COMMENT. Hashimoto encephalopathy (HE) is characterized by seizures, neurologic and psychiatric manifestations, and elevated titers of serum anti-thyroid antibodies. It is responsive to corticosteroids. Plasmapheresis is a novel method of acute treatment. HE should be considered, along with anti-N-methyl-D-aspartate-receptor, voltage-gated potassium channel antibody-associated limbic encephalitis, and herpes simplex virus encephalitis, in the differential diagnosis of a child with acute personality changes and seizures resistant to antiepileptic medication. The pathogenesis of HE is associated with high serum anti-thyroid antibody titers; thyroid hormone levels are usually normal or slightly low. An autoimmune disease process is likely.

POLG NOVEL MUTATION WITH ALPERS SYNDROME

Researchers at University Hospital, Berne, Switzerland describe the molecular genetic analysis of *POLG* in a 3.5 years old boy with VPA-induced fatal liver failure and encephalopathy (Alpers-Huttenlocher syndrome, AHS). Mutations in the *POLG* gene are a common cause of inherited mitochondrial disease in children and adults. They are involved with various neurodegenerative diseases, including Alpers syndrome, and result in accumulation of multiple mtDNA deletions and/or depletions of mtDNA in muscle, brain and liver. Some *POLG* mutations lead to a range of clinical phenotypes that predispose to fatal liver failure after exposure to VPA. *POLG* analysis in mitochondrial diseases helps in confirmation of AHS and optimizes clinical management. (Schaller A, Hahn D, Jackson CB, et al. Molecular and biochemical characterization of a novel mutation in *POLG* associated with Alpers syndrome. **BMC Neurology** 2011;11:4-11). (Respond: Dr Andre Schaller. E-mail: andre.schaller@insel.ch).

COMMENT. The study extends the list of *POLG* mutations associated with VPA hepatoxicity. A report of reversible valproate hepatotoxicity due to mutations in mitochondrial DNA polymerase gamma *(POLG1*) is cited. (McFarland R et al. **Arch Dis Child** 2008;93(2):151-153).

DEMYELINATING DISEASE

HEAD AND BRAIN SIZE IN PEDIATRIC MULTIPLE SCLEROSIS

Researchers at the Montreal Neurological Institute, the Hospital for Sick Children, Toronto, Canada; and Department of Neurology, Rennes, France conducted MRI measurements of whole brain and regional white matter, gray matter, and deep gray matter structure volumes in 38 patients (mean age 15.2+/-2.4 years) with pediatric-onset relapsing-remitting multiple sclerosis (MS). Mean age at MS onset was 12.1 years; mean disease duration 3.1 years. Values obtained from sex-matched healthy controls enrolled in the MRI Study of Normal Brain Development were used as controls. The intracranial volume and normalized brain volume z scores were significantly lower in patients with MS compared with controls. Thalamic volumes in MS patients were lower even after correction for global brain volume decreases. Reduced thalamic and brain volumes