

ARCHIVES OF NEUROLOGY

JANUARY 2003 VOLUME 60, NUMBER 1 PAGES 1-148

EDITORIAL

- Cyp46 (24S-Cholesterol Hydroxylase):
A Genetic Risk Factor for Alzheimer Disease** 16
Benjamin Wolozin, MD, PhD

NEUROLOGICAL REVIEW

- Neurological Aspects of Biological and Chemical
Terrorism: A Review for Neurologists** 21
Coleman O. Martin, MD; Harold P. Adams, Jr, MD

ORIGINAL CONTRIBUTIONS

- Increased Brain β -Amyloid Load,
Phosphorylated Tau, and Risk of Alzheimer Disease
Associated With an Intronic CYP46 Polymorphism** 29
*Andreas Papassotiropoulos, MD;
Johannes R. Streffer, MD;
Magdalini Tsolaki, MD;
Simon Schmid, MD;
Dietmar Thal, MD;
Francesca Nicosia, PhD;
Vassiliki Iakovidou, MD;
Alessia Maddalena, MD;
Dieter Lütjohann, PhD;
Estifanos Ghebremedhin, MD;
Thomas Hegi, MD;
Thomas Pasch, MD;
Muriel Träxler, BSc;
Annette Brühl, MD;
Luisa Benussi, PhD;
Giuliano Binetti, MD;
Heiko Braak, MD;
Roger M. Nitsch, MD;
Christoph Hock, MD*

- Prediction of Early Clinical Severity and Extent
of Neuronal Damage in Anterior-Circulation
Infarction Using the Initial Serum
Neuron-Specific Enolase Level** 37
*Seung-Hun Oh, MD; Jin-Goo Lee, MD;
Sang-Jun Na, MD; Ji-Hyung Park, MD;
Young-Chul Choi, MD; Won-Joo Kim, MD*

- Antiganglioside Antibodies in Multifocal
Acquired Sensory and Motor Neuropathy** 42
*Armin Alaedini, PhD; Howard W. Sander, MD;
Arthur P. Hays, MD; Norman Latov, MD, PhD*

- Increased Metabolic Muscle Fatigue
Is Caused by Some but Not All
Mitochondrial Mutations** 50

*Wilhelm J. Schulte-Mattler, MD;
Tobias Müller, MD;
Marcus Deschauer, MD;
Frank N. Gellerich, PhD;
Paul A. Iaizzo, PhD;
Stephan Zierz, MD*

- Elevated Plasma Homocysteine Levels
in Patients Treated With Levodopa:
Association With Vascular Disease** 59

*John D. Rogers, MD;
Anna Sanchez-Saffon, BA;
Alan B. Frol, PhD;
Ramon Diaz-Arrastia, MD, PhD*

- Lower Levels of N-Acetylaspartate
in Multiple Sclerosis Patients** 65

With the Apolipoprotein E ϵ 4 Allele
*Christian Enzinger, MD; Stefan Ropele, PhD;
Siegfried Strasser-Fuchs, MD; Peter Kapeller, MD;
Helena Schmidt, MD; Birgit Poltrum, MD;
Reinhold Schmidt, MD; Hans-Peter Hartung, MD;
Franz Fazekas, MD*

- Drop Attacks and Vertigo Secondary
to a Non-Meniére Otologic Cause** 71

*Gail Ishiyama, MD;
Akira Ishiyama, MD;
Robert W. Baloh, MD*

- Effects of Subthalamic Nucleus Stimulation
and Levodopa Treatment on Gait Abnormalities
in Parkinson Disease** 80

*Pierre Krystkowiak, MD;
Jean-Louis Blatt, MD, PhD;
Jean-Louis Bourriez, PhD;
Alain Duhamel, PhD;
Miriam Perina, MD;
Serge Blond, MD;
Jean-Daniel Guieu, PhD;
Alain Destée, MD;
Luc Defebvre, MD, PhD*

- Survival Study of Vascular Dementia
in Rochester, Minnesota** 85

*David S. Knopman, MD; Walter A. Rocca, MD, MPH;
Ruth H. Cha, MS; Steven D. Edland, PhD;
Emre Kokmen, MD*

