

# Neurology<sup>®</sup> Neuroimmunology & Neuroinflammation

## Table of Contents

Neurology.org/nn • Online ISSN: 2332-7812  
Volume 2, Number 5, October 2015



### EDITOR'S CORNER

e159 Name a brain protein, and an autoantibody shall be found!

*J. Dalmau*

**OPEN**

### EDITORIAL

e152 Subtype-specific therapy for autoimmune neuropathies?

*E. Lancaster and S.S. Scherer*

**OPEN** Companion article, e149

### ARTICLES

e149 Rituximab in treatment-resistant CIDP with antibodies against paranodal proteins

*L. Querol, R. Rojas-García, J. Diaz-Manera, J. Barcena, J. Pardo, A. Ortega-Moreno, M.J. Sedano, L. Seró-Ballesteros, A. Carvajal, N. Ortiz, E. Gallardo, and I. Illa*

**OPEN** ▲ Editorial, e152

e139 Macrophage migration inhibitory factor promotes resistance to glucocorticoid treatment in EAE

*N. Ji, A. Kovalovsky, G. Fingerle-Rowson, M.N. Guentzel, and T.G. Forsthuber*

**OPEN**

e138 CSF cytokine profile distinguishes multifocal motor neuropathy from progressive muscular atrophy

*T. Furukawa, N. Matsui, K. Fujita, H. Nodera, F. Shimizu, K. Miyamoto, Y. Takahashi, T. Kanda, S. Kusunoki, Y. Izumi, and R. Kaji*

**OPEN**

e145 Direct MRI detection of impending plaque development in multiple sclerosis

*M. Absinta, G. Nair, P. Sati, I.C.M. Cortese, M. Filippi, and D.S. Reich*

**OPEN**

e155 Gray matter-related proteins are associated with childhood-onset multiple sclerosis

*V. Singh, E.D. van Pelt, M.P. Stoop, C. Stingl, I.A. Ketelslegers, R.F. Neuteboom, C.E. Catsman-Berrevoets, T.M. Luiders, and R.Q. Hintzen*

**OPEN**

e148 Deregulated Fc $\gamma$  receptor expression in patients with CIDP

*I. Quast, F. Cueni, F. Nimmerjahn, B. Tackenberg, and J.D. Lünemann*

**OPEN**

e141 NMDA receptor antibodies: A rare association in inflammatory demyelinating diseases

*M. Ramberger, G. Bsteh, K. Schanda, R. Höftberger, K. Rostásy, M. Baumann, F. Aboulenein-Djamshidian, A. Lutterotti, F. Deisenhammer, T. Berger, and M. Reindl*

**OPEN**

e143 New type of encephalomyelitis responsive to trimethoprim/sulfamethoxazole treatment in Japan

*Y. Sakiyama, N. Kanda, Y. Higuchi, M. Yoshimura, H. Wakaguri, Y. Takata, O. Watanabe, J. Yuan, Y. Tashiro, R. Saigo, S. Nozuma, A. Yoshimura, S. Arishima, K. Ikeda, K. Shinohara, H. Arata, K. Michizono, K. Higashi, A. Hashiguchi, Y. Okamoto, R. Hirano, T. Shiraishi, E. Matsuura, R. Okubo, I. Higuchi, M. Goto, H. Hirano, A. Sano, T. Iwasaki, F. Matsuda, S. Izumo, and H. Takashima*

**OPEN**

e144 Immunologic monitoring during a phase 2a trial of the GNBAC1 antibody in patients with MS

*M. Zimmermann, N.S.R. Sanderson, M. Rasenack, P.H. Lalive, A.B. Lang, F. Curtin, R.L.P. Lindberg, L. Kappos, and T. Derfuss*

**OPEN** ▲

e147 Effects of diazoxide in multiple sclerosis: A randomized, double-blind phase 2 clinical trial

