27th international symposium on ALS/MND

Dublin, Republic of Ireland
7 – 9 December 2016

Programme

Host: Irish Motor Neurone Disease Association (IMNDA)

Organised by the Motor Neurone Disease Association in co-operation with the International Alliance of ALS/MND Associations
The 27th International Symposium on ALS/MND has been approved by the Federation of the Royal College of Physicians of the United Kingdom for 18 category 1 (external) CPD credit(s).

Host for the symposium:

Irish Motor Neurone Disease Association
Coleraine House, Coleraine Street, Dublin 7
Tel: (-) 01 873 0422
Email: info@imnda.ie
Website: www.imnda.ie

Live educational activities, occurring outside of Canada, recognized by the UEMS-EACCME for ECMEC credits are deemed to be Accredited Group Learning Activities (Section 1) as defined by the Maintenance of Certification Program of The Royal College of Physicians and Surgeons of Canada.

CME Accreditation

The 27th International Symposium on ALS/MND is accredited by the European Accreditation Council for Continuing Medical Education (EACCME) to provide the following CME activity for medical specialists. The EACCME is an institution of the European Union of Medical Specialists (UEMS), www.uems.net.

The ‘27th International Symposium on ALS/MND’ is designated for a maximum of 16 hours of European external CME credits. Each medical specialist should claim only those hours of credit that he/she actually spent in the educational activity.

Through an agreement between the European Union of Medical Specialists and the American Medical Association, physicians may convert EACCME credits to an equivalent number of AMA PRA Category 1 Credits™. Information on the process to convert EACCME credit to AMA credit can be found at www.ama-assn.org/go/internationalcme.

Organiser of the symposium:

Motor Neurone Disease Association
10-15 Notre Dame Mews,
Northampton NN1 2BG, UK
Tel: (-) 44 1604 611845 or 611822
Fax: (-) 44 1604 611858
Email: symposium@mndassociation.org
Website: www.mndassociation.org

Held in co-operation with:

The International Alliance of ALS/MND Associations
Tel: (-) 1 215 568 2426
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Email: alliance@als-mnd.org
Website: www.alsmndalliance.org
Welcome

Fáilte go Baile Átha Cliath – Welcome to Dublin!

The Irish Motor Neurone Disease Association extends a warm welcome to you to Dublin for the 27th International Symposium on ALS/MND and the accompanying 24th Annual Meeting of the International Alliance of ALS/MND Associations. Participation in these important meetings allows us to work together towards our global vision ‘A world free from Motor Neurone Disease’.

The name Dublin comes from the Gaelic words “Dubh Linn”, referring to a black pool at the time of the city’s foundation over 1000 years ago. Combining a solid reputation for education and innovation, with great accessibility, it is no coincidence that Dublin is now the European headquarters of many of the world’s leading corporations, and a centre for academic research and innovation.

The Irish Motor Neurone Disease Association (IMNDA) is the only organisation of its kind in the country. We are dedicated to working on behalf of people living with MND and their families and carers. Our key services include home visiting by an MND nurse, financial assistance towards home care help and the supply of specialised equipment on loan. We also fund and promote research into the causes and treatments of MND.

The IMNDA works very closely and supports the Irish MND Research Group based at Beaumont Hospital, and Trinity College Dublin, led by Professor Orla Hardiman. The work of the Irish MND Research Group focuses on deep phenotyping, biomarker development and genomics. In collaboration with researchers across the world, it seeks to expand our understanding of MND, by recognising and studying all aspects of disease heterogeneity and by developing new models of disease classification that will support a precision medicine approach towards new treatments.

Dublin has not forgotten its roots and is steeped in a rich cultural heritage that begs to be explored. All year round, the city is alive with music, art and theatre of all kinds and, of course, art galleries and museums abound. All complemented by an impressive range of restaurants and cafés, the best golf courses, exciting surfing spots, scenic coastlines and picturesque mountains.

As the famous Irish poet William Butler Yeats wrote ‘There are no strangers here, only friends that have not yet met.’ Together we will fight for a world free from MND/ALS.

Aisling Farrell
CEO, Irish Motor Neurone Disease Association

Foreword

Welcome to the 27th International Symposium on ALS/MND. This year’s programme continues the tradition of showcasing the most important developments in both science and clinical practice. The remarkable discoveries in the genetics of amyotrophic lateral sclerosis in the last 10 years have provided a platform for studies which are now revealing important insights into the key mechanisms leading to motor neuron degeneration and the overlap with frontotemporal dementia. Although it is encouraging to see more clinical trials than ever being presented at the symposium, ALS/MND is a clinical syndrome with complex biological determinants. We have sessions highlighting how future treatments may well be based on more rational target discovery and personalised medicine. This requires an understanding of the ‘big data’ emerging from high throughput next generation sequencing, epigenetics and the study of environmental exposures.

Advances in technology also feature in the sessions on patient based biomarker research and also clinical practice. The ability to study the disease before clinical onset is now becoming a reality due to advances in imaging and the willingness of subjects at risk of ALS/MND to engage with the research community. Telehealth and E-learning demonstrate how we can engage with clinical management in ever more flexible and creative ways.

Data is everywhere and the more we can share our knowledge and resources the greater our capacity to improve the lives of people living with MND. The annual symposium is where this sharing begins. On behalf of the Programme Committee I wish you a happy and productive meeting.

Prof Kevin Talbot
Programme Committee Chair

Aisling Farrell
CEO, Irish Motor Neurone Disease Association
Programme

Wednesday 7 December 2016

SESSION 1 LOCATION: THE LIFFEY B

JOINT OPENING SESSION

Chairs: S Light (UK) T Talbot (UK)

08.30 – 08.35
Welcome – S Light (UK) T Talbot (UK)

08.35 – 08.40
Welcome from Host Association – Special Guest

08.40 – 09.10
C1 Insights into the ALS/MND exposome – R Vermeulen (The Netherlands)

09.10 – 09.40
C2 Untangling the X-files of ALS – R Bedlack (USA)

09.40 – 09.55
International Alliance Humanitarian Award, International Alliance Forbes Norris Award

09.55 – 10.15
IPG Award and winner’s research presentation

10.30 – 11.00 COFFEE Locations: The Forum and Level 3 Foyer

SESSION 2A LOCATION: THE LIFFEY A

RNA PROCESSING AND DYSREGULATION

Chairs: J Rothstein (USA) J Ule (UK)

11.00 – 11.30
C3 Using iCLIP to study the assembly of protein-RNA complexes associated with MND – J Ule (UK)

11.30 – 11.50
C4 ALS and Artificial Intelligence: IBM Watson suggests novel RNA binding proteins altered in ALS – N Bakkar (USA)

11.50 – 12.10
C5 Matrin-3 regulates TDP-43 levels via its 3'UTR region – E Rodriguez-Lebron (USA)

12.10 – 12.30
C6 Muscleblind protects against transcriptomic dysregulation and neurodegeneration in FUS mediated ALS – U Pandey (USA)

SESSION 2B LOCATION: THE LIFFEY B

MULTIDISCIPLINARY MANAGEMENT

Chairs: H Mitsumoto (USA) D Oliver (UK)

11.00 – 11.30
C7 Developing and implementing the NICE guideline on MND – D Oliver (UK)

11.30 – 11.50
C8 Longitudinal analysis of patient communication and treatment preferences in an ALS clinic – Z Simmons (USA)

11.50 – 12.10
C9 Determinants of therapeutic decision making in ALS in Germany, Sweden and Poland – D Lulé (Germany)

12.10 – 12.30
C10 Comparison of survival of people with ALS by diagnostic cohort – S Martin (UK)

12.30 – 14.00 LUNCH Locations: The Forum and Level 3 Foyer

SESSION 3A LOCATION: THE LIFFEY A

RNA AND NEURODEGENERATION

Chairs: E Hornstein (Israel) C Shaw (UK)

14.00 – 14.30
C11 Vulnerability of microRNAs in FTD-ALS – E Hornstein (Israel)

14.30 – 14.50
C12 Transcellular spread of motor degeneration via microRNAs in genetic models of ALS – A Parker (Canada)

14.50 – 15.10
C13 Circular RNA biogenesis is dependent on FUS and is impaired in an ALS model system – S Dini Modigliani (Italy)

15.10 – 15.30
C14 Loss of TDP-43 contributes to non-coding RNA mediated toxicity – E Lee (USA)

