INC2016 and GBS100 Glasgow Programme

Tuesday, June 21, 2016

17.00 onwards Registration opens in Wolfson Medical School Building

18.30-20.00 Opening Reception (Drinks and canapés) Hunterian Museum

20.00 Dinner on your own in local restaurants (Glasgow West End)

Wednesday, June 22, 2016

08.00-09.00 INC meeting (open to all) Main discussion topics: INC call to order (Pieter van Doorn), PeriNomS, and Imagine

09.00-10.30 Kedrion sponsored Plenary Symposium 1: The nodal complex as a disease target in inflammatory neuropathies Chaired by Hans-Peter Hartung and Eduardo Nobile-Orazio

09.00-09.30 The Nodal Complex Peter Brophy

Peter Brophy is a Professor of Neuroscience in the Centre for Neuroregeneration, Edinburgh University. The goal of the Brophy laboratory is to understand how interactions between neurons and glia underpin rapid nerve impulse conduction in the vertebrate nervous system. Myelination of axons is a key event that stimulates the clustering of voltage gated sodium channels at nodes of Ranvier. The concentration of these sodium channels with associated proteins at nodes promotes rapid propagation of the action potential.

We are addressing three specific questions:
1. What determines the site of node of Ranvier assembly?
2. How are voltage-gated sodium channels clustered at the node?
3. What is the role of the axon initial segment?

Here I will focus on our recent studies on visualizing node assembly by live imaging.

09.30-10.00 Nodal proteins, assembly, adhesion and disruption Jerome Devaux

Dr Devaux is full time research scientist at the Center for Neurobiology and Neurophysiology of Marseille. His major research interests are 1) to uncover the mechanism regulating the axonal trafficking of ion channels in neurons using cellular and molecular approaches; 2) to determine the implication of autoantibodies in dysimmune neuropathies using animal models; and 3) to pose the strategic bases for improving axon conduction in human demyelinating pathologies. He will present the prevalence and pathogenic function of autoantibodies directed against cell adhesion molecules in patients with chronic inflammatory demyelinating polyneuropathy.

The node of Ranvier has long been suspected of being a target for antibodies but those targets remained elusive. Greater understanding of the protein components of the node of Ranvier, their interactions with one another, their roles in adhesion, structural integrity and impulse propagation as well as the discovery of antibodies making nodal protein epitopes targets for disruption has resulted in significant advances in our understanding of the pathophysiology of inflammatory diseases of the peripheral nerves.

10.00-10.30 Antibody effects at the node of Ranvier Rhona McGonigal

Dr Rhona McGonigal is a Research Associate in Neuroimmunology at the University of Glasgow whose interests are mechanisms mediating pathophysiology at the node of Ranvier in the variants of the autoimmune peripheral neuropathy, Guillain-Barré syndrome.

She will discuss complement-mediated antibody effects at the node of Ranvier in novel mouse models of Guillain-Barré syndrome and the consequent impact on axonal integrity. Loss of axonal integrity influences the severity of patient outcome and as such it is critical to understand the signals directing these events and identify new therapeutic targets.

10.30-11.00 Refreshment break

11.00-12.30 Short and Speed Poster Presentations Chaired by Thomas Harbo and Jong Kuk Kim

11.00-11.15 Detection of auto-antibodies against the paranodal protein Caspr in patients with painful inflammatory neuropathy Kathrin Doppler

11.15-11.30 The spectrum of neurological diseases associated with mycoplasma pneumoniae infection and anti-glycolipid antibodies Motoi Kuwahara

11.30-12.30 Speed Poster Presentations

SP1.1 IVIg treatment induces a transient expansion of antibody secreting cells and a switch from IgG1 to IgG2 in peripheral blood of Guillain-Barré syndrome patients Maarten Brem

SP1.2 Neuronal hydrogen peroxide promotes nerve terminal regeneration at the neuromuscular junction Elisa Duregotti
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**SP1.3** Effect of intravenous immunoglobulins on natural killer cells in peripheral blood of patients with chronic inflammatory demyelinating polyneuropathy
Maximilian Heininger

**SP1.4** Interleukin-10 promoter polymorphisms in patients with Guillain–Barré syndrome in Bangladesh
Zahirul Islam

**SP1.5** Intra-ganglionic delivery of nitric oxide donor induces mitochondrial damage and distal axonopathy
Abhijeet Joshi

