DEVELOPMENTAL DISORDERS

POSTMIGRATIONAL EVOLUTION OF POLYMICROGYRIA

A first in vivo MRI documentation of the postmigrational postnatal evolution of a case of polymicrogyria is reported from the Children's Hospital, Boston, MA. The infant was born premature at 27 weeks' gestation. He suffered respiratory distress and cerebral ischemia, and was maintained on a ventilator for 17 days. Ultrasound scans showed bilateral intraventricular hemorrhages at 1 day, and a cystic periventricular leukomalacia that evolved over 3 weeks. MRIs with ultrafine slices were performed at 4 postnatal weeks, term, and at 6 months of age. A minor perisylvian abnormality noted at 4 weeks had developed into extensive bilateral cortical polymicrogyria by 6 months of age, in addition to mineralization of the cystic leukomalacia changes noted earlier. The postnatal evolution of polymicrogyria, associated with the simultaneous occurrence of periventriucular leukomalacia, points to an ischemic encephaloclastic mechanism. (Inder TE, Huppi PS, Zientara GP et al. The postmigrational development of polymicrogyria documented by magnetic resonance imaging from 31 weeks' postconceptional age. Ann Neurol June 1999;45:798-801). (Respond: Dr Joseph J Volpe, Neurology Department, Fegan 1103, Children's Hospital, 300 Longwood Ave, Boston, MA 02115).

COMMENT. Congenital bilateral perisylvian polymicrogyria presents with facial diplegia, dysarthria, pseudobulbar palsy, seizures, and retardation. The present case report associated with premature birth, cerebral ischemia, and periventricular leukomalacia is described as unique, although the histopathology is not known.

RM Norman, in Greenfield's Neuropathology (London, Edward Arnold, 1958), refers to *sclerotic microgyria* (ulegyria) and polygyria among the neuropathological sequelae of birth injury, cerebral ischemia and anoxia. A review of these earlier neuropathological reports is recommended reading.

Genetics of bilateral perisylvian polymicrogyria (BPPMG) is discussed in a report of 6 affected members of 3 consecutive generations of a family. (Borgatti R, Triulzi F, Zucca C et al. Neurology June 1999;52:1910-1913). The severity of the manifestations in one boy affected and the transmission of the disorder through women suggest an X-linked dominant trait. BPPMG is a heterogeneous disorder, in some cases genetically determined, and in others, related to ischemic injury.

MOSAICISM IN TUBEROUS SCLEROSIS COMPLEX

Six families with mosaicism in a series of 62 unrelated families with a mutation in one of the two tuberous sclerosis complex (TSC) genes, TSC1 or TSC2, are reported from the Erasmus University and Hospital, Rotterdam, The Netherlands. In one family the parents showed no clinical signs, but gonadal mosaicism was determined after the diagnosis of TSC was made in 3 children. The exclusion of signs of TSC in the parents reduces the likelihood of a mosaic mutation parental carrier from 10% to 2%. In 5 families with somatic mosaicism, TSC was diagnosed in the parent after the child's case was identified. The genetic counseling implications of mosaicism are discussed. (Verhoef S, Bakker L, Tempelaars AMP et al. High rate of mosaicism in tuberous sclerosis complex. Am J Hum Genet June 1999;64:1632-1637). (Reprints: Dr S Verhoef, MGC Department of Clinical Genetics, Erasmus Academic Hospital, PO Box 1738, 3000 DR Rotterdam, The Netherlands).

COMMENT. The authors advise complete clinical screening (including brain CT scan) of parents of sporadic TSC patients. A mild phenotype associated with somatic mosaicism might otherwise be missed. In the absence of signs of TSC, only 2% of parents will show gonadal mosaicism.

SEIZURE DISORDERS

NEONATAL EPILEPSIES AND SEIZURE EVOLUTION

The evolution of epileptic syndromes in 75 children with EEG-confirmed epilepsies of neonatal onset was studied at Nagoya University, Japan. Seizures were partial in 63 (84%) patients followed for a minimum of 3 years, including 23 with benign neonatal convulsions; generalized in 9; and both generalized and partial in 3. Partial seizures occurred with idiopathic and symptomatic epilepsies, whereas generalized seizures were present mainly in early infantile epileptic encephalopathy. Seizures were cryptogenic in 7 infants, despite intractable partial seizures, and none developed into other epileptic syndromes. Of 44 patients with symptomatic epilepsies, 18 (41%) developed West syndrome (WS). Fifteen of these WS patients had presented with localization-related epilepsy as neonates, and of these, 7 developed localization-related epilepsy after WS was diagnosed. (Watanabe K, Miura K, Natsume J, Hayakawa F, Furune S, Okumura A. Epilepsies of neonatal onset: seizure type and evolution. Dev Med Child Neurol May 1999;41:318-322). (Respond: Dr Kazuyoshi Watanabe, Department of Pediatrics, Nagoya University School of Medicine, 65 Tsurumai-cho, Showa-ku, Nagoya 466-8550, Japan).

COMMENT. Benign neonatal convulsions, classified as generalized epilepsies in the International Classification, should be reclassified as partial, localization-related epilepsy. The term "age-dependent epileptic encephalopathy" is usually applied to cases of early infantile epileptic encephalopathy which evolve into West syndrome and later Lennox-Gastaut syndrome. Symptomatic localization-related epilepsy with transient West syndrome in infancy is recognized as an additional age-dependent epileptic syndrome.

The burst-suppression electroencephalogram is discussed by Niedermeyer E et al (Clinical EEG July 1999;30:99-105). Generalized burst-suppression (BS) in the EEG may occur with 'Early Infantile Epileptic Encephalopathy' (Ohtahara syndrome) and 'Early Myoclonic Encephalopathy' (Aicardi and Goutieres). It is also observed in deep stages of anesthesia and sedative overdose, with cardiorespiratory arrest, anoxia, and undercutting of the cortex. "The term BS should not be applied to the brief flat stretches that may occur during NREM sleep in infants with hypsarrhythmia."

PARENTS' FEAR OF FEBRILE SEIZURES

Parents' perceptions and knowledge about fever and febrile seizures were determined by a questionnaire study at the Sophia Children's Hospital, Rotterdam, The Netherlands. Of 230 parents of children who participated in a randomized controlled trial of ibuprofen to prevent recurrence of febrile seizures, 181 (79%) responded to the questionnaire. Each child had been treated in the emergency room because of a febrile seizure, and the risk factors for seizure recurrence included a positive family history of febrile seizures, a multiple type seizure, a temperature below 40.0°C at the initial seizure, and previous febrile seizure recurrence. The parents were informed of the generally benign nature of the