28th international symposium on ALS/MND

Boston USA
8 - 10 December 2017

Programme

Hosts:
ALS Hope Foundation
ALS Therapy Development Institute

Organised by the Motor Neurone Disease Association in co-operation with the International Alliance of ALS/MND Associations
CME Accreditation
The 28th 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).
Welcome to Boston!

It is with great pleasure that the ALS Hope Foundation and the ALS Therapy Development Institute welcome you to Boston for the 28th International Symposium on ALS/MND. We invite you to explore this year’s extraordinary program.

Last year’s meeting welcomed more than 1100 attendees from 30 countries around the globe, underscoring its value to the global community. This year’s meeting will feature more innovative programming and educational offerings than ever before, giving you unparalleled access to groundbreaking research and a wide array of presentations on biomedical and clinical research advances in our field.

We would like to specifically recognize the participation of people with ALS/MND in that research. Thanks to their efforts of collaboration with scientists in both preclinical and clinical research endeavors, advancement in this fight can occur.

Boston is home to the greatest hospitals and universities in the world, as well as highly regarded pharmaceutical and biotechnology companies, cultural institutions and parklands. Boston embodies a remarkable tradition of high quality research and pioneering approaches to problems affecting healthcare. We live in what is described as ‘the intersection of urgent need and lasting impact’.

Our two organizations have entered into a partnership as local hosts, to facilitate this important event. Both founded in 1999, the ALS Hope Foundation funds the first multidisciplinary ALS clinic in the USA and the Neuromuscular Research Laboratory at Temple University College of Medicine in Philadelphia. ALS TDI is the world’s first and largest nonprofit biotech 100% dedicated to the discovery and development of effective treatments for ALS. Our organizations share the same mission: to see an end to this disease.

We hope your time in Boston for this year’s meeting leads to new ideas and helps spur innovation and partnership in this fight, in cooperation with scientists, organizations, funders and most importantly those we aim to serve: people living with ALS/MND.

Boston is a welcoming and diverse city: in your free time we encourage you to explore it!

Sincerely,

Terry Heiman-Patterson, M.D., Ph.D.,
President of the ALS Hope Foundation

Steve Perrin, Ph.D.,
CEO of the ALS Therapy Development Institute

Foreword

The 28th International Symposium begins with the question of how we define ALS/MND. This is probably not a discussion which would have been prominent at the time of the first Symposium, when most people would have considered it to be one biological entity, with the prospect of the same treatment for all those diagnosed with this devastating disease. Since then research has unearthed a bewildering level of biological complexity behind the clinical syndrome of ALS. As is usually the case in neurology, it pays to go back to clinical basics, and revisit the question of what we mean when we use the term ALS or MND.

Multiple biological pathways contribute to the process of neurodegeneration, and we still have a significant gap in our understanding of what initiates the pathological process. However, we have some major clues pointing to why motor neurons and their connecting networks are vulnerable. In this meeting you will hear from world leaders in the cell biology of ALS how RNA processing, stress granule assembly, neuroinflammation and other pathways are now credible targets for drug discovery.

Since the last meeting, spinal muscular atrophy, a lower motor neuron disorder mostly affecting children, has been the subject of landmark therapeutic trials using antisense oligonucleotides to alter the way in which the causative gene is processed. The results are remarkable and inspire genuine hope that ALS could be treated or even prevented in a similar way in the significant minority of patients carrying genetic mutations. The development of antisense based therapies for motor neuron diseases is therefore the subject of one of our plenary lectures.

In our clinical practice, we depend on evidence based medicine to guide us when making treatment decisions. However, we are increasingly faced with well-educated and highly informed patients who quite rightly find the delays in getting drugs from the ‘bench to the bedside’ frustrating, and for whom the idea of ‘trying anything’ is more attractive than ‘doing nothing’. In which circumstances should people be allowed to take treatments for which there is no evidence of benefit? Is the principle of ‘compassionate use’ a good reason to lower our scientific standards when assessing treatments? We explore this in a special session on pre-approval access to medicines.

It is great to be in Boston, the home of so much great ALS science over the years. In the 25 years since the discovery of SOD1 as the first genetic cause of the disease, progress has at times been slow, but there are now genuine reasons to believe we are at the beginning of a new era of ALS therapeutics.

Prof Kevin Talbot
Programme Committee Chair
Programme

Friday 8 December 2017

**SESSION 1 LOCATION: GRAND BALLROOM**

**JOINT OPENING SESSION**

_Chairs: S Light (UK) and K Talbot (UK)_

<table>
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<tr>
<th>Time</th>
<th>Event</th>
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<tbody>
<tr>
<td>08.45 – 09.00</td>
<td>Welcome – S Light (UK) K Talbot (UK) Welcome from Host Associations – S Perrin (USA)</td>
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<tr>
<td>09.00 – 09.45</td>
<td>C1 ALS/MND: Defining the disease – J Rosenfeld (USA)</td>
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**10.30 – 11.00 REFRESHMENTS AND NETWORKING:** Grand Ballroom Foyer and Galleria

<table>
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<tr>
<th>Time</th>
<th>Event</th>
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<tbody>
<tr>
<td>09.45 – 10.00</td>
<td>International Alliance Humanitarian Award International Alliance Forbes Norris Award</td>
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<tr>
<td>10.00 – 10.20</td>
<td>IPG Award and winner’s research presentation</td>
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**SESSION 2A LOCATION: MARINA BALLROOM**

**CELL BIOLOGY AND PATHOLOGY**

_Chairs: H Durham (Canada) and C Bendotti (Italy)_

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<tr>
<th>Time</th>
<th>Event</th>
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<tbody>
<tr>
<td>11.00 – 11.30</td>
<td>C2 The ‘lateral sclerosis’ half of ALS: Corticospinal (‘upper’) motor neurons from Charcot to their molecular development, diversity, circuitry, and growth cones – J Macklis (USA)</td>
</tr>
<tr>
<td>11.30 – 11.50</td>
<td>C3 Synapse dysfunction of layer V pyramidal neurons precedes neurodegeneration in a mouse model of TDP-43 proteinopathies – E Handley (Australia)</td>
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<tr>
<td>11.50 – 12.10</td>
<td>C4 Mechanisms of FUS mediated ALS: Insights from mouse genetics – L Dupuis (France)</td>
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<tr>
<td>12.10 – 12.25</td>
<td>C5 Functional analysis of the ALS-associated miR-1825 – A Helferich (Germany)</td>
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<td>12.25 – 12.40</td>
<td>C6 Identification of target mRNA transported to axons by TDP-43 – S Nagano (Japan)</td>
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**SESSION 2B LOCATION: GRAND BALLROOM A/B**

**AUTONOMY AND QUALITY OF LIFE**

_Chairs: T Heiman-Patterson (USA) and O Hardiman (Ireland)_

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<tr>
<th>Time</th>
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<tr>
<td>11.00 – 11.30</td>
<td>C7 Communication in serious illness: An evidence-based approach – J Tulsky (USA)</td>
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<td>11.30 – 11.50</td>
<td>C8 Health status perspective in ALS – S Pinto (Portugal)</td>
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<tr>
<td>11.50 – 12.10</td>
<td>C9 ALS patients with locked-in syndrome: Quality of life, depression and medical decision making – D Lulé (Germany)</td>
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<tr>
<td>12.10 – 12.30</td>
<td>C10 A systematic review of decision making among patients and their family in ALS care – G Foley (Ireland)</td>
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**12.30 – 14.00 LUNCH:** Grand Ballroom Foyer and Galleria
SESSION 3A LOCATION: MARINA BALLROOM

RNA BINDING AND TRANSPORT

Chairs: J Rothstein (USA) and L Van Den Bosch (Belgium)

14.00 – 14.30
C11 RNA-binding proteins and nucleocytoplasmic transport defects in ALS – L Van Den Bosch (Belgium)

14.30 – 14.50
C12 The nuclear pore complex is compromised in sALS and ALS/FTD – J Grima (USA)

14.50 – 15.10
C13 Safety and efficacy of SRSF1-dependent nuclear export inhibition of C9ORF72 repeat-transcripts: Moving towards therapies – L Castelli (UK)

15.10 – 15.30
C14 Phase separation of FUS is suppressed by the nuclear import receptor Transportin and FUS arginine methylations - D Dormann (Germany)

SESSION 3B LOCATION: GRAND BALLROOM A/B

TECHNOLOGY AND ALS

Chairs: T Meyer (Germany) and C McDermott (UK)

14.00 – 14.30
C15 Enhancing neurological care through telemedicine – R Dorsey (USA)

14.30 – 15.00
C16 Will telehealth revolutionize clinical care for ALS patients? – S Perrin (USA)

15.00 – 15.30
C17 BrainGate: Toward restoring communication and mobility – L Hochberg (USA)

15.30 – 16.00 REFRESHMENTS AND NETWORKING: Grand Ballroom Foyer and Galleria

SESSION 4A LOCATION: MARINA BALLROOM

RNA AND STRESS RESPONSE

Chairs: J Robertson (Canada) and C Shaw (UK)

16.00 – 16.30
C18 Neuroprotective effects of angiogenin-induced tRNA cleavage – P Anderson (USA)

16.30 – 16.50
C19 Mild chronic stresses sensitise neurons to the acute strong stress by reducing their capacity to maintain stress granule assembly – V Buchman (UK)

16.50 – 17.10
C20 Dynamics and nature of inclusions of TDP-43 and its isoforms – A Weichert (Canada)

17.10 – 17.30
C21 Role of RNA G-quadruplex structures in the molecular pathology of C9orf72-ALS – J Gallo (UK)

SESSION 4B LOCATION: GRAND BALLROOM A/B

CLINICAL TRIALS

Chairs: L van den Berg (Netherlands) and A Genge (Canada)

16.00 – 16.20
C22 Masitinib as an add-on therapy to riluzole is safe and effective in the treatment of ALS – J S Mora (Spain)

16.20 – 16.40
C23 VITALITY-ALS: Results of a phase 3 trial of tirasemtiv, a fast skeletal muscle troponin activator, as a potential treatment for patients with ALS – J Shefner (USA)

16.40 – 16.55
C24 Efficacy, safety and tolerability study of 1mg rasagiline in ALS: A prospective, randomized, parallel-group, double-blind trial – A Ludolph (Germany)

16.55 – 17.10
C25 Ibudilast: Bi-modal therapy with riluzole in early and advanced ALS patients – B Brooks (USA)

17.10 – 17.25
C26 Towards more efficient clinical trial designs in ALS: Lessons from the Edaravone Development Programme – J Palumbo (USA)

17.25 - 17.45
Discussion

LOCATION: GALLERIA 17.45 – 19.30

POSTER SESSION A

18.00 – 18.20
Theme 1: Genetics and genomics
Theme 2: In vitro experimental models

18.20 – 18.40
Theme 3: In vivo experimental models
Theme 4: Human cell biology and pathology

18.40 – 19.00
Theme 5: Epidemiology

19.00 – 19.20
Theme 6: Biomarkers
Theme BW: Biomedical work in progress
# 28th International Symposium on ALS/MND

## Saturday 9 December 2017

### SESSION 5A LOCATION: MARINA BALLROOM

**THERAPEUTIC STRATEGIES**

*Chairs: L. Bruijn (USA) and R. Mead (UK)*

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<th>Presenter(s)</th>
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<tbody>
<tr>
<td>08.30 – 09.00</td>
<td>C27</td>
<td>Antisense oligonucleotide-based therapies for motor neuron diseases – F. Bennett (USA)</td>
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<tr>
<td>09.00 – 09.30</td>
<td>C28</td>
<td>Improving drug access to the CNS – F. Walsh (USA)</td>
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<tr>
<td>09.30 – 09.45</td>
<td>C29</td>
<td>Harnessing machine learning and artificial intelligence to identify novel ALS therapeutics – M. Stopford (UK)</td>
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<tr>
<td>09.45 – 10.00</td>
<td>C30</td>
<td>Meta-analysis of pharmacogenetics interactions in ALS clinical trials – R. van Eijk (Netherlands)</td>
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### SESSION 5B LOCATION: GRAND BALLROOM A/B