*P. Villoslada, A. Rovira, X. Montalban, R. Arroyo, F. Paul, V. Meca-Lallana, C. Ramo, O. Fernandez, A. Saiz, A. Garcia-Merino, L. Ramió-Torrentà, B. Casanova, C. Oreja-Guevara, D. Muñoz, J.E. Martinez-Rodriguez, E. Lensch, J.M. Prieto, S.G. Meuth, X. Nuñez, C. Campás, and M. Pugliese, for the NeuroAdvan study*

**OPEN** ▲

e153 *GRIN1* polymorphisms do not affect susceptibility or phenotype in NMDA receptor encephalitis  
G.S. Day, H. Prüss, S.M. Benseler, T.A. Paton, A.D. Paterson, and D.M. Andrade

**OPEN**

e154 Pharmacogenomic study in patients with multiple sclerosis: Responders and nonresponders to IFN- $\beta$   
M.F. Bustamante, C. Morcillo-Suárez, S. Malhotra, J. Rio, L. Leyva, O. Fernández, U.K. Zettl, J. Killestein, D. Brassat, J.A. García-Merino, A.J. Sánchez, E. Urcelay, R. Alvarez-Lafuente, L.M. Villar, J.C. Alvarez-Cermeño, X. Farré, J. Lechner-Scott, K. Vandebroek, A. Rodríguez-Antigüedad, J.S. Drulovic, F. Martinelli Boneschi, A. Chan, J. Oksenberg, A. Navarro, X. Montalban, and M. Comabella

**OPEN**

### CLINICAL/SCIENTIFIC NOTES

e146 Faciobrachio-crural dystonic seizures in LGI1 limbic encephalitis: A treatable cause of falls  
F.S. Ismail, S. Popkirov, J. Wellmer, and W. Grönheit

**OPEN** 

e142 Improvement of GAD65-associated autoimmune epilepsy with testosterone replacement therapy  
M. Heiry, P. Afra, F. Matsuo, J.E. Greenlee, and S.L. Clardy

**OPEN** 

e150 Pediatric anti-Hu-associated encephalitis with clinical features of Rasmussen encephalitis  
B.R. Aravamuthan, I. Sánchez Fernández, J. Zurawski, H. Olson, M. Gorman, and M. Takeoka

**OPEN**

e151 Incomplete Susac syndrome exacerbated after natalizumab  
L. Zhovtis Ryerson, I. Kister, M. Snuderl, C. Magro, and B. Bielekova

**OPEN**

e156 Plasticity-related gene 5: A novel surface autoantigen in paraneoplastic cerebellar degeneration  
M.H. van Coevorden-Hameete, E. de Graaff, M.J. Titulaer, E. Hulsboom, L. Sabater, C.C. Hoogenraad, and P.A. Sillevius Smitt

**OPEN**

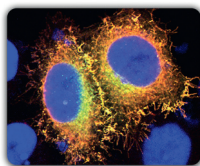
 Video  LOE classification  LOE recommendation  Podcast



[tinyurl.com/NeurologyNN](http://tinyurl.com/NeurologyNN)



[twitter.com/GreenJournal](https://twitter.com/GreenJournal)



**Cover image:** HeLa cells expressing PRG5 tagged with GFP (green) were permeabilized and stained with a patient serum that strongly recognizes PRG5 (red). See "Plasticity-related gene 5: A novel surface autoantigen in paraneoplastic cerebellar degeneration."

# Neurology® Neuroimmunology & Neuroinflammation

Neurology.org/nn • Online ISSN: 2332-7812  
Volume 2, Number 5, October 2015



**VISION:** *Neurology: Neuroimmunology & Neuroinflammation* will be the premier peer-reviewed journal for experts in the fields of Neuroimmunology and Neuroinflammation.

**MISSION:** *Neurology: Neuroimmunology & Neuroinflammation* will provide neurologists with peer-reviewed articles, editorials, and reviews to enhance patient care, education, and clinical & translational research.