**SP1.6** Tracking the interaction of neuritogenic T cells and myelin in an ex vivo model of the peripheral nervous system
Anne Mausberg

**SP1.7** ATP released by injured neurons activates Schwann cells
Samuele Negro

**SP1.8** Valuable objective markers for predicting anti-neurofascin 155 antibody status among CIDP patients
Hidenori Ogata

**SP1.9** The pathogenic effects of disialosyl antibodies are revealed by functional assays using differentiated human induced pluripotent stem cells
Simon Rinaldi

**SP1.10** A novel model of Miller Fisher syndrome to study nerve terminal regeneration
Umberto Rodella

**SP1.11** Complement-fixing anti-ganglioside antibodies in acute canine polyradiculoneuritis
Angie Rupp

**SP1.12** Myelin basic protein regulates pain from light touch in females but not males
Veronica Shubayev

**SP1.13** Diagnostic and prognostic value of anti-GD1b antibodies in immune-mediated neuropathies
Anne Tio

**SP1.14** IgG degrading enzyme of Streptococcus pyogenes improves a rabbit model of acute motor axonal neuropathy
Yuzhong Wang

**SP1.15** Transglutaminase 6 antibodies in gluten neuropathy
Panagiotis Zis

12.30-14.00
**Lunch break and poster viewing**
INC Board Committee Meeting

14.00-15.30
*Grifols sponsored Plenary Symposium 2: Clinical phenotyping of the inflammatory neuropathies*
Chaired by Yusuf Rajabally and Ingemar Merkies (followed by SPIN Award ceremony)

14.00-14.30
**CIDP phenotypes: similarities in the laboratory and clinic**
Luis Querol

Dr Luis Querol is a Spanish neurologist and scientist working at the Hospital de la Santa Creu i Sant Pau in Barcelona, at Prof Isabel Illa’s lab. He is interested in autoimmune neurological disorders in general but his main research focus is in the role of autoantibodies against proteins of the node of Ranvier in chronic inflammatory neuropathies. Dr Querol has described the presence of autoantibodies against contactin-1 in a subset of severe CIDP patients and the clinical features associated to anti-neurofascin 155 antibodies in CIDP. He has also collaborated in the description of the basic mechanisms leading to pathology in these patients. His talk will be focused on the implications of current diagnostic approaches to inflammatory neuropathy care, how the description of highly specific biomarkers may lead to more accurate and targeted therapeutic strategies and which would be the ideal research approach and cooperative environment to address the need of better biomarkers in inflammatory neuropathies.

14.30-15.00
**Clinical phenotyping of inflammatory neuropathies with electrophysiology**
Hessel Franssen

Hessel Franssen attended medical school from 1969-1977. He trained for Neurology and Clinical Neurophysiology at a large non-academic hospital in The Hague. For one and a half years he served the Dutch army as a first lieutenant medical officer, although he would have preferred serving the artillery. In 1986 he defended his PhD thesis on cortical regulation of eye movements at the Catholic University of Nijmegen, The Netherlands. Since 1987 he is Associate Professor of Neurology and Clinical Neurophysiology at the University Hospital Utrecht. He specialised in neuromuscular diseases and electromyography. For 7 years, he served as president of the Dutch Society for Clinical Neurophysiology and was one of the founders of the Dutch Clinical Neurophysiology Days, a yearly recurring teaching and scientific event.

He has written papers on criteria for demyelination and conduction block, temperature and nerve conduction, chronic idiopathic axonal neuropathy, multifocal motor neuropathy, and paraproteinaemic neuropathies.
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His research includes mechanisms of axonal degeneration, computer simulation of myelinated axon pathophysiology and nerve excitability studies.

His lecture for the 2016 PNS/INC meeting will deal with the background of electrophysiological criteria for inflammatory neuropathies, practical tips and tricks for their diagnosis and the relationship between symptoms and pathophysiology.

15.00-15.30 Paraproteinemias – certainties and uncertainties
Christopher Klein

Dr Christopher Klein is a Professor of Neurology at Mayo Clinic and has major interest in the pathogenesis of peripheral neuropathies. He has expertise in both inherited and paraneoplastic disorders and has published extensively on molecular causes of these disorders.

