



Cover image: Brain tree.

#### Editor

Matthew C Kiernan (Australia)

#### Deputy Editors

Karen L Furie (USA)  
Satoshi Kuvabara (Japan)  
Lawrence Wong (China)

#### Associate Editors

Alan Carson (UK)  
Nick Ward (UK)  
Peter Warnke (USA)

#### Commissioning Editor

Martin Turner (UK)

#### Web Editors

Clare Caldwell  
Arun Krishnan  
Cindy Lin  
Steve Vucic

#### Podcast Editor

Elizabeth Highton-Williamson

#### JNNP Blog Editors

Jose Manuel Matamala  
Thanuja Dharmadasa

#### JNNP Facebook Editors

Cindy Lin  
William Huynh

#### Editors, Practical Neurology

Geraint Fuller (UK)  
Phil Smith (UK)

#### Editorial Office

*Journal of Neurology Neurosurgery & Psychiatry*

BMJ Publishing Group Ltd

BMA House

Tavistock Square

London WC1H 9JR, UK

E: [jnnp@bmj.com](mailto:jnnp@bmj.com)

Twitter: @JNNP\_BMJ

ISSN: 0022-3050 (print)

ISSN: 1468-330X (online)

Impact factor: 7.144

**Disclaimer:** JNNP is published by BMJ Publishing Group Ltd, a wholly owned subsidiary of the British Medical Association. The BMJ Group grants editorial freedom to the Editor of JNNP. JNNP follows guidelines on editorial independence produced by the World Association of Medical Editors and the code on good publication practice of the Committee on Publication Ethics.

JNNP is intended for medical professionals and is provided without warranty, express or implied. Statements in the journal are the responsibility of their authors and advertisers and not authors' institutions, the BMJ Publishing Group or the BMA unless otherwise specified or determined by law. Acceptance of advertising does not imply endorsement.

To the fullest extent permitted by law, the BMJ Publishing Group shall not be liable for any loss, injury or damage resulting from the use of JNNP or any information in it whether based on contract, tort or otherwise. Readers are advised to verify any information they choose to rely on.

**Copyright:** © 2019 BMJ Publishing Group. All rights reserved; no part of this publication may be reproduced, stored in a retrieval system, or transmitted in any form or by any means, electronic, mechanical, photocopying, recording, or otherwise without prior permission.

JNNP is published by BMJ Publishing Group Ltd, typeset by Exeter Premedia Services Private Ltd, Chennai, India and printed in the UK on acid-free paper from sustainable forests.

*Journal of Neurology Neurosurgery & Psychiatry* (ISSN No: 0022-3050) is published monthly by BMJ Publishing Group and is distributed in the USA by Air Business Ltd. Periodicals postage paid at Jamaica NY 11431 POSTMASTER: send address changes to *Journal of Neurology Neurosurgery & Psychiatry*, Air Business Ltd, c/o Worldnet Shipping Inc., 156-15, 146th Avenue, 2nd Floor, Jamaica, NY 11434, USA

## Contents

Volume 90 Issue 2 | JNNP February 2019

### Editorial commentaries

- 121** Atypical chronic inflammatory demyelinating polyneuropathies  
*S Kuvabara, S Misawa, M Mori*

- 122** The reunification of amyotrophic lateral sclerosis  
*M R Turner*

- 124** New teased-fibre definitions represent specific mechanisms of neuropathy  
*H Koike, M Matsuno, G Sobue*

### Neuromuscular

- 125** Atypical CIDP: diagnostic criteria, progression and treatment response. Data from the Italian CIDP Database

*P E Doneddu, D Cocito, F Manganelli, R Fazio, C Briani, M Filosto, L Benedetti, A Mazzeo, G A Marfia, A Cortese, B Fierro, S Jann, E Beghi, A M Clerici, M Carpo, A Schenone, M Luigetti, G Lauria, G Antonini, T Rosso, G Siciliano, G Cavaletti, G Liberatore, L Santoro, E Peci, S Tronci, M Ruiz, S Cotti Piccinelli, A Toscano, G Mataluni, L Piccolo, G Cosentino, M Sabatelli, E Nobile-Orazio, on behalf of the Italian CIDP Database study group*

- 133** Proposal of new clinical diagnostic criteria for POEMS syndrome  
*T Suichi, S Misawa, Y Sato, M Beppu, E Sakaida, Y Sekiguchi, K Shibuya, K Watanabe, H Amino, S Kuvabara*

- 138** Expanded teased nerve fibre pathological conditions in disease association  
*M Xu, M Pinto, C Sun, J K Engelstad, P James Dyck, P J Dyck, C J Klein*