## Editor

*Neurology: Neuroimmunology & Neuroinflammation*

Josep O. Dalmau, MD, PhD  
ICREA-University of Barcelona  
Barcelona, Spain  
University of Pennsylvania  
Philadelphia, PA  
Tel: 612-928-6400  
Toll-free: 800-957-3182 (US)  
Fax: 612-454-2748  
nnjournal@neurology.org

## Editor-in-Chief

*Neurology*®  
Robert A. Gross, MD, PhD, FAAN  
Professor of Neurology and of Pharmacology and Physiology  
Strong Epilepsy Center  
University of Rochester Medical Center  
Rochester, NY  
Tel: 612-928-6400  
Toll-free: 800-957-3182 (US)  
Fax: 612-454-2748  
journal@neurology.org

## Editorial Inquiries

Tel: 612-928-6400  
Toll-free: 800-957-3182 (US)  
Fax: 612-454-2748  
nnjournal@neurology.org

## Academy Officers

Terrence L. Cascino, MD, FAAN, President  
Ralph L. Sacco, MD, MS, FAAN, President Elect  
James C. Stevens, MD, FAAN, Vice President  
Aaron E. Miller, MD, FAAN, Secretary  
Lisa M. Shulman, MD, FAAN, Treasurer  
Timothy A. Pedley, MD, FAAN, Past President

## Executive Office, American Academy of Neurology

Catherine M. Rydell, CAE, Executive Director/CEO  
201 Chicago Ave  
Minneapolis, MN 55415  
Tel: 612-928-6100

## Editorial Office

Patricia K. Baskin, MS, Executive Editor  
Kathleen M. Pieper, Managing Editor, *Neurology*  
Sandra A. Moriarity, Senior Production Editor  
Sharon L. Quimby, Managing Editor, *Neurology*® *Clinical Practice*  
Morgan S. Sorenson, Managing Editor, *Neurology: Neuroimmunology & Neuroinflammation*  
Lee Ann Kleffman, Managing Editor, *Neurology*® *Genetics*  
Cynthia S. Abair, MA, Senior Graphics Editor  
Robert J. Witherow, Senior Editorial Associate  
Karen Skaja, Editorial Associate  
Andrea R. Wagner, Editorial Assistant  
Kaitlyn Aman Ramm, Editorial Assistant  
Kristen Swendsrud, Editorial Assistant

## Publisher

Wolters Kluwer  
Baltimore, MD

## Publishing Staff

Kim Jansen, Executive Publisher  
Alexandra Lazerow, Production Team Leader, AAN Journals  
Kayla Bawroski, Production Editor  
Nick Strickland, Production Associate

# Neurology® Neuroimmunology & Neuroinflammation

Neurology.org/nn • Online ISSN: 2332-7812

Volume 2, Number 5, October 2015



## EDITOR

### *Neurology: Neuroimmunology & Neuroinflammation*

Josep O. Dalmau, MD, PhD  
ICREA-University of Barcelona  
Barcelona, Spain  
University of Pennsylvania  
Philadelphia, PA

## EDITOR-IN-CHIEF

### *Neurology*

Robert A. Gross, MD, PhD, FAAN  
Professor of Neurology and of Pharmacology and Physiology  
Strong Epilepsy Center  
University of Rochester Medical Center  
Rochester, NY

## DEPUTY EDITORS

### *Neurology*

David S. Knopman, MD, FAAN  
Professor of Neurology  
Department of Neurology  
College of Medicine  
Mayo Clinic  
Rochester, MN

### *Neurology: Neuroimmunology & Neuroinflammation*

Scott S. Zamvil, MD, PhD, FAAN  
Department of Neurology  
University of California, San Francisco  
San Francisco, CA

## ASSOCIATE EDITORS

### *Neurology: Neuroimmunology & Neuroinflammation*

Michael Heneka, MD  
University of Bonn  
Bonn, Germany  
Specialties: Neurodegeneration, stroke

Robyn S. Klein, MD, PhD  
Washington University School of Medicine  
St. Louis, MO  
Specialty: Infectious disease

Friedmann Paul, MD  
Charite University Hospital  
Berlin, Germany  
Specialties: Multiple sclerosis, neuromyelitis optica, neuroimaging  
(magnetic resonance imaging, optical coherence tomography)