He will be discussing the plasma cell paraneoplastic disorder POEMS syndrome and the major new advancements in its treatment and understanding. Outlined is the success story in POEMS. Curative therapy is now possible for the majority of patients, whereas previously major neurological disability including death from neuromuscular failure were common. Emphasized is how collaborative efforts between peripheral nerve experts, hematologists, transplantation scientists, and immunologists made these advances possible.

Current algorithmic treatment recommendations will be reviewed as well as our understanding of the molecular pathogenesis of this disorder. The current deficiencies in our understanding of pathogenesis will be discussed and how still remaining is the need for better fundamental investigations into this complex pathology. Emphasized will be how improved understanding may expand insights into other MGUS neuropathies and potentially idiopathic demyelinating disorders.

15.30-18.30 INC meeting (open to all)
Main discussion topic: Clinical Trials

18.30-19.30 Poster viewing

20.00 Dinner on your own in local restaurants
(Glasgow West End)

Thursday, June 23, 2016

08.00-09.00 INC meeting (open to all)
Main discussion topic: IGOS and any other business

09.00-10.30 CSL Behring sponsored Plenary Symposium 3:
Therapeutic immunology of inflammatory neuropathies
Chaired by Ivo van Schaik and Hans Katzberg

09.00-09.30 Update on the immunology of CIDP, MMN and GBS
Helmar Lehmann

Helmar C. Lehmann, M.D., is a clinician-scientist at the Department of Neurology at the University of Cologne, Germany. He received clinical training at the Department of Neurology in Düsseldorf, Germany between 2003 and 2010. In 2004 and 2008-2009, Lehmann completed a postdoctoral research fellowship in the Neuromuscular Division, Department of Neurology, Johns Hopkins Hospital, Baltimore, USA. Lehmann is leading a research group that focuses on pathogenesis, diagnosis and treatment of peripheral neuropathies.

Over the last years significant progress has been achieved in understanding the pathological processes underlying immune mediated neuropathies. In his talk Lehmann will provide an overview about current knowledge in the immune pathogenesis of Guillain-Barré syndrome (GBS), multifocal motor neuropathy (MMN) and chronic inflammatory demyelinating polyneuropathy (CIDP).

09.30-10.00 Biological depletion therapies
Iain McInnes

Professor Iain B McInnes is Director of the Institute of Infection Immunity and Inflammation, Director of the Scottish MRC Clinical Pharmacology and Pathology Clinical PhD Training Programme, Chief Investigator for the Scottish Early RA Cohort (SERA) and the related SMS-IC biomarker discovery programme, PROMISERA, and Chief Investigator of numerous global phase II and III clinical trials of novel immune therapies. He has served as member or chair of numerous national grant-funding panels and as Chair of several EULAR Scientific Committees. He is immediate past chairman of the Foreum (Foundation for European Rheumatology Research) Scientific Committee, leads the European Roadmap programme that is defining the research agenda for rheumatology for the next decade and has just been elected Treasurer of EULAR. He was elected a Fellow of the Royal Society of Edinburgh in 2008 and the Academy of Medical Sciences in 2012.

17.30-18.30 INC meeting (open to all)
Main discussion topic: Clinical Trials

18.30-19.30 Poster viewing

20.00 Dinner on your own in local restaurants
(Glasgow West End)
His research interests include understanding the role of cytokines in inflammatory synovitis. He leads a trials unit specialising in the use of biologic agents in early clinical trials in inflammatory arthritis. Professor McInnes has published widely in the areas of Immunobiology and rheumatology, and is an Associate Editor of the Annals of Rheumatic Diseases and a member of the executive Editorial Board of European Journal of Immunology. His work, together with that of his colleagues at the University of Glasgow, has been recognised in receipt of many prizes and lectureships including most recently the Sir James Black medal 2015 from the Royal Society of Edinburgh.

He will discuss biological depletion therapies in relation to both rheumatological and neurological diseases.

