

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 Information Access Company.

The Journal of the American Medical Association (JAMA)

Archives of Dermatology
Archives of Family Medicine
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 11 600 readers in over 77 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 \$168 for 1 year (12 issues) in the United States and US possessions; \$200 in the Americas; £129 outside the Americas. The institution rates for 1 year are \$237 in the United States; \$274 in the Americas; £177 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, contact JAMA & Archives Journals, Reader Services Centre, PO Box 299, London WC1H 9TD, England. Phone: +44 (0)171-383-6270. Fax: +44 (0)171-383-6402. E-mail: bmjsubs@dial.pipex.com.

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, 500 Fifth Ave, #2210, New York, NY 10110. Phone (212) 354-0050. Fax: (212) 354-1169. E-mail: wanda_bartolotta@ama-assn.org. For reprint orders in limited quantities for distribution by educational organizations and for author reprints, please contact Joseph R. Rekish, 515 N State St, Chicago, IL 60610. Phone: (312) 464-4594. Fax: (312) 464-4849.

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

NEUROLOGY HOME PAGE—<http://www.ama-assn.org/neuro>.

PERMISSIONS—Contact Ada Jimenez-Walker, 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.

Publication Staff

Offices: 515 N State St
Chicago, IL 60610

Editorial Processing Department, Archives Journals

Director: Paula Glitman
Manager: Stacy Christiansen
Freelance Manager: Diane L. Cannon
Freelance Coordinator:
Connie Manno

Electronic Editing Specialist:

Paul Frank

Assistant Electronic Editing Specialist:

Monica Mungle

Senior Copy Editor:

Dee E. Egger

Copy Editors:

Hemranee Bhoyroo

Barbara Owczarek

Editorial Assistant:

Celina Canchola

Scientific Information and Multimedia Group

Administrative Assistant: Helga Fritz

New Media Editorial Office

Editorial Director, New Media:

William M. Silberg

Associate Editor:

Marty Suter

Assistant Editors:

Deborah Flapan

Neal Shankman

Staff Assistant: Marla A. Hall

Production & Distribution Division

Manager, Budgets & Costs:

Bonnie Van Clevon

Publishing Manufacturing Analyst:

Diane Darnell

Office Manager:

Karen Branham

Senior Secretary:

Shannon Epplert

Advertising & Production Department

Director:

Karen Adams-Taylor

Manager, Advertising Services:

Carole Piszker

Manager, Production Services:

Susan Price

Production Associates:

Debbie Camp

Betty Frigerio

Mari Guizzetti

Sarah Powell

Jennifer Reiling

Chad Sanderson

Jo Anne Turner

Carmela Ware

Distribution

Distribution Manager:

Paul Gasielki

Senior Traffic Control Specialist:

Michael McGraw

Traffic Control Specialist:

Victoria Buckner



Electronic Production Department

Director:

Linda Knott

Manager, Composition:

Phil Arko

Electronic Production Operators:

Kim Buy

Brenda Chandler

Amy Evers

Mary Ann Kuranda

Sandra Lopez

Manager, Graphics:

Charl Richey

Graphics Operators:

Joe Amft

JoAnne Weiskopf

Alicja Wojcik-Dunn

Manager, Proofreading:

Teresa H. Omiotek

Proofreaders:

David Antos

Daniel James

Mary Kay Tinerella

Barbara Wojtowicz

Production Assistant:

Ruth Sprague

Database & New Media

Manager, Database & New Media:

Mary Ellen Johnston

Indexer:

Kathy Gaydar

Database Assistants:

Gail Barrett

La'son J. Diggs

Melanie Parenti

Specialty Journal Division Office

Assistant to the Publisher:

Christine Hearne

Circulation & Content Marketing Division

Administrative Assistant:

Millette Jackson-Bates

Circulation Processing Department

Director:

Beverly Martin

Circulation Development Department

Director:

Bina Oza

Licensing & Permissions Department

Director:

Norman Frankel

Permissions:

Ada Jimenez-Walker

International Coordinator:

Sharon Ali

Reprints

Reprint Coordinator:

Joseph Rekish



ARCHIVES OF NEUROLOGY

VOL 55 NO. 6, JUNE 1998

NEUROLOGICAL REVIEW

- Recent Insights Into Neurofibromatosis** 778
Type 1: Clear Genetic Progress
David H. Gutmann, MD, PhD

