

lesions of systemic lupus erythematosus and supportive of an immunopathogenic mechanism for dyslexia.

### LANGUAGE DISORDERS

#### OUTCOME OF ACQUIRED APHASIA

The effects of age at onset, etiology, severity, and type of aphasia on the course and outcome were investigated in a group of 28 aphasic children at the Department of Neurology, University Hospital Rotterdam-Dijkzigt, the Netherlands. Head injury was the cause in eight patients with onset between 4 and 11 years, vascular diseases accounted for seven cases with age of onset between 3 and 13 years, infectious diseases were present in 5 patients with onset between 4 and 12 years, Landau-Kleffner syndrome occurred in six cases with onset at 4-6 years, and cerebral tumor was present in four patients 9-13 years of age. There was no difference in recovery for those children aged above or below 11 years. The very young children with Landau-Kleffner syndrome had a bad prognosis. Six of the eight children with head injury had a favorable outcome, in contrast to those with vascular or infectious disease. Only one of six children with Landau-Kleffner syndrome recovered completely despite normal CT scans. The severity and bilaterality of the lesions showed no significant relation to prognosis of the aphasia. The severity of the cerebral lesion was assessed using a rating scale for CT scans. Most of the children had not recovered completely one year after onset of the aphasia. Recovery was significantly different according to etiological categories. (Loonen MCB, van Dongen HR. Acquired childhood aphasia. Outcome 1 year after onset. Arch Neurol Dec 1990; 47:1324-1328).

COMMENT. In general the outcome of acquired aphasia in childhood is good after mild head injury and a poor outcome may be expected in aphasia due to infectious disorders or Landau-Kleffner syndrome. Bilateral lesions on CT scan are frequently found in patients with infectious and vascular diseases in which aphasia is associated with a poor outcome.

#### EEG IN LANDAU-KLEFFNER SYNDROME

The EEG was studied in five children with Landau-Kleffner syndrome at the Service de Neurologie I, Hôpital Central, Strasbourg, France, and Department de Neurologie C.H.U. Sart Tilman, Liege, Belgium. Day and nighttime EEG video monitoring was performed before and after each change of therapy. One hundred EEGs of 30 minute to two hour duration were recorded on awake patients. Sleep EEGs were obtained during five spontaneous daytime naps, 15 naps induced by amitriptyline, and 65 complete nights of sleep. Spike-wave duration was measured as a percentage of the total sleep period. The EEG in waking patients showed focal and generalized spike-wave discharges on a normal background rhythm. During sleep, discharges increased and bilateral spike-waves occurred more than 85% of the sleep period. The abnormal EEG and the impairment of higher cognitive function developed and

regressed together. It was difficult to differentiate the Landau-Kleffner syndrome and "Epilepsy with continuous spike-waves during slow wave sleep". Only the neuropsychological features differed. They are probably variations of a single syndrome. (Hirsch E et al. Landau-Kleffner syndrome: A clinical and EEG study of five cases. Epilepsia Nov/Dec 1990; 31:756-767).

COMMENT. In the classical Landau-Kleffner syndrome aphasia is acquired and other higher cortical functions do not usually deteriorate. In the "continuous spike-wave activity during sleep syndrome" speech is disturbed in 50% of cases but intellectual deterioration occurs and psychiatric disorders develop. In three of the five patients studied by the authors, only treatment by corticosteroids had lasting effects in correcting the regression of higher functions and EEG abnormalities. Phenobarbital, carbamazepine, and phenytoin were either ineffective or exacerbated the EEG and neuropsychological abnormalities. (Marescaux C et al. Landau-Kleffner syndrome: A pharmacological study of five cases. Epilepsia Nov/Dec 1990; 31:768-777). These authors recommend that high doses of corticosteroids should be administered early at the first appearance of symptoms and preferably before total aphasia or intellectual regression occur and treatment should be maintained at lower doses for a period of months. An abnormal pattern of cerebral glucose utilization demonstrated in three children with Landau-Kleffner syndrome by PET study is reported by the same group of authors from Liege, Belgium and Strasbourg, France (Maquet P et al. Cerebral glucose utilization during sleep in Landau-Kleffner syndrome: A PET study. Epilepsia Nov/Dec 1990; 31:778-783). The specificity of the metabolic abnormalities and their relationship to the aphasia and EEG remain to be determined.

#### CNS TUBERS AND TUMORS

##### MRI AND EEG IN TUBEROUS SCLEROSIS

The relationship between MRI cortical lesion sites and the topographic distribution of EEG spike-and-wave foci was studied in 34 children with tuberous sclerosis at the Service de Neuropediatrie, Hopital Saint Vincent de Paul, Paris, France and the Istituto di Neuropsichiatria Infantile, Roma, Italy. Seizures had appeared in the first year of life in 30 patients, they were partial in ten and infantile spasms in 11, or both in nine. MRI revealed localized cortical areas of high intensity signal, or tubers, in all but two patients. EEG performed at the same age as MRI showed focal spikes and slow waves in all but three patients. Of 34 patients 26 had both MRI cortical large or intermediate tubers and EEG foci (76%). There was a significant correlation between the number of large tubers and the number of EEG foci. Patients with frontal lobe involvement on MRI showed frontal EEG foci after age two. Secondary bilateral synchrony in the EEG was associated with frontal tubers. (Cusmai R et al. Topographic