10.00-10.30  Future immunotherapeutics for inflammatory neuropathy
Jane Owens

Dr Jane Owens is Senior Director and Head of the Neuromuscular Portfolio within the Rare Disease Research Unit at Pfizer and is project leader of the GL-2045 (a novel recombinant immunotherapeutic) Development Team. Discovery of therapeutics for rare hematology and neuromuscular indications is a key area of focus for the unit. An opportunity to target indications potentially in both categories arose from the license of a biologic molecule from Gliknik Inc. – GL-2045, a multimerized human IgG1 Fc being developed to treat rare autoimmune diseases. GL-2045 is a recombinant mimetic of pooled human immune globulin (IVIG). There is a significant unmet need for patients with these debilitating diseases, especially those suffering with the autoimmune neuropathies and being treated primarily with IVIG. That’s because, although highly effective, trading plasma-derived treatment comes with blood-borne pathogen transmission and supply interruption risks, along with a burdensome administration regimen which warrants improvement.

Dr Owens will discuss the future of immunotherapeutics in Neuropathy, by introducing molecules with potential to replace IVIG or expand the marketplace, including the proposed mechanism of action and evidence of efficacy of GL-2045, which demonstrates significant potency improvements compared to IVIG in several rodent models.

10.30-11.00  Refreshment break

11.00-12.30  Short and Speed Poster Presentations
Chaired by Marinella Carpo and Alexander Rossor

11.00-11.15  Systemic IGF-1 gene delivery by rAAV9 improves spontaneous autoimmune peripheral polyneuropathy (SAPP)
Tong Gao

11.15-11.30  Macrophage imaging in experimental nerve injury by 68Ga-DOTATATE: a combined autoradiography/PET study
Guido Stoll

11.30-12.30  Speed Poster Presentations

SP2.1  Guillain-Barré syndrome related to Zika virus infection in Brazil
Amilton Antunes Barreira

SP2.2  Guillain-Barré syndrome associated to Zika virus outbreak in Recife, northeast Brazil
Maria Lucia Brito Ferreira

SP2.3  MRI findings of the central and peripheral nervous system in patients with Zika virus infection
Ivan Rocha F. da Silva

SP2.4  Investigation of Guillain-Barré syndrome outbreak – Bahia State, Brazil, 2016
James Sejvar

SP2.5  Does varicella zoster virus infection trigger the demyelinating form of Guillain-Barré syndrome?
Badrul Islam

SP2.6  Different axonal excitability properties between acute inflammatory demyelinating polyneuropathy and acute motor sensory axonal neuropathy
Jong Seok Bae

SP2.7  Sural sparing is specific of demyelinating Guillain-Barré syndrome
Fu Liong Hiew

SP2.8  Differences in response to intravenous immunoglobulin in immune-mediated neuropathy: the importance of conduction block
Nichi Garg

SP2.9  Electrophysiologic errors contribute to CIDP mis-diagnosis
Jeffrey Allen

SP2.10  Sensory nerve conduction studies should be included in the electrodiagnostic criteria of Guillain-Barré syndrome.
Christen Sheng Jie Lim

SP2.11  Differences in biophysical properties between motor and sensory axons
Boudewijn THM Sleutjes

SP2.12  Optimizing the electrodiagnosis of Guillain-Barré syndrome subtypes: criteria sets versus a linear discriminant analysis model
Antonino Uncini

SP2.13  The marriage of high-resolution sonography and nerve conduction studies in IgM MGUS polyneuropathy: more generalised than length dependent?
Stephan Goedee
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12.30-14.00 Lunch break and poster viewing

14.00-15.30 Baxalta-sponsored Plenary Symposium 4: Clinical biomarkers, surrogates and diagnostic devices for inflammatory neuropathies Chaired by Pieter van Doorn and Osvaldo Nascimento

14.00-14.30 CSF and serum biomarkers in PNS disease/ultrasensitive measurements of neurofilament light in the Simoa platform Åsa Sandelius

Dr Åsa Sandelius is a Post doctoral fellow at the Institute for Neuroscience and Physiology, Department for Psychiatry and Neurochemistry at Sahlgrenska Academy, Gothenburg University, Sweden, whose major research interests are fluid biomarker research for neurodegenerative disorders and neuronal injury, focusing on Alzheimer’s disease and immunoassay development. She will present a novel immunoassay platform for ultrasensitive protein quantification, the Single Molecule Array (Simoa), in which the enhancement of sensitivity makes it possible to measure proteins of very low abundance and in the case of CNS proteins measure in blood instead of CSF. Dr Sandelius will present a comparison of neurofilament light (NFL) quantification in serum, plasma and CSF using the Simoa, regular ELISA and an electrochemiluminescent method. Furthermore, she will discuss the biomarker potential of NFL and its relationship to clinical biomarkers, surrogates and diagnostic devices for inflammatory neuropathies.