SESSION 3B LOCATION: THE LIFFEY B

ALS/FTD

Chairs: T Bak (UK) O Hardiman (Ireland)

14.00 – 14.30
C15 ALS/FTD as a disorder of connectivity: Why are cognitive symptoms an integral part of the ALS spectrum – T Bak (UK)

14.30 – 14.50
C16 Cognitive and behavioural profiles in frontotemporal dementia with and without amyotrophic lateral sclerosis – J Saxton (UK)

14.50 – 15.10
C17 Cognitive impairment in MNDs: Expanding from ALS to PLS & PMA – E de Vries (The Netherlands)

15.10 – 15.30
C18 Beyond the motor system: Exploring psychosis in ALS – E Devenney (Australia)

15.30 – 16.00 COFFEE Locations: The Forum and Level 3 Foyer

The Opening Session will be busy, so please arrive promptly. Once the room capacity is reached, delegates will follow the session in an adjoining room (audio and PowerPoint only).
SESSION 4A LOCATION: THE LIFFEY A

PROTEIN MISFOLDING AND AGGREGATION

Chairs: H Durham (USA) J Ravits (USA)

16.00 – 16.20
C19 Proteins found in ALS inclusions are supersaturated indicating proteostasis collapse in motor neurons – J Yerbury (Australia)

16.20 – 16.40
C20 TDP-43 and SOD1: A toxic pas de deux in ALS – E Pokrishevsky (Canada)

16.40 – 17.00
C21 CCNF mutations in ALS and FTD lead to dysfunctional protein homeostasis – I Blair (Australia)

17.00 – 17.20
C22 Transfer of ALS protein aggregates between motor neurons in the zebrafish spinal cord – M Morsch (Australia)

17.20 – 17.40
C23 Modelling neuroanatomic propagation of ALS in the spinal cord – B Drawert (USA)

Late breaking news: Mutant SOD1 aggregates from human ventral horn transmit templated aggregation and fatal ALS-like disease – S Marklund (Sweden)

LOCATION: THE FORUM 17.45 – 19.30

POSTER SESSION A

Only Poster Session A posters will be on display throughout this session. The presenters of each poster theme will be available for discussion at the following times:

18.00 – 18.20
Theme 1: Epidemiology
Theme 7: Improving diagnosis and prognosis

18.20 – 18.40
Theme 5: Human cell biology and pathology
Theme 8: Imaging and electrophysiology

18.40 – 19.00
Theme 3: In vitro experimental models
Theme 9: Therapeutic strategies

19.00 – 19.20
Theme CW: Clinical work in progress
Theme BW: Biomedical work in progress

Thursday 8 December 2016

SESSION 5A LOCATION: THE LIFFEY A

THERAPEUTIC STRATEGIES

Chairs: L Brujin (USA) P Shaw (UK)

08.30 – 08.50
C28 Designing kinase inhibitors to combat ER stress-mediated apoptosis in a stem cell model of ALS – E Lowry (USA)

08.50 – 09.10
C29 Modulation of UPR response in IPS cell-derived motor neurons from ALS-patients – Y Rudhard (Germany)

09.10 – 09.30
C30 Identification of therapeutic targets for cytoskeletal defects in ALS – A Javaherian (USA)

09.30 – 10.00
C31 Challenges in CNS drug discovery – P Brennan (UK)

SESSION 5B LOCATION: THE LIFFEY B

PRE/EARLY SYMPTOMATIC DISEASE

Chairs: P Andersen (Sweden) M Turner (UK)

08.30 – 09.00
C32 Detecting early changes in FTD – J Rohrer (UK)

09.00 – 09.20
C33 Quantitative motor testing: Biomarker of pre-symptomatic ALS? – M Benatar (USA)

09.20 – 09.40
C34 Cortical dysfunction is a global phenomenon in ALS – P Menon (Australia)

09.40 – 10.00
C35 Blood biomarkers of carbohydrate and lipid metabolism and risk of amyotrophic lateral sclerosis: A more than 20 year follow-up of the Swedish AMORIS cohort – D Mariosa (Sweden)

SESSION 5C LOCATION: LIFFEY HALL 2

AUTONOMY AND QUALITY OF LIFE: THE PATIENT-CARER DYAD

Chairs: D Lule (Germany) M Ogino (Japan)

08.30 – 09.00
C36 Physical and psychological influences upon quality of life in motor neurone disease/ALS – C Young (UK)

09.00 – 09.20
C37 Subjective perception of health in ALS: A moving target? – N Thakore (USA)

09.20 – 09.40
C38 Are caregivers able to correctly predict ALS patients’ wish for hastened death and their well-being – J Keller (Germany)

09.40 – 10.00
C39 Journey to ALS diagnosis: Caregiver perspectives – M Galvin (Ireland)

10.00 – 10.30 COFFEE Locations: The Forum and Level 3 Foyer
**SESSION 6A** LOCATION: THE LIFFEY A

**CELL BIOLOGY AND PATHOLOGY**

Chairs: M Hafezparast (UK) J Prehn (Ireland)

10.30 – 11.00
C40 Angiogenin, tRNA and vascular integrity in health and disease – J Prehn (Ireland)

11.00 – 11.20
C41 Apical dendrite degeneration, a new cellular pathology in amyotrophic lateral sclerosis – H Ozdinler (USA)

11.20 – 11.40
C42 Dyshomeostasis of copper proteins is a common feature of sporadic human MND and transgenic mouse models: Outcomes from a novel metalloproteomic analysis – P Crouch (Australia)

11.40 – 12.00
C43 Nuclear export inhibition of C9ORF72 repeat transcripts prevents neuronal death and associated motor deficits – G Hautbergue (UK)

12.00 – 12.20
C44 The DNA damage response (DDR) is induced by the C9ORF72 repeat expansion in ALS – M Farg (Australia)

12.20 – 12.40
C45 The RNA-binding protein, hnRNP K, forms a critical nexus between TDP-43 pathology and oxidative stress in ALS – D Moujalled (Australia)

12.40 – 13.00
C46 Delineating mechanisms of dysphagia in ALS – E Plowman (USA)

13.00 – 13.20
C47 Eating and cognition across the amyotrophic lateral sclerosis-frontotemporal dementia spectrum: Effect on survival – E Devenney (Australia)

13.20 – 13.40
C48 A decrease in blood cholesterol after gastrostomy could impact survival in ALS – H Blasco (France)

13.40 – 14.00
C49 Gut appetite regulatory and metabolic hormones in ALS: Relationship to body composition, energy expenditure and survival – E Kasarskis (USA)

14.00 – 14.20
C50 Changes in energy metabolism in ALS are associated with alterations in glucose and fatty acid flux – S Ngo (Australia)

**SESSION 6B** LOCATION: THE LIFFEY B

**NUTRITIONAL MANAGEMENT AND METABOLISM**

Chairs: J-P Loeffler (France) R Tandan (USA)

10.30 – 10.50
C46 Delineating mechanisms of dysphagia in ALS – E Plowman (USA)

10.50 – 11.10
C47 Eating and cognition across the amyotrophic lateral sclerosis-frontotemporal dementia spectrum: Effect on survival – E Devenney (Australia)

11.10 – 11.30
C48 Does percutaneous endoscopic gastrostomy lengthen survival in patients with weight loss when bulbar function is preserved? – L Jenkins (USA)

11.30 – 11.50
C49 A decrease in blood cholesterol after gastrostomy could impact survival in ALS – H Blasco (France)

11.50 – 12.10
C50 Gut appetite regulatory and metabolic hormones in ALS: Relationship to body composition, energy expenditure and survival – E Kasarskis (USA)

12.10 – 12.30
C51 Changes in energy metabolism in ALS are associated with alterations in glucose and fatty acid flux – S Ngo (Australia)

**SESSION 6C** LOCATION: LIFFEY HALL 2

**NEUROIMAGING**

Chairs: P Bede (Ireland) J Grosskreutz (Germany)

10.30 – 10.50
C52 Gray matter correlates of cognitive decline in ALS: A multi-atlas based MRI study – M França Jr (Brazil)

10.50 – 11.10
C53 Cortical profile of C9orf72 gene expression associated with cortical thinning in amyotrophic lateral sclerosis – R Schmidt (The Netherlands)

11.10 – 11.30
C54 Functional and structural connectivity in ALS – insights from MRI connectome analyses and TMS – N Geevasinga (Australia)

