early brain biopsy as a prerequisite for diagnosis and treatment of HSE. This view is supported by Hanley et al, Johns Hopkins Hospital, but is considered invalid by Fishman RA, Univ of California, San Francisco (Arch Neurol 1987; 44: 1289-1292). Brett EM, at the Hospital for Sick Children, Great Ormond Street, London, has spoken against routine diagnostic brain biopsy for suspected HSE in children (Br Med J 1986; 293: 1388). Noninvasive magnetic resonance imaging (MRI) may provide early diagnosis without the immediate risks of biopsy and its later complications (Schroth G et al. Neurology 1987; 37: 179).

NEUROCUTANEOUS SYNDROMES

TUBEROUS SCLEROSIS

A 14-year-old girl with bilateral renal cell carcinoma (hypernephroma) complicating a previously unrecognized tuberous sclerosis is reported from Cornell Univ Med College, NY, and North Shore Univ Hosp, Manhasset, NY, together with a review of 6 similar cases culled from the literature. The patient presented with a 6-month history of progressive weight loss and anemia. She had an acne-like rash on her face and a nodule on her tongue. Her father and paternal uncle were institutionalized for convulsive and psychiatric disorders. Abdominal sonogram and CAT scan showed a large mass arising from the right kidney and a smaller mass in the left kidney. Areas of sclerosis and periventricular calcifications were found in skull X-rays and CAT scan of the head. Renal cell carcinoma and hamartomatous nodules were diagnosed at surgery. Epithelial-lined cysts of the kidney and adenoma sebaceum of the face were typical of tuberous sclerosis. All abnormal hematological and chemistry values, including hypercalcemia, anemia, thrombocytopenia, and hypoalbuminemia returned to normal and the patient was asymptomatic at 3 years after surgery. (Weinblatt ME et al. Renal cell carcinoma in patients with tuberous sclerosis. Pediatrics 1987; 80: 898-903).

COMMENT. Tumors occur more commonly in the kidney than in any other organ as a manifestation of tuberous sclerosis, their frequency being estimated at 80% (Critchley M, Earl CJC. Brain 1932; 55: 311). Hamartoma or multiple angiomyolipoma is the most frequent variety but hypernephroma, liposarcoma, adenosarcoma, myosarcoma, and perithelioma are also described (Wilson SAK. Neurology 1955, Williams and Wilkins, Baltimore). Mostly bilateral and often multiple, they sometimes undergo cystic degeneration. Of 29 cases of tuberous sclerosis reported by Fowler JS and Dickson WEC (Quart J Med 1910; 4: 43), 17 (58%) had renal tumors.

The present authors recommend that all patients with tuberous sclerosis should have periodic sonography of the kidney and frequent urinalysis, especially in adolescence and adult life when the incidence of renal masses begins to increase. Fortunately,

most renal tumors associated with tuberous sclerosis are relatively benign and patients with renal cell carcinoma localized to the kidney have a 75% chance of recovery following surgery alone.

DEGENERATIVE DISORDERS

PROGRESSIVE MYOCLONUS EPILEPSY (LAFORA TYPE)

The diagnosis of Lafora's syndrome, progressive myoclonus epilepsy and intracytoplasmic periodic acid-Schiff-positive inclusions (Lafora bodies), was made by skin biopsy in a 16-year-old girl at the Depts of Pathology and Dermatology, University of Texas Medical Branch, Galveston, TX. She presented because of refractory generalized convulsions. In good health until 8 months previously, she developed progressive incoordination, slurred speech, weakness, and impaired school performance followed after 2 months by her first generalized tonic-clonic seizure and progressive mental deterioration. The family history was negative. Metabolic, endocrine, and infectious disorders were excluded. CT scan was normal. EEG showed "generalized cerebral dysfunction." Round to oval, intracytoplasmic inclusions, strongly PAS-positive, in eccrine duct cells and peripheral nerve bundles of dermis were demonstrated histologically in cryostat- and paraffin-embedded sections and by electronmicroscopy. Skin biopsy, the least invasive method of identifying the Lafora body, was first proposed by Carpenter S. Karpati G (Neurology 1981; 31; 1564). A summary of other disorders and their characteristic inclusions that may be diagnosed by skin biopsy includes neuronal ceroid lipofuscinoses (Bielschowski and Spielmeyer-Vogt), glycogenosis II (Pompe's), metachromatic leukodystrophy and occasionally, globoid leukodystrophy (Krabbe). (Newton GA et al. Lafora's disease. The role of skin biopsy. Arch Dermatol 1987; 123: 1667-1669).

COMMENT. A number of syndromes of progressive myoclonus epilepsy, frequently autosomal recessive in inheritance, have been described. The Lafora type is characterized by a rapidly progressive dementia, myoclonus, and intracytoplasmic Lafora body inclusions demonstrated in neurons, especially localized in the substantia nigra and dentate nucleus, in the heart, liver, muscle, retina, nerves and now in skin. The EEG shows discharges of fast spike-waves and polyspike-waves, photosensitivity, deterioration of background activity, and multifocal abnormalities especially posteriorly. The onset occurs between 6 and 19 years of age and the patient dies within an average of 5.5 years after onset. No enzymatic defect has yet been identified.

The Unverricht-Lundborg types have a slower rate of progression than the Lafora myoclonic epilepsy, with onset at about age 10 years, variable severity of myoclonus, associated cerebellar ataxia, and milder mental symptoms, patients surviving for 15 years and more. This variety known as the Finnish or "Baltic"