14.30-15.00 Skin biopsy as a diagnostic tool and biomarker for disease progression and therapy Sung-Tsang Hsieh

Dr Sung-Tsang Hsieh, Professor of Neurology, National Taiwan University Hospital, has been working on small fiber neuropathy for 2 decades. His laboratory is one of the early groups developing skin biopsy as an innovative diagnostic method by measuring intraepidermal nerve fiber density (IENF density) as the major criterion of diagnosing small fiber neuropathy. Dr Hsieh further explored the clinical significance of skin innervation, i.e. the functional correlations of IENF density. He also established and combined physiology (contact heat evoked potential) and imaging (functional MRI) methods as an integrated approach to examine and investigate neuropathic pain mechanisms after peripheral nerve degeneration. In addition to examining nociceptive nerves in the epidermis, skin biopsy also provides opportunities to study autonomic innervation of sweat glands in the dermis. Skin biopsy has been applied for following up the progression of small fiber neuropathy prospectively. This presentation will focus on the potential roles of skin biopsy as a biomarker for therapeutic responses.

15.00-15.30 Imaging for the evaluation of peripheral neuropathies Roberto Gasparotti

Dr Roberto Gasparotti is a Professor of Neuroradiology at the University of Brescia medical school and serves as chief of the Neuroradiology Unit at the University Hospital Spedali Civili, Brescia, Italy. His major research interests include Magnetic Resonance Imaging in neurodegenerative disorders and peripheral nerve diseases. He will discuss the current role of advanced Magnetic Resonance techniques, such as Neurography and Diffusion Tensor Imaging in the evaluation of inflammatory neuropathies.

15.30-15.45 Refreshment break

15.45-16.45 Speed Poster Presentations Chaired by Jane Pritchard and Christine Verboon

SP3.1 The International Guillain-Barré Study (IGOS): a prospective study on clinical and biological predictors of disease course and outcome Bianca van den Berg

SP3.2 IGOS-Kids: children with Guillain-Barré syndrome Joyce Roodbol

SP3.3 An overview of clinical features of Guillain-Barré syndrome: a retrospective analysis based on 566 patients in Northeast China Ying Wang

SP3.4 Construction and validation of the chronic acquired polyneuropathy patient-reported index, "CAP-PRI:" a disease-specific, health-related quality of life instrument Kelly Gwathmey

SP3.5 Observer-blind randomised controlled trial to evaluate the efficacy of a twelve week tailored home exercise programme in the management of inflammatory neuropathy Claire White

SP3.6 Pharmacokinetics of intravenous immunoglobulin maintenance treatment in patients with CIDP Willem-Jan Fokkink

SP3.7 Immunoglobulin treatment for patients with mild Guillain-Barré syndrome: an international prospective observational study Christine Verboon
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SP3.8 Prevalence of anti-neurofascin-155 and anti-contactin-1 antibodies in chronic inflammatory demyelinating poly-radiculoneuropathy: a serological multicenter study in Italy
Andrea Cortese

SP3.9 How often is Fisher syndrome overlapped with Guillain-Barré syndrome or Bickerstaff brainstem encephalitis?
Yukari Sekiguchi

SP3.10 Incidence and treatment of A-CIDP in second IVIg dose in Guillain-Barré syndrome patients with poor prognosis (SID-GBS) trial – a case series
Christa Walgaard

SP3.11 Presentation of familial amyloid polyneuropathies fulfilling clinical and electrophysiological criteria for a demyelinating polyneuropathy.
Pierre Lozeron

SP3.12 Diffusion tensor imaging of the brachial plexus as an aid to the diagnosis of inflammatory neuropathies: preliminary results
Jos Oudeman

SP3.13 Corneal confocal microscopy in chronic inflammatory demyelinating polyneuropathies
Mark Stettner

SP3.14 Difference in small fiber involvement in CIDP and POEMS syndrome: An intra-epidermal electrical stimulation pain-SEP study
Sagiri Isose

SP3.15 Intravenous immunoglobulin therapy for small fiber neuropathy
Bianca de Greef

