Keep an eye on the prion – the spreading pathology of MSA

Reviewers: Roger A Barker, Professor of Clinical Neuroscience, University of Cambridge and Honorary Consultant Neurologist at Addenbrooke’s Hospital, Cambridge.

Over the last 50 years the origin of prion diseases has become more secure even if the mechanism by which the abnormal protein accumulation kills cells is still unresolved. These disorders include genetic conditions such as Gerstmann-Sträussler-Scheinker disease as well as sporadic diseases such as CJD. Of late there has been a great deal of interest in whether other, more common, neurodegenerative disorders of the CNS may have a similar basis – namely that the pathogenic protein that lies at the core of the condition can act in a prion like disease. This has been explored in most detail with tau and alpha synuclein and are the subject of two papers from the Prusiner lab that have just been published in PNAS.

These studies involve in vitro studies using human embryonic kidney cells along with the in vivo work involving the TgM83 +/- mice that express alpha synuclein (AS3T). In each case the cells/animals are “inoculated” with “prions” purified from the brains of patients dying with PSP, MSA or PD. In the first study they show that both MSA and PSP brains can infect cells in vitro, while PD brain extracts did not display such behaviour. This was then confirmed in the second paper with respect to MSA versus PD synuclein extracts in the mouse model. The authors therefore conclude that “alpha synuclein is the first human prion to be identified, to our knowledge, which again has been suggested by other studies (e.g. Pellaerts et al Nature 2015). Thus these papers are of great interest to those working in this field, although as far as I know, it has still not been shown that anyone has developed an alpha synucleinopathy from exposure to “infected” human tissue as has been the case for CJD.

This is a bold statement although fits with an emerging literature showing that alpha synuclein can spread from cell to cell and seed pathology in some instances. Thus finding adds to that existing literature, although does draw a distinction between the alpha synuclein “strains” found in PD and MSA which again has been suggested by other studies (e.g. Pellaerts et al Nature 2015). Thus these papers are of great interest to those working in this field, although as far as I know, it has still not been shown that anyone has developed an alpha synucleinopathy from exposure to “infected” human tissue as has been the case for CJD.

To list your event in this diary, email brief details to Rachael Hansford at Rachael@acmr.co.uk by 6th October, 2015.

Lucy Church, R.improprofessionals@rsm.ac.uk T. 0207 290 3928, www.rsm.ac.uk/liveaucinow

Modern Thinking in MS Management
7pm Friday 20 November - 16.40 Saturday 21 November 2015, The Palace Hotel, Manchester, UK www.modernthinkingsms.com

11th Essential Neuro MRI Course
Saturday 21st November, 2015, Liverpool Medical Institute, UK

One day intensive course in how to interpret MRI Brain & Spine6 Cat 1 CPD
Contact: Sam Pickup T. 0517 709 622 E. essentialcourses@hotmail.com

Consultant PD Masterclass – Sheffield
Module 1 - 2, 3rd & 4th June 2015
Module 2 - 26th November 2015
(Both modules must be attended)
www.participantsacademy.co.uk for further details.

Ketogenic Study Evening
35 November, 2015, Liverpool, UK
Jacqui McAleer, E. jmassociates1@rme.com

The 2nd British Symposium on the History of Neurology and Psychiatry
A commemoration of the centenary of the death of Sir William Gowers
November 25th, 2015, Institute of Neurology, London, UK
Programme and registration details: Liz Beddeman at www.hnps.co.uk2016

History of Neurology and Psychiatry in London
November 26th, 2015
Institute of Neurology, Queen Square, London, UK
Programme and registration details: Liz Beddeman at www.hnps.co.uk2016

23rd Annual Meeting of the European Charcot Foundation
26–28 November, 2015, Milan, Italy
www.charcot-ms.org

2015

September

Paediatric Oncology Solid Tumours Study Day
14 September, 2015, London, UK
www.royalmansfield.nhs.uk/childrens-paeds-chs
7pm Friday 20 November - 16.40 Saturday 21 November 2015, The Palace Hotel, Manchester, UK
www.modernthinkingsms.com

11th Essential Neuro MRI Course
Saturday 21st November, 2015, Liverpool Medical Institute, UK

One day intensive course in how to interpret MRI Brain & Spine6 Cat 1 CPD
Contact: Sam Pickup T. 0517 709 622 E. essentialcourses@hotmail.com

Consultant PD Masterclass – Sheffield
Module 1 - 2, 3rd & 4th June 2015
Module 2 - 26th November 2015
(Both modules must be attended)
www.participantsacademy.co.uk for further details.

Ketogenic Study Evening
35 November, 2015, Liverpool, UK
Jacqui McAleer, E. jmassociates1@rme.com

The 2nd British Symposium on the History of Neurology and Psychiatry
A commemoration of the centenary of the death of Sir William Gowers
November 25th, 2015, Institute of Neurology, London, UK
Programme and registration details: Liz Beddeman at www.hnps.co.uk2016

History of Neurology and Psychiatry in London
November 26th, 2015
Institute of Neurology, Queen Square, London, UK
Programme and registration details: Liz Beddeman at www.hnps.co.uk2016

23rd Annual Meeting of the European Charcot Foundation
26–28 November, 2015, Milan, Italy
www.charcot-ms.org

December

BNPA Neurology & Psychiatry SpRs
Teaching Weekend
11, 12, 13 December, 2015, Oxford, UK
E. admin@bpna.org.uk T. 01865 438 191, m. 07940 591096

History of Neurology and Psychiatry in London
November 26th, 2015
Institute of Neurology, Queen Square, London, UK
Programme and registration details: Liz Beddeman at www.hnps.co.uk2016

23rd Annual Meeting of the European Charcot Foundation
26–28 November, 2015, Milan, Italy
www.charcot-ms.org

2016

February

24 February, 2016, Cambridge, UK
www.partnershipsincare.co.uk or contact samantha.coburn-kett@partnershipsincare.co.uk