**PRE-APPROVAL ACCESS**

*Chairs: R. Miller (USA) and S. Perrin (USA)*

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<tr>
<td>08.30 – 08.55</td>
<td>C31</td>
<td>Compassionate use of unapproved medicines: Law, ethics and policy – A. Bateman-House (USA)</td>
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<tr>
<td>08.55 – 09.20</td>
<td>C31A</td>
<td>Improving access to investigational treatment: It’s not about the FDA – J. Rabourn (USA)</td>
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<tr>
<td>09.20 – 09.45</td>
<td>C32</td>
<td>Neurologists’ views on ‘Right to Try’: Salem Witch Trials revisited – R. Bedlack (USA)</td>
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<tr>
<td>09.45 – 10.00</td>
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<td>Discussion</td>
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### SESSION 6A LOCATION: MARINA BALLROOM

**SOD1 ALS: FROM PATHOLOGY TO THERAPY**

*Chairs: J. Beckman (USA) and R. Brown (USA)*

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<tr>
<td>10.30 – 10.50</td>
<td>C33</td>
<td>The familial G93A SOD1 mutation alters intrinsic electrical properties and morphological development of cortical interneurons – T. Dickson (Australia)</td>
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<tr>
<td>10.50 – 11.10</td>
<td>C34</td>
<td>Misfolded SOD1 pathology in sporadic ALS – B. Paré (Canada)</td>
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<tr>
<td>11.10 – 11.30</td>
<td>C35</td>
<td>Distinct neuronal inclusions containing misfolded SOD1 in patients with mutations in C9ORF72 and other ALS- and FTD-associated genes – K. Forsberg (Sweden)</td>
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<tr>
<td>11.30 – 11.50</td>
<td>C36</td>
<td>Development of peptides that specifically recognize misfolded SOD1 proteins in ALS – E. Tokuda (Japan)</td>
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<tr>
<td>11.50 – 12.10</td>
<td>C37</td>
<td>Peptide-directed selective knockdown of misfolded SOD1 as a therapy for ALS – T. Guan (Canada)</td>
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<tr>
<td>12.10 – 12.30</td>
<td>C38</td>
<td>A promising small molecule lead in the search for a SOD1-targeted drug for ALS – G. Wright (UK)</td>
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### SESSION 6B LOCATION: GRAND BALLROOM A/B

**RESPIRATORY ASSESSMENT AND MANAGEMENT**

*Chairs: C. Jackson (USA) and M. de Carvalho (Portugal)*

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<tbody>
<tr>
<td>10.30 – 10.50</td>
<td>C39</td>
<td>Characterization of ALS patients based on MIP and FVC at first visit – K. Bommireddipalli (USA)</td>
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<tr>
<td>10.50 – 11.10</td>
<td>C40</td>
<td>Comparison between slow and forced vital capacities on survival prediction in ALS – S. Pinto (Portugal)</td>
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<tr>
<td>11.10 – 11.30</td>
<td>C41</td>
<td>How good is the respiratory subscore of ALSFRS-R? – N. Thakore (USA)</td>
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<tr>
<td>11.30 – 11.50</td>
<td>C42</td>
<td>Cough assist using the flow and pressure graphics to improve patient outcomes – J. Nilsestuen (USA)</td>
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<tr>
<td>11.50 – 12.10</td>
<td>C43</td>
<td>Mechanical insufflation exsufflation and lung volume recruitment in ALS: A prospective study of the prescription process, the outcomes and the experience – R. McConnell (Ireland)</td>
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<tr>
<td>12.10 – 12.30</td>
<td>C44</td>
<td>A US randomized trial of DPS in ALS: The outcome differs from two European trials – J. Katz (USA)</td>
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### SESSION 6C LOCATION: GRAND BALLROOM C-E

**BIOFLUID MARKERS**

*Chairs: A. Malaspina (UK) and R. Bowser (USA)*

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<tbody>
<tr>
<td>10.30 – 10.50</td>
<td>C45</td>
<td>Urinary p75 neurotrophin receptor extracellular domain: A biomarker relevant to ALS therapy development – M. Rogers (Australia)</td>
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<tr>
<td>10.50 – 11.10</td>
<td>C46</td>
<td>Blood and CSF neurofilament levels as biomarkers of pre-symptomatic disease – M. Benatar (USA)</td>
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<tr>
<td>11.10 – 11.30</td>
<td>C47</td>
<td>CSF pNfH as a diagnostic and prognostic biomarker in ALS: Experience with a colorimetric sandwich immunoassay – E. Gray (UK)</td>
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<tr>
<td>11.30 – 11.50</td>
<td>C48</td>
<td>Lipidomics reveals cerebrospinal-fluid signatures of ALS – H. Blasco (France)</td>
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<tr>
<td>11.50 – 12.10</td>
<td>C49</td>
<td>Unravelling phenotypic heterogeneity in ALS using quantitative proteomics: From animal models of the disease to human pathology – A. Malaspina (UK)</td>
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<tr>
<td>12.10 – 12.30</td>
<td>C50</td>
<td>Longitudinal analysis of the CSF proteome in ALS: Emerging microglial markers – A. Thompson (UK)</td>
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### 10.00 – 10.30 REFRESHMENTS AND NETWORKING: Grand Ballroom Foyer and Galleria

### 12.30 – 14.00 REFRESHMENTS AND NETWORKING: Grand Ballroom Foyer and Galleria
SESSION 7A LOCATION: MARINA BALLROOM

TDP-43

Chairs: M Polymenidou (Switzerland) and B Turner (Australia)

14.00 – 14.20
C51 Dynamic polymerization of TDP-43 in health and disease – M Polymenidou (Switzerland)

14.20 – 14.40
C52 TDP-43 splicing repression: Target identification and validation – P Wong (USA)

14.40 – 15.00
C53 A bona fide TDP-43 knock-in mouse demonstrates perturbed TDP-43 regulation and helps yield candidate suppressors of cognitive dysfunction in ALS-FTD – J Sreedharan (UK)

15.00 – 15.15
C54 Low expression of mutant Ubiquilin-2 exacerbates ALS-FTD features in a TDP-43 mouse model – V Picher-Martel (Canada)

15.15 – 15.30
C55 Unregulated expression of TDP-43 leads to divergent neurodegeneration in cortex and spinal cord in mice – Z Xu (USA)

SESSION 7B LOCATION: GRAND BALLROOM A/B

EPIDEMIOLOGY

Chairs: A Al-Chalabi (UK) and C Armon (USA)

14.00 – 14.20
C56 Presymptomatic lifestyle classified according to C9orf72 genotype – H Westeneng (Netherlands)

14.20 – 14.40
C57 Genetic mutations shorten the multistep process in ALS – A Chiò (Italy)

14.40 – 15.00
C58 ALS and food intake in Italy – E Pupillo (Italy)

15.00 – 15.15
C59 The effects of duration and intensity of cigarette smoking on the risk of ALS – S Peters (Netherlands)

15.15 – 15.30
C60 Spatial analysis of ALS cases in the United States and their proximity to multidisciplinary ALS clinics, 2013 – K Horton (USA)

SESSION 7C LOCATION: GRAND BALLROOM C-E

EMERGING MARKERS

Chair: M Weber (Switzerland) and P van Damme (Belgium)

14.00 – 14.20
C61 Imaging denervation in ALS for clinical trials: A longitudinal cohort study – T Jenkins (UK)

14.20 – 14.40
C62 Cortical excitability index: A novel diagnostic biomarker in ALS – N Geevasinga (Australia)

14.40 – 15.00
C63 Cortical dysfunction appears to be a regional feature in ALS – P Menon (Australia)

15.00 – 15.15
C64 Metabolic changes in asymptomatic C9orf72 carriers compared with non-carriers in the same family assessed by brain 7T MRSI – H Westeneng (Netherlands)

15.15 – 15.30
C65 The metabolic signature of ApoE genotype in ALS: A 18F-FDG-PET study – A Chiò (Italy)

15.30 – 16.00 REFRESHMENTS AND NETWORKING: Grand Ballroom Foyer and Galleria
SESSION 8A LOCATION: MARINA BALLROOM

GENETICS
Chairs: P Andersen (Sweden) and M Zatz (Brazil)

16.00 – 16.20
C66 Characterisation of a novel ALS-associated candidate gene identified from whole exome sequencing – C Shaw (UK)

16.20 – 16.40
C67 Targeted genetic screen of RNA-binding proteins in ALS reveals novel genetic variants with synergistic effect on clinical phenotype – J Cooper-Knock (UK)

16.40 – 16.55
C68 Integrating copy number analysis with structural variation detection in whole genome sequenced ALS UK cohort – A Al Khleifat (UK)

16.55 – 17.10
C69 Genome wide association study of genetic modifiers in ALS carriers of repeat expansions in C9orf72 gene – I Fogh (UK)

17.10 – 17.25
C70 DNA methylation age-acceleration is associated with disease duration and age at onset in C9orf72 patients – E Rogaeva (Canada)

17.25 – 17.40
C71 Discovery of previously unknown relationships between ALS patients increases power to identify causal disease genes – K Williams (Australia)

SESSION 8B LOCATION: GRAND BALLROOM A/B

DISEASE MANAGEMENT
Chairs: C Young (UK) and Z Simmons (USA)

16.00 – 16.20
C72 NEALS Bulbar Subcommittee: Protocol design for speech and swallowing – G Pattee (USA)

16.20 – 16.40
C73 The Oral Secretion Scale (OSS) predicts tolerance of noninvasive ventilation (NIV), the need for hospice or transition to tracheostomy ventilation (TV) and prognostic factors for survival in patients with ALS/MND – P Cazzolli (USA)

16.40 – 17.00
C74 Trial of resistance and endurance exercise in ALS – N Maragakis (USA)

17.00 – 17.20
C75 The 100 collars project: A multi-centre evaluation of the HeadUp cervical orthosis – C McDermott (UK)

17.20 – 17.40
C76 Treatment for cramps in ALS/MND: An updated Cochrane review – B Oskarsson (USA)

SESSION 8C LOCATION: GRAND BALLROOM C-E

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

16.00 – 16.20
C77 Tracing disease progression in ALS: A multimodal longitudinal imaging study of structural brain involvement – R Walhout (Netherlands)

16.20 – 16.40
C78 Patterns of cortical atrophy in ALS and implications on prognosis – M Rafiq (UK)

16.40 – 17.00
C79 Perfusion imaging signatures of pathological spread across TDP-43 proteinopathies – P Ferraro (USA)

17.00 – 17.20
C80 Spinal cord gray matter atrophy as MRI biomarker for ALS patients – P Pradat (France)

17.20 – 17.40
C81 Hypothalamic structure alterations in presymptomatic and symptomatic ALS – M Gorges (Germany)

LOCATION: GALLERIA 17.45 – 19.30

POSTER SESSION B

18.00 – 18.20
Theme 7: Electrophysiology
Theme 8: Imaging

18.20 – 18.40
Theme 9: Clinical trials
Theme 10: Therapeutic strategies

18.40 – 19.00
Theme 11: Improving diagnosis, prognosis and disease progression
Theme 12: Cognitive and psychological assessment and support

19.00 – 19.20
Theme 13: Respiratory and nutritional management
Theme 14: Multidisciplinary care and improving quality of life
Theme CW: Clinical work in progress
Sunday 10 December 2017

SESSION 9A LOCATION: MARINA BALLROOM
NEUROINFLAMMATION AND GLIAL SIGNALLING
Chairs: P Crouch (Australia) and L Barbeito (Uruguay)
08.30 – 09.00
C82 Imaging glial activation in people with ALS – N Atassi (USA)
09.00 – 09.20
C83 MicroRNAs secreted by C9orf72 patient-derived astrocytes contribute to impairment in axonal growth and cell death in vitro – L Ferraiuolo (UK)
09.20 – 09.40
C84 Slowing disease progression in the SOD1 mouse model of ALS by blocking neuregulin-induced microglial activation – F Song (USA)
09.40 – 10.00
C85 The role of microglia in TDP-43 clearance and redistribution in the zebrafish spinal cord – M Morsch (Australia)

SESSION 10A LOCATION: MARINA BALLROOM
INTEGRATED OMICS AND PATHWAY ANALYSIS
Chairs: P Shaw (UK) and J Veldink (Netherlands)
10.30 – 11.00
C91 Analyzing biological networks to identify novel disease pathways – E Fraenkel (USA)
11.00 – 11.30
C92 Early analysis of clinical and iPSC motor neuron multi-omics signature from a large population of sporadic and familial ALS patients reveals verifiable subgroups and molecular pathways – J Rothstein (USA)
11.30 – 11.50
C93 Molecular phenotyping of human neurons with TDP-43 pathology reveals derepression of transposable elements – E Lee (USA)
11.50 – 12.10
C94 Extensive RNA sequencing study in brain tissue obtained from patients harbouring a C9ORF72 repeat expansion – M van Blitterswijk (USA)
12.10 – 12.30
C95 Integrated molecular landscape of ALS provides insights into disease etiology – G Poelmans (Netherlands)

