



*A proud earthly tree with little bunches, c.1919. Beyond Reason. Art and Psychosis, 2006.*

#### Editor

Martin Rossor (UK)

#### Deputy Editor

Michael Hanna (UK)

#### Associate Editors

Simon Fleming (UK)

Didier Leys (France)

Philip Scheltens (The Netherlands)

Peter Warnke (USA)

#### Web Editor

Rustam Al-Shahi (UK)

#### Editor, Practical Neurology

Charles Warlow (UK)

#### Editorial board

See page iv

#### Editorial Office

Journal of Neurology Neurosurgery & Psychiatry

BMJ Publishing Group Ltd

BMA House

Tavistock Square

London WC1H 9JR, UK

T: +44 (0)20 7874 7024

F: +44 (0)20 7383 6668

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

ISSN: 0022-3050 (print)

ISSN: 1468-330X (online)

Impact factor 3.63

**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:** © 2008 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 The Charlesworth Group, and printed in the UK on acid-free paper from sustainable forests by Cambrian Printers Limited, Aberystwyth, UK.

Periodicals postage paid, Rahway, NJ. Postmaster: send address changes to: Journal of Neurology Neurosurgery & Psychiatry, c/o Mercury Airfreight International Ltd, 365 Blair Road, Avenel, NJ 07001, USA.

### Editorial commentaries

- 111** Avoiding deep brain stimulation failures in Tourette syndrome  
*M S Okun, H H Fernandez, K D Foote, T K Murphy, W K Goodman*
- 112** When the MRI scan suggests multiple sclerosis but the symptoms do not  
*J Chataway*
- 113** Small neurons may be preferentially affected in ganglionopathy  
*H Koike, G Sobue*



### Cochrane neurological network corner

- 114** Multidisciplinary rehabilitation for adults with multiple sclerosis  
*F Khan, L Turner-Stokes, L Ng, T Kilpatrick*

### Review

- 115** Recommendations on diagnostic strategies for chronic inflammatory demyelinating polyradiculoneuropathy  
*The French CIDP Study Group*

### Papers

- 119** Prolonged activation EEG differentiates dementia with and without delirium in frail elderly patients  
*C Thomas, U Hestermann, S Walther, U Pfueller, M Hack, P Oster, C Mundt, M Weisbrod*
- 126** Factors associated with survival probability in autopsy-proven frontotemporal lobar degeneration  
*S X Xie, M S Forman, J Farmer, P Moore, Y Wang, X Wang, C M Clark, H B Coslett, A Chatterjee, S E Arnold, H Rosen, J H T Karlawish, V M Van Deerlin, V M-Y Lee, J Q Trojanowski, M Grossman*
- 130** Longitudinal analysis of regional grey matter loss in Huntington disease: effects of the length of the expanded CAG repeat  
*H H Ruocco, L Bonilha, L M Li, I Lopes-Cendes, F Cendes*
- 136** Deep brain stimulation in 18 patients with severe Gilles de la Tourette syndrome refractory to treatment: the surgery and stimulation  
*D Servello, M Porta, M Sassi, A Brambilla, M M Robertson*
- 143** Cognitive complaints in the early phase after stroke are not indicative of cognitive impairment  
*A Duits, T Munnecom, C van Heugten, R J van Oostenbrugge*
- 147** *Chlamydia pneumoniae* seropositivity in aetiological subtypes of brain infarction and carotid atherosclerosis: a case-control study

*S Alamowitch, J Labreuche, P-J Touboul, F Eb, P Amarenco, for the GENIC Investigators*

- 152** A longitudinal study of serum 25-hydroxyvitamin D and intact parathyroid hormone levels indicate the importance of vitamin D and calcium homeostasis regulation in multiple sclerosis  
*M Soilu-Hänninen, M Laaksonen, I Laitinen, J-P Erälinna, E-M Lilius, I Mononen*
- 158** A genome scan in a single pedigree with a high prevalence of multiple sclerosis  
*D A Dymnt, M Z Cader, B M Herrera, S V Ramagopalan, S M Orton, M Chao, C J Willer, A D Sadovnick, N Risch, G C Ebers*
- 163** Non-length dependent small fibre neuropathy/ganglionopathy  
*K C Gorson, D N Herrmann, R Thiagarajan, T H Brannagan, R L Chin, L J Kinsella, A H Ropper*
- 170** Intraoperative brainstem auditory evoked potential pattern and perioperative vasoactive treatment for hearing preservation in vestibular schwannoma surgery  
*B Bischoff, J Romstöck, R Fahlbusch, M Buchfelder, C Strauss*

### Short reports

- 176** A comparison of unawareness in frontotemporal dementia and Alzheimer's disease  
*E Salmon, D Perani, F Collette, D Feyers, E Kalbe, V Holthoff, S Sorbi, K Herholz*
- 180** Variant Creutzfeldt-Jacob disease: the second case in Portugal and in the same geographical region  
*Á Machado, H Soares, H Antunes, Z Magalhães, C Ferreira, I Baldeiras, M H Ribeiro, I Santana, J Ramalheira, L Castro, S Carpenter*
- 183** Frequency of *GCH1* deletions in Dopa-responsive dystonia  
*B Zirn, D Steinberger, C Troidl, K Brockmann, M von der Hagen, C Feiner, L Henke, U Müller*
- 187** Irregularity distinguishes limb tremor in cervical dystonia from essential tremor  
*A G Shaikh, H A Jinnah, R M Tripp, L M Optican, S Ramat, F A Lenz, D S Zee*

