confusion, disorientation, behavioral changes, or other cognitive impairments, with or without inflammation of brain tissue. Encephalitis is characterized by brain inflammation resulting from direct infection of the brain parenchyma (e.g. Bartonella or influenza), a post-infectious process as in ADEM or a noninfectious condition such as NMDAR encephalitis. The definition covers infectious and noninfectious encephalitis and encephalopathy of presumed infectious etiology. Specific etiologies are identified in <50% of cases.

Proteomes in plasma and CSF of children with cerebral malaria were found to differ from those with acute bacterial meningitis and nonspecific encephalopathies. Pathogenic states in children with impaired consciousness in malaria endemic areas could be reflected by changes in protein biomarkers in both plasma and CSF. (Gitau EN et al. J Infect Dis 2013 Nov 1;208(9):1494-503).

POSTERIOR REVERSIBLE ENCEPHALOPATHY SYNDROME

Investigators at Children's Hospital of Montefiore, Albert Einstein College of Medicine, NY, determined the incidence of posterior reversible encephalopathy syndrome (PRES) in a pediatric critical care unit. Ten patients <21 years of age with PRES (incidence of 1 in 259 admissions, 0.4%) were studied. Nine patients presented with generalized tonic and/or clonic seizures. Continuous EEG showed generalized slowing but no epileptiform activity. Risk factors included hypertension, cytotoxin medication use, and anemia. Comorbidities included systemic lupus erythematosus, inflammatory bowel disease, ulcerative colitis, liver cirrhosis, pulmonary embolism, renal insufficiency, septic shock, and acute chest syndrome. One-year follow-up showed no residual neurological deficits and resolution of white matter signal abnormalities on neuroimaging. (Raj S, Overby P, Erdfarb A, Ushay HM. Posterior reversible encephalopathy syndrome; incidence and associated factors in a pediatric critical care population. Pediatr Neurol 2013 Nov;49(5):335-9). (Response: Dr Raj, The Children's Hospital at Montefiore, Bronx, NY 10467. E-mail: drshashiraj@gmail.com).

COMMENT. PRES, also referred to as hypertensive encephalopathy or reversible posterior leukoencephalopathy syndrome, is a clinical syndrome that results from disruption of the blood-brain barrier and vasogenic edema, demonstrated on MRI with hyperintense signals in the posterior cerebral white matter.

PRES in an infant 35 days old is reported from the Mayo Clinic. The syndrome is rare in children less than 1 year. The infant had a history of obstructive sleep apnea, laryngomalacia, and choanal atresia. While undergoing bronchoscopy, she developed apneic episodes with stiffening of extremities. EEG revealed occipital lobe onset seizures, and MRI showed hyperintense T2 signal in both posterior temporal and parietooccipital lobes. A labile blood pressure was normalized and seizures abated with fosphenytoin and levetiracetam. At 3 month of age, resolution of MRI abnormality confirmed the diagnosis of PRES. (Mrelashvili A, Watson RE, Wong-Kisiel LC. **Pediatr Neurol** 2013 Nov;49(5):387-8). **PRES and risk of epilepsy.** The incidence of subsequent epilepsy was 2.25 fold higher in patients with hypertensive encephalopathy (HE) than in controls, in a nationwide population-based study in Taiwan. The incidence of epilepsy was higher in men, younger patients with HE, and in those with brain disorders. (Chung TT, et al. **Epilepsy Behav** 2013 Nov;29(2):374-8).

SEIZURE DISORDERS

SCN1A AND SUSCEPTIBILITY TO MTL EPILEPSY, HIPPOCAMPAL SCLEROSIS AND FEBRILE SEIZURES

Investigators at the Department of Clinical and Experimental Epilepsy, Institute of Neurology, Queen Square, London, and other centers in the UK and Europe conducted a genome-wide association study in 1018 people with mesial temporal lobe epilepsy (MTLE) with hippocampal sclerosis and 7552 control subjects, with (n=757) and without (n=803) a history of febrile seizures. Meta-analysis revealed a genome-wide significant association for MTLE with hippocampal sclerosis with febrile seizures at the sodium channel gene cluster on chromosome 2q24.3. No genetic association with febrile seizures was found in a cohort of 172 individuals with febrile seizures who did not develop epilepsy during follow-up to age 13 years. The findings suggest SCN1A involvement and common genetic variation in the epilepsy syndrome of MTLE, hippocampal sclerosis with febrile seizures. (Kasperaviciute, D, Catarino CB, Matarin M, et al. Epilepsy, hippocampal sclerosis and febrile seizures linked by common genetic variation around SCN1A. **Brain** 2013 Oct;136(Pt 10):3140-50). (Response: Sanjay M Sisodiya PhD, FRCP. Department of Clinical and Experimental Epilepsy, UCL Institute of Neurology, 33 Queen Square London, WC1N 3BG, UK. E-mail: s.sisodiya@ucl.ac.uk).

COMMENT. In addition to MTLEHS + FS, genetically-determined, epilepsy syndromes in which febrile seizures are a prominent feature include Dravet syndrome, and genetic epilepsy with febrile seizures plus. The authors suggest that focusing on clinically recognized syndromes or constellations (Berg AT, et al. **Epilepsia** 2010 Apr;51(4):676-85) could reduce heterogeneity before genomic analyses and lead to discovery of more narrowly-defined syndromes. Genetic association studies should uncover the cause of some epilepsies and facilitate prevention or a cure.

TNK2 mutations in severe autosomal recessive infantile onset epilepsy with intellectual disability. The proband, a girl, presented at age 19 months with focal seizures resembling MTLE, and characterized by unresponsiveness, hypertonia, automatisms and secondary generalization. Seizures recurred several times a day and were refractory to medication. Birth and early development were normal, and cognitive regression with autistic features occurred soon after seizure onset. MRI was normal. Video-EEG recording and PET scan showed right anteromedial temporal lobe seizure onset, but temporal lobectomy at age 4.5 years failed to control seizures. The resected tissue showed no abnormality. Two younger brothers had a similar history to that of the proband. The cognitive regression with absence of myoclonus, normal MRI, and unremarkable interictal EEG distinguish this phenotype from known infantile onset