SESSION 9B LOCATION: GRAND BALLROOM A/B
CORRELATES OF CLINICAL PROGRESSION
Chairs: J Rosenfeld (USA) and L Cui (China)
08.30 – 08.50
C86 Hypermetabolism is associated with lower motor neurone burden, functional decline and predicts survival in ALS – F Steyn (Australia)
08.50 – 09.10
C87 Lipid metabolism and survival across the ALS-FTD spectrum – R Ahmed (Australia)
09.10 – 09.30
C88 Clinical characteristics and associated factors in ALS patients with longer survival – Q Wei (China)
09.30 – 09.45
C89 Biomarker mixtures predict ALSFRS-R at time of diagnosis – P Factor-Litvak (USA)
09.45 – 10.00
C90 Blood vitamin D levels correlate with ALS severity: A prospective study – N Pageot (France)

SESSION 10B LOCATION: GRAND BALLROOM A/B
COGNITIVE CHANGE
Chairs: M Kiernan (Australia) and C Lomen-Hoerth (USA)
10.30 – 11.00
C96 The ALS-FTD continuum – C Lomen-Hoerth (USA)
11.00 – 11.20
C97 A population-based study of cognition in the ALS-FTDS: The incidence and nature of language changes – M Pinto-Grau (Ireland)
11.20 – 11.40
C98 Language is the cognitive function which is most vulnerable to change in ALS – D Lulé (Germany)
11.40 – 12.00
C99 Behavioural changes in bvFTD and ALS-FTD: A prospective study – J Saxon (UK)
12.00 – 12.20
C100 Longitudinal assessment in ALS using the Edinburgh Cognitive and Behavioural ALS Screen (ECAS) – B Poletti (Italy)
12.20 – 12.40
C101 Development and clinical implications of the brief Dimensional Apathy Scale (b-DAS) – R Radakovic (UK)

10.00 – 10.30 REFRESHMENTS AND NETWORKING: Grand Ballroom Foyer and Galleria

SESSION 11 LOCATION: GRAND BALLROOM A/B
JOINT CLOSING SESSION
Chairs: K Talbot (UK) and S Chandran (UK)
14.00 – 14.40
C102 ALS: Lessons from SOD1 and prospects for SOD1 and C9orf72 gene silencing – R Brown (USA)
14.40 – 14.45
Poster Prize presentation
14.45 – 14.55
Patient Impact Award
14.55 – 15.05
Invitation to Glasgow 2018
15.05 – 15.15
Late breaking news

12.30 – 14.00 LUNCH: Grand Ballroom Foyer and Galleria
Posters sessions

THEME 1

Genetics and Genomics

GEN-01 Intermediate-length CAG repeat in ATXN2 is associated with later onset in Brazilian patients with amyotrophic lateral sclerosis

GEN-02 Does the P56S- VAP-B mutation decrease the cancer risk in ALS patients?

GEN-03 Homozygous mutations in ALS in a homogeneous population

GEN-04 The identification of novel mutations causing familial ALS and the elucidation of common disease mechanisms
Salman M, Morris A, Topp S, Smith B, Shaw C, de Bellerocche J

GEN-05 A comprehensive analysis of telomere length in ALS
Al Khleifat A

GEN-06 Occurrence of multiple mutations in ALS-associated genes in an Italian cohort

GEN-07 TBK1 mutations in Italian patients with amyotrophic lateral sclerosis: genetic and functional characterization

GEN-08 TBK1 variants in Chinese familial and sporadic patients with amyotrophic lateral sclerosis
Liu X, He J, Li J, Chen L, Zhang N, Liu X, Ma Y, Fan D

GEN-09 NEK1 Arg261His variant is rare in amyotrophic lateral sclerosis from China
Gu X, Chen Y, Zhou Q, Shang H

GEN-10 Co-occurrence of amyotrophic lateral sclerosis in the index patient of a X-linked Charcot-Marie-Tooth type 1 pedigree in mainland China
Zou Z, Feng S

GEN-11 High-throughput sequencing revealed a SETX mutation in a Chinese patient with sporadic amyotrophic lateral sclerosis
Zou Z, Che C, Liu C, Huang H

GEN-12 SCFδ1 genes variant rs10391514 could modulate ALS phenotype in a Chinese cohort
Chen Y, Zhou Q, Wei Q, Shang H

GEN-13 CHCHD10 mutations in patients with amyotrophic lateral sclerosis in mainland China
Shen S, He J, Tang L, Zhang N, Fan D

GEN-14 Next generation sequencing of 41 ALS related genes in Chinese ALS patients
Li X, Cui L, Liu M, Ding Q, Guan Y, Zhang X, Liu Q

GEN-15 Targeted next-generation sequencing of ALS-related genes in Indian ALS patients

GEN-16 Investigating CCNF mutations in a Taiwanese cohort with amyotrophic lateral sclerosis
Lee Y-C, Liao Y-C

GEN-17 The SMN2 gene copy number states can affect the onset risk and survival time in Japanese ALS

GEN-18 A multidisciplinary approach to identify novel familial amyotrophic lateral sclerosis genes

GEN-19 Genome-wide and targeted analysis of DNA methylation in C9orf72 and SOD1 ALS/FTD cohorts

GEN-20 Local population structure correction in ALS rare variant analysis
Byrne R, Martiniano R, Gerard Bradley D, Hardiman O, McLaughlin R

GEN-21 Identical twins discordant for motor neuron disease: insights from genome and methyleome data

GEN-22 The project MinE data browser: bringing whole-genome sequencing data in ALS to researchers and the public
Project MinE

GEN-23 A high throughput gene, environment and epigenetics database and analysis system for international ALS research
Lacapreggia A, Newhouse S, Dobson R, Al-Chalabi A

GEN-24 Facilitating data transfer from a patient community to the ALS online genetics database (ALSoD)
Wicks P, Cerrato D, Martin S, Kulka A, Abel O

GEN-25 Polygenic link between blood lipids and amyotrophic lateral sclerosis
Chen X, Yazdani S, Piehl F, Magnusson PK, Fang F

GEN-26 Research of C9orf72 intermediate-length GGGGCC repeat expansions in Chinese patients with amyotrophic lateral sclerosis
Wang A, He J, Tang L, Zhang N, Fan D

GEN-27 Intermediate C9orf72 repeat numbers are not ALS risk factors

GEN-28 Analysis of C9orf72 repeat expansion in a large cohort of Italian ALS patients and its association with a poor prognosis
Mosca L, Tarlati C, Lizio A, Riva N, Sansone V, Lunetta C

GEN-29 Age-related penetration of the C9orf72 repeat expansion
Murphy N, Arthur K, Tienari P, Houlden H, Chio A, Traynor B

GEN-30 Burden analysis of ALS-gene variants in patients with and without C9orf72 expansion

GEN-31 Genetic testing of sporadic ALS patients reveals pathogenetic mutations in non-ALS genes
Valente M, Zucca S, Palmieri I, Garau J, Rey F, Gagliardi S, Diamanti L, Cerioni M, Cereda C

GEN-32 POSTER WITHERDRAWN

GEN-33 Investigation of antisense long non-coding RNAs in sporadic amyotrophic lateral sclerosis patients
Gagliardi S, Zucca S, Arigoni M, Pandini C, Diamanti L, Pansarasara O, Cerioni M, Calogero R, Cereda C

GEN-34 Genetic factors associated with frontal disease in ALS: neuroimaging and neuropsychological evidence

GEN-35 A novel p.Leu106fs*15 SOD1 mutation leading to the formation of a premature stop codon in an apparently sporadic ALS patient: insights into the underlying pathomechanisms

GEN-36 Clinical and experimental studies of a novel p.S258R FUS mutation in amyotrophic lateral sclerosis

GEN-37 The difference of molecular mechanisms between SOD1G41S and G41D in ALS mouse neuroblastoma N2a cell models
Shi M, Zhao H, Niu Q, Jin Q

GEN-38 Identification of disease modifier genes in the g93A5SOD1 mouse model of amyotrophic lateral sclerosis (ALS)
Heiman-Patterson T, Blankenhorn E, Cort L, Hennelsey N, Dixon M, Alexander G

GEN-39 The role of DPRs in C9orf72 pathology: from ES cells to mouse models
Milioto C, Devoy A, Davies B, Fisher E, Isaacs A

GEN-40 Identification of common protective biological pathways and processes in amyotrophic lateral sclerosis models in D. melanogaster

GEN-41 Characterizing the C9orf72 hexanucleotide repeat expansion in neurologically normal Caucasian population
Lin Z

THEME 2

In Vitro Experimental Models

IVT-01 Neurotoxicity of the cyanotoxin BMAA through axonal degeneration and intercellular spreading
Barbagallo P, Candalija A, Cowley S, Dafinca R, Talbot K

IVV-45 Inosine supplementation increases C9orf72 astrocyte ATP levels and leads to increased motor neuron survival
Allen S, Hall B, Myszczynska M, Ferraiuolo L, Shaw P

THEME 3
In Vivo Experimental Models

IVV-01 Increased aggregated SOD1 in spinal cord from SOD1(G93A) transgenic mice correlates with later disease onset and improved longevity

IVV-02 Misfolded SOD1 levels in the blood of SOD1(G93A) transgenic mice as indicators of ALS disease progression

IVV-03 Pharmacological inhibition or genetic ablation of complement C5a receptor, C5aR1, ameliorates disease pathology in the hSOD1G93A mouse model of amyotrophic lateral sclerosis
Lee J, Kumar V, Fung J, Rutenberg M, Noakes P, Woodruff T

IVV-04 Ablation of free fatty acid receptor 2 (FFAR2) signaling accelerates early disease progression in the SOD1G93A mouse model of amyotrophic lateral sclerosis

IVV-05 Fibroblast growth factor-2 (FGF-2) dependent interplay of neurotrophic factors and signalling cascades in amyotrophic lateral sclerosis
Kefalakes E, Sarikidi A, Bursch F, Hersel N, Grothe C, Petri S

IVV-06 The influence of neurotrophic factors on in vivo axonal transport in the SOD1G93A mouse
Tosolini A, Sleigh J, Schiavo G

IVV-07 Muscle type specific abnormalities in terminal Schwann cell morphology following partial denervation in the SOD1 G93A mouse
Harrison J, Rafuse V

IVV-08Astrocyte-derived extracellular vesicles contribute to the propagation of pathogenic proteins in ALS
Endo F, Yamanaka K

IVV-09 Innate immune adaptor TRIF slows disease progression of ALS mice by eliminating aberrantly activated astrocytes
Komine O, Yamashita H, Fujimori-Tonou N, Uematsu S, Akira S, Yamanaka K

IVV-10 The NF-kB signaling pathway is activated by the SOD1-G93A mouse model of amyotrophic lateral sclerosis by eliminating aberrantly accumulated SOD1 in spinal cord from SOD1(G93A) transgenic mice correlates with later disease onset and improved longevity

IVV-11 Circadian rhythm dysfunction accelerates disease progression and increases intestinal cyano bacteria in an amyotrophic lateral sclerosis model
Huang Z, Liu Q, Dai J, He B, Xie Y, Yao X, Su H

IVV-12 Beneficial effect of oxaloacetate for the neuromuscular function of SOD1G93A mice

IVV-13 Increasing urate levels may delay disease onset in the SOD1 G93A mouse model of amyotrophic lateral sclerosis

IVV-14 Identifying molecular drivers of ALS in an ALS model: failed recovery and enhanced ventral horn inflammation after peripheral nerve injury
Bracken B, Song F, Kems J, Gonzalez M, Loeb J

IVV-15 Early dysfunction of premotor glycinergic interneurons in a zebrafish model of amyotrophic lateral sclerosis
Cielo G, Shaw P, Ramesh T, McDearmid J

IVV-16 Corticospinal motor neuron degeneration precedes spinal motor neuron degeneration and involves a new set of molecular players

IVV-17 Environmental and genetic contributions in an ALS model: failed recovery and enhanced ventral horn inflammation after peripheral nerve injury
Schram S, Song F, Kems J, Gonzalez M, Loeb J

