936). (Respond: Dr K Otani, Department of Pediatric Neurology, Osaka Medical Center, 840 Murodo-cho, Izumi, Osaka 590-02, Japan).

COMMENT. The authors cite 4 reports including eight previous patients with secondary sensory seizures published in the last 40 years, the first involving 2 patients of Penfield and Jasper (1954). One study was entitled 'Sensory seizures mimicking a psychogenic seizure.' (Lessor RP et al. Neurology 1983;33:800). It is certainly conceivable that the diagnosis is sometimes overlooked and the symptoms misinterpreted as psychogenic.

EEG MONITORING OF NEONATAL SEIZURES

Sixty-three neonates were investigated using prolonged video/EEG monitoring to identify seizures and determine the diagnostic efficiency of clinical observation and short duration EEGs at the Department of Paediatric Neurology, Prince of Wales Children's Hospital, Sydney, NSW, Australia. Thirty-two patients had seizures confirmed. Clinical observations after anticonvulsant treatment identified seizures in 66%, and a 60 min EEG revealed electrographic seizures in 76%, after phenobarbital treatment, and in 50% after addition of phenytoin. Short duration EEG avoids misdiagnoses in most patients with ambiguous clinical signs and aids substantially in the identification of neonatal seizures. (Bye A, Flanagan D. Electroencephalograms, clinical observations and the monitoring of neonatal seizures. J Paediatr Child Health December 1995;31:503-507). (Respond: Dr A Bye, Prince of Wales Children's Hospital, High St, Randwick, NSW 2031, Australia).

COMMENT. When clinical signs of seizures are controlled by anticonvulsants, a 60 min EEG is required to uncover subclinical neonatal seizures, and in some cases, especially when phenytoin has been given in addition to phenobarbital, prolonged video/EEG monitoring may be necessary in diagnosis. An EEG after infusion of anticonvulsant does not gaurantee seizure identification, but the probability of diagnosis increases in relation to the length of the recording. In a study at the Magee-Womens Hospital, Pittsburgh, PA, more than 50% of 92 neonates with seizures had only electrographic expression of seizures, and 16% exhibited electroclinical dissociation. (see Progress in Pediatric Neurology II, PNB Publishers, 1994, pp 11-16).

DEGENERATIVE AND METABOLIC DISORDERS

CARNITINE DEFICIENCY SYNDROMES

Carnitine deficiency syndromes are reviewed from the Departments of Neurology and Pediatrics, Columbia Presbyterian Medical Center, New York, NY. *Primary carnitine deficiency* is a decrease of intracellular carnitine that impairs fatty acid oxidation and is not associated with another systemic illness. It may be systemic or muscular, presenting as progressive cardiomyopathy, hypoketotic hypoglycemic encephalopathy, or myopathy. Age at onset is 1 month to 7 years, with a mean of 2 years. In encephalopathy, carnitine levels in plasma and tissues are below 10% of normal, and acylcarnitines are proportionately reduced. Acylcarnitine to free carnitine ratio is normal. Treratment is oral carnitine at daily doses of 100 to 200 mg/kg. Intermittent diarrhea and a fishy body odor are described as side effects of carnitine replacement. Muscle carnitine deficiency is characterized by severe reduction

in muscle carnitine levels and normal serum carnitine. *Secondary carnitine deficiency* is manifested by a decrease in levels of plasma or tissue carnitine, and is associated with genetically determined metabolic errors, acquired medical conditions, or iatrogenic factors. Metabolic errors involve fatty acid oxidation, B-oxidation cycle, aminoacidurias, and mitochondrial disorders. Acquired disorders include cirrhosis, malnutrition, vegetarian diet, celiac disease, extreme prematurity, AIDS, and Fanconi syndrome. Anticonvulsant treatment with valproate has been linked to some carnitine deficiencies. Several mechanisms are proposed for valproic acid-induced carnitine deficiency, some involving an underlying primary metabolic inborn error. The authors recommend prophylactic carnitine in all children under 2 years of age who are treated with valproate. Secondary carnitine deficiencies are managed by high carbohydrate, low fat frequent feedings, and vitamin/cofactor supplements (carnitine, glycine, and riboflavin). (Pons R, De Vivo DC. Primary and secondary carnitine deficiency syndromes. J Child Neurol November 1995;10(Suppl):2S8-2S24). (Respond: Dr Darryl C De Vivo, Neurological Institute, 710 West 168 Street, New York, NY 10032).

COMMENT. In the same issue, Coulter DL, from the Boston City Hospital, discusses the risk factors and treatment of carnitine deficiency in epilepsy. (J Child Neurol 1995;10(Suppl):2S32-2S39). Carnitine deficiency in epilepsy results from metabolic diseases, poor nutrition, and anticonvulsants, especially but not exclusively valproate. Carnitine supplements benefit high-risk, symptomatic patients and those with free carnitine deficiency, but not the low-risk, asymptomatic patients and those with normal carnitine levels.

PRIMARY LATERAL SCLEROSIS WITH GAZE PARALYSIS

Three children in a Jordanian family, with consanguinous parents, who met the Stark and Moersch (1945) criteria for the diagnosis of primary lateral sclerosis (PLS) are reported from the King Faisal Specialist Hospital and Research Centre, and King Khalid Eye Specialist Hospital, Riyadh, Saudi Arabia, and Northwestern University Medical School, Chicago, Illinois, USA. In addition they had a diffuse conjugate saccadic gaze paralysis, especially on down-gaze. A chronic progressive weakness beginning in late infancy, associated with spastic quadriplegia and pseudobulbar palsy, led to wheelchair dependence by adolescence and later loss of speech, while intellect was preserved. CT, MRI, EEG, EMG, NCS, and laboratory tests, including enzyme and amino acid assays, were normal. All patients had absent transcranial magnetic motor-evoked potentials in abductor pollicis and anterior tibial brevis muscles. Molecular testing, using DNA blood extracts, showed no linkage to chromosome 2q33 juvenile amyotrophic lateral sclerosis locus, the 8q recessive familial spastic paraplegia locus, or the 5q13 spinal muscular atrophy locus. The clinical course and absence of specific neuropathological etiologies support the diagnosis of familial, autosomal recessive, primary lateral sclerosis. (Gascon GG, Siddique T et al. Familial childhood primary lateral sclerosis with associated gaze paralysis. Neuropediatrics 1995;26:313-319). (Respond: Dr Generoso G Gascon, Division of Pediatric Neurology, Rhode Island Hospital/Brown University, Physicians Office Building, Suite 438, 110 Lockwood Ave, Providence, RI 02903).

COMMENT. All three of these patients were referred with a diagnosis of cerebral palsy, despite the familial and progressive nature of the disorder. The authors cite only one other case of childhood primary lateral sclerosis in the literature. The present report is presented as the first of familial cases. Ford FR