H-IE is in progress, with a view to prevention of West syndrome by early administration of ACTH [Millichap JJ, prepublication observations].

References

1. Millichap JG, Bickford RG. JAMA. 1962;182(5):523-527.
2. Lux AL, et al. Lancet Neurol. 2005 Nov;4(11):712-7.
3. Gano D, et al. Pediatr Neurol. 2013 Dec;49(6):401-5.
4. Suzuki M, et al. Epilepsia. 2003 Mar;44(3):443-6.
5. Kato T, et al. Epilepsia. 2010 Dec;51(12):2392-6.

LONG-TERM OUTCOME OF JUVENILE MYOCLONIC EPILEPSY

Investigators at Epilepsy Centers in Berlin, Germany, performed a retrospective study of seizure outcome in 66 patients with juvenile myoclonic epilepsy (JME) after a mean follow-up time of 44.6 years (20-69 years); 59.1% of patients remained seizure-free for at least 5 years before the last contact. Of seizure-free patients, 28 (71.8%) remained on AEDs and 11 (28.2%) were off medication for at least the last 5 years. Absence seizures at onset were an independent predictor of an unfavorable outcome and JME persistence. (Senf P, et al. Prognosis of juvenile myoclonic epilepsy 45 years after onset. Seizure outcome and predictors. **Neurology** 2013 Dec 10;81(24):2128-33).

COMMENTARY. JME is usually described as a chronic disorder requiring lifelong therapy [1]. In contrast, recent long-term follow-up studies point to a more favorable prognosis, allowing cautious withdrawal of medication after long seizure control [2]. In an editorial comment, a trial of older medications, including primidone and acetazolamide, is recommended in patients with refractory JME [3].

References

1. Penry JK, et al. Epilepsia. 1989;30 Suppl 4:S19-23.
2. Camfield CS, Camfield PR. Neurology. 2009 Sep 29;73(13):1041-5.
3. So NK. Neurology. 2013 Dec 10;81(24):2132.

ENCEPHALOPATHIES

ENCEPHALOPATHIC SUSAC SYNDROME

Investigators from Tubingen and Munster, Germany, report the case of a 32-year-old woman who at 32 weeks of pregnancy developed a change in personality, disorientation, ataxia, dysarthria, and hemispasticity. MRI showed multiple diffuse T2-intense lesions, many involving the corpus callosum. CSF showed mild lymphocytic pleocytosis (13 cells/mcl) and elevated protein (1,800 mg/l) and no oligoclonal bands. A bluish, net-like exanthema on trunks and legs was diagnosed as livedo racemosa. Weeks later, she was readmitted with visual field loss and ischemic damage to both retinae, and bilateral hearing loss. With a diagnosis of Susac syndrome, IV cyclophosphamide, the standard treatment, was not instituted because of risk of permanent infertility. A combination of prednisolone, IV immunoglobulins, mycophenolate mofetil, and methotrexate provided a sustained control of symptoms. (Engeholm M, et al.