IVV-18 Environmental and genetic contributions in an ALS model: failed recovery and enhanced ventral horn inflammation after peripheral nerve injury
Schram S, Song F, Kems J, Gonzalez M, Loeb J

IVV-19 Identifying molecular drivers of ALS in transgenic TDP-43 mice

IVV-20 Using in-cell NMR to study the protein folding and structural dynamics of TAR DNA binding protein-43
Alam SM, Deckert A, Wang X, Cabrita LD, Christodoulou J

IVV-21 Identifying physiologically relevant targets of TDP-43 translational regulation
Lehmkuhl E, Siddeogowda BB, Alsup E, Joardar A, Jensen K, Zarnescu D

IVV-22 P13K/mTOR pathway rescues TDP-43 toxicity in the spinal motor neuron in zebrafish
Akawara K, Kawakami K

IVV-23 ALS as an aggregate pathological phosphorylation of Thr175 tau induces a tau proteinopathy in vivo

IVV-24 CSF and serum neurofilament light chain levels as a biomarker for diagnosis and disease progression in a canine disease model of ALS
Patze H, Sah O, Toedebusch C, Coates J

IVV-25 Embryonic exposure to the environmental neurotoxin BMAA negatively impacts early neuronal development and progression of neurodegeneration in the Sod1-G93R zebrafish model of amyotrophic lateral sclerosis

IVV-26 Assessing the role of sense and antisense foci in Drosophila models of C9orf72 ALS/FTD

THEME 4
Human Cell Biology and Pathology

HCB-01 Mosaicism of repeat expansions in C9orf72 modulates its expression and produces RNA foci and dipeptide repeat proteins in a 90-year-old case without neurodegeneration

HCB-02 The C9orf72 protein regulates actin dynamics in neurons
Jagaraj C, Sundaramoorthy V, Walker A, Atkin J

IVV-27 Activation of BMP signaling in non-motor neurons rescues motor dysfunction in a Drosophila model of amyotrophic lateral sclerosis
Held A, Major P, Lipscombe D, Wharton K

IVV-28 Stress leads to neurodegeneration in single-copy models of amyotrophic lateral sclerosis in C. elegans
Baskoylu S, Yersak J, O’Hearn P, Grosser S, Simon J, Hart A

IVV-29 Identification of suppressors of stress-induced neurodegeneration in a knockin SOD1 model
Yanagi K, Lins J, Stinson L, Walsh M, Mahapatra A, Char S, Hart A

IVV-30 Profound muscular pathology in mice expressing WT and F115C mutant matrin 3 is not directly linked to motor dysfunction

IVV-31 Disease models of ALS/FTD — a human pathological perspective
Tan R, Ke Y, Ittner L, Halliday G

IVV-32 Micro-CT for non-invasive evaluation of muscle wasting in mouse models
Pasetto L, Olivari D, Nardo G, Chiara Troiele M, Bendotti C, Piccirillo R, Bonetto V

IVV-33 Modelling ALS in the visual system

IVV-34 Scientific background for developing oral levisimendan (ODM-109) for the treatment of amyotrophic lateral sclerosis

IVV-35 Lead identification and optimization in an in vivo tunicamycin assay
Hatzipetros T, Tassinari V, Kidd J, Moreno A, Thompson K, Vieira F

IVV-36 Development of an AAV gene therapy targeting SOD1 for the treatment of ALS: translation of delivery
HCB-03 Investigation of arginine methylation of poly-2GR inclusions in C9orf72 FTD patient cortex

HCB-04 Neuropathological characterization of the motor cortex in familial and sporadic amyotrophic lateral sclerosis

HCB-05 Heterogeneity of pathology in the primary motor cortex of amyotrophic lateral sclerosis and post mortem MRI correlates
Gamaralage MP, Menke RAL, Foxley S, Jenkinson M, Qi F, Tendler D, Turner MR, Miller K, Ansorge O

HCB-06 A novel TDP-43 mutation: the role of TDP-43 RNA binding in TDP-43 proteinopathy and ALS
Chen H-J, Topp S, Hui HS, Smith B, Katarya M, Shaw C

HCB-07 TDP-43 is ubiquitylated by the Skp1-cullin-cyclop E3 ubiquitin ligase complex
Rayner S, Lee A, Williams K, Blair I, Molloy M, Chung R

HCB-08 Differential expression of small RNAs in muscle tissue of patients with amyotrophic lateral sclerosis

HCB-09 Validation of IBM Watson’s prediction of heterogeneous nuclear ribonucleoprotein U as a novel protein linked to ALS
Vu L, Bakkar N, Bowser R

HCB-10 Phosphorylation by casein kinase 2 regulates the RNA-binding activity of FUS

HCB-11 A novel method for understanding the mutant FUS inclusion interactome demonstrates sequestration of proteins critical for mRNA metabolism
Kamalgar M, Chen J, Gal J, Zhu H

HCB-12 Characterization of FUS post-translational modifications
Arenas A, Gal J, Zhu H

HCB-13 Mutant FUS disrupts immune response via abnormal regulation of paraspeckle components
An H, Kukharsky M, Highley JR, Buchman V, Shilkovnikova T

HCB-14 Histoine deacetylase expression is altered in amyotrophic lateral sclerosis

HCB-15 Alterations in hippo/YAP signaling as a pathogenic mechanism in amyotrophic lateral sclerosis
Mueller K, Granucci E, Dios A, Berry J, Vakili K, Sadri-Vakili G

HCB-16 Motor neuron disease-associated mutations in the mitochondrial protein CHCHD10 act by loss of function

HCB-17 Itoathy dysfunction in PBMCs of sporadic ALS patients
Bordoni M, Pansaras O, Fantini V, Crippa V, Garau J, Diamanti L, Ceroni M, Cereda C

HCB-18 Initiation, propagation and inhibition of SOD1 misfolding in amyotrophic lateral sclerosis
Pokrishesvsky E, Hong RH, Nan J, Cashman N

HCB-19 Copper dysfunction unifies sporadic ALS and progressive multiple sclerosis
Hilton J, Kyseniuk K, Mercer S, Roberts B, Hare D, McLean C, Donnelly P, White A, Crouch P

HCB-20 Iron accumulation in the sporadic ALS-affected motor cortex: ceruloplasmin, biomarkers and ferroptosis

HCB-21 Declines in synaptic adhesion and MusSK signaling at neuromuscular synapses may underlie early muscle weakness in ALS patients

HCB-22 ERVK integrase impairs anti-viral IRF3 signaling in ALS
Mainghera M, Douville R

HCB-23 Changes in brainstem cytokines in normal ageing and motor neuron disease
Tennakoon A, Johnson I, Katheresan V

HCB-24 Complex inflammation response in ALS: based on a case and control study
Chen Y, Gu X, Shang H

HCB-25 Localizations of activated microglia/macrophages and dendritic cells are distinct in ALS spinal white matter and exert different impacts on clinical progression
Hayashi S, Yamasaki R, Okamoto K, Murai H, Kira J-I

HCB-26 Identification of distinct extracellular vesicles in plasma of ALS patients

HCB-27 Role of vesicle secretion in ALS transmission

HCB-28 Secretion of toxic exosomes by muscle cells of ALS patients: role in ALS pathogenesis

HCB-29 Chronic traumatic encephalopathy within the motor cortex of amyotrophic lateral sclerosis and water quality in northern New England

HCB-30 Generating a collection of induced pluripotent stem cells from primary fibroblast cultures isolated from ALS patients
Tsolias A, Dane T, Maxwell M, Lukashev M

THEME 5
Epidemiology

EPI-01 Amyotrophic lateral sclerosis in Africa: a multi-center cohort study, ALS and related syndromes under the tropics (TROPALS) collaboration

EPI-02 Spatial correlation of ALS mortality and the role of environmental variables in Chile
Zinco P, Valenzuela D, Lillo P

EPI-03 The incidence of amyotrophic lateral sclerosis (ALS) and its influence factors in Beijing, China, 2010-2015
Zhou S, Qian S, Chang W, Wang L

EPI-04 An odyssey in epidemiology - from snow to hill to ALS: acquired somatic mutations may trigger ALS onset
Armon C

EPI-05 The Epidemiology of ALS in Massachusetts, 2008—2012: results from the first comprehensive capture, population-based ALS registry in the US
Fraser A, Abille V, Paganoni S, Berry J, Atassi N, Chad D, Nicholson K, Knorr R

EPI-06 National amyotrophic lateral sclerosis (ALS) biorepository, USA
Kaye W, Wagner L, Stein T, Traynor B, Orr M

EPI-07 Designing and implementing an international web-based questionnaire to look for risk factors for ALS/motor neuron disease
Kullmann JP, Pamphlett R

EPI-08 ARREST ALS: an extension of the ALS COSMOS study

EPI-09 MND Register for England, Wales and Northern Ireland
Martin S, Ossher L, Kulka A, Talbot K, Al-Chalabi A

EPI-10 The Swedish MND quality registry

EPI-11 POSTER WITHDRAWN

EPI-12 Epidemiological survey of SBMA in Italian north-east regions
Bertolín C, Querin G, Martinelli I, Meo G, Pegoraro E, Soraru G

EPI-13 A high-incidence cluster of ALS in the French Alps: common environment and multiple exposures
Laigrange E, Bonneterre V, Talbot K, Couratier P, Bernard E, Camu W

EPI-14 Assessing cyanobacterial harmful algal blooms as risk factors for amyotrophic lateral sclerosis
Torick B, Zintz B, Stomme E

EPI-15 Geospatial association between amyotrophic lateral sclerosis and its influence factors in Beijing, China, 2010-2015
Zhou S, Qian S, Chang W, Wang L

EPI-04 An odyssey in epidemiology - from snow to hill to ALS: acquired somatic mutations may trigger ALS onset
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Martin S, Ossher L, Kulka A, Talbot K, Al-Chalabi A

EPI-10 The Swedish MND quality registry

EPI-11 POSTER WITHDRAWN
EPI-16 Predictors of survival among US military veterans with ALS
Larson T, Mehta P, Horton K

EPI-17 Exposure to electric shocks and extremely-low-frequency magnetic fields: the risk of amyotrophic lateral sclerosis

EPI-18 Highways of NH/VT and ALS patients: a geospatial analysis using case-control addresses and census blocks
Guetti B, Heneghan P, Andrew A, Torbick N, Facippio D, Stommel E, Bradley W

EPI-19 Smoking is a risk factor for ALS in a UK population - a case control study
Martin S, Shaw P, Pearce N, Shaw C, Morrison K, Al-Challabi A

EPI-20 Do people with ALS have lower index-to-ring finger length ratios (2D:4D)?
Kullmann JP, Pamphlett R

EPI-21 The role of pre-morbid diabetes in developing amyotrophic lateral sclerosis
Calvo A, D’Ovidio F, Costa G, Derrico A, Carnà P, Chio A

EPI-22 Dysphagia in ALS: an Italian population-based study
Moglia C, Calvo A, Canosa A, Cammarosano S, Manera U, Pisanò F, Mora G, Mazzini L, Chiò A

EPI-23 Medical cost of ALS in Japan
Uchino A, Toimina N, Ogino Y, Ogino M

EPI-24 Are people with ALS really nicer? An online international study of the big five personality traits
Kullmann JP, Pamphlett R, Hayes S

EPI-25 Modelling individual amyotrophic lateral sclerosis disease courses in the PRO-ACT database using the D50 progression model

EPI-26 Serum creatine kinase in neuromuscular disease/post-polio syndrome (PPS)
Quadros AA, Corrêa MDSS, Mota MF, de Campos KM, Viana CF, Ferreira LS, Munhoz CT, Helou AS, Piovesan RH, Oliveira AS

THEME 6

Biomarkers

BIO-01 TDP-43 based biomarker development in ALS

BIO-02 Circulating neurofilament-containing hetero-aggregates as a test-bed for novel biomarkers and therapeutics in neurodegeneration
Adiutori R, Aarum J, Zubir I, Leoni E, Di Benedetto S, Bremang M, Pike I, Sheer D, Malaspina A

BIO-03 Characterisation of CSF extracellular vesicles and their proteome in ALS

BIO-04 Vesicle secretion in ALS transmission: the VITAL consortium

BIO-05 Circulating exosomes as a novel source of biomarkers for ALS progression

BIO-06 Micro-RNA carrying exosomes in motor neuron disease patients
Del Carratore R, Dolciotti C, Bendinelli S, Pelagatti A, Falleni A, Cavalli L, Da Prato I, Carboncini M, Bongionanni P

BIO-07 Different signatures and pathways of mRNA/microRNAs in extracellular vesicles of ALS patients

BIO-08 Identification of circulating non-coding RNAs as biomarkers of amyotrophic lateral sclerosis

BIO-09 Chitotriosidase as a biomarker for ALS

BIO-10 A prospective study of two distinct epigenetic signatures for ALS diagnosis and prognosis prediction

BIO-11 Lipocalin-2 levels are increased in amyotrophic lateral sclerosis and activate microglia

BIO-12 The Kynurenine Pathway as a biomarker for ALS progression
Tan V, Lim C, Borotkanics R, Gilles J

BIO-13 Blood hemoglobin A1c levels and amyotrophic lateral sclerosis survival
Wei Q-Q, Cao B, Shang H

BIO-14 Blood inflammatory markers in motor neuron disease patients: pattern changes over time along disease progression

BIO-15 C-reactive protein (CRP) is significantly higher in amyotrophic lateral sclerosis (ALS) patients on non-invasive ventilation (NIV) and tracheostomy–invasive ventilation (TIV) compared with ALS patients at intake clinic evaluation and decreases following riluzole administration — is CRP potentially a biomarker for treatment responsiveness?

