MA, Tai P. Mitochondrial encephalopathy with lactic acidosis and stroke-like episodes (MELAS) may respond to adjunctive ketogenic diet. **Pediatr Neurol** 2014 May;50(5): 498-502).

COMMENTARY. A ketogenic diet should be considered for treatment of intractable seizures and stroke-like episodes related to mitochondrial respiratory chain complex (MRC) defects. In a Korean study of 14 patients with MRC defects and various seizure types (5 with infantile spasms, 4 with Lennox-Gastaut syndrome, 1 with Landau-Kleffner syndrome), 50 – 90% seizure control was obtained with the ketogenic diet [1]. A subsequent study in Korea involving 24 cases of MRC defect with seizures, the ketogenic diet controlled seizures in 75% patients [2].

References.
1. Kang HC, et al. Epilepsia. 2007 Jan;48(1):82-8.
2. Lee YM, et al. Epilepsia. 2008 Apr;49(4):685-90.

HEADACHE DISORDERS

HEADACHE AND MIGRAINE WITH SICKLE CELL DISEASE

Investigators from University of Texas Southwestern Medical Center, Dallas, TX, and other centers in the US and London, UK, studied risk factors for headache and migraine in 872 children, age 5 to 15 years (mean age, 9.1 years), with sickle cell disease (hemoglobinSS or hemoglobinSb-thalassemia) and no history of overt stroke or seizures. Recurrent headaches were reported in 317 (36.4%) and migraines in 132 (15.1%). Both were associated with lower hemoglobin and higher rate of hospitalization for pain events requiring hospitalization for treatment with opioids in the previous 3 years. Only six of 317 (1.9%) children reporting recurrent headaches were receiving medication for headache prophylaxis. The prevalence of silent cerebral infarct, diagnosed by MRI and neurological examination, was similar in patients with recurrent headaches and in those without headaches (32.8% and 29%, respectively; P=0.241). Older age, lower Hgb concentration, and higher pain event rate were associated with recurrent headaches and migraines. (Dowling MM, Noetzel MJ, Rodeghier MJ, et al. Headache and migraine in children with sickle cell disease are associated with lower hemoglobin and higher pain event rates but not silent cerebral infarction. **J Pediatr** 2014 May;164(5):1175-1180.e1).

COMMENTARY. Isolated recurrent headaches or migraine in neurologically normal children with sickle cell disease (SCD) might not necessitate additional evaluation with imaging studies, but new severe headaches presenting acutely warrant further investigation, especially when associated with acute CNS events. In a study of children with SCD who presented acutely with headache, headache was the chief complaint in 3.8% of acute care visits, and acute CNS events occurred in 16.9%. Factors associated with acute CNS events included older age, history of stroke, TIA, or seizure, focal neurological findings, and elevated platelets [1].

References.
1. Hines PC, et al. J Pediatr. 2011 Sep;159(3):472-8.