11.30 – 11.50
C55 The progression of cerebral pathology in ALS: A six-monthly multi-modal MRI study over two years – R Menke (UK)

11.50 – 12.10
C56 Development of an automated MRI-based diagnostic protocol based on disease-specific pathognomonic features in amyotrophic lateral sclerosis: A quantitative disease-state classification study – C Schuster (Ireland)

12.10 – 12.30
C57 Data-driven modelling of diffusion MRI changes in ALS indicates evolution of distal prior to proximal corticospinal tract pathology – M Gabel (UK)

12.30 – 14.00 LUNCH Locations: The Forum and Level 3 Foyer
SESSION 7A LOCATION: THE LIFFEY A

EPIGENETICS AND GENOMICS

Chairs: J Kirby (UK) J Veldink (The Netherlands)

14.00 – 14.30

C58 Epigenetic pathways to neuropsychiatric and neurological disease – J Mill (UK)

14.30 – 14.50

C59 Epigenetic modelling and therapeutic targeting of the expanded C9ORF72 locus – Z Zeier (USA)

14.50 – 15.10

C60 Changes in expression levels of homeobox genes and transthyretin in patients with C9ORF72 repeat expansions – W Van Blitterswijk (USA)

15.10 – 15.30

C61 A gene signature for amyotrophic lateral sclerosis associated with TDP-43 pathology – C Jackson (UK)

15.30 – 16.00

Coffee Locations: The Forum and Level 3 Foyer

SESSION 7B LOCATION: THE LIFFEY B

SYMPTOMATIC TREATMENTS

Chairs: O Gredal (Denmark) C Jackson (USA)

14.00 – 14.30

C62 The CANALS study: A randomized, double-blind, placebo-controlled, multi-centre study to assess the safety and efficacy on spasticity symptoms of a Cannabis Sativa extract in motor neuron disease patients – N Riva (Italy)

14.30 – 14.50

C63 Aerobic exercise therapy in patients with amyotrophic lateral sclerosis (FACTS-2-ALS): A randomized clinical trial – A van Groenestijn (The Netherlands)

14.50 – 15.10

C64 Meditation training for people with amyotrophic lateral sclerosis: A randomized clinical trial – F Paginni (Italy)

15.10 – 15.30

C65 Early treatment with NIPPV: factors affecting compliance over time – C Jackson (USA)

15.30 – 16.00

SESSION 7C LOCATION: LIFFEY HALL 2

MOUSE MODELS

Chairs: G Nardo (Italy) F René (France)

14.00 – 14.20

C66 Phenotypic characterization of a new CHMP2Btransgenic mouse that develops histological and behavioural features of amyotrophic lateral sclerosis and frontotemporal dementia – R Waeggaert (France)

14.20 – 15.00

C67 AAV9-mediated C9orf72 experimental modelling of ALS/FTD in mice – M Azouz (UK)

15.00 – 15.15

C68 Degeneration of serotonin neurons is necessary to elicit spasticity in amyotrophic lateral sclerosis – L Dupuis (France)

15.15 – 15.30

C70 Robust beneficial effects of a non-competitive AMPA receptor antagonist in an ALS mouse model – M Akamatsu (Japan)

15.30 – 16.00

Coffee Locations: The Forum and Level 3 Foyer

SESSION 8A LOCATION: THE LIFFEY A

CLINICAL GENETICS

Chairs: I Blair (Australia) P Corcia (France)

16.00 – 16.30

C71 Genetic pleiotropy – D Goldstein (USA)

16.30 – 16.50

C72 Genetic screening of 18,926 samples reveals new risk alleles for familial and sporadic ALS – K Kenna (USA)

16.50 – 17.10

C73 Project MiniE GWAS: Genome-wide association analyses identify new risk variants and the genetic architecture of amyotrophic lateral sclerosis – W Van Blitterswijk (The Netherlands)

17.10 – 17.30

C74 Shared Novel Variant Analysis Identifies Novel Genes in Familial ALS from Whole Exome Sequencing – S Topp (UK)

SESSION 8B LOCATION: THE LIFFEY B

TECHNOLOGY AND ALS

Chairs: C McDermott (UK) P Wicks (USA)

16.00 – 16.20

C75 The power of sharing data: 10 years experience with PatientsLikeMe – P Wicks (USA)

16.20 – 16.40

C76 Telehealth in motor neuron disease: A mixed methods, randomised controlled, pilot study of the use of the TiM telehealth system to deliver highly specialised care in motor neuron disease, at a distance – E Hobson (UK)

16.40 – 17.00

C77 Optimising care through telemonitoring in ventilated patients with motor neuron disease: A pilot study – S Rutkove (USA)

17.00 – 17.20

C78 E-learning for ALS health care providers – C Roos (The Netherlands)

17.20 – 17.40

C79 Augmentative and alternative communication for locked-in state patients – P Fedele (Italy)

SESSION 8C LOCATION: LIFFEY HALL 2

EVOLVING BIOMARKERS

Chairs: M De Carvalho (Portugal) M Weber (Switzerland)

16.00 – 16.20

C80 Tissue-enhanced proteomic analysis of plasma samples reveals new mechanistic biomarker candidates for the stratification of amyotrophic lateral sclerosis patients – I Zubiri (UK)

16.20 – 16.40

C81 Electrical impedance myography for early diagnosis and assessment of ALS progression: results of a multicenter clinical trial – S Rutkove (USA)

16.40 – 17.00

C82 MEG cortico-muscular coherence to assess corticospinal tract integrity in ALS – M Meier (USA)

17.00 – 17.20

C83 Auditory mismatch negativity in amyotrophic lateral sclerosis – J Mill (The Netherlands)

17.20 – 17.40

C84 Glial activation measured by [11C]-PBR28 PET correlates with 1H-MRS brain metabolites in amyotrophic lateral sclerosis – E Ratai (USA)
Friday 9 December 2016

SESSION 9A LOCATION: THE LIFFEY A

NEURON-GLIA INTERACTIONS

Chairs: L Barbeito (Uruguay) S Przedborski (USA)

08.30 – 09.00
C85 Astrocyte toxicity in models of ALS – S Przedborski (USA)

09.00 – 09.20
C86 Early stage motor neurons neuroprotection via astrocytes restricted NF-κB activation – N Ouali Alami (Germany)

09.20 – 09.40
C87 Neuronal pathophysiology in a human iPSC model of ALS involves interplay between astrocytes and motor neurons – A Chouhan (UK)

09.40 – 10.00
C88 Predicting novel relationship of glia to the caudal distribution of lower motor neuron in relation to the region of clinical onset in sporadic ALS patients – F Song (USA)

SESSION 9B LOCATION: THE LIFFEY B

CLINICAL TRIALS

Chairs: M Cudkowicz (USA) J Mora (Spain)

08.30 – 08.50
C89 Long-term safety and efficacy of Edaravone (MCI-186) for the treatment of amyotrophic lateral sclerosis (ALS) – J Palumbo (USA)

08.50 – 09.10
C90 Adaptive design single center phosphodiesterase type 4 (PDE-4) inhibitor-(MN-166 (Ibudilast)) phase 1b/2a clinical trial – interim-blinded analysis – behavior of creatinine as a biomarker in short clinical trials (NCT02238626) – B Brooks (USA)

09.10 – 09.30
C91 Rasagiline for the treatment of ALS: A randomized controlled study – R Barohn (USA)

09.30 – 09.45
C92 VITALITY-ALS, a Phase 3 trial of the fast skeletal muscle troponin activator, Tirasemtiv, for the potential treatment of amyotrophic lateral sclerosis (ALS): Study design and baseline characteristics – J Shefner (USA)

09.45 – 10.00
C93 Can pyrimethamine lower CSF SOD1 levels in familial ALS? Results from a multicenter Phase II trial – D Lange (USA)

10.00 – 10.30 COFFEE Locations: The Forum and Level 3 Foyer
SESSION 10A LOCATION: THE LIFFEY A

NEUROINFLAMMATION

Chairs: S Appel (USA) M Lynch (Ireland)

10.30 – 11.00
C94 The contribution of inflammation to neurodegeneration – M Lynch (Ireland)

11.00 – 11.20
C95 Suppressing neuroinflammation: A key to therapy in amyotrophic lateral sclerosis – S Appel (USA)