BIO-16 Up-regulation of neuronal apoptosis inhibitory protein associates with slower disease progression in amyotrophic lateral sclerosis: implication of a novel prognostic biomarker

BIO-17 LRP4 antibodies in Chinese patients with ALS

BIO-18 Correlation of testosterone levels with progression of amyotrophic lateral sclerosis: a cross sectional study

BIO-19 The fecal microbiome ALS patients

BIO-20 Gut microbiome assessment in people with ALS: an interval analysis

THEME 7

Electrophysiology

ELE-01 Increased rate of sensory abnormalities in C9orf72-associated ALS

ELE-02 Peripheral sensory nerve disorder in amyotrophic lateral sclerosis

ELE-03 Functional involvement of the motor corpus callosum in amyotrophic lateral sclerosis
Hubers A, Böckler B, Kammer T, Kassubek J, Ludolph AC

ELE-04 Deconstructing motor neuron disease: site-of-origin and patterns of disease spread
Dharmadasa T, Matamala JM, Howells J, Simon NG, Vucic S, Kiernan MC

ELE-05 Imbalance in cortical inhibition-excitation networks underlies the development of cortical hyperexcitability in ALS
Van den Bos M, Geervaisinga N, Menon P, Kiernan M, Vucic S

ELE-06 Spinal interneuronopathy in patients with ALS
Marchand-Pauvert V, Pradat P-F

ELE-07 Changes in nerve axonal function in patients with amyotrophic lateral sclerosis treated with edaravone

ELE-08 Acute effects of riluzole and retigabine on axonal excitability in patients with amyotrophic lateral sclerosis: a randomized, double-blind, placebo-controlled, three-way cross-over trial

ELE-09 Creatine kinase level and its relationship with quantitative electromyography analysis in amyotrophic lateral sclerosis
Tai H, Cui L, Liu M, Guan Y, Li X, Shen D, Ding Q, Zhang K, Liu S

ELE-10 EMG evaluation of ALS: fasciculation potentials are not so common
ELE-11 Motor unit number estimation recording from the tongue: a pilot study
Mclduff C, Martucci M, Shin C, Qi K, Boegle A, Rutkove S

ELE-12 Reduction in motor unit number index (MUNIX) in phase I, phase II and phase III of the D50 ALS disease model occurs first in APB, with EDB, ADM and TA following in that order

ELE-13 Motor unit loss in amyotrophic lateral sclerosis as detected by MSCAN occurs mainly in phase II of the D50 disease progression model
Hohmann M, Appelfeller M, Ringer TM, Gunke A, Stubendorff B, Witte OW, Grosskreutz J

ELE-14 Computational analysis of ultrasound imaging to quantify spatial and temporal characteristics of fasciculations in healthy and MND affected participants
Bibbigs K, Harding P, Combnes N, Loram I, Tole E

ELE-15 A novel automated pipeline accurately counts fasciculations detected by serial high-density surface EMG
Batchford J, Wickham A, Drakakis E, Boutelle M, Mills K, Shaw C

ELE-16 The origin of fasciculation in ALS: direct correlation between axonal excitability and fasciculation

ELE-17 Longitudinal surveillance study in ALS patients correlating orthopnea with changes in the electrocardiogram QRS axis

ELE-18 Accelerometers as non-invasive tools for the objective measurement of limb specific range of motion and force in ALS
Premasiri A, Mosko O, McNally M, Vieira F

THEME 8

Imaging

IMG-01 SPG11-related motor neuron disease is a mixed neurodevelopmental white matter and neurodegenerative grey matter condition

IMG-02 Phenotype-specific imaging signatures along the ALS-FTD spectrum
Finegan E, Omer T, Hutchinson S, Doherty M, Vajda A, McCluskey L, Pender N, Hardiman O, Bede P

IMG-03 Virtual brain biopsies in ALS: a diagnostic framework based on in-vivo pathological patterns
Bede P, Iyer P, Finegan E, Omer T, Hardiman O

IMG-04 Imaging and physiology markers of disease progression in C9orf72 mutation carriers
Floeter MK, Daniellian LE, Bageac D, Lehky T, Offit M, Clark MG, Smallwood R, Wu T

IMG-05 MRI and DTI predictors of reduced survival across the ALS-FTSD continuum

IMG-06 Beyond C9ORF72: neuroradiological characterisation of hexanucleotide repeat negative ALS-FTD patients
Finegan E, Omer T, Hutchinson S, Doherty M, Vajda A, McLaughlin R, Pender N, Hardiman O, Bede P

IMG-07 Structural and functional Papez circuit integrity in amyotrophic lateral sclerosis

IMG-08 Neuroimaging changes in the first 5 years of symptoms in patients with primary lateral sclerosis
Clark M, Huang C, Bageac D, Daniellian L, Smallwood R, Floeter MK

IMG-09 Functional connectivity changes associated with disease progression in ALS
Loewe K, Machts J, Petri S, Heinze H-J, Veißhauer S, Schoenfeld MA

IMG-10 Integration of progressive white matter structural and functional MRI changes in motor neuron disease
Menke R, Douard G, Talbot K, Turner M

IMG-11 Brain-behaviour correlates of bulbar ALS: gray and white matter regional analyses
Shelliken S, Myers M, Black SE, Zinman L, Yunusova Y

IMG-12 Frontostriatal grey matter atrophy in ALS—detection on individual patient level
Radakovic R, Flanagan E, Kiernan M, Mioshi E, Hornberger M

IMG-13 Voxel-based morphometry (VBM) subcortical white matter changes correlate with D50 model disease progression in amyotrophic lateral sclerosis
Batybekova M, Pfehl T, Stubendorff B, Bokemeyer M, Mayer T, Hartung V, Witte OW, Grosskreutz J

IMG-14 Progression of cerebellar involvement in amyotrophic lateral sclerosis as seen by SUIT/ CAT12 voxel-based morphometry and D50 disease modelling
Batybekova M, Pfehl T, Stubendorff B, Bokemeyer M, Mayer T, Hartung V, Witte OW, Grosskreutz J

IMG-15 Sensitivity and specificity of neurite orientation dispersion and density magnetic resonance imaging (NODDI) at the single patient level in amyotrophic lateral sclerosis
Barritt AW, Broad R, Leigh PN, Cercignani M

IMG-16 Selective alteration of thalamic motor structural connectivity in ALS
Tu S, Menke R, Talbot K, Kiernan M, Turner M

IMG-17 Region-specific impairment of the cervical spinal cord in amyotrophic lateral sclerosis: a preliminary study using SC templates and quantitative MRI (DTI/ihMT)

IMG-18 Spinal cord MRI: is it an effective classification tool for the diagnosis of motor neuron disease conditions?
Querin G, El Mendili MM, Delphine S, Lenglet T, Marchand-Pauvert V, Pradat P-F

IMG-19 Ultra-short echo time magnetic resonance spectroscopy of multiple metabolites in amyotrophic lateral sclerosis, preliminary findings
Blacher J, Staermose T, Figlewski K, Maller AT, Near J

IMG-20 Elemental imaging of post-mortem CNS in MND using laser ablation-ICP-MS
Kysuens K, Paul B, Hare D, Crouch P

IMG-21 Relationship between brain metabolism and cognitive/behavioral functioning in ALS

IMG-22 [18F]GE180 compared to [11C]PBR28 in detecting in vivo glial activation in people with ALS

IMG-23 PET imaging studies show enhanced expression of mGluR5 and inflammatory response during progressive degeneration in ALS mouse model expressing SOD1-G93A gene

IMG-24 Development of positron emission tomography radiotracer for imaging cannabinoid receptor type 2 (CB2) in ALS

IMG-25 Diaphragm ultrasound in ALS: a case demonstrating an important role for this technique

THEME 9

Clinical Trials

CLT-01 ALS clinical research: limitations for physicians
Ross M, Burge M, Dalrymple J, Muzyka I, Chang A, Smith B

CLT-02 Survival and disease progression in SOD1 familial ALS in Catalonia

CLT-03 People living with ALS and their caregiver's input into drug development

CLT-04 An argument for factorial design in ALS trials
Thakore N, Lapin B, Pioro E, Schoenfeld D

CLT-05 Analytic strategies for combined assessment of function and mortality in ALS clinical trials
van Eijk RPA, Eijkemans MJC, Rizopoulos D, van den Berg LH, Nikolakopoulos S

CLT-06 ALS at home: measuring longitudinal patient outcomes without study visits
Shelfer J, Liss J, Berisha V, Shelton K, Qi K, Rutkove S

CLT-07 How much does distance limit the pool of potential clinical trial participants in the United States?
Collet MC
CLT-08 Mood enhancement using repetitive transcranial magnetic stimulation (rTMS) to the left dorsolateral prefrontal cortex (LTDLPCF) as an adjuvant therapeutic technique may improve quality-of-life and disease progression
Mariel A, Parmenter M, Rashed H, Pavlakis P, Holzberg S, Mona S, Lange D

CLT-09 Modulation of miRNAs in the CSF of ALS patients treated with MSC-NTP cells (NurOwn)

CLT-10 Hopelessness and depression in patients with amyotrophic lateral sclerosis undergoing a mesenchymal stem cell clinical trial. Preliminary results
Gonzalves Gedo M, Oliveira MAF, Ruivo Maximino J, Chadi G

CLT-11 ALS patients who receive edaravone treatment over 10 years
Yoshino H

CLT-12 Pharmacokinetics and safety of TW001 (the oral formulation of edaravone)
Hulsokte E, Mols R, Lagarauw M, van den Berg L, van der Geest R

CLT-13 Edaravone in ALS: the effect of potential drug—drug interactions via P450
Nakamura Y, Kawaguchi A

CLT-14 Onset of detectable effect of edaravone: a post-hoc analysis
Takei K, Takahashi F, Liu S, Tsuda K, Palumbo J

CLT-15 Edaravone in ALS clinical trials: an assessment of safety, tolerability and treatment persistence
Hubbel J, Tsuda K, Kalin A, Ji M

CLT-16 Initial findings from a first in human trial of BiIB067, an antisense oligonucleotide targeting SOD1 mRNA, support continued development of BiIB067 for ALS

CLT-17 Safety and tolerability of expanded autologous Regulatory T lymphocyte infusions in amyotrophic lateral sclerosis
Thonhoff J, Beers D, Zhao W, Pleitez M, Simpson E, Berry J, Cudkowicz M, Appel S

CLT-18 Oral levosimendan (ODM-109): key placebo-controlled results from the phase 2 study in ALS
Motta MP, Motta MP, Quadros AAJ, Oliveira ASB

CLT-23 A phase 1 study of CuATSM in amyotrophic lateral sclerosis
Rowe DB, Mathers S, Joel K, Rosenfeld CS

CLT-24 A pharmacometabolomics approach in a clinical trial of oxosoxime in ALS

CLT-25 Adverse events and therapeutic adherence of the L-carnitine+piracetam association in patients with motor neuron disease/post-polio syndrome — randomized clinical trial
De Souza Brito Conti M, Quadros AAJ, Oliveira ASB

CLT-26 Therapeutic effect of L-carnitine+piracetam on fatigue and muscle strength in patients with post-polio syndrome — randomized, double-blind, placebo-controlled clinical study
Motta MP, Quadros AAJ, Oliveira ASB

