1999;45:154-161), Clark GD and Naebels JL of Baylor College of Medicine, Houston, point out the importance of genotype-phenotype correlations in understanding the overlapping of clinical/pathological manifestations of migration disorders and the value in genetic counseling. Asymptomatic mothers of children with *DCX* mutations are at risk of further transmitting the disorder, whereas no germline transmission of *LIS1* mutations have been described.

NODULAR NEURONAL HETEROTOPIA

Brain tissue from 4 children with intractable epilepsy and subcortical or periventricular nodular heterotopia of different etiologies (megalencephaly, cortical dysplasia, polymicrogyria) was examined at the University of Oxford, Radcliffe Infirmary, UK. Histological and carbocyanine dye (DiI) tracing techniques showed fibers surrounding nodules and connectivity between adjacent nodules. Immunohistochemical tests for calretinin and neuropeptide Y (NPY), normally expressed in GABAergic cortical interneurons, found numerous calretinin-positive neurons (CPN) within nodules with incomplete differentiation, abnormal clusters of CPN in the overlying cortical plate, and many cell processes positive for NPY. Heterotopic nodules were associated with malformation in the overlying cortex and had limited connectivity with other brain regions. Abnormal connectivity could affect the balance of excitation and inhibition in neuronal circuits, leading to epileptogenic activity. (Hannan AJ, Servotte S, Katsnelson A et al. Characterization of nodular neuronal heterotopia in children. Brain Feb 1999;122:219-238). (Dr Zoltan Molnar, Institut de Biologie Cellulaire et de Morphologie, Rue du Bugnon 9, 1005 Lausanne, Switzerland).

COMMENT. The abnormal structure, composition, and connections between nodules and the overlying cortex may explain the epileptogenicity of neuronal heterotopias and its propagation to other brain regions.

POSTERIOR FOSSA MALFORMATIONS AND EPILEPSY

Risk factors for epilepsy in children with posterior fossa malformations (PFM) were studied in 22 cases of PFM with epilepsy (41%), and 32 without epilepsy (59%), seen at the University of Bologna, Italy. The most common PFMs were cerebellar hypoplasia (43%), Dandy-Walker (D-W) complex (37%), and Arnold-Chiari (A-C) malformation (13%). Epilepsy recurred most frequently in A-C malformation (71%), compared to 35% in D-W complex, and 35% in cerebellar hypoplasia. Risk factors for epilepsy in PFM cases were cerebellar lesions plus familial antecendents for epilepsy and/or febrile convulsions; 50% in PFM cases with epilepsy, and 9% in PFM without epilepsy. Epilepsy was mainly partial in 77%; benign epilepsies and febrile convulsions occurred in 27%. (Parmeggiani A, Posar A, Scaduto MC et al. Posterior fossa malformations and epilepsy. J Child Neurol Feb 1999;14:113-117). (Respond: Dr Antonia Parmeggiani, Department of Child Neurology and Psychiatry, Neurological Institute, University of Bologna, via Ugo Foscolo 7, 40123 Bologna, Italy).

COMMENT. Epilepsy may occur in 40% of children with posterior fossa malformation. A family history of epilepsy and/or febrile convulsions are significant risk factors for epilepsy in children with posterior fossa malformation.

Posterior fossa pathology other than malformation can predispose to epilepsy. Among 291 children treated for intracranial tumor at the Mayo Clinic over a 10 year period, seizures occurred in 17% - in 25% of patients with supratentorial tumors and in 12% of those with infratentorial tumors. The

diagnosis of supratentorial tumors was delayed for an average of 2 years, whereas infratentorial tumors were diagnosed within 3 months of the initial seizure (Backus RE, Millichap JG. Pediatrics 1962;29:978-984).

SEIZURE DISORDERS

PROGNOSIS OF INFANTILE SPASMS AND L-G SYNDROME

The occurrence, outcome, and prognostic factors of infantile spasms (IS) and Lennox-Gastaut syndrome (LGS) were determined in children treated in the Department of Pediatrics, University of Oulu, Finland, from Jan 1976 to Dec 1993. Thirty seven had IS (0.41/1000 live births) and 25 had LGS (0.28/1000 live births). Ten (27%) of the patients with IS who later developed LGS (40% of LGS cases) had symptomatic epilepsy, were mentally retarded, and their seizures were uncontrolled at 10 year follow-up. Symptomatic epilepsy (30 (81%) IS and 17 (68%) LGS) had congenital or genetic etiologies in almost all cases (87% of IS, 100% of LGS). Cryptogenic epilepsy in 7 (19%) of the IS cases had a favorable prognosis, whereas in 8 (32%) of LGS cases, a cryptogenic etiology did not decrease the risk for a poor outcome. The majority received ACTH and polytherapy. (Rantala H, Putkonen T. Occurrence, outcome, and prognostic factors of infantile spasms and Lennox-Gastaut syndrome. Epilepsia March 1999;40:286-289). (Reprints: Dr H Rantala, Department of Pediatrics, University of Oulu, FIN90220 Oulu 22, Finland).

COMMENT. The prevalence of infantile spasms (IS) in a primary university pediatric population in Finland is 0.4/1000 live births and that of Lennox-Gastaut syndrome (LGS) is similar. IS evolves into LGS in 27% of cases and these are symptomatic epilepsies, with a poor prognosis. Cryptogenic etiology has a favorable prognosis for IS but not in LGS cases.

Vigabatrin in the treatment of infantile spasms has been studied retrospectively in 25 infants (19 symptomatic, 6 cryptogenic cases) followed at the Children's Hospital of Michigan, Detroit, MI. (Koo B. Pediatr Neurol Feb 1999;20:106-110). Clinical improvement was obtained in 16 (64%), and EEG improvement in 17 (68%). EEG and cognitive decline and/or more frequent spasms occurred in 7 (28%), often associated with larger VGB doses (>100 mg/kg daily). Smaller doses and EEG monitoring are recommended, since EEG and cognitive deterioration may occur despite clinical control of spasms.

Efficacy of Lamotrigine in refractory neonatal seizures of unknown etiology is reported in a single newborn treated at the Royal Alexandra Hospital for Children, Parramutta, NSW, Australia. (Barr PA, Buettiker VE, Antony JH. Pediatr Neurol Feb 1999;20:161-163). The EEG showed a burst-suppression pattern, and seizures were mainly generalized. Conventional AEDs were ineffective, vigabatrin (105 mg/kg/d) was partially effective, and the addition of lamotrigine (4.4 mg/kg/d) was followed by a sustained seizure control.

GELASTIC EPILEPSY AND HYPOTHALAMIC HAMARTOMA

The causes, clinical manifestations, and evolution of gelastic seizures (GS) were studied, using video-EEG and MRI, in 9 patients observed between 1986 and 1997 at the Epilepsy Center, Federico II University, Naples, Italy. Seizures were frequent (several/day) and characterized by laughing attacks, sometimes with facial flushing, and rarely with loss of contact. Age at onset was less than a year in 3, and < 12 years in 8. All older patients reported feelings of embarrassment,