

ARCHIVES OF NEUROLOGY

The ARCHIVES OF NEUROLOGY is a member of the consortium of AMA journals listed below. The complete text of all AMA journals is available online from Dialog Information Services and Ovid Technologies, Inc.

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 Pathology & Laboratory Medicine
Archives of Pediatrics & Adolescent Medicine
Archives of Surgery

The ARCHIVES OF NEUROLOGY (ISSN 0003-9942) is published monthly by the American Medical Association, 515 N State St, Chicago, IL 60610, 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, IL, 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.

SUBSCRIPTION RATES—The personal subscription rates for the ARCHIVES OF NEUROLOGY are \$225 for 1 year (12 issues) in the United States and US possessions; \$325 in the Americas; £195 outside the Americas. The institution rates for 1 year are \$350 in the United States; \$450 in the Americas; £295 outside the Americas. Special rates for residents and medical students are available. Address all subscription communications to: Subscriber Services Center, American Medical Association, PO Box 10946, Chicago, IL 60610-0946. Phone: (800) 262-2350. Fax: (312) 464-5831. E-mail: ama-subs@ama-assn.org. For mailing addresses outside the United States and US possessions, see International Subscription Information.

CHANGE OF ADDRESS—POSTMASTER, send all address changes to ARCHIVES OF NEUROLOGY, c/o Subscriber Services, American Medical Association, 515 N State St, Chicago, IL 60610. Please notify us of address change at least 6 weeks in advance to ensure uninterrupted service. Include both old and new addresses, a recent mailing label, and new ZIP code. For mailing addresses outside the United States and US possessions, see International Subscription Information.

SUBSCRIBER SERVICES—For information about subscribing to any of the AMA publications, change of address, missing issues, or purchasing back issues, please contact Subscriber Services Center, American Medical Association, PO Box 10946, Chicago, IL 60610-0946, or call (800) 262-2350 or (312) 670-SUBS (670-7827) between 8:30 AM and 4:30 PM CST. Fax: (312) 464-5831. E-mail: ama-subs@ama-assn.org. For mailing addresses outside the United States and US possessions, see International Subscription Information.

INTERNATIONAL SUBSCRIPTION INFORMATION—Subscriptions outside the United States and US possessions are served according to geographic region. Please address correspondence to the following 2 offices based on delivery address: (1) For delivery in North America, Central America and South America, contact Subscriber Services, AMA, PO Box 10946, Chicago, IL 60610-0946. Phone: (312) 670-7827. Fax: (312) 464-5831. E-mail: ama-subs@ama-assn.org. (2) For delivery outside the Americas: Subscription Department, JAMA & Archives Journals, American Medical Association, Tower House, Sovereign Park, Market Harborough, Leicestershire LE15 9EF, England. Phone: +44 (0) 1858-438880. Fax: +44 (0) 1858-434958.

REPRINTS—Authors place their reprint order at the time the edited typescript is reviewed and should allow 4 to 6 weeks for delivery following publication. Requests for individual reprints should be sent directly to the author at the address shown in the article.

For bulk reprint orders for distribution by commercial organizations, please contact Wanda Bartolotta, 2441 84th St, Brooklyn, NY 11214. Phone (718) 946-7424. Fax: (718) 946-7372. E-mail: wanda_bartolotta@ama-assn.org. For reprint orders in limited quantities for distribution by educational organizations and for author reprints, please contact Reprints Coordinator, 515 N State St, Chicago, IL 60610. Phone: (312) 464-4594. Fax: (312) 464-4849.

PERMISSIONS—Contact Rhonda Bailey, Permissions Assistant, 515 N State St, Chicago, IL 60610. Phone: (312) 464-2513.

ADVERTISING PRINCIPLES—Each advertisement in this issue has been reviewed and complies with the principles governing advertising in AMA scientific publications. A copy of these principles is available on request. 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.