ARCHIVES OF NEUROLOGY

Survival Study of Parkinson Disease in Olmsted County, Minnesota 91
Alexis Elbaz, MD, PhD; James H. Bower, MD; Brett J. Peterson, BS; Demetrius M. Maraganore, MD; Shannon K. McDonnell, MS; J. Eric Ahlskog, PhD, MD; Daniel J. Schaid, PhD; Walter A. Rocca, MD, MPH

A Novel Central Nervous System–Enriched Spinocerebellar Ataxia Type 7 Gene Product 97
David D. Einum, PhD; Anna M. Clark, PhD; Jeannette J. Townsend, MD; Louis J. Ptacek, MD; Ying-Hui Fu, PhD

OBSERVATIONS

A Fatal Case of Coxsackievirus B4 Meningoencephalitis 107
Bruce C. Cree, MD, PhD; Gary L. Bernardini, MD, PhD; Arthur P. Hays, MD; Gina Lowe, MD

Hearing Symptoms in Migrainous Infarction 113
Hyung Lee, MD; Gregory T. Whitman, MD; Jeong Geung Lim, MD; Sang Doe Yi, MD; Yong Won Cho, MD; Sarah Ying, MD; Robert W. Baloh, MD

The Fragile X Premutation Presenting as Essential Tremor 117
Maureen A. Leehey, MD; Renato P. Munhoz, MD; Anthony E. Lang, MD; James A. Brunberg, MD; Jim Grigsby, PhD; Claudia Greco, MD; Sebastian Jacquemont, MD; Flora Tassone, PhD; A. M. Lozano, MD, PhD; Paul J. Hagerman, MD, PhD; Randi J. Hagerman, MD

IMAGES IN NEUROLOGY

Spinal Arteriovenous Malformation 125
Frank W. Drislane, MD; Jean K. Matheson, MD; David Dubuisson, MD, PhD

HISTORY OF NEUROLOGY

The History of Dermatome Mapping 126
Steven A. Greenberg, MD

HYPOTHESIS

Are Astrocytes Central Players in the Pathophysiology of Multiple Sclerosis? 132
Jacques De Keyser, MD; Esther Zeinstra, MD; Elliot Frohman, MD

BOOK REVIEWS

Ethical Issues In Neurology 138
Thomas D. Bird, MD

Neuroanatomy and Neurophysiology: A Review 138
Dewey K. Ziegler, MD

Channelopathies of the Nervous System 139
Cameron Adams, MD

Neurology and General Medicine 140
Thomas P. Bleck, MD, FCCM

REGULAR DEPARTMENTS

Reviewers for 2002 11

This Month in Archives of Neurology 14

Call for Papers 25, 35

CME Announcement 70

Calendar 142

Physician Marketplace 144

Index to Advertisers 146

Contact Information 147

Instructions for Authors are available on our Web site at www.archneurolog.com. If you do not have access to the Web, contact Jenny Kang at (214) 648-9383.

ARCHIVES OF NEUROLOGY

Mission Statement: The mission of the *Archives of Neurology* is to publish scientific information primarily important for those physicians caring for people with neurologic disorders but also for those interested in the structure and function of the normal and diseased nervous system. These specific aims are (1) to make timely publication of original research of the nervous system, (2) to record observations of single patients or groups of patients that will provide new information and insights, (3) to report more basic research that is pertinent to the understanding of disease, (4) to introduce topics of practice, ethics, teaching, and history that are useful, (5) to provide a forum for discussion on topics that may be controversial in this field. This information will be published only after extensive peer review so that originality, clarity, and precision are insured.

Editor: **Roger N. Rosenberg, MD**
Department of Neurology
University of Texas Southwestern Medical Center
5323 Harry Hines Blvd
Dallas, TX 75390-9108

Associate Editors:

Steven T. DeKosky, MD Pittsburgh, Pa
Hassan M. Fathallah-Shaykh, MD Chicago, Ill
Christopher G. Goetz, MD Chicago, Ill
Susan T. Iannaccone, MD Dallas, Tex
David E. Pleasure, MD Philadelphia, Pa
E. S. Roach, MD Dallas, Tex
Ira Shoulson, MD Rochester, NY

Assistant Editors:

Lawrence S. Honig, MD, PhD New York, NY
Matthew Menken, MD Princeton, NJ

Editorial Assistant: **Jenny Kang** Dallas, Tex

Editorial Office:

