Predictors of tolerance and survival after noninvasive positive-pressure ventilation in ALS

Lo Coco et al. prospectively studied 71 patients with ALS on noninvasive positive-pressure ventilation. Survival was related to ventilatory use (≥4 hours/day) and to the modifications of pulmonary function decline after treatment initiation. Furthermore, bulbar involvement and body mass index predicted tolerance and survival in tolerant patients.

see page 761

Paraoxonase (*PON*) gene polymorphisms and sporadic ALS

Slowik et al. report that R-C haplotype of *PON1* and *PON2* genes increased the odds of developing SALS by threefold. This association suggests that exposure to environmental factors enhancing lipid peroxidation in individuals with genetically determined decreased PON activity may predispose to SALS.

see page 766

Saeed et al. demonstrate the association in a large cohort of patients with sporadic ALS with polymorphisms in paraoxonase genes. These enzymes detoxify agents used in the Gulf War. This study supports the hypothesis that environmental toxicity in a susceptible host contributes to ALS.

see page 771

The editorial by Shaw and Al-Chalabi about these two articles notes that there is still no proof that genetic variants other than SOD1 play a major role in susceptibility to SALS. Moreover, we do not have a functional SNP or haplotype that has improved understanding of disease pathogenesis of SALS in the way that the APOE ε4 allele does in Alzheimer disease. Although there is some evidence of an association between susceptibility to ALS and the PON cluster in both of the studies in this issue, the interpretation requires thought as it is not clear why different SNPs would be associated with ALS in the two studies. They argue for large sample sizes to improve power, and combining results from different populations.

see page 738

Cluster headache in the general population

In a two-stage cohort study of 31,750 Swedish twins, Ekbom et al. found that lifetime prevalence of cluster headache is 151 per 100,000 with a male-to-female ratio of 5:1. Twin concordance occurred in 2 out of 12 monozygous pairs but in none out of 25 dizygous pairs.

see page 798

The STARD statement: Have we improved reports of diagnostic testing?

Smidt et al. examined the quality of reporting of 265 diagnostic accuracy studies, as measured by compliance with the STARD statement. Quality of reporting in major general and neurology journals (including *Neurology*) has improved only slightly since publication of the STARD statement. Authors should include a flow diagram in their manuscript, to clarify their study design and patient flow.

see page 792

The editorial by Johnston and Holloway notes that since STARD guidelines were only published in 2003, it may be too early to see the hoped for improvement in 2004, however, these data are cause for concern. Journal editors and reviewers should demand a higher level of detail prior to publication and readers should insist on this detail prior to making clinical decisions. The availability of on-line supplements in many journals may offer an option for recording the higher level of detail that is critical for peer review and for subsequent clinical decision making.

see page 740

Blood pressure measures and risk of stroke

In a prospective cohort study of 11,466 men, Bowman et al. evaluated the ability of various blood pressure values to predict stroke and stroke subtypes. While all blood pressure measures were significant predictors of stroke risk, systolic blood pressure was the best predictor for total, ischemic, and hemorrhagic stroke.

see page 820

Excessive daytime sleepiness (EDS) in PD: Drugs or disease?

Gjerstad et al. studied associations between demographic and clinical correlates and EDS in a longitudinal study of PD followed for over 8 years. The authors found that EDS is a frequent and highly persistent feature in PD, with multifactorial underlying pathophysiology.

see page 853

A novel familial *MECP2* mutation in a young boy

Ventura et al. describe a boy with moderate mental retardation, autistic features, and epilepsy, carrying a new missense mutation on the *MECP2* gene. Male patients with unexplained mental retardation and pervasive developmental disorders deserve screening for mutations in *MECP2*.

see page 867

Hemicraniectomy for massive ischemic stroke

In a study of 42 patients with massive hemispheric infarction treated with decompressive hemicraniectomy, Rabinstein et al. found that older age predicted poor outcome, whereas degree of preoperative midline shift or timing of surgery did not.

see page 891

Gaucher mutations in synucleinopathies

Genotyping autopsy samples from subjects with different synucleinopathies, Goker-Alpan et al. identified glucocerebrosidase mutations in 23% of Lewy body dementias, implicating this mutant enzyme as a risk factor.

see page 908

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