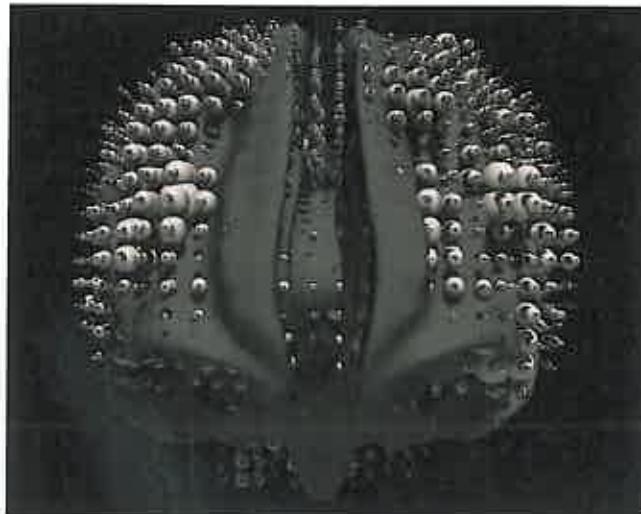


BRAIN

A JOURNAL OF NEUROLOGY



Cover image: Rendering of the effect of focal brain damage on gaze direction, derived from 1172 patients with acute ischaemic stroke lesions. The diameter and orientation of each glyph is proportional to the median magnitude and median degree of deviation. The green and purple 'pupils' represent deviation to the left and right of the midline, respectively. From Xu et al. High-dimensional therapeutic inference in the focally damaged human brain. Pp. 48–54.

Contents

Editorial

Editorial

D. M. Kullmann

1

Scientific Commentaries

The rise of a new associationist school for lesion-symptom mapping

M. Thiebaut de Schotten and C. Foulon

2

Holding down the pain

A. H. Dickenson

5

Early nucleus basalis of Meynert degeneration predicts cognitive decline in Parkinson's disease

J. P. Gratwick and T. Foltyne

7

Spread of tau deposits: can we trust *in vivo* findings?

K. Herholz

10

Review article

Movement disorders with neuronal antibodies: syndromic approach, genetic parallels and pathophysiology

B. Balint, A. Vincent, H.-M. Meinck, S. R. Irani and K. P. Bhatia

13

Update

The cerebellum in Alzheimer's disease: evaluating its role in cognitive decline

H. I. L. Jacobs, D. A. Hopkins, H. C. Mayrhofer, E. Bruner, F. W. van Leeuwen, W. Raaijmakers and J. D. Schmahmann

37

Reports**High-dimensional therapeutic inference in the focally damaged human brain**

T. Xu, H. Rolf Jäger, M. Husain, G. Rees and P. Nachev

48

Heterozygous SSBP1 start loss mutation co-segregates with hearing loss and the m.1555A>G mtDNA variant in a large multigenerational family

P. J. Kulik, A. Gomez-Duran, P. A. Gammie, C. Garone, M. Minczuk, Z. Golder, J. Wilson, J. Montoya, S. Häkli, M. Kärppä, R. Horvath, K. Majamaa and P. F. Chinnery

55

Original Articles**Mutations affecting glycinergic neurotransmission in hyperekplexia increase pain sensitivity**

P. H. Vuilleumier, R. Fritzsche, J. Schliessbach, B. Schmitt, L. Arendt-Nielsen, H. U. Zeilhofer and M. Curatolo

63

Plasma oxysterols: biomarkers for diagnosis and treatment in spastic paraparesis type 5

C. Marelli, F. Lamari, D. Rainteau, A. Lafourcade, G. Banneau, L. Humbert, M.-L. Monin, E. Petit, R. Debs, G. Castelnovo, E. Ollagnon, J. Lavie, J. Pilliod, I. Coupry, P. J. Babin, C. Guissart, I. Benyounes, U. Ullmann, G. Lesca, C. Thauvin-Robinet, P. Labauge, S. Odent, C. Ewenczyk, C. Wolf, G. Stevanin, D. Hajage, A. Durr, C. Goizet and F. Mochel

72

Clemastine rescues myelination defects and promotes functional recovery in hypoxic brain injury

B. A. C. Cree, J. Niu, K. K. Hoi, C. Zhao, S. D. Caganap, R. G. Henry, D. Q. Dao, D. R. Zollinger, F. Mei, Y.-A. A. Shen, R. J. M. Franklin, E. M. Ullian, L. Xiao, J. R. Chan and S. P. J. Fancy

85

Macrophage enzyme and reduced inflammation drive brain correction of mucopolysaccharidosis IIIB by stem cell gene therapy

R. J. Holley, S. M. Ellison, D. Fil, C. O'Leary, J. McDermott, N. Senthivel, A. W. W. Langford-Smith, F. L. Wilkinson, Z. D'Souza, H. Parker, A. Liao, S. Rowston, H. F. E. Gleitz, S.-H. Kan, P. I. Dickson and B. W. Bigger

99

A fingerprint of the epileptogenic zone in human epilepsies

O. Grinenko, J. Li, J. C. Mosher, I. Z. Wang, J. C. Bulacio, J. Gonzalez-Martinez, D. Nair, I. Najm, R. M. Leahy and P. Chauvel