### General neurology

- 141** Cognitive and behavioural changes in PLS and PMA: challenging the concept of restricted phenotypes  
*B S de Vries, L M M Rustemeijer, L A Bakker, C D Schröder, J H Veldink, L H van den Berg, T C W Nijboer, M A van Es*

- 148** Lifetime risk of common neurological diseases in the elderly population  
*S Licher, S K L Darveesh, F J Wolters, L Fani, A Heshmatollah, U Mutlu, P J Koudstaal, J Heeringa, M J G Leening, M K Ikeram, M A Ikeram*

### Neurodegeneration

- 157** Neurofilament light chain in serum for the diagnosis of amyotrophic lateral sclerosis  
*F Verde, P Steinacker, J H Weishaupt, J Kassubeck, P Oeckl, S Halbgebauer, H Tumani, C A F von Arnim, J Dorst, E Feneberg, B Mayer, H-P Müller, M Gorges, A Rosenbohm, A E Volk, V Silani, A C Ludolph, M Otto*

- 165** Parkinson's disease: evolution of cognitive impairment and CSF Aβ<sub>1-42</sub> profiles in a prospective longitudinal study  
*S Lerche, I Wurster, B Röben, G Machetanz, M Zimmermann, F Bernhard, E Stransky, C Deuschle, C Schulte, O Hansson, H Zetterberg, T Gasser, D Berg, W Maetzler, K Brockmann*

- 171** Cognitive decline in Parkinson's disease: the impact of the motor phenotype on cognition  
*J Wojala, I A Heber, P Neuser, J Heller, E Kalbe, S P Rehberg, A Storch, K Linse, C Schneider, S Gräber, D Berg, J Dams, M Balzer-Geldsetzer, R Hilker-Roggenhoff, C Oberschmidt, S Baudrexel, K Witt, N Schmidt, G Deuschl, B Mollenhauer, C Trenkwalder, I Liepelt-Scarfone, A Spotke, S Roeske, U Wüllner, H-U Wittchen, O Riedel, R Dodel, J B Schulz, K Reetz*

- 180** Elevated YKL-40 and low sAPPβ:YKL-40 ratio in antemortem cerebrospinal fluid of patients with pathologically confirmed FTL  
*D Alcolea, D J Irwin, I Illán-Gala, L Muñoz, J Clarimón, C T McMillan, J Fortea, R Blesa, E B Lee, J Q Trojanowski, M Grossman, A Lleó*

**MORE CONTENTS ►**



EDITOR'S CHOICE

This article has been chosen by the Editor to be of special interest or importance and is freely available online.



PATIENT'S CHOICE

This article has been chosen in conjunction with the BMJ Patient Forum to be of particular interest to patients and is freely available online.



OPEN ACCESS

This article has been made freely available online under the BMJ Journals open access scheme. See <http://authors.bmj.com/open-access/>



The article is part of the Impact Commentaries series



Member since 2008  
JM00010

This journal is a member of and subscribes to the principles of the Committee on Publication Ethics  
<http://publicationethics.org/>



When you have finished with this magazine please recycle it.



The online version of this article contains multiple choice questions hosted on BMJ Learning.

## Neurogenetics

- 187** X linked Charcot-Marie-Tooth disease and multiple sclerosis: emerging evidence for an association

*G Koutsis, M Breza, G Velonakis, J Tzartos, D Kasselimis, C Katanou, E Karavasilis, D Tzanetakos, M Anagnostouli, E Andreadou, M-E Evangelopoulos, C Kilidireas, C Potagas, M Panas, G Karadima*

- 195** Genetic profile and onset features of 1005 patients with Charcot-Marie-Tooth disease in Japan

*A Yoshimura, J-H Yuan, A Hashiguchi, M Ando, Y Higuchi, T Nakamura, Y Okamoto, M Nakagawa, H Takashima*

- 203** Genetic risk factors for modulation of age at onset in Machado-Joseph disease/spinocerebellar ataxia type 3: a systematic review and meta-analysis

*E P de Mattos, M Kolbe Muszkopf, V Bielefeldt Leotti, M L Saraiva-Pereira, L B Jardim*

- 211** Spinal cord involvement in adult-onset metabolic and genetic diseases

*C Marelli, E Salsano, L S Politi, P Labauge*

## Multiple sclerosis

- 219** Structural network disruption markers explain disability in multiple sclerosis

*T Charalambous, C Tur, F Prados, B Kanber, D T Chard, S Ourselin, J D Clayden, C A M Gandini Wheeler-Kingshott, A J Thompson, A T Toosy*

## Movement disorders

- 227** Unravelling of the paroxysmal dyskinesias

*R Erro, K P Bhatia*

## Occasional essay

- 235** Adolescent brain development and gender: predictors of future reading habits

*C E Caldwell*

## PostScript

- 238** Letters