WORLD WIDE WEB ADDRESS—<http://www.ama-assn.org>

NEUROLOGY HOME PAGE—<http://www.archneurol.com>

Publication Staff

Offices: 515 N State St, Chicago, IL 60610

Editorial Processing Department

Division Director: Cheryl Iverson
Director: Paula Glitman
Copyediting Manager: Lise M. Stevens
Freelance Copyediting Manager: Diane L. Cannon
Freelance Copyediting Coordinators: Gwen Chaffin, Connie Manno
Electronic Editing Specialist: Paul Frank
Assistant Electronic Editing Specialist: Monica Mungle
Senior Copy Editors: Dee E. Egger, Vicky Golden
Copy Editors: Jean Kang, John McFadden, Donald S. Smith
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
Web Development Editor: Keith Huff
Web Copy Editor: Erin M. Giannini
Web Office Manager: Marla A. Hall
Editorial Assistant: Gemma Wall

Specialty Journal Division Office

Manager, Marketing Services: Geoffrey A. Flick
Assistant to the Publisher: Christine Hearne

Circulation & Content Marketing Division

Administrative Assistant: Millette Jackson-Bates

Circulation Processing Department

Director: Robert Herling
Circulation Systems Manager: Alfred Tomaselli
Circulation Specialists: Reginald Jordan, Kathryn Osten

Circulation Development Department

Director: Bina Oza
Circulation Marketing Managers: Vida Damijonaitis, Donna Goldman, Michael McGraw
Circulation Marketing Specialist: Sandra Garzon

Licensing & Permissions Department

Director: Norman Frankel
Permissions: Rhonda Bailey

Production & Distribution Division

Directors: Karen Adams-Taylor, Paul Gasielcki, Vanessa Hayden
Managers: David Antos, Karen Branham, Debbie Camp, Diane Darnell, Karl Elvin, Betty Frigerio, Terri Hanley, Mary Ellen Johnston, Carole Piszker, Susan Price, Charl Richey, Carmela Ware
Production Assistants: Kristy Kambanis, Ruth Sprague
Production Associates and Specialists Advertising and Production Services: LaWanda Brown, Susan Chiara, Mary Anne Donley, Sarah Powell, Denise Steinhauer, Ellen Yoshihara
Composition: Rebecca Aiken, Brenda Chandler-Grayer, Amy Evers, Fred Furtner, Mary Ann Kuranda, Sandra Lopez
Database & New Media: Bruce McGregor (indexer), Clifford O. Barrett, Gail Barrett, Regina Brownlee, Rowena F. Dagang, La'son Diggs Sledge, Angela Smits
Distribution: Linda Alexander, Michelle Robinson
Graphics: Joe Amft, Noel Chaparro, Carolyn Hall, Maria Kowalkowski, Chris Meyer, JoAnne Weiskopf
Proofreading: Daniel James, Teresa H. Omiolek, Mary Kay Tinerella, Barbara Wojtowicz, Agnes A. Zarkadas

Advertising

Director, Advertising Sales: Peter G. Messina
AMA Classified Advertising Department: Thalia Moss, 800-262-2260.
Advertising Office:
East: Kelly Lezynski, 119 Cherry Hill Rd, 3rd Flr, Parsippany, NJ 07054 (973) 263-9191.
Midwest/West: J. P. Cahill & Associates, 341 S Waiola Ave, La Grange, IL 60525 (708) 352-1450.



ARCHIVES OF NEUROLOGY

JUNE 2001

VOLUME 58, NUMBER 6

PAGES 849-1036

EDITORIALS

Titin, Thymoma, and Myasthenia Gravis 869
Johan A. Aarli, MD

The Challenge of Respiratory Dysfunction in Guillain-Barré Syndrome 871
Angelika F. Hahn, MD, FRCPC

BASIC SCIENCE SEMINARS IN NEUROLOGY

Plasticity, Hippocampal Place Cells, and Cognitive Maps 874
Matthew Shapiro, PhD

ORIGINAL CONTRIBUTIONS

Anti-Titin Antibodies in Myasthenia Gravis: Tight Association With Thymoma and Heterogeneity of Nonthymoma Patients 885
Ana Maria Yamamoto, PhD;
Philippe Gajdos, MD, PhD;
Bruno Eymard, MD, PhD;
Christine Tranchant, MD, PhD;
Jean-Marie Warter, MD, PhD;
Lucienne Gomez, BSc; Charles Bourquin, MD;
Jean-François Bach, MD, PhD;
Henri-Jean Garchon, MD, PhD

Anticipating Mechanical Ventilation in Guillain-Barré Syndrome 893
Nicholas D. Lawn, FRACP; Dade D. Fletcher, MD;
Robert D. Henderson, FRACP; Troy D. Wolter, MS;
Eelco F. M. Wijdicks, MD