11.20 – 11.40
C96 Activated immune response in the peripheral nervous system is instrumental to delay the disease progression in ALS mouse models – G Nardo (Italy)

11.40 – 12.00
C97 Early- and late-activated microglia show distinct localizations and exert different impacts on TDP-43 pathology in amyotrophic lateral sclerosis spinal cord – S Hayashi (Japan)

12.00 – 12.20
C98 Post-paralysis treatment with masitinib significantly slows disease progression in transgenic SOD1G93A rats – L Barbeito (Uruguay)

12.20 – 12.40
C99 An unexpected role for microglia during recovery from motor neuron disease in a new mouse model of TDP-43 proteinopathy – K Spiller (USA)

SESSION 10B LOCATION: THE LIFFEY B

DISEASE PROGRESSION AND PROGNOSTIC MODELLING

Chairs: A Al-Chalabi (UK) J Rosenfeld (USA)

10.30 – 10.45
C100 Baseline predictors of survival in a large cohort of ALS patients: The ALS COSMOS study – P Factor-Litvak (USA)

10.45 – 11.00
C101 Development and external validation of a prognostic model estimating survival in individual ALS patients – H Westeneng (The Netherlands)

11.00 – 11.15
C102 Retrospective analysis of data from a Phase III trial of Edaravone in amyotrophic lateral sclerosis (ALS) using two ALS Clinical Staging Systems – W Agnese (USA)

11.15 – 11.30
C103 ALSFRS-R patterns of disease onset and progression through the spine – D Cerrato (USA)

11.30 – 11.45
C104 Autonomic dysfunction in ALS: Sympathetic overactivity predicts velocity of disease progression – G Mora (Italy)

11.45 – 12.00
C105 Making sense of the ALSFRS-R using joint longitudinal and survival models of functional dimension subscores – J Rooney (Ireland)

12.00 – 12.15
C106 In silico block randomization of ALS patients using a machine learning algorithm – J Berry (USA)

12.15 – 12.30
C107 Rate of change and linearity of ALSFRS-R and its subscales in the PRO-ACT database – N Thakore (USA)

12.30 – 13.45 LUNCH Locations: The Forum and Level 3 Foyer

SESSION 11 LOCATION: THE LIFFEY B

JOINT CLOSING SESSION

Chairs: T Heiman-Patterson (USA) and K Talbot (UK)

13.45 – 14.00
C108 Airlie House Clinical Trials Guidelines Workshop update 2016 – H Mitsumoto (USA)

14.00 – 14.30
C109 Entering the era of precision medicine: Realising the value of MND data at scale – W Hide (UK)

14.30 – 15.00
C110 A precision medicine approach to ALS: What will it take? – A Chio (Italy)

15.00 – 15.05
Poster Prize presentation

15.05 – 15.10
Invitation to Boston 2017

15.10 – 15.20
Late breaking news: Safety, tolerability and efficacy of intrathecal autologous mesenchymal stromal cells secreting neurotrophic factors (MSC-NTF) for patients with ALS from a Phase Ila randomized double blind placebo controlled trial – J Berry (USA)

15.20 – 15.30
Late breaking news: A Phase II trial of arimoclomol in SOD1 ALS – M Benatar (USA)
Poster sessions

THEME 1: Epidemiology

P1 Nutritional Therapy Guideline for amyotrophic lateral sclerosis
Okamoto K, Egami M, Fujinawa N, Kihiro T

P2 Season and time impact on ALS death: What might be the story behind it?
Pinto S, de Carvalho M

P3 Mortality rates for motor neuron disease at the state and county levels in the U.S. are associated with the use of well water

P4 BMAA ALS: is there a link between ALS and BMAA exposure?

P5 Environmental risk factors for ALS/neurodegeneration via aerosol exposure in NH, VT, and NY

P6 Association between alcohol consumption and risk of ALS in the Euro-MOTOR case-control study

P7 A population-based study on the impact of smoke and vascular risk factors on ALS outcome

P8 Occupational formaldehyde and amyotrophic lateral sclerosis: A population-based study in the Danish registries
Seals R, Hansen J, Gredal O, Weiskopf M

P9 Military service and the risk of amyotrophic lateral sclerosis: A meta analysis
Tal H, Cui YX, Shen DCH, Cui B, Fang J, Li DW

P10 Amyotrophic lateral sclerosis and the military: A population-based study in the Danish registries

P11 Psychiatric and neurodegenerative diseases among patients with amyotrophic lateral sclerosis and their families

P12 Establishing the true rates of familial ALS: A population based study over 20 years
Heverin M, Vajda A, Hardiman O

P13 Age of onset differentially influences the progression of regional dysfunction in sporadic amyotrophic lateral sclerosis

P14 Handedness and side of onset in limb-onset ALS: Is there a connection?
Piero E, Thakore N

P15 Effects of sex and family history on the amyotrophic lateral sclerosis (ALS) multistep model
Siu J, Perkins E, Cashman N

P16 Phenotypic characterization and clinical course of late and early disease onset in ALS patients
Garcia Molina E, Espinosa T, Dominguez R, Turon J, Povedano M, Paipa Merchán A

P17 Secure trends of ALS incidence in an Italian population-based register, 1995-2014, Is there a cohort effect?

P18 Mortality trends of amyotrophic lateral sclerosis in Norway 1951-2014: An age-period-cohort study
Nakken O, Lindstrom JC, Tynes OB, Holmøy T

P19 Amyotrophic lateral sclerosis incidence and cluster identification in New Brunswick, Canada, over a 10-year period
Jean J, O’Connell C, Wang H, McCullum S

P20 Prevalence of amyotrophic lateral sclerosis (ALS) in the United States, 2012-2013

P21 Incidence of motor neurone disease in the Scottish population: A 25 year perspective

P22 Co-morbidities in people with motor neurone disease/ALS
Sangheli A, Mills RJ, Young CA, on behalf of TONIC group

P23 The World Health Organisation disability assessment scale in the measurement of activity and participation in motor neurone disease/ALS
James E, Tennant A, Young C, on behalf of TONIC group

P24 Rates of decline in functional parameters and survival in ALS patients

P25 An examination of baseline factors predictive of patient completion status in ALS clinical trials
Liu D, Ferguson TA, Han S, Lindborg SR, Johns DR

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P49 MATR3 mutation analysis in a Chinese cohort of ALS cases
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P50 MATR3 mutation analysis in a Chinese cohort of ALS cases
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Two new risk loci for sporadic ALS in the Han Chinese population

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SOD1 mutations are the most common in Han Chinese populations with amyotrophic lateral sclerosis

Wei QQ, Zhou QQ, Chen Y, Ou R, Cao B, Xu Y, Yang J, Chen X, Shang H

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H46R SOD1 mutation is consistently associated with a relatively benign form of amyotrophic lateral sclerosis with very slow progression

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Common polymorphisms of the CX3CR1 gene are modifiers of ALS outcomes: A population-based study

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The HFE His63Asp polymorphism is a modifier of ALS outcomes in Italian and French patients with SOD1 mutations


Association analysis of polymorphisms in the VMA and TMEM106B genes in Parkinson’s disease, amyotrophic lateral sclerosis and multiple system atrophy in Chinese populations

Chen Y, Wei Q, Ou R, Cao B, Shang H

Novel mutation in the Vesicle-Trafficking Protein VAPB of one Chinese familial amyotrophic lateral sclerosis patient

Chen Y, Dong Y, Sun YM, Lu JH, Wu JJ

Mutations in FUS are the most frequent genetic cause in juvenile sporadic ALS patients of Chinese origin

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FUS mutations of familial amyotrophic lateral sclerosis patients in east China and their clinical characteristics

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A novel Optineurin truncation mutation identified in a consanguineous Palestinian family with Amyotrophic lateral sclerosis confirms loss of function as a disease mechanism


691_692insAG is a founder mutation of OPTN


Mutant Cyclin F inhibits endoplasmic reticulum (ER)-associated degradation (ERAD), ER-Golgi trafficking and autophagy, perturbing ER proteinostasis and inducing toxicity in ALS

Harmon M, Skehel P

An optical method for detecting endoplasmic reticulum and mitochondrial associated membranes

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A cellular model of ALS with SQSTM1 mutations exhibits autophagy defects; a new platform for evaluating genetic interactions and drug screening


Alteration of oligomeric states and subcellular localization of ALS1-LINKED motor neuron diseases