CLT-27 Laboratorial parameters in the L-carnitine+piracetam association in patients with neuron motor disease/post-polio syndrome — randomized clinical trial
De Souza Brito Conti M, Quadros AAJ, Oliveira ASB

CLT-28 Evaluation of the efficacy of the reduction of iron charge in the cold intolerance of patients with post-polyneuritis syndrome: preliminary results
Motta MP, Campos KM, dos Santos VR, Oliveira ASB, Quadros AAJ

THEME 10

Therapeutic Strategies

TST-01 Potential role of gut microbiota in ALS pathogenesis

TST-02 Variation of gut microbiome rescues paralysis and neurodegeneration profiles in C elegans ALS models
Labarre A, Guittard E, Parker JA

TST-03 Single chain antibodies against TDP-43—RRM1 domain as therapeutic approach for TDP-43 proteinopathy
Pozzi S, Thammissetty SS, Gravel C, Julien J-P

TST-04 Accumulation of neuronal cytoplasmic TDP-43 activates cell stress signalling pathways in genetically modified TDP-43 mice
Le S, Wright A, Mehta P, Berning B, Gul H, Hedel T, Riddell W, Atkin J, Walker A

TST-05 An ALS treatment strategy based on ligand-mediated stabilization of the native hSOD1 conformation
Santur KB, Mohler UJ, Willbold D

TST-06 Therapeutic vaccines for ALS directed against misfolding specific epitopes of Cu/Zn superoxide dismutase 1
Zhaob M, Marciniuk K, Yousell M, Gibbs E, Napper S, Cashman N

TST-07 Treatment of the SOD1transgenic mouse model of amyotrophic lateral sclerosis (ALS) with an all-D-enantiomeric peptide

TST-08 Multifunctional effects of endogenous hematopoietic stem cells mobilization on pathogenesis of ALS in a mouse model of the disease
Solomon B

TST-09 Conservative iron chelation as a neuroprotective strategy in amyotrophic lateral sclerosis

TST-10 Retrograde axonal migration and neuroprotection of tetanus toxin fragments and Bcl-2 fusion proteins
Watanabe Y, Matsuba T, Nakamishi M, Une M, Hanajima R

TST-11 Mesenchymal stem cell-derived exosomes shuttle miRNAs that affect the activated microglia phenotype in SOD1transgenic mice
Giunti D, Milanesi M, Marin C, del Rosko NK, Bonifacio T, Uccelli A, Bonanno G

TST-12 Metabotropic glutamate receptor type 5 (mGlu5) plays a key role in glial activation in the SOD1G93A mouse model of MND

TST-13 Masitinib prevents sciatric nerve and sensory afferent pathology in a SOD1transgenic rat model of amyotrophic lateral sclerosis
Trias E, Ibarburu S, Barreto-Núñez R, Mansfield C, Hermine O, Beckman J, Barbeito L

TST-14 A novel mast cell related pathogenic mechanism in the SOD1transgenic rat model of ALS that can be therapeutically targeted by masitinib
Trias E, Ibarburu S, Barreto-Núñez R, Hermine O, Beckman J, Barbeito L

TST-15 Targeting of the retinoid pathway in SOD1transgenic mice by delivery of engineered polymeric nanoparticles
Medina D, Chung E, Ceton R, Bowser R, Sirianni R

TST-16 Ataxin-2 in stress granules regulates nucleocytoplasmic transport in ALS
Zhang K, Daigle G, Cunningham K, Coyne A, Grima J, Wadhwaa H, Bowen K, Rothstein J, Lloyds T

TST-17 Peripheral blood lymphocytes in ALS patients are defined by a distinct profile of deregulated chemokine receptors
Pemer C, Förster M, Heidel F, Witte OW, Prett L, Grosskreutz J

TST-18 Beneficial effects of RNS60 in cellular and animal models of amyotrophic lateral sclerosis

TST-19 The histone deacetylase inhibitor RGFP109 enhances efficacy of heat shock protein inducers in motor neurons and glia
Kuta R, Minotti S, Larochelle N, Nalbantoglu J, Durham H

TST-20 Promotion of the M2 microglial state and enhanced neuronal trophic support extend survival in the murine model of ALS
Snyder A, Neely E, Mrowczynski O, Payne R, Gerominio A, Simmons Z, Conner J

TST-21 Intra-spinal delivery of AAV2-NRTN for ALS — a dose-ranging study of safety, tolerability, biodistribution and efficacy
Gross S, Shim B, Peterson B, Bartus R, Boulis N, Maragakis N

TST-22 Intravenous delivery of AAV gene therapy to regions of the CNS critical for the treatment of ALS

TST-23 POSTER WITHDRAWN

TST-24 Immunogenes for targeted neurotrophic gene delivery to motor neurons

THEME 11
Improving Diagnosis, Prognosis and Disease Progression

IDP-01 ‘CONFIDO’: a pilot study of dog-assisted therapies in ALS patients
Vignolo M, Zuccarino R, Rao F, Trinchero C, Giove E, Cippolina I, Ferraris I, Caponnetto C

IDP-02 The twin cities ALS research consortium: a model for regional collaboration in advancing research for people living with ALS

IDP-03 Dynamic weighting of old and new information for predicting future condition of ALS patients
Nahon A, Lerner B

IDP-04 Displaced reality: the challenges of creating an ALS clinical study in which all data collection takes place in the patient’s home
Rutkove S, Shifer J

IDP-05 NeuroGUIDization of PALS population as a necessary condition for patient-centric research and care

IDP-06 PRO-AC: meta-analysis of concomitant medications and active ingredients on disease progression
Sherman A, Sinani E, Walker J, Macklin E

IDP-07 The ALS stratification challenge: using big data and predictive computer models to identify clinically significant ALS patients sub-populations

IDP-08 Stratifying ALS patients by disease progression patterns
Gordon J, Lerner B

IDP-09 Enriched clinical trial cohorts improve study power
Fournier C, Taylor A, Ennist D

IDP-10 Modeling of ALS progression using a temporal machine-learning algorithm
Nahon A, Gordon J, Lerner B

IDP-11 Modeling neuroanatomic propagation of ALS in the spinal cord
Drewert B, Thakore N, Mitchell B, Piero E, Ravits J, Petzold L

IDP-12 Validation of predictive ALS machine learning models with a contemporary, external dataset
Taylor A, Beaulieu D, Jahandideh S, Meng L, Bian A, Andrews J, Ennist D

IDP-13 Machine learning models for the assessment of potential ALS biomarkers
Jahandideh S, Ennist D

IDP-14 Utility of the ALSFRS-R to measure function in advanced ALS: the VA biorepository brain bank
Braunfeld M, Ron I, Rissoni N

IDP-15 The ALS mobile analyzer: monitoring ALS disease progression via smartphone app and identifying novel digital biomarkers
Braunfeld M, Ron I, Rissoni N

IDP-16 An exploratory study to investigate the use of biotelometry to identify markers of disease progression in subjects with amyotrophic lateral sclerosis — pilot phase

IDP-17 Effects of socio-economic and cultural factors on the ALSFRS-R in South African ALS patients: a pilot study
Braga A, Henning F

IDP-18 The aim and development of the primary lateral sclerosis functional rating scale (PLSFRS)
McHale B, Huf J, Ibagon C, Mitsumoto H

IDP-19 Clinical relevance of quantitative upper motor neuron burden (UMNB) scales in ALS
Babs B, Alshikho MJ, Cernasov PM, Reynolds BV, Pijanowski OR, Paganoni S, Chan J, Atassi N

IDP-20 Multistate modeling of ALS stages: estimating risks of transition and death
Thakore N, Lapin B, Pioro E

IDP-21 Estimating QALYs from the ALSFRS-R in South African ALS patients: a pilot study
Braga A, Henning F

IDP-22 Six-minute walk test (6MWT) correlate with serum creatinine (SC) in ambulatory individuals
Ikeda T

IDP-23 Lingual and jaw kinematic abnormalities precede speech and swallowing symptoms in ALS
Perry B, Yunusova Y, Martino R, Plowman E, Green J

IDP-24 Correlations between slow vital capacity and measures of respiratory function on the ALSFRS-R
Jackson C, de Carvalho M, Genge A, Heiman-Patterson T, Shefner J

IDP-25 Changes of supine forced vital capacity is the best respiratory predictor of disease progression in ALS patients

IDP-26 Alteration of forced vital capacity is a prognostic factor for survival of amyotrophic lateral sclerosis patients

IDP-27 Lingual and jaw kinetic abnormalities: a clinical evaluation
Perry B, Yunusova Y, Martino R, Plowman E, Green J

IDP-28 A study on relation of body mass index to survival in Chinese sporadic amyotrophic lateral sclerosis patients
Yang L, Liu X, Chen L, Tang L, Fan D

THEME 12
Cognitive and Psychological Assessment and Support

COG-01 POSTER WITHDRAWN

COG-02 It’s NICE to ECAS: the impact of training health professionals to identify cognitive and behavioural change in ALS/MND
Hodgins F, Bell S, Abrahams S

COG-03 A new eye-tracking based measure of cognitive flexibility in amyotrophic lateral sclerosis

COG-04 Apathy in ALS in the context of depression or FTD — a clinical evaluation
Wüst M, Keller J, Abo-Ozhan HE, Uttner I, Ludolph AC, Lušk D

COG-05 Wechsler measures of reasoning correlate with interpretation of emotional expressions and to family rating of change in apathy and executive functioning in the FTD prodrome
Flaherty C, Hotz A, Slinkard K, Kraft J, Marino A, Simmons Z

COG-06 Use of coping strategies in motor neuron disease/amyotrophic lateral sclerosis: association with demographic and disease-related characteristics
Schnurer D, Mills R, Young C

THEME 12
COG-07 Empathy and trust among people with MND sharing information, experiences and emotions in an online discussion forum
Hargreaves S, Bath P, Duffin S, Ellis J, Lovatt M

COG-08 Physical and mental factors affecting perceived stigma amongst people with motor neurone disease/amyotrophic lateral sclerosis
Edge R, Tannant A, Hadad S, Young C

COG-09 Problems and metaphors shown in narratives of family caregivers of patients with amyotrophic lateral sclerosis
Muokaaka K

COG-10 Factor analysis of the Zarit burden interview for amyotrophic lateral sclerosis (ALS) caregivers
Carney S, Galvin M, Pender N, Staines A, Hardiman O

COG-11 Frequency of neuropsychiatric syndromes in first and second degree relatives of patients with amyotrophic lateral sclerosis (ALS)
Ryan M, Costello E, Heverin M, Hardiman O

COG-12 Symptoms of psychiatric disorders in people living with ALS and their family members

THEME 13
Respiratory and Nutritional Management

RNM-01 Understanding the complexity and instability of respiratory control in ALS/MND involves more than just forced vital capacity

RNM-02 Importance of the criteria of indication and adequacy of ventilation of patients with motor neuron disease
Oda AL, Carvalho EV, Ferreira FB, Holsapfel SGA, Salvioni CCS, Chieia MAT, Oliveira ASB

RNM-03 Bulbar function tests (alternate motion rates, single-breath counts, single breath sustained phonation, timed 90mL swallow) compared with oro-facial motor scales and respiratory function tests — longitudinal surveillance in bulbar and limb onset ALS patients — effect of non-invasive ventilation

RNM-04 Respiratory function, peak cough flow and history of respiratory tract infection in people with motor neuron disease and other neuromuscular disorders
Sheers N, Howard M, Rautela L, Chao C, Rochford P, Nicholls C, Berlowitz D

RNM-05 Relationship between effectiveness of cough peak flow and maximum phonation time in patients with motor neuron disease
Oda AL, Braga TER, Salvioni CCS, Alves PCL, Borges RM, Sierra HNM, Neves JWC, Frabasile L, Chieia MAT, Oliveira ASB

RNM-06 Profile of patients with amyotrophic lateral sclerosis at the time of non-invasive ventilation
Carvalho EV, Holsapfel SGA, Caromano FA, Oda AL, Oliveira ASB

RNM-07 Predicting factors of survival after onset on non-invasive ventilation in ALS patients
Gómez-Mendietta MA, Salvador M, González G, Carpio C, Martínez-Redondo M, Rodríguez de Rivera F, Santiago A

RNM-08 Baseline risk factors associated with non-invasive ventilation use in ALS

RNM-09 Non-invasive ventilation in motor neurone disease/amyotrophic lateral sclerosis: an Australasian perspective
Chow WK, Rowe D, Ing A