Neurologic Findings in Machado-Joseph Disease: Relation With Disease Duration, Subtypes, and (CAG)_n 899

Laura B. Jardim, PhD; Maria L. Pereira, PhD;
Isabel Silveira, PhD; Anabela Ferro;
Jorge Sequeiros, PhD; Roberto Giugliani, PhD

Response to Levodopa Treatment in Dopa-Responsive Dystonia 905
John G. Nutt, MD; Torbjørn G. Nygaard, MD

Early Electrodiagnostic Findings in Guillain-Barré Syndrome 913
Paul H. Gordon, MD; Asa J. Willbourn, MD

Cellular Distribution of Torsin A and Torsin B in Normal Human Brain 921
Marina Konakova, PhD; Duong P. Huynh, PhD;
William Yong, MD; Stefan M. Puls, MD

[¹⁸F]FDG-PET Reveals Temporal Hypometabolism in Patients With Temporal Lobe Epilepsy Even When Quantitative MRI and Histopathological Analysis Show Only Mild Hippocampal Damage 933
Salla Lamusuo, MD; Leena Jutila, MD;
Aarne Ylinen, MD, PhD;
Reetta Kälviäinen, MD, PhD;
Esa Mervaala, MD, PhD;
Merja Haaparanta, PhD;
Satu Jääskeläinen, MD, PhD;
Kaarina Partanen, MD, PhD;
Matti Vapalahti, MD, PhD;
Juha Rinne, MD, PhD

American Medical Association

Physicians dedicated to the health of America



Copyright 2001 by the American Medical Association. All rights reserved.
Reproduction without permission is prohibited.

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.

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

Peter L. Payerli
Interim Vice President, Publishing
Publisher

Cheryl Iverson
Managing Editor

E. Ratcliffe Anderson, Jr, MD
Executive Vice President,
Chief Executive Officer

Robert A. Musacchio, PhD
Senior Vice President,
Publishing & Business Services

Oral Almotriptan vs Oral Sumatriptan in the Abortive Treatment of Migraine: A Double-blind, Randomized, Parallel-Group, Optimum-Dose Comparison

Egilius L. H. Spierings, MD, PhD;
Baltazar Gomez-Mancilla, MD, PhD;
Daniel E. Grosz, MD;
Clayton R. Rowland, PhD;
Fredrick S. Whaley, PhD;
Kathleen J. Jirgens

944

Use of the Multiple Sclerosis Functional Composite as an Outcome Measure in a Phase 3 Clinical Trial

Jeffrey A. Cohen, MD; Gary R. Cutter, PhD;
Jill S. Fischer, PhD; Andrew D. Goodman, MD;
Fedora R. Heidenreich, MD; Amy J. Jak, MA;
Judith E. Kniker, MA; Mariska F. Kooijmans, MD, PhD;
Julia M. Lull, BA; Alfred W. Sandrock, MD, PhD;
Jack H. Simon, MD; Nancy A. Simonian, MD;
John N. Whitaker, MD; for the IMPACT Investigators

961

Autopsy-Confirmed Familial Early-Onset Alzheimer Disease Caused by the L153V Presenilin 1 Mutation

John C. Janssen, MRCP;
Peter L. Lantos, MD, PhD, DSc, FRCPath;
Nicholas C. Fox, MRCP;
Richard J. Harvey, MD, MRCPsych;
Jonathan Beck, BSc;
Andrew Dickinson, BSc;
Tracey A. Campbell, BSc;
John Collinge, MD, FRCP, FRCPath;
Diane P. Hanger, PhD;
Lisa Cipolotti, PhD;
John M. Stevens, FRCP;
Martin N. Rossor, MD, FRCP

953

Mutation Analysis and the Correlation Between Genotype and Phenotype of Arg778Leu Mutation in Chinese Patients With Wilson Disease

Zhi-Ying Wu, MD, PhD; Ning Wang, MD, PhD;
Min-Ting Lin, MD; Ling Fang, MD, PhD;
Shen-Xing Murong, MD; Long Yu, MD, PhD

971

Attention and Fluctuating Attention in Patients With Dementia With Lewy Bodies and Alzheimer Disease

Clive Ballard, MRCPsych, MD;
John O'Brien, MRCPsych, DM; Alistair Gray, BSc;
Franchesca Cormack, BSc; Gareth Ayre, PhD;
Elise Rowan, PhD; Peter Thompson, MRCPsych;
Romola Bucks, PhD; Ian McKeith, FRCPsych, MD;
Matthew Walker, PhD; Martin Tovee, PhD