An optical method for detecting endoplasmic reticulum and mitochondrial associated membranes

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Mutant Cyclin F inhibits endoplasmic reticulum (ER)-associated degradation (ERAD), ER-Golgi trafficking and autophagy, perturbing ER proteinostasis and inducing toxicity in ALS

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P86 Impairment of growth hormone signalling significantly delays the onset of hind-limb weakness in a mouse model of ALS

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P94 Partial cytoplasmic mislocalization of truncated FUS leads to cellular autonomous late onset motor neuron degeneration

P95 Systemic delivery of Riluzole does not affect disease onset or increase lifespan in FUS (1-359) or TDP-43A315T mice
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P220 Monitoring amyotrophic lateral sclerosis disease progression with plasma creatinine levels
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P223a Monitoring amyotrophic lateral sclerosis disease progression with plasma creatinine levels
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P223b Monitoring amyotrophic lateral sclerosis disease progression with plasma creatinine levels
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P224 Monitoring amyotrophic lateral sclerosis disease progression with plasma creatinine levels
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P225b Monitoring amyotrophic lateral sclerosis disease progression with plasma creatinine levels
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P225c Monitoring amyotrophic lateral sclerosis disease progression with plasma creatinine levels
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P223 New insights into amyotrophic lateral sclerosis using quantitative magnetization transfer (qMT)
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P224 Effects of motor neuron disease progression on cortical beta rhythms: A single case study of amyotrophic lateral sclerosis using magnetoencephalography
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P225 Brain sodium MRI depicts upper motor neuron involvement in amyotrophic lateral sclerosis

P226 Altered functional connectivity in young-onset amyotrophic lateral sclerosis patients with early stage: a resting-state fMRI study
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P227 Selective white matter vulnerability in ALS: Implications for diagnostic classification
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P228 Prominent brain MRI white matter signal changes in an ALS patient
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P229 Susceptibility-weighted imaging as a potential biomarker in assessing whole brain gray matter damage in amyotrophic lateral sclerosis
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P230 Loss of tract integrity in amyotrophic lateral sclerosis: A longitudinal evaluation using MR diffusion tensor imaging

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P232 Postural instability in ALS is linked to cortico-basal ganglia and brainstem involvement
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P237 Deep phenotyping of frontotemporal dementia (FTD) and FTD-MND in Ireland (a clinico-based cohort longitudinal study)
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P238 Biophysical differences between motor axons in patients with amyotrophic lateral sclerosis and healthy controls

P239 Changes in peripheral axonal excitability in hereditary spastic paraplegia
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P240 Conduction slowing of peripheral motor nerves may occur in amyotrophic lateral sclerosis
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P241 The characteristics of F-waves in patients with Kennedy’s disease
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Sane H, Parapanje A, Sawant D, Inamdar S, Gokulchandran N, Badhe P, Sharma A

P267 Long-term G-CSF treatment on a named patient basis

P268 AlCAR treatment ameliorates skeletal muscle and neuromuscular junction pathology without any improvement in motoneuron survival and clinical outcome in a mouse model of spinal muscular atrophy
Cervérot C, Tarabal O, Piedrafita L, Casanovas A, Hernández S, Esquerra JE, Calderón J

P269 High speed FACS identifies novel neurotrophic factor combinations for disease-relevant motor neurons

P270 Stabilization of SOD1 native structure as a strategy for ALS prevention and treatment
Mohrülker J, Willbold D, Santer KB, Seimich M

P271 SOD1 reduction and preclinical efficacy of BIIB067, a SOD1 antisense oligonucleotide in Phase I testing in ALS

P272 Macrocyclic lactones, a novel drug class with therapeutic potential for ALS

P273 Gene therapy for familial ALS using AAV9-mediated silencing of mutant SOD1
Shaw P, McDearmid J, Ramesh T

P274 Increasing ALS clinical trial efficiency using group sequential trial designs

P275 Morpholino antisense oligomers as a therapeutic approach for amyotrophic lateral sclerosis

P276 TGF-β2 improved impaired neuromuscular transmission in the hG93A-SOD1 mouse model of motor neuron disease
McIntosh J, Miles GB, Bivin V

P277 Protective effects of novel engineered MET agonists on astrocyte-spinal neuron co-cultures from SOD1G93A transgenic mice

P278 MicroNeurotrophins improve survival in motor neuron-astrocyte co-cultures but do not improve disease phenotypes in a mutant SOD1 mouse model of amyotrophic lateral sclerosis

P279 Effect of CDNF administration in SOD1- G93A mouse model of amyotrophic lateral sclerosis

P280 Targeting of the retinoid pathway in SOD1G93A transgenic mice by delivery of engineered polymeric nanoparticles

P281 Selective knockdown of misfolded SOD1 through chaperone-mediated autophagy-based lysosomal degradation
Zhang X, Guan T, Li X, Wang Yu T, Namaka M, Marzan H, Jong K

P282 Impaired activity of Nrf2 is restored by Cu-ATSM

P283 Copper delivery by CuATSM derivatives and survival of SOD-G93A/CX3C5 mice
Beckman J

P284 Copper malfunction is common to sporadic MND and animal models of familial MND: Implications for copper-ATSM as a potential therapeutic and PET imaging agent

P285 The GSK-3 inhibitor AZD1080 delays onset and improves motor function in SOD1G93A transgenic mouse model of MND

P286 Characterization of the therapeutic potential of the potassium channel blocker 4-aminopyridine in ALS

P287 Interleukin-6 blockade improves inflammatory but not metabolic condition in ALS

P288 Targeting extracellular cyclinophilin A extends survival in the SOD1G93A mouse model of ALS

P289 Disease specific changes in sirtuin 3 levels in a mouse model of amyotrophic lateral sclerosis
Barth E, Bayer H, Hanselmann J, Wedyp P, Lindenberg KS, Witting A

P290 Riluzole rescues the early pre-clinical changes in ALS: Will early use of riluzole be beneficial in the clinic?

P291 Riluzole and a macrocyclic lactone restricts interneuron pathophysiology and delays the early motor dysfunction in the SOD1G93A mouse model of ALS: Is early intervention important in treating ALS?

P292 Histone deacetylase inhibitors enhance efficacy of drugs to maintain protein quality control
Durham H, Minotti S, Larochelle N, St. Louis K, Nalbantoglu J

P293 Efficient CNS targeting in adult mice by a single lumbar intrathecal AAV9-eGFP administration: application for neurological disorders
Bey K, Ciron G, Dubreil L, Deniaud J, Cristini J, Blouin V, Aouburg P, Colle MA

P294 Dosing and time course of targeted non-viral gene delivery to motor neurons in-vivo
Rogers ML, Smith KS, Turner BJ

P295 Cell-sheet transplantation in a mouse model of amyotrophic lateral sclerosis
Nakashish M, Une M, Watanabe Y, Nakashima K

P296 Pharmacologic and pathophysiologic readouts of C9orf72 therapy in iPS neurons
Hayes L, Gendron T, Petruzzelli L, Dixney M, Rothstein J

P297 Identification of small molecules with therapeutic relevance for C9orf72-ALS/FTD

P298 CRISPR/Cas9 genome editing results in precise genotypic and phenotypic correction in C9orf72 mutant IPSC-derived motor neurons

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Cognitive and Psychological Assessment and Support

P299 Communication in CLIS ALS patients using a vibrotactile p300 and motor imagery based brain computer interface
Spataro R, Allison B, Guger C, La Bella V

P300 Changes in cognitive profile during the course of 6 months in ALS

P301 Coping strategies, gender and disease subtype in MND/ALS
Young CA, Mills RJ, Tannant A, on behalf of TONIC group
P302 Cognitive changes and impact on clinical choices in ALS

P303 Overcoming verbal-motor limitations in ALS: A new Eye-Tracker based neuropsychological battery

P304 Selective attention in amyotrophic lateral sclerosis patients: Neuropsychological evaluation by using an eye-tracking system approach

P305 The relationship among onset type, neuropsychology and FDG-PET brain metabolic change in amyotrophic lateral sclerosis
Cui B, Cui L, Tai H, Shen D, Liu M, Li X, Li F

P306 Bulbar, motor and language impairment interactions in ALS

P307 The Edinburgh cognitive and behavioural ALS screen in a Chinese amyotrophic lateral sclerosis population
Ye S, Ji Y, Fan D

