

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

defect in intramitochondrial enzyme processing. Neurology Nov 1991; 41:1815-1821).

COMMENT. In a review of the literature the authors found 26 additional cases of adult onset Reye's syndrome. Viral studies showed influenza A or B in 11, varicella in 1, dengue in 1, and not reported in 14. 12 recovered.

The incidence of Reye's syndrome has decreased since 1981 and in 1988 there were only 20 cases reported. The relationship between Reye's syndrome and aspirin is debated (Casteels-Van Daele M. Lancet Nov 30, 1991; 338:1408) and the choice of alternative antipyretic management of fever is discussed in an editorial (Lancet Oct 26, 1991; 338:1049). Chronic paracetamol poisoning is reported in Australia and ibuprofen is suggested as an alternative.

CORTICOSTEROIDS IN PNEUMOCOCCAL MENINGITIS

The records of 97 infants and children with pneumococcal meningitis, 41 receiving corticosteroid therapy, were examined at the Department of Pediatrics, University of Texas Southwestern Medical Center, Dallas, Texas. When steroid-therapy was given before or concurrently with antibiotic therapy, none of 30 steroid-treated versus 16 of 52 non-steroid-treated patients developed neurologic or cardiovascular instability after the first parenteral antibiotic dose was given. Bilateral moderate or greater hearing loss occurred in 9% of steroid-treated group compared to 21% of non-steroid group. The incidence of hearing loss was significantly lower in those survivors of overwhelming meningeal infection who had been treated with steroids (Kennedy WA et al. The role of corticosteroid therapy in children with pneumococcal meningitis. AJDC Dec 1991; 145:1374-1378).

COMMENT. The data from this study were obtained retrospectively and a placebo-controlled double-blind study of dexamethasone in pneumococcal meningitis is indicated. The beneficial effect of corticosteroid adjunctive therapy has been demonstrated previously only in patients with Haemophilus meningitis. Since the incidence of H. influenzae meningitis will be reduced by immunization in the future, S. pneumoniae or N. meningitidis will be the causal agent in a larger percentage of children (Schuster V, Kreth HW, N Engl J Med Dec 5, 1991; 325:1654). Dexamethasone in childhood meningitis is discussed in the correspondence section of this issue of The N Engl J Med. A retrospective evaluation of 12 years experience in the treatment of neonatal meningitis is reported from The Departments of Paediatrics and Bacteriology, Goteborg and Molndal, University of Goteborg, Sweden (Tessin I et al. Acta Paediatr Scand Oct 1991; 80:911-916). Initial treatment with ampicillin-aminoglycoside combination failed in 6 of 34 patients, despite organism sensitivity to the antibiotics.