not rule out the diagnosis and treatment with immunosuppression. (Eibers J, Halliday W, Hawkins C, Hutchinson C, Benseler SM. Brain biopsy in children with primary small-vessel central nervous system vasculitis. **Ann Neurol** Nov 2010;68:602-610). (Respond: Dr Benseler, Division of Rheumatology, Dept Paediatrics, Hospital for Sick Children, University of Toronto, 555 University Ave, Toronto, Ontario, M5G 1X8, Canada. E-mail: susanne,benseler@sickkids.ca).

COMMENT. PACNS is an immune-mediated, acquired inflammatory disease involving either small or large CNS blood vessels. In children, the disease may present acutely with seizures and refractory status epilepticus, subacutely with focal neurological deficits, or chronically with headaches and cognitive decline. In an editorial, Hunder GG and Brown RD Jr of the Mayo Clinic, Rochester, MN, and Salvarani C of Italy question whether childhood PACNS is a single disease entity. (**Ann Neurol** 2010:68:573-574). They report 8 adult cases with similar findings to those in children, except in adults, focal neurological abnormalities occur in all 8 (only 2 of 13 in children), seizures are absent, and ESR is normal. (Salvarini C et al. **Medicine** (Baltimore) 2008;87:264-271). Histological findings are also different in adult cases. Small vessel PCNSV in adults is less clearly defined than in children.

Diagnosis of cPACNS should be considered and brain biopsy performed in a child presenting with new-onset seizures, severe headaches, and cognitive decline, associated with elevated ESR, CSF pleocytosis and elevated protein, and MRI evidence of vasculitis.

PAROXYSMAL DISORDERS

COURSE OF ALTERNATING HEMIPLEGIA OF CHILDHOOD

The natural history and long-term outcome of alternating hemiplegia of childhood (AHC) was studied by questionnaire within a large cohort of 157 patients, as part of the European Network for Research on Alternating Hemiplegia (ENRAH) project. The study was largely retrospective and, for 2 years, prospective. Patients were aged from 9 months to 52 years at time of inclusion. Median age at diagnosis was 20 months. All had hemiplegic attacks, 91% had abnormal ocular movements, 86.5% reported episodes of bilateral weakness, 88% tonic/dystonic attacks, 53% epileptic seizures, 72% had chorea, and 92% mental retardation. Premonitory signs or aura were reported in 41% patients, and sleep inhibited attacks in the majority (83%) of cases. A relaxing environment (music, massage) had a beneficial effect. Children with abnormal ocular movements and hypotonia improved in adulthood, whereas the severity of other symptoms in the whole cohort did not change over the course of the illness. Gait was unsteady in 84%, and school attendance and employment were severely impacted. Seven patients died, some during severe plegic attacks or seizures. Severe hemiplegic/dystonic attacks did not increase risk of poor outcome. The natural history of AHC in individual patients was highly variable and fluctuating and, as a group, did not indicate a progressive and degenerative course. Risk of sudden death in a minority was associated with more severe neurological impairment. Various treatments were employed, all receiving flunarizine as

prophylactic agent of non-epileptic events, partially effective in 74%. AEDs had no significant effect on non-paroxysmal events and only occasional control of paroxysmal symptoms. Diazepam sometimes reduced the duration of dystonic attacks. (Panagiotakaki E, Gobbi G, Neville B, et al. Evidence of a non-progressive course of alternating hemiplegia of childhood: study of a large cohort of children and adults. **Brain** December 2010;133:3598-3610). (Respond: Pr Alexis Arzimanoglou, Institute for Children and Adolescents with Epilepsy, Hopital Femme Mere Enfant, Hospices Civils de Lyon, 59 Boulevard Pinel, 69677, Lyon, France. E-mail: aarzimanoglou@orange.fr).

COMMENT. AHC is characterized by episodes of hemiplegia, dystonic and other non-epileptic paroxysmal events (paroxysmal nystagmus, autonomic disorder), epileptic seizures, and global neurological impairment. Between episodes, patients are neurologically impaired, with ataxia, athetosis or chorea, the majority developing mental retardation. Onset is in infancy, sometimes in the neonatal period. Consciousness is preserved during episodes. The pathophysiology has been compared to hemiplegic migraine, but differs in the development of fixed neurological impairment and retardation.

NEONATAL DISORDERS

HYPOXIC-ISCHEMIC-ENCEPHALOPATHY STUDIED BY EEG AND MR SPECTROSCOPY

Amplitude-integrated EEG (a-EEG) time course during the first 24 hrs of life was related to brain metabolism changes detected by proton MR spectroscopy (H-MRS) at 7-10 days of post-natal life in non-cooled term newborns with hypoxic-ischemic-encephalopathy (HIE). In a study at the Neonatology Unit, University of Bologna, Italy, a-EEG at 6, 12 and 24 hrs of life in 27 of 31 patients who survived was correlated with outcome; the a-EEG showed improvement in newborns with normal H-MRS and good outcome and a deterioration in those with abnormal H-MRS and poor outcome. a-EEG time course in the first 24 hrs of life may document the severity and evolution of cerebral damage following a perinatal IH event. Both H-MRS and a-EEG show a good correlation with outcome. (Ancora G, Soffritti S, Lodi R, et al. A combined a-EEG and MR spectroscopy study in term newborns with hypoxic-ischemic-encephalopathy. **Brain Dev** 2010;32:835-842). (Respond: E-mail: gina.ancora@unibo.it).

COMMENT. Prompt evaluation of newborns with HI encephalopathy to determine severity of brain damage and prognosis is important in therapy and follow-up. Seizure activity detected by a-EEG was associated with poor outcome only in patients with abnormal a-EEG background pattern. A normal a-EEG background at 24 hrs of life was predictive of normal outcome, a finding in agreement with that of van Rooij et al. 2005 (cited by authors), who reported that the background activity at onset of seizures is the best predictor of outcome. H-MRS shows a better correlation with outcome compared to conventional MRI. a-EEG and H-MRS have a similar sensitivity and specificity for prediction of outcome.