P308 Edinburgh Cognitive and Behavioural Amyotrophic Lateral Sclerosis Screen (ECAS) versus extensive neuropsychological examination: A comparative study in a Spanish ALS cohort
Cazorla S, Montoliu A, Salvado M, Jacas C, Gavira J

P309 Validation of the Edinburgh ALS Cognitive and Behavioural Screen (ECAS) in Canada

P310 Japanese version of the ALS-FTD-questionnaire (ALS-FTD-Q-J)
Watanabe Y, Beeldman E, Raaphorst J, the ALS-FTD-Q-J research group, Ito S, Adachi T, Takigawa T, Watanabe Y, Beeldman E, Raaphorst J

P311 Clinical usefulness of MoCA for the detection of cognitive impairment in amyotrophic lateral sclerosis patients
Nagashima K, Makika K, Fujita Y, Ikeda M, Ikeda Y

P312 New insight into the corticospinal correlates of extra-motor clinical profiles in non-demented ALS patients
Consonni M, Contaroni V, Catricalà E, Dalla Bella E, Bruzzone MG, Lauria G, Cappa SF

P313 Three single case study comparison of apathy on the FTD-MND spectrum

P314 POSTER WITHDRAWN

P315 Distinct patterns of cognitive behavioral change in emerging FTDL in the presence and absence of MND support: A 'bottom-up' model of FTDL onset
Kraft J, Flaherty C, Hotz A, Slinkard K, Simmons Z

P316 A population-based biopsychosocial investigation of caregiver quality of life in ALS
Burke T, Galvin M, Pinto-Grau M, Lonergan K, Hardiman O, Pender N

P317 Need for psychotherapy in patients with ALS and their relatives
Keck M, Kettemann D, Funke A, Kobel M, Meyer T

P318 Exploring patient and public involvement in motor neurone disease
Hobson EV, Musson L, McDermott CJ

P319 Stigma in people with motor neurone disease/ALS
Young CA, Mcsloy-Poli C, Taintant A, on behalf of TONIC group

P320 Depression and anxiety in people with MND/ALS
James E, Mills RJ, Taintant A, Young CA, on behalf of TONIC group

P321 Symptoms of psychological trauma resulting from being given a diagnosis of motor neurone disease
Marchant D, Goldstein LH, Al-Chalabi A

P322 Depression before the diagnosis in amyotrophic lateral sclerosis patients with cognitive dysfunctions: Two independent events or a preview of the same?
De Marchi F, Bersano E, Solara V, Mennilli MF, Cantello R, Mazzoni L

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Respiratory and Nutritional Management

P323 Using lung volume recruitment therapy in motor neurone disease (MND): A community service evaluation
Day H,Magee C

P324 Breathing stacking using a lung recruitment bag with ALS patients in north Scotland
Fraser D

P325 Impact of combined inspiratory-expiratory exercise on respiratory and bulbar function in an individual with ALS
Howman E, Robinson R, Tabor L, Wymes J

P326 Respiratory muscle unloading (RMU) to treat exertional related dyspnea (ERD) in ambulatory patients with Amyotrophic Lateral Sclerosis (ambALS)
Sanjak M, Mally D, Holsten S, Langford VL, Braver E, Brooks BR

P327 False negative, negative inspiratory tests in an ALS patient
Ross M, Burge M, Miller B, Dalrymple J, Wessels L

P328 Evaluation of the respiratory function in ALS patients by diaphragm echomography
Bongioanni P, Pelagatti A, Del Gamba C, Dolciotti C, Cavalli L, Santin M, Rossi B, Sartucci F

P329 Motor neuron disease: Assessment and management of respiratory complications
Humby J

P330 Diaphragm pacing: Survival data for patients implanted since FDA approval continues to show promising results
Onders R, Katriji B, Elmo M, Kaplan C, Schnitz R

P331 Novel trial design in a clinical study of diaphragm pacing (DPS) for ALS
Miller R, Ennist D, Thompson J, Fritz M, Moore D

P332 Clinical results of diaphragm pacing in Japanese patients with amyotrophic lateral sclerosis
Ito H, Fukutake S, Kohriki S, Kawachi J, Kamei T, Onders R

P333 The use of a hand held ventilator to supplement NIV for patients with respiratory insufficiency
Oliver D, Vincent-Smith L, Banerjee S, Kindred J, Martin K

P334 Changes in NIV ventilation over time - a longitudinal study
Oliver D, Banerjee S, Vincent-Smith L, Kindred J, Martin K, Rogers C

P335 Prolonged survival of non-invasive ventilation in Japanese patients with ALS
Hasegawa Y, Hirose T, Nakamura Y, Shigekiyo T, Tani H, Ishida S, Nakaiha M

P336 Spontaneous breath cycling is impaired in patients with ALS using non-invasive ventilation

P337 Differences in achievement of tidal volumes and rapid shallow breathing between PS and VAPS modes of non-invasive ventilation

P338 Effectiveness of automatic intratracheal suctioning system for amyotrophic lateral sclerosis patients with tracheostomy-invasive ventilation

P339 Spumon substance P concentration and peak cough experimental flow in patients with ALS after administration of enalapril

P340 Use of a water protocol in ALS patients with thin liquid dysphagia
Begg K, Marcoux C

P341 Dysphagia in amyotrophic lateral sclerosis and possible impact on riluzole management
Inghillini M, Onesti E, Schettino I, Gori MC, Frasca V, Cambieri C, Ceccanti M, Ruoppolo G

P342 Evaluating the potential of diet and food components as disease modifiers in amyotrophic lateral sclerosis (ALS)
Dawczynski C, Ringer TM, Prell T, Stubendorff B, Witte OW, Lorkowski S, Grosskreutz J

P343 Analysis of the interface between dysphagia and nutritional implications in patients with motor neuron disease
Alves PCL, Oda AL, Vecina ALL, Neves JWC, Oliveira ASB

P344 Percutaneous endoscopic gastrostomy in patients with ALS at the Ljubljana ALS Centre – a retrospective analysis

P345 Gastroscopy and survival in ALS patients
Kwan J, Domingo C, Diaz-Abad M, Epps D, Delussio A

P346 Gastroscopy placement in ALS patients—outcomes after changing clinical practice from percutaneous endoscopic gastrostomy (PEG) to radiographically inserted gastrostomy (RIG)

P347 Changing practice from radiologically inserted gastrostomy (RIG) to nasal unsedated seated gastrostomy (NuPEG): Our experience
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**Symptom Management**

P354 What DO people living with MND think about dysphagia? Lisiecka D, Kelly H, Jackson J

P355 The clinical utility of a self-reported swallowing outcome measure Sterling L, Allred P


P357 Comparison of driving capacity with distraction using the lane change task in drivers with amyotrophic lateral sclerosis compared with healthy controls Hayes H, Anderson D, Mathy P, Berggren K, Gibson S, Bromberg M

P358 Eye-tracking training for ALS patients using serious games Itó F, Kikuta S, Kawaguchi Y


P360 Mobility equipment use by people with ALS/MND in Australia Gibb R, Connors K, Mahony L, Davies R, Mathers S, Morgan P

P361 Common powered mobility components for the ALS/MND population: The US experience Allred P, Feldman S

P362 Telemedicine in ALS: Perspectives from the clinic team during a pilot program Morris A, Walsh S, Simmons Z

**THEME 13**

**Palliative Care and Decision Making**

P380 Support group for next of kin to patients diagnosed with ALS/MND Bjorkquist M, Stahl D

P381 The relationships between fatigue, sleep and disability in motor neurone disease Mills RJ, Tennant A, Young CA, on behalf of the TONIC study group

P382 Economic evaluations, cost studies and utility studies in motor neurone disease/amyotrophic lateral sclerosis: A systematic methodological review Moore A, Hughes D, Young CA, on behalf of the TONIC study group


P384 Personnel costs of a multidisciplinary ALS clinic Sawicki D, Drake K, Paganoni S, Berry J, Cudkowicz M

P385 Emotional experience in patients with amyotrophic lateral sclerosis withdrawing from long-term ventilation Kettemann D, Funke A, Maier A, Spittel S, Meyer T

P386 Determining the impact of a designated multidisciplinary care centre in Cambridge on the natural history and management of motor neurone disease Copsey H, Roberts R

P387 Motor neurone disease: Staff perspectives of the goal setting process within a community multidisciplinary team Prema R

