COMMENT. MRIs repeated at age 10 months may disclose delayed myelination in infants with WS. The delayed myelination is not always explained by ACTH therapy and may reflect the organic brain lesion causing the seizures.

FRAGILE X MUTATIONS AND EPILEPSY

A posiible link between predisposition for epilepsy and mutations in the fragile X mental retardation-1 gene (FMR) was investigated in the Neuropediatric Department, Behandlungszentrum Vogtareuth; and Laboratory of Genetic Diagnostics, Munchen, Germany. EEGs performed on 14 patients with an amplification in the FMR-1 gene showed focal sharp waves and partial seizures in sleep in 8 boys, aged 4-8 years. Of 16 children with rolandic epilepsy (BECT) studied for FMR-1 gene mutations, 1 boy was positive. (Kluger G, Bohm I, Laub MC, Waldenmaier C. Epilepsy and fragile X gene mutations. Pediatr Neurol Nov 1996;15:358-360). (Respond: Dr Kluger,Neuropediatric Department, Behandlungszentrum Vogtareuth, Krankenhausstrabe 20, D-83569 Vogtareuth, Germany).

COMMENT. A higher incidence of seizures or EEG abnormalities may be expected in boys with fragile X-1 gene mutations.

ANTIEPILEPTIC DRUGS

PHENYTOIN AND CARBAMAZEPINE TERATOGENICITY

In a prospective, controlled, and blinded study of 36 mother-child pairs exposed to carbamazepine (CBZ) monotherapy, 34 pairs exposed to phenytoin (DPH) monotherapy, and 9 nonmedicated epileptic women and their children, the patterns of malformations in the children exposed to potential teratogenic factors were compared with matched mother-child pairs exposed to nonteratogens, at the Hospital for Sick Children, Toronto, Canada. There was no correlation between the daily dose of DPH or CBZ and number of malformations. Microcephaly occurred in 6% of children exposed to DPH and 8.8% of those exposed to CBZ, but not in medicated nonepileptic or nonmedicated epileptic subgroups. Malformations in 8.8% of DPH and 5.7% of CBZ exposed children were not significantly different from controls. Minor anomalies in children exposed to either AED were more frequent than in controls, with a relative risk of 2.1. Hypertelorism was more frequent among DPH-exposed offspring; 25% incidence vs 11% in controls. High forehead, frontal bossing, malar hypoplasia, epicanthus and micrognathia occurred in association with untreated epilepsy, as well as DPH and CBZ treatment. (Nulman I, Scolnik D, Chitayat D, Farkas LD, Koren G. Findings in children exposed in utero to phenytoin and carbamazepine monotherapy: independent effects of epilepsy and medications. Am J Med Genet Jan 1997;68:18-24). (Respond: Gideon Koren MD, The Hospital for Sick Children, 555 University Avenue, Toronto, Ontario, M5G 1X8, Canada).

COMMENT. AEDs and epilepsy have teratogenic effects that are independent and result in minor anomalies in infants exposed in utero. Previous studies have shown that valproate and carbamazepine are associated predominantly with spina bifida and hypospadias, whereas barbiturates and phenytoin may induce congenital heart malformations and facial clefts. None of the AEDs is free of possible adverse effects on the fetus. Experience with