(214) 648-9383
fax: (214) 648-9388
e-mail: archneuro@mednet.swmed.edu

Section Editors

Neurological Review Editor: **David E. Pleasure, MD** Philadelphia, Pa
Controversy in Neurology/Consensus in Neurology Editor:
Steven T. DeKosky, MD Pittsburgh, Pa
Basic Science Seminars in Neurology Editor:
Hassan M. Fathallah-Shaykh, MD Chicago, Ill
Neurotherapeutics Editor: **Ira Shoulson, MD** Rochester, NY
History of Neurology Editor: **Christopher G. Goetz, MD** Chicago, Ill
Neurology and Public Health Editor: **Matthew Menken, MD** Princeton, NJ
Book Review Editor: **Susan T. Iannaccone, MD** Dallas, Tex
Web Editor: **Lawrence S. Honig, MD, PhD** New York, NY
CME Editor: **Matthew Menken, MD** Princeton, NJ

Editorial Board

James I. Ausman, MD, PhD Chicago, Ill
Louis R. Caplan, MD Boston, Mass
Mahlon R. DeLong, MD Atlanta, Ga
James A. Ferrendelli, MD Houston, Tex
John H. Growdon, MD Boston, Mass
Vladimir Hachinski, MD, DSc London, Ontario
Audrey S. Penn, MD Bethesda, Md
Stanley B. Prusiner, MD San Francisco, Calif
Michael Racke, MD Dallas, Tex
Bruce R. Ransom, MD, PhD Seattle, Wash
Thomas R. Swift, MD Augusta, Ga
Kenneth L. Tyler, MD Denver, Colo
Paul C. Van Ness, MD Dallas, Tex

International Advisory Committee

Johan A. Aarli, MD Bergen, Norway
Oded Abramsky, MD, PhD Jerusalem, Israel
Samir F. Atweh, MD Beirut, Lebanon
Paula Coutinho, MD, PhD Porto, Portugal
Alexandra Dürr, MD Paris, France
Bruno Estanol, MD Mexico City, Mexico
Cesare Fieschi, MDN Rome, Italy
Hans J. Freund, MD Düsseldorf, Germany
Wang Guo-Xiang, MD Beijing, China
Sang-Bok Lee, MD, PhD Seoul, Korea
Colin Masters, MD Melbourne, Victoria, Australia
Sergio Novis, MD Rio de Janeiro, Brazil
Michael Piradov, MD Moscow, Russia
Nippon Pongvarin, MD, FRCP Bangkok, Thailand
Anthony H. V. Schapira, DSc, MD, FRCP London, England
Bhim S. Singhal, MD Bombay, India
Erik Stalberg, MD Uppsala, Sweden
Eduardo Tolosa, MD Barcelona, Spain
Shoji Tsuji, MD, PhD Niigata, Japan

Publication Staff

Offices: 515 N State St, Chicago, IL 60610

Catherine D. DeAngelis, MD, MPH
Editor in Chief,
Scientific Publications & Multimedia Applications
Cheryl Iverson Managing Editor

Editorial Processing Department

Department Director: Paula Glitman
Copyediting Manager: Brenda Gregoline
Freelance Copyediting Manager: Diane L. Cannon
Freelance Copyediting Coordinators: Lauren B. Fischer, Connie Manno
Editorial Systems Managers: Paul Frank, Monica Mungle
Systems Programmer/Internet Specialist: J. D. Neff
Senior Copy Editor: Vickey Golden
Copy Editors: Juana N. Llorens, Rachel Novickas,
Cheryl L. Tobey, Rachel Bodzin Young
Editorial Assistant: Wanda Hill

Scientific Publications & Multimedia Applications

Assistant to the Editor: Helga Fritz

Scientific Online Resources

Director: Margaret Winker, MD
Electronic Media Department Director: Michelle Kurzynski
Associate Web Editors: Katherine Behar, Erin M. Giannini
Web Office Manager: Marla A. Hall

Publishing & Business Staff

Group Publisher: Nawin Gupta, PhD
VP Office Staff: James Clinton, Geoffrey Flick, Terri Hanley,
Maureen Archey, Anna Goins
International Business and Development:
William Fox (director), Norman Frankel, PhD, Rhonda Bailey,
Millette Jackson-Bates
Online Business and CME: Bonnie Chi-Lum, MD, MPH (publisher)

