

had elicited diverse cognitive syndromes (i.e. acalculia, agraphia, alexia, anomia, constructional apraxia, finger agnosia, and right-left disorientation). Stimulation of adjacent loci, between the angular and supra-marginal gyri, had produced a complete Gerstmann syndrome without accompanying deficits.

### **THALAMIC LESIONS IN INFANCY**

The clinical, pathologic and etiologic characteristics of thalamic lesions in infancy are reviewed in an editorial. During the past 30 years, two distinct patterns of thalamic hemorrhagic insult in infants have been described, with different etiology, clinical presentation, scan appearance and prognosis. Intrauterine infection with cytomegalovirus, rubella, or toxoplasma and streptococcal and pneumococcal meningitis have also been associated with bilateral thalamic calcification and spastic quadriplegia. Genetic factors have been described occasionally, including a rare autosomal recessive encephalopathy; and chromosomal trisomies 21 and 13 have also been linked with thalamic echogenicity. An asphyxial insult may occur before birth, perinatally or postnatally. Most affected infants were born at term, the thalamic changes were always bilateral, and the MRI was the most sensitive technique in diagnosis.

In one pattern of thalamic hemorrhagic or asphyxial insult, the neurologic abnormalities presented at birth. Many infants died in the neonatal period or early childhood and all survivors were severely handicapped. In another pattern of thalamic lesion, previously healthy term infants with primary thalamic hemorrhage presented acutely at age 11-14 days with seizures, opisthotonos and facial weakness. There was eye deviation to the side of the hemorrhage and a sunseting phenomenon. In these infants the neurologic outcome was good: 1 child was normal at 20 months and 3 had only mild spastic hemiparesis. In almost 2/3 of term infants with intraventricular hemorrhage the primary lesion is in the thalamus. (Editorial. Thalamic lesions in infancy. The Lancet May 9, 1992; 339:1143-1145.)

**COMMENT.** Thalamic lesions may occur independently of cortical lesions but are always found when cortical or subcortical damage is present. *Etat marbre* (status marmoratus), characteristically associated with athetoid forms of cerebral palsy, is seen as a hypermyelination in association with striatal marbling or as an isolated phenomenon. Thalamic involvement underlies the severe mental deficiency found in some cases of congenital athetosis. (Norman RM. In Greenfield's Neuropathology, Baltimore, Williams and Wilkins, 1963, 391-393.)

### **SEIZURE DISORDERS**

#### **VALPROATE-INDUCED PUBERTAL ARREST**

A 12-year-old girl with complex partial seizures who had pubertal arrest of both growth and secondary sexual development while receiving VPA is reported from the Department of Pediatrics, Wright State University, Dayton,

OH and the University of Iowa, Iowa City, IA. She had received Depakote from 10 years 7 months of age in doses ranging from 750 mg/day (17 mg/kg/day) to 1,500 mg/day (36 mg/kg/day) and a maximum VPA level of 135.5 ug/dl. Her growth rate during treatment decreased to 1.9 cm/yr compared to 4.9 cm/yr prior to treatment. When treatment was discontinued, normal pubertal growth and maturation resumed with a growth rate of 10 cm/yr. Cranial MRI with gadolinium and CT were normal (Cook JS et al. Pubertal arrest associated with valproic acid therapy. Pediatr Neurol May/June 1992; 8:229-231). (Correspondence: Dr. Cook, Department of Pediatrics, Wright State University, Dayton, OH 45402.)

**COMMENT.** The authors recommend that children and adolescents on VPA therapy should have height, weight and Tanner staging of genitalia and pubic hair assessed at each clinic visit to detect arrest of growth and development.

A causal association between maternal use of VPA and fetal spina bifida has been reported (Omtzigt JGC et al. The risk of spina bifida aperta after first trimester exposure to valproate in a prenatal cohort. Neurology 1992; 42(suppl 5):119-125.) Of 261 pregnant women receiving antiepileptic drugs, 6.3% of fetuses with VPA exposure had spina bifida aperta, whereas other antiepileptic drugs were not implicated in this developmental abnormality. High serum levels of VPA in the mother may have increased the risk which might be diminished by reduction of the daily dose.

### **OCCIPITAL LOBE SEIZURES AND CELIAC DISEASE**

The electroclinical ictal findings of 4 epileptic patients with clinically asymptomatic celiac disease are reported from the University of Bologna, Italy. All patients had paroxysmal visual manifestations and EEG seizure discharges arising from the occipital lobe. CT in 2 patients showed occipital calcifications. A gluten-free diet was not beneficial and polytherapy with antiepileptic drugs aggravated the seizures in 3 cases. Epilepsy and evidence of steatorrhea and growth failure were present in early childhood. The diagnosis of celiac disease was made by tests for serum folate level, a d-xylose absorption test, anti-gliadin antibody test and jejunal biopsy which showed villous atrophy (Ambrosetto G et al. Occipital lobe seizures related to clinically asymptomatic celiac disease in adulthood. Epilepsia May/June 1992; 33:476-481). (Reprints: Professor G. Ambrosetto, Institute of Neurology, Via Ugo Foscolo 7, 40123, Bologna, Italy.)

**COMMENT.** The same authors had investigated 10 patients with epilepsy and occipital cerebral calcification and had found evidence of celiac disease in 6. All patients with epilepsy and posterior cerebral calcification should be investigated for possible celiac disease. (Gobbi G, Ambrosetto P et al. Celiac disease, posterior cerebral calcifications and epilepsy. Brain Dev Jan 1992; 14:23-29.)