BASIC SCIENCE SEMINARS IN NEUROLOGY

- Complementary DNA Libraries and** 785
Neurological Disease
Benjamin Joseph, BA; Henry Furneaux, PhD

ORIGINAL CONTRIBUTIONS

- The GTP Cyclohydrolase I Gene in Russian** 789
Families With Dopa-Responsive Dystonia
Sergei N. Illarioshkin, MD; Elena D. Markova, MD;
Pyotr A. Slominsky, PhD; Natalya I. Miklina, MD;
Svetlana N. Popova, PhD; Svetlana A. Limborska, PhD;
Shoji Tsuji, MD; Irina A. Ivanova-Smolenskaya, MD

- Increased Production of Tumor Necrosis** 793
Factor α , and Not of Interferon γ ,
Preceding Disease Activity in Patients
With Multiple Sclerosis
Bob W. van Oosten, MD; Frederik Barkhof, MD;
Petra E. T. Scholten; B. Mary E. von Blomberg, PhD;
Herman J. Adèr, PhD; Chris H. Polman, MD

- Awareness of Involuntary Movements** 801
in Huntington Disease
Julie S. Snowden, PhD; David Craufurd, MSc;
Helen L. Griffiths, MSc; David Neary, MD

- Apolipoprotein E $\epsilon 4$ Allele and Familial** 810
Aggregation of Alzheimer Disease

Maria Martinez, PhD;
Dominique Campion, PhD, MD;
Alexis Brice, MD; Didier Hannequin, MD;
Bruno Dubois, MD; Olivier Didierjean, RA;
Agnès Michon, MD; Catherine Thomas-Anterion, MD;
Michèle Puel, MD; Thierry Frebourg, PhD, MD;
Yves Agid, PhD, MD; Françoise Clerget-Darpoux, PhD

- Anticardiolipin Antibodies Are** 817
Frequently Present in Patients With
Idiopathic Intracranial Hypertension
Ronon R. Leker, MD; Israel Steiner, MD

- Diagnostic Criteria for Essential** 823
Tremor: A Population Perspective
Elan D. Louis, MD, MS; Blair Ford, MD;
Helen Lee, MS; Howard Andrews, PhD;
Gabiella Cameron, MD

- Severity of Cognitive Impairment in Juvenile** 835
and Late-Onset Huntington Disease
Estrella Gómez-Tortosa, MD;
Antonio del Barrio, PhD; Pedro J. García Ruiz, MD;
Rosario Sánchez Pernaute, MD; Javier Benítez, MD;
Alicia Barroso, PhD; F. Javier Jiménez, MD;
Justo García Yébenes, MD

- Neuropsychological Impairment From Acute** 845
Low-Level Exposure to Carbon Monoxide
Yona Amitai, MD; Zoli Zlotogorski, PhD;
Vered Golan-Katzav, MA; Anya Wexler, MD;
Ditza Gross, PhD

American Medical Association

Physicians dedicated to the health of America



Copyright 1998 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.

Lynn Jensen, PhD
Chief Operating Officer &
Interim Executive Vice President

George D. Lundberg, MD
Editor in Chief, Scientific
Information and Multimedia

Robert L. Kennett
Interim Senior Vice President,
Publishing & Business Services

Peter L. Payerli
Publisher

Vanessa Hayden
Director, Production &
Distribution Division

Cheryl Iverson
Director, Editorial Processing
Division

James A. Skowrenski
Director, Circulation &
Content Marketing Division

Geoffrey A. Flick
Manager, Marketing Services

Advertising Offices: East: Kelly
Lezynski, 119 Cherry Hill Rd, 3rd Flr,
Parsippany, NJ 07054 (973) 263-
9191. **Midwest/West:** J. P. Cahill & As-
sociates, 341 S Waiola Ave, La Grange,
IL 60525 (708) 352-1450. **AMA Clas-**
sified Advertising Department: Thalia
Moss, 800-262-2260.