Circulation and Classified Advertising Sales

Director: Curt Reilly
Managers: Vida Damijonaitis, Kim Krebs, Thalia Moss
Staff: Kathy Braun, Ana Dirksen, Sandra Garzon,
Christine Hearne, Amy Postlewait, Trina Rizzo,
Barbara Van Wagner

Advertising Sales and Marketing

Director: Peter J. Murphy
Sales Representatives: Barbara Napoli
Marketing Services: Nancy Bell
Staff: Victoria Anne Acot, Susan Lovenberg, Mirna Monal,
Debra Ann Sardo

Reprints

Director: Mark Kuhns
Staff: Wanda Bartolotta, Joseph Reکش

Publishing Operations Division

Division Director: Vanessa Hayden
Directors: Karen Adams-Taylor, Paul Gasielcki,
Sue Sherrill
Managers: David Antos, Karen Branham, Debbie Camp,
Diane Darnell, Karl Elvin, Betty Frigerio, Linda Gardner,
Mary Ellen Johnston, Mark Matura, Michael McGraw,
Phil O'Leary, Susan Price, Charl Richey, Alfred Tomaselli,
Carmela Ware
Advertising and Production Services: Linda Alexander,
LaWanda Brown, Susan Chiara, Mary Anne Donley,
Euwanaha Joiner-Smith, Brian McCartney,
Sean O'Donnell, Noemi Olmo, Carole Piszker,
Sarah Powell, Denise Steinhauer, Ellen Yoshihara
Circulation Processing: Bernadette Bauer, Julie Burton,
Rosa Eng, Felicia Kyles, Lawrence Majkszak,
Kathryn Osten
Composition and New Media: Rebecca Aiken,
Clifford Barrett, Gail Barrett, Regina Brownlee,
Brenda Chandler-Grayer, Rowena Dagang,
Amy Evers, Mary Ann Kuranda, Sandra Lopez,
John Marcinek, Bruce McGregor, La'son Diggs Sledge,
Angela Smits
Graphics: Joe Amft, Fred Furtner, Carolyn Hall,
Maria Kowalkowski, Chris Meyer, JoAnne Weiskopf
Proofreading: Carol Joy Farrell, Daniel James,
Judith A. Liteskis, Teresa H. Omiolek,
Barbara Wojtowicz, Agnes A. Zarkadas

JAMA
&
ARCHIVES
JOURNALS
American Medical Association

All articles published, including editorials, letters, and book reviews, represent the opinions of the authors and do not reflect the policy of the American Medical Association, the Editorial Board, or the institution with which the author is affiliated, unless this is clearly specified.

Copyright 2003 by the American Medical Association. All rights reserved. Reproduction without permission is prohibited.

AMA Executives

Michael D. Maves, MD, MBA
Executive Vice President, Chief Executive Officer
Robert A. Musacchio, PhD
Senior Vice President, Publishing & Business Services

ARCHIVES OF NEUROLOGY

CONTACT INFORMATION

EDITORIAL OFFICE

Manuscript Submission: All manuscripts should be sent to Editor, *Archives of Neurology*, Department of Neurology, University of Texas Southwestern Medical Center, 5323 Harry Hines Blvd, Dallas, TX 75390-9108, USA; phone: (214) 648-9383; fax: (214) 648-9388; e-mail: archneurol@mednet.swmed.edu.
Instructions for Authors: Available online at www.archneurol.com.

CONSORTIUM OF AMA JOURNALS

The *Archives of Neurology* is a member of the consortium of AMA journals listed below.

The Journal of the American Medical Association (JAMA) Archives of Dermatology Archives of Facial Plastic Surgery Archives of General Psychiatry Archives of Internal Medicine Archives of Neurology	Archives of Ophthalmology Archives of Otolaryngology–Head & Neck Surgery Archives of Pediatrics & Adolescent Medicine Archives of Surgery
---	--

JAMA/ARCHIVES MEDIA RELATIONS

Jann Ingmire; phone: (312) 464-5262; e-mail: jamaarchmedia@ama-assn.org.

ARCHIVES OF NEUROLOGY ONLINE

Archives of Neurology is available online at www.archneurol.com.

