Pediatrics Jan 2002;109:116-123). (Reprints: Dr JK Lynch, Bldg 10, Room 5S220 10 Center Dr, MSC 1447, Bethesda, MD 20892).

COMMENT. The authors conclude that pediatric stroke registries, similar to the one in Canada, are needed to provide data regarding the impact of maternal and perinatal factors on the causes and outcome of childhood stroke in the US.

DEVELOPMENTAL DISORDERS

X-LINKED LISSENCEPHALY WITH ABSENT CORPUS CALLOSUM

Three new cases of the congenital syndrome consisting of X-linked lissencephaly, absent corpus callosum, and genital anomalies (XLAG) are reported from the University Hospital, Angers, France. Male sex, intractable epilepsy, hypotonia, and early mortality are additional characteristics. MRI findings include anterior pachygyria, posterior agyria, thickened cerebral cortex, dysplastic basal ganglia, and agenesis of the corpus callosum. Pathologically, the brain shows a trilayered cortex, neuronal migration defect, dysplastic basal ganglia, and gliotic and spongy white matter. Females related to affected boys may be mentally retarded, suffer from epilepsy, and have agenesis of the corpus callosum. (Bonneau D, Toutain A, Laquerriere A, et al. X-linked lissencephaly with absent corpus callosum and ambiguous genitalia (XLAG): Clinical, magnetic resonance imaging, and neuropathological findings. Ann Neurol March 2002;51:340-349). (Respond: Dr Bonneau, Sevice de Genetique Medicale, CHU d'Angers, 4 rue Larrey, 49100 Angers, France).

COMMENT. First described by Berry-Kravis and Israel in 1994, XLAG is characterized by lissencephaly, complete agenesis of the corpus callosum, and hypogenitalism. A posterior agyria, an anterior pachygyria, and an intermediate thickening of the cortex distinguish this syndrome from lissencephaly type I, in which the cortex is thicker, and the corpus callosum may be hypoplastic but not absent. The MRI and neuropathological features are also distinct. Abnormal MRI findings in carrier females also expand the XLAG phenotype.

MYELOMENINGOCELES AND INCIDENCE OF SHUNTING

The distribution of postnatally repaired myelomengocele (MMC) lesions, characterized by neurologic and radiologic assessment, and the incidence of shunting were determined and correlated by a retrospective chart review of 297 patients followed at the spina bifida clinic at the Children's Hospital of Philadelphia. The rate of ventricular shunting was 81%, and was correlated with the level of the lesion. The more cephalad the MMC, the higher the incidence of shunting. Levels determined by functional neurologic examination were generally higher or equal to the vertebral level of the lesion defined by spine radiographs. Sacral lesions categorized radiologically rather than neurologically had a higher shunt rate. (Rintoul NE, Sutton LN, Hubbard AM, et al. A new look at myelomeningoceles: Functional level, vertebral level, shunting, and the implications for fetal intervention. Pediatrics March 2002;109:409-413). (Reprints: Leslie N Sutton MD, Division of Neurosurgery, Children's Hospital of Philadelphia, 34th St and Civic Center Blvd, Philadelphia, PA 10194).

COMMENT. In this series of patients, the proportion of lumbar MMC was 62% in contrast to 80% reported by Emery and Lendon in a study published in 1973. The authors of the Philadelphia study comment that differences in the

distribution of MMC lesions in this compared to earlier studies may reflect prenatal diagnosis and termination or the effects of maternal folic acid supplementation. Their findings, emphasizing the relation of rate of shunting and functional level of lesion, are considered important in counseling and also in the design of fetal intervention studies. Fetal MMC closure may be associated with a delayed development of symptomatic hydrocephalus and lower rate of shunting than postnatal closure. Reversal of the hindbrain hernia in fetal Chiari II malformation might open the cerebrospinal fluid drainage and prevent the obstructive hydrocephalus (McLone DG, Knepper PA. 1989).

Risk of major birth defects is more than doubled in infants conceived after intracytoplasmic sperm injection or in vitro fertilization, according to data obtained from birth registries in Western Australia, between 1993 and 1997 (Hansen M, Kubinczuk JJ, Bower C, Webb S. N Engl J Med March 7, 2002;346:725-730). The principal defects were musculoskeletal and chromosomal.

SEIZURE DISORDERS

PREVALENCE OF HYPOPIGMENTED AND CAFE-AU-LAIT SPOTS IN IDIOPATHIC EPILEPSY

The prevalences of hypopigmented maculae and cafe-au-lait spots were investigated in 210 children with idiopathic epilepsy, between 2 and 17 years of age, and 2754 health controls children, at the Departments of Pediatrics and Dermatology, Hacettepe University and Inonu University Medical Schools, Turkey. In epileptic children, hypopigmented maculae and cafe-au-lait spots occurred in 14.3% and 30%, respectively, compared to 1.6% and 2.8%, in healthy children (P<.0001). Hypopigmented maculae were polygonal, ash leaf, and fingerprint in shape. Cafe-au-lait spots were discrete, round or oval, and uniformly hyperpigmented. These skin lesions should be considered a concomitant risk factor for epilepsy. (Karabiber H, Sasmaz S, Turanh G, Yakinci C. Prevalence of hypopigmented maculae and cafe-au-lait spots in idiopathic epileptic and healthy children. J Child Neurol Jan 2002;17:57-59). (Respond: Dr Hamza Karabiber, Kahramanmaras Sutcu Imam Universitesi Tip Facultesi Cocuk Hastaliklari ABD, 46050 Kahramanmaras, Turkey).

COMMENT. The diagnostic criteria for type 1 neurofibromatosis include 6 or more cafe-au-lait spots greater than 5 mm in prepubertal and 15 mm in postpubertal children. They may be localized in any region except the palms and soles. Hypopigmented maculae are found in tuberous sclerosis and also in albinism, Waardenburg's syndrome, and vitiligo. The incidence is higher when examined with a Wood lamp. The figure 3 is sometimes regarded as a significant number of cafe-au-lait spots in patients failing to meet criteria for the diagnosis but sufficient to indicate a partial penetrance of NF-1 (Whitehouse D. Arch Dis Child 1966;41:316). In the above study, 3 spots were counted in 10% of 78 healthy children with spots and in 17% of 63 with epilepsy and cafe-au-lait spots. This number may indicate a trend toward increased risk of idiopathic epilepsy.

CLINICAL, EEG, AND MRI DIFFERENCES IN FRONTAL AND TEMPORAL LOBE EPILEPSY

Children who underwent video-EEG monitoring between 1995 and 2000, and were classified as frontal lobe epilepsy (FLE) (n=39) or mesial temporal lobe epilepsy (MTLE) (n=17), were examined for clinical, EEG, and quantitative MRI