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 Instituto 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

comparative study of magnetic resonance imaging and electroencephalography in 34 children with tuberous sclerosis. Epilepsia Nov/Dec 1990; 31:747-755).

COMMENT. The MRI is more efficient than CT in detecting and localizing tubers of tuberous sclerosis which appear as high intensity signal areas in T2-weighted sequence. The cortical tubers shown on MRI are epileptogenic whereas subependymal calcifications as detected by CT are non-epileptogenic. Frontal lesions present from the first year of life often have a delayed EEG expression, in keeping with the posteroanterior migration of epileptic foci in childhood, the result of maturational changes.

VON HIPPEL-LINDAU DISEASE AND GADOLINIUM MRI

Gadolinium-enhanced MRI was used to determine the frequency and distribution of CNS lesions in 50 patients with Von Hippel-Lindau Disease (VHL) at the National Institutes of Health, Bethesda, MD; University of Louisville, KY; and the Massachusetts General Hospital, Boston, MA. CNS hemangioblastomas were found in 36 (72%). The cerebellum was affected in 52%, spinal cord in 44%, and the brainstem in 18%. One-half of these tumors were asymptomatic and without clinical signs. The youngest patient was 11 years old and asymptomatic. The youngest symptomatic patient with VHL was 2 years of age with symptoms of ocular lesions. The mean age of all VHL patients was 34 years with a range of 11-62 years. (Filling-Katz MR et al. Central nervous system involvement in Von Hippel-Lindau disease. Neurology Jan 1991; 41:41-46).

COMMENT. Hemangioblastomas and VHL may be detected by 11 years of age and the screening of patients at-risk for VHL should begin at 11 years using gadolinium-enhanced MRI; ophthalmic examination should be initiated within the first two years of life. Diagnosis of VHL is important for genetic counseling as well as early detection of unsuspected ocular and visceral tumors which may lead to blindness or early death unless treated.

SUBARACHNOID METASTATIC TUMORS AND MRI

Subarachnoid metastatic disease was evaluated in 17 children with primary intracranial neoplasms at the Departments of Neurology and Radiology, The Children's Hospital of Philadelphia, PA. The tumors were principally neuroectodermal, ependymomas, and astrocytomas and the majority originated in the posterior fossa. The gadolinium MRI was superior in delineating spinal cord nodules and "sugar coating" whereas myelography and CT more readily revealed nerve root sleeve filling defects. (Kramer ED et al. Comparison of myelography with CT follow-up versus gadolinium MRI for subarachnoid metastatic disease in children. Neurology Jan 1991; 41:46-50).

COMMENT. Gadolinium-enhanced MRI appears to be a safe non-invasive test and the technique of choice for the diagnosis of subarachnoid metastatic disease.