepileptic agents (topiramate, valproate)(Olmez A et al. Pediatr Neurol 2006;35(5):348-351)(Millichap JG, Lomboiroso CT, Lennox WG. Pediatrics 1955;15(6):705-714).