OBSERVATIONS

- Magnetic Resonance Spectroscopy: Use in Monitoring MELAS Treatment** 849
Steven G. Pavlakis, MD; Peter B. Kingsley, PhD; Gary P. Kaplan, MD, PhD; Peter W. Stacpoole, MD, PhD; Michael O'Shea, MEE; Dana Lustbader, MD
- Familial Idiopathic Intracranial Hypertension With Spinal and Radicular Pain** 854
Raffaele Santinelli, MD; Carlo Tolone, MD; Roberto Toraldo, MD; Gianfranco Canino, MD; Alfredo De Simone, MD; Michele D'Avanzo, MD
- Patients With Epilepsy Who Die Suddenly Have Cardiac Disease** 857
Benjamin H. Natelson, MD; Ronald V. Suarez, MD; Christopher F. Terrence, MD; Rosa Turizo, MD

IMAGES IN NEUROLOGY

- Disc Edema and Retinal Periphlebitis as the Initial Manifestation of Sarcoidosis** 862
Jennifer E. Thorne, MD; Steven L. Galetta, MD

HISTORY: NEUROLOGY WAS THERE

- Neurology Was There in 1848** 867
Richard Satran, MD

OBITUARY

- Luigi Amaducci, MD, 1932-1998** 876
Sandro Sorbi, MD

LETTERS TO THE EDITOR

- The Central Visual Field in Homonymous Hemianopia: Foveal or Macular Representation?** 881
Peter Wellings, FRCS, FRACO
- In Reply** 881
Lawrence G. Gray, OD; Steven L. Galetta, MD; Norman J. Schatz, MD
- Hemisphere-Specific Cognitive and Motor Changes After Unilateral Posteroventral Pallidotomy** 881
Lisa Trépanier, MA; Jean Saint-Cyr, PhD; Anthony Lang, MD; Andres Lozano, MD, PhD
- In Reply** 883
Vicki M. Soukup, PhD; Fred Ingram, PhD; Mya C. Schiess, MD; James G. Bonnen, MD; Haring J. W. Nauta, MD, PhD; John R. Calverley, MD

REGULAR DEPARTMENTS

- This Month in the Archives of Neurology** 775
- Book Reviews** 879
- Calendar** 884
- Classified Advertising** 886
- Index to Advertisers** 888
- Instructions for Authors—**
See September 1997 issue, page 1172, or visit the Web site at www.ama-assn.org.

ARCHIVES

OF

NEUROLOGY

Editor:
Roger N. Rosenberg, MD
Department of Neurology
University of Texas Southwestern
Medical Center
5323 Hines Blvd
Dallas, TX 75235-9108

Associate Editors:
Vladimir Hachinski, MD, FRCPC, DScMed
London, Ontario

Susan T. Iannaccone, MD
Dallas, Tex

David E. Pleasure, MD
Philadelphia, Pa

E. S. Roach, MD
Dallas, Tex

Assistant Editors:
Christopher Goetz, MD
Chicago, Ill

Lawrence S. Honig, MD, PhD
Dallas, Tex

Ira Shoulson, MD
Rochester, NY

Editorial Assistants:
Jenny Kang
Dallas, Tex

Constance S. Adkins
Dallas, Tex

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

Section Editor:
*Controversies in Neurology/
Consensus in Neurology Editor:*
Vladimir Hachinski, MD, FRCPC, DScMed
London, Ontario

History Editor:
Christopher Goetz, MD
Chicago, Ill

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

Web Editor:
Lawrence S. Honig, MD, PhD
Dallas, Tex

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

Hassan Fathallah-Shaykh, MD
Dallas, Tex

James A. Ferrendelli, MD
Houston, Tex

John H. Growdon, MD
Boston, Mass

Matthew Menken, MD
Somerset, NJ

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

John N. Whitaker, MD
Birmingham, Ala

International Advisory Committee

Johan A. Aarli, MD
Bergen, Norway

Oded Abramsky, MD, PhD
Jerusalem, Israel

Paula Coutinho, MD, PhD
Porto, Portugal

Alexandra Dürr, MD
Paris, France

Colin Masters, MD
Melbourne, Victoria, Australia

Michael Piradov, MD
Moscow, Russia

Anthony H. V. Schapira, DSc, MD, FRCP
London, England

Erik Stalberg, MD
Uppsala, Sweden

Eduardo Tolosa, MD
Barcelona, Spain

Shoji Tsuji, MD, PhD
Niigata, Japan