apnea, aspiration pneumonia, episodic muscular rigidity and near miss SIDS. Nose tapping to elicit the hyperekplexic startle response should be included in the routine examination of newborns. See Ped Neurol Briefs May 1991; 5:36. The primary pysiological abnormality in hyperekplexia involves spinal as well as brainstem hyperexcitability (Matsumoto J et al. Ann Neurol July 1992; 32:41-50).

## LISSENCEPHALY: CAUSAL HETEROGENEITY

Clinical, cytogenetic and molecular studies in 65 patients with isolated lissencephaly sequence (ILS) are reported from Indiana University School of Medicine, Indianapolis; Tufts New England Medical Center, Boston; Eastern Virginia Medical School, Norfolk; University of Washington School of Medicine, Seattle; and Baylor College of Medicine, Houston. All patients had type I lissencephaly of varying severity and a grossly normal cerebellum; 17% had agenesis of the corpus callosum and 21% had cavum septi pellucidi. The facial appearance was described as essentially normal, but subtle abnormalities were observed, including microcephaly (71%), bitemporal hollowing (70%), abnormal nasal bridge (49%), and small jaw (58%). All were severely mentally retarded, and 76% had a mixed seizure disorder that progressed to infantile spasms in 35%. Seizures were uncommon during the first few months of life, but opisthotonos was often reported. Hypotonia evolved into spasticity, and feeding difficulties usually improved after several days or weeks. During pregnancy, prolonged or heavy vaginal bleeding occurred in 12% and flu-like syndromes in the mothers of 12%. CP and MRI appearances showed a smooth cerebral surface with open sylvian region, a typical "figure-8" appearance on axial images and enlarged posterior lateral ventricles (colpocephaly). Molecular studies showed microdeletions in chromosome band 17p (6 patients). Other causes included autosomal recessive inheritance, intrauterine infection and intrauterine perfusion failure. The calculated risk of recurrence in sibs was 7% (Dobyns WB et al. Causal heterogeneity in isolated lissencephaly. Neurology July 1992; 42:1375-1388). (Reprints: Dr. William B. Dobyns, Division of Pediatric Neurology, University of Minnesota, P.O. Box 380, 420 Delaware Street SE, Minneapolis, MN 55455.)

**COMMENT.** For further reports on lissencephaly see Ped Neur Briefs August 1991; 5:59-60; June 1992; 6:48. The role of human fetal ependyma in the pathogenesis of some cerebral malformations such as lissencephaly/pachygyria and holoprosencephaly is reviewed by Dr. Harvey B. Sarnat in Pediatr Neurol May/June 1992; 8:163-78. Lissencephaly is a primary disturbance of neuroblast migration associated with abnormal gyration of the cerebral cortex. Ependymal abnormalities include persistence of the fetal pseudostratified columnar organization and subventricular rosettes of ependymal cells. The author provides an excellent account of the pathogenesis of cerebral dysgenesis. (Correspondence after August 15: Dr. Sarnat, Children's Hospital, CH-49, 4800 Sand Point Way N.E., Seattle, WA 98105.)

## CONGENITAL HYDROCEPHALUS AND SEIZURES

The frequency of seizures and long-term outcome in 68 children with congenital hydrocephalus not associated with myelomeningocele were

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