[MORE CONTENTS ►](#)



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



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



Articles carrying the Unlocked Logo are freely available online under the BMJ Journals unlocked scheme. See <http://jnnp.bmj.com/info/unlocked.dtl>



[www.publicationethics.org.uk](http://www.publicationethics.org.uk)

This journal is a member of and subscribes to the principles of the Committee on Publication Ethics

- 190** Executive functions are impaired in patients with Parkinson's disease with visual hallucinations  
*J Barnes, L Boubert*
- 193** Emotional facial palsy following striato-capsular infarction  
*L Michel, P Derkinderen, D Laplaud, B Dumas-Duport, E Auffray-Calvier, T Lebouvier*
- 195** Unexpected multiple sclerosis: follow-up of 30 patients with magnetic resonance imaging and clinical conversion profile  
*C Lebrun, C Bensa, M Debouverie, J De Seze, S Wiertliewski, B Brochet, P Clavelou, D Brassat, P Labauge, E Rouillet, on behalf of CFSEP*
- 199** Fatigue is associated with cerebral white matter hyperintensities in patients with systemic lupus erythematosus  
*E Harboe, O J Greve, M Beyer, L G Gøransson, A B Tjensvoll, S Maroni, R Omdal*
- 202** Hypothermia in VGKC antibody-associated limbic encephalitis  
*S Jacob, S R Irani, Y A Rajabally, A Grubneac, R J Walters, M Yazaki, L Clover, A Vincent*
- 205** Autosomal-dominant distal myopathy with a *myotilin* S55F mutation: sorting out the phenotype  
*J Berciano, E Gallardo, R Domínguez-Perles, E Gallardo, A García, R García-Barredo, O Combarros, J Infante, I Illa*
- 209** Causes of death in pedigrees with the 3243A>G mutation in mitochondrial DNA  
*K Majamaa-Voltti, J Turkka, M-L Kortelainen, H Huikuri, K Majamaa*
- 212** Effect of daily repetitive transcranial magnetic stimulation for treatment of tinnitus: comparison of different stimulus frequencies  
*E M Khedr, J C Rothwell, M A Ahmed, A El-Atar*
- 216** Serial yes/no reliability after traumatic brain injury: implications regarding the operational criteria for emergence from the minimally conscious state  
*R Nakase-Richardson, S A Yablon, M Sherer, C C Evans, T G Nick*
- 219** Neurological evaluation of patients with Gaucher disease diagnosed as type 1  
*J L Capablo, A Saenz de Cabezón, J Fraile, P Alfonso, M Pocovi, P Giraldo, on behalf of the Spanish Group on Gaucher Disease*
- 224** A small deep infarct revealing leukoencephalopathy, calcifications and cysts in an adult patient  
*I Wargon, M-C Lacour, D Adams, C Denier*
- 225** Measuring the effect of amantadine in chronic anoxic minimally conscious state  
*C Schnakers, R Hustinx, G Vandewalle, S Majerus, G Moonen, M Boly, A Vanhaudenhuyse, S Laureys*
- 227** Acute necrotizing encephalopathy: a relapsing case in a European adult  
*A Fasano, G F Natoli, A Cianfoni, D Ferraro, G Loria, A R Bentivoglio, S Servidei*
- 229** A novel mutation (c.64 65delGGinsAACC [p.G21fsX66]) in the *GTP cyclohydrolase 1* gene that causes Segawa disease  
*M von Mering, H Gabriel, T Opladen, G F Hoffmann, A Storch*
- 229** Iatrogenic Creutzfeldt–Jakob disease 22 years after human growth hormone therapy: clinical and radiological features  
*M Furtner, E Gelpi, S Kiechl, M Knoflach, A Zangerl, T Gotwald, J Willeit, H Maier, T Ströbel, U Unterberger, H Budka*
- 231** Skin reactions after intramuscular injection of Botulinum toxin A: a rare side effect  
*N Brueggemann, L Doegnitz, L Harms, A Moser, J M Hagenah*
- 232** Parkinson's disease with Onuf's nucleus involvement mimicking multiple system atrophy  
*S S O'Sullivan, J L Holton, L A Massey, D R Williams, T Revesz, A J Lees*
- 234** Complete deletion of the aprataxin gene: ataxia with oculomotor apraxia type 1 with severe phenotype and cognitive deficit  
*G Yoon, R Westmacott, L MacMillan, N Quercia, P Koutsou, A Georghiou, K Christodoulou, B Banwell*
- 
- Historical note**
- 129** A note on X linked adrenoleukodystrophy (Addison–Schilder syndrome)  
*J M S Pearce*
- 
- Neurological pictures**
- 198** Embolic spread of tuberculomas in the brain in multidrug resistant tubercular meningitis  
*A Kohli, R Kapoor*
- 222** Medulla compression caused by vertebral artery dolichoectasia  
*J H Roh, Y S Koo, S-H Jang, M H Park*
- 236** Identifying aortic plaque inflammation as a potential cause of stroke  
*R R Moustafa, D Izquierdo, P L Weissberg, J-C Baron, E A Warburton*

## PostScript

- 223** Assessment of visual pursuit in post-comatose states: use a mirror  
*A Vanhaudenhuyse, C Schnakers, S Brédart, S Laureys*