

COMMENT. Reading epilepsy, a form of reflex epilepsy, first described by Bickford R at the Mayo Clinic in 1956, occurs in 2 forms, primary or idiopathic and secondary and attributed to a structural brain lesion. Levetiracetam may prove superior to valproate and clonazepam in treatment, and spontaneous remission of primary reading epilepsy may occur in idiopathic cases. Generalized tonic-clonic seizures may be avoided by stopping reading at onset of mouth jerking, but anticonvulsant medications are usually required. The left predominance of the EEG seizure discharge in cases of reading epilepsy is in keeping with reports of reduced white matter integrity in the left arcuate fasciculus of dyslexics. (Vandermosten M et al. *Brain* 2012;135:935-948; see *Ped Neur Briefs* 2012 April;26(4):32).

**Neurocognitive endophenotype with rolandic epilepsy.** Children with rolandic epilepsy (RE) have reading, language, and attention disorders. In 13 probands with RE and 11 epilepsy-free siblings who completed a neurocognitive evaluation, 9% of siblings and 31% of probands were reading impaired, 36% of siblings and 54% of probands were language impaired, and 70% of siblings and 67% of probands had attention impairments. Probands and siblings showed a similar profile of deficits in language and attention. Early psychological evaluation and academic intervention may benefit children with RE. (Smith AB, et al. *Epilepsia* 2012 March;53(4):705-711).

## INFLAMMATORY DISORDERS

### **HEMOPHAGOCYtic LYMPHOHISTIOCYTOSIS**

Investigators at the Hopital Bicetre and other hospitals in Paris, France studied the CNS symptoms at onset of primary hemophagocytic lymphohistiocytosis (HLH), and differentiated these from other CNS inflammatory diseases. At disease onset, 46 patients included in the study had a median age of 2.5 months (range 0-190 months), and 34 (74%) were under 12 months. Familial HLH was the most frequent genetic defect. Neurologic symptoms were present at onset in 29 children (63%) and were associated with fever, hepato-splenomegaly or lymphadenopathy in 26. The 3 main neurologic symptoms were seizures, impaired consciousness, and meningismus. Microcephaly was diagnosed at birth in 6 boys and developed shortly after birth in 1. CSF was abnormal in 50%. MRI was abnormal in 7 (15%). Unlike patients with ADEM, MRI showed symmetric periventricular lesions, without thalamic and brainstem involvement and with infrequent hyposignal intensity on T1. At end of follow-up (3.6 +/- 3.6 years), 18 (39%) patients had died; 17 of 28 (61%) surviving patients were normal neurologically, 5 (18%) had a severe neurologic outcome, and 6 (21%) had mild cognitive deficits. Risk of abnormal neurologic outcome was related to neurologic symptoms, MRI abnormalities, or abnormal CSF at onset, and was not influenced by age or type of genetic defect. (Deiva K, Mahlaoui N, Beaudonnet F, et al. CNS involvement at the onset of primary hemophagocytic lymphohistiocytosis. *Neurology* 2012 April 10;78:1150-1156). (Response and reprints: kumaran.deiva@bet.aphp.fr).

COMMENT. Neurologic symptoms are frequent at onset of primary HLH and 50% patients have abnormal CSF. Early diagnosis is essential for a favorable outcome.