977

ARCHIVES
OF
NEUROLOGY

Editor:

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

Associate Editors:

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

John N. Whitaker, MD
Birmingham, Ala

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: archneurol@email.swmed.edu

Section Editors:

Neurological Review Editor:
David E. Pleasure, MD
Philadelphia, Pa

*Controversy in Neurology/
Consensus in Neurology Editor:*

John N. Whitaker, MD
Birmingham, Ala

*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

Steven T. DeKosky, MD
Pittsburgh, Pa

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

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, MD
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

Niphon 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

OBSERVATIONS

- An Apparently Sporadic Case With *Parkin* Gene Mutation in a Korean Woman** 988
Beom S. Jeon, MD, PhD; Jong-Min Kim, MD; Dong-Soo Lee, MD, PhD; Nobutaka Hattori, MD, PhD; Yoshikuni Mizuno, MD, PhD
- Diffusion-Weighted Magnetic Resonance Imaging in Nonconvulsive Status Epilepticus** 993
Kon Chu, MD; Dong-Wha Kang, MD; Joo-Yong Kim, MD; Kee-Hyun Chang, MD; Sang Kun Lee, MD
- A Family With X-linked Dystonia-Deafness Syndrome With a Novel Mutation of the *DDP* Gene** 1004
Hiroshi Ujike, MD, PhD; Yasuyuki Tanabe, MD, PhD; Yasushi Takehisa, MD; Toshiyuki Hayabara, MD, PhD; Shigetoshi Kuroda, MD, PhD

IMAGES IN NEUROLOGY

- Macular Star in Neuroretinitis** 1008
M. Tariq Bhatti, MD; Reem Asif, MD; Lubna B. Bhatti, MBBS
- Clinical Brainstem Death With Preserved Electroencephalographic Activity and Visual Evoked Response** 1010
Machiel J. Zwarts, MD, PhD; Francois H. M. Kornips, MD; Oscar M. Vogels, MD, PhD

HISTORY OF NEUROLOGY: SEMINAL CITATIONS

- Emergence of Temporal Lobe Surgery for Epilepsy** 1011
Kimford J. Meador, MD
- Charcot-Marie-Tooth Disease** 1014
A. Gordon Smith, MD

HISTORY OF NEUROLOGY: NEUROLOGY WAS THERE

- Neurology Was There in the 1930s** 1017
Richard Satran, MD

BOOK REVIEWS

- Prognosis of Neurological Disorders, 2nd ed** 1020
Eleftherios S. Papathanasiou, MS
- Arteriovenous Malformations in Functional Areas of the Brain** 1020
Robert D. Brown, Jr, MD

LETTERS TO THE EDITOR

- Coexistence of 2 Different Type I Repeats** 1022
Takashi Kageyama, MD; Shun Shimohama, MD, PhD; Jun Kawamata, MD
- Statin-Alzheimer Disease Association Not Yet Proven** 1022
Gerson T. Lesser, MD; Leslie S. Libow, MD
- In Reply** 1023
Benjamin Wolozin, MD, PhD; George Siegel, MD
- Statin Therapy and the Prevention of Dementia** 1023
Edward L. E. M. Bollen; Allan Gaw, PhD; Brendan M. Buckley, MD
- In Reply** 1024
Benjamin Wolozin, MD, PhD; George Siegel, MD
- Neurology Was There: 1929** 1024
Richard F. Mayer, MD
- T2-Hyperintense Lateral Rim and Hypointense Putamen Are Typical but Not Exclusive of Multiple System Atrophy** 1024
Frédéric Macia, MD; Farid Yekhleif, MD; Guillaume Ballan, MD; Olivia Delmer, MD; François Tison, MD, PhD
- In Reply** 1026
Eduard Kraft; Johannes Schwarz; Claudia Trenkwalder; Wolfgang H. Oertel

REGULAR DEPARTMENTS

- This Month in Archives of Neurology** 866
- Call for Papers** 958
- Calendar** 1027
- ARCHIVES CME** 1028
- Classified Advertising** 1031
- Index to Advertisers** 1035
- Instructions for Authors—**
See January 2001 issue, page 144, or visit the Web site at www.archneurol.com