

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.