headaches in 3. The frequency and duration of cluster periods increased with age in 14. (Maytal J et al. Childhood onset cluster headaches. Headache June 1992; 32:275-279.) (Reprints: Dr. Joseph Maytal, Division of Pediatric Neurology, Schneider Children's Hospital, Long Island Jewish Medical Center, New Hyde Park, New York 11042.)

COMMENT. Familial cluster headache occurring in 3 generations is reported from the Brigham and Women's Hospital, Boston (Spierings ELH, Vincent AJPE. Neurology July 1992; 42:1399-1400). The child, an 8-year-old boy, had suffered from headaches since age 4; the father age 42, and the paternal grandfather age 73, had cluster headache also. Three pairs of identical twins with cluster headache have been reported, suggesting a genetic factor in predisposition. (Roberge C et al. Headache 1987; 27:299; Couturier EGM et al. Neurology 1991; 41:761.)

BRAIN DAMAGE SYNDROMES

DELAYED COGNITIVE ABNORMALITIES AND FRONTAL LOBE DAMAGE

The consequences of early frontal lobe damage, restricted to the polar and mesial portion of the left prefrontal cortex and deep white matter, on higher cognitive and psychologic development are reported in a 33-year-old woman who sustained injury at 7 years of age and was evaluated at the Division of Behavioral Neurology and Cognitive Neuroscience, University of Iowa College of Medicine, Iowa City. The birth and milestones of development were normal, and academic achievement was average before she sustained a spontaneous intraparenchymal cerebral hemorrhage at 7 years. Her neurological exam was normal and generalized tonic clonic seizures were controlled with medication. MRI revealed the lesion in the left prefrontal cortex and deep white matter, and cerebral blood flow studies were abnormal in left and right frontal regions. Defects in higher cognition were noted especially in self regulation of emotion and affect and in social behavior. These were of delayed onset and were followed by a period of progression and finally an arrest of development in adolescence. The progressive impairment resulted from a discrepancy between the demands of adolescent development and the altered maturation of frontal lobe neural and cognitive systems. The patient failed to acquire the executive and self regulatory processes associated with frontal lobe function. (Eslinger PJ et al. Developmental consequences of childhood frontal lobe damage. Arch Neurol July 1992; 49:764-769.) (Reprints: Dr. Eslinger, Division of Neurology, The Milton S. Hershey Medical Center, 500 University Drive, Hershey, PA 17033.)

COMMENT. A variety of neuropsychological syndromes including Gerstmann's Syndrome are described as a consequence of lesions or stimulation to the left posterior perisylvian territory (Benton AL. Gerstmann's syndrome. Arch Neurol May 1992; 49:445-447). A cortical stimulation study in a 17-year-old epileptic boy had shown that repeated stimulation of different loci in the posterior perisylvian region

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 sunsetting 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,