117

Oestrogen receptor β ligand acts on CD11c $^{+}$ cells to mediate protection in experimental autoimmune encephalomyelitis

R. Y. Kim, D. Mangu, A. S. Hoffman, R. Kovash, E. Jung, N. Itoh and R. Voskuhl

132

Altered caudate connectivity is associated with executive dysfunction after traumatic brain injury

S. De Simoni, P. O. Jenkins, N. J. Bourke, J. J. Fleminger, P. J. Hellyer, A. E. Jolly, M. C. Patel, J. H. Cole, R. Leech and D. J. Sharp

148

In vivo cholinergic basal forebrain atrophy predicts cognitive decline in de novo Parkinson's disease

N. J. Ray, S. Bradburn, C. Murgatroyd, U. Toseeb, P. Mir, G. K. Kountouriotis, S. J. Teipel and M. J. Grothe

165

Neuronal inhibition and synaptic plasticity of basal ganglia neurons in Parkinson's disease

L. Milosevic, S. K. Kalia, M. Hodale, A. M. Lozano, A. Fasano, M. R. Popovic and W. D. Hutchison

177

Structural connectivity of right frontal hyperactive areas scales with stuttering severity

N. E. Neef, A. Anwander, C. Bütferring, C. Schmid-Samoa, A. D. Friederici, W. Paulus and M. Sommer

191

A human prefrontal-subthalamic circuit for cognitive control

R. Kelley, O. Flouty, E. B. Emmons, Y. Kim, J. Kingyon, J. R. Wessel, H. Oya, J. D. Greenlee and N. S. Narayanan

205

Two critical brain networks for generation and combination of remote associations

D. Bendetowicz, M. Urbanski, B. Garcin, C. Foulon, R. Levy, M.-L. Bréchemier, C. Rosso, M. Thiebaut de Schotten and E. Volle

217

Obligatory and facultative brain regions for voice-identity recognition

C. Roswandowitz, C. Kappes, H. Obrig and K. von Kriegstein

234

The cerebellar cognitive affective/Schmahmann syndrome scale

F. Hoche, X. Guell, M. G. Vangel, J. C. Sherman and J. D. Schmahmann

248

Widespread brain tau and its association with ageing, Braak stage and Alzheimer's dementia

V. J. Lowe, H. J. Wiste, M. L. Senjem, S. D. Welgand, T. M. Therneau, B. F. Boeve, K. A. Josephs, P. Fang, M. K. Pandey, M. E. Murray, K. Kantarci, D. T. Jones, P. Vemuri, J. Graff-Radford, C. G. Schwarz, M. M. Machulda, M. M. Mielke, R. O. Roberts, D. S. Knopman, R. C. Petersen and C. R. Jack, Jr

271

Asymmetry of post-mortem neuropathology in behavioural-variant frontotemporal dementia

D. J. Irwin, C. T. McMillan, S. X. Xie, K. Rascovsky, V. M. Van Deerlin, H. B. Coslett, R. Hamilton, G. Aguirre, E. B. Lee, V. M. Y. Lee, J. Q. Trojanowski and M. Grossman

288

Longitudinal structural and molecular neuroimaging in agrammatic primary progressive aphasia
K. A. Tetzloff, J. R. Duffy, H. M. Clark, E. A. Strand, M. M. Machulda, C. G. Schwarz, M. L. Senjem, R. I. Reid, A. J. Spychalla, N. Tosakulwong, V. J. Lowe,
C. R. Jack, Jr, K. A. Josephs and J. L. Whitwell
302

Dorsal column

Grey Matter

Dr Harrison Martland and the history of punch drunk syndrome
A. R. Changa, R. A. Vietrogoski and P. W. Carmel
318

Letters to the Editor

POLR3A variants in hereditary spastic paraplegia and ataxia

L. Gauquelin, M. Tétreault, I. Thiffault, E. Farrow, N. Miller, B. Yoo, E. Bareke, G. Yoon, O. Suchowersky, N. Dupré, M. Tarnopolsky, B. Brais, N. I. Wolf,
J. Majewski, G. A. Rouleau, Z. Gan-Or and G. Bernard
e1

Reply: POLR3A variants in hereditary spastic paraplegia and ataxia

M. Minnerop, D. Kurzwelly, T. W. Rattay, D. Timmann, H. Hengel, M. Synofzik, C. Stendel, R. Horvath, R. Schüle and A. Ramirez
e2

DGUOK recessive mutations in patients with CPEO, mitochondrial myopathy, parkinsonism and mtDNA deletions

L. Caporali, L. Bello, F. Tagliavini, C. La Morgia, A. Maresca, L. Di Vito, R. Liguori, M. L. Valentino, D. Cecchini, E. Pegoraro and V. Carelli
e3

Reply: DGUOK recessive mutations in patients with CPEO, mitochondrial myopathy, parkinsonism and mtDNA deletions

D. Ronchi, D. Piga, S. Lamberti, M. Sciacco, S. Corti, M. Moggio, N. Bresolin and G. Pietro Comi
e4

CLIPPERS, a possible symptomatic lymphohistiocytic immune reaction

G. Taieb and P. Labauge
e5

Reply: CLIPPERS, a possible symptomatic lymphohistiocytic immune reaction

W. O. Tobin, Y. Guo and B. M. Keegan
e6

Corrigendum

e7