RNM-10 Supporting MND patients using NIV: experiences of professional caregivers
Cousins R, Ando H, Young C

RNM-11 Case series: graphic analysis allows titration of negative pressures during MIE to prevent airway collapse in bulbar patients
Nilstuen J

RNM-12 Home telemonitoring for ALS in early and late ventilated patients: a validation study
Pinto A, Braga AC, Guedes S, de Carvalho M

RNM-13 Dyspnoea, orthopnoea and ventilation therapy in ALS — a systematic analysis of 10 years of managed care

RNM-14 Use of specialized ventilator to increase the maximum insufflation capacity (MIC) in patients with ALS
Dorca A, Schneider F

RNM-15 Sniff nasal inspiratory pressure (SNIP) in amyotrophic lateral sclerosis: relevance of the methodology for respiratory function evaluation
Pinto S, de Carvalho M

RNM-16 Influence of ventilatory strategy on oral communication of patients with tracheostomized amyotrophic lateral sclerosis — case report
Dorca A, Sisterolli D

RNM-17 Reviewing studies of diaphragm pacing results to identify the correct phenotype of ALS/MND patients for which diaphragm pacing could help in ventilation and in which patients diaphragm pacing should not be utilized: the devil is in the details

RNM-18 Induction rates of PEG and NPPV for ALS in Japanese ALS centers
Ogino Y, Matsumura T, Arutsu N, Tateishi T, Morita M, Itami T, Ogino M

RNM-19 Hydration in motor neuron disease/amyotrophic lateral sclerosis
Salvioni C, Oda AL, Chieia MAT, Oliveira ASB

RNM-20 Effects of dietary counseling on nutrient consumption from the electronic health application system to measure outcomes remotely (EAT MORE) study

RNM-21 Progression of dysphagia in patients with motor neuron disease: analysis by volume and consistency of food
Oda AL, Salvioni CCS, Alves PCL, Borges RM, Sierra HNM, Neves JWC, Frabasile L, Rocha MSG, Chieia MAT, Oliveira ASB

RNM-22 The effect of a multidisciplinary approach in the management of percutaneous radiological gastrostomy: a single center experience in a large cohort of ALS patients

RNM-23 Dysphagia and nutrition care in ALS — a systematic analysis of 10 years of managed care

RNM-24 Factors affecting 1-month mortality after gastrostomy placement

RNM-25 An analysis of prognostic factors after percutaneous endoscopic gastrostomy placement in Japanese patients with amyotrophic lateral sclerosis
Nagashima K, Furuta N, Makioaki K, Fujita Y, Ikeda M, Ikeda Y

RNM-26 Nutritional status and relationship with dysphagia in patients with amyotrophic lateral sclerosis with alternative feeding route
Salvioni C, Oda AL, Pauli MC, Alves P, Borges RM, Sierra HNM, Neves JWC, Stanich P, Chieia MAT, Oliveira ASB

THEME 14
Multidisciplinary Care and Improving Quality of Life

MDC-01 Canadian ALS best practice recommendations

MDC-02 ALS multidisciplinary care units: evaluating the implementation and economic cost of a one-stop model

MDC-03 Multidisciplinary care improves survival of patients with ALS — evidence from the Ljubljana ALS centre

MDC-04 The positive impact of exercise for people with MND
Carey H

MDC-05 Sinusoidal electrical muscle stimulation and passive arm/leg cycling exercise for rehabilitative treatment of patients with motor neurone disease
Dini M, Corbianco S, Baldereschi G, Bongioanni P

MDC-06 Why people with ALS (PALS) choose not to participate in ALS patient education courses
Brandstüpp L, With H, Vægter M, Jeppesen J

MDC-07 Risk factors for social withdrawal in motor neurone disease/amyotrophic lateral sclerosis
Schlüter D, Tannant A, Young C
MDC-08 Change of the speed and emotional burden of nursing students’ use of a letter board as a simulation of an ALS patient with severe difficulty of communication
Narita Y, Shindo A, Nishikawa Y

MDC-09 Effect of sentence length on intelligibility and speech motor performance in ALS
Allison K, Yunusova Y, Green J

MDC-10 Associations between pain, mood and quality-of-life in motor neuron disease/amyotrophic lateral sclerosis
Edge R, Yeung JA, Young C

MDC-11 The impact of apathy on quality-of-life in ALS
Caga J, Hsieh S, Highton-Williamson E, Zoirng MC, Ramsey E, Devenney E, Ahmed RM, Kiernan M

MDC-12 G-tube placement in people with ALS: data from the ceftriaxone clinical trial and clinic-based evaluations

MDC-13 The effect of the percutaneous endoscopic gastrostomy (PEG) tube placement on the quality-of-life of ALS patients and their caregivers

MDC-14 Changes in taste perception in ALS patients with gastrostomy feeding tubes and the impact on their quality-of-life
Tarlanni C, Greco LC, Lizio A, Gerardi F, Sansone VA, Lunetta C

MDC-15 Quantifying constipation in ALS: an analysis from the microbiome assessment in people with ALS study
Nicholson K, Jeon M, Chan J, Bjornevik K, Ascherio E, Berry J

MDC-16 Decision-making of TV in Japan
Tomina Ni, Matsumura T, Atsuta N, Tateshi T, Morita M, Imai T, Ogino Y, Ogino M

MDC-17 A new tool to improve oral communication during non-invasive ventilation in amyotrophic lateral sclerosis patients

MDC-18 Patients’ perspectives of multidisciplinary home-based telehealth for amyotrophic lateral sclerosis
James N, Power E, Hogden A, Vucic S

MDC-19 Efficacy of teleBCI for training and engaging in brain-computer interface communication
Geronimo A, Simmons Z

MDC-20 Brain-computer interface with P300-Speller: usability for disabled patients with amyotrophic lateral sclerosis
Sorani MH, Guy V, Papadopoulos T, Bruno M, Desnuelle C, Clerc M

MDC-21 Usability of eye tracking technology for increased communication aimed at patients with motor neuron disease
Ferreira LS, Quadros AAJ, Rogério dos Santos V, Fávero FM, Andrews Portes L, Oliveira ASB

MDC-22 Timeline for provision of power wheelchair (PWC) prescription to patients with amyotrophic lateral sclerosis (pALS) enrolled at Carolinas Neuromuscular/ALS-MDA Care Center at Carolinas Healthcare System (CHS)
Holsten SE, Ward AL, Sanjak MS, Crosby-Johnson M, Sanders T, Williamson TA, Lucas NM, Braver EK, Bockenek WL, Mabe C, Brooks B

MDC-23 ‘Freddy by a wheelchair’ — How patients use and experience their power wheelchairs
Wicks P, Cerrato D, Eaneff S, Leike K, Andersson- Svanh A

MDC-24 Caregiver experience, health-related quality-of-life and life satisfaction in informal caregivers to patients with ALS
Kierkgaard M, Sandstedt P, Littorin S, Gottberg K, Ytterberg C, Olsson M

MDC-25 A new framework to increase participation in peer-to-peer support for spouse carers of people with ALS (PALS)
Brandstrup L, Vaeger M, With H, Jeppesen J

MDC-26 Needs of informal ALS caregivers across the caregiving course: a qualitative analysis
Galvin M, Carney S, Corr B, Pender N, Hardiman O

MDC-27 Knowledge about ALS/MND among young caregivers in the US and South Africa: Implications for education and family support
Kavanaugh M, Woodley J, Mochan A, Henning F

MDC-28 Patients with ALS find value in genetic testing
Wagner K, Nagaraja H, Allain D, Quick A, Kolb S, Roggenbuck J

MDC-29 Information seeking and ALS communications: impacts and preferences from diagnosis to end of life
Moir M, Luth W, Bubela T, Johnston W

MDC-30 Do not resuscitate/do not intubate status in ALS patients with gastrostomy feeding tubes and the impact on their quality-of-life
Moir M, Luth W, Bubela T, Johnston W

MDC-31 Considerations of organ donation from patients with ALS

THEME BW

Biomedical Work in Progress

BW-01 C9ORF72 patient specific iPSC-derived lines as ALS in vitro model
M Nizzardo, R Federica, T Michela, R Paola, M Bucchia, S Brjakovic, N Bresolin, S Corti, GP Comi

BW-02 NeuroLINCS: Identifying ALS-specific signatures from iPSC-derived motor neuron using multi-omic integration
J Li, VJ Dardov, RG Lim, JG Daigle, C Svendsen, J Van Eyk, E Fraenkel, LM Thompson, J Rothstein, NeuroLINCS Consortium

BW-03 A microfluidic co-culture system to study the neuromuscular junctions formed by human motor neurons derived from ALS patient iPSCs
C Franz, E Hosseinian, P Mukherjee, A Domenighetti, E Krickin

BW-04 MiRNA profiling of ALS iPSCs and iPSC-derived motor neurons: molecular and therapeutic implications
M Rizzuti, M Nizzardo, V Melzi, G Filosa, L Dionis, L Calandrelli, N Bresolin, GP Comi, S Barabino, S Corti

BW-05 MicroRNA-183-5p couples cell stress sensing and cell death programming in the development of amyotrophic lateral sclerosis
C Li, H Shang

BW-06 Early gene expression profiling of spinal motor neuron vulnerability pathways in a mouse model of ALS
F Zanganah, C Bye, B Turner

BW-07 Humanising the mouse TardaBP gene
F De Giorgio, A Devoy, C Milioto, F Zhu, K McKenzie, A Acevedo-Arozena, E Fisher

BW-08 Mitochondrial dysfunction associated with a SOD1-ALS knock in model
B Steinitz, K Wharton

BW-09 Nuclear pore complex composition in the mammalian CNS: Regional and cell type specific differences
JC Grima, VJ Dardov, AN Coyne, JG Daigle, K Zhang, T Philips, JV Eyk, TE Lloyd, MJ Matunis, JD Rothstein

BW-10 Developing a novel in vivo model of cell-to-cell protein transmission in MND
M Haidar, L Lau, B Turner, C Bye

BW-11 Comparison of autoimmune comorbidities among different motor neuron disease subtypes: A retrospective study
PP Pavlakis, M Shahbazi, DJ Lange

BW-12 Do chloroviruses contribute to ALS?
G Pattee

BW-13 Immigration study on Amyotrophic lateral sclerosis (ALS) and Parkinsonism-dementia complex (PDC) of the Kii peninsula, Japan
Y Kokubo, R Sasaki, S Morimoto, M Mimura, H Ishiura, M Hasegawa, M Yoshida, S Tsujii, S Kuzuhara

BW-14 Longitudinal ALS registries: methods, objectives, and results
X Arcilia, D Walk, K Goslin, P Vade, A Sherman

BW-15 POSTER WITHDRAWN

BW-16 Detection of C9orf72 allele expansions in a cohort of 277 ALS patients and control subjects
A Calo, A Tsilas, T Dane, M Lukashev

BW-17 Breast cancer susceptibility in patients with spinal bulbar muscular atrophy: a case report.
G Querin, I Martellini, C Bertolin, E Pegoraro, P Mara, G Soraru

BW-18 Latent cluster analysis of ALS phenotypes: identification of prognostically differing groups - objectives, and results
S Orlandi, A Walk, K Goslin, P Vade, A Sherman

BW-19 Mutations in the ARRP21 gene are associated with familial and sporadic amyotrophic lateral sclerosis
CH Wong, SD Topp, YB Lee, S Mueller, O Baron, G Cocks, M Fanto, BN Smith, N Ticozzi, J Landers, CE Shaw
Clinical Work in Progress

CW-01 ALS prefer initiative - a platform for patient engagement in drug development
R Bedack, J Ravits, M Benatar, C Heatwole, C Balas, A Durham, D Zook, S Rudnicki, B Charpentier, L Bruijn, J Berry, J Andrews

CW-02 A pilot study of Safety of Caprylic Triglycerides in ALS
D Lange, M Shahbazi, S Holzberg

CW-03 Biomarker supervised fibrastim (G-CSF) response in ALS patients

CW-04 COMMEND: A randomized, double-blind, controlled, parallel group Ph2 study assessing FLX-767 for the treatment of muscle cramps in motor neuron disease
G Short, J Szege, D Cabral-Lilly, B Hegarty, W McVicar, T Wessel