REPRINTS

Author and Educational Reprints: Journal Products coordinator: Joseph R. Rehash; phone: (312) 464-4594; fax: (312) 464-4849. Orders of individual articles must be for a minimum of 100 reprints.
Commercial Reprints: Wanda Bartolotta; phone: (718) 946-7424; fax: (718) 946-7372; e-mail: Wanda_Bartolotta@ama-assn.org. Orders of individual articles must be for a minimum of 300 reprints.

PERMISSIONS

Information about obtaining permission to republish, reprint, or adapt material published in *Archives of Neurology*, is available from Rhonda Bailey; phone: (312) 464-2513; fax: (312) 464-5835; e-mail: Rhonda_Bailey@ama-assn.org.

DOCUMENT DELIVERY

Copies of individual articles are available online at www.archneurol.com and from the following: Infotrieve, phone: (800) 422-4633; fax: (310) 208-5971; URL: www.infotrieve.com; e-mail: orders@infotrieve.com; Institute for Scientific Information (ISI), phone: (800) 603-4367; fax: (856) 787-1679; URL: www.isidoc.com; e-mail: service@isidoc.com; Dialog Information Services; Proquest; Ovid Technologies, Inc; and EBSCO.

The ARCHIVES OF NEUROLOGY (ISSN 0003-9942) is published monthly by the American Medical Association, 515 N State St, Chicago, IL 60610-0946, and is an official publication of the Association. The ARCHIVES OF NEUROLOGY reaches more than 13 800 readers in over 79 countries each month. Periodicals postage paid at Chicago, Ill, and at additional mailing offices. GST registration number 12622 5556 RT. Canada Post International Publications Mail (Canadian Distribution) Sales Agreement No. 318973. Printed in the USA.

ADVERTISING

Information about advertising is available from the following sales representatives and online at www.archneurol.com.
Account Managers: Barbara Napoli (nationally); phone: (973) 263-9191 ext 203.
Manager, Marketing Services: Nancy Bell; phone: (973) 263-9191 ext 216.
Advertising Office: 119 Cherry Hill Rd, Suite 330, Parsippany, NJ 07054-1114; phone: (973) 263-9191.
Physician Marketplace: Thalia Moss, 515 N State St, Chicago, IL 60610; phone: (312) 464-4169; e-mail: Thalia_Moss@ama-assn.org.

ADVERTISING PRINCIPLES

Advertisements in this issue have been reviewed to comply with the principles governing advertising in AMA publications. A copy of these principles is available on request and online at www.archneurol.com. The appearance of advertising in AMA publications is not an AMA guarantee or endorsement of the product or the claims made for the product by the manufacturer.

ADDRESS CHANGES

Postmaster, send all address changes to *Archives of Neurology*, c/o the AMA Unified Service Center, American Medical Association, 515 N State St, Chicago, IL 60610. Notification of address changes must be made at least 6 weeks in advance; include both old and new addresses and a recent mailing label if sending via mail.

ANNUAL SUBSCRIPTION RATES

Subscription rates are shown in the table below. For service in the US and Americas, inquiries on pricing, address changes, missing issues, subscription billing, subscription renewal, and back issues, contact the AMA Unified Service Center, 24 hours a day, 7 days a week.

AMA members: phone: (800) 262-3211; e-mail: mssc@ama-assn.org. **Other physicians and nonphysician customers, including subscription agents:** phone: (800) 262-2350 or (312) 670-7827; fax: (312) 464-5831; e-mail: ama-subs@ama-assn.org. **Mail inquiries:** Unified Service Center, American Medical Association, PO Box 10946, Chicago, IL 60610-0946. For subscribers with mailing addresses outside the US and the Americas: Contact JAMA/Archives Journals, American Medical Association, Tower House, Sovereign Park, Market Harborough, Leicestershire LE15 9EF, UK. Phone: +44 1858-438880; fax: +44 1858-434958; e-mail: ama@subscription.co.uk.

Annual Subscription Rates

	United States	Americas	UK	Rest of World
INDIVIDUAL				
Print and Online	\$235	\$290	£205	€295
Online Only	\$180	\$180	£130	€210
INSTITUTION				
Print and Online	\$365	\$475	£310	€485
Online Only	\$275	\$275	£295	€320

In Canada, US rate plus GST. Rates for residents and medical students available at pubs.ama-assn.org, or from the AMA Unified Service Center.