

before DNA analyses are performed and genetic counseling undertaken. (N Engl J Med Dec 12, 1991; 325:1736-37).

NEUROCHEMICAL MARKERS IN RETT'S SYNDROME

The levels of endogenous biogenic amines and neurotransmitter receptors in the brains of 5 patients with Rett's syndrome and 6 normal controls were examined at The Johns Hopkins University, Kennedy Institute for Handicapped Children, The Department of Neurology and Pediatrics, The Johns Hopkins Hospital, Baltimore, MD and The National Institute of Mental Health, Washington, D.C. The patients with Rett's syndrome were all female and they had died at ages 4, 10, 12, 15, 21 years. The cause of death was unknown in 2, asphyxiation in 1, drowning in 1 and pneumonia, 1. The level of choline acetyltransferase activity was lower in many cortical and subcortical regions in the Rett syndrome brains as compared with the mean level in the controls. Endogenous dopamine levels in the superior frontal and superior temporal gyri, occipital cortex, and putamen were reduced. The authors note that these results suggest neurochemical features in Rett's syndrome similar to those found in Parkinson's and Alzheimer's diseases (Wenk GL, Moser H et al. Altered neurochemical markers in Rett's syndrome. Neurology Nov 1991; 41:1753-1756).

COMMENT. The dopamine agonist, bromocriptine, used to treat girls with Rett's syndrome, has made them more relaxed, sociable, affectionate and attentive, the stereotyped hand-washing movements diminished and some patients spoke for the first time (Zappella M, Genazzani A. Wien Med Wochenschr 1986; 122:98). The hypothesis that a disturbance in the function of central dopaminergic systems in Rett's syndrome is related to a defect in maturation may also be extended to include the cholinergic neural system. The search for a biochemical marker for Rett's syndrome continues.

INFECTIOUS DISORDERS

MITOCHONDRIAL ENZYMES IN REYE'S SYNDROME

A nonuniform decrease in several mitochondrial residual enzyme activities in the liver and brain of a 42 year old woman who died with Reye's syndrome is reported from the Departments of Neurology and Pediatrics, Columbia Presbyterian Medical Center, New York, NY. Pyruvate carboxylase activity was negligible whereas subunits II and IV of cytochrome c oxidase were in normal quantities. There was no evidence for a specific insult to mitochondrial DNA or intramitochondrial protein synthesis. The trigger that precipitates the initial disturbance in the chemical micro environment of the mitochondrial matrix remains unknown. Restoration of mitochondrial ATP concentration repairs the disturbance of intramitochondrial enzyme processing and is followed by recovery of organ function (Van Coster RN, De Vivo DC et al. Adult Reye's syndrome: a review with new evidence for a generalized