Lan Zhou, MD, PhD  
Mount Sinai School of Medicine  
New York, NY  
Specialty: Neuromuscular

## EDITORIAL BOARD

Rohit Aggarwal, MD, MS, University of Pittsburgh, Pittsburgh, PA  
Sergio E. Baranzini, PhD, University of San Francisco, San Francisco, CA  
Susanne Benseler, MD, Calgary, Alberta, Canada  
Jeffrey L. Bennett, MD, PhD, FAAN, University of Colorado, Aurora, CO  
Séverine Boillée, PhD, Hôpital de la Salpêtrière, Paris, France  
Matthijs C. Brouwer, MD, PhD, Academic Medical Center, Amsterdam, The Netherlands  
David B. Clifford, MD, FAAN, Washington University School of Medicine, St. Louis, MO  
Russell C. Dale, MBChB, MRCP, PhD, University of Sydney, Sydney, Australia  
Oscar H. Del Brutto, MD, FAAN, Clinica, Kennedy, Guayaquil, Ecuador  
Thomas G. Forsthuber, MD, University of Texas, San Antonio, TX  
Joseph El Khoury, MD, Massachusetts General Hospital, Boston, MA  
Markus P. Kummer, PhD, University of Bonn, Bonn, Germany  
Jun Li, MD, PhD, Vanderbilt University, Nashville, TN  
Lin Mei, MD, PhD, Georgia Regents University, Augusta, Georgia  
Christopher Power, MD, University of Alberta, Alberta, Canada  
Richard M. Ransohoff, MD, Biogen Idec, Boston, MA  
Christine Stadelmann, MD, University of Göttingen, Göttingen, Germany  
Israel Steiner, MD, Rabin Medical Center, Petach Tikva, Israel  
Lawrence Steinman, MD, FAAN, Stanford University, Stanford, CA  
Jesper Tegnér, PhD, MSc, Karolinska University Hospital, Stockholm, Sweden  
Maarten J. Titulaer, MD, PhD, Erasmus Medical Center, Rotterdam, The Netherlands  
Hugh J. Willison, MBBS, PhD, University of Glasgow, Glasgow, UK

## LEVEL OF EVIDENCE REVIEW TEAM

Melissa J. Armstrong, MD, Baltimore, MD  
Richard L. Barbano, MD, PhD, FAAN, Rochester, NY  
Richard M. Dubinsky, MD, MPH, FAAN, Kansas City, KS  
Jeffrey J. Fletcher, MD, MSc, Ann Arbor, MI  
Gary M. Franklin, MD, MPH, FAAN, Seattle, WA  
David S. Gloss II, MD, MPH&TM, Danville, PA  
John J. Halperin, MD, FAAN, Summit, NJ  
Jason Lazarou, MSc, MD, Toronto, Ontario, Canada  
Steven R. Messé, MD, FAAN, Philadelphia, PA  
Pushpa Narayanaswami, MBBS, DM, FAAN, Boston, MA  
Alex Rae-Grant, MD, Cleveland, OH

# Neurology® Neuroimmunology & Neuroinflammation

Neurology.org/nn • Online ISSN: 2332-7812  
Volume 2, Number 5, October 2015



## Publication Information

**Neurology®** is a registered trademark of the American Academy of Neurology (registration valid in the United States).

**Neurology® Neuroimmunology & Neuroinflammation** (eISSN 2332-7812) is an open access journal published online for the American Academy of Neurology, 201 Chicago Ave, Minneapolis, MN 55415, by Wolters Kluwer, 16522 Hunters Green Parkway, Hagerstown, MD 21740-2116. Business offices are located at Two Commerce Square, 2001 Market St., Philadelphia, PA 19103. Production offices are located at 351 West Camden Street, Baltimore, MD 21201-2436. © 2015 American Academy of Neurology.

