

Neurotrophins and their relevance to neurologic disease are reviewed by Kernie SG, Parada LF (Arch Neurol May 2000;57:654-657). Neurotrophin therapy has potential in diseases of the peripheral nervous system especially because its function has been studied in peripheral sensory ganglia. Trials of subcutaneous injections of nerve growth factor (NGF), the prototypical neurotrophin, in diabetic polyneuropathy have shown promising results.

RETT SYNDROME

MECP2 MUTATIONS AND RETT SYNDROME PHENOTYPES

Seventy-one sporadic and 7 familial Rett syndrome (RTT) patients were screened for *MECP2* mutations by direct sequencing and the pattern of X chromosome inactivation (XCI) was determined in 39 RTT patients at the Baylor College of Medicine, Houston, TX. Twenty-three different disease-causing *MECP2* mutations were identified in 54 of 71 (76%) sporadic and in 2 of 7 (29%) familial cases. Thirty-one of 34 patients (91%) with classic RTT had random XCI. Nonrandom XCI was associated with milder phenotypes. RTT is caused by a partial loss of *MeCP2* function. (Amir RE, van den Veyver IB, Schultz R et al. Influence of mutation type and X chromosome inactivation on Rett syndrome phenotypes. Ann Neurol May 2000;47:670-679). (Respond: Dr Huda Y Zoghbi, Howard Hughes Medical Institute, Baylor College of Medicine, One Baylor Plaza, Houston, TX 77030).

COMMENT. Different *MECP2* mutations have similar Rett syndrome phenotypic consequences, and random X chromosome inactivation plays a role in the full phenotypic spectrum of classic RTT.

Amino acid receptors in frontal cortex in RTT syndrome. A study at Johns Hopkins University, Kennedy Krieger Institute, showed that the densities of N-methyl-D-aspartate, AMPA, and GABA, measured autoradiographically in the superior frontal gyrus, were higher in younger patients and lower in older patients when compared with controls. The age-related changes in amino acid receptor density could be correlated with the stages of RTT syndrome, younger age-stage II/III regression and seizures to a less epileptic plateau stage in older girls. (Blue ME, Naidu S, Johnston MV. Development of amino acid receptors in frontal cortex from girls with Rett syndrome. Ann Neurol 1999;45:541-545).

ATTENTION DEFICIT DISORDERS

DAMP DIAGNOSIS

A simplified pediatric school entry screening examination for the syndrome of deficits in attention, motor control and perception (DAMP) is suggested from the Goteborg University, Sweden. A population-based cohort of 113 children, 6-7 years of age (62 with and 51 without DAMP), were compared on measures of attention, motor function, language, and cognition. Attention deficits were identified by both parents and pediatrician. Four of nine motor function tests and visual reaction times discriminated between the DAMP and control groups. Design copying for diagnosing perceptual disorders was better than block design and object assembly WISC subtests. Full-scale WISC IQs were lower in the DAMP group, and children with DAMP had greater phonological processing difficulties. (Landgren M, Kjellman B, Gillberg C. Deficits in attention, motor control and perception (DAMP): a simplified school entry examination. Acta Paediatr March 2000;89:302-309). (Respond: Christopher Gillberg MD, Department of Child