

Multifocal motor neuropathy (MMN) with and without conduction block

Delmont et al. reviewed the clinical features and response to IVIg treatment of 20 patients with MMN vs 13 patients without CB at diagnosis. After a median follow-up of 7 years there were no differences between the two groups in term of age, sex, time from onset to diagnosis, anti-GM1 antibody titers, or CSF data. IVIg benefit was similar in patients without CB (8/13) vs with CB (14/20).

see page 592

The editorial by Chaudhry and Swash notes that MMN has a prevalence of 1 to 2/100,000. It was initially recognized as a disorder that could be confused with ALS. If one accepts that CB was adequately excluded in the Delmont et al. cases, the following question is raised: "Is there an axonal form of MMN?" The term multifocal acquired motor axonopathy (MAMA) has been used by other authors to describe such an axonal MMN variant.

A trial of IVIg seems justified in patients with progressive distal asymmetric weakness in a multifocal peripheral nerve distribution with or without CB.

see page 558

SSEPs and prognosis in postanoxic coma

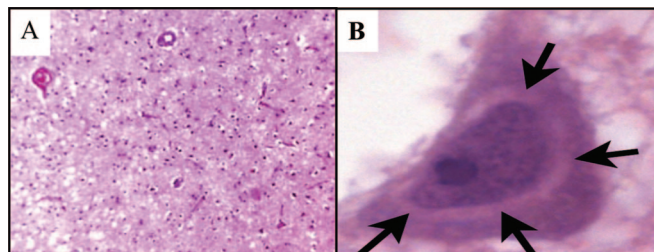
Zandbergen et al. found that determination of the N70-response of the somatosensory evoked potentials in addition to N20 may, under certain circumstances, improve the prediction of poor outcome in patients with postanoxic coma.

see page 583

The editorial by Thomas P. Bleck notes that the success of cardiopulmonary resuscitation has had the unintended consequence of increasing the number of patients in coma because of hypoxic-ischemic encephalopathy. Prognostication is usually based on physical findings elicited at defined time points after resuscitation. There have been many attempts to improve prognostication with electrophysiologic tests. Studies of the N70 potential in single centers have found the N70 SSEP to have as high a sensitivity as 94% and specificity of 97% for predicting good outcome in patients with preserved N20 potentials. In the Zandbergen et al. multicenter study, local neurophysiologists used equipment already available at each hospital. Here, there was only moderate interobserver agreement on the interpretation of the evoked potentials. In this setting, the addition of the N70 data to the N20 results was not as useful as it appeared to be in earlier investigations.

see page 556

VCP mutations, frontotemporal dementia, Paget disease, and inclusion body myopathy



(A) Diffuse microvacuolization of cortical frontal superficial layers, severely decreased neuronal density, and loss of lamination. (B) Intraneuronal lesions: arrows indicate the perinuclear vacuole.

Guyant-Maréchal et al. found valosin-containing protein gene mutations present in two families in which FTD is the most prominent symptom. The histopathologic study performed in patients harboring the R155C mutation supports the hypothesis that this mutation disrupts normal valosin-containing protein function, leading to diffuse accumulation of ubiquitinated proteins.

see page 644

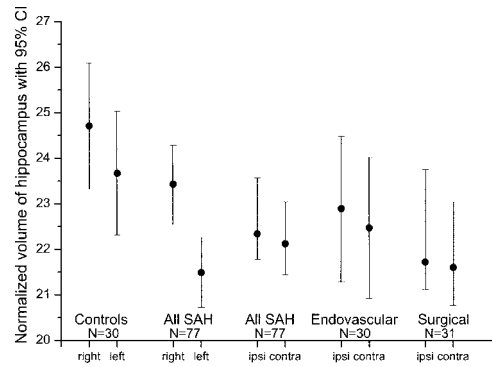
The editorial by Ghetti and Goebel notes that FTD has been found to be linked to at least three chromosomal loci: chromosomes 9, 17, and 3. The condition described by Guyant-Marechal et al. may be considered a member of the growing number of protein aggregate diseases, which includes Alzheimer disease, Parkinson disease, and prion diseases. It is also one of the protein aggregate myopathies, which include the myofibrillar myopathies and the inflammatory and hereditary inclusion body myopathies. Two regions on chromosome 17 have been linked to FTD. Mutations in the MAPT gene, located in chromosome 17q21-22, are associated with FTD. Recently, a dominant form of FTD has been mapped to a pericentromeric region of 12 cM on chromosome 3. Mutations have been found in the charged multivesicular body protein (CHMP2B) gene, which encodes a component of the endosomal sorting complexes required for transport (ESCRTIII complex).

see page 560

SAH is followed by temporomesial volume loss: An MRI study

Bendel et al. found that subarachnoid hemorrhage and its treatment may be followed by atrophy in temporomesial structures. Furthermore, the amygdala ipsilateral to the ruptured aneurysm was smaller in patients after surgical than endovascular treatment. A clear correlation was demonstrated between neuropsychological performance and reduced temporomesial volumes.

see page 575



WWW.NEUROLOGY.ORG OFFERS IMPORTANT INFORMATION TO PATIENTS AND THEIR FAMILIES

The *Neurology* Patient Page provides:

- a critical review of ground-breaking discoveries in neurologic research that are written especially for patients and their families
- up-to-date patient information about many neurologic diseases
- links to additional information resources for neurologic patients.

All *Neurology* Patient Page articles can be easily downloaded and printed, and may be reproduced to distribute for educational purposes. Click on the Patient Page icon on the home page (www.neurology.org) for a complete index of Patient Pages.

CME

NEUROLOGY ONLINE CME PROGRAM

Earn CME while reading *Neurology*. This program is available only to online *Neurology* subscribers. Simply read the articles marked CME, go to www.neurology.org, and click on CME Information. This will provide all of the information necessary to get started. The American Academy of Neurology (AAN) is accredited by the Accreditation Council for Continuing Medical Education (ACCME) to sponsor continuing medical education for physicians. *Neurology* is planned and produced in accordance with the ACCME Essentials. For more information, contact AAN Member Services at 800-879-1960.

Neurology[®]

August 22 Highlights
Neurology 2006;67;554-555
DOI 10.1212/01.wnl.0000237006.16065.c8

This information is current as of August 21, 2006

Updated Information & Services	including high resolution figures, can be found at: http://n.neurology.org/content/67/4/554.full
Subspecialty Collections	This article, along with others on similar topics, appears in the following collection(s): Vascular dementia http://n.neurology.org/cgi/collection/vascular_dementia
Permissions & Licensing	Information about reproducing this article in parts (figures, tables) or in its entirety can be found online at: http://www.neurology.org/about/about_the_journal#permissions
Reprints	Information about ordering reprints can be found online: http://n.neurology.org/subscribers/advertise

Neurology® is the official journal of the American Academy of Neurology. Published continuously since 1951, it is now a weekly with 48 issues per year. Copyright . All rights reserved. Print ISSN: 0028-3878. Online ISSN: 1526-632X.

