ATTENTION DEFICIT DISORDERS

BRAIN MRI CHANGES AND MEASURES OF ATTENTION IN ADHD

The relation between measures of inhibition and sustained attention and MRI structural changes in the caudate and frontal lobe was evaluated in 10 boys with ADHD (aged 8-17) and 11 male controls, examined at the Massachusetts General Hospital, and McLean Hospital, Boston, MA. Children with ADHD had reversed asymmetry of the head of the caudate nucleus, smaller volume of the caudate head, and smaller volume of the white matter of the right frontal lobe. They scored more poorly on measures of inhibition and sustained attention but not on IQ, achievement, or motor speed. Those with reversed caudate asymmetry had lower scores on inhibition tests and more externalizing behavior disorder. Attention deficits were related to smaller volume of right hemisphere white matter. (Semrud-Clikeman M, Steingard RJ, Filipek P, Biederman J, Bekken K, Renshaw PF. Using MRI to examine brain-behavior relationships in males with attention deficit disorder with hyperactivity. J Am Acad Child Adolesc Psychiatry April 2000;39:477-484). (Reprints: Dr Semrud- Clikeman, SZB 504, University of Texas at Austin, Austin, TX 78712).

COMMENT. The symptom of disinhibition in children with ADHD is correlated with reversed caudate asymmetry whereas deficits in attention are related to right hemisphere structural changes. Both disorders need to be addressed in treatment that should include additional time for processing information and untimed tests.

PHONOLOGICAL PROCESSING AND INHIBITION IN ADHD AND RD

The cognitive profile of 4 groups of children (2 of ADHD v No ADHD; and 2 of reading disability (RD) v no RD), aged 7 to 11 years, was examined using two measures of inhibitory control and 3 phonological processing measures, in a study at the Hospital for Sick Children, Toronto, Canada. All phonological measures were impaired in both RD groups relative to the non-RD groups, and measures of inhibition were impaired in the ADHD groups relative to non-ADHD groups. An RD effect on inhibitory control was present in one inhibition measure. The group with co-morbid ADHD and RD showed additive deficits of both single groups. (Purvis KL, Tannock R. Phonological processing, not inhibitory control, differentiates ADHD and reading disability. J Am Acad Child Adolesc Psychiatry April 2000;39:485-494). (Reprints: Dr Tannock, Department of Psychiatry Research, Hospital for Sick Children, 555 University Ave, Toronto, Ontario, Canada M5G 1X8).

COMMENT. Measures of phonological processing may differentiate children with and without reading disability, whereas impairment of inhibitory control may not distinguish between ADHD and RD.

MENTAL RETARDATION SYNDROMES

ATRX GENE MUTATION, MENTAL RETARDATION AND EPILEPSY

A pedigree including 4 mentally retarded family members with a nonsense mutation in the *ATRX* gene is reported from the John Radcliffe Hospital, University of Oxford, UK. Two patients had the typical features of ATR-X syndrome, with facial dysmorphism, genital abnormalities and thalassemia, and 3 had seizures. (Guerrini R, Shanahan JL, Carrozzo R, Bonanni P, Higgs DR, Gibbons RJ.

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