Neurology Oct 1997;49:1042-1047). (Reprints: Dr William B Dobyns, Department of Neurology, Box 486 UMHC, 420 Delaware Street SE, Minneapolis, MN 55455).

COMMENT. In contrast to this new multiple congenital anomaly-mental retardation syndrome described by Dobyns et al, the classical BPNH syndrome of bilateral periventricular nodular heterotopia with epilepsy affects mainly girls with normal intelligence. One gene associated with classical BPNH is mapped to chromosome Xq28, similar to the finding in one case of Dobyns syndrome. Other syndromes associated with BPNH include: 1) BPNH with Ehler-Danlos Syndrome, in females with normal intelligence; and 2) BPNH with frontonasal dysplasia, with nephrosis, the short-gut syndrome, and Vles agenesis of the corpus callosum syndrome, all affecting males with mental retardation, as in Dobyns syndrome. Most have X-linked inheritance patterns, and 3 are localized to Xq28. (see Sarnat HB, in Progress in Pediatric Neurology III, 1997;pp365-369, for reference to X-linked periventricular heterotopia and markers in the distal Xq28 locus).

OCULOCEREBROCUTANEOUS SYNDROME: SIGNS & ETIOLOGIES

Three additional cases, and a review of clinical features of a total of 26 cases of oculocerebrocutaneous (OCC) syndrome, are reported from Maastricht University, and University of Groningen, The Netherlands. Common features of OCC syndrome are 1) *Ocular:* orbital cysts, eyelid coloboma, microphthalmia/anophthalmia; 2) *Cutaneous:* skin appendages, dermal hypoplasia and punch-like defects; and 3) *Cerebral:* psychomotor retardation, seizures, dilated ventricles, cysts, callosal agenesis, and cerebellar hypoplasia. The probable pathogenic mechanism is a disruption of the anterior neuroectodermal plate resulting in neurocristopathy with craniofacial dysmorphism. Possible etiological hypotheses include a lethal mutation with survival by mosaicism, an autosomal dominant inheritance with high mutation rate, and exposure to a potential teratogen, as yet unrecognized. (Moog U, de Die-Smulders C, Systermans JMJ, Cobben JM. Oculocerebrocutaneous syndrome: report of three additional cases and aetiological considerations. Clin Genet October 1997;52:219-225). (Respond: Dr Ute Moog MD, Clinical Genetics Center Maastricht, PO Box 1475, 6201 BL Maastricht, The Netherlands).

COMMENT. The oculocerebrocutaneous syndrome, with its triad of clinical features, is frequently manifested by mental retardation and seizures. No familial cases have been reported. Encephalocraniocutaneous lipomatosis (ECCL) is also characterized by a triad of cutaneous, ocular and cerebral anomalies associated with retardation and seizures, and may be considered in the differential diagnosis. One patient described with OCC syndrome also showed lipomatosis of the spinal dura and a lipoma in the region of the zygoma. A mutation factor responsible for cell growth and cell migration has been proposed in the pathogenesis of ECCL, similar to that for hamartoses.

MIGRAINE

DIVALPROEX CF. PROPANOLOL FOR MIGRAINE PROPHYLAXIS

Migraine frequency was reduced in two-thirds of adult patients treated at the Allegheny Hdche Ctr, Pittsburgh, with either divalproex or propanolol cf to 19% with placebo. (Kaniecki RG. Arch Neurol Sept 1997;54:1141-1145).