CW-06 Pivotal phase 3 clinical trial of ultra-high dose methylcobalamin for ALS: The first trial using Awaji criteria (JET-ALS Study)
R Kaji, Y Izumi, S Kuwabara

CW-07 Blinded post-trial selection of outcome measures increases efficiency of ALS clinical trials
E Macklin, S Rutkove, S Schoenfeld

CW-08 Assessment of longitudinal changes in ALS using diffusion MRI
P Pisharody, C Lenglet, D Walk

CW-09 Longitudinal assessment of biochemical changes in ALS by in vivo magnetic resonance spectroscopy at ultra-high field
I Cheong, G Oz, M Marjanska, D Walk

CW-10 Precentral and postcentral cortical thickness and their relation to ALSFRS-R and neuropsychological biomarker MUNIX in ALS

CW-11 Multimodal assessment of ALS using ultra-high field MR spectroscopy and diffusion MRI
I Cheong, P Pisharody, G Oz, C Lenglet, G Manousakis, D Walk

CW-12 Chronic laryngeal nerve stimulation for swallowing preservation in ALS: A preclinical feasibility study
I Deninger, J Allen, B Ballenger, O Ohihausen, B Zitsch, V Caywood, K Osman, N Khodaparast, TE Lever

CW-13 A novel dynamic neck brace for ALS patients: Characterizing EMGs during synchronized neck motion
S Agrawal, H Zhang, B-C Chang, J Andrews, H Mitsumoto

CW-14 Impairment of cortico-muscular communication in motor neuron disease
A Coffey, S Dukic, R McMackin, M Heverin, M Lowery, R Carson, E Lalor, D Halliday, B Nasserolaslami, E Lalor, O Hardiman

CW-15 Investigation of dysfunction in cognitive brain networks in ALS by localisation of the sources of mismatch negativity
R McMackin, S Dukic, M Broderick, K Mohr, P Iyer, C Schuster, A Coffey, B Gavin, M Heverin, P Bede, N Pender, M Muthuraman, B Nasserolaslami, E Lalor, O Hardiman

CW-16 Detection of hand movement from EEG in ALS-patients and healthy individuals
S Alakhyborrosenaibadi, J Blicher, K Dremstrup, N Jiang, D Farina, N Mrachacz-Kersting

CW-17 Cortical unresponsiveness in bulbar onset motor neuron disease
H Rashed, P Pavlakos, A Marei, S Holzberg, M Shahbazi, D Lange

CW-18 Maladaptation of intracortical processes underlies development of exercise-induced fatigue in ALS
T Trinh, M Kiernan, M Lee

CW-19 Symptom monitoring application in real time for ALS (SMART-ALS): A pilot study using the beive smartphone application
J Berry, M Hussain, K Carlson, M Simoneu, J Barback, S Galbiati, J Barnett, P Staples, S Paganoni, J-P Onnela

CW-20 Prize4Life: Infrastructure and resources for ALS research
N Davis, M Bronfeld, I Ron, S Rishoni

CW-21 Augmentative Communication: Proactive preparation of low tech tools and message banking in collaboration with people with MND
J Costello

CW-22 Merging clinical work and research to improve care of patients with ALS/MND and calculate cost of care
E Locatelli, M Cudkowicz

CW-23 Retrospective study of nursing staff’s use of electronic records for ALS patients at a teaching hospital in Japan
Y Nishikawa, Y Nanta, A Shindo, H Tomimoto

CW-24 Does activation of brown adipose tissue participate to hypermetabolism in ALS patients?
A Hesters, D Bonnefont-Rousselot, F Salachas, L Lacomblez, M-O Habert, A Kas, G Bruneteau

CW-25 Dysphagia and dysarthria in Facial Onset Sensory Motor Neuronopathy (FOSMN): a case report
S Feroldi, F Bianchi, C Gasperoni, D Ginocchio, G Mora

CW-26 Achieving independent lives for people with ALS connected to artificial respirators through the process of accepting care from non-family members
Y Hasegawa, S Tateiwa

CW-27 The importance of mealtime assessment in ALS patients
N Pizzorni, D Ginocchio, F Bianchi, S Feroldi, C Gasperoni, M Falco, C Limonta, G Mora, A Schindler

CW-28 Successful percutaneous gastrostomy tube placement with fluoroscopy in ALS patient requiring 24/7 ventilation
C Burian, L Wolfe, J-M Li, S Ajroud-Driss

CW-29 Addressing quality of life concerns that could influence survival through the innovative gadgetry of tubes and buttons that help manage the inflow and outflow
R Onders, M Elmo, B Katirji, C Kaplan

CW-30 Supraephbic catheter in motor neuron diseases: a case series
K Patel, N Rome, S Shroff, E Simpson

CW-31 A trial of laryngeal exercises and diet among people with ALS
V Flood, S Vucic, H Bogaardt, P Menon

CW-32 Patient-reported outcomes in ALS: Evaluation of physical therapy, occupational therapy and speech-language therapy from the patient perspective
A Maier, S Spittel, A Funke, D Kettlemann, B Walter, C Munch, T Meyer

CW-33 POSTER WITHDRAWN

CW-34 How can family members keep working while providing care for ALS/MND patients?
I Ishijima, Y Kawaguchi, K Adachi, T Nakajima

CW-35 Behavioural subphenotypes in amyotrophic lateral sclerosis and their contribution to caregiver burden
T Burke, M Pinto-Grau, K Lonergan, M Heverin, M Galvin, O Hardiman, N Pender

CW-36 Changes in the event-related auditory potentials in amyotrophic lateral sclerosis patients with spinal onset
C Dolcotti, A Pelagatti, I Ghicopulos, F Sartucci, MC Carboncini, P Bongioanni

CW-37 Longitudinal changes in cognition and behaviour in ALS

CW-38 Cognitive-behavioral assessment and disclosure practices across the northeast amyotrophic lateral sclerosis (NEALS) Consortium
T Haines, A Alteiro, C Reichwein, A Morris, S Walsh, Z Simmons

CW-39 Frequency of cognitive impairment in first and second degree relatives of patients with amyotrophic lateral sclerosis
M Ryan, E Costello, E Corr, R McMahon, M Heverin, O Hardiman

CW-40 Depictions of people with ALS in Canadian newspaper coverage of assisted death
W Luth, M Moir, W Johnston, T Bubela
# Programme of events/locations

## Friday 8 December

<table>
<thead>
<tr>
<th>Time</th>
<th>Event</th>
<th>Location</th>
</tr>
</thead>
<tbody>
<tr>
<td>06.45 – 08.15</td>
<td>The ALS Expert Perspective* (Sponsored by Mitsubishi Tanabe Pharma America, Inc.)</td>
<td>Otis Lobby Level</td>
</tr>
<tr>
<td>07.00 – 18.00</td>
<td>Registration International Symposium</td>
<td>Grand Ballroom Foyer  Concourse -1</td>
</tr>
<tr>
<td>07.00 – 19.00</td>
<td>Speaker Room</td>
<td>Elm 1-2                Concourse -1</td>
</tr>
<tr>
<td>08.30 – 10.15</td>
<td>Symposium Joint Opening Session</td>
<td>Grand Ballroom         Concourse -1</td>
</tr>
<tr>
<td>11.00 – 17.40</td>
<td>Symposium Biomedical Session 2A/3A/4A</td>
<td>Marina Ballroom Lobby Level</td>
</tr>
<tr>
<td>11.00 – 17.30</td>
<td>Symposium Clinical Session 2B/3B/4B</td>
<td>Grand Ballroom A/B Concourse -1</td>
</tr>
<tr>
<td>10.30 / 15.30</td>
<td>Networking and Refreshments am/pm</td>
<td>Grand Ballroom Foyer and Galleria Concourse -1 and North Wing -1</td>
</tr>
<tr>
<td>12.30 – 14.00</td>
<td>Lunch</td>
<td>Grand Ballroom Foyer and Galleria Concourse -1 and North Wing -1</td>
</tr>
<tr>
<td>13.00 – 14.00</td>
<td>Pan-Asian Consortium for Treatment and Research in ALS (PACTALS)**</td>
<td>Otis Lobby Level</td>
</tr>
<tr>
<td>17.45 – 19.30</td>
<td>Poster Session A</td>
<td>Galleria Harbor Wing -1</td>
</tr>
<tr>
<td>17.30 – 21.00</td>
<td>Neurofilaments in ALS diagnosis and prognosis: Considerations for clinical use and use in drug development (closed meeting)</td>
<td>Otis Lobby Level</td>
</tr>
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## Saturday 9 December

<table>
<thead>
<tr>
<th>Time</th>
<th>Event</th>
<th>Location</th>
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<tbody>
<tr>
<td>07.00 – 18.00</td>
<td>Registration International Symposium</td>
<td>Grand Ballroom Foyer  Concourse -1</td>
</tr>
<tr>
<td>07.00 – 19.00</td>
<td>Speaker Room</td>
<td>Elm 1-2                Concourse -1</td>
</tr>
<tr>
<td>08.30 – 10.00</td>
<td>Symposium Biomedical Session 5A</td>
<td>Marina Ballroom         Concourse -1</td>
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<tr>
<td>08.30 – 10.00</td>
<td>Symposium Clinical Session 5B</td>
<td>Grand Ballroom A/B Concourse -1</td>
</tr>
<tr>
<td>10.30 – 17.40</td>
<td>Symposium Biomedical Session 6A/7A/8A</td>
<td>Marina Ballroom         Concourse -1</td>
</tr>
<tr>
<td>10.30 – 17.40</td>
<td>Symposium Clinical Session 6B/7B/8B</td>
<td>Grand Ballroom A/B Concourse -1</td>
</tr>
<tr>
<td>10.30 – 17.40</td>
<td>Symposium Alternative Sessions 6C/7C/8C</td>
<td>Grand Ballroom C-E Concourse -1</td>
</tr>
<tr>
<td>10.00 / 15.30</td>
<td>Networking and Refreshments am/pm</td>
<td>Grand Ballroom Foyer and Galleria Concourse -1 and North Wing -1</td>
</tr>
<tr>
<td>12.30 – 14.00</td>
<td>Lunch</td>
<td>Grand Ballroom Foyer and Galleria Concourse -1 and North Wing -1</td>
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<tr>
<td>17.45 – 19.30</td>
<td>Poster Session B</td>
<td>Galleria Harbor Wing -1</td>
</tr>
<tr>
<td>19.15 – 21.15</td>
<td>The Innovation Landscape in ALS/MND: Beyond Pharmacotherapies (Sponsored by Cytokinetics)</td>
<td>Commonwealth Ballroom Concourse -1</td>
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## Sunday 10 December

<table>
<thead>
<tr>
<th>Time</th>
<th>Event</th>
<th>Location</th>
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<tbody>
<tr>
<td>07.00 – 08.30</td>
<td>Western ALS Investigator Meeting (closed meeting)</td>
<td>Otis Lobby Level</td>
</tr>
<tr>
<td>07.00 – 14.00</td>
<td>Registration International Symposium</td>
<td>Grand Ballroom Foyer  Concourse -1</td>
</tr>
<tr>
<td>07.00 – 14.00</td>
<td>Speaker Room</td>
<td>Elm 1-2                Concourse -1</td>
</tr>
<tr>
<td>08.30 – 12.40</td>
<td>Symposium Biomedical Sessions 9A/10A</td>
<td>Marina Ballroom         Concourse -1</td>
</tr>
<tr>
<td>08.30 – 12.30</td>
<td>Symposium Clinical Sessions 9B/10B</td>
<td>Grand Ballroom A/B Concourse -1</td>
</tr>
<tr>
<td>10.00</td>
<td>Networking and Refreshments am/pm</td>
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</tr>
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<td>12.30 – 14.00</td>
<td>Lunch</td>
<td>Grand Ballroom Foyer and Galleria Concourse -1 and North Wing -1</td>
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<tr>
<td>14.00 – 15.30</td>
<td>Symposium Joint Closing Session 11</td>
<td>Grand Ballroom A/B Concourse -1</td>
</tr>
</tbody>
</table>

*This meeting is open to delegates from the US, Japan and South Korea only

**This meeting is open to delegates from the Asia-Pacific Region
Locations

Lobby Level
(Ground Floor)

Concourse
Level (-1)

North Wing,
Galleria Level (-1)
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For further information please contact
Motor Neurone Disease Association
10 - 15, Notre Dame Mews, Northampton, NN1 2BG
Tel: +44 (0) 1604 250505
Email: symposium@mndassociation.org
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