Friedreich's ataxia (FRDA): Mitochondrial dysfunction and oxidative stress

FRDA is caused by mutations in the gene that codes for the mitochondrial protein frataxin. There is evidence that mitochondria in FRDA accumulate iron and that this results in free-radical accumulation. Two papers provide complementary evidence that patients with FRDA have evidence of oxidative stress: Schulz et al. (p. 1719) note that patients with FRDA have increased urinary concentration of a marker of DNA oxidative damage (8 OH²dG) and Emond et al. (p. 1752) found increased plasma levels of a product of lipid peroxidation (malondialdehyde). Both groups explored whether administration of an antioxidant could reverse the abnormality: Schulz et al. found that it did improve the urine abnormalities. ♦ The accompanying editorial by Sherer and Greenamyre (p. 1600) reviews the pathogenesis of FRDA and emphasizes that these two papers not only provide the laboratory tests that may be useful for monitoring the FRDA, but also point to a new treatment strategy—antioxidants such as idebenone, which are now entering clinical trials in FRDA.

Clinical tests for identifying and distinguishing dementias: The MMSE, ACE, and FAB

Three papers and an accompanying editorial address important issues in assessing patient cognition. Dufouil et al. (p. 1609) studied Mini-Mental State Examination (MMSE) score in 2106 subjects age 75 or older and establish norms for a population by age, sex, and education level. For example, at

age 95 the lowest 10% scored 10, the top 10% scored 27.

♦ Mathuranath et al. (p. 1613) studied a much lengthier (100-item) test—Addenbrooke's Cognitive Examination (ACE)—and found it able to distinguish AD from frontotemporal dementia. ♦ Dubois et al. (p. 1621) designed the Frontal Assessment Battery (FAB), which takes 10 minutes to administer, to assess frontal lobe function—which is not well-assessed by other brief tests.

♦ The Cummings editorial (p. 1601) places these tests into the context of the need to assess the ever-increasing number of very old subjects, and the growing recognition that a large proportion of demented patients have degeneration disorders other than AD. It is becoming possible to distinguish the different disorders by simple clinical testing.

Predicting AD from complaints of memory loss of patients versus families

Two articles address this issue. Carr et al. (p. 1724) followed 382 nondemented and mildly demented subjects for at least 2 years with tests of cognitive performance and with assessment of complaints of memory loss in subjects versus a family member (or other informant). Subject complaints did not predict dementia whereas informants did. Subject complaints of memory loss correlated with depression.

♦ Lin et al. (p. 1758) also showed that caregivers were better at predicting memory loss and further found that patients with more severe cases of AD had a decreased awareness of their memory loss.

rt-PA: Effect of timing on outcome

Two papers provide new information on rt-PA use. Marler

et al. (p. 1649) looked at the original NINDS rt-PA Stroke Study database, combining the 0 to 90 minute and 90 to 180 minute treatment groups from the study to assess patient outcomes. They found that patients treated at 0 to 90 minutes for stroke onset were more likely to improve at 24 hours and to have a better 3-month outcome than those treated at 90 to 180 minutes. There was no detectable difference in rate of hemorrhage.

Transcranial Doppler (TCD) for detecting risk of ischemia after carotid dissection

Molina et al. (p. 1738) used serial TCD in 28 patients with internal carotid dissection. The detection of microembolic signals by TCD was predictive of recurrent ischemia.

Subarachnoid hemorrhage (SAH): Rebleeding and ischemia

Brilstra et al. (p. 1656) studied 346 consecutive patients with aneurysmal SAH for the relationships between early (within 4 days) versus late (after 10 days) surgery. Early surgery was associated with greater risk of ischemia; a high rebleed rate was noted at days 11 to 21.

Glatiramer acetate (GA-Copolymer-1): Immunomodulatory effects

Gran and Tranquill et al. (p. 1704) studied possible ways GA may produce its beneficial effect in MS. They found that GA inhibits T cell clones that proliferate in response to myelin basic protein, with evidence that this effect may in turn be mediated by several mechanisms, including effects on the T cell receptor as well as by inducing T-helper immune modulator cells.

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