P388 ALS medical needs and the regional medical resource survey in Japan Iwaki M, Komayakawa Y, Kira J

P389 How 95 percent of all Danish ALS patients use a national centre of rehabilitation expertise With H, Vaegter M, Brandstrup L, Jeppesen J

P390 Evaluation of the quality of life and the functionality scales in patients with MND Oda AL, Alves PCL, Oliveira ASB

P391 Attitudes towards life-prolonging measures and use of interventions in a multidisciplinary ALS clinic Saunders N, Salmon K, Magnussen C, Bertone D, Vitala A, Genge A

P392 Practice pattern of non-invasive ventilation and its impact on end-of-life care in patients with amyotrophic lateral sclerosis among Canadian care providers Chun M, Golton T, Shoesmith C

P393 Re-examining the utility value of mechanical ventilators to enable the long-term survival of ALS patients Hasegawa Y, Kirihara N, Nishida M, Sakai M

P394 Living organ donation in patients with amyotrophic lateral sclerosis Gibson S, Bromberg M, Ansari S
THEME BW

Biomedical Work in Progress

BW1 Multicenter data collection for assessing the natural history of ALS
Arcila-Londono X, Vader P, Walk D, Sherman A

BW2 Patterns of disease progression in SOD1 familial motor neuron disease: a retrospective study of 42 patients with long-term follow-up

BW3 The Icebucket Challenge Sporadic ALS
Australia Systems Genomics Consortium: SALSA-SGC

BW4 Assessment of variant carriers on whole-genome sequence and MiSEQ data of ALS patients
Iacangelo A, Sproviero W, Shatunov A, Al-Chalabi A, Jones A, Dobson R, Newhouse S, Al-Chalabi A

BW5 Integrating copy-number analysis with structural-variation detection in 50 ALS patients with two extreme survival phenotypes
Al-Khleifat A, Iacoangeli A, Shatunov A, Sproviero W, Al-Chalabi A

BW6 Genome-wide analysis of polymorphic tandem repeats through the development of a NGS method in a cohort of ALS patients

BW7 Shared novel variant analysis identifies novel genes in familial ALS from whole exome sequencing

BW8 Whole genome sequencing as a tool to unravel rare variants associated with ALS survival
Morsiani M, Robberecht W, Lambrechts D, Pulit S, van den Berg L, Veldink J, Consortium Project Mine Sequencing, Van Dammme P

BW9 Functional analysis of TDP43: Interaction with the epigenetic machinery
Sanna S, Esposito S, Masala A, Manca MA, Rassu M, Iacarrino C, Crocilo C

BW10 Genome-wide DNA methylation profiling in sporadic ALS

BW11 Multicentric referral-based study of ALS-related genes in an Argentine ALS/FTD cohort

BW12 Analysis of the protective effect of genetic admixture in amyotrophic lateral sclerosis
Mclaughlin R, Byrne R, Hardiman O

BW13 Functional and genetic characterisation of TBK1 mutations in a large cohort of familial ALS patients
de Majo M, Smith B, Gkazi A, Topp S, Nishimura A, Miller J, Vance C

BW14 Heterozygous deficiency of TBK1 in the high copy number SOD1-G93A transgenic mouse model
Brenner D, Bruno C, Ludolph AC, Weishaupt JH

BW14A A HuTDP-43△E31K mouse model shows signs of both motor neuron disease (MND) and frontotemporal dementia (FTD)

BW15 Humanising the Tardbp locus in the mouse

BW16 Understanding the link between cortical injury and ALS
Lagrimas A, Kozlowski D, Odziner PH, Jara J

BW17 Patterns of cortical atrophy at diagnosis in amyotrophic lateral sclerosis and implications on prognosis
Abualia M, Rafq M

BW18 An autopsy case of amyotrophic lateral sclerosis presented pallido-nigro-lusigen degeneration with TDP-43 pathology
Uchino A, Ogino M, Fujigasaki J, Nishiyama K, Murayama S

BW19 Assays of clinical importance in relation to the clinical course in ALS
Mitre Roperito B, Rosén H, Persson L

BW20 Bioenergetic profiling of cellular models of motor neuron disease to identify new approaches for supporting motor neuron health
Allen S, Francis L, Myszczynska M, Ferraiuolo L, Shaw P

BW21 Analysis of axonal transport in cultured neurons derived from an ALS mouse model by using the microfluidic cell culture system

BW22 Corf072 G4C2 HRE-mediated nucleocytoplasmic trafficking defects alters autophagic targeting
Marin J, Gleixner A, Marks M, Pandey U, Donnelly C

BW23 Systematic evaluation of the potential for repurposing autophagy targeting drugs in the treatment OF ALS-FTLD
Servante J, Scott D, Goode A, Cox A, Layfield R

BW24 Gene therapy for amyotrophic lateral sclerosis with migration of bone marrow-derived cells

BW25 A pilot study on the effects of plasma exchange with Albutein® 5% on motor and cognitive function of ALS patients

BW26 Cannabinoids for symptom management in amyotrophic lateral sclerosis: A pilot study
Magnussen C, Seguin R, O’Connell C, Genge A, Ware M

BW27 A registry-based randomized controlled, double-blinded clinical trial of Pimozide in patients with ALS

BW28 Interim results from an open-label, single-center, hybrid-virtual 12-month trial of a Lunasin regimen for patients with amyotrophic lateral sclerosis (ALS)

BW29 The Genervon case: Analysis and implications on the right-to-treat debate in ALS
Ringkamp G, Zoughalam A

THEME CP

Care Practice

CP1 Investigating the use of digital legacies with people affected by MND
Clabburm O, O’Brien M, Jack B, Knighting K

CP2 The Carers’ Alert Thermometer (CAT): Identifying the support needs of family carers of people living with MND
O’Brien M, Knighting K, Jack B, Fairfield H, Drinkwater N

CP3 Well-being and care burden of close relatives to persons with ALS-FTD
Grealad G, Hovmand B

CP4 Psychosocial support for ALS informal caregivers: Study protocol of a randomized controlled trial
de Witt J, Schroder C, Beelen A, van den Berg LH, Visscher-Meily A

CP5 A prospective study of quality of life in newly diagnosed ALS patients
Jakobsson Larsson B, Nygren I, Englund T

CP6 Influences on quality of life for people with MND/ALS: Progress of the trajectories of outcome in neurological conditions study
Young C, Dyas-Wollf L, Tennant A, on behalf of the TONIC group

CP7 People living with motor neuron disease facing their own mortality
Harris D

CP8 Preparing for end-of-life after Carter: A review of end-of-life experiences and perspectives of people with ALS, their families and health care providers

Moir M, Bubela T, Johnston W

CP10 Palliative care in a US Veterans Hospital
ALS Program: Structure, process and outcomes
Ratner E, Bradshaw K, Greenwood D
CP11 Streamlining primary care for veterans with amyotrophic lateral sclerosis  
Sluder K, Johnson J

CP12 Proving our worth – developing outcome measures for the motor neuron disease multi-disciplinary clinic  

CP13 Improving the clinic experience for patients: Decreasing wait time in multidisciplinary clinics  
Shahbazi M

CP14 Care, Audit, Research and Evaluation (CARE-MND) in the Scottish motor neuron disease population  

CP15 Scotland the Brave – Changes in funding of MND Clinical Specialists in Scotland  
Newton J, Seithell A

CP16 Supporting lifelong care in the community using the Long Term Conditions Register: Patient feedback  
Prema R, Canova C

CP17 The “Uppsala Model”: The care and treatment of MND patients at Uppsala University Hospital entails a multidisciplinary MND program  
den Dulk C, Franke C, Cidh Å, Banieghbal B

CP18 Physiotherapy, Occupational Therapy, Speech and Language Therapy in amyotrophic lateral sclerosis – experience of 5 years managed care in Germany  

CP19 Case study in research: A viable method for enhancing understanding of MND within Occupational Therapy  
Carey H

CP20 What drives driving habits in patients with ALS?  
Ciani G, Shabazi M, Holzberg S, Lange D

CP21 teleBCI – an online platform for brain-computer interface training  
Geronimo A, Simmons Z

CP22 A preliminary evaluation of exoskeletal training for ALS patients in an ambulatory setting  
Funke A, Kettemann D, Keck M, Spittel S, Meyer T

