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CONGENITAL MALFORMATIONS

CEREBELLAR VERMIS HYPOPLASIA IN FRAGILE X SYNDROME

Investigators at the Kennedy Krieger Institute, Johns Hopkins University School of Medicine, have compared posterior vermis size (cross-sectional area) measured by MRI in 32 males with fragile X syndrome (fra X), 28 males with other causes of cognitive disability (CD), 38 males with normal development (ND), and 37 females with fra X and 53 female control subjects. Neurocognitive correlates of posterior vermis size were also examined in females with fra X. Posterior vermis size was decreased in fra X subjects, males more than females, and the decrease was greater after adjusting for intracranial area. Performance on the WISC-R, WAIS-R, block design, Wisconsin Card Sorting Test, and the Rey inventory correlated with posterior vermis size in female fra X patients. The correlation was stronger for the verbal IQ than the performance IQ scores. (Mostofsky SH, Mazzocco MMM, Aakalu G, Warsofsky IS, Denckla MB, Reiss AL. Decreased cerebellar posterior vermis size in fragile X syndrome. Correlation with neurocognitive performance. Neurology Jan 1998;50:121-130). (Reprints: Dr Stewart H Mostofsky, Behavioral Neurogenetics and Neuroimaging Research Center, Kennedy Krieger Institute, 707 N Broadway, Baltimore, MD 21205).

COMMENT. Cerebellar posterior vermal hypoplasia is a developmental anomaly in patients with fragile X syndrome, and males are more severely affected than females. Decreases in size of the posterior vermis in female fra X patients correlate with impaired performance on selected neurocognitive tests. The cerebellar vermis is involved in cognitive processes, especially in tasks dependent on visual-spatial perception, areas deficient in female fra X subjects.

Hypoplasia of the cerebellum and of the splenium of the corpus callosum, and other cerebral anomalies have been reported in children with attention deficit hyperactivity disorder, a frequent complication of fragile X syndrome. (Progress in Pediatric Neurology III, PNB Publishers, 1997;212-213).

CLINICAL CORRELATES OF CORPUS CALLOSAL AGENESIS

The clinical correlates of agenesis of the corpus callosum were examined in

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