16.45-17.30 Debate: Why my database is better than yours.
Speakers: Stephen Reddel, Filip Eftimov, Luis Querol, Susumu Kusunoki, Christopher Klein, Eduardo Nobile-Orazio.
Moderator: Richard Lewis

17.30-18.30 Poster viewing

19.00-20.00 Civic Reception in Glasgow City Chambers with group photo

20.00 Dinner on your own in Glasgow downtown restaurants (Merchant City)

Friday, June 24, 2016

09.00-09.05 Welcome
Hugh Willison, local organiser

09.05-09.15 Selected readings from the 1916 paper
Jean-Marc Léger

09.15-09.35 My life with GBS – the doctor perspective
Richard AC Hughes

09.35-10.00 My life with GBS – the patient perspective
Estelle Benson, Glennys Sanders

10.00-10.20 Axonal GBS: heretic or visionary?
Tom Feasby, chaired and questioned by Robert Hadden

10.20-10.40 Gangliosides and anti-ganglioside antibodies
Susumu Kusunoki, chaired and questioned by Ruth Huizinga

10.40-11.00 International cooperation in GBS research: IGOS
Bart Jacobs, chaired and questioned by Judith Spies

11.00-11.20 Refreshment Break

11.20-11.50 In conversation: animal models of GBS – do they inform or mislead?
Introduction: Kazim Sheikh
Discussants: John Pollard, Madeleine Cunningham, Anne Mausberg, Guido Stoll
Summing up: Tom Feasby
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| 11.50-12.10 | Delivering specialist care to patients with GBS and research: challenges and rewards in a mixed resource setting.  
*Sponsored by the Chandra Mehta Foundation*  
Arun Taly interviewed by Luis Querol   |                                                                                         |
| 12.10-13.00 | **GBS World Tour: global perspectives**                                                           |                                                                                         |
| 12.10-12.25 | GBS in children – the East and West perspective  
Joyce Roodbol (Holland) & Badrul Islam (Bangladesh)                                              |                                                                                         |
| 12.25-12.40 | GBS and Zika virus infection in Pernambuco, Brazil  
Lucia Brito                                                                                  |                                                                                         |
| 12.40-12.55 | GBS in a tertiary general hospital at Yangon, Myanmar  
Ohnmar                                                                                  |                                                                                         |
| 13.00-14.00 | **Lunch Break**                                                                                  | Video interviews will be shown on screens throughout lunch                                |
| 14.00-14.20 | Fisher syndrome and other GBS variants  
Allan Ropper, chaired and questioned by Nortina Shahrizaila                                     |                                                                                         |
| 14.20-14.50 | Treatment of GBS: past, present & future. From PE/IVIG to ICA-GBS and JET-GBS  
Pieter van Doorn, Amy Davidson, Satoshi Kuwabara   |                                                                                         |
| 14.50-15.10 | The top hits in the GBS bibliography: analysis and clinical commentary on the lead articles  
Susan Ashworth and Emilien Delmont                                 |                                                                                         |
| 15.10-15.40 | Case reports: what they have told us about patient care: Selected presentations  
Stephen Reddel and Umapathi Thirugnanam |                                                                                         |
| 15.40-16.00 | **Launch of GBS100 Monograph with selected readings/comments by Authors**                      | John Goodfellow and others.                                                           |
| 16.00-16.30 | Short Break, including a book launch toast                                                      |                                                                                         |
| 16.30-17.30 | In search of the world’s expert on GBS: a quiz  
Quizmasters: Simon Rinaldi and Ken Gorson                                                      |                                                                                         |
| 17.30-17.55 | **GBS – The Next 100 Years**  
Coordinated by Ken-ichi Kaida and Christine Verboon  
Immunopathogenesis – Madeleine Cunningham;  
Diagnosis – Nortina Shahrizaila; Prognostic modelling – Christa Walgaard;  
GBS in developing countries – Badrul Islam; International cooperation – Bianca van den Berg |                                                                                         |
| 17.55-18.00 | Closing remarks: Michael Lunn, programme chair                                                  |                                                                                         |
| 19.00     | **Centenary Banquet and Ceilidh, Oran Mor, Byers Road**  
Presentation of PNS/INC Awards                                                                  |                                                                                         |