**Neurology® Neuroimmunology & Neuroinflammation** is an official journal of the American Academy of Neurology.  
Journal website: Neurology.org/nn, AAN website: AAN.com

**Copyright and Permission Information:** Please go to the journal website ([www.neurology.org/nn](http://www.neurology.org/nn)) and click the “©Request Permissions” icon for the relevant article. Alternatively, send an email to [healthpermissions@wolterskluwer.com](mailto:healthpermissions@wolterskluwer.com). General information about permissions can be found here: [http://www.lww.com/webapp/wcs/stores/servlet/content\\_resources\\_permissions-journals\\_11851\\_-1\\_12551](http://www.lww.com/webapp/wcs/stores/servlet/content_resources_permissions-journals_11851_-1_12551).

**Translation Rights & Licensing queries:** Silvia Serra, Global Director of Rights, Licensing, Permissions, Wolters Kluwer Citi Building, 25 Canada Square, E14 5LB, England, UK. tel: +44-(0)203-1976643; [translationrights@wolterskluwer.com](mailto:translationrights@wolterskluwer.com).

**Disclaimer:** Opinions expressed by the authors and advertisers are not necessarily those of the American Academy of Neurology, its affiliates, or of the Publisher. The American Academy of Neurology, its affiliates, and the Publisher disclaim any liability to any party for the accuracy, completeness, efficacy, or availability of the material contained in this publication (including drug dosages) or for any damages arising out of the use or non-use of any of the material contained in this publication.

**Advertising Sales Representatives:** Wolters Kluwer, 333 Seventh Avenue, New York, NY 10001. Contacts: Eileen Henry, tel: 732-778-2261, fax: 973-215-2485, [eileen.henry@wolterskluwer.com](mailto:eileen.henry@wolterskluwer.com) and Valentin Torres, tel: 646-674-6354, cell: 646-628-4530, fax: 917-677-8420, [valentin.torres@wolterskluwer.com](mailto:valentin.torres@wolterskluwer.com). In Europe: Avia Potashnik, Wolters Kluwer, tel: +44 207 981 0722; +44 7919 397 933 or e-mail: [avia.potashnik@wolterskluwer.com](mailto:avia.potashnik@wolterskluwer.com).

**Careers & Events:** Ryan Magee, Wolters Kluwer, Two Commerce Square, 2001 Market Street, Philadelphia, PA 19103, tel: 215-521-8804, fax: 215-521-8801; [ryan.magee@wolterskluwer.com](mailto:ryan.magee@wolterskluwer.com).

**Reprints:** Ben Beit-Zuri, Wolters Kluwer, 333 Seventh Avenue, New York, NY 10001, [ben.beit-zuri@wolterskluwer.com](mailto:ben.beit-zuri@wolterskluwer.com), [reprintsolutions@wolterskluwer.com](mailto:reprintsolutions@wolterskluwer.com).

**Special projects:** US & Canada: Alan Moore, Wolters Kluwer, Two Commerce Square, 2001 Market Street, Philadelphia, PA 19103, tel: 215-521-8638, [alan.moore@wolterskluwer.com](mailto:alan.moore@wolterskluwer.com). International: Silvia Serra, Global Director of Rights, Licensing, Permissions, Wolters Kluwer Citi Building, 25 Canada Square, E14 5LB, England, UK. tel: +44-(0)203-1976643; [silvia.serra@wolterskluwer.com](mailto:silvia.serra@wolterskluwer.com).

# Neurology<sup>®</sup> Neuroimmunology & Neuroinflammation

2 (5)

*Neurol Neuroimmunol Neuroinflamm* 2015;2;

**This information is current as of January 1, 2015**

**Updated Information & Services**

including high resolution figures, can be found at:  
<http://nn.neurology.org/content/2/5.full.html>

**Permissions & Licensing**

Information about reproducing this article in parts (figures, tables) or in its entirety can be found online at:  
<http://nn.neurology.org/misc/about.xhtml#permissions>

**Reprints**

Information about ordering reprints can be found online:  
<http://nn.neurology.org/misc/addir.xhtml#reprintsus>

*Neurol Neuroimmunol Neuroinflamm* is an official journal of the American Academy of Neurology. Published since April 2014, it is an open-access, online-only, continuous publication journal. Copyright . All rights reserved. Online ISSN: 2332-7812.