NEALS 1 The effects of Nuedexta on speech pause time  

NEALS 2 Mouthmeters: A tool for assessing bulbar motor involvement using a low-cost, 3D depth sensing system  
Green JR, Richburg BD, Markan S, Berry J

THEME CW

Clinical Work in Progress

CW1 Preliminary results from the Breathe MND-1 trial: Natural history of sleep disordered breathing in motor neuron disease; and randomised controlled trial of a new mode of non-invasive ventilation  

CW2 Review of personalised ventilation programmes and changes in pressure support over time in patients with MND/ALS  
Rogers C, Banerjee S, Oliver D

CW3 Understanding the use of noninvasive ventilation in the treatment of amyotrophic lateral sclerosis: Results of an international physician survey  

CW4 Does NIV and age influence survival rate in patients with amyotrophic lateral sclerosis: Experience in a multidisciplinary clinic - a retrospective review  
Magan N, Vitala T, Genge A, Salmon K

CW5 Body composition and disease progression in patients with motor neuron disease  
Salvoni C, Stanich P, Lellis R, Oliveira AB

CW6 Gain of body fat in amyotrophic lateral sclerosis patients: The great nutritional challenge  
Stanich P, Salvoni C, Lellis R, Oliveira AB

CW7 Understanding the impact of gastrostomy and quality of life in patients with ALS  
Ciani G, Holzberg S, Shahbazi M, Lange D

CW8 Clinical management of oral hygiene for patients with ALS  
McDonagh M, Riggs M, Banker-Horner L, Barkhaus P, Stich D, Domagala A, Fee D

CW9 Standard of Care for dysphagia management in ALS patients  
Epps D, Kitila M, Diaz-Abad M, Kwan J

CW10 Videofluoroscopic assessment of swallowing dysfunction in ALS  
Epps D, Alapati J, Kitila M, Diaz-Abad M, Kwan J

CW11 ALS Functional Communication Scale: A tool for standardizing and expanding speech therapy interventions and documenting improvement in communication  
Román A

CW12 Mobile technology towards automatic detection of early-stage ALS from short speech samples  
Wang J, Kothalkar P, Herndon B, Cao B, Heitzman D

CW13 Language changes in ALS: Preliminary results on a population-based study  
Pinto-Grau M, Burke T, Lonergan K, Murphy L, Elamin M, Hardiman O, Pender N

CW14 The role of neuropsychology within a multidisciplinary team in an acute care setting  
Meldrum S, Kersel D, Gorrie G

CW15 Neuropsychiatric symptoms in people living with motor neuron disease and their family members  

CW16 Application of neuropsychological measures for patients with amyotrophic lateral sclerosis (ALS)  
Parmentier M, Lange D, Shahbazi M

CW17 Investigating cognitive profiles in motor neuron disease – initial findings of the Cognitive And Behavioural Impairment in ALS (CABIA) study  

CW18 Cognitive impairment in amyotrophic lateral sclerosis (ALS): Screening tools, experiences and prognosis in Norway  
Taule T, Morland AS, Assmus J, Tynes OB, Rekand T

CW19 ALS Testing through Home-based Outcome Measures (The AT HOME Study)  
Shefner J, Mackline E, Narayanaswami P, Rutkove S

CW20 The ALS Early Recognition Timeline (ALERG) Project: Methods for a work in progress  
Nicholson K, Halsey K, Castro V, Gainer V, Murphy S, Schoenfeld D, Ferguson T, Atassi N

CW21 Feasibility and reliability of modified oculobulbar facial respiratory score (mOFRS) in amyotrophic lateral sclerosis (ALS) and sporadic inclusion body myositis (sIBM)  
Gebert N, Wencel M, Rai S, Tierry P, Mozaffar T, Goyal N

CW22 Modelling ALS progression using an artificial neural network-based computational system  
Uberti M, Galvani G, Vieide F, Doretti A, Maderna L, Peccarreta R, Silani V, Borgenova E, Ticozzo N

CW23 A retrospective study of patients diagnosed with amyotrophic lateral sclerosis and concurrent cases of peripheral thrombi  
Kaiser M, Holzberg S

CW24 The diagnostic yield of laboratory investigations in the work up for a suspected diagnosis of amyotrophic lateral sclerosis  
Minaan A, Korngut L

CW25 Serial high-density surface electromyography (HDSEMg) recordings in motor neuron disease: Fasciculations as a biomarker of motor neurone health  
Bashford J, Wickham A, Drakakis E, Bouteille M, Mills K, Shaw C

CW26 Skeletal muscle mri in spinal and bulbar muscular atrophy – a study in an animal model and patients  

CW27 A systematic review and meta-analysis of the diagnostic utility of cerebrospinal fluid biomarkers in motor neuron disease  
Akyol L, Soane T, Yeo JM, Green A, Chandran S, Pal S

CW28 Isolation and characterisation of circulating neurofilament-containing aggregates in health and disease  

CW29 Microglia cell-type specific NF-κB biomarker of motor neurone health  
Gebert N, Wencel M, Rahi S, Tierney P, Mozaffar T, Goyal N

CW30 High-affinity vital staining of microglia cell-type specific NF-κB in amyotrophic lateral sclerosis mouse model  
Béland L-C, Bouteil H, Kriz J

CW31 High-affinity vital staining of neuromuscular junctions for confocal endomicroscopy  
Resol C, Jones R, Dissanayake Y, Gillingwater TS, Skehel R, Ribchester R

POSTER SESSIONS | 27TH INTERNATIONAL SYMPOSIUM ON ALS/MND
# Programme of events

## Wednesday 7 December

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<td>07.00 – 19.00</td>
<td>Registration International Symposium</td>
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<td>Level 1</td>
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<tr>
<td>07.00 – 19.00</td>
<td>Speaker Room</td>
<td>Liffey Boardroom 3</td>
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<tr>
<td>08.30 – 10.15</td>
<td>Symposium Joint Opening Session</td>
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<tr>
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<td>Symposium Biomedical Sessions 2A/3A/4A</td>
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<tr>
<td>11.00 – 17.30</td>
<td>Symposium Clinical Sessions 2B/3B/4B</td>
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<tr>
<td>10.30 / 15.30</td>
<td>Refreshment breaks am/pm</td>
<td>The Forum and Level 3 Foyer</td>
<td>Ground and Level 3</td>
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<tr>
<td>12.30 – 14.00</td>
<td>Lunch</td>
<td>The Forum and Level 3 Foyer</td>
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<tr>
<td>12.30 – 14.00</td>
<td>Neurofilaments in the ALS clinic and beyond: From assays to clinical practise (closed meeting)</td>
<td>Liffey Meeting Room 2b</td>
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<td>13.00 – 14.00</td>
<td>Pan-Asian Consortium for Treatment and Research in ALS (PACTALS)*</td>
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<td>17.45 – 19.30</td>
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<td>19.30 – 21.45</td>
<td>Assessing disease progression in ALS: What are the most relevant measures?</td>
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## Thursday 8 December

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<td>08.30 – 17.30</td>
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<tr>
<td>08.30 – 17.40</td>
<td>Symposium Clinical Sessions 5B/6B/7B/8B</td>
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<tr>
<td>08.30 – 17.40</td>
<td>Symposium Alternative Sessions 5C/6C/7C/8C</td>
<td>Liffey Hall 2</td>
<td>Level 1</td>
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<tr>
<td>10.00 / 15.30</td>
<td>Refreshment breaks am/pm</td>
<td>The Forum and Level 3 Foyer</td>
<td>Ground and Level 3</td>
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<tr>
<td>12.30 – 14.00</td>
<td>Lunch</td>
<td>The Forum and Level 3 Foyer</td>
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<tr>
<td>12.30 – 14.00</td>
<td>Biogen External Investigator meeting</td>
<td>Liffey Meeting Room 2</td>
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<tr>
<td>17.45 – 19.30</td>
<td>Poster Session B</td>
<td>The Forum</td>
<td>Ground</td>
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<tr>
<td>18.00 – 19.00</td>
<td>Cochrane Neuromuscular/ALS Group</td>
<td>Liffey Meeting Room 2b</td>
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## Friday 9 December

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<td>Western ALS Investigator Meeting (closed meeting)</td>
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<tr>
<td>07.00 – 14.00</td>
<td>Registration International Symposium</td>
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<td>Symposium Joint Closing Session 11</td>
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</table>

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