

examined at the Washington University School of Medicine, St. Louis, MO. Mental retardation was a risk factor for the development of seizures and was diagnosed 4 times more frequently in children with seizures (76% compared to 17%). Cortical CNS malformation diagnosed on CT, such as agenesis of the corpus callosum, was another seizure predictive factor (48% v. 17%). The absence of mental retardation, older age and non-paroxysmal EEG at seizure onset, and absence of CNS malformation correlated with seizure remission. Seizures were adequately controlled by anticonvulsants in 42% and medication was successfully discontinued in patients of normal intelligence who had been seizure free for 3 years. (Noetzel MJ, Blake JN. Seizures in children with congenital hydrocephalus: long-term outcome. Neurology July 1992; 42:1277-1281.) (Reprints: Dr. Noetzel, Department of Neurology, Washington University School of Medicine, 400 S. Kingshighway, St. Louis, MO 63110.)

**COMMENT.** The clinical and neuroradiologic findings in a male infant with congenital hydrocephalus due to intrauterine HTLV-1 infection are reported from the Division of Child Neurology, Institute of Neurological Sciences, Tottori University Faculty of Medicine, Yonago, Japan. A 20 day old male infant was admitted with macrocephaly. The mother developed human T-cell lymphotropic virus type I (HTLV-1) - associated myelopathy shortly after the birth of the infant. The infant's serum HTLV-1 antibody was elevated, suggesting an intrauterine route of infection. (Tohyama J et al. Neurology July 1992; 42:1406-1408.)

Factors affecting the prognosis of intrauterine hydrocephalus diagnosed in the third trimester are reviewed from the Department of Neurosurgery, Kobe University, School of Medicine, Japan (Oi S, Surg Neurol 1992; 37:66-68). Four patients underwent transabdominal or transvaginal cephalocentesis with measurement of intracranial pressure and intrauterine pressure. Another 4 patients had pre- and postnatal CT or MRI measurements of the head performed shortly before and after birth. The results indicated extremely high intracranial pressure in the fetal brain, whereas after birth the macrocephaly was accompanied by a relatively low intracranial pressure. Fetal hydrocephalus is extremely hypertensive and an impairment of neuronal functional development prenatally can be irreversible. The fetal ventricular amniotic shunt was not appropriate for maintaining the decompressive effect and a more reliable drainage system is required.

## **HEADACHE DISORDERS**

### **CHILDHOOD CLUSTER HEADACHES**

Cluster headaches similar to the typical adult form occurred in 35 patients at or before 18 years of age in a study from the Montefiore Medical Center and Schneider Children's Hospital, New York. The mean age at onset was 14 years (range 5-18). The delay from onset to diagnosis ranged from 0 to 34 years (mean 8 years). Pain was ocular in 88% and lacrimation occurred in 85%. Ptosis was present in 48% and nasal congestion in 60%. A family history of migraine occurred in 9 patients and a family history of cluster

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