A nonsense mutation of the *ATRX* gene causing mild mental retardation and epilepsy. Ann Neurol Jan 2000;47:117-121). (Respond: Dr Gibbons, Nuffield Department of Clinical Biochemistry and Cellular Science, University of Oxford, John Radcliffe Hospital, Headington, Oxford, OX3 9DU, UK).

COMMENT. ATR-X syndrome is clinically heterogeneous. The absence of typical facial dysmorphism does not preclude the diagnosis in a child with mental retardation.

BILATERAL FRONTAL POLYMICROGYRIA WITH MENTAL RETARDATION AND EPILEPSY

Thirteen patients with symmetric polymicrogyria of both frontal lobes are reported from King's College, University of London, UK; University of Pisa, Italy; University of California-San Francisco, CA; UAE University, United Arab Emirates, and University of Chicago, IL. Clinical characteristics included developmental delay, spastic quadriparesis, impaired language development, mental retardation, and epilepsy. Two had consanguineous parents. Head circumference was normal in all but one. Age at examination ranged from 10 months to 32 years. MRI showed irregular infoldings of the cerebral surface with abnormally thick cortex of the frontal lobes bilaterally. EEGs in 5 with epilepsy showed bilateral frontal slowing, sharp waves and spike-and-wave activity. (Guerrini R, Barkovich AJ, Sztriha L, Dobyns WB. Bilateral frontal polymicrogyria; a newly recognized brain malformation syndrome. Neurology February (2 of 2) 2000;54:909-913). (Dr Renzo Guerrini, Academic Neuroscience Centre, King's College Hospital, Denmark Hill, London SE 59RS, UK).

COMMENT. Bilateral frontal polymicrogyria is described as a new syndrome, distinct from polymicrogyria involving perisylvian and parasagittal parieto-occipital regions. The majority are detected by MRI in early childhood during investigation for mental and motor retardation or spastic quadriparesis and epilepsy, and some are genetically determined.

DOUBLE CORTEX SYNDROME WITH MENTAL RETARDATION AND EPILEPSY

Magnetic resonance imaging was used to differentiate 30 female sporadic patients with double cortex (DC) syndrome examined at Beth Israel Deaconess Medical Center, Children's Hospital, Boston. Ages ranged from 4 to 46 years. Age at onset of seizures was 6 months to 12 years. Mental retardation ranged from mild to moderate or severe. MRI and genetic tests differentiated patients into four groups: anterior biased/global DC with doublecortin mutation (53%); anterior biased/global DC without mutation (27%); posterior biased DC without mutation (10%); and limited/unilateral DC without mutation (10%). Other genetic loci or mosaicism at the doublecortin locus may be responsible for the heterogeneity of DC syndrome. (Gleeson JG, Luo RF, Grant PE et al. Genetic and neuroradiological heterogeneity of double cortex syndrome. Ann Neurol February 2000;47:265-269). (Respond: Dr CA Walsh, Division of Neurogenetics, Beth Israel Deaconess Medical Center/Harvard Medical School, 77 Avenue Louis Pasteur, Boston, MA 02115).

COMMENT. Double cortex syndrome can represent a genetically heterogeneous group of mental retardation syndromes, 50 per cent showing an identifiable DCX mutation. MRI scans in patients with mutation have an anterior biased subcortical band and overlying pachygyria, whereas some without

mutation have posterior biased MRI changes.

HEAD GROWTH IN RETT SYNDROME

The longitudinal development of head growth was determined in 82 girls with Rett syndrome (RS) by plotting measurements from the Swedish RS register on normal growth charts, in a study at SU/Ostra Hospital, Goteborg University, Sweden. RS was classic in 69 and forme fruste in 13. In classic RS, mean head circumference fell successively to 2 SD below the norm at age 4 years, and stabilized at -3 SD after age 8 years. In forme fruste cases, mean head circumference was -0.8 SD below but within normal limits. Height measurements at -2 SD at age 6 years was correlated to decrease in head growth. Marked deceleration in head growth was correlated with maximum impairment of gross and fine motor function. (Hagberg G, Stenbom Y, Engerstrom IW. Head growth in Rett syndrome. Acta Paediatr February 2000;89:198-202). (Respond: Dr G Hagberg, Department of Pediatrics, SU/Ostra Hospital, Goteborg University, S-416 85 Goteborg, Sweden).

COMMENT. By age 6 years, children with RS show impairments of gross and fine motor function that are correlated with the rate of head growth deceleration. Forme fruste cases with almost normal head growth have well-preserved gross and fine motor function.

MOVEMENT DISORDERS

CHILDHOOD HEAD TREMOR

Four children, ages 15 months to 11 years, with head tremor were followed longitudinally for 1 to 8 years in a study at Connecticut Children's Medical Center, Hartford, CT. Head tremor (1-2 Hz) characterized by "yes-yes" or "no-no" movements began at 5 to 10 months of age. Tremor was increased by sitting up without head support, and by movement, and was absent while lying down or sleeping. Mild leg dystonia developed in 2 children, and shuddering spells preceded onset of tremor in 3. A family history of tremor was elicited in 2, maternal epilepsy in 1, and infantile shuddering in the father of 1. Neurologic exam was otherwise normal, and MRI, CT, and laboratory tests, including aminoacids and organic acids, ceruloplasmin and copper levels, were also normal. One child responded to timolol and trihexyphenidyl, 1 to primidone, and 2 remitted spontaneously. (DiMario FJ Jr. Childhood head tremor. J Child Neurol January 2000;15:22-25). (Respond: Dr Francis J DiMario Jr, Department of Pediatrics, Connecticut Children's Medical Center, 282 Washington St, Hartford, CT 06106).

COMMENT. The differential diagnosis of head tremor in infants includes spasmus nutans and bobble-head doll syndrome. *Spasmus nutans* affects infants of 6 months to 2 years and consists of a rhythmic nodding or rotatory tremor of the head, a fine, rapid, pendular nystagmus, and tilting of the head. Nystagmus may be unilateral or dissociated, usually horizontal, occasionally vertical, or rotatory; it is increased by visual fixation or forceful control of the head tremor. Closure of the eyes may reduce the tremor. Head nodding bears no constant relation to the nystagmus. The cause is unknown, sometimes ascribed to rickets, living in dark environments, or of viral origin. Onset is in winter months. It is rare, benign and self limited. *Bobble-head doll syndrome* is associated with hydrocephalus, with obstruction around the IIIrd ventricle or aqueduct, and